Clinical Information for Medical Necessity Review

CMS Policy Hierarchies & Application

Guideline Definitions

GENERAL	
Preface	Abdomen
Breast	Cardiac
Cardiac Implantable Devices (CID)	Chest
Head	Musculoskeletal
Neck	Oncology
Pelvis	Peripheral Nerve and Neuromuscular Disorders (PNND)
Peripheral Vascular Disease (PVD)	
Spine	
PEDIATRIC	
Pediatric Abdomen	Pediatric Cardiac
Pediatric Chest	Pediatric Head
Pediatric Musculoskeletal	Pediatric Neck
Pediatric and Special Populations Oncology	
Pediatric Pelvis	
Pediatric Peripheral Nerve and Neuromuscular Disorders (PNND)	
Pediatric Peripheral Vascular Disease (PVD)	
Pediatric and Special Populations Spine	

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Clinical Information to Establish Medical Necessity Guideline

Guideline

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Clinical Information to Establish Medical Necessity Guideline

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The clinical guidelines apply an evidence-based approach to evaluate the most appropriate medically necessary care for each individual. This evaluation requires submission of legible medical records pertinent to the test or treatment being requested by the provider.

If the medical records provided do not provide sufficiently detailed information to understand the individual's current clinical status or cannot be read, then medical necessity for the request cannot be established and cannot be approved.

Specific elements of an individual's medical records commonly required to establish medical necessity include, but are not limited to:

- Virtual or in-person clinical evaluation which includes a detailed history and physical examination since the onset or change in symptoms
- Laboratory studies
- Imaging studies
- Pathology reports
- Procedure reports
- Reports from other providers participating in treatment of the relevant condition

Clinical Information to Establish Medical Necessity Guideline

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Medicaid and Medicare Hierarchy and Application

Guideline

Medicaid and Medicare Hierarchy and Application
Medicaid
Medicare Policy
Medicare/Medicaid Dual Membership

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Medicaid and Medicare Hierarchy and Application

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Medicaid

Medicaid Hierarchy

1. Medicaid state-specific policy
2. The evidence-based guidelines or the appropriate alternative guideline utilized by a program/health plan in place of these guidelines
3. Early and Periodic Screening, Diagnostic and Treatment (EPSDT) guidelines should also be reviewed for individuals under 21 years of age as applicable for coverage determination

Application of Medicaid Policy

The state Medicaid policy will be reviewed first to determine if the information provided is instructive to the clinical case at hand. For the purpose of this policy, sufficient clinical criteria to render a medical necessity decision is defined as the presence of a state Medicaid policy that addresses the service/procedure/test/equipment and the member condition (indication) and supplies sufficient clinically relevant detail to be instructive to the case. See the numbered items below for specific guidance:

1. State Medicaid policy addresses clinical scenario: (service/procedure/test/equipment AND member condition AND Medical Necessity criteria):
   a. Medicaid policy will be utilized and cited for determining medical necessity
   b. If specific medical necessity criteria are present in the state policy, but are limited or less detailed than eviCore or alternative guideline, the state policy would still be applied and cited, as this would be considered sufficient information to render a decision
2. State policy exists for service/procedure/test/equipment AND member condition but NO medical necessity criteria exist for this indication. The state policy would still be instructive to the case clinical scenario and Medicaid policy will be utilized and cited.
3. State Medicaid policy addresses service/procedure/test/equipment, but does NOT cover member condition in question or provide specific medical necessity criteria for this indication and does not contain relevant clinical information to be instructive to the case/clinical scenario, eviCore guidelines or alternative guidelines, as appropriate, will be utilized and cited

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a. (Example: Service is requested to address a member condition and the state policy indicates that the service is indicated for this member condition without providing specific criteria regarding under which circumstances the service would be indicated for this condition. This is considered instructive, and the state policy would be applied/cited.)

4. If no clinical information is provided with a request, the above hierarchy is still applied. If there is an applicable state-specific Medicaid policy, the Medicaid policy would be utilized and cited to request clinical information/documentation. If there is no applicable state-specific Medicaid policy, eviCore guidelines or alternative guidelines, as appropriate, will be utilized and cited

Note: The scope of this policy is to outline the default order in which policy sources will be used during a delegated medical necessity determination. The Medicaid hierarchy outlined here may be superseded by Plan- and State-specific Hierarchy policies, where applicable, based on contracts with a state Medicaid entity.

Application and citation of policy

State Medicaid Policy exists	State Medicaid policy addresses service/ procedure/test/ equipment	State policy addresses member condition and/ or is instructive to the case	State Medicaid Policy provides specific medical necessity criteria relevant to service and member condition	Application and Citation of Policy
Y	Y	Y	Y	State Medicaid policy applied and cited
Y	Y	Y	N	State Medicaid policy applied and cited
Y	Y	N	N	EviCore criteria applied and cited

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State Medicaid Policy exists	State Medicaid policy addresses service/ procedure/test/ equipment	State policy addresses member condition and/ or is instructive to the case	State Medicaid Policy provides specific medical necessity criteria relevant to service and member condition	Application and Citation of Policy
Y	N	N	N	EviCore criteria applied and cited

Medicare Policy

CMS Coverage Hierarchy: Medicare Advantage Medical Policy Development and Application

Medicare Advantage medical policies identify the clinical criteria for determining when medical services are considered 'reasonable and necessary' (medically necessary). Medicare Advantage plans are required by CMS to provide the same medical benefits to Medicare Advantage members as original Medicare.

Medicare Advantage plans must comply with national coverage determinations (NCD), local coverage determinations (LCD), and general coverage and benefit conditions included in traditional Medicare laws. This includes criteria for determining whether an item or service is a benefit available under traditional Medicare. When coverage criteria are not fully established in Medicare statute, regulation, NCD, or LCD, Medicare Advantage organizations may create publicly accessible internal coverage criteria that are based on current evidence in widely used treatment guidelines or clinical literature.

If additional criteria are needed to interpret or supplement generalized provisions within an NCD, LCD or other Medicare coverage policy, or there is flexibility allowed in the NCD/LCD, or there is no applicable NCD, LCD, or LCA (used in concert with an LCD) available to determine medical necessity, then other evidence-based criteria may be applied.

Coverage criteria are not fully established when (42 CFR 422.101(6) (i) :

(i) Coverage criteria not fully established. Coverage criteria are not fully established when:

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(A) Additional, unspecified criteria are needed to interpret or supplement general provisions in order to determine medical necessity consistently. The MA organization must demonstrate that the additional criteria provide clinical benefits that are highly likely to outweigh any clinical harms, including from delayed or decreased access to items or services;

(B) NCDs or LCDs include flexibility that explicitly allows for coverage in circumstances beyond the specific indications that are listed in an NCD or LCD; or

(C) There is an absence of any applicable Medicare statutes, regulations, NCDs or LCDs setting forth coverage criteria.

The following hierarchy is used to determine Medicare Advantage Medical Policy:

1. CMS Coverage Manuals or other CMS-Based Resource: Coverage provisions in interpretive manuals are instructions that are used to further define when and under what circumstances items or services may be covered (or not covered)
2. National Coverage Determinations (NCD)
3. Local Coverage Determinations (LCD)
4. Local Coverage Articles (LCA) may be used in conjunction with LCDs, or if the LCA is stated as 'Not an LCD reference article' within the LCD , the LCA may be cited alone.
5. EviCore's evidence-based guidelines or the appropriate alternative guideline utilized by a program/health plan in place of EviCore's guidelines.

- Rationale supporting the adoption and use of internally developed coverage guidelines: When coverage criteria are not fully established (as defined in 42 CFR 422.101(6) (i)) in Medicare policy, an NCD, or an LCD, CMS allows a Medicare Advantage Organization (MAO) to create and use internally developed coverage criteria. See 'Coverage criteria are not fully established when (42 CFR 422.101(6) (i) above on page 3 for the full definition of 'not fully established'.
- NCDs with Coverage with Evidence Development (CED) contain fully established Medicare coverage criteria for purpose of 42 CFR § 422.101(b)(6). Therefore, Medicare does not permit MA organizations to apply internal coverage criteria to limit, expand, or modify the scope of the NCD with CED (42 CFR § 422.101(b)(6), November 25, 2025).
- EviCore will exhaust all traditional Medicare policies, NCDs, and LCDs prior to using internal criteria policies. When an internally created coverage policy is utilized for a specific condition where a Medicare policy, an NCD, or LCD is not fully established, the general provisions supplemented by the internal criteria will include when the specific clinical presentation of the enrollee, for the request under review is not addressed in the Medicare coverage policy; or there are no specific coverage criteria included in a Medicare policy because the policy provides broad provisions, which may be instructive, but are not detailed enough to be used to determine the medical necessity of the request. Additionally, internal coverage policy may be used when

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Medicare policy includes statements within the policy that allow for coverage beyond what is written in the Medicare policy.

- Under the guidance outlined above EviCore's evidence-based guidelines or the appropriate alternative guideline utilized by a program/health plan in place of EviCore's guidelines, EviCore may apply internally developed clinical coverage guidelines to the request under review. Each EviCore clinical coverage policy is developed following an objective, evidence-based process based on scientific evidence, generally accepted and current standards of medical practice, and authoritative clinical practice guidelines. Each coverage policy developed and used outlines clinical benefits, addresses any clinical harm, and access to services. In addition, each member's unique clinical situation is considered in conjunction with current CMS guidelines and EviCore clinical coverage policy, as applicable.

Note: Where a Medicare Administrative Contractor (MAC) has adopted the Palmetto GBA MolDX® Program's criteria for the LCDs governing molecular and genomic tests within their jurisdiction, EviCore's Laboratory Management program will follow the MolDX criteria published by the MACs for those jurisdictions.

Medicare/Medicaid Dual Membership

Hierarchy and Application for dual eligible Medicare/Medicaid

Individuals enrolled in both Medicare and Medicaid are considered to be dually eligible. For individuals with both Medicare and Medicaid, the following hierarchy should be applied.

1. CMS Coverage Manuals
2. National Coverage Determinations (NCD)
3. Local Coverage Determinations (LCD)
4. Local Coverage Articles (LCA) – when used in conjunction with an LCD
5. Evidence based clinical policies (EviCore) or the appropriate alternative guideline utilized by a program/health plan in place of EviCore's guidelines

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Guideline Definitions

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Experimental, Investigational, or Unproven

Certain studies, treatments, procedures, or devices may be considered experimental, investigational, or unproven for **any** condition, illness, disease, injury being treated if one of the following is present:

- if there is a paucity of supporting evidence;
- if the evidence has not matured to exhibit improved health parameters;
- if clinical utility has not been demonstrated in any condition; OR
- the study, treatment, procedure, or device lacks a collective opinion of support.

Supporting evidence includes standards that are based on credible scientific evidence published in peer-reviewed medical literature (such as well conducted randomized clinical trials or cohort studies with a sample size of sufficient statistical power) generally recognized by the relevant medical community. Collective opinion of support includes physician specialty society recommendations and the views of physicians practicing in relevant clinical areas when physician specialty society recommendations are not available.

Medically Necessary

Healthcare services or supplies needed to diagnose, treat, or evaluate a condition or prevent an injury, illness, condition or disease that meets accepted standards of medicine based on evidenced-based clinical standards of care based on supporting evidence and/or collective opinion of support that is:

- Clinically appropriate in terms of type, frequency, extent, site, and duration, and considered effective for the individual's illness, injury or disease;
- Clinical utility of the technology, drug, device, treatment or procedure has been demonstrated for a diagnosis, treatment, evaluation or prevention of an illness, condition or disease based on evidence-based clinical standards of care;
- Not more costly than an alternative service or sequence of services that are at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of a condition;
- Not primarily for the convenience of the individual seeking medical services, health care provider, or other physicians or healthcare providers;
- Supporting evidence and/or collective opinion of support does not demonstrate that there is an alternative that is more appropriate/effective for diagnosis, treatment, or evaluation of a condition.

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Supporting evidence includes standards that are based on credible scientific evidence published in peer-reviewed medical literature (such as well conducted randomized clinical trials or cohort studies with a sample size of sufficient statistical power) generally recognized by the relevant medical community. Collective opinion of support includes physician specialty society recommendations and the views of physicians practicing in relevant clinical areas when physician specialty society recommendations are not available. Determination of medical necessity is based on specific clinical guidelines.

Not Medically Necessary

Certain studies, treatments, procedures, or devices may be considered not medically necessary if there is supporting evidence but one of the following is present:

- Not clinically appropriate in terms of type, frequency, extent, site, and duration, and/or not considered effective for the individual's illness, injury or disease;
- Clinical utility of the technology, drug, device, treatment or procedure has not been demonstrated for a diagnosis, treatment, evaluation or prevention of the specific illness, condition or disease based on evidence-based clinical standards of care;
- More costly than an alternative service or sequence of services that are at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of a condition;
- Primarily for the convenience of the individual seeking medical services, health care provider, or other physicians or healthcare providers;
- Supporting evidence and/or collective opinion of support demonstrates that there is an alternative that is more appropriate/effective for diagnosis, treatment, or evaluation of a condition.

Supporting evidence includes standards that are based on credible scientific evidence published in peer-reviewed medical literature (such as well conducted randomized clinical trials or cohort studies with a sample size of sufficient statistical power) generally recognized by the relevant medical community. Collective opinion of support includes physician specialty society recommendations and the views of physicians practicing in relevant clinical areas when physician specialty society recommendations are not available. Determination of not medically necessary is based on specific clinical guidelines.

Special Considerations for Laboratory Testing

Laboratory-based testing is defined in terms of both the underlying technology used and, the indication for testing (i.e.: syndrome, condition, etc.). This is due to the ubiquitous use of specific technologies in laboratory medicine.

References

1. **https://www.cigna.com/health-care-providers/coverage-and-claims/policies/medical-necessity-definitions** .

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2. **Medically necessary - Glossary | HealthCare.gov**
3. **H-320.953 Definitions of "Screening" and "Medical Necessity" | AMA**
4. Per Compliance Internal policy: Clinical Certification of Services – Initial UM 0045:
   a. Experimental/Investigational: The use of a technology, drug, device, treatment, or procedure that has not been proven or recognized as having proven benefit in clinical medicine for any condition, illness, disease, or injury being treated.
   b. Medical Necessity: Refers to services or supplies for diagnosing, evaluating, treating or preventing an injury, illness, condition or disease, based on evidence-based clinical standards of care. Medically necessary services are accepted health care services and supplies provided by health care entities, appropriate to evaluation and treatment of a disease, condition, illness or injury and consistent with the applicable standard of care. Determination of medical necessity is based on specific clinical guidelines. (NCQA 2025 Standards; CMS; American College of Medical Quality)

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Guideline Development (Preface-1)

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Guideline Development (Preface-1.1)

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Guideline Development (Preface-1.1)

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- These evidence-based, proprietary clinical guidelines evaluate a range of advanced imaging and procedures, including NM, US, CT, MRI, PET, Radiation Oncology, Sleep Studies, Gastrointestinal Endoscopy, as well as Cardiac, musculoskeletal and Spine interventions.
- EviCore reserves the right to change and update the guidelines. The guidelines undergo a formal review annually. These clinical guidelines are based on current evidence supported by major national and international association and society guidelines and criteria, peer-reviewed literature, major treatises as well as, input from health plans, and practicing academic and community-based physicians.
- These guidelines are not intended to supersede or replace sound medical judgment, but instead, should facilitate the identification of the most appropriate imaging or other designated procedure given the individual's clinical condition. These guidelines are written to cover medical conditions as experienced by the majority of individuals. However, these guidelines may not be applicable in certain clinical circumstances, and physician judgment can override the guidelines.
- These guidelines provide evidence-based, clinical benefits with a focus on health care quality and patient safety.
- Clinical decisions, including treatment decisions, are the responsibility of the individual and his/her provider. Clinicians are expected to use independent medical judgment, which takes into account the clinical circumstances to determine individual management decisions.

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Benefits, Coverage Policies, and Eligibility Issues (Preface-2)

Guideline

Benefits, Coverage Policies, and Eligibility Issues (Preface-2.1)
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Benefits, Coverage Policies, and Eligibility Issues (Preface-2.1)

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Medicare Coverage Policies

- See EviCore's **Medicaid and Medicare Hierarchy and Application**.

Investigational and Experimental Studies

- See EviCore's **Guideline Definitions.**

Clinical and Research Trials

- Similar to investigational and experimental studies, clinical trial imaging requests are reviewed to determine whether they meet Health Plan coverage and these evidence-based clinical guidelines.
- Imaging studies which are inconsistent with established clinical standards, or are requested for data collection and not used in direct clinical management are not supported.

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References (Preface-2)

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1. Prospective Payment Systems - General Information. Centers for Medicare and Medicaid Services. https://www.cms.gov/medicare/payment/prospective-payment-systems#:~:text=A%20Prospective%20Payment%20System%20(PPS,on%20a%20predetermined%2C%20fixed%20amount
2. Medicares Coverage With Evidence Development: A Policy-Making Tool in Evolution. *J Oncol Pract*. 2007;3(6):296-301. doi:10.1200/jop.0763501
3. Coverage of Clinical Trials under the Patient Protection and Affordable Care Act; 42 U.S.C.A. § 300gg-8

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Clinical Information (Preface-3)

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Clinical Information (Preface-3.1)
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Clinical Information (Preface-3.1)

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Clinical Documentation and Age Considerations

- These clinical guidelines use an evidence-based approach to determine the most appropriate procedure for each individual, at the most appropriate time in the diagnostic and treatment cycle. These clinical guidelines are framed by:
  - clinical presentation of the individual, rather than the studies requested
  - adequate clinical information that must be submitted to EviCore in order to establish medical necessity for advanced imaging or other designated procedures includes, but is not limited to the following:
    - Pertinent clinical evaluation since the onset or change in symptoms including a detailed history, physical examination, appropriate laboratory studies, and appropriate prior imaging studies.
      - Condition-specific guideline sections may describe additional clinical information which is required for a pertinent clinical evaluation.
      - The Spine and Musculoskeletal guidelines require x-ray studies from when the current episode of symptoms has started or changed.
      - Advanced imaging or other designated procedures should not be ordered prior to clinical evaluation of an individual by the physician treating the individual. This may include referral to a consultant specialist who will make further treatment decisions.
      - Other meaningful technological contact (telehealth visit, telephone or video call, electronic mail or messaging) since the onset or change in symptoms by an established individual can serve as a pertinent clinical evaluation.
        - Some conditions may require a face-to-face evaluation as discussed in the applicable condition-specific guideline sections.
    - A recent clinical evaluation may be unnecessary if the individual is undergoing a guideline-supported, scheduled follow-up imaging or other designated procedural evaluation. Exceptions due to routine surveillance indications are addressed in the applicable condition-specific guideline sections.
  - the evidence-based approach to determine the most appropriate procedure for each individual requires submission of medical records pertinent to the requested imaging or other designated procedures.
- Many conditions affecting the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.

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- Individuals who are 18 years old or younger should be imaged according to the Pediatric Imaging Guidelines if discussed in the condition-specific guideline sections. Any conditions not specifically discussed in the Pediatric Imaging Guidelines should be imaged according to the General Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Imaging Guidelines, except where directed otherwise by a specific guideline section.

General Imaging Information

- "Standard" or "conventional" imaging is most often performed in the initial and subsequent evaluations of malignancy. Standard or conventional imaging includes plain film, CT, MRI, or US.
  - Often, further advanced imaging is needed when initial imaging, such as ultrasound, CT, or MRI does not answer the clinical question. Uncertain, indeterminate, inconclusive, or equivocal may describe these situations.
- Appropriate use of contrast is a very important component of evidence-based advanced imaging use.
  - The appropriate levels of contrast for an examination (i.e., without contrast, with contrast, without and with contrast) is determined by the evidence-based guidance reflected in the condition-specific guideline sections.
  - If, during the performance of a non-contrast imaging study, there is the unexpected need to use contrast in order to evaluate a possible abnormality, then that is appropriate.

Ultrasound

- Diagnostic ultrasound uses high-frequency sound waves to evaluate soft tissue structures and vascular structures utilizing grey scale and Doppler techniques.
- Ultrasound allows for dynamic real-time imaging at the bedside.
  - Ultrasound is limited in areas where there is dense bone or other calcification.
  - Ultrasound also has a relatively limited imaging window so may be of limited value in evaluating very large abnormalities.
  - In general, ultrasound is highly operator-dependent, and proper training and experience are required to perform consistent, high-quality evaluations.
- Indications for ultrasound may include, but are not limited to, the following:
  - Obstetric and gynecologic imaging
  - Soft tissue and visceral imaging of the chest, abdomen, pelvis, and extremities
  - Brain and spine imaging when not obscured by dense bony structures
  - Vascular imaging when not obscured by dense bony structures
  - Procedural guidance when not obscured by dense bony structures
  - Initial evaluation of ill-defined soft tissue masses or fullness and differentiating adenopathy from mass or cyst. Prior to advanced imaging, ultrasound can be

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very beneficial in selecting the proper modality, body area, image sequences, and contrast level that will provide the most definitive information for the individual.

- More specific guidance for ultrasound usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

Computed Tomography (CT)

- The AMA CPT® manual does not describe nor assign any minimum or maximum number of sequences for any CT study. CT imaging protocols are often influenced by the individual's clinical situation and additional sequences are not uncommon. There are numerous CT protocols that may be performed to evaluate specific clinical questions, and this technology is constantly undergoing development.
- CT utilizes ionizing radiation to create cross-sectional and volumetric images of the body.
  - Advantages over ultrasound include a much larger field of view and faster completion time in general. Disadvantages compared to ultrasound include lack of portability and exposure to ionizing radiation.
  - Advantages over MRI include faster imaging and a more spacious scanner area limiting claustrophobia. Disadvantages compared to MRI include decreased soft tissue definition, especially with non-contrast imaging, and exposure to ionizing radiation.
- CT can be performed without, with, or without and with intravenous (IV) contrast depending on the clinical indication and body area.
  - In general, non-contrast imaging is appropriate for evaluating structures with significant tissue density differences such as lung parenchyma and bony structures, or when there is a contraindication to contrast.
  - In general, CT with contrast is the most common level of contrast and can be used when there is need for improved vascular or soft tissue resolution, including better characterization of known or suspected malignancy, as well as infectious and inflammatory conditions.
  - CT without and with contrast has a limited role as the risks of doubling the ionizing radiation exposure rarely outweigh the benefits of multiphasic imaging, though there are some exceptions which include, but are not limited to, the following:
    - Characterization of a mass
    - Characterization of arterial and venous anatomy
    - CT with contrast may be used to better characterize findings on a very recent (within two weeks) inconclusive non-contrast CT where the guidelines would support CT without and with contrast.
  - More specific guidance for CT contrast usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.
- Shellfish allergy:

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  - It is commonly assumed that an allergy to shellfish indicates iodine allergy, and that this implies an allergy to iodinated contrast media used with CT. However, this is NOT true. Shellfish allergy is due to tropomyosins. Iodine plays no role in these allergic reactions. Allergies to shellfish do not increase the risk of reaction to iodinated contrast media any more than that of other allergens.
- Enteric contrast (oral or rectal) is sometimes used in abdominal imaging. There is no specific CPT® code which refers to enteric contrast.
- The appropriate contrast level and anatomic region in CT imaging is specific to the clinical indication, as listed in the condition-specific guideline sections.
- CT should not be used to replace MRI in an attempt to avoid sedation unless it is listed as a recommended study in the appropriate condition-specific guideline.
- There are significant potential adverse effects associated with the use of iodinated contrast media. These include hypersensitivity reactions, thyroid dysfunction, and contrast-induced nephropathy (CIN). Individuals with impaired renal function are at increased risk for CIN.
- Both contrast CT and MRI are considered to have the same risk profile with renal failure (GFR <30 mL/min).
- The use of CT contrast should proceed with caution in pregnant and breastfeeding individuals. There is a theoretical risk of contrast toxicity to the fetal and infant thyroid. The procedure can be performed if the specific need for that contrast-enhanced procedure outweighs risk to the fetus. Breastfeeding individuals may reduce this risk by choosing to pump and discard breast milk for 12-24 hours after the contrast injection.
- CT without contrast is medically necessary if clinical criteria for CT with contrast are met AND the individual has/is:
  - elevated blood urea nitrogen (BUN) and/or creatinine
  - renal insufficiency
  - allergies to iodinated contrast
  - thyroid disease which could be treated with I-131
  - diabetes
  - very elderly
  - urgent or emergent settings due to availability
  - trauma
- CT is superior to other imaging modalities in certain conditions including, but not limited to, the following:
  - Screening following trauma
  - Imaging pulmonary disease
  - Imaging abdominal and pelvic viscera
  - Imaging of complex fractures

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  - Evaluation of inconclusive findings on Ultrasound or MRI, or if there is a contraindication to MRI
- More specific guidance for CT usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

Magnetic Resonance Imaging (MRI)

- The AMA CPT® manual does not describe nor assign any minimum or maximum number of sequences for any MRI study. MRI protocols are often influenced by the individual's clinical situation and additional sequences are not uncommon. There are numerous MRI sequences that may be performed to evaluate specific clinical questions, and this technology is constantly undergoing development.
- Magnetic Resonance Imaging (MRI) utilizes the interaction between the intrinsic radiofrequency of certain Molecules in the body (hydrogen in most cases) and a strong external magnetic field.
  - MRI is often superior for advanced imaging of soft tissues and can also define physiological processes in some instances (e.g., edema, loss of circulation [AVN], and increased vascularity [tumors]).
  - MRI does not use ionizing radiation, and even non-contrast images have much higher soft tissue definition than CT or Ultrasound.
  - MRI typically takes much longer than either CT or Ultrasound, and for some individuals may require sedation. It is also much more sensitive to individual motion that can degrade image quality than either CT or Ultrasound.
- MRI Breast and MRI Chest are not interchangeable, as they focus detailed sequences on different adjacent body parts.
- MRI may be utilized either as the primary advanced imaging modality, or when further definition is needed based on CT or ultrasound imaging.
- Most orthopedic and dental implants are not magnetic. These include hip and knee replacements; plates, screws, and rods used to treat fractures; and cavity fillings. Yet, all of these metal implants can distort the MRI image if near the part of the body being scanned.
  - Other implants, however, may have contraindications to MRI. These include the following:
    - Pacemakers
    - ICD or heart valves
    - Metal implants in the brain
    - Metal implants in the eyes or ears
    - Infusion catheters and bullets or shrapnel
  - CT can therefore be an alternative study to MRI in these scenarios.
- The contrast level and anatomic region in MRI imaging is specific to the clinical indication, as listed in the specific guideline sections.

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- MRI utilizing Xenon Xe 129 (CPT® C9791) for contrast is considered investigational and experimental at this time. MRI with or with and without contrast in these guidelines refers to MRI utilizing gadolinium for contrast.
- MRI is commonly performed without, without and with contrast.
  - Non-contrast imaging offers excellent tissue definition.
  - Imaging without and with contrast is commonly used when needed to better characterize tissue perfusion and vascularization.
    - Most contrast is gadolinium based and causes T2 brightening of the vascular and extracellular spaces.
    - Some specialized gadolinium and non-gadolinium contrast agents are available, and most commonly used for characterizing liver lesions.
  - MRI with contrast only is rarely appropriate and is usually used to better characterize findings on a recent inconclusive non-contrast MRI, commonly called a completion study.
  - MRI contrast is relatively contraindicated in pregnant individuals.
  - More specific guidance for MRI contrast usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.
- MRI may be preferred in individuals with renal failure and in individuals allergic to intravenous CT contrast.
  - Both contrast CT and MRI are considered to have the same risk profile with renal failure (GFR <30 mL/min).
  - Gadolinium can cause Nephrogenic Systemic Fibrosis (NSF). The greater the exposure to gadolinium in individuals with a low GFR (especially if on dialysis), the greater the chance of individuals developing NSF.
  - Multiple studies have demonstrated potential for gadolinium deposition following the use of gadolinium-based contrast agents (GBCAs) for MRI studies. The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
- A CT is medically necessary in place of an MRI when clinical criteria are met for MRI AND there is a contraindication to having an MRI (pacemaker, ICD, insulin pump, neurostimulator, etc.).
  - When replacing MRI with CT, contrast level matching should occur as follows:
    - MRI without contrast → CT without contrast
    - MRI without and with contrast → CT with contrast or CT without and with contrast
- The following situations may impact the appropriateness for MRI and/or MR contrast:

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  - Caution should be taken in the use of gadolinium in individuals with renal failure.
  - The use of gadolinium contrast agents is relatively contraindicated during pregnancy unless the specific need for that procedure outweighs risk to the fetus.
  - MRI can be performed for non-ferromagnetic body metals (i.e., titanium), although some imaging facilities will consider it contraindicated if recent surgery, regardless of the metal type.
- MRI should not be used as a replacement for CT for the sole reason of avoidance of ionizing radiation when MRI is not supported in the condition-based guidelines, since it does not solve the problem of overutilization.
- MRI is superior to other imaging modalities in certain conditions including, but not limited to, the following:
  - Imaging the brain and spinal cord
  - Characterizing visceral and musculoskeletal soft tissue masses
  - Evaluating musculoskeletal soft tissues including ligaments and tendons
  - Evaluating inconclusive findings on ultrasound or CT
  - Individuals who are pregnant or have high radiation sensitivity
  - Suspicion, diagnosis, or surveillance of infections
- More specific guidance for MRI usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

Positron Emission Tomography (PET)

- PET is a nuclear medicine study that uses a positron emitting radiotracer to create cross-sectional and volumetric images based on tissue metabolism.
- Conventional imaging (frequently CT, sometimes MRI or bone scan) of the affected area(s) drives much of initial and restaging and surveillance imaging for malignancy and other chronic conditions. PET is not medically necessary for surveillance imaging unless specifically stated in the condition-specific guideline sections.
- PET/MRI is generally not supported, see **PET-MRI (Preface-5.3)**.
- PET is rarely performed as a single modality, but is typically performed as a combined PET/CT.
  - The unbundling of PET/CT into separate PET and diagnostic CT CPT® codes is not supported, because PET/CT is done as a single study.
- PET/CT lacks the tissue definition of CT or MRI, but is fairly specific for metabolic activity based on the radiotracer used.
- Indications for PET/CT may include the following:
  - Oncologic Imaging for evaluation of tumor metabolic activity
  - Cardiac Imaging for evaluation of myocardial metabolic activity
  - Brain Imaging for evaluation of metabolic activity for procedural planning
- More specific guidance for PET usage, including exceptions to this general guidance, can be found throughout the condition-specific guidelines.

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Overutilization of Advanced Imaging

- A number of reports describe overutilization in many areas of advanced imaging and other procedures, which may include the following:
  - High-level testing without consideration of less invasive, lower cost options which may adequately address the clinical question at hand
  - Excessive radiation and costs with unnecessary testing
  - Defensive medical practice
  - CT without and with contrast (so called “double contrast studies") requests, which have few current indications
  - MRI requested in place of CT to avoid radiation without considering the primary indication for imaging
  - Adult CT settings and protocols used for smaller people and children
  - Unnecessary imaging procedures when the same or similar studies have already been conducted
- A review of the imaging or other relevant procedural histories of all individuals presenting for studies has been recognized as one of the more important processes that can be significantly improved. By recognizing that a duplicate or questionably medically necessary imaging study has been ordered for individuals, it may be possible to avoid exposing them to unnecessary risks. To avoid these unnecessary risks, the precautions below should be considered:
  - The results of initial diagnostic tests or radiologic studies to narrow the differential diagnosis should be obtained prior to performing further tests or radiologic studies.
  - The clinical history should include a potential indication such as a known or suspected abnormality involving the body part for which the imaging study is being requested. These potential indications are addressed in greater detail within the applicable guidelines.
  - The results of the requested imaging procedures should be expected to have an impact on individual management or treatment decisions.
  - Repeat imaging studies are not generally necessary unless there is evidence of disease progression, recurrence of disease, and/or the repeat imaging will affect an individual’s clinical management.
- Pre-operative imaging/pre-surgical planning imaging/pre-procedure imaging is not medically necessary if the surgery/procedure is not medically necessary. Once the procedure has been approved or if the procedure does not require prior authorization, the appropriate pre-procedural imaging may be approved.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants

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of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Coding Issues (Preface-4)

Guideline

3D Rendering (Preface-4.1)
CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2)
Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)
Unilateral Versus Bilateral Breast MRI (Preface-4.4)
CPT® 76380 Limited or Follow-up CT (Preface-4.5)
SPECT/CT Imaging (Preface-4.6)
CPT® 76140 Interpretation of an Outside Study (Preface-4.7)
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HCPCS Codes (Preface-4.9)
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3D Rendering (Preface-4.1)

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CPT® 76376 and CPT® 76377:

- Both codes require concurrent supervision of the image post-processing 3D manipulation of the volumetric data set and image rendering.
  - Concurrent supervision is defined as active physician participation in and monitoring of the reconstruction process including design of the anatomic region that is to be reconstructed; determination of the tissue types and actual structures to be displayed (e.g., bone, organs, and vessels); determination of the images or cine loops that are to be archived; and monitoring and adjustment of the 3D work product. The American College of Radiology (ACR) recommends that it is best to document the physician's supervision or participation in the 3D reconstruction of images.
- These two codes differ in the need for and use of an independent workstation for post-processing.
  - CPT® 76376 reports procedures not requiring image post-processing on an independent workstation.
  - CPT® 76377 reports procedures that require image post-processing on an independent workstation.
- These 3D rendering codes should not be used for 2D reformatting.
- Two-dimensional reconstruction (e.g., reformatting an axial scan into the coronal plane) is now included in all cross-sectional imaging base codes and is not separately reimbursable.
- The codes used to report 3D rendering for ultrasound and echocardiography are also used to report the 3D post processing work on CT, MRI, and other tomographic modalities.
- Providers may be required to obtain prior authorization on these 3D codes even if prior authorization is not required for the echocardiography and/or ultrasound procedure codes. It may appear that EviCore pre-authorizes echocardiography and/or ultrasound when, in fact, it may only be the 3D code that needs the prior authorization.
- CPT® codes for 3D rendering should not be billed in conjunction with computer-aided detection (CAD), MRA, CTA, nuclear medicine SPECT studies, PET, PET/CT, stereotactic localization (CPT® 77011 or CPT® 70486 if used), Mammogram, MRI Breast, US Breast, CT Colonography (virtual colonoscopy), Cardiac MRI, Cardiac CT, or Coronary CTA studies.

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- CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT® 76376 (3D rendering not requiring image post-processing on an independent workstation) can be considered in the following clinical scenarios:
  - Bony conditions:
    - Evaluation of congenital skull abnormalities in newborns, infants, and toddlers (usually for pre-operative planning)
    - Complex fractures (comminuted or displaced)/dislocations of any joint (for pre-operative planning when conventional imaging is insufficient)
    - Spine fractures, pelvic/acetabulum fractures, intra-articular fractures (for pre-operative planning when conventional imaging is insufficient)
    - Pre-operative planning for other complex surgical cases
    - Complex facial fractures
  - Pre-operative planning for other complex surgical cases
  - Cerebral angiography
  - Pelvis conditions:
    - Uterine intra-cavitary lesion when initial US is equivocal: See **Abnormal Uterine Bleeding (AUB) (PV-2.1)** and **Leiomyoma/Uterine Fibroids (PV-12.1)** in the Pelvis Imaging Guidelines.
    - Hydrosalpinxes or peritoneal cysts when initial US is indeterminate: See **Complex Adnexal Masses (PV-5.3)** in the Pelvis Imaging Guidelines.
    - Lost IUD (inability to feel or see IUD string) with initial US: See **Intrauterine Device (PV-10.1)** in the Pelvis Imaging Guidelines.
    - Uterine anomalies with initial US: See **Uterine Anomalies (PV-14.1)** in the Pelvis Imaging Guidelines.
    - Infertility: See **Initial Infertility Evaluation, Female (PV-9.1)** in the Pelvis Imaging Guidelines.
  - Abdomen conditions:
    - CT Urogram: See **Hematuria and Hydronephrosis (AB-39)** in the Abdomen Imaging Guidelines.
    - MRCP: See **MR Cholangiopancreatography (MRCP) (AB-27)** in the Abdomen Imaging Guidelines.

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CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2)

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- CT-, MR-, and Ultrasound-guidance procedure codes contain all of the imaging necessary to guide a needle or catheter. It is inappropriate to routinely bill a diagnostic procedure code in conjunction with a guidance procedure code.
- Imaging studies performed as part of a CT-, MR-, or Ultrasound-guided procedure should be reported using the CPT® codes in the following table:

TABLE: Imaging Guidance Procedure Codes

CPT®	Description
19085	Biopsy, breast, with placement of breast localization device(s), when performed, and imaging of the biopsy specimen, when performed, percutaneous; first lesion, including MR guidance
19086	Biopsy, breast, with placement of breast localization device(s), when performed, and imaging of the biopsy specimen, when performed, percutaneous; each additional lesion, including MR guidance
75989	Imaging guidance for percutaneous drainage with placement of catheter (all modalities)
76942	Ultrasonic guidance for needle placement
77011	CT guidance for stereotactic localization
77012	CT guidance for needle placement
77013	CT guidance for, and monitoring of parenchymal tissue ablation
77021	MR guidance for needle placement
77022	MR guidance for, and monitoring of parenchymal tissue ablation

CPT® 19085 and CPT® 19086

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- The proper way to bill an MRI-guided breast biopsy is CPT® 19085 (Biopsy, breast, with placement of breast localization device(s), when performed, and imaging of the biopsy specimen, when performed, percutaneous; first lesion, including MR guidance). Additional lesions should be billed using CPT® 19086.
  - **CPT® 77021** (MR guidance for needle placement) is not an appropriate code for a breast biopsy.

CPT® 75989

- This code is used to report imaging guidance for a percutaneous drainage procedure in which a catheter is left in place.
- This code can be used to report whether the drainage catheter is placed under fluoroscopy, Ultrasound-, CT-, or MR-guidance modality.

CPT® 77011

- A stereotactic CT localization scan is frequently obtained prior to sinus surgery. The dataset is then loaded into the navigational workstation in the operating room for use during the surgical procedure. The information provides exact positioning of surgical instruments with regard to the individual's 3D CT images.
- In most cases, the pre-operative CT is a technical-only service that does not require interpretation by a radiologist.
  - The imaging facility should report CPT® 77011 when performing a scan not requiring interpretation by a radiologist.
  - If a diagnostic scan is performed and interpreted by a radiologist, the appropriate diagnostic CT code (e.g., CPT® 70486) should be used.
  - It is not appropriate to report both CPT® 70486 and CPT® 77011 for the same CT stereotactic localization imaging session.
  - 3D Rendering (CPT® 76376 or CPT® 76377) should not be reported in conjunction with CPT® 77011 (or CPT® 70486 if used). The procedure inherently generates a 3D dataset.

CPT® 77012 (CT) and CPT® 77021 (MR)

- These codes are used to report imaging guidance for needle placement during biopsy, aspiration, and other percutaneous procedures.
- They represent the radiological supervision and interpretation of the procedure and are often billed in conjunction with surgical procedure codes.
  - For example, CPT® 77012 is reported when CT guidance is used to place the needle for a conventional arthrogram.
  - Only codes representing percutaneous surgical procedures should be billed with CPT® 77012 and CPT® 77021. It is inappropriate to use with surgical codes for open, excisional, or incisional procedures.

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- **CPT® 77021** (MR guidance for needle placement) is not an appropriate code for breast biopsy.
  - CPT® 19085 would be appropriate for the first breast biopsy site and CPT® 19086 would be appropriate for additional concurrent biopsies.

CPT® 77013 (CT) and CPT® 77022 (MR)

- These codes include the initial guidance to direct a needle electrode to the tumor(s), monitoring for needle electrode repositioning within the lesion, and as necessary for multiple ablations to coagulate the lesion and confirmation of satisfactory coagulative necrosis of the lesion(s) and comparison to pre-ablation images.
  - **NOTE:** CPT® 77013 should only be used for non-bone ablation procedures.
    - CPT® 20982 includes CT guidance for bone tumor ablations.
  - Only codes representing percutaneous surgical procedures should be billed with CPT® 77013 and CPT® 77022. It is inappropriate to use with surgical codes for open, excisional, or incisional procedures.
- CPT® 77012 and CPT® 77021 (as well as guidance codes CPT® 76942 [US], and CPT® 77002 - CPT® 77003 [fluoroscopy]) describe radiologic guidance by different modalities.
  - Only one unit of any of these codes should be reported per individual encounter (date of service). The unit of service is considered to be the individual encounter, not the number of lesions, aspirations, biopsies, injections, or localizations.

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Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)

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Unlisted Procedures

CPT®	Description
78999	Unlisted procedure, diagnostic nuclear medicine
76498	Unlisted MR procedure (e.g., diagnostic or interventional)
76497	Unlisted CT procedure (e.g., diagnostic or interventional)

- For general information related to unlisted procedures, please refer to **Management of Unlisted Codes**.
- These unlisted codes should be reported whenever a diagnostic or interventional CT or MR study is performed in which an appropriate anatomic site-specific code is not available.
  - A Category III code that describes the procedure performed must be reported rather than an unlisted code if one is available.
- CPT® 76497 or CPT® 76498 (Unlisted CT or MRI procedure) is medically necessary in the following clinical scenarios:
  - Studies done for navigation and planning for neurosurgical procedures (i.e. Stealth or Brain Lab Imaging)
  - Custom joint arthroplasty planning (not as an alternative recommendation): See **Osteoarthritis (MS-12.1)** in the Musculoskeletal Imaging Guidelines.
  - Any procedure/surgical planning if thinner cuts or different positional acquisition (than those on the completed diagnostic study) are needed. These could include navigational bronchoscopy. See **Navigational Bronchoscopy (CH-1.7)** in the Chest Imaging Guidelines.

Therapy Treatment Planning

- Radiation Therapy Treatment Planning: See **Unlisted Procedure Codes in Oncology (ONC-1.5)** in the Oncology Imaging Guidelines.

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Unilateral Versus Bilateral Breast MRI (Preface-4.4)

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- Diagnostic MRI of both breasts should be coded as CPT® 77049 regardless of whether both breasts are imaged simultaneously or whether unilateral breast MRI is performed in two separate imaging sessions.

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CPT® 76380 Limited or Follow-up CT (Preface-4.5)

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- CPT® 76380 describes a limited or follow-up CT scan. The code is used to report any CT scan, for any given area of the body, in which the work of a full diagnostic code is not performed.
- Common examples include, but are not limited to, the following:
  - Limited sinus CT imaging protocol
  - Limited or follow-up slices through a known pulmonary nodule
  - Limited slices to assess a non-healing fracture (such as the clavicle)
- Limited CT (CPT® 76380) is not medically necessary for treatment planning purposes. See **Unlisted Procedure Codes in Oncology (ONC-1.5)** in the Oncology Imaging Guidelines.
- It is inappropriate to report CPT® 76380, in conjunction with other diagnostic CT codes, to cover 'extra slices' in certain imaging protocols.
  - There is no specific number of sequences or slices defined in any CT CPT® code definition.
  - The AMA, in *CPT® 2019*, does not describe nor assign any minimum or maximum number of sequences or slices for any CT study.
    - A few additional slices or sequences are not uncommon.
    - CT imaging protocols are often influenced by the individual's clinical situation. Sometimes the protocols require more time and sometimes less.

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SPECT/CT Imaging (Preface-4.6)

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- SPECT/CT involves SPECT (Single Photon Emission Computed Tomography) nuclear medicine imaging and CT for optimizing location, accuracy, and attenuation correction and combines functional and anatomic information.
  - Common studies using this modality include ^{123}I- or ^{131}I-Metaiodobenzylguanidine (MIBG) and octreotide scintigraphy for neuroendocrine tumors.
- Hybrid Nuclear/CT scan can be reported as CPT® 78830 (single area and single day), CPT® 78831 (2 or more days), or CPT® 78832 (2 areas with one day and 2-day study).
- CPT® 78072 became effective January 1, 2013 for SPECT/CT parathyroid nuclear imaging.

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CPT® 76140 Interpretation of an Outside Study (Preface-4.7)

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- It is inappropriate to use diagnostic imaging codes for interpretation of a previously performed exam that was completed at another facility.
  - If the outside exam is being used for comparison with a current exam, the diagnostic code for the current examination includes comparison to the prior study.
  - CPT® 76140 is the appropriate code to use for an exam which was completed elsewhere and a secondary interpretation of the images is requested.

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Quantitative MR Analysis (Preface-4.8)

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- Category III CPT® codes for quantitative analysis of multiparametric-MR (mp-MRI) data with and without an associated diagnostic MRI have been established. Quantitative mp-MRI uses software to analyze tissue physiology of visceral organs and other anatomic structures non-invasively.
- For criteria associated with these types of studies, please see the condition-specific guidelines.

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HCPCS Codes (Preface-4.9)

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- Healthcare Common Procedure Coding System (HCPCS) codes are utilized by some hospitals in favor of the typical Level-III CPT® codes. These codes are typically 4 digits preceded by a C or S.
  - Many of these codes have similar code descriptions to Level-III CPT® codes (i.e., C8931 – MRA with dye, Spinal Canal; and, CPT® 72159 – MRA Spinal Canal).
  - If cases are submitted with HCPCS codes with similar code descriptions to the typical Level-III CPT® codes, those procedures should be managed in the same manner as the typical CPT® codes.
  - HCPCS code management is discussed further in the applicable guideline sections.
- Requests for many Healthcare Common Procedure Coding System (HCPCS) codes, including non-specific codes such as S8042 (Magnetic resonance imaging [MRI], low-field), should be redirected to a more appropriate and specific CPT® code. Exceptions are noted in the applicable guideline sections.

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Whole-Body Imaging (Preface-5)

Guideline

Whole Body CT Imaging (Preface-5.1)
Whole-Body MR Imaging (Preface-5.2)
PET/MRI (Preface-5.3)
References (Preface-5)

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Whole Body CT Imaging (Preface-5.1)

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- Whole-body CT or LifeScan (CT Brain, Chest, Abdomen, and Pelvis) for screening of asymptomatic individuals is not a covered benefit. The performance of whole-body screening CT examinations in healthy individuals does not meet any of the current validity criteria for screening studies and there is no clear documentation of benefit versus radiation risk.
- Whole-body low-dose skeletal CT is supported for oncologic staging in Multiple Myeloma. See **Multiple Myeloma and Plasmacytomas (ONC-25)** in the Oncology Imaging Guidelines.

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Whole-Body MR Imaging (Preface-5.2)

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- Whole-body MRI (WBMRI) is, with the exception of select cancer predisposition syndromes and autoimmune conditions discussed below, generally not supported at this time due to lack of standardization in imaging technique and lack of evidence that WBMRI improves outcome for any individual disease state.
  - While WBMRI has the benefit of whole-body imaging and lack of radiation exposure, substantial variation still exists in the number of images, type of sequences (STIR vs. diffusion weighting, for example), and contrast agent(s) used.
- Coding considerations:
  - There are no established CPT® or HCPCS codes for reporting WBMRI.
  - WBMRI is at present only reportable using CPT® 76498. All other methods of reporting whole-body MRI are inappropriate including the following:
    - Separate diagnostic MRI codes for multiple individual body parts
    - MRI Bone Marrow Supply (CPT® 77084)
- Disease-specific considerations:
  - Cancer screening:
    - Interval WBMRI is recommended for cancer screening in individuals with select cancer predisposition syndromes. Otherwise, WBMRI has not been shown to improve outcomes for cancer screening.
      - For additional information, see **Li-Fraumeni Syndrome (LFS) (PEDONC-2.2)**, **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)**, **Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)**, **Hereditary Paraganglioma-Pheochromocytoma (HPP) Syndromes (PEDONC-2.13)**, **Constitutional Mismatch Repair Deficiency (CMMRD or Turcot Syndrome) (PEDONC-2.15)**, **Infantile Myofibromatosis (PEDONC-2.18)**, or **Bloom Syndrome (PEDONC-2.19)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
  - Cancer staging and restaging:
    - Whole-body MRI has limited indications in staging and restaging of multiple myeloma. See **Multiple Myeloma and Plasmacytomas (ONC-25)** in the Oncology Imaging Guidelines for additional details.
    - Evidence has not been published establishing WBMRI as a standard evaluation for any other type of cancer.
  - Autoimmune disease:
    - WBMRI can be approved in some situations for individuals with chronic recurrent multifocal osteomyelitis.

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- For additional information, see **Chronic Recurrent Multifocal Osteomyelitis (PEDMS-10.2)** in the Pediatric Musculoskeletal Imaging Guidelines.

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PET/MRI (Preface-5.3)

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- PET/MRI is generally not supported for a vast majority of oncologic and neurologic conditions due to lack of standardization in imaging technique and interpretation. However, it is medically necessary in select circumstances when the following criteria are met:
  - The individual meets condition-specific guidelines for PET/MRI OR
  - The individual meets ALL of the following:
    - The individual meets guideline criteria for PET/CT, **AND**
    - PET/CT is not available at the treating institution, **AND**
    - The provider requests PET/MRI in lieu of PET/CT
- When the above criteria are met, PET/MRI is reported using the code combination of PET Whole-Body (CPT® 78813) and MRI Unlisted (CPT® 76498). All other methods of reporting PET/MRI are inappropriate.
  - When clinically appropriate, diagnostic MRI codes can be medically necessary at the same time as the PET/MRI code combination.
- For more information, please see the appropriate condition-based guideline.

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- Complete reference citations for the journal articles are embedded within the body of the guidelines and/or may be found on the Reference pages at the end of some guideline sections.

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Abbreviations for Abdomen Imaging Guidelines

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Abbreviations for Abdomen Imaging Guidelines	
AAA	abdominal aortic aneurysm
AASLD	American Association for the Study of Liver Diseases
ACE	angiotensin-converting enzyme
ACG	American College of Gastroenterology
ACR	American College of Radiology
ACTH	adrenocorticotropic hormone
AFP	alpha-fetoprotein
AGA	American Gastroenterological Association
ALT	alanine aminotransferase
ASGE	American Society for Gastrointestinal Endoscopy
AST	aspartate aminotransferase
AUA	American Urological Association
BEIR	Biological Effects of Ionizing Radiation
BUN	blood urea nitrogen
CAG	Canadian Association of Gastroenterology
CNS	central nervous system

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Abbreviations for Abdomen Imaging Guidelines	
CT	computed tomography
CTA	computed tomography angiography
CTC	computed tomography colonography (aka: virtual colonoscopy)
DVT	deep vein thrombosis
ERCP	endoscopic retrograde cholangiopancreatography
EUS	endoscopic ultrasound
FNH	focal nodular hyperplasia
GFR	glomerular filtration rate
GGT	gamma glutamyl transferase
GI	gastrointestinal
HCC	hepatocellular carcinoma
HCPCS	Healthcare Common Procedural Coding System (commonly pronounced: “hix pix”)
HU	Hounsfield units
IAA	iliac artery aneurysm
IV	intravenous
KUB	kidneys, ureters, bladder (plain frontal supine abdominal radiograph)
LFT	liver function tests
MASLD	metabolic dysfunction associated steatotic liver disease (formerly known as NAFLD)

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Abbreviations for Abdomen Imaging Guidelines	
MRCP	magnetic resonance cholangiopancreatography
MRA	magnetic resonance angiography
MRE	magnetic resonance elastography
MRI	magnetic resonance imaging
mSv	millisievert
NAFLD	nonalcoholic fatty liver disease (now known as MASLD)
PA	posteroanterior projection
PET	positron emission tomography
RAS	renal artery stenosis
RBC	red blood cell
SBFT	small bowel follow through
SPECT	single photon emission computed tomography
VC	virtual colonoscopy (CT colonography)
PFT	pulmonary function tests
WBC	white blood cell
ZES	Zollinger-Ellison Syndrome

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General Guidelines (AB-1.0)

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- A pertinent clinical evaluation since the new onset or change in symptoms is required prior to advanced imaging:
  - A pertinent clinical evaluation should include the following:
    - A detailed history and physical examination relevant to the current complaint
    - Appropriate laboratory studies
    - Non-advanced imaging modalities such as plain x-ray or ultrasound of the area of concern
  - For an established individual, a meaningful technological contact (telehealth visit, telephone call, electronic mail, or messaging) since the onset of change in symptoms can serve as a pertinent clinical evaluation.

Red Flag Findings

- The following signs and symptoms can be indicative of more serious conditions. Documentation of abdominal pain along with ANY of the following warrants exclusion from prerequisites to advanced imaging:
  - History of malignancy with a likelihood or propensity to metastasize to abdomen
  - Immunocompromised individual (e.g., on immunosuppressive therapy, history of HIV)
  - Fever (≥101 degrees Fahrenheit)
  - Elevated WBC >10,000, or above the upper limit of normal for the particular lab reporting the result
  - Low WBC (absolute neutrophil count <1000)
  - Palpable mass of clinical concern and/or without benign features
  - GI bleeding, overt or occult, not obviously hemorrhoidal
  - Abdominal tenderness documented as moderate or severe
  - Peritoneal signs, such as guarding or rebound tenderness
  - Suspected complication of bariatric surgery
  - Notation by the ordering provider that the individual has a "surgical abdomen"
  - Age ≥60 years with unintentional weight loss of ≥10 lbs. or ≥5% of body weight over 6 months or less, without an identifiable reason
- See the condition-specific sections for when the above list of exclusionary criteria apply and lead directly to advanced imaging.

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Imaging Recommended Per Drug Manufacturer

- When follow-up imaging for the purposes of monitoring or screening is recommended in the package insert for a particular drug therapy or medication, that imaging may be medically necessary.

Complications Related to COVID-19

- Please refer to the appropriate condition-specific guideline relevant to the presenting signs or symptoms in individuals with potential sequelae of COVID-19.
  - Examples include:
    - For suspected acute mesenteric ischemia, see: **Mesenteric Ischemia (AB-6.1)**
    - For suspected renal failure, see: **Renal Failure (AB-36.1)**

Pre-operative Radiologic Imaging

- If imaging is requested by the operating surgeon to support planned surgery, the imaging is considered medically necessary.
- Please refer to the appropriate condition-specific guideline relevant to the clinical condition for pre-operative imaging indications (e.g., **Percutaneous Gastrostomy (AB-9.2)**)
- Radiologic therapeutic intervention is addressed elsewhere in this Guideline.
  - Radiologic management of lower GI bleeding, see: **Small Bowel Bleeding Suspected (AB-22.2)**
  - Radiologic management of mesenteric ischemia, see: **Mesenteric/Colonic Ischemia (AB-6.1)**
  - Radiologic management of portal hypertension, see: **Portal Hypertension (AB-26.3)**

3D Rendering

- CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT® 76376 (3D rendering not requiring image post-processing on an independent workstation) is medically necessary in the following clinical scenarios:
  - Pre-operative planning for complex surgical cases
  - CT Urogram (See: **Hematuria and Hydronephrosis (AB-39)**)
  - MRCP (See: **MR Cholangiopancreatography (MRCP) (AB-27)**)
- CPT® codes for 3D rendering should not be billed in conjunction with computer-aided detection (CAD), MRA, CTA, nuclear medicine SPECT studies, PET, PET/CT, or CT Colonography (virtual colonoscopy).

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social

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conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

Evidence Discussion

Except as noted in condition-specific sections of these Abdominal Guidelines, initial evaluation by ultrasound is generally prerequisite to advanced imaging modalities. Ultrasound requires no ionizing radiation, is cost effective, helps determine most appropriate next advanced imaging study (CT vs. MRI) and contrast level, is readily accessible, and often can be scheduled same day.

When Red Flag signs and symptoms are present, literature supports early use of computer tomography (CT) and/or magnetic resonance imaging (MRI) without need for a prior ultrasound. Red Flags include:

- Risk of metastases: Liver, lung, and regional lymph nodes are frequent metastatic targets readily identified by advanced abdominal imaging. Metastatic foci are less readily identified by ultrasound in the hollow viscus than solid abdominal organs - e.g., in high prevalence metastatic spread to the gas-filled stomach by breast cancer (27%), lung cancer (23%), renal cell cancer (7.6%), and malignant melanoma (7%).[20]
- Fever: Accompanied by abdominal pain, or in combination with vomiting, bloody stools, unexplained weight loss, or persistent fever requires urgent imaging evaluation. CT and MRI are better suited than ultrasound in localizing and characterizing gut-related urgencies such as bowel blockage, abdominal ischemia, acute inflammatory conditions (diverticulitis, flares of inflammatory bowel disease, perforation), and obstructing tumors.[21]
- Abnormal white cell number: Neutropenia or leukocytosis warrants definitive advanced imaging to avoid delays in diagnosis and treatment, especially in immunocompromised settings, for life-threatening pathology such as neutropenic enterocolitis (typhilitis) or the various infectious, inflammatory, or injurious conditions described in the Abdominal Guideline sections in which an elevated white cell count is seen.[22]
- Concerning palpable mass: The imaging approach to diagnosis varies by location and clinician-concern. For intra-abdominal masses, contrast-enhanced CT and ultrasound examination have demonstrated accuracy. For abdominal wall masses, which may arise from muscle, subcutaneous tissue, or connective tissue, MRI, CT, and ultrasound all provide diagnostic value. When mass is accompanied by abdominal pain, advanced imaging modalities may facilitate care.[23]

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- GI bleeding: When the source of bleeding is unidentified after upper endoscopy and/or colonoscopy, subsequent diagnostic modalities should be guided by clinical presentation, hemodynamic stability, and local expertise. CT angiography demonstrates a sensitivity of 86% and specificity of 95% in acute GI bleeding and is useful in directing definitive hemostatic treatment.[24]
- Significant abdominal tenderness, with or without peritoneal signs: Rapid onset of severe abdominal pain with significant tenderness, an acute abdomen or surgical abdomen, may indicate a potentially life-threatening condition requiring urgent surgical intervention for which accurate and timely diagnosis is critical. Advanced imaging also offers greater accuracy than ultrasound in the setting of a painless acute abdomen seen in older people, children, the immunocompromised, and in the last trimester of pregnancy.[25]
- Suspected complication of bariatric surgery: Early advanced imaging followed by emergent intervention avoids morbidity in Roux-en-Y individuals with internal hernias or in balloon recipients with bowel obstruction or perforated gastrojejunal ulcer.[26]
- Unexplained weight loss: Problematic weight loss in the older adult is defined by the United States Omnibus Budget Reconciliation Act of 1987 (Title IV: subtitle C: Nursing Home Reform) as a loss of 5% of body weight in one month or 10% over a period of six months or longer. Unintentional weight loss is associated with an increased risk of death among older adults.[27]

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Overview (AB-1.1)

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- GI Specialist evaluations can be helpful, particularly in determining mesenteric/colonic ischemia, diarrhea/constipation, irritable bowel syndrome (IBS), or need for MRCP.
- Abdominal imaging begins at the diaphragm and extends to the umbilicus or iliac crest.
- Pelvic imaging begins at the iliac crest and extends to the pubis.
  - The uterus, fallopian tubes and ovaries arise out of the pelvis and are considered pelvic organs. If the uterus or adnexa rise out of the pelvic cavity, above the level of the iliac crest or umbilicus on physical exam or previous imaging, abdominal imaging may be added for the completion of imaging.
- Imaging for clinical concerns at the dividing line can be providers' choice (abdomen and pelvis; abdomen or pelvis).

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CT Imaging (AB-1.2)

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- CT imaging is a more generalized modality. CT Abdomen is usually performed with contrast (CPT® 74160):
  - Oral contrast has no relation to the IV contrast administered. Coding for contrast only refers to IV contrast. There is no coding for oral contrast.
  - Exceptions are noted in these guidelines and include:
    - CT Abdomen with contrast (CPT® 74160) or without and with contrast (CPT® 74170) with suspicion of a solid organ lesion (liver, kidney, pancreas, spleen)
      - Please refer to the specific guideline for the lesion in question for specific guidance.
    - CT Abdomen without contrast (CPT® 74150) or CT Abdomen and Pelvis without contrast (CPT® 74176) if there is renal insufficiency/failure, or a documented allergy to contrast. It can also be considered medically necessary for diabetics or the very elderly.
    - CT Abdomen and Pelvis without and with contrast (CPT® 74178 – CT Urogram) for certain urologic conditions (e.g. hematuria).
  - Shellfish allergy:
    - It is commonly assumed that an allergy to shellfish infers iodine allergy, and that this implies an allergy to CT iodinated contrast media. However, this is NOT true. Shellfish allergy is due to tropomysins. Iodine plays no role in these allergic reactions. Allergies to shellfish do not increase the risk of reaction to IV contrast any more than that of other allergens.
  - CT Abdomen and Pelvis, usually with contrast (CPT® 74177), is considered medically necessary when signs or symptoms are generalized, or involve a lower quadrant of the abdomen.
  - CT Enterography (CPT® 74177) combines CT imaging with large volumes of ingested neutral bowel contrast material to allow visualization of the small bowel.
  - CT Enteroclysis
    - A tube is placed through the nose or mouth and advanced into the duodenum or jejunum. Bowel contrast material is infused through the tube and CT imaging is performed either with or without intravenous contrast.
    - CT Enteroclysis is used to allow visualization of the small bowel wall and lumen. CT Enteroclysis may allow better or more consistent distention of the small bowel than CT Enterography.
    - Report by assigning: CPT® 74176 or CPT® 74177
  - Triple-phase CT
    - 3 phases of a triple-phase CT are:

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- 1) Hepatic arterial phase,
- 2) Portal venous phase, and
- 3) Washout or delayed acquisitions phase.

▪ It should be noted that, in general, a pre-contrast or non-contrast CT is usually not needed in a standard triple-phase CT, except in those individuals previously treated with locoregional embolic or ablative therapies. Other specific instances in which a prior non-contrast CT is medically necessary for the evaluation of liver lesions are noted in **Liver Lesion Characterization (AB-29.1)**.

- CT Colonography (CTC)
  - There are 3 CPT® codes for CTC:
    - CPT® 74263: Screening CTC (only used for screening procedures)
    - CPT® 74261: CTC without contrast
    - CPT® 74262: CTC with contrast
  - See: **CT Colonography (CTC) (AB-25)** for further indications for these procedures

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MR Imaging (AB-1.3)

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- MRI may be preferred as a more targeted study in cases of renal failure, in individuals allergic to intravenous CT contrast, and as noted in these guidelines.
  - MRI Abdomen with contrast only is essentially never performed. If contrast is indicated, MRI Abdomen without and with contrast (CPT® 74183) should be considered medically necessary.
  - For pregnant individuals ultrasound or MRI without contrast should be used to avoid radiation exposure. The use of gadolinium contrast agents is limited during pregnancy, as gadolinium contrast agents cross the placenta and enter the amniotic fluid with unknown long-term effects on the fetus.
    - See: **Pregnancy Considerations for Imaging (AB-1.12)** for additional discussion of this issue
- MR Elastography (CPT® 76391) replaces MRI Abdomen (CPT® 74183 or CPT® 74181) for requests for MR Elastography liver (See: **Liver Elastography (AB-45)**)

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MR Enterography and Enteroclysis Coding Notes (AB-1.4)

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- MR Enterography or Enteroclysis is reported in one of two ways:
  - MRI Abdomen without and with contrast (CPT® 74183), or
  - MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis with and without contrast (CPT® 72197)

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Ultrasound (AB-1.5)

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- Ultrasound, also called sonography, uses high frequency sounds waves to image body structures.
  - The routine use of 3D and 4D rendering (post-processing) in conjunction with ultrasound is not medically necessary.
  - All ultrasound studies require permanently recorded images either stored on film or in a Picture Archiving and Communication System (PACS).
  - The use of a hand-held or any Doppler device that does not create a hard-copy output is considered part of the physical examination and is not separately billable. This exclusion includes devices that produce a record that does not permit analysis of bi-directional vascular flow.
- Duplex scan describes an ultrasonic scanning procedure for characterizing the pattern and direction of blood flow in arteries and veins with the production of real-time images integrating B-mode 2D vascular structures, Doppler spectral analysis, and color flow Doppler imaging.
  - The minimal use of color Doppler alone, when performed for anatomical structure identification during a standard ultrasound procedure, is not separately reimbursable.

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Abdominal Ultrasound (AB-1.6)

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- Complete abdominal ultrasound (CPT® 76700) includes all of the following required elements:
  - Liver, gallbladder, common bile duct, pancreas, spleen, kidneys, upper abdominal aorta, and inferior vena cava
  - If a particular structure or organ cannot be visualized, the report should document the reason.
- Limited abdominal ultrasound (CPT® 76705) is without all of these required elements and can refer to a specific study of a single organ, a limited area of the abdomen, or a follow-up study.
  - Further, CPT® 76705 should:
    - Be assigned to report follow-up studies once a complete abdominal ultrasound (CPT® 76700) has been performed; and
    - Be assigned to report ultrasonic evaluation of diaphragmatic motion; and
    - Be reported only once per individual imaging session; and
      - Not be reported with CPT® 76700 for the same individual for the same imaging session

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Retroperitoneal Ultrasound (AB-1.7)

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- Complete retroperitoneal ultrasound (CPT® 76770) includes all of the following required elements:
  - Kidneys, lymph nodes, abdominal aorta, common iliac artery origins, inferior vena cava
  - For urinary tract indications, a complete study can consist of kidneys and bladder
- Limited retroperitoneal ultrasound (CPT® 76775) studies are without all of these required elements and can refer to a specific study of a single organ, a limited area of the abdomen, or a follow-up study.
  - Further, CPT® 76775 should:
    - Be assigned to report follow-up studies once a complete retroperitoneal ultrasound (CPT® 76770) has been performed; and
    - Be reported only once per individual imaging session; and
      - Not be reported with CPT® 76770 for the same individual for the same imaging session

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CT-, MR-, Ultrasound-guided Procedures (AB-1.8)

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See: **CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2)** in the Preface Imaging Guidelines

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Contrast-Enhanced Ultrasound (AB-1.9)

AB.GG.0001.9.A

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Ultrasound with contrast (CEUS, CPT® 76978, CPT® 76979) is an emerging technology that may be as good, if not better, than CT or MRI in certain circumstances. Abdominal Imaging Guidelines address its use as appropriate. CPT® 76978 refers to the initial imaging of the first lesion, and CPT® 76979 refers to additional lesions that are imaged subsequently.

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Quantitative MRI (AB-1.10)

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- Quantitative MRI analysis of tissue composition (CPT® 0648T, 0649T, 0697T and 0698T)
  - These CPT codes are experimental and investigational.
  - See: **Quantitative MR Analysis of Tissue Composition (Preface-4.8)** and **Fatty Liver (Metabolic Associated Steatotic Liver Disease (MASLD), Formerly Known as NAFLD) (AB-29.2)** for further discussion of these modalities.

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RADCAT Grading System (AB-1.11)

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- The RADCAT (Radiology Report Categorization) Grading System was developed in order to communicate to ordering physicians (most commonly in the ER setting) the relative urgency of a radiologic finding. It is not related to the LI-RADs reporting system, nor does it necessarily imply the need for follow-up imaging, as opposed to clinical follow-up. The rating system is as follows:
  - RADCAT 1: Normal Result
  - RADCAT 2: Routine Result
  - RADCAT 3: Result with recommendation for non-urgent routine follow-up
  - RADCAT 4: Priority Result
  - RADCAT 5: Critical Result

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Pregnancy Considerations for Imaging (AB-1.12)

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The American College of Obstetricians and Gynecologists has issued guidelines with regards to imaging during pregnancy and lactation. Their recommendations are as follows:[15]

- Ultrasonography and magnetic resonance imaging (MRI), which are not associated with exposure to ionizing radiation, are the imaging modalities of choice for pregnant individuals. These modalities should be limited to situations in which the results are expected to address a clinically relevant question or otherwise provide medical benefit to the individual.
- With few exceptions, radiation exposure through radiography, computed tomography (CT) scan, or nuclear medicine imaging techniques is at a dose much lower than the exposure associated with fetal harm.
  - If these techniques are necessary in addition to ultrasound or MRI or are more readily available for the diagnosis in question, they should not be withheld from a pregnant individual.
- The use of gadolinium contrast with MRI should be limited; it may be used as a contrast agent in a pregnant individual only if it significantly improves diagnostic performance and is expected to improve fetal or maternal outcome.
- With regards to iodinated IV contrast media, “it is generally recommended that contrast only be used if absolutely required to obtain additional diagnostic information that will affect the care of the fetus or woman during pregnancy.”

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Abdominal Pain (AB-2)

Guideline

Abdominal Pain (AB-2.2)
Right Upper Quadrant and Epigastric Pain (AB-2.3)
References (AB-2)

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Abdominal Pain (AB-2.2)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- For pregnant individuals, see **Pregnancy Considerations for Imaging (AB-1.12)**
  - Abdominal and/or Pelvic and/or Transvaginal ultrasound (CPT® 76700 and/or CPT® 76856 and/or CPT® 76830) is medically necessary initially to avoid ionizing radiation.
  - MRI Abdomen and/or MRI Pelvis without contrast (CPT® 74181 and or CPT® 72195) is medically necessary if the initial US is considered non-diagnostic.
- For the evaluation of left lower (including suspected diverticulitis), right lower (including suspected appendicitis), left upper, mid-abdominal, and/or non-specific abdominal pain regardless of duration (acute or chronic, defined respectively as less than or to equal to 6 months, or greater than or equal to 6 months):
  - CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if ANY of the following are present:
    - Age ≥65
    - Prior Abdominal and/or Pelvic US appropriate to the area of concern have been performed and demonstrate a need for additional imaging OR do not explain the source of pain
    - Recent laboratory studies related to the current episode have been performed and are non-diagnostic OR do not point to a specific etiology
      - Laboratory studies may include: CBC with differential, CMP, BMP, chemistry profile including electrolytes, glucose, creatinine, BUN with liver chemistries, ESR, amylase and lipase, CRP, inflammatory markers, pregnancy testing for reproductive age individuals, and urinalysis
        - All the specific laboratory studies listed are not required, but a relevant work-up should be performed in order to direct further advanced imaging appropriately.
- For follow-up imaging of diverticulitis:
  - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for ANY of the following:
    - acute diverticulitis if symptoms or elevated WBC persists despite treatment
    - diverticulitis treated with radiologic intervention (e.g., drainage procedure)
    - complicated diverticulitis, including confirmed abscess, fistulae, free fluid, or perforation. (See: **Abdominal Sepsis/Suspected Abdominal Sepsis) (AB-3)**
- For the evaluation of non-operative treatment of acute appendicitis:

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  - One-time CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for EITHER of the following:
    - symptoms suspicious for recurrent appendicitis OR
    - post non-operative treatment follow-up
- For the evaluation of splenic etiologies such as suspected infarct or abscess (severe pain and tenderness, fever, history of atrial fibrillation):
  - CT Abdomen without and with contrast or with contrast (CPT® 74170 or CPT® 74160)
- For the evaluation of suspected small or large bowel etiologies (e.g., ischemia, obstruction, volvulus, or perforation):
  - CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177)
- For the evaluation of suspected gastroenteritis and/or enterocolitis in the presence of any red flag findings:
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
    - If no red flag findings are present, initial evaluation should include history and physical exam, as well as relevant laboratory testing and/or abdominal x-ray.
    - For suspected ischemic enterocolitis, see: **Mesenteric Ischemia (AB-6.1)** or **Colonic Ischemia (Including Ischemic Colitis) (AB-6.2)**
    - For known or suspected IBD, see: **IBD (Crohn's Disease or Ulcerative Colitis) (AB-23.1)**
- For pain described as pelvic, see: **Pelvic Pain/Dyspareunia, Female (PV-11.1)**, **Male Pelvic Disorders (PV-19.1)** or other appropriate sections based on likely etiology.

CPT® Codes for Abdominal Pain (AB-2.2)			
CPT® 74150	CT Abdomen without contrast	CPT® 76700	Ultrasound, complete Abdomen
CPT® 74160	CT Abdomen with contrast	CPT® 76705	Ultrasound, limited Abdomen
CPT® 74176	CT Abdomen and Pelvis without contrast	CPT® 76830	Ultrasound, Transvaginal
CPT® 74177	CT Abdomen and Pelvis with contrast	CPT® 76856	Ultrasound, complete Pelvis
CPT® 74181	MRI Abdomen without contrast	CPT® 72195	MRI Pelvis without contrast

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CPT® Codes for Abdominal Pain (AB-2.2)			
CPT® 74183	MRI Abdomen without and with contrast	CPT® 72197	MRI Pelvis without and with contrast

Evidence Discussion

There are many potential causes of abdominal pain. Standard-of-care evaluation generally begins with complete history, physical examination, and directed laboratory testing. The initial approach establishes differential diagnoses and clinical options. It also clarifies what imaging modality and body region are medically necessary to achieve the highest diagnostic yield while reducing radiation exposure and overutilization.[1,3,5]

In the absence of red flags, Abdominal and/or Pelvic ultrasound is generally cost effective when performed early in the diagnostic paradigm. Although image quality may degrade due to overlying gas in the gut, US offers advantages in that it requires no contrast agents nor radiation exposure, can be used in pregnancy, and is readily available in evaluation of most conditions manifesting with abdominal pain. [1,3,5]

Per ASCRS, colonic endoscopic evaluation is recommended to confirm the diagnosis after resolution of acute diverticulitis to exclude malignancy, especially when initial CT scan supports abscess, shouldering, or shelf-like appearance of a presumed inflammatory mass, obstruction, mesenteric or retroperitoneal adenopathy.[18]

Non-operative treatment of acute appendicitis offers a unique imaging opportunity. CT Abdomen and Pelvis with contrast is preferred to assess post-treatment response, to identify coexisting pathology masked by appendiceal inflammation, and when surgery is considered for symptoms that recur or fail to improve due to progression of disease or complications.[7]

Advanced imaging is further warranted, especially in follow-up to non-diagnostic ultrasound, when both acute and non-acute signs and symptoms pose a diagnostic and therapeutic challenge. The optimal choice of imaging modality is guided by contemporary medical literature and specialty societal recommendations.

- CT imaging of the Abdomen and Pelvis provides high diagnostic value as initial imaging in red flag circumstances and/or in follow-up when serial imaging is necessary as defined by guidelines or ultrasound is indeterminate. CT imaging can characterize gut-related urgencies including, but not limited to, bowel blockage, abdominal ischemia, and obstructing tumors. CT is highly sensitive for acute inflammatory conditions, such as diverticulitis and appendicitis. CT offers relatively brief imaging acquisition times but requires use of ionizing radiation typically in combination with intravenous contrast, making it non-ideal in pregnant individuals.[3,5,7]

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- MRI is typically reserved for pregnant individuals when ultrasound proved non-diagnostic. MRI is also relatively contraindicated for individuals who cannot tolerate confined space, long image acquisition times, and loud repetitive noise. Relative to evaluation of abdominal pain, the American College of Radiology opined "MRI is less sensitive for extraluminal air and urinary tract calculi, is more time-consuming to perform, and requires an active screening process for indwelling devices and metal, and more subject to motion artifacts in symptomatic individuals."[3,5,7]

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Right Upper Quadrant and Epigastric Pain (AB-2.3)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- For pregnant individuals, see: **Pregnancy Considerations for Imaging (AB-1.12)**
- For the evaluation of right upper quadrant pain in the absence of red flag findings:
  - Abdominal ultrasound (complete or limited) (CPT® 76700 or CPT® 76705) is the initial diagnostic test
  - CT Abdomen with contrast (CPT® 74160) or MRCP/MRI (MRI Abdomen without or without and with contrast) (CPT® 74181 or CPT® 74183) if ultrasound is equivocal or nondiagnostic
- For the evaluation of epigastric pain or dyspepsia in the absence of red flag findings:
  - Ultrasound Abdomen (CPT® 76700 or CPT® 76705) to assess for biliary/ pancreatic disease is the initial study
  - CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183), or MRCP (CPT® 74181 or CPT® 74183), is medically necessary to further evaluate non-diagnostic findings on ultrasound
  - CT Abdomen with contrast (CPT® 74160), or MRI Abdomen without and with contrast (CPT® 74183) is medically necessary for persistent symptoms after a negative or inconclusive ultrasound as well as ONE of the following:
    - Test and treat for Helicobacter pylori (H. pylori) and a trial of acid suppression with a proton pump inhibitor (PPI) for 4-8 weeks if eradication is successful, but symptoms do not resolve OR
    - An empiric trial of acid suppression with a PPI for 4-8 weeks
- Hepatobiliary System Imaging (HIDA) with OR without pharmacologic intervention (CPT® 78226 or CPT® 78227) is medically necessary for ANY of the following:
  - If there is right upper quadrant pain or epigastric pain and there is a suspicion of gallbladder disease, with a normal, or equivocal or non-diagnostic recent ultrasound, CT, or MRI
    - If findings on US suggest acute cholecystitis in a symptomatic individual (presence of gallstones with gallbladder wall thickening, Murphy's sign, and peri-cholecystic fluid) then a HIDA scan is generally not medically necessary.
    - If the HIDA without pharmacologic intervention (CPT® 78226) is initially performed and is normal or inconclusive, the site can convert the study to HIDA with pharmacologic intervention (CPT® 78227). The member will not need to return for a second study with injection of a pharmaceutical.

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- Suspected bile leak after trauma or surgery
- Monitoring of liver regeneration
- Assessment of liver transplant
- Assessment of choledochal cyst
- Pre-operative assessment prior to partial hepatectomy
- Chronic acalculous cholecystitis
- Biliary dyskinesia
- Functional gallbladder disease
- Sphincter of Oddi dysfunction

Special Considerations for Suspicion of Pancreatic Cancer

CT Abdomen with contrast (CPT® 74160), CT Abdomen and Pelvis with contrast (CPT® 74177), or MRI Abdomen without and with contrast (CPT® 74183) is appropriate for suspicion of pancreatic cancer in individuals aged ≥60 years with weight loss and any ONE of the following:

- diarrhea
- back pain
- abdominal pain
- nausea
- vomiting
- constipation
- new onset diabetes
- abnormal lab results raising the possibility of pancreatic cancer (e.g., elevated CA 19-9, GGTP, alkaline phosphate, or bilirubin)
- non-diagnostic or negative prior US

If none of the above signs or symptoms applies, follow criteria for epigastric pain and dyspepsia.

See also: **Pancreatic Cancer-Suspected Diagnosis (ONC-13.2)** in the Oncology Imaging Guidelines

Evidence Discussion

Many specific conditions may manifest with pain in the right upper abdominal quadrant and/or epigastrium. Dyspepsia is common and often best evaluated by empiric response to acid suppression therapy, assessment of treatment efficacy when H.Pylori is identified, and endoscopy.[2,4,6,7]

For non-peptic causation of right upper and epigastric abdominal pain, ultrasound is the initial imaging study. Ultrasound is a readily available modality that offers good diagnostic utility while eliminating exposure to ionizing radiation. Ultrasound when

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diagnostic may confirm diagnoses of biliary and/or pancreatic disease, of space-occupying lesions, and of vascular concerns.[2,4,6,7]

When indeterminate, ultrasound may help direct what advanced imaging modality and contrast is medically necessary—e.g., MRCP/ERCP for dilated biliary ducts, CT for pancreatitis, MRI/CT with and without contrast to better characterize abdominal mass lesions. While CT is highly sensitive for acute inflammatory conditions and offers relatively brief image acquisition times, it requires use of ionizing radiation. MRI is typically reserved for pregnant individuals when ultrasound proved non-diagnostic but is relatively contraindicated for individuals who cannot tolerate confined space, prolonged image acquisitions, loud repetitive noise. Relative to evaluation of abdominal pain, the American College of Radiology opined "MRI is less sensitive for extraluminal air and calculi, is more time-consuming to perform, and requires an active screening process for indwelling devices and metal, and more subject to motion artifacts in symptomatic individuals."[2,4,6,7]

Hepatobiliary System Imaging (HIDA) with radionuclide uptake and scan in suspected hepatic, biliary, and pancreatic disease may add prognostic and functional information when ultrasound is inconclusive. HIDA scan has particular pre-operative value in the setting of gallbladder dysfunction, bile leaks, and choledochal cyst.[2,4,6,7]

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Abdominal Sepsis (Suspected Abdominal Abscess) (AB-3)

Guideline

Abdominal Sepsis/Abscess (AB-3.1)
Reference (AB-3)

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Abdominal Sepsis/Abscess (AB-3.1)

AB.AS.0003.1.A

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- For abdominal symptoms associated with fever and/or elevated white blood cell count ANY of the following are medically necessary:
  - CT Abdomen with contrast (CPT® 74160)
  - CT Pelvis with contrast (CPT® 72193)
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
- For intraperitoneal abscess:
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
    - Interval imaging as requested
- For follow-up imaging of known abnormal fluid collections after medical management or catheter drainage, ANY of the following are medically necessary:
  - Serial Ultrasound (CPT® 76705)
  - CT Abdomen with contrast (CPT® 74160)
  - CT Pelvis with contrast (CPT® 72193)
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
    - The interval can be days, weeks, or months based on the clinical course of the individual

Evidence Discussion

- Individuals presenting with potential abdominal sepsis or an abscess represent an urgent clinical concern. Therefore, individuals exhibiting abdominal symptoms accompanied by fever or an elevated WBC count (or any red flag) should proceed directly to advanced imaging without further evaluation. A CT scan of the abdomen and/or pelvis with contrast is typically the appropriate study for such evaluations.[2]
- Interval imaging may be medically necessary for abscesses or other fluid collections, particularly after catheter drainage. Both ultrasound and CT imaging are appropriate for serial imaging. The timing of serial imaging is not specified and should be based on the individual's unique clinical course.[1]

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Flank Pain, Rule Out or Known Renal/ Ureteral Stone (AB-4)

Guideline

Ultrasound (AB-4.0)
Suspected Renal/Ureteral Stone(s) (AB-4.1)
Observation of Known Renal/Ureteral Stone(s) (AB-4.2)
Follow-Up of Treated Renal/Ureteral Stone (AB-4.3)
Annual Surveillance (AB-4.4)
Nuclear Kidney Imaging (AB-4.5)
References (AB-4)

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Ultrasound (AB-4.0)

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- For the evaluation of flank pain and/or suspected renal or ureteral calculi, retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) is medically necessary in place of CT Abdomen and Pelvis for any initial or follow-up indications, if requested by provider.

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Suspected Renal/Ureteral Stone(s) (AB-4.1)

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- CT Abdomen and Pelvis without contrast (CPT® 74176) is medically necessary for ANY of the following:
  - Suspected renal/ureteral stone with symptoms in non-pregnant adults (flank pain/ renal colic) OR
  - Suspected staghorn calculi
- CT Abdomen and Pelvis without contrast (CPT® 74176) or CT Urogram (CPT® 74178) is medically necessary for the following:
  - Suspicion renal/ureteral stones (flank pain/renal colic) with hematuria
- Ultrasound (CPT® 76770 or CPT® 76775) or MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195) is medically necessary for the following:
  - Suspected renal/ureteral stone in pregnant individuals (flank pain/renal colic)
    - The use of gadolinium contrast agents is contraindicated during pregnancy unless the specific need for that procedure outweighs risk to the fetus.
- Suspected renal/ureteral stone in children (flank pain/renal colic)
  - See: **Flank Pain, Renal Stone (PEDAB-4)** in the Pediatric Abdomen Imaging Guidelines

Evidence Discussion

Non-contrast CT (NCCT) is the imaging study of choice for initial evaluation of individuals with acute onset of flank pain and suspicion of stone disease without known prior stone disease. Non-contrast CT (NCCT) can reliably characterize the location and size of an offending ureteral calculus, identify complications, and diagnose alternative etiologies of abdominal pain. For individuals with known disease and recurrent symptoms of urolithiasis, NCCT remains the test of choice for evaluation.[17]

Although less sensitive in the detection of stones, ultrasound may have a role in evaluating for signs of obstruction. Radiography potentially has a role, although has been shown to be less sensitive than NCCT.[1]

In pregnancy, given radiation concerns, ultrasound is recommended as the initial modality of choice with potential role for non-contrast MRI.[4]

In scenarios where stone disease is suspected and initial NCCT is inconclusive, contrast-enhanced imaging, either with MRI or CT/CT Urogram may be medically necessary.[17]

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Observation of Known Renal/Ureteral Stone(s) (AB-4.2)

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- Radiopaque Stones
  - Initial follow-up imaging:
    - Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) and KUB X-ray
  - Subsequent follow-up imaging:
    - If initial follow-up ultrasound and KUB are negative, and there is no hematuria and individual is asymptomatic:
      - See: **Annual Surveillance (AB-4.4)**
    - If initial follow-up ultrasound and KUB demonstrates hydronephrosis, retained stone, or if the individual has persistent hematuria, or is symptomatic:
      - CT Abdomen and Pelvis without contrast (CPT® 74176)
- Non-radiopaque Stones (i.e. radiolucent)
  - Initial follow-up imaging:
    - CT Abdomen and Pelvis without contrast (CPT® 74176)
  - Subsequent follow-up imaging:
    - If CT is negative:
      - See: **Annual Surveillance (AB-4.4)**
    - If CT demonstrates a retained stone, hydronephrosis, or if the individual is being evaluated for surgery:
      - Further imaging can be considered on an individual basis
- ANY of the following are medically necessary for surgical/procedural evaluation of staghorn calculi:
  - CT Abdomen and Pelvis (contrast as requested)
  - 3-D reconstruction (CPT® 76377 or CPT® 76376)
  - Nuclear kidney imaging (CPT® 78707, CPT® 78708, or CPT® 78709) when there is concern for a poorly functioning kidney

Background and Supporting Information

- Radiopaque versus radiolucent stones on plain radiograph:
  - Radiopaque
    - Calcium-based stones (70-80%)
    - Struvite stones (triple phosphate) (usually opaque but variable – 15-20%)
  - Radiolucent

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- Uric acid (5-10%)
- Cystine (1-3%)
- Medication stones (e.g. indinavir) (1%)

Evidence Discussion

Serial imaging can be used to follow the progress of a passing stone and might also be used by the urologist and/or nephrologist as they monitor non-obstructing stones for growth. No evidence was found on the optimum frequency of imaging in people who have or have had renal or ureteric stones.[15]

Non-contrast CT of the abdomen and pelvis consistently provides the most accurate diagnosis but also exposes individuals to ionizing radiation. Traditionally, ultrasonography has a lower sensitivity and specificity than CT but does not require use of radiation. However, when these imaging modalities were compared in a randomized controlled trial they were found to have equivalent diagnostic accuracy. Both modalities have advantages and disadvantages. Kidney, ureter, bladder (KUB) plain film radiography is most helpful in evaluating for interval stone growth in individuals with known stone disease and is less useful in the setting of acute stones. MRI provides the possibility of 3D imaging without exposure to radiation, but it is costly and currently stones are difficult to visualize.[16]

Follow-up imaging for asymptomatic individuals with radiopaque stones should be with retroperitoneal ultrasound and plain film radiography. Follow-up for radiolucent stones, hydronephrosis or retained stone on ultrasound, or symptomatic individuals, non-contrast CT is medically necessary.[16]

Individuals with staghorn calculi who are being considered for surgery, CT Abdomen and Pelvis (any contrast level), with or without 3-D reconstruction can be performed. Additionally, nuclear imaging may be medically necessary when there is concern for poor kidney function.[12]

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Follow-Up of Treated Renal/Ureteral Stone (AB-4.3)

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- Post-shock wave lithotripsy (SWL):
  - Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) is the appropriate initial follow-up imaging.
  - Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) and/or CT Abdomen and Pelvis (contrast as requested) is medically necessary for ANY of the following:
    - Individuals who are symptomatic
    - Individuals with hydronephrosis
    - Individuals who have residual fragments
  - Individuals treated by SWL who have passed fragments, are asymptomatic, and without hydronephrosis can be followed according to **Annual Surveillance (AB-4.4)**.
- Post-medical expulsive therapy (MET):
  - Retroperitoneal ultrasound is medically necessary for individuals treated by MET who have passed a stone and are symptomatic
  - CT Abdomen and Pelvis (contrast as requested) is medically necessary if hydronephrosis is demonstrated with ultrasound
  - Individuals treated by MET who have passed a stone and are asymptomatic can be followed according to **Annual Surveillance (AB-4.4)**.
- Post-ureteroscopic extraction with an intact stone:
  - Retroperitoneal ultrasound is medically necessary for individuals without symptoms.
  - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for individuals with symptoms or hydronephrosis demonstrated on ultrasound.
  - Individuals without symptoms or without hydronephrosis demonstrated on ultrasound can be followed according to **Annual Surveillance (AB-4.4)**.
- Post-ureteroscopic extraction requiring fragmentation of the stone(s):
  - Retroperitoneal ultrasound is medically necessary for individuals without symptoms.
  - CT Abdomen and Pelvis without contrast (CPT® 74176) is medically necessary for individuals without symptoms, but hydronephrosis demonstrated on ultrasound.
  - Individuals without symptoms or without hydronephrosis demonstrated on ultrasound can be followed according to **Annual Surveillance (AB-4.4)**.

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  - Retroperitoneal ultrasound and KUB for individuals with symptoms and a radiopaque stone
  - CT Abdomen and Pelvis without contrast (CPT® 74176) is medically necessary for individuals with symptoms and a non-radiopaque stone.
- Post-surgical/procedural treatment of staghorn calculi:
  - CT Abdomen and Pelvis without contrast (CPT® 74176)
  - Retroperitoneal ultrasound and/or CT Abdomen and Pelvis (contrast as requested) is medically necessary for individuals with persistent symptoms and/or hydronephrosis.

Evidence Discussion

Following treatment for renal stones, retroperitoneal ultrasound is the recommended initial modality for follow-up. CT scan is medically necessary in individuals with symptoms or if hydronephrosis identified on ultrasound. Ultrasound is subsequently recommended for annual surveillance in asymptomatic individuals.[7]

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Annual Surveillance (AB-4.4)

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- Annual surveillance is medically necessary for stable individuals with a history of stones to assess for stone growth or formation of new stones:
  - Plain x-ray (KUB) is medically necessary for individuals with radiopaque stones.
  - Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) is the preferred modality for individuals with non-radiopaque stones.

Evidence Discussion

Plain x-ray is cost-effective and readily available for surveillance of radiopaque stones. Ultrasound is preferred for most individuals with radiolucent stones. One year imaging interval is recommended for stable individuals, but this may be tailored on stone activity or clinical signs.[7]

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Nuclear Kidney Imaging (AB-4.5)

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- Nuclear kidney imaging (CPT® 78707, CPT® 78708, or CPT® 78709) is medically necessary for the evaluation of the following:
  - Recurrent flank pain when CT and ultrasound are non-diagnostic OR
  - Prior imaging (CT or ultrasound) shows hydronephrosis and to determine if this is truly obstructive in nature

Evidence Discussion

Renal scintigraphy is used for evaluation of renal perfusion and function as well as renal anatomy. Regarding anatomy, renal scintigraphy is currently used when there is an allergy to CT or MRI contrast material.[5] The use of IV contrast in CT, as well as MRI, is avoided in cases of abnormal renal function and altered GFR. Renal scintigraphy has a role in the diagnosis of obstructive uropathy. It can be used to differentiate true obstruction from non-obstructive simulators causing urinary tract dilation. Nuclear renal scanning is also an excellent modality for the qualitative as well as quantitative assessment of renal transplant function. Because radiation exposure from renal scintigraphy is very low as compared to a CT scan, it maintains a role in the evaluation of pediatric renal anatomy whether normal, anomalous, or pathologic.[6]

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Mesenteric/Colonic Ischemia (AB-6)

Guideline

Mesenteric Ischemia (AB-6.1)
Colonic Ischemia (Including Ischemic Colitis) (AB-6.2)
References (AB-6)

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Mesenteric Ischemia (AB-6.1)

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Acute Mesenteric Ischemia

- Suspicion of acute mesenteric ischemia, ONE of the following:
  - CTA Abdomen and/or Pelvis (Mesenteric) (CPT® 74175, or CPT® 74174, or CPT® 72191) (preferable), OR
  - MRA Abdomen and/or Pelvis (CPT® 72198 and/or CPT® 74185), OR
  - CT Abdomen and Pelvis with contrast (CPT® 74177)

Chronic Mesenteric Ischemia

- Suspicion of chronic mesenteric ischemia:
  - Mesenteric Artery Duplex Ultrasound (CPT® 93975 or CPT® 93976) and/or ONE of the following:
    - CTA Abdomen and Pelvis (CPT® 74174) OR
    - MRA Abdomen and Pelvis (CPT® 74185 and CPT® 72198)
- For clinical concern of median arcuate ligament syndrome, see: **Median Arcuate Ligament Syndrome, Nutcracker Syndrome and other Abdominal Vascular Compression Syndromes (PVD-18)** in the Peripheral Vascular Disease (PVD) Imaging Guidelines
- For suspicion of superior mesenteric artery syndrome, see: **Superior Mesenteric Artery (SMA) Syndrome (AB-20.4)**

Pre- and Post-Treatment for Mesenteric Ischemia

- Pre-operative evaluation, if not already performed (including prior to endovascular intervention):
  - CTA Abdomen and Pelvis (CPT® 74174)
- Post-procedure surveillance imaging following invasive treatment for mesenteric ischemia (celiac, superior mesenteric, and inferior mesenteric angioplasty with or without stenting, or mesenteric artery bypass grafting):
  - Baseline Duplex Ultrasound (CPT® 93975 or CPT® 93976) within 1 month of the procedure
  - Duplex Ultrasound (CPT® 93975 or CPT® 93976) at 6 months, 12 months, 18 months, and 24 months, then annually thereafter
  - CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) or CTA Abdomen (CPT® 74175) or CTA Abdomen and Pelvis (

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CPT® 74174) or MRA Abdomen (CPT® 74185) and if requested, MRA Pelvis (CPT® 72198):

- For symptoms suggesting recurrent ischemia OR
- In the absence of symptoms, following a Duplex Ultrasound if, on the Duplex study:
  - Celiac axis:
    - PSV >370 cm/s or a substantial increase from the post-treatment baseline PSV (substantial increase has not been defined) or demonstration of restenosis ≥70%
  - Superior mesenteric artery:
    - PSV >420 cm/s, or a substantial increase from the post-treatment baseline PSV (substantial increase has not been defined) or demonstration of restenosis of ≥70%
  - Inferior mesenteric artery:
    - Substantial increase from the post treatment baseline PSV (substantial increase has not been defined)

Surveillance of Asymptomatic Mesenteric Artery Occlusive Disease

- Annual Mesenteric Artery Duplex Ultrasound (CPT® 93975 or CPT® 93976)

Evidence Discussion

- Mesenteric ischemia reflects decreased intestinal blood flow through the mesenteric vessels. Causes include: mesenteric artery embolism (often seen with atrial fibrillation), mesenteric artery thrombosis (typically from progressive atherosclerosis that may range from non-occlusive low flow to frank occlusion), and mesenteric vein thrombosis (commonly due to hyper-coagulable states).[5]
- Typical presentation of acute mesenteric ischemia is based on severe abdominal pain out of proportion to findings on physical exam, usually in individuals with a combination of the following risk factors: advanced age, hyperlipidemia, heart disease, hypercoagulability, renal failure, inflammatory conditions (e.g., vasculitis, pancreatitis, diverticulitis), recent vascular catheterization, substance use (e.g., tobacco smoking, cocaine).[5]
- Chronic mesenteric ischemia (CMI) is a syndrome related to inadequate blood flow, typically related atherosclerotic occlusive disease affecting the mesenteric circulation. Blood flow to the bowel is from the celiac artery, superior mesenteric artery, and inferior mesenteric artery. Ischemia may occur when there is significant disease affecting at least two of three arteries; however, symptoms related to severe disease isolated to one artery is also possible. Symptoms may be characterized by postprandial abdominal pain, "food fear," diarrhea, or weight loss. Revascularization

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is typically recommended once CMI is diagnosed; this may be done via an endovascular approach (angioplasty and stenting) or through open reconstruction.[14]

- Duplex ultrasound provides an excellent screening tool for mesenteric artery occlusive disease. Duplex ultrasound is recommended for regular evaluation of individuals treated for mesenteric ischemia. Duplex ultrasound requires no ionizing radiation and is readily available. Duplex ultrasound findings help to determine the next most appropriate advanced imaging study if needed. Duplex ultrasound has a high negative predictive value of 99% with overall accuracy of 96% in ruling out significant stenosis.[10-13]
- CTA is recommended as an additional diagnostic tool in chronic mesenteric ischemia because it provides excellent image detail and helps to better define mesenteric lesions. Disadvantages of CTA include ionizing radiation, expense, and the need for a contrast agent.[1]
- MRA is considered an alternative modality to CTA. MRA boasts sensitivity and specificity of over 95% for detection of significant stenosis. However, it is limited in its ability to characterize degree of calcification, requires contrast administration, is not as widely available, and presents limitation in individuals with metallic implants.[12]

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Colonic Ischemia (Including Ischemic Colitis) (AB-6.2)

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- If colonic ischemia is suspected in the setting of significant abdominal pain, CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary as the first imaging modality if ANY of the following:
  - rectal bleeding OR
  - moderate or severe tenderness OR
  - fever (≥101 degrees) OR
  - guarding, rebound tenderness, or other peritoneal signs OR
  - elevated WBC as per the testing laboratory's range
- Repeat imaging for asymptomatic or improving individuals, including routine post-operative imaging, is generally not medically necessary.
- CTA Abdomen (CPT® 74175) or CTA Abdomen and Pelvis (CPT® 74174) or MRA Abdomen (CPT® 74185) AND if requested, MRA Pelvis (CPT® 72198) is medically necessary for the suspicion of right sided or pancolonic ischemia (per the initial CT Abdomen and Pelvis).

Background and Supporting Information

- Suspicion of colonic ischemia based on sudden cramping abdominal pain accompanied by urgency to defecate and passage of bright red blood, maroon blood, or bloody diarrhea, with risk factors including cardiovascular disease, diabetes mellitus, kidney disease, previous abdominal surgery, use of constipating medications, COPD, and atrial fibrillation.

Evidence Discussion

- Based on ACG Clinical Guideline: "In contrast to AMI (acute mesenteric ischemia) in which conventional mesenteric angiography or CTA plays an essential role, vascular imaging studies are not medically necessary in most individuals with suspected CI (colonic ischemia) because by the time of presentation, colon blood flow has usually returned to normal and the observed changes are not from ongoing ischemia but rather reflect the ischemic insult with or without reperfusion injury."[4]
- CT scan is recommended as first-line imaging for individuals with ischemic colitis. CT allows for identification and/or exclusion of other causes of abdominal pain; may suggest diagnosis of colonic ischemia, including distribution of disease; and may allow assessment of disease severity.[4]

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- CT-angiogram (CTA) is generally not medically necessary, since in most cases, blood flow has returned to normal by the time of clinical presentation. CTA may be helpful in distinguishing between acute mesenteric ischemia (AMI) and ischemic colitis. In diagnosing AMI, sensitivity and specificity are reported to be over 90%. Isolated right sided colonic ischemia (IRCI) carries a worse prognosis than other distributions of colitis and may represent evidence of significant SMA disease; as such, CTA is medically necessary to fully evaluate the vasculature and potentially prevent catastrophic associated complications.[4]
- Radiation and contrast related complications are risks associated with CT and CTA.[11]
- MRA also allows for evaluation of the proximal celiac artery and SMA. Advantages include high sensitivity and specificity. Disadvantages include poor visualization of distal vessels and non-occlusive ischemia, long acquisition times, and motion susceptibility artifact which could potentially delay treatment. In contrast to CTA, MRA is "less likely to show ischemic findings within the bowel itself."[11]
- Alternative imaging studies include non-contrast CT scan, ultrasound, and barium enema:
  - Non-contrast CT scan – there is a lack of literature related to this imaging modality; however, signs of ischemia, including evaluation of bowel and vasculature, rely on use of contrast.[11]
  - Ultrasound – Experience "in the setting of CI is very limited;" also, there is a low specificity, high false negative rate.[4]
  - Duplex US (arterial study) – there may be a role; however, various factor, including difficulty evaluating distal vessels and non-occlusive ischemia, as well as acquisition time, and individual discomfort limit utility in evaluating for acute mesenteric ischemia.[11]
  - Barium enema – originally described in diagnosis of CI in the 1960s. There is a very limited role today, as CT and colonoscopy are preferred. Modern usage is mainly to follow ischemic strictures in a chronic setting.[4]

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Post-Operative Pain Within 60 Days Following Abdominal Surgery – Abdominal Procedure (AB-7)

Guideline

Post-Op Pain and/or Complication Within 60 Days (AB-7.1)
References (AB-7)

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Post-Op Pain and/or Complication Within 60 Days (AB-7.1)

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- CT Abdomen and/or Pelvis with contrast (CPT® 74177, or CPT® 74160, or CPT® 72193) is medically necessary for suspected post-operative/post-procedure complications (e.g., bowel obstruction, abscess, anastomotic leak, or post-endoscopic complication).
- Beyond 60 days post-operatively, see: **Abdominal Pain (AB-2)**.
- For liver post-transplant indications and imaging, see: **Liver Transplant, Post-Transplant Imaging (AB-42.3)**
- For kidney post-transplant indications and imaging, see: **Kidney Transplant, Post Transplant (AB-42.6)**
- For suspected ureter or bladder injury indications and imaging, see **Ureteral and/or Bladder Trauma of Injury (PV-25.1)**

Evidence Discussion

Early investigation with advanced imaging is medically necessary to identify post-operative/post-procedural complications. Most complications manifest within the first 2 months.[1,2]

CT imaging is the mainstay for abdominal imaging in the post-operative period due to its high resolution and speed. It is particularly effective at identifying abdominal fluid collections in the peri-hepatic and peri-splenic areas, as well as in the pelvis. CT may also differentiate between post-operative seromas, hematomas, and abscesses, aiding in the drainage of these collections. The use of contrast is recommended to enhance diagnostic accuracy. [1,2]

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Abdominal Lymphadenopathy (AB-8)

Guideline

Abdominal Lymphadenopathy (AB-8.1)
Inguinal Lymphadenopathy (AB-8.2)
Sclerosing Mesenteritis and Mesenteric Panniculitis (AB-8.3)
References (AB-8)

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Abdominal Lymphadenopathy (AB-8.1)

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- History of malignancy
  - Refer to oncology guidelines specific for known malignancy
  - Biopsy may be considered
- For suspected lymphoproliferative disorder:
  - Imaging modality for biopsy is at the discretion of the interventional radiologist
    - Imaging studies performed as part of a CT-, MR-, or Ultrasound-guided procedure should be reported using appropriate CPT® codes, see: **CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2)**
  - PET/CT (CPT® 78815) is medically necessary prior to biopsy in order to determine a more favorable site for biopsy, when a prior biopsy was nondiagnostic, or a relatively inaccessible site is contemplated which would require invasive surgical intervention for biopsy attempt.
- If clinical, laboratory findings, biopsy, or PET suggest benign etiology, and no history of malignancy:
  - First follow-up imaging at 3 months
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary
  - Additional follow-up imaging at 6 and 12 months, if no improvement at 3 months
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary
  - Further imaging is NOT medically necessary if clinically significant changes are not seen within one year of follow-up imaging.
- If a follow-up CT demonstrates a concerning change, biopsy should be performed.
  - If biopsy is inconclusive, PET/CT (CPT® 78815) is medically necessary

Evidence Discussion

Abdominal lymphadenopathy can be associated with infectious, autoimmune, and malignant etiologies.[12] Lymph node size is a key indicator of potential malignancy. Lymph nodes exceeding 10 mm in short-axis diameter are generally considered suspicious, particularly in individuals with a history of cancer or systemic symptoms. Whenever possible, tissue pathology is preferred in the diagnosis of enlarged lymph nodes.[13]

CT remains the main modality for evaluation of intra-abdominal lymph nodes. This can be used for identification, follow-up, and guidance for percutaneous biopsy. Serial CT should be done with consideration of radiation exposure.[6,12]

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PET/CT, although not specific for malignancy, can assist in identifying alternate sites for biopsy in individuals with a previously non-diagnostic biopsy or when lymph nodes are relatively inaccessible and biopsy would require an invasive surgical intervention.[9]

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Inguinal Lymphadenopathy (AB-8.2)

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There is no evidence-based support for advanced imaging of clinically evidenced isolated inguinal lymphadenopathy without biopsy. Advanced imaging should be directed by results of biopsy. If biopsy results are negative or benign, then advanced imaging is not medically necessary.

- High suspicion of lymphoma: See **Non-Hodgkin Lymphomas (ONC-27)** and **Hodgkin Lymphoma (ONC-28)** in the Oncology Imaging Guidelines
- Prior history of malignancy: See **Metastatic Cancer, Carcinoma of Unknown Primary Site, and Other Types of Cancer (ONC-31)** in the Oncology Imaging Guidelines
- If biopsy is positive for malignancy, advanced imaging is guided by sections specific to the histological diagnosis.

Background and Supporting Information

- Localized inguinal lymphadenopathy should prompt:
  - search for adjacent extremity injury or infection
  - 3 to 4 weeks of observation if clinical picture is benign
  - excisional or image guided core needle biopsy under ultrasound or CT guidance of most abnormal lymph node if condition persists or malignancy suspected
- Generalized inguinal lymphadenopathy should prompt:
  - diagnostic work-up, including serological tests, for systemic diseases and
  - excisional or image guided core needle biopsy under ultrasound or CT guidance of most abnormal lymph node if condition persists or malignancy suspected

Evidence Discussion

Inguinal adenopathy is benign and self-limited in most individuals. History and physical alone can often identify the cause of the adenopathy. Biopsy remains the primary diagnostic tool in evaluation of undiagnosed inguinal adenopathy. This can be done with fine needle aspiration or core needle biopsy. Diagnostic rates can be improved with the use of ultrasound.[7]

There is no evidence-based support for advanced imaging of inguinal adenopathy in the absence of biopsy results that would direct that imaging. If benign, no further work-up is necessary.

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Sclerosing Mesenteritis and Mesenteric Panniculitis (AB-8.3)

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- For new or worsening clinical symptoms with a diagnosis of sclerosing mesenteritis or mesenteric panniculitis:
  - CT Abdomen and Pelvis without and with contrast (CPT® 74178) is medically necessary
- Requests for follow-up imaging in asymptomatic individuals or for sequential imaging to monitor for the development of malignancy is based upon size of lymph nodes:
  - Asymptomatic individuals with lymph nodes less than 10mm meet medical necessity for annual CT Abdomen and/or Pelvis with and without contrast OR MRI Abdomen and/or Pelvis with and without contrast for 2 consecutive years to ensure benign stability.
  - If lymph nodes are greater than or equal to 10mm, PET/CT (CPT® 78815) or biopsy is considered medically necessary.

Background and Supporting Information

- Sclerosing mesenteritis and mesenteric panniculitis are rare, incompletely understood entities that are characterized by an idiopathic inflammatory condition of the mesentery. The diagnosis is typically suggested by characteristic radiologic findings known as Coulier's criteria. To support the diagnosis, at least three of the five criteria should be met including:
  - mesenteric mass that encases or displaces adjacent structures without evidence of direct invasion
  - increased attenuation of mesenteric fat (hyperattenuating fat)
  - presence of soft-tissue nodules or mesenteric lymph nodes with the affected area
  - fatty halo (low-attenuation ring) surrounding lymph nodes or mesenteric vessels
  - hyperattenuating pseudocapsule encasing the mesenteric lesion
- Sclerosing mesenteritis may represent a spectrum of diseases (e.g., retractile mesenteritis, mesenteric panniculitis, and mesenteric lipodystrophy) or may be stages of one disease with progression.
- The chronic inflammation may result in fibrosis with a mass effect and can involve the gut (causing obstruction), the mesenteric vessels, and other intra-abdominal or retroperitoneal organs. The etiology is uncertain but may be secondary to trauma (previous abdominal surgery), an autoimmune process, ischemia, infection, and possibly may represent a paraneoplastic syndrome secondary to a malignancy, though this is controversial.

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- There is an increased prevalence of malignancy in individuals with sclerosing mesenteritis, and this has resulted in requests for sequential imaging in stable or asymptomatic individuals. In addition, requests may be made to assess the clinical response in those undergoing active treatment.
- However, studies have reported that the data on potentially developing a subsequent malignancy is inconclusive and thus “it does not seem justified to subject individuals with mesenteric panniculitis, especially those in whom other associations such as abdomino-pelvic surgery may explain the mesenteric panniculitis findings, to multiple follow-up CT scans with the aim of detecting a future malignancy.”
- In addition, there is no correlation between radiologic and clinical findings, and management decisions are guided by the severity and type of symptoms. Thus, sequential radiologic imaging to assess treatment response is not recommended.

Evidence Discussion

Recommendation for follow-up imaging in individuals with suspected malignancy-associated conditions vary significantly, ranging from no imaging to annual examination over five years. While some meta-analyses show no detectable increase in cancer risk, other studies suggest a slight long-term increased risk of lymphoma in individuals without initial malignancy indicators, with most cases detected within the first year. Imaging is also used to monitor inflammation progression, although data remains limited. In general, individuals with stable disease and no radiologic progression after two years do not require frequent imaging. However, if new suspicious features arise, such as significant growth in mesenteric masses or new lymphadenopathy, a biopsy is advised. For asymptomatic individuals without initial signs of malignancy, annual CT scans for two years are recommended to ensure disease stability, and thereafter performed for concerning symptoms.[10]

CT scan of the Abdomen and Pelvis is the preferred modality in the diagnosis of new or worsening symptoms. [11]

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Bariatric Surgery and Percutaneous Gastrostomy (AB-9)

Guideline

Bariatric Post-Operative Complications (AB-9.1)
Percutaneous Gastrostomy (AB-9.2)
References (AB-9)

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Bariatric Post-Operative Complications (AB-9.1)

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- For the evaluation of post-operative complications:
  - CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen with contrast (CPT® 74160), or CTA Abdomen and Pelvis (CPT® 74174) is medically necessary for individuals who have had weight loss surgery and present with suspected complications evidenced by:
    - nausea or vomiting OR
    - abdominal pain/tenderness OR
    - fever OR
    - abdominal distension OR
    - suspected hernia
- Internal hernias in individuals who have had Roux-en-Y gastric bypasses may have intermittent and relatively mild abdominal symptoms which require immediate evaluation with CT imaging.
- See: **Post-Operative Pain Within 60 Days Following Abdominal Surgery – Abdominal Procedure (AB-7)**

Background and Supporting Information

- Bariatric procedures include gastric banding, gastric bypass, sleeve gastrectomy, and biliopancreatic diversion procedures.

Evidence Discussion

- Pre-operative assessment:[4,11,16]
  - In individuals with previous surgery of the foregut, imaging may be medically necessary for surgical planning. This is addressed in **Abdomen Imaging Guidelines: General Guidelines (AB 1.0)** under pre-operative radiology imaging. "If imaging is requested by the operating surgeon to support planned surgery, the imaging may be approved."
  - Routine use of preoperative advanced imaging is considered "usually not appropriate" by the American College of Radiology ACR Appropriateness Criteria® for Preoperative and Postoperative Imaging for Bariatric Procedures.
- Post-operative complications:
  - Bariatric surgery can result in numerous complications that may not be apparent after initial evaluation or ultrasound. These include internal hernias, marginal

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ulceration, intussusception, stenosis, perforations, and leaks. A systematic review of splenic complications showed that splenic abscess and splenic infarct were the most common splenic complications.[12] Symptoms concerning for complications include weight loss failure, heartburn, nausea and vomiting, abdominal pain, fever, abdominal distention, abdominal tenderness, leukocytosis, and suspicion of a hernia.[12]

- CT is highly sensitive in the setting of symptomatic patients with suspected complications and is considered the gold standard imaging.[12,14,15,16] Contrast is helpful to evaluate for anastomotic leaks.[16]
- CTA is a useful imaging option for post-operative complications where blood supply is potentially compromised, such as suspected splenic infarct.[13]
- Internal hernias in individuals who have had Roux-en-Y gastric bypasses may have intermittent and relatively mild abdominal symptoms which require immediate evaluation with CT imaging.[17]
- Though abdominal pain in post-operative bariatric individuals may be gallbladder induced and an ultrasound would be helpful for this diagnosis, the complications of bariatric surgery can become quickly life-threatening, so any request for CT imaging in the post-operative bariatric individual should not be delayed with recommendations for ultrasound, even if the examination does not indicate any signs or symptoms of more serious or complicated disease.[4,10,11]

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Percutaneous Gastrostomy (AB-9.2)

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- Percutaneous Endoscopic Gastrostomy (PEG)
  - CT Abdomen with or without contrast (CPT® 74160 or 74150) is medically necessary:
    - For pre-operative assessment in the presence of:
      - abdominal wall defects such as an open abdomen OR
      - the presence of “ostomy” sites or drain tubes OR
      - abdominal surgical scars or prior major abdominal surgery (e.g. laparotomy, laparoscopy) OR
      - known situs inversus OR
      - known paraesophageal hernia OR
      - previous endoscopic attempt did not achieve adequate transillumination through the abdominal wall or compression and a suitable site for PEG placement could not be determined
  - Percutaneous Gastrostomy via Interventional Radiologist using CT guidance
    - A pre-operative CT Abdomen with or without contrast (CPT® 74160 or 74150) is medically necessary for complicated cases in which a safe window cannot be determined via fluoroscopy.
    - See above indications for CT prior to endoscopic or surgical gastrostomy tube placement for pre-operative indications.
  - Suspected complication of an endoscopically or IR-placed gastrostomy or jejunostomy tube:
    - CT Abdomen with or without contrast (CPT® 74160 or 74150) or CT Abdomen and Pelvis with or without contrast (CPT® 74177 or 74176)

Background and Supporting Information

- A percutaneous endoscopic gastrostomy utilizes endoscopic guidance in order to place the feeding tube.
- The optimal site for gastrostomy placement is determined by illuminating the abdominal wall from the stomach using the scope and simultaneously indenting the wall with the finger and visualizing that indention endoscopically.
  - Routine CT prior to this is generally not medically necessary.
  - A study retrospectively compared complication rates between individuals who underwent a pre-procedure CT vs. those that did not and found no difference in the rate of bleeding events, need for operative intervention, or accidental tube dislodgement.

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- One individual in the non-CT group had an injury due to the tube being placed through the colon, but in that case, there was failure of transillumination through the abdominal wall.
- The authors concluded, “routine CT to evaluate for unfavorable anatomy such as overlying liver or transverse colon prior to PEG tube placement does not result in a reduced complication rate. Safe site selection utilizing the correct technique of transillumination of the abdominal wall and visualization of the indentation of the operator’s finger is essential for safe PEG tube placement.”

Evidence Discussion

The use of routine pre-procedure CT scans does not result in lower complication rates for endoscopic percutaneous gastrostomy. A retrospective study comparing complication rates between individuals who underwent pre-procedure CT scans and those who did not found no difference in the rate of bleeding events, need for operative intervention, or accidental tube dislodgement. Thus, pre-procedure CT of the abdomen is reserved for complex placement scenarios.[1-9]

Post-procedure, the role of CT imaging is to assist in identifying complications, allowing fast visualization of issues such as a migrated internal bumper or injury to internal viscera.[1-9]

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Blunt Abdominal Trauma (AB-10)

Guideline

Blunt Abdominal Trauma (AB-10.1)
References (AB-10)

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Blunt Abdominal Trauma (AB-10.1)

AB.BA.0010.1.A

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- Abdominal and/or Pelvic ultrasound (CPT® 76700 and/or CPT® 76856) is medically necessary for the evaluation of blunt abdominal trauma when requested.
- CT Abdomen and/or Pelvis with contrast (CPT® 74160, or CPT® 72193, or CPT® 74177) is medically necessary for:
  - High probability of intra-abdominal injury after blunt abdominal trauma, with new onset of ANY of the following conditions:
    - Abdominal pain or tenderness
    - Pelvic or femur fracture
    - Lower rib fracture
    - Costal margin tenderness or evidence of thoracic wall trauma
    - Diminished breath sounds
    - Vomiting
    - Pneumothorax
    - Hematocrit <30%
    - Hematuria
    - Elevated AST
    - Non-examinable individual (intoxicated, less than fully conscious, Glasgow Coma Scale Score <13, etc.)
  - Evidence of abdominal wall trauma or presence of a seat-belt sign
  - If an ultrasound demonstrates definitive abnormalities or yields inconclusive findings

Evidence Discussion

Intra-abdominal injury is an indication for ultrasound (US) and/or advanced imaging. Advanced imaging in acute trauma is generally with CT of the Abdomen and/or Pelvis with contrast. Both US and CT can be completed rapidly. CT with contrast can provide more detailed images of blood vessels and tissues, helping to better identify areas of bleeding, inflammation, or injury.[1] A seat belt sign is a linear area of bruising, abrasion or erythema across the abdomen, chest, or neck in the distribution of a seat belt. The presence of a seat belt sign is strongly associated with intra-abdominal injury.[8]

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Gaucher Disease and Hemochromatosis (AB-11)

Guideline

Gaucher Disease (AB-11.1)
Hereditary (Primary) Hemochromatosis (HH) and Other Iron Storage Diseases (AB-11.2)
References (AB-11)

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Gaucher Disease (AB-11.1)

AB.GD.0011.1.A

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- For evaluation of neurologic involvement in Gaucher Disease, see: **Gaucher Disease (Storage Disorders) (PN-8.6)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.

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Hereditary (Primary) Hemochromatosis (HH) and Other Iron Storage Diseases (AB-11.2)

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- MRI Abdomen without contrast (CPT® 74181) for iron quantification
  - If transferrin iron saturation (TS) ≥45% OR elevated serum ferritin (males [assigned at birth] >300 ng/ml, females [assigned at birth] >200 ng/ml) **AND**
  - Genetic studies for hemochromatosis have been performed and results are ANY of the following:
    - Negative for hemochromatosis
    - C282Y/H63D compound heterozygote
    - C282Y heterozygote
    - Non-C282Y homozygote

- **Note:**

  For C282Y/C282Y homozygote, iron quantification generally not indicated. Workup is as follows:

  - If serum ferritin >1000 ug/L or elevated liver enzymes:
    - Liver biopsy for fibrosis staging and rule out concurrent liver disease
  - If serum ferritin <1000 ug/L and normal liver enzymes:
    - Therapeutic phlebotomy

    Note:

    Studies indicate that measurements of hepatic iron concentration by MRI may be more useful in ruling out than diagnosing clinically significant iron overload. MRI can distinguish between primary and secondary iron overload based on iron uptake in the reticuloendothelial system.)

- For the evaluation of suspected hepatic iron overload in chronic transfusional states (e.g., sickle cell disease, thalassemia, oncology patients, bone marrow failure, and stem cell transplant individuals):
  - MRI Abdomen without contrast (CPT® 74181) for iron quantification can be performed annually.

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- See: **Transfusion-Associated (Secondary) Hemochromatosis (PEDAB-18.2)** in the Pediatric Abdomen Imaging Guidelines regarding transfusion-associated hepatic iron deposition.
- If clinical, biopsy, or radiological findings suggest advanced fibrosis or cirrhosis and HCC surveillance is requested, then follow HCC Screening Guidelines – See: **Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)**.
- Role of MR Elastography (CPT® 76391):
  - The role of MR Elastography to assess the degree of fibrosis in the setting of hemochromatosis is not yet clearly defined and thus not currently considered medically necessary.

Background and Supporting Information

- An elevated serum ferritin >1000 mcg/l is associated with an increased risk of cirrhosis and mortality in C282 homozygotes, while a serum ferritin <1000 mcg/l is associated with a very low likelihood of cirrhosis.
- The role of serial MRI for monitoring hepatic iron concentration in hemochromatosis has not been defined. Treatment is phlebotomy and results are monitored by serum ferritin.
- One of the main limitations of MR Elastography is that artifact from excess iron deposition degrades signal intensity in MRE sequences, leading to technical failure of elastography and a decrease in MRE's diagnostic reliability. The latest ACG Clinical Guideline (2019) indicated that MRI for the purpose of estimating hepatic iron concentration is appropriate in the circumstances described above. However, "if there is a concomitant need to stage hepatic fibrosis, then liver biopsy is the preferred method." The ACG diagnostic algorithm for the workup of hemochromatosis does not include MR Elastography at any stage, including the evaluation for the presence, absence, or degree of fibrosis.

Evidence Discussion

The ACG Clinical Guideline indicated that MRI without contrast is the preferred modality for assessing hepatic iron concentration in iron overload conditions, including primary hereditary hemochromatosis (HH) as well as in secondary, multi-transfusion conditions, such as sickle cell disease, thalessemia, and in oncology individuals and those with bone marrow failure, in whom it can be done annually. MRI offers several key advantages. MRI can distinguish between primary and secondary iron overload based on uptake in the reticuloendothial system, is non-invasive, radiation-free, and has the ability to be performed on both liver and heart. In addition, it is useful for screening, as noted, in the appropriate populations.[9]

CT has been used but presents the negatives of radiation exposure. Dual-energy scans are required to compensate for background attenuation, so its use is reserved for individuals without access to MRI.[15]

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Ultrasound-based elastography can assess the need for biopsy. However, Magnetic Resonance Elastography (MRE) is not preferred due to MRI signal degradation by excess iron and is not recommended by the ACG at any stage of the work-up.[9]

For individuals with iron indices indicative of classic HH, iron mobilized by well-controlled phlebotomy can provide an alternative estimate of total body iron comparable to liver iron quantification. Serial MRI monitoring of hepatic iron concentration has not been defined; instead, serum ferritin levels are monitored during phlebotomy.[16]

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Hernias (AB-12)

Guideline

Inguinal or Femoral Hernia, or Indeterminate Groin Pain (AB-12.1)
Spigelian, Ventral, Umbilical, or Incisional Hernia (AB-12.2)
Hiatal Hernia (AB-12.3)
References (AB-12)

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Inguinal or Femoral Hernia, or Indeterminate Groin Pain (AB-12.1)

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- Clinical examination alone is usually sufficient for confirming the diagnosis of an evident groin hernia.
- If musculoskeletal ailments such as osteitis pubis or athletic pubalgia are in the differential, see: **Joint Instability and Dysfunction (MS-30.1)** or **Muscle and Tendon Injury (MS-11.1)** in the Musculoskeletal Imaging Guidelines.
- Ultrasound, pelvic limited (CPT® 76857) or pelvic complete (CPT® 76856) is the initial imaging study if:
  - vague groin swelling with diagnostic uncertainty OR
  - poor localization of swelling (as might be seen with a small hernia and prominent overlying fat) OR
  - intermittent swelling not present on examination OR
  - other/indeterminate groin complaints without swelling
- If ultrasound is indeterminate or non-diagnostic, ONE of the following is medically necessary:
  - CT Pelvis with contrast (CPT® 72193) or without contrast (CPT® 72192)
  - MRI Pelvis without contrast (CPT® 72195) or with and without contrast (CPT® 72197)
- For suspected incarceration or strangulation (**initial ultrasound is not required**):
  - CT Pelvis with contrast (CPT® 72193) or without contrast (CPT® 72192)
- For chronic post-surgical groin pain (after hernia repair):
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) or US-guided nerve block
  - CT Pelvis with contrast (CPT® 72193) or without contrast (CPT® 72192) or MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) can be approved if either ultrasound or ultrasound-guided nerve block is indeterminate or non-diagnostic, to assess for other, non-neuropathic causes.

Evidence Discussion

- Diagnosis of inguinal and femoral hernias is usually possible by history and physical alone. When the diagnosis is in question because physical exam is inconclusive or symptoms are vague, ultrasound should be the initial imaging study. Ultrasound can provide useful information without the risk of radiation. It is readily available, easily performed and can be used in conjunction with provocative maneuvers such as valsalva to help delineate a hernia. These provocative maneuvers are more difficult to perform during CT scanning, which gives a more static image.[2]

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- In the event of an inconclusive ultrasound or if there is a concern for a complicated hernia, imaging of the pelvis with either CT or MRI is appropriate. Abdominal imaging is not medically necessary for evaluation of an inguinal or femoral hernia.[2]
- Post-surgical pain can be associated with neuropathy, recurrence, or mesh complications. These problems should be evaluated with US and/or nerve block as well prior to proceeding to advanced imaging if these studies are indeterminate.[8]

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Spigelian, Ventral, Umbilical, or Incisional Hernia (AB-12.2)

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- For known or suspected primary or recurrent Spigelian hernia (anterior abdominal wall hernia through the semilunar line), ventral hernia, umbilical, or incisional hernia:
  - CT Abdomen without or with contrast (if at or above the umbilicus) (CPT® 74150 or CPT® 74160) OR
  - CT Pelvis without or with contrast (if below the umbilicus) (CPT® 72192 or CPT® 72193) OR
  - CT Abdomen and Pelvis without or with contrast (if above and below the umbilicus, or indeterminate) (CPT® 74176 or CPT® 74177)

Evidence Discussion

- Hernias of the abdominal wall can have a variable presentation and a challenging physical exam. In addition, there may be secondary hernias that are not noted on physical exam or the hernia may track through different layers of the abdominal wall. The size of the hernia defect is also an important consideration in determining operative approach. Ultrasound is limited in being able to evaluate size and extent of hernia through various tissue planes. Advanced imaging may be medically necessary for both diagnosis and in planning treatment. Limits to imaging only involve targeting imaging to the appropriate body region.[7]

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Hiatal Hernia (AB-12.3)

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- CT Chest with contrast (CPT® 71260) and/or CT Abdomen with contrast (CPT® 74160) is medically necessary to evaluate ANY of the following:
  - For pre-operative treatment planning
  - Suspected complication of primary disease or surgery

Background and Supporting Information

- Complications of hiatal hernia may include a gastric volvulus (torsion) within the chest cavity, vomiting, chest pain, or difficulty in swallowing.

Evidence Discussion

- Hiatal hernias can become symptomatic. If so, evaluation should follow the guidelines for the specific symptom complex (such as reflux, cough, abdominal or chest pain, vomiting, dysphagia, abnormal chest x-ray, etc.).[9]
- To avoid unnecessary testing and radiation exposure, advanced imaging for hiatal hernias should be reserved for specialist requests for pre-operative evaluation or for complications of the primary disease or surgery.[9]

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Abdominal Mass (AB-13)

Guideline

Abdominal Wall Mass (AB-13.1)
Indeterminate Intra-Abdominal Mass (AB-13.2)
Abnormal Findings on Endoscopy/Colonoscopy (AB-13.3)
References (AB-13)

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Abdominal Wall Mass (AB-13.1)

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- Abdominal ultrasound and/or Pelvic ultrasound (CPT® 76700 or CPT® 76705 and/or CPT® 76856) is medically necessary as the initial imaging study to assess an abdominal wall or subcutaneous mass.
- MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160) is medically necessary to further characterize a suspected malignant or indeterminate mass detected on ultrasound.
  - Pelvic imaging may be medically necessary depending on the location of the mass.

Evidence Discussion

- Mass lesions of the subcutaneous tissue and abdominal wall are generally benign and can be diagnosed through physical examination (such as lipomas, fibromas, epidermal inclusion cysts, etc.). For lesions that require imaging for further delineation, ultrasound is the initial study of choice. Ultrasound allows for real-time imaging, and the addition of Doppler techniques can help identify vascular lesions. It is highly specific for benign lesions.[1] If the ultrasound image is inconclusive, it can guide the choice of additional imaging modalities, body areas, and contrast levels.
- Subsequent or second-line imaging for indeterminate ultrasound findings includes CT with contrast or MRI with and without contrast. ACR Appropriateness Criteria® stated, "CT is widely considered fast and accurate for excluding or confirming a mass" and "may be additionally helpful in the setting of suspected hernia, congenital abnormalities, hematomas, and infections."[1] It is not necessary to perform CT without and with contrast as little diagnostic value is gained by the additional imaging without contrast.[5] MRI is particularly useful for evaluating masses that appear sarcomatous prior to biopsy. ACR stated, "Although helpful in defining relationship of masses to adjacent fascia, muscles, and vessels, MRI is often not specific enough to establish a definitive diagnosis."[1] The appropriate body region for imaging depends on the location of the mass.

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Indeterminate Intra-Abdominal Mass (AB-13.2)

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- For a palpable abdominal mass on physical examination:
  - Abdominal ultrasound (CPT® 76700) and/or Pelvic ultrasound (CPT® 76856) is medically necessary in lieu of CT, if requested
  - CT Abdomen with contrast (CPT® 74160) if above the umbilicus
  - CT Abdomen and Pelvis with contrast (CPT® 74177) if extending below the umbilicus
  - CT Pelvis with contrast (CPT® 72193) if involving the pelvis
- Indeterminate findings on a prior CT or ultrasound:
  - MRI Abdomen without and with contrast (CPT® 74183)
    - MRI Pelvis without and with contrast (CPT® 72197) is medically necessary to evaluate if the mass extends below the umbilicus or involves the pelvis
- For a pulsatile abdominal mass, suspected aortic aneurysm: See: **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)** in the Peripheral Vascular Disease (PVD) Imaging Guidelines.
- For females (assigned at birth) with a suspected adnexal mass or fibroid: See: **Adnexal Mass/Ovarian Cysts (PV-5)** or **Leiomyomata/Uterine Fibroids and Other Uterine Masses(PV-12)** in the Pelvis Imaging Guidelines.
- Pregnant individual:
  - Abdominal and/or Pelvic and/or Transvaginal ultrasound (CPT® 76700 and/or CPT® 76856 and/or CPT® 76830) is appropriate for initial imaging.

Evidence Discussion

- The origins and characteristics of a palpable intra-abdominal mass are difficult to determine on physical exam. For intra-abdominal masses, contrast-enhanced CT and ultrasound examination have demonstrated accuracy. Although ultrasound may be limited by body habitus or bowel gas, it offers several advantages.[1] Ultrasound requires no ionizing radiation, is cost effective, helps determine most appropriate next advanced imaging study (CT vs. MRI), is readily accessible, and often can be scheduled same day.
- ACR Appropriateness Criteria® stated, "CT demonstrated high positive predictive value (99%) and negative predictive value (97%) for determining the presence or absence of a mass and correctly identified the organ of origin in 93% of individuals with palpable abnormalities on clinical examination." (2019) MRI is useful for

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further delineation of an indeterminate mass found on US or CT due to its excellent sensitivity for soft-tissue differentiation.[1]

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Abnormal Findings on Endoscopy/ Colonoscopy (AB-13.3)

AB.AM.0013.3.A
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The following criteria applies to findings on endoscopy or colonoscopy that have NOT been diagnosed or biopsy-proven as malignant. For biopsy-proven malignancy, refer to the appropriate Oncology Imaging Guideline.

- For suspicious (not clearly benign) submucosal lesion(s) noted in the stomach or small bowel or gastric extrinsic compression on colonoscopy, ONE of the following is medically necessary:
  - CT Abdomen with contrast (CPT® 74160) OR
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
- For suspicious (not clearly benign) lesion(s) noted above the rectum (in the colon or small bowel), unexplained colonic extrinsic compression on colonoscopy, or pre-operative planning for complete surgical removal of a polypoid lesion the following are medically necessary,
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
  - MRI Pelvis without and with contrast (CPT® 72197) or MRI Pelvis without contrast (CPT® 72195) is medically necessary in addition to CT when there is an additional suspicious lesion(s) in the distal sigmoid or rectum
- For submucosal rectal lesions or unexplained extrinsic compression in the rectum:
  - MRI Pelvis without and with contrast (CPT® 72197), or if requested, MRI Pelvis without contrast (CPT® 72195)
- For criteria specific to biopsy-proven colorectal cancer, See **Colorectal Cancer – Initial Work-up/Staging (ONC-16.2)** in the Oncology Imaging Guidelines.
- For criteria specific to biopsy-proven Gastrointestinal Stromal Tumor (GIST): See **Gastrointestinal Stromal Tumor (GIST) (ONC-12.5)** in the Oncology Imaging Guidelines.
- For criteria specific to biopsy-proven gastric cancer: See **Gastric Cancer - Initial Work-up/Staging (ONC-14.9)** in the Oncology Imaging Guidelines.

Evidence Discussion

- Endoscopic ultrasound with or without fine-needle aspiration is the preferred initial imaging modality to further characterize a gastric submucosal lesion detected on endoscopy. Rectal endoscopic ultrasound is the preferred initial imaging study to further characterize submucosal rectal lesions.[2,3]

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- Radiographic modalities may be warranted when endoscopic and/or rectal ultrasound cannot be performed (e.g. stricture or severe inflammatory process prohibit passage of probe, etc.), is indeterminate, findings indicate need for further characterization, there is external impression(s) against the gut wall, and/or therapeutic need to understand extent of visualized disease and/or of the origin of an endoscopically-apparent malignancy. Choice of the optimal imaging modality requires consideration of factors such as age, gender, fertility, co-morbidities, medications, and allergies.
- Ultrasound can provide high resolution imaging of the liver, gallbladder, bile ducts, pancreas, spleen, kidneys, and abdominal vasculature. It can also provide information regarding phase and direction of blood flow in arteries and veins via Duplex scanning. Ultrasound requires no ionizing radiation, is readily available being mobile, cost effective, and easier to schedule for same day testing. However, image quality may be limited due to bowel gas (a particular disadvantage in assessment of endoscopically-identified gut lesions), poor acoustic window acquisition, obesity, and sonographer experience level.
- Computed tomography (CT) of the abdomen offers excellent 3-dimensional resolution of the gut and its surrounding structures, especially when performed with use of oral and/or intravenous (IV) contrast agents. CT scan requires a significant dose of ionizing radiation but is ideally suited to characterizing lesions within the gut because the quick speed of image acquisition reduces the potential for motion artifact. The NCCN-recommended imaging of a suspected metastatic adenocarcinoma includes CT imaging of the chest, abdomen, and pelvis.[4]
- Magnetic resonance imaging (MRI) uses a magnetic field to capture excellent 3-dimensional soft tissue resolution. As with CT scans, the technique is often performed with IV contrast agents, and with specialized techniques, can be directed either at whole or parts of the abdomen or at specific abdominal structures (e.g., MR elastography of liver, MR enterography of small bowel, MR cholangiopancreatography [MRCP] of the biliary and pancreatic system). MRI yields better soft contrast resolution than CT and does not expose individuals to ionizing radiation, but due to longer image time is motion artifact-prone and thus less suited to resolving gastrointestinal detail. MRI has disadvantages in that it may require sedation in those with claustrophobia and in young individuals who may be unable to hold still and follow directions. MRI also cannot be performed in those with ferrous magnetic implants or non-removable foreign bodies.[8]

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Lower Extremity Edema (AB-14)

Guideline

Lower Extremity Edema (AB-14)

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Lower Extremity Edema (AB-14)

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In the setting of IVC thrombus secondary to the filter or concern for proximal DVT (iliofemoral), see: **Acute Limb Swelling (PVD-12)** and for evaluation of chronic limb swelling/May-Thurner Syndrome, see: **Chronic Limb Swelling Due to Venous Insufficiency/Venous Stasis Changes/Varicose Veins (PVD-13)** in the Peripheral Vascular Disease Imaging Guidelines.

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Zollinger-Ellison Syndrome (ZES-Gastrinoma) (AB-15)

Guideline

Zollinger-Ellison Syndrome (ZES-Gastrinoma) (AB-15.1)

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Zollinger-Ellison Syndrome (ZES-Gastrinoma) (AB-15.1)

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- For suspected gastrinoma, see: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines.

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Adrenal Cortical Lesions (AB-16)

Guideline

Adrenal Cortical Lesions (AB-16)
Asymptomatic Adrenal Cortical Lesions (AB-16.1)
References (AB-16.1)
Adrenal Hormone Excess/Symptomatic Adrenal Lesions (AB-16.2)
References (AB-16.2)
Adrenal Insufficiency (AB-16.3)
References (AB-16.3)
Adrenal Nuclear Imaging (AB-16.4)
References (AB-16.4)

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Adrenal Cortical Lesions (AB-16)

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Procedure Code	Description
CPT® 74150	CT Abdomen without contrast
CPT® 74160	CT Abdomen with contrast
CPT® 74170	CT Abdomen without and with contrast
CPT® 74181	MRI Abdomen without contrast
CPT® 74183	MRI Abdomen without and with contrast
CPT® 78812	PET, Skull Base to Mid-Thigh
CPT® 78815	PET/CT, Skull Base to Mid-Thigh

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Asymptomatic Adrenal Cortical Lesions (AB-16.1)

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Overall Considerations

- The following recommendations are for asymptomatic individuals:
  - For symptomatic individuals, see: **Symptomatic Adrenal Cortical Lesions (AB-16.2)**.
  - US is not a prerequisite study for advanced imaging in the evaluation of any adrenal abnormality.
- Abdominal pain may be present in large or rapidly expanding adrenal tumors due to mass effect or hemorrhage.
  - If the source of abdominal pain is suspected to be an incidental adrenal mass and initial imaging was indeterminate, immediate reimaging with a dedicated adrenal protocol study (see 3 imaging modalities below) is reasonable irrespective of the size of the mass.
  - See: **Abdominal Pain (AB-2)** in the Abdomen Imaging Guidelines for imaging recommendations if abdominal pain is unrelated to the adrenal mass.
- The three imaging modalities that can be used for definitive benign characterization of an adrenal mass are:
  - CT Abdomen without contrast (CPT® 74150)
  - CT Abdomen without and with contrast (CPT® 74170)
  - CS-MRI (chemical shift MRI, CPT® 74181)
- The following list represents definitively benign characteristics of the adrenal gland. This list applies wherever "benign characteristics" are mentioned in the table below:
  - ≤10 HFU on CT
  - ≥60% absolute washout or ≥40% relative washout on CT abdomen without and with contrast with calculated washout (adrenal protocol CT, CPT® 74170)
    - An important exception to the washout rule: Non-adenomatous adrenal masses that may show elevated washout on adrenal protocol CT but are not benign include:
      - adrenal metastasis from hypervascular tumors (e.g. RCC and HCC)
      - pheochromocytoma
      - adrenocortical carcinoma
      - clinical suspicion should be used in these cases to guide further investigation

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  - Decreased signal on Chemical Shift MRI (CS-MRI, CPT® 74181)
  - Cyst (if imaging was completed with and without contrast and "no enhancement"- defined as <10HFU change between unenhanced and enhanced/contrasted CT)
  - Adrenal myelolipoma (macroscopic fat)
- If definitively benign diagnosis cannot be made during follow up imaging using dedicated CT adrenal protocol (If <60% absolute washout or <40% relative washout) or lack of signal drop out on MRI chemical shift:
  - Additional imaging is medically necessary at 6-12 months from initial follow up, OR
  - Consider resection for possible primary adrenocortical carcinoma after biochemical evaluation and exclusion of pheochromocytoma.
    - For individuals who are poor surgical candidates, if ordered by or in consultation with an endocrinologist, endocrine surgeon, or urologist:
      - Imaging as requested
- CT Abdomen without and with contrast (CPT® 74170) may be medically necessary in place of any below recommended CT Abdomen without contrast for the following:
  - Facility protocol is to cease imaging if adrenal mass is found to have HFU<10 on initial non-contrasted images
- MRI Abdomen without contrast (CPT® 74181) is medically necessary in place of CT for the following:
  - Clips that cause artifacts when using CT
  - Allergy to CT contrast
  - Individuals in whom radiation exposure should be limited (children, pregnant individuals, individuals with known germline mutations, and individuals with recent excessive radiation exposure)
- CS MRI may not detect the intracellular lipid in an adrenal mass if HFU is 30 HU or more on CT without contrast. CS MRI is less effective than CT without and with contrast with calculated washout for adenomas with unenhanced attenuation of more than 20 HU
- Below imaging can be applied to bilateral adrenal masses, with each lesion addressed separately.

Mass Characteristics and Appropriate Imaging

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Mass Details	Imaging Study
• Asymptomatic AND • Incidentally found on US, CT, or MRI of area OTHER than the abdomen or if seen only on US of the abdomen AND • Any size AND • No history of cancer	• CT Abdomen without contrast (CPT® 74150)
• Asymptomatic AND • Incidentally found on CT Chest without contrast, entirely imaged, and fully characterized as indeterminate by HFU score AND • >2 cm AND • No history of cancer	• CT Abdomen without and with contrast (CPT® 74170) in lieu of above recommended CT Abdomen without contrast
• Asymptomatic AND • Incidentally found on CT or MRI of the Abdomen or Abdomen and Pelvis AND • <1 cm in short axis AND • No history of cancer	• No further imaging is medically necessary ◦ It is uncertain as to whether subcentimeter nodularity or adrenal thickening qualifies as an adrenal mass on radiology reports
• Asymptomatic AND • Incidentally found on CT or MRI of the Abdomen or Abdomen and Pelvis AND • No prior imaging for comparison AND • Diagnostic with benign imaging characteristics AND • ≥1 cm AND • No history of cancer	• No further imaging is medically necessary, regardless of size ◦ The risk of malignancy in a mass with diagnostically benign findings on imaging is extremely low

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Mass Details	Imaging Study
• Asymptomatic AND • 1 cm to 2 cm AND • Incidentally detected and indeterminate on any CT or MRI Abdomen or Abdomen and Pelvis AND • No prior imaging for comparison AND • No history of cancer	• Reimaging is medically necessary at 12 months from the initial indeterminate study, as follows*: ◦ CT Abdomen without and with contrast (CPT® 74170 - adrenal protocol), CT Abdomen without contrast (CPT® 74150), or CS-MRI (chemical shift MRI, CPT® 74181) ▪ No further imaging is medically necessary after initial 12 month study if ANY of the following: - Definitively benign characteristics - Stable in size (change <8mm) over >1 year (likely benign adenoma) *NOTE: These instructions are regarding indeterminate lesions without prior studies to compare, in asymptomatic individuals. If prior imaging exists for comparison and radiology report shows stability over 1 year or if the imaging study already shows definitively benign characteristics, no further imaging is medically necessary

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Mass Details	Imaging Study
• Asymptomatic AND • >2 cm to <4 cm AND • Incidentally detected and indeterminate on any CT or MRI Abdomen or Abdomen and Pelvis AND • No prior imaging for comparison AND • No history of cancer	• Reimaging is medically necessary immediately after initial indeterminate study, as follows*: ◦ CT Abdomen without and with contrast (CPT® 74170 - adrenal protocol), or CS-MRI (chemical shift MRI, CPT® 74181) ▪ Further follow-up imaging is medically necessary at 6 and 12 months ▪ No further imaging is medically necessary if the initial study or follow up study has definitively benign characteristics or if follow up study shows stability in size (change <8mm) over >1 year (as likely benign adenoma) *NOTE: These instructions are regarding indeterminate lesions without prior studies to compare, in asymptomatic individuals . If prior imaging exists for comparison and radiology report shows stability over 1 year or if the imaging study already shows definitively benign characteristics no further imaging is medically necessary.
• Asymptomatic AND • ≥4 cm AND • Incidentally detected and indeterminate on any CT or MRI Abdomen or Abdomen and Pelvis AND • No prior imaging for comparison AND • No history of cancer	• Reimaging is medically necessary immediately after initial indeterminate study, as follows: ◦ CT Abdomen without and with contrast (CPT® 74170) or chemical shift MRI (CPT® 74181) • Consider resection for possible primary adrenocortical carcinoma ◦ See: **Adrenocortical Carcinoma (ONC-15.13)** in the Oncology Imaging Guidelines

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Mass Details	Imaging Study
• History of cancer with a likelihood or propensity to metastasize to the adrenal gland or abdomen • Incidentally detected and indeterminate on any CT or MRI Abdomen or Abdomen and Pelvis	• See: **Adrenal Gland Metastases (ONC-31.4)** in the Oncology Imaging Guidelines
• Known adrenal mass with benign characteristics, but newly symptomatic or new hormonal excess	• Repeat imaging per **Adrenal Hormone Excess/Symptomatic Adrenal Lesions (AB-16.2)**

Background and Supporting Information

Benign Adenoma Imaging Characteristics		
	Findings consistent with Adenoma:	Indeterminate for Adenoma:
CT Abdomen without contrast	≤10 Hounsfield Units	>10 Hounsfield Units
CT Abdomen WWO with calculated washout	≥60% absolute washout or ≥40% relative washout	<60% absolute washout <40% relative washout
Chemical Shift MRI	Signal drop out	Lack of signal drop out

- Endocrine guidelines recommend biochemical evaluation in all incidental adrenal lesions (with the exception of myelolipomas and cysts). However laboratory results are NOT required for imaging in an asymptomatic individual.
- Most benign adenomas, which account for up to 75% of adrenal incidentalomas, are lipid rich and thus easily characterized because they measure 10HFU or less on CT without contrast. CT Abdomen without and with contrast with calculated washout and chemical shift MRI help identify lipid poor adenomas, which are the next most common group. Masses which remain indeterminate include pheochromocytomas (up to 7%) and primary adrenal cancers or metastases to the adrenal glands (approximately 4%).
- Adrenal masses are often found incidentally on CT scans performed WITH contrast to evaluate abdominal symptoms. While CT scans performed with contrast only may report the HFU of an adrenal mass, most benign adenomas are labeled "indeterminate" originally because non-contrasted HFU and HFU after washout cannot be measured or calculated.

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- An "Adrenal Protocol CT" measures pre-contrast HFU of an adrenal mass as well as the HFU during "wash out" of contrast medium after 60 to 90 seconds [early] and 10 to 15 minutes [delayed]. Benign adenomas show more rapid and efficient contrast washout as compared to malignant adrenal masses.
- When an adrenal mass shows avid enhancement on CT scan (>110 – 120 HU), a pheochromocytoma should be considered.
- In addition to the imaging features in the grid which are considered "diagnostic" of a benign adrenal mass, other radiographic characteristics "suggestive" of a benignity include: smooth/round shape, homogeneous content, lack of calcification/ hemorrhage/necrosis, growth rate <1cm/year, lack of FDG avidity on PET, and <4cm.
- Radiographic characteristics "suggestive" of malignancy include: irregular margins/ shape, heterogeneous content, presence of calcification/hemorrhage/necrosis, growth rate >1cm/year, presence of FDG avidity on PET, and >4-6cm.
- Malignancies most likely to metastasize to the adrenal glands include lung cancer, gastrointestinal cancer, melanoma, and renal-cell carcinoma.

Evidence Discussion

- CT scan of the abdomen is the recommended initial study to evaluate adrenal gland nodules. A CT scan may expose patients to radiation; however, it takes less time to perform and is less costly than an MRI. Additionally, CT scans are superior to MRI when evaluating lesions with higher density, particularly when using an adrenal CT protocol for washout measurements.[8]
- 75% of adrenal incidentalomas are benign, nonfunctioning adenomas. They are lipid-rich, with low density, exhibit Hounsfield Units (HU) of 10 or less, and have other benign characteristic appearances that make them easily identifiable on an unenhanced CT of the abdomen. The sensitivity and specificity for adenoma characterization are 71% and 98%, respectively, when using unenhanced CT scan for lesions having a density of 10 or less HU.[8]
- Unenhanced CT scans of lesions with a density greater than 30 HU had a 66.6% chance of remaining indeterminate, even after evaluation with chemical shift MRI.[8]
- A chemical shift MRI (CS-MRI) of the abdomen is also useful for characterizing adrenal gland masses with lower density. It is an alternative for follow-up studies when there is a contraindication to CT or contrast or during pregnancy. However, it should be cautioned that MRI may not detect intracellular lipid when the adrenal mass has a HU > 30. MRI is also less sensitive in evaluation of masses with higher HU over 20 compared to CT scans that calculate contrast wash out times. [8]
- Adrenal protocol CT, with its high sensitivity (98%) and specificity (92%), should be the study of choice to differentiate between adenomas and non-adenomas when an adrenal mass remains indeterminate.[8]

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Adrenal Hormone Excess/Symptomatic Adrenal Lesions (AB-16.2)

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Overall Considerations

- Prior to advanced imaging, adrenal hormone excess must be clinically suspected and then biochemically confirmed via testing listed in the table below.
- The following imaging recommendations can also be followed in asymptomatic individuals with an adrenal incidentaloma who are found to have abnormalities at initial hormonal evaluation.
- For severe hormone elevation or rapidly progressing symptoms for which adrenocortical carcinoma is suspected, see: **Adrenocortical Carcinoma (ONC-15.13)** in the Oncology Imaging Guidelines.

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Condition	Signs/Symptoms (not required to be documented for imaging)	Laboratory requirements PRIOR to initial adrenal imaging	Indicated Imaging
• Suspected cortisol excess (adrenal Cushing's Syndrome)	• Weight gain • Hyperglycemia/ diabetes • Low bone mineral density/fractures • Hyperpigmented Striae • Lipodystrophy ("buffalo hump")	• ACTH low/ suppressed AND • Cortisol elevation documented by any of the following: ◦ Elevated AM cortisol following overnight 1mg dexamethasone suppression (cortisol >1.8 mcg/dL) ◦ Elevated late night salivary cortisol ◦ Elevated urine free cortisol	• CT Abdomen without contrast (CPT® 74150) ◦ If CT Abdomen without contrast shows an indeterminate adrenal mass, the following is medically necessary immediately: ▪ CT Abdomen without and with contrast adrenal protocol (CPT® 74170) OR ▪ MRI Abdomen without contrast chemical shift (CPT® 74181)

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Condition	Signs/Symptoms (not required to be documented for imaging)	Laboratory requirements PRIOR to initial adrenal imaging	Indicated Imaging
• Suspected adrenal hyper-androgenism/ virilizing adrenal tumor	• Hirsutism • Virilization (voice deepening, clitoromegaly)	• Elevated serum DHEAS AND/OR • Elevated testosterone	• CT Abdomen without contrast (CPT® 74150) ◦ If CT Abdomen without contrast shows an indeterminate mass, the following is medically necessary immediately: ▪ CT Abdomen without and with contrast adrenal protocol (CPT® 74170) OR ▪ MRI Abdomen without contrast chemical shift (CPT® 74181) • In individuals with an elevated testosterone level and an ovarian etiology is suspected, see: **Polycystic Ovary Syndrome (PV-8.1)** in the Pelvis Imaging Guidelines and **Ovarian Cancer-Suspected/ Diagnosis (ONC-21.2)** in the

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Condition	Signs/Symptoms (not required to be documented for imaging)	Laboratory requirements PRIOR to initial adrenal imaging	Indicated Imaging
			Oncology Imaging Guidelines.
• Suspected feminizing adrenal tumor	• Gynecomastia • Testicular atrophy	• Elevated serum estradiol AND • Non-elevated serum LH AND • No testicular mass seen on dedicated imaging	• CT Abdomen without contrast (CPT® 74150) ◦ If CT Abdomen without contrast shows an indeterminate adrenal mass, the following is medically necessary immediately: ▪ CT Abdomen without and with contrast adrenal protocol (CPT® 74170) OR ▪ MRI Abdomen without contrast chemical shift (CPT® 74181)

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Condition	Signs/Symptoms (not required to be documented for imaging)	Laboratory requirements PRIOR to initial adrenal imaging	Indicated Imaging
• Suspected primary aldosteronism (Conn's Syndrome)	• HTN • Hypokalemia	• Serum aldosterone >15-20ng/dL in the setting of suppressed renin* and spontaneous hypokalemia (K<3.5mEq/L) OR • Confirmatory testing** showing lack of aldosterone suppression. • (See **Background and Supporting Information** on renin* levels and confirmatory testing**)	• CT Abdomen without contrast (CPT® 74150) ◦ If CT Abdomen without contrast shows an indeterminate adrenal mass, the following is medically necessary immediately: ▪ CT Abdomen without and with contrast adrenal protocol (CPT® 74170) OR ▪ MRI Abdomen without contrast chemical shift (CPT® 74181)

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Condition	Signs/Symptoms (not required to be documented for imaging)	Laboratory requirements PRIOR to initial adrenal imaging	Indicated Imaging
• Suspected pheo-chromocytoma/ paraganglioma	• HTN • Palpitations • Tremor • Pallor • Flushing • Hyperadrenergic spells	• Elevated plasma free metanephrines OR • Elevated urinary fractionated metanephrines	• CT Abdomen and Pelvis without and with contrast (CPT® 74178), CT Abdomen and Pelvis with contrast (CPT® 74177), or MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast • See also: **Adrenal Nuclear Imaging (AB-16.4)** and **Adrenal Tumors (ONC-15.10)** in the Oncology Imaging Guidelines and **Hereditary Paraganglioma-Pheochromocytoma Syndromes (PEDONC-2.13)** in the Pediatric and Special Populations Oncology Imaging Guidelines
• Suspected adrenocortical carcinoma	• Rapidly progressive symptoms • Elevation of multiple adrenal hormones	• NA	• See: **Adrenocortical Carcinoma (ONC-15.13)** in the Oncology Imaging Guidelines

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Condition	Signs/Symptoms (not required to be documented for imaging)	Laboratory requirements PRIOR to initial adrenal imaging	Indicated Imaging
• Confirmed adrenal hormone excess AND • Requested for surgical planning AND • Requested by or in consultation with an endocrinolo-gist, endocrine surgeon, or urologist	NA	NA	• Repeat imaging as requested

Background and Supporting Information

- Surgery is the management of choice for individuals with virilizing adrenal tumors, feminizing adrenal tumors, pheochromocytoma/PGL, and suspected adrenocortical carcinoma due to an increased risk of malignancy and/or comorbidity. Adrenal masses that secrete excess cortisol (adrenal Cushing's syndrome) or aldosterone (primary hyperaldosteronism/Conn's syndrome) are rarely malignant; however, surgery is also definitive management.

Suspected cortisol excess (adrenal Cushing's syndrome)

- Low or suppressed ACTH levels (<10 pg/mL) are consistent with an adrenal source.
- DHEAS levels are also low in adrenal Cushing's syndrome.
- The diagnosis of Cushing's syndrome can be delayed for years due to the insidious nature of clinical presentation and the complexity of diagnostic testing.

Suspected adrenal hyperandrogenism/virilizing adrenal tumor

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- Testosterone is produced by both the ovary (primary source) and adrenal gland while DHEA and DHEAS are produced almost exclusively by the adrenal gland.
- The magnitude of the androgen level is of poor predictive value for tumors, although a very high testosterone (adult-male range) or DHEAS level (>700 µg/dL) is suggestive.

Suspected feminizing adrenal tumor

- Adrenal tumors, mainly carcinomas (extremely rare, 0.5–2.0 per million), can secrete both estrogens and high amounts of adrenal androgens, which aromatize to estrogens. In this case, gynecomastia is usually of recent onset, progresses rapidly and testicular atrophy can also be seen.
- Common causes of excessive endogenous estrogens should be excluded prior to adrenal imaging. These include increased secretion from testis (Leydig cell or Sertoli cell tumors, stimulation of normal Leydig cells by LH or hCG) and increased aromatization of androgens to estrogens (aging, obesity, alcoholic cirrhosis, hyperthyroidism, drugs, hCG-secreting tumors, aromatase excess syndrome).

Suspected primary aldosteronism (Conn's syndrome)

- A positive screen for primary aldosteronism is an aldosterone level >15-20ng/dL in the setting of suppressed renin* (plasma renin activity <0.6-1.0ng/mL/hour or plasma renin concentration <5-8.2 mU/L) and spontaneous hypokalemia (K<3.5mEq/L).
- The most common dynamic confirmatory tests include the oral sodium suppression test, the seated intravenous saline suppression test, the fludrocortisone suppression test, and the captopril challenge test, and results that indicate a "positive" result are unique to the each test. For example, if oral sodium loading is used, a 24-hour urine aldosterone excretion of more than 12 mcg in the setting of 24-hour urine sodium excretion of more than 200 mEq is diagnostic of primary aldosteronism (and values of more than 10 mcg/24 hours are strongly suggestive).
- Primary hyperaldosteronism may be managed medically with mineralocorticoid receptor antagonists (spironolactone and eplerenone) in cases of bilateral adrenal disease or poor surgical candidacy. If there has been no recent adrenal imaging, reimaging can be considered in cases of diagnostic uncertainty or poor response to medical therapy.

Suspected pheochromocytoma/paraganglioma

- A pheochromocytoma (85% of chromaffin tumors) arises from the chromaffin cells in the adrenal medulla and commonly produces one or more of the following catecholamines: epinephrine, norepinephrine and dopamine.
- A paraganglioma (15-20% of chromaffin tumors) arises from the extra-adrenal chromaffin cells of the sympathetic paravertebral ganglia of the thorax, abdomen, and pelvis (catecholamine producing) or the parasympathetic ganglia along the glossopharyngeal and vagal nerves in the neck and base of skull (not catecholamine producing).

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- Cases of pheochromocytoma/paraganglioma can be sporadic but one-third are hereditary and due to germ-line mutations that may increase malignant potential

Suspected adrenocortical carcinoma

- Adrenocortical carcinoma may be suspected radiographically or clinically. Approximately 60% of individuals present with evidence of adrenal steroid hormone excess, with or without virilization. Hormonally inactive ACCs typically produce symptoms related to tumor burden, including abdominal pain, back pain, early satiety, and weight loss.
- See: **Adrenocortical Carcinoma (ONC-15.13)**

Evidence Discussion

- Advanced imaging is medically necessary when there is biochemical confirmation of adrenal hormone excess.[2,7,10,13]
- CT of the abdomen is the initial imaging study of choice to identify adrenal adenomas when adrenal hormone excess is confirmed. CT scans are readily available and can identify if adrenal lesions are present and can show characteristics of the lesions that help to distinguish benign lesions from indeterminate lesions.[2,7,10,13]
- MRI with chemical shift can further help characterize lesions that are indeterminate on CT scan.[2,7,10,13]
- Including the pelvis in CT scan imaging is medically necessary when evaluating for pheochromocytomas or paragangliomas as these tumors can appear in both the abdominal and pelvis areas and also indicated for staging purposes when adrenal carcinoma is suspected.[2,7,10,13]

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Adrenal Insufficiency (AB-16.3)

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- CT Abdomen (contrast as requested) is medically necessary if the cause of primary adrenal insufficiency is unclear.
  - MRI Abdomen (contrast as requested) is medically necessary if CT is contraindicated.
- Imaging is NOT medically necessary if clinical presentation and labs are consistent with ANY of the following:
  - Primary autoimmune destruction of the adrenal cortex (Addison's disease)
  - Congenital adrenal hyperplasia
  - Adrenoleukodystrophy

Background and Supporting Information

- Imaging can detect infiltrative disease, adrenal hemorrhage, infections, and malignant tumors which may be the cause of adrenal dysfunction.

Evidence Discussion

A CT scan of the abdomen is medically necessary to evaluate the cause of primary adrenal insufficiency when it is unclear. [1-4]

- If screening tests for autoimmune or genetic causes of primary adrenal insufficiency are positive, then imaging is not medically necessary.
- Other causes of primary adrenal insufficiency include adrenal hemorrhage, infiltrative diseases, infections such as tuberculosis, and tumors. All of these can be identified by a CT scan of the abdomen.
- The CT scan is usually readily available, relatively quick to process, and therefore preferred over MRI as the initial study unless contraindicated.
- It can accurately identify the size, location, and appearance of adrenal tumors, as well as the presence of local or vascular invasion, lymph node involvement, and distant metastases in the majority of individuals.
- The CT scan can also accurately identify hemorrhage of the adrenal gland.
- While an abdominal ultrasound is less expensive, it does not provide the precise anatomic definition seen on a CT scan, making the CT scan the preferred study.

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Adrenal Nuclear Imaging (AB-16.4)

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Nuclear medicine imaging can assist in the evaluation of adrenal masses not adequately characterized by CT or MRI.

- Evaluation of SUSPECTED pheochromocytoma or paraganglioma:
  - MIBG (Any ONE of the following codes can be considered medically necessary: CPT® 78801, CPT® 78802, or CPT® 78804).
    - Any ONE of the following codes may also be considered medically necessary, individual or in combination with CPT® 78801, 78802, 78804: SPECT studies (CPT® 78803 or CPT® 78831), or hybrid SPECT/CT studies (CPT® 78830 or CPT® 78832).
  - Octreotide scans can be considered medically necessary in place of MIBG scans (with the same CPT codes) as requested in rare clinical circumstances including head and neck paragangliomas.
- For PET/CT indications and for cases of KNOWN pheochromocytoma or paraganglioma, see: **Adrenal Tumors (ONC-15.10-15.12)** in the Oncology Imaging Guidelines.
- Evaluation of SUSPECTED neuroblastoma, ganglioneuroblastoma, or ganglioneuromas:
  - MIBG (Any ONE of the following codes can be considered medically necessary: CPT® 78801, CPT® 78802, or CPT® 78804).
    - Any ONE of the following codes may also be considered medically necessary, individual or in combination with CPT® 78801, 78802, 78804: SPECT studies (CPT® 78803 or CPT® 78831), or hybrid SPECT/CT studies (CPT® 78830 or CPT® 78832).
- For KNOWN neuroblastoma, ganglioneuroblastoma, or ganglioneuroma, see **Neuroblastoma (PEDONC-6)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Adrenal Nuclear Imaging of the cortex and/or medulla (single site, planar imaging of the adrenal gland only) (CPT® 78075) includes the adrenal scintigraphy scans for 131I-iodocholesterol (NP-59) as well as MIBG (Iodine i-123 iobenguane and Iodine i-131 iobenguane sulfate) scans.
  - 131I-iodocholesterol (NP-59) scans for adrenal cortex imaging can be useful in cases of suspected hyperaldosteronism and adrenal Cushing's, however NP-59 is not readily available for use in the United States.

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  - MIBG (Iodine i-123 iobenguane and Iodine i-131 iobenguane sulfate) scans for adrenal medulla imaging can be helpful in cases of known pheochromocytoma or neuroblastoma.
    - CPT® 78075 is insufficient for the initial evaluation of a suspected pheochromocytoma, paraganglioma or neuroblastoma as this study does not evaluate extra-adrenal sites of disease, but can be considered in rare circumstances.
    - SPECT and SPECT/CT codes as listed above for MIBG can be added to CPT® 78075 as requested.
- History of multiple endocrine neoplasia syndromes: See **Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- History of neurofibromatosis: there is insufficient evidence to support routine imaging of adult individuals with Neurofibromatosis in asymptomatic individuals. See: **Adrenal Hormone Excess/Symptomatic Adrenal Lesions (AB-16.2)** if there is concern for pheochromocytoma. Labs would be required before imaging as stated in guideline.
- History of von Hippel-Lindau disease: See **Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

Evidence Discussion

- Nuclear medicine studies provide functional imaging that helps to further characterize adrenal masses not adequately detailed on CT or MRI.[1-5]
- A meta-analysis found I-123 MIBG sensitivity of 96% in individuals with non-metastatic pheochromocytoma or paraganglioma and 79% in individuals with metastatic pheochromocytoma or paraganglioma.[2]
- Studies have shown excellent lesion-based sensitivity in detecting pheochromocytoma and paraganglioma, often more than 92%, when using ^{68}Ga-DOTATATE (somatostatin analog-SSA) PET/CT.[2]
- A meta-analysis comparing the sensitivity of ^{18}F-FDG and 68Ga-DOTA-SSA found that the sensitivity of ^{68}Ga-DOTA-SSA (95%) was superior to that of ^{18}F-FDG (85%) in detecting pheochromocytoma and paraganglioma.[2]
- Nuclear medicine studies are very useful in head and neck paragangliomas (HNPGL) that prove to be difficult to detect on standard CT or MRI. The sensitivity of ^{68}Ga-DOTATATE was 100% for HNPGL, with identification of additional lesions not visualized with other modalities.[2]
- MIBG or SSA nuclear scans are also very helpful in identifying neuroblastoma, ganglioneuroblastoma, or ganglioneuromas, often associated with Von Hippel-Lindau Syndrome.[1-5]

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Abdominal Aortic Aneurysm (AAA), Iliac Artery Aneurysm (IAA), and Visceral Artery Aneurysms Follow-Up of Known Aneurysms and Pre-Op Evaluation (AB-17)

Guideline

Abdominal Aortic Aneurysm (AAA) (AB-17.1)
Iliac Artery Aneurysm (IAA) (AB-17.2)
Visceral Artery Aneurysm (AB-17.3)

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Abdominal Aortic Aneurysm (AAA) (AB-17.1)

AB.17.1.A

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- For the evaluation of an abdominal aortic aneurysm or pulsatile abdominal mass, see: **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)** in the Peripheral Vascular Disease Imaging Guidelines.

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Iliac Artery Aneurysm (IAA) (AB-17.2)

AB.17.2.A
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- For the evaluation of iliac artery aneurysm (IAA) see: **Iliac Artery Aneurysm (IAA) (PVD-6.4)** in the Peripheral Vascular Disease Imaging Guidelines.

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Visceral Artery Aneurysm (AB-17.3)

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- For the evaluation of known or suspected visceral artery aneurysm, see: **Visceral Artery Aneurysm (PVD-6.5)** in the Peripheral Vascular Disease Imaging Guidelines.

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Abdominal Aortic Aneurysm (AAA) and Iliac Artery Aneurysm (IAA)-Post Endovascular or Open Aortic Repair (AB-18)

Guideline

AAA, IAA, Post Endovascular or Open Aortic Repair (AB-18.1)

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AAA, IAA, Post Endovascular or Open Aortic Repair (AB-18.1)

AB.18.1.A

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- For open aortic abdominal aneurysm repair and additional imaging indications, see: **Post Aortic Endovascular/Open Surgery Surveillance Studies (PVD-6.8)** in the Peripheral Vascular Disease Imaging Guidelines.

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Aortic Dissection and Imaging for Other Aortic Conditions (AB-19)

Guideline

Aortic Dissection and Other Aortic Conditions (AB-19.1)
Imaging for Other Aortic Conditions (AB-19.2)

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Aortic Dissection and Other Aortic Conditions (AB-19.1)

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- For the evaluation of aortic dissection and other aortic conditions, see: **Aortic Imaging** in the Peripheral Vascular Disease Imaging Guidelines.

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Imaging for Other Aortic Conditions (AB-19.2)

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- For imaging indications for other aortic conditions, see: **Aortic Imaging** in the Peripheral Vascular Disease Imaging Guidelines.

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Bowel Obstruction, Gastroparesis, and Bloating (AB-20)

Guideline

Bowel Obstruction (AB-20.1)
Gastroparesis and Dumping Syndrome (AB-20.2)
Nausea and Vomiting as the Primary Symptom (AB-20.3)
Superior Mesenteric Artery (SMA) Syndrome (AB-20.4)
Bloating, Gas, and Distention (AB-20.5)
References (AB-20)

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Bowel Obstruction (AB-20.1)

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- Suspected bowel obstruction:
  - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary
  - Pediatric individuals:
    - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary in lieu of CT, if requested.
  - Pregnant individuals:
    - MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195) is medically necessary
  - If the etiology or level of suspected intermittent or low-grade small bowel obstruction remains undetermined and additional imaging is needed after CT Abdomen and Pelvis ONE of the following is medically necessary:
    - CT Enteroclysis (CPT® 74176 or CPT® 74177) OR
    - CT Enterography (CPT® 74177) OR
    - MR Enteroclysis (CPT® 74183 and CPT® 72197) OR
    - MR Enterography (CPT® 74183 and CPT® 72197)
- CT Enterography (CPT® 74177) is medically necessary if there is a suspected small bowel tumor as a cause of the small bowel obstruction (including a history of no prior abdominal or pelvic surgery, no known hernia and/or concomitant obscure GI bleeding):
- When Crohn's Disease is the suspected cause of small bowel obstruction:
  - See: Suspected **IBD (Crohn's Disease or Ulcerative Colitis) (AB-23.1)** and **Known IBD (AB-23.2)**
- For small bowel obstruction in individuals with a history of bariatric surgery, see: **Bariatric Surgery Post-Operative Complications (AB-9.1)**

Background and Supporting Information

- Complete or high-grade obstruction can be defined as no fluid or gas passing beyond the site of obstruction. In incomplete or partial obstruction (low-grade), some fluid or gas passes beyond the point of obstruction. A plain film is not required prior to advanced imaging for suspicion of either high- or low- grade obstruction.

Evidence Discussion

In individuals suspected of having small or large bowel obstruction, the best imaging modality is CT of the abdomen and pelvis. Such imaging plays a crucial role in both diagnosis and management. Computed tomography (CT) is more useful than plain

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radiographs especially in identifying the severity, location, etiology, inflammation, and complications of bowel obstructions including ischemia, necrosis, and perforation. Magnetic resonance imaging (MRI) can be a useful alternative to CT imaging in special populations for whom radiation exposure needs to be limited, but the higher prevalence of motion artifact may make images more difficult to interpret.[1]

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Gastroparesis and Dumping Syndrome (AB-20.2)

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Gastroparesis

- Gastric Emptying Study (CPT® 78264) is medically necessary for the suspicion of delayed gastric emptying and ONE of the following:
  - Nausea, or vomiting of old food ingested several hours earlier
  - Bloating
  - Early satiety, or postprandial fullness
  - Recurrent aspiration
  - Unexplained poor glucose control in diabetes
  - Gastroesophageal reflux refractory to medical management
  - Non-ulcer dyspepsia
  - Retained gastric contents on endoscopy
- For the evaluation of suspected abnormalities in both total and regional times for small bowel transit
  - Gastric emptying study with small bowel transit (CPT® 78265) is medically necessary
- For the evaluation of suspected abnormalities in both total and regional times for colon transit
  - Gastric emptying study with small bowel and colon transit (CPT® 78266) is medically necessary

Dumping Syndrome

- Gastric Emptying Study (CPT® 78264) to evaluate signs or symptoms of dumping syndrome is not medically necessary.

Note: For both solid-phase and liquid-phase gastric emptying studies
- If performed on the same day by any protocol, CPT® 78264 should not be reported with two units, only one unit.
- If a solid-phase study is performed, and then on a later date a liquid-phase study is performed, one unit of CPT® 78264 should be reported for each date of service.

Evidence Discussion

Gastric emptying scintigraphy uses a radiolabeled solid meal to measure the rate of gastric emptying. This is the conventionally best accepted method to measure gastric

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emptying. It is performed two to four hours after ingestion of a radiolabeled meal. Performing the test for the longer duration is proposed to increase the accuracy of testing. Gastric emptying with small bowel or colonic transit time can provide further information regarding intestinal and colonic transit time. Gastric emptying scintigraphy has limited value in the evaluation of dumping syndrome, but remains the preferred method for diagnosis of gastroparesis.[3,4,6]

Dumping syndrome is a common complication of gastric and bariatric surgery in which changes in anatomy and innervation promote a rapid emptying of gastric contents into the small bowel. This triggers a series of physiologic responses. "Early dumping," occurs within the first hour after a meal and is characterized by abdominal pain, bloating, gassiness, nausea, vomiting, and diarrhea as well as vasomotor symptoms such as flushing, sweatiness, tachycardia, and hypotension. "Late dumping," symptoms occur between 1 and 3 hours after meals and are usually related to hypoglycemia (e.g. weakness, confusion, syncope).[24]

Dumping syndrome is usually a clinical diagnosis and the recommended diagnostic testing is an oral glucose tolerance test.[24]

Evidence-based guidelines have recently concluded that gastric emptying tests have low sensitivity and specificity for dumping syndrome, and that a gastric emptying test showing a rapid emptying rate would not be used to confirm a diagnosis of dumping syndrome. Rapid emptying can occur in other conditions, and it has been demonstrated that the initial rapid emptying in dumping may produce symptoms such as nausea, which they delays gastric emptying in dumping may produce symptoms such as nausea, which delays gastric emptying, such that the results of a gastric emptying study are in the normal range. Because of these limitation, recent guidelines have concluded that, "gastric emptying testing seems to be of low utility in diagnosing dumping syndrome."[18]

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Nausea and Vomiting as the Primary Symptom (AB-20.3)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- Nausea and vomiting as the primary symptom
  - An initial assessment should be performed prior to imaging requests. Diagnostic testing for nausea and vomiting should be targeted at finding the etiology suggested by a thorough history and physical examination. In the absence of more complicated or serious disease, if the cause is not obvious or suggestive from the history and physical, laboratory data including a CBC, chemistry profile, and, in a reproductive-age individual, pregnancy testing, should be performed prior to advanced radiographic imaging. Imaging is based on the findings of the initial evaluation as follows:
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for ANY of the following:
      - If the initial assessment does not suggest a specific cause
      - If the evaluation proves unproductive
    - Symptoms suggesting mucosal disease (e.g. GERD, suspicion of ulcer disease):
      - EGD prior to advanced imaging
    - If nausea and vomiting remains unexplained despite workup and CT Abdomen and Pelvis is negative:
      - Gastric emptying study (CPT® 78264)
    - Symptoms suggesting an intracranial etiology (vertigo/nystagmus, associated headache, or neurogenic vomiting suggested by a positional nature and/or associated with other neurologic signs and symptoms):
      - See: **Headache (HD-11)** , **Dizziness, Vertigo and Syncope (HD-23)** , or other Head Imaging Guidelines depending on the predominant neurologic presentation
      - See: **General Guidelines – Other Imaging Situations (HD-1.7)** in the Head Imaging Guidelines for persistent, unexplained nausea and vomiting, when GI evaluation is negative.
    - Nausea and vomiting associated with RUQ pain, dyspeptic symptoms or epigastric pain, see: **Right Upper Quadrant and Epigastric Pain (AB-2.3)**

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Evidence Discussion

Nausea and vomiting are common symptoms encountered in medicine. Prior to imaging studies, an evaluation including a detailed history including duration, frequency, and severity should be performed. Diagnostic testing for nausea and vomiting should focus on finding the etiology of the symptoms. In addition to a detailed history and physical examination, laboratory work up and pregnancy testing may reveal the etiology of symptoms. The purpose of the initial assessment is to define whether the symptom complex suggests a central (neurologic), endocrine (e.g. pregnancy, thyroid disorder), iatrogenic (chemotherapy/medication-induced), obstructive (e.g., low-grade small bowel obstruction), or a mucosal (gastritis, peptic ulcer disease) etiology. If mucosal disease causing vomiting is suspected, upper endoscopy should be performed prior to advanced imaging. If gallbladder disease is suspected, right upper quadrant ultrasound should be performed. If neurologic symptoms are present, advanced brain imaging may be indicated depending on symptoms and presentation. If the initial evaluation of nausea and vomiting does not reveal a specific cause, advanced imaging may be pursued. CT abdomen and pelvis with contrast provides valuable information regarding abdominal and pelvic anatomy such as obstruction or inflammation and may be used to evaluate nausea and vomiting when clinically appropriate.[7,10,11,12]

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Superior Mesenteric Artery (SMA) Syndrome (AB-20.4)

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- CTA Abdomen (CPT® 74175) or MRA Abdomen (CPT® 74185) are medically necessary for clinical suspicion of SMA syndrome and ANY of the following:
  - Recent significant weight loss which leads to a loss of retroperitoneal fat
  - Presence of a severe debilitating illness such as malignancy, malabsorption syndromes, AIDS, high-level trauma, and extensive burns
  - History of corrective spine surgery for scoliosis
  - Anorexia Nervosa
  - Abdominal surgery
  - Congenital short ligament of Treitz
  - Radiologic findings or history suggestive of duodenal obstruction
  - Failure to diagnose persistent nausea and/or vomiting despite the workup outlined in **Nausea and Vomiting as the Primary Symptom (AB-20.3)**

Background and Supporting Information

- SMA syndrome is a rare cause of duodenal obstruction in which there is a decrease in the aortomesenteric angle with resulting compression of the duodenum by the SMA.
- The typical clinical scenario includes an episode of weight loss followed by chronic food intolerance with nausea and vomiting, further weight loss, and epigastric pain, and can be relieved by lying prone or in the left lateral decubitus position.
- The diagnosis can be suspected with barium studies demonstrating delayed passage of contrast beyond the duodenum, dilatation of the first and second portions of the duodenum, anti-peristaltic flow of barium proximal to the obstruction, and relief of obstruction when placed in the prone, knee-chest, or left lateral position, or with an upper endoscopy revealing pulsatile extrinsic compression of the duodenum, or plain films suggesting duodenal obstruction.

Evidence Discussion

The gold standard test for suspicion of SMA syndrome is a CTA of the abdomen or an MRA of the abdomen, which confirms the diagnosis and provides a measurement of the angle between the SMA and the abdominal aorta. All other investigative modalities may suggest an obstruction at the third portion of the duodenum but are not diagnostic.[21-23]

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Bloating, Gas, and Distention (AB-20.5)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- For bloating as the primary symptom, present for at least 3 months, see: **Irritable Bowel Syndrome (AB-21.4)**
- For documented suspicion of bowel obstruction (e.g., individuals with prior abdominal surgery, previous history of SBO, known adhesions, history of Crohn's Disease, etc.) see: **Bowel Obstruction (AB-20.1)**.
- If associated with constipation, see: **Constipation (AB-21.3)**
- If associated with dyspeptic symptoms, see: **Right Upper Quadrant and Epigastric Pain (AB-2.3)**
- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if ANY of the following present:
  - History of malignancy with a likelihood or propensity to metastasize to abdomen
  - Fever (≥101 degrees Fahrenheit)
  - Elevated WBC >10,000, or above the upper limit of normal for the particular lab reporting the result
  - Low WBC (absolute neutrophil count <1000)
  - Palpable mass of clinical concern and/or without benign features
  - GI bleeding, overt or occult, not obviously hemorrhoidal
  - Abdominal tenderness documented as moderate or severe
  - Peritoneal signs, such as guarding or rebound tenderness
  - Suspected complication of bariatric surgery
  - Notation by the ordering provider that the patient has a "surgical abdomen"
  - Age >60 years with unintentional weight loss of ≥10 lbs. or ≥5% of body weight over 6 months or less, without an identifiable reason

Background and Supporting Information

Bloating and distension are among the most common gastrointestinal complaints, and appears in 96% of individuals with IBS, and 20-30% of the general population. Bloating is the subjective perception of increased abdominal pressure. Distension is the objective finding of increased abdominal girth.

The following approaches were offered by the American Gastroenterological Association (AGA)[20] as Best Practice Advice in evaluation and management of belching, abdominal bloating, and distension:

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- Clinical history and physical examination findings and impedance pH monitoring can help to differentiate between gastric and supra-gastric belching.
- Rome IV criteria (see also: **Irritable Bowel Syndrome [AB-21.4]**) should be used to diagnose primary abdominal bloating and distention.
- Carbohydrate enzyme deficiencies may be ruled out with dietary restriction and/or breath testing. In a small subset of at-risk patients, small bowel aspiration or biopsy may be warranted.
- Serologic testing may rule out celiac disease in individuals with bloating and, if serologies are positive, a small bowel biopsy should be done to confirm the diagnosis.
- Abdominal imaging and upper endoscopy should be restricted to individualss with alarm features, recent worsening symptoms, or an abnormal physical examination.
- Gastric emptying studies should not be ordered routinely for bloating and distention, but may be considered if nausea and vomiting are present. See also: **Gastroparesis and Dumping Syndrome (AB-20.2)**
- Whole gut motility and radiopaque transit studies should be restricted to individuals with refractory lower GI symptoms and suspected neuromyopathic conditions.
- When abdominal bloating and distention may be related to constipation or difficult evacuation, anorectal physiology testing is suggested to rule out a pelvic floor disorder. See also: **Constipation (AB-21.3)**

Evidence Discussion

Determining when symptoms of bloating, gas, and distention require imaging is done by risk stratification using demographics factors such as patient age as well as concomitant signs and symptoms.

- Computer tomography (CT) of the abdomen offers excellent 3-dimensional resolution of the gut and its surrounding structures, especially when performed with use of oral and/or intravenous (IV) contrast agents. CT imaging captures all of the abdominal organs and the surrounding cavity and mesentery. It is central to the evaluation of this condition because it can accurately diagnose the presence and location of obstruction, malignancy, vascular insufficiency, or infection, which are important pathologic diagnoses to identify or exclude in the subset of high-risk individuals. CT scan requires a significant dose of ionizing radiation but is ideally suited to imaging lesions within the gut because the speed of image acquisition reduces the potential for motion artifact. Typically performed with IV contrast in individuals with normal kidney function, there is the added risk of allergic reaction to contrast; however the contrast enhances the ability to evaluate for both infectious and vascular conditions.[8,9,19]

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Diarrhea, Constipation, and Irritable Bowel (AB-21)

Guideline

Acute and Persistent Diarrhea (Up to 30 Days) (AB-21.1)
Chronic Diarrhea (More than 30 Days) (AB-21.2)
Constipation (AB-21.3)
Irritable Bowel Syndrome (AB-21.4)
References (AB-21)

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Acute and Persistent Diarrhea (Up to 30 Days) (AB-21.1)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- Routine advanced imaging is not medically necessary for uncomplicated diarrhea (up to 30 days in duration).
- Prior to advanced imaging, individuals with travel and/or dysenteric (including bloody) diarrhea should undergo biological assessment and/or antimicrobial treatment.
- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if:
  - Suspected ischemia (See: **Mesenteric Ischemia (AB-6.1)** and **Colonic Ischemia (AB-6.2)**) OR
  - Older (>50) individuals with significant abdominal pain OR
  - Previous gastric bypass OR
  - Immunocompromised OR
  - Obstruction, toxic megacolon, or perforation suspected

Evidence Discussion

Acute or persistent diarrhea is a common complaint that most often results from self-limited infectious or digestive causes, and for this reason, imaging is generally not medically necessary. Four weeks or more is used as the time frame to describe when diarrhea is no longer considered acute because "intestinal infections, the most common cause of acute diarrhea, typically resolve in 1 week or 4 weeks at the most".[14] However, in a subset of individuals and in the setting of clinical suspicion, imaging is medically necessary to exclude vascular insufficiency, perforation, obstruction and severe metabolic derangement. Determining the situations in which imaging is medically necessary is based on provider concern for such conditions in addition to demographic factors such as age and prior medical and surgical history. When imaging is medically necessary, CT scan with contrast is the modality of choice.[2,11]

- Computer tomography (CT) of the abdomen offers excellent 3-dimensional resolution of the gut and its surrounding structures, especially when performed with use of oral and/or intravenous (IV) contrast agents. CT imaging captures all of the abdominal organs and the surrounding cavity and mesentery. It is central to the evaluation of this condition because it can accurately diagnose the presence and location of obstruction, malignancy, vascular insufficiency, toxic megacolon, and perforation in the subset of high-risk individuals. CT scan requires a significant dose of ionizing radiation but is ideally suited to imaging lesions within the gut because the speed of

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image acquisition reduces the potential for motion artifact. Typically performed with IV contrast in individuals with normal kidney function, there is the added risk of allergic reaction to contrast, however the contrast enhances the ability to evaluate for both infectious and vascular conditions.[2,11]

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Chronic Diarrhea (More than 30 Days) (AB-21.2)

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- Prior to advanced imaging, basic lab work should include routine CBC, chemistries, as well as stool tests for pathogens.
- CT Abdomen with contrast (CPT® 74160), CT Abdomen and Pelvis with contrast (CPT® 74177), CT Enterography (CPT® 74177), or MR Enterography (CPT® 74183 or CPT® 74183 and CPT® 72197), is considered medically necessary if ALL of the following have been performed:
  - Colonoscopy has been performed and is nondiagnostic or suggestive of inflammatory bowel disease
  - Fecal calprotectin or fecal lactoferrin
  - Testing for giardia antigen or PCR for giardia
  - Testing for celiac disease with serum IgA tissue transglutaminase (tTG)
- See: **Suspected IBD (Crohn's Disease or Ulcerative Colitis) (AB-23.1)** for concerns regarding inflammatory bowel disease.

Evidence Discussion

Chronic diarrhea is defined as "a condition in which diarrhea that persists or recurs for four weeks or more causing various problems in daily life." Four weeks or more is used as the time frame because "intestinal infections, the most common cause of acute diarrhea, typically resolve in 1 week or 4 weeks at the most".[14]

The initial evaluation of chronic diarrhea (more than 30 days) involves non-imaging modalities (blood tests, stool tests, and colonoscopy), to evaluate for celiac disease, giardia, and inflammatory bowel disease. If these evaluations are non-diagnostic, imaging can be considered to identify more unusual causes of chronic diarrhea such as obstruction, malignancy, biliary causes, and small bowel disorders such as small bowel Crohn's disease.[6,11]

- Computer tomography (CT) of the abdomen offers excellent 3-dimensional resolution of the gut and its surrounding structures, especially when performed with use of oral and/or intravenous (IV) contrast agents. CT imaging captures parts or the whole of the abdomen, or can be directed to interrogate with specialized techniques a specific organ. Depending on clinical suspicion, for this condition, CT of the abdomen, CT of the abdomen and pelvis or specialized CT enterography of the small bowel may be employed. CT scan requires a significant dose of ionizing radiation, but is ideally

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suited to imaging lesions within the gut because the speed of image acquisition reduces the potential for motion artifact.[6,11]

- Magnetic resonance imaging (MRI) uses a magnetic field to capture excellent 3-dimensional resolution. As with CT scans, the technique is often performed with IV contrast agents, and can with specialized techniques be directed either at whole or parts of the abdomen or at specific abdominal structures. For this condition MR enterography delivers high resolution images of small bowel mucosa to evaluate for the subtle inflammatory changes such as those seen in small bowel Crohn's disease. MRI yields better soft contrast resolution than CT and does not expose individuals to ionizing radiation, but due to longer image time is motion artifact-prone and thus less suited to resolving gastrointestinal detail. In addition, and especially in youths, MRI may require sedation.[6,11]

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Constipation (AB-21.3)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if:
  - Concern for obstruction
- MRI (MRI Pelvis without contrast CPT® 72195) for Defecography is considered investigational/experimental by UHC.

Background and Supporting Information

- The work-up and treatment of constipation usually proceeds with a history and physical followed by empiric medication or dietary trials.
  - In general, a colonoscopy is performed prior to advanced imaging in an individual presenting with chronic constipation if the alarm symptoms of blood in the stool, anemia, or weight loss are present.

Evidence Discussion

Clinical presentation and results of minimally invasive testing determine the situations in which constipation requires imaging.

- Computer tomography (CT) of the abdomen offers excellent 3-dimensional resolution of the gut and its surrounding structures, especially when performed with use of oral and/or intravenous (IV) contrast agents. CT imaging captures all of the abdominal organs and the surrounding cavity and mesentery. It is central to the evaluation of individuals with constipation alongside red flag symptoms that suggest infection or malignancy. CT scan requires a significant dose of ionizing radiation but is ideally suited to imaging lesions within the gut because the speed of image acquisition reduces the potential for motion artifact. Typically performed with IV contrast in individuals with normal kidney function, there is the added risk of allergic reaction to contrast; however, the contrast enhances the ability to evaluate for both infectious and malignant conditions.[3,4,5]

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Irritable Bowel Syndrome (AB-21.4)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- Advanced imaging in the absence of alarm symptoms has a very low yield, but can be considered medically necessary in the following circumstances:
  - CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary in the following circumstances:
    - Presence of ANY of the following alarm symptoms:
      - Weight loss
      - Frequent nocturnal awakenings due to gastrointestinal symptoms
      - Fever
      - Blood in the stool or iron deficiency anemia (See: **GI Bleeding (AB-22)** for appropriateness of imaging in this circumstance)
      - New onset and progressive symptoms
      - Onset of symptoms after age 50
      - Family history of colon cancer or inflammatory bowel disease
      - Findings of an abdominal mass
      - Presence of lymphadenopathy
    - After ALL of the following studies have been performed:
      - Fecal calprotectin ≥50ug/g or fecal lactoferrin ≥4.0ug/g or CRP >0.5 in individuals with diarrhea-predominance
      - Testing for giardia antigen or PCR for giardia
      - Celiac testing should also be performed in individuals with diarrhea-predominance IBS, and if positive see: **Celiac Disease (AB-24.1)** for imaging guidance.
        - See also: Background and Supporting Information in IBD **(Crohn's Disease or Ulcerative Colitis) (AB-23.1)**

Background and Supporting Information

- Irritable bowel syndrome is characterized by abdominal pain associated with altered bowel habits, abdominal distention, and bloating. It is important to understand that IBS is a positive diagnosis, not a diagnosis of exclusion. ACG guidelines (2021) strongly suggest that IBS be assessed with a “positive diagnostic strategy as compared to a diagnostic strategy of exclusion”. Subtypes include IBS-C (constipation-predominant), IBS-D (diarrhea-predominant), IBS-M (mixed), and unclassified IBS. Rome IV Criteria for the diagnosis of irritable bowel syndrome are:

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- Recurrent abdominal pain, on average ≥1 d/wk in the past 3 months, related to ≥2 of the following:
  - Defecation
  - Change in stool frequency
  - Change in stool appearance (form)

Evidence Discussion

Risk stratification (using demographics factors such as individual age, family history, timing of symptoms, concomitant symptoms, and physical exam findings) determines the situations in which imaging is medically necessary for irritable bowel syndrome. In a subset of individuals, imaging is medically necessary to exclude inflammatory conditions such as Crohn's disease and malignant conditions such as bowel cancer.[7,8,9]

- Computer tomography (CT) of the abdomen offers excellent 3-dimensional resolution of the gut and its surrounding structures, especially when performed with use of oral and/or intravenous (IV) contrast agents. CT imaging captures all of the abdominal organs and the surrounding cavity and mesentery. It is central to the evaluation of this condition because it can accurately identify both the presence and location of inflammatory conditions and malignant conditions in the appropriately identified subset of high-risk individuals. CT scan requires a significant dose of ionizing radiation but is ideally suited to imaging lesions within the gut because the speed of image acquisition reduces the potential for motion artifact. Typically performed with IV contrast in individuals with normal kidney function, there is the added risk of allergic reaction to contrast; however, the contrast enhances the ability to evaluate for both inflammatory and malignant conditions.[7,8,9]

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GI Bleeding (AB-22)

Guideline

GI Bleeding (AB-22.1)
Small Bowel Bleeding Suspected (AB-22.2)
References (AB-22)

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GI Bleeding (AB-22.1)

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- CTA Abdomen (CPT® 74175), CTA Abdomen and Pelvis (CPT® 74174), or CT Abdomen and Pelvis with contrast (CPT® 74177) are medically necessary for the initial evaluation of ANY of the following:
  - If therapeutic angiography is being considered
  - If colonoscopy cannot be performed in an individual with active lower GI bleeding
  - If endoscopy cannot be performed in an individual with active upper GI bleeding
  - If surgery is being considered for treatment of GI bleeding
  - GI bleeding and moderate to severe abdominal pain and/or tenderness
  - GI bleeding and hemodynamic instability
  - If there is concern for an aorto-enteric fistula (known or suspected aortic aneurysm, history of any type of aortic aneurysm repair)
- If there is brisk active bleeding AND endoscopy does not reveal a source
  - Gastrointestinal Bleeding Scintigraphy (CPT® 78278) is medically necessary
- Iron Deficiency Anemia:
  - If the bleeding is of suspected gastrointestinal origin:
    - Upper endoscopy and colonoscopy should be performed, unless contraindicated
    - Small bowel video capsule endoscopy is medically necessary, if endoscopies are negative
    - If small bowel video capsule endoscopy is negative OR for further evaluation of abnormal video capsule findings ONE of the following is medically necessary:
      - CT Abdomen and Pelvis with contrast (CPT® 74177)
      - CT Enterography (CPT® 74177)
      - MR Entergraphy (CPT® 74183 or CPT® 74183 and CPT® 72197) is medically necessary if there is a contraindication to CT Enterography.
- If the bleeding is determined to be non-gastrointestinal, refer to the appropriate condition-based guideline.

Evidence Discussion

In individuals suspected of having GI bleeding, after initial endoscopic evaluation if feasible, the best imaging modality is CT or CTA of the abdomen and pelvis. Such imaging plays a crucial role in both diagnosis and management. Computed tomographic angiography (CTA) is more expedient and accurate at localizing the site of bleeding as compared to gastrointestinal bleeding scintigraphy (tagged RBC scintigraphy) which can be a useful alternative in the setting of active GI bleeding, especially if it is slow or

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intermittent. CTA is the exam of choice for potential causes of catastrophic bleeding such as aortoenteric fistula, transmural bowel injuries, and mesenteric hemorrhage.[24]

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Small Bowel Bleeding Suspected (AB-22.2)

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- If the small bowel is the suspected source of bleeding, and if endoscopic studies (upper endoscopy, colonoscopy and if available video capsule endoscopy) are unrevealing:
  - CT Enterography (CPT® 74177) is medically necessary
  - MR Enterography (CPT® 74183 or CPT® 74183 and CPT® 72197) is medically necessary, if there is a contraindication to CT Enterography
- Meckel's scan (CPT® 78290) is medically necessary if bleeding is suspected from a Meckel's diverticulum.

Evidence Discussion

The goal of identifying the source of GI tract bleeding is to identify lesion, location, and ability to perform therapeutic intervention. Bleeding from the small bowel is uncommon, accounting for approximately 5–10% of all individuals presenting with gastrointestinal (GI) bleeding. The initial diagnostic modality of choice is endoscopy or colonoscopy to help identify lesions and execute appropriate interventions.[6,19, 20,21,22,23]

Video capsule endoscopy (VCE) is considered a first-line modality for small bowel investigation. Its main advantages are that it is noninvasive and allows examination of the entire length of the small bowel in 70-90% individuals with diagnostic yield of 38–83% in individuals with suspected small bowel bleeding. The main utility of this test lies in its high positive (94–97%) and negative predictive value (83–100%) in the evaluation of GI bleeding. Findings on VCE leading to endoscopic or surgical intervention or a change in medical management have been reported in 37–87% of individuals.[6,19, 20,21,22,23]

Providers occasionally request a CT or MR Enterography prior to the administration of a VCE, in order to assess whether there is pathology that might impede passage of the capsule and cause retention. This is not supported as a routine procedure prior to VCE. It should be noted that a patency capsule is available, and that this may identify individuals at higher risk of retention. However, guidance from the consensus group of the American College of Gastroenterology recommends that in individuals with obstructive symptomatology, imaging (MR Enterography or CT Enterography) should be performed prior to VCE. This group would also include high risk individuals with a known history of Crohn's Disease, known history of strictures or other obstruction, history of previous pelvic or abdominal radiation, or suspected tumor.[6,19, 20,21,22,23]

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Computed tomographic enterography is indicated in individuals with suspected obstruction before VCE or after negative VCE examinations, women who are pregnant, and individuals who are unable to swallow the VCE capsule.[6,19, 20,21,22,23]

Cross-sectional imaging techniques optimized for imaging the small bowel are advantageous due to ability to see all bowel loops without superimposition and the visualization of extra-luminal structures. Enterography can be performed with either CT or MR. CT is more widely used in the setting of GI bleeding because of the superior temporal and spatial resolution compared with MR and is more widely available. CT can detect vascular and inflammatory abnormalities, which may be missed on VCE. Because of the small number of studies regarding MR enterography, this exam is not routinely recommended in lieu of CT enterography, but can be considered medically necessary in individuals aged <40 years because of lower radiation exposure.[6,19,20,21,22,23]

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Inflammatory Bowel Disease (AB-23)

Guideline

Suspected IBD (Crohn’s Disease or Ulcerative Colitis) (AB-23.1)
Known IBD (AB-23.2)
Perirectal/Perianal Crohn's Disease (AB-23.3)
Primary Sclerosing Cholangitis (PSC) (AB-23.4)
References (AB-23)

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Suspected IBD (Crohn's Disease or Ulcerative Colitis) (AB-23.1)

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- Suspected Crohn's Disease or Ulcerative Colitis:
  - CT Abdomen and Pelvis with contrast (CPT® 74177) OR CT Enterography (CPT® 74177) OR MR Enterography (CPT® 74183 or CPT® 74183 and CPT® 72197) is medically necessary for ANY of the following:
    - History of malignancy with a likelihood or propensity to metastasize to abdomen
    - Fever (≥101 degrees Fahrenheit)
    - Elevated WBC >10,000, or above the upper limit of normal for the particular lab reporting the result
    - Palpable mass of clinical concern and/or without benign features
    - GI bleeding, overt or occult, not obviously hemorrhoidal
    - Abdominal tenderness documented as moderate or severe
    - Peritoneal signs, such as guarding or rebound tenderness
    - Suspected complication of bariatric surgery
    - Notation by the ordering provider that the patient has a "surgical abdomen"
    - Age >60 years with unintentional weight loss of ≥10 lbs. or ≥5% of body weight over 6 months or less, without an identifiable reason
  - Chronic diarrhea without the above signs or symptoms, see: **Diarrhea, Constipation, and Irritable Bowel (AB-21)**
  - CT Enterography (CPT® 74177) OR MR Enterography (CPT® 74183 or CPT® 74183 and CPT® 72197) is medically necessary for the evaluation of chronic abdominal pain with diarrhea in the absence of red flag criteria (as noted in **General Guidelines AB-1.0**) when there is documented concern for inflammatory bowel disease and if ANY of the following:
    - There is a positive family history of inflammatory bowel disease, **OR**
    - There are endoscopy or colonoscopy findings suggestive of inflammatory bowel disease, **OR**
    - Elevated inflammatory markers (fecal lactoferrin ≥4.0 ug/g, CRP >0.5 mg/dL, or fecal calprotectin ≥50 ug/g), **OR**
    - Diagnosis is still in doubt after colonoscopy and evaluation of inflammatory markers, and Crohn's disease is suspected
  - CT Abdomen and Pelvis with or without contrast (CPT® 74177 or CPT® 74176) can be performed prior to endoscopy if requested by or in consultation with the provider who will be performing the endoscopy.

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Note:

Serologic markers and genetic markers are currently under investigation with regards to their value in diagnosing inflammatory bowel disease, and are sometimes used as a screening test for IBD in which other examinations are negative. At the current time they are not considered suitable as a screening test for inflammatory bowel disease in individuals with GI symptoms, and the routine use of serologic or genetic markers for the diagnosis of IBD is not indicated. Thus, an isolated positive marker result in an individual without any other findings to suggest IBD, especially in the presence of negative inflammatory markers and endoscopic examinations, is not, in and of itself, an indication for advanced imaging.

- Serologic markers include anti-glycan antibodies, such as ASCA, ACCA, ALCA, AMCA, Anti-L, Anti-C, Anti-OmpC, Anti-Is, Anti-Cbir, pANCA, PAB, GAB

Background and Supporting Information

Studies have demonstrated the negative predictive value of a low fecal calprotectin and CRP with regards to inflammatory bowel disease. Chey, et al. in a meta-analysis demonstrated that a fecal calprotectin <40mcg/g or a CRP ≤0.5 mg/dl effectively excludes inflammatory bowel disease in individuals with IBS. Katsinelos, et al. reviewed wireless capsule endoscopy results in individuals with abdominal pain and diarrhea. The diagnostic yield of capsule endoscopy in individuals with abdominal pain and diarrhea with positive inflammatory markers was 90.1%, and 0% in indviduals with abdominal pain and diarrhea with negative inflammatory markers. This led the Canadian Association of Gastroenterology to recommend against the use of capsule endoscopy in individuals with chronic abdominal pain or diarrhea as their only symptoms and no evidence of biomarkers associated with Crohn's Disease, stating "CE (capsule endoscopy) is not warranted in most indviduals who present with chronic abdominal pain in the absence of positive tests for inflammatory markers or abnormal findings on endoscopy or imaging".

Evidence Discussion

In individuals with suspected inflammatory bowel disease, cross-sectional imaging can be performed after initial endoscopy is suggestive of inflammatory changes or if abnormal inflammatory markers concerning for IBD, or positive family history of IBD. Cross-sectional imaging methods such as computed tomography and magnetic resonance imaging are complementary to endoscopy, which allows diagnosis of disease when endoscopy is negative and diagnosis is still in doubt.[1,2,3,4,16]

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Known IBD (AB-23.2)

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- CT Abdomen and Pelvis with contrast (CPT® 74177), CT Enterography (CPT® 74177), or MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195) is medically necessary for known Crohn's Disease or Ulcerative Colitis and ANY of the following:
  - To determine change in treatment
  - Monitoring response to therapy
  - Suspected complications including abscess, perforation, fistula, or obstruction

Evidence Discussion

Cross-sectional imaging methods such as computed tomography and magnetic resonance imaging are utilized to evaluate IBD disease activity, extra-enteric complication and response to therapy with a great impact on patient management. Magnetic resonance imaging (MRI) either with or without contrast or diffusion weighted has now emerged as suitable radiation-free alternative to CT imaging, with comparable diagnostic accuracy. [1,2,3,4,20,21,22,23]

MRE should be used preferentially in young individuals and in individuals in whom it is likely that serial exams will need to be performed, because of the absence of any radiation exposure.[1,2,3,4,20,21,22,23]

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Perirectal/Perianal Crohn's Disease (AB-23.3)

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- Perirectal/Perianal Fistula:
  - MRI Pelvis without and with contrast (CPT® 72197)
  - Endoscopic ultrasound is preferential to CT in this setting.
  - CT Pelvis with contrast (CPT® 72193) is an inferior study in this setting, and should be used when MRI or Endoscopic ultrasound cannot be performed.
- Perirectal/Perianal Abscess:
  - MRI Pelvis without and with contrast (CPT® 72197)
  - CT Pelvis with contrast (CPT® 72193) is inferior but can be considered medically necessary as an alternative if desired.

This section is applicable to individuals with Crohn's Disease. See: **Fistula in Ano (PV-21.1)** and **Perirectal Abscess (PV-21.2)** in the Pelvis Imaging Guidelines for non-Crohn's related perirectal and/or perianal fistulae.

Evidence Discussion

Cross-sectional imaging methods such as magnetic resonance imaging and computed tomography are utilized to evaluate Crohn's related complications like perirectal and/or perianal fistulae or abscess. CT is useful in evaluating abscesses and inflammation; however, due to its limited resolution, defining fistulas may be difficult. MRI, which has better resolution, along with endoscopic ultrasound, are highly accurate in defining perianal and perirectal fistulas and are the preferred modalities for diagnosing fistulas secondary to Crohn's disease.[4]

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Primary Sclerosing Cholangitis (PSC) (AB-23.4)

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- Primary Sclerosing Cholangitis (PSC):
  - MRCP is medically necessary to assess for PSC in those:
    - with IBD and any elevated liver study (including alkaline phosphatase, GGTP, bilirubin, AST, or ALT)
    - without IBD, but with persistent cholestatic liver tests. (See: **Abnormal Liver Chemistries (AB-30)**)
  - Ultrasound (CPT® 76700 or 76705) or MRI/MRCP is medically necessary as surveillance for cholangiocarcinoma in individuals with PSC every 6 months.

Background and Supporting Information

Primary sclerosing cholangitis (PSC) is a chronic liver and biliary tract disease that can result in stricturing and fibrosis of the intra- and extra- hepatic biliary ducts, as well as end-stage liver disease. It is most often associated with inflammatory bowel disease. Biliary obstruction can occur anywhere along the biliary tree, resulting in cholangitis, and there is a high risk of the development of cholangiocarcinoma, which must be strongly considered in individuals with PSC and a dominant stricture, as well as an increased risk of gallbladder polyps and other malignancies. As such, imaging plays an important role in the diagnosis and follow-up of PSC.

See: **Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)** Background and Supporting Information PSC (Primary Sclerosing Cholangitis) vs PBC (Primary Biliary Cholangitis)

Evidence Discussion

The diagnosis of primary sclerosing cholangitis can be confirmed via magnetic resonance cholangiography (MRCP) when suspected, in individuals with IBD or in individuals with persistent cholestasis, in the absence of known IBD. Surveillance for cholangiocarcinoma in individuals with PSC is important because it allows for disease progression as well as monitoring for the development of cancer. Studies show that there is survival benefit to routine surveillance with MRCP, although ultimately the reason for this survival benefit is not clear.[24] MRI has been shown to have better diagnostic performance than ultrasound in detecting early stage cholangiocarcinoma (a potential risk for individuals with PSC) and earlier detection is associated with more favorable prognostic features.[25] Data is variable on the optimal timeframe for

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surveillance imaging with some sources using benchmarks of annual imaging; this guideline allows for regular cross-sectional imaging with ultrasound or MR every 6 months.[5,18,19,24,25]

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Celiac Disease (Sprue) (AB-24)

Guideline

Celiac Disease (AB-24.1)
References (AB-24)

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Celiac Disease (AB-24.1)

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- CT Abdomen and Pelvis with contrast (CPT® 74177), CT Enteroclysis (CPT® 74176 or CPT® 74177), or CT Enterography (CPT® 74177), or MR Enterography (CPT® 74183, or CPT® 74183 and CPT® 72197) is medically necessary for:
  - one-time study after initial, confirmed diagnosis of celiac disease OR
  - confirmed celiac disease and new or continued symptoms (e.g., bloating, diarrhea, abdominal pain, weight loss, distention, evidence of malabsorption, anemia) despite adherence to 6 months of a gluten free diet

Background and Supporting Information

- Celiac is an autoimmune disease in which the villi of the small intestine are damaged from eating gluten (found in wheat, barley, and rye).
- Complications of celiac disease include ulcerative jejunitis, lymphoma, and small intestinal adenocarcinoma.
- Diagnosis is made by blood testing:
  - Anti-tissue transglutaminase antibody [anti-tTG], anti-endomysium antibody (EMA), total IgA count, CBC to detect anemia, ESR, C-reactive protein, complete metabolic panel, vitamin D, E, B12 levels.
- Endoscopy with biopsy of the small bowel is performed to confirm the diagnosis of celiac disease if anti-tTG and/or EMA tests are positive.
- Capsule endoscopy may be used to confirm diagnosis of celiac disease in individuals with positive serology and negative biopsy, or when there is contraindication to biopsy or EGD.

Evidence Discussion

Serologic studies with antibody testing and upper endoscopy and small bowel biopsies are usually performed to confirm the diagnosis of celiac disease.[1] The findings on standard barium examination are often not specific. Abdominal pain, bloating, diarrhea, and evidence of malabsorption are frequent symptoms of celiac disease, as well as indications for CT imaging. The use of standard CT abdominal imaging, as well as CT Enteroclysis and CT Enterography, allow for the noninvasive assessment of the small bowel to evaluate the extent of disease and identify complications of the disease (including ulcerative jejunoileitis, lymphoma, and small bowel tumors). Early diagnosis of these disorders allows specific treatment to be initiated to prevent increased morbidity and mortality. Added advantages of CT imaging for the diagnosis of celiac disease are simultaneous visualization of the small and large bowel, as well as visualization of mesenteric lymph nodes to determine the presence of mesenteric adenopathy.[5,7]

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CT Colonography (CTC) (AB-25)

Guideline

CTC (AB-25.1)
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Note:

A screening CTC (CPT® 74263) can ONLY be used for an individual who is a candidate for average risk screening as defined below. It cannot be used for any other indication. If the request for a CTC is for any other reason than average risk screening, please refer to diagnostic CTC indications. A diagnostic CTC would be the appropriate code, if approvable, for any other reason than average risk screening. This would include surveillance for a history of colon polyps, the evaluation of a change in bowel habits, abdominal pain, bleeding, etc. Please refer to the definition below of an average-risk individual, as well as the circumstances for which a diagnostic CTC is appropriate.

- Screening CTC (CPT® 74263) for colorectal cancer is NOT medically necessary if:
  - FIT-DNA (multi-targeted stool DNA test) within the last 3 years, OR
  - colonoscopy within the last 10 years
- Screening CTC (CPT® 74263) is medically necessary every 5 years for colorectal cancer for:
  - Average-risk individuals ages 45 to 75
    - Average risk is defined as:
      - no previously diagnosed colorectal cancer, or colonic adenomas, or inflammatory bowel disease involving the colon
  - Individuals between 76 to 85 if there is no history of a previously negative colonoscopy or CTC, or, if in the opinion of the provider, the benefits of screening outweigh the risks.
  - Individuals with a SINGLE first-degree relative diagnosed at age >60 years with colorectal cancer or an advanced adenoma can be screened with CTC beginning at age 40.
    - If there are 2 or more first degree relatives at any age with CRC or an advanced adenoma, or a first degree relative <60, the individual should be screened via colonoscopy, not CTC.
- Diagnostic CTC without contrast (CPT® 74261) is medically necessary for:
  - Failed conventional colonoscopy due to a known colonic lesion, structural abnormality, or technical difficulty, and/or
  - Conventional colonoscopy is medically contraindicated. Contraindications may include:
    - Coagulopathy
    - Intolerance to sedation

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    - Elderly ≥80 years of age
    - Recent (within the last 60 days) myocardial infarction (MI)
- Diagnostic CTC with contrast (CPT® 74262) is medically necessary if:
    - there is a known obstructing colorectal malignancy so that staging prior to surgery can be performed, if desired.
    - there is a clearly stated indication for IV contrast to evaluate extra-colonic organs. When performed in this setting, a CTC with contrast will substitute for a CT Abdomen and Pelvis such that an additional CT Abdomen and Pelvis would generally not be medically necessary.
- MRI Colonography:
    - Currently, no published society-endorsed guideline with respect to colorectal cancer screening lists MRI Colonography as an alternative screening study. As such, requests for MRI Colonography would be considered investigational at this time. There is no specific CPT assigned for this procedure. It is sometimes requested as an MRI Abdomen and MRI Pelvis.

Background and Supporting Information

CT Colonography is routinely performed without contrast, and IV contrast is not needed in most cases.

Evidence Discussion

When it comes to screening with CT colonography, guidelines differ regarding the best approach for colorectal cancer (CRC) screening in asymptomatic, average-risk individuals. Generally, CTC is not advised for screening in individuals at an increased risk for CRC. This includes those with a history of adenomas or CRC, inflammatory bowel disease, or familial CRC syndromes.[1,2,3,4]

CTC is comparable to colonoscopy in terms of sensitivity and specificity, takes only about 15 minutes, is non-invasive, and often requires no sedation. However, the cathartic agents recommended for CTC are the same as those for conventional colonoscopy. Additionally, CTC imaging is associated with considerable radiation exposure and detected polyps cannot be removed during the procedure. Therefore, those with positive findings on their CTC will require a follow-up colonoscopy.[1,2,3,4]

Notably, the American Cancer Society and US Preventive Services Task Force recommend CTC for screening.[3,8,9]

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Cirrhosis and Liver Screening for Hepatocellular Carcinoma (HCC); Ascites and Portal Hypertension (AB-26)

Guideline

Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)
Ascites (AB-26.2)
Portal Hypertension (AB-26.3)
Monitoring After Fontan Procedure (AB-26.4)
References (AB-26)

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Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)

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HCC Screening and Surveillance

- Ultrasound (CPT® 76700 or CPT® 76705) every 6 months for HCC screening is medically necessary in ANY of the following circumstances:
  - All individuals, regardless of etiology, with cirrhosis or advanced fibrosis (e.g., Fibrosis Score F3 or greater on an elastography study, or results of a lab study such as FIB-4 or a biopsy indicative of severe activity or advanced fibrosis).
  - All individuals with Hepatitis B, regardless of the presence of cirrhosis or advanced fibrosis.
- See: **Hepatic Arteries and Veins (AB-43.1)** for individuals with Chronic Budd-Chiari Syndrome (BCS) and HCC surveillance in Budd-Chiari Syndrome/Hepatic Vein Thrombosis.
- See: **Monitoring After Fontan Procedure (AB-26.4)** for individuals who have undergone the FONTAN procedure.
- See: **General Guidelines (AB-1.0)** for use of medications or treatments which increase risk of HCC.

Evaluation of Liver Nodules

- If liver nodule is identified in the setting of chronic liver disease:
  - Less than 1cm
    - Repeat US in 3 months, then every 3 to 6 months
    - If stable for 2 years, then return to US every 6 months
  - Greater than or equal to 1cm
    - Multiphase CT Liver (either CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) should be performed.
      - If negative: Return to routine surveillance via US in 6 months.
      - If Li-RADS NC (non-categorizable): Repeat the same study or an alternative diagnostic imaging ≤3 months. (Note: non-categorizable refers to a technical problem with the study, such as image omission or severe degradation)
      - If Li-RADS 1 (definitely benign): Return to routine surveillance via US in 6 months.

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  - If Li-RADS 2 (probably benign): CT or MRI in 6 months can be approved (US requests are approvable if desired). If unchanged, return to routine surveillance via US.
  - If Li-RADS 3 (intermediate): CT or MRI in 3-6 months, and can be repeated every 6 months 2 more times, for a total of 18 months from the initial finding. If no change by 18 months, return to US surveillance every 6 months.
  - If Li-RADS 4 (probable HCC): Repeat or alternative imaging in ≤3 months. If HCC confirmed: See: **Upper GI Cancers (ONC-14)** in the Oncology Imaging Guidelines.
  - If Li-RADS 5 (HCC confirmed): See: **Upper GI Cancers (ONC-14)** in the Oncology Imaging Guidelines.
  - If Li-RADS M (Malignant, not definitely HCC): Repeat or alternative imaging in ≤3 months, and follow appropriate Oncology guidelines upon diagnosis.

- Exceptions to the above algorithms:
  - Advanced imaging (Multiphase CT Liver (either CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) for surveillance may be substituted for US in the following circumstances:
    - Obesity (BMI >35)
    - Marked parenchymal heterogeneity (suggestive of advanced liver fibrosis) noted on US
    - Visualization limitations noted on US which could be technical (such as obscuration by intestinal gas, chest wall deformity, etc.), or those related to structural or parenchymal changes in the liver
  - For individuals on the Liver Transplant list: See: **Liver Transplant, Pre-Transplant (AB-42.1)**
  - For Magnetic Resonance Elastography (MRE) indications, see: **Liver Elastography (AB-45)**
- Alpha-fetoprotein ≥20 ng/mL: Multiphasic CT or MRI Abdomen:
  - Further imaging should follow the above Li-RADS algorithm, depending on the findings of the CT or MRI.
  - If the initial CT or MRI does not reveal a lesion, but the AFP increases on subsequent testing, additional advanced imaging by CT or MRI may be approved.
- Contrast-Enhanced Ultrasound (CEUS)
  - Further studies are needed to assess the value of CEUS in this setting, and it is not medically necessary at this time.

Background and Supporting Information

When performed for liver lesion evaluation, a multiphase CT protocol may include non-contrast imaging as well as arterial, portal venous, and delayed-phase post-contrast imaging. However, these protocols do not always require non-contrast imaging which

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may not provide additional information in many scenarios. Therefore, a multiphase CT for liver lesion evaluation can be requested as CPT® 74160 (CT Abdomen with contrast) or CPT® 74170 (CT Abdomen without and with contrast).

The presence of liver disease in the absence of advanced fibrosis or cirrhosis, with the exception for those circumstances indicated above, is not an indication for screening. This would include, for example, MASLD (metabolic dysfunction associated steatotic liver disease, formerly known as NAFLD), the presence of which is not an indication for screening in the absence of either advanced fibrosis or cirrhosis

The American Association for the Study of Liver Diseases (AASLD) revised its guidelines with respect to surveillance for HCC in individuals with cirrhosis in 2018. The recommended algorithm now includes either US alone or US with serum AFP every 6 months. It should be noted that “modification of this surveillance strategy based on the etiology of liver diseases or risk stratification models cannot be recommended at this time.”

In addition, the AASLD also issued a subsequent Practice Guidance in 2018 and this document forms the basis of these guidelines. The AASLD has adopted the Li-RADS classification of liver lesions with respect to HCC surveillance imaging for individuals with advanced liver disease, and follow-up imaging protocols are based on this system. In view of this, the Li-RADS classification now informs imaging protocols used in this guideline.

Note: PSC (Primary Sclerosing Cholangitis) vs. PBC (Primary Biliary Cholangitis)

These 2 entities sound similar, and both are cholestatic, but they are different diseases, and as such have different monitoring requirements.

PSC is an idiopathic cholestatic disease characterized by chronic inflammation, progressive fibrosis, and stricturing of the *medium and large-sized* extra-hepatic or intra-hepatic bile ducts. Segmental bile duct dilation proximal to areas of stricturing creates the characteristic beaded appearance on a cholangiogram, such as MRCP. This may progress and eventually lead to cirrhosis as well. It is most commonly associated with inflammatory bowel disease. From a surveillance standpoint, PSC may be complicated by disease-associated malignancies, including cholangiocarcinoma, hepatocellular carcinoma, and pancreatic cancer. Thus, follow-up imaging in this setting is generally via MRCP +/- MRI Abdomen (CPT® 74181 or CPT® 74183) – See: **Primary Sclerosing Cholangitis (PSC) (AB-23.4)**.

PBC is a complex, chronic, and slowly progressive autoimmune liver disease that predominately affects women, and is characterized by cholestatic liver biochemistries as well as the presence of AMA (Anti-Mitochondrial Antibodies), and results in T-lymphocyte-mediated destruction of *small* intrahepatic bile ducts. This may ultimately lead to cirrhosis, and thus an increased risk of hepatocellular carcinoma. Because of this, surveillance via US screening protocols for HCC are followed in PBC.

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It may be necessary, when the diagnosis of PBC is uncertain, for an MRCP to be performed in order to distinguish between PBC and PSC. However, MRI or MRCP is not used for serial monitoring for PBC, once the diagnosis is established. This is in contradistinction to PSC, in which MRCP is used to surveil for cholangiocarcinoma, as discussed above.

Evidence Discussion

Ultrasound has several advantages over advanced imaging techniques such as computed tomography (CT) and magnetic resonance imaging (MRI). Ultrasound requires no ionizing radiation, is readily available, cost-effective, and often allows for same-day scheduling. The reproducibility of results has made it the initial modality of choice for imaging hepatobiliary conditions and screening for hepatocellular carcinoma (HCC) for the past 20 years. Ultrasound also helps to determine the next appropriate advanced imaging study - whether CT, MRI, or magnetic resonance cholangiopancreatography (MRCP) - along with contrast levels.[19,21,24,25]

Disadvantages include image quality degradation due to bowel gas, challenges in acquiring an acoustic window, obesity, and sonographer inexperience.[19,21,24,25]

Although emerging data may support CT and MRI-based liver surveillance, AASLD does not currently recommend their routine use in individuals at risk for HCC. Studies from Asia suggest that both two-phase CT and hepatobiliary contrast-enhanced MRI are more sensitive for early-stage HCC detection compared to US-based surveillance, with sensitivities of 83% and 86% versus 28%–29%, respectively. However, neither CT nor MRI has been validated in Western patient cohorts without chronic viral hepatitis B. Additionally, CT-based surveillance raises concerns about radiation and contrast exposure, especially if conducted semiannually. Similarly, MRI contrast agents present concerns regarding radiology service capacity, patient acceptance, and cost-effectiveness.[19,21,24,25]

Relative to surveillance, AASLD acknowledges the suboptimal performance of CT or MRI in accurately diagnosing HCC in lesions <1cm. AASLD recommends observing individuals with sub-centimeter liver lesions on ultrasound by repeat short-interval surveillance using ultrasound and AFP in 3-6 months. Imaging by multiphase CT or contrast-enhanced MRI is advised for those with new or enlarging solid liver lesions >1 cm and individuals with unequivocally elevated AFP independent of ultrasound results.[19,21,24,25]

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Ascites (AB-26.2)

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- Abdominal ultrasound (CPT® 76700 or CPT® 76705) and/or Doppler (CPT® 93975) with diagnostic paracentesis required for all initial evaluations of ascites to determine the need for further or advanced imaging.
  - Further advanced imaging is determined by the nature of etiology of the ascites (e.g., portal hypertension secondary to cirrhosis, malignancy such as ovarian or pancreatic, heart failure, etc.).
- Peritoneal-venous shunt patency study (CPT® 78291) is considered medically necessary for evaluation of shunt patency and function in an individual with ascites.

Background and Supporting Information

- Guidance from the American Association for the Study of Liver Diseases (2021) indicates that the initial evaluation of individuals with ascites should include a medical history, physical examination, abdominal US with Doppler, lab studies including CBC, Liver function tests, serum and urine electrolytes and paracentesis with ascitic fluid analysis, which then guides further management. They specifically note that "A diagnostic paracentesis should be performed in all individuals with new-onset ascites that is accessible for sampling".

Evidence Discussion

According to AASLD guidance for ascites management, Doppler ultrasound is the preferred initial radiologic test. Ultrasound is highly sensitive for diagnosing ascites and does not expose individuals to radiation.[15]

Depending on the analysis of the ascitic fluid, further imaging such as CT (to evaluate for malignancy or cirrhosis) or an echocardiogram (for heart failure) may be warranted.[3,12,13]

For individuals with refractory ascites and a LaVeen Shunt, a nuclear peritoneal-venous shunt study is the recommended imaging choice.[3,12,13]

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Portal Hypertension (AB-26.3)

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- In evaluating the cause of portal hypertension:
  - Abdominal US (CPT® 76700 or CPT® 76705) and/or Duplex Doppler US [CPT® 93975] of the liver and upper abdomen is medically necessary to distinguish ANY of the following:
    - pre-hepatic causes (including portal vein thrombosis, extrinsic compression from a tumor)
    - intrahepatic causes (including cirrhosis)
    - post-hepatic (including hepatic vein thrombosis)
- CT Abdomen with contrast (CPT® 74160) or MRI Abdomen with contrast (CPT® 74183) is medically necessary for ANY of the following indications:
  - Ultrasound is technically limited in assessing hepatic and/or portal vessels
  - Suspicion of portal vein thrombosis, cavernous transformation, or extrinsic impression, but ultrasound was indeterminate
  - Portal hypertension along with secondary signs are present (including splenomegaly or porto-systemic collaterals)
- For TIPS (transjugular intrahepatic portosystemic shunt)
  - See: **Hepatic Arteries and Veins (AB-43.1)**

Background and Supporting Information

- Causes of portal hypertension can be divided into pre-hepatic (e.g., portal vein thrombosis, extrinsic compression from a tumor), intrahepatic (e.g. cirrhosis) and post-hepatic (e.g., hepatic vein thrombosis).
- Most cases of portal hypertension are caused by cirrhosis. Cirrhosis is a histologic diagnosis. The term compensated advanced chronic liver disease (cACLD) encompasses a spectrum of fibrosis and cirrhosis that may be difficult to distinguish on clinical grounds alone since cACLD individuals may manifest with few signs or symptoms.
- When cirrhosis progresses to "decompensated advanced chronic liver disease" (dACLD) individuals may present with ascites, encephalopathy, varices. The most feared complication of portal hypertension is the development of gastro-esophageal varices with hemorrhage
- The gold standard for the assessment of portal hypertension is the Hepatic Venous Pressure Gradient (HPVG [pressure gradient between portal vein and the inferior vena cava]), which is an invasive test. This is the pressure gradient between portal vein and the inferior vena cava. Clinically significant portal hypertension (CSPH) is defined by HPVG of > 10 mm Hg. Measurement of HPVG requires invasive testing.

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There is however a non-invasive test alternative to guide imaging and to risk stratify between CSPH and cACLD. This test combines the Platelet Count with measurement of liver stiffness (LSM), typically from transient elastography (e.g., FibroScan). Risk is best summarized by the "Rule of Five."

- The "Rule of Five," thresholds are as follows:
  - LSM < 15 kPa and platelet count > 150,000/mm³: Low risk of CSPH — endoscopy can be avoided.
  - LSM between 15–25 kPa or platelet count < 150,000/mm³: Intermediate risk — further evaluation or endoscopy may be needed.
  - LSM ≥ 25 kPa: High likelihood of CSPH — endoscopy is recommended.

Evidence Discussion

Initial evaluation of individuals suspected of portal hypertension (PH) should always include a detailed history and physical exam, as well as appropriate lab studies and non-invasive assessment of liver parenchyma by ultrasound. Valuable information is added about alterations in hepatic vascular flow per Doppler ultrasound.[27,28]

In cases of uncertainty, abdominal CT scan or MRI may be warranted. Individual specific factors however should be herein considered such as pregnancy, tolerance to long image acquisition time and confined spaces, high cost, as well as exposure to contrast agents and ionizing. [21,22,23]

Surrogate markers of clinically significant portal hypertension (CSPH) include the presence of gastroesophageal varices or portosystemic collaterals. When such markers are not apparent, clarification may require invasive studies such as liver biopsy or hepatic venous pressure gradient (HVPG) measurement.[27]

However, non-invasive studies have emerged as preferred assessment tools when cACLD is suspected. Transient elastography (TE) correlates liver stiffness (LSM) with the degree of liver fibrosis. Patients with LSM 5-10 kPa do not have cACLD and can be monitored for liver disease progression by serial LSM every 2-3 years.[27,28]

More recent refinement of non-invasive risk assessment of CSPH combines LSM and platelet count. Individuals at low risk of gastro-esophageal varices can be monitored annually. For individuals with higher risk for cACLD or CSPH the "Rule of Five" guides further decision-making.

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Monitoring After Fontan Procedure (AB-26.4)

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- Abdominal ultrasound (CPT® 76700 or CPT® 76705) and Doppler (CPT® 93975) are medically necessary every 6 months or per institution protocol.
- MR Elastography (CPT® 76391) is medically necessary every 6 months
- If any sized lesions are detected on ultrasound:
  - MRI Abdomen without contrast, or without and with contrast (CPT® 74181 or CPT® 74183) with follow-up timeframes as requested.
- If advanced fibrosis or cirrhosis is detected on any imaging modality:
  - HCC monitoring every 6 months after advanced fibrosis or cirrhosis is detected with MRI Abdomen without contrast, or without and with contrast (CPT® 74181 or CPT® 74183) is medically necessary.
- CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen with contrast (CPT® 74160), or other elastography techniques (i.e., Fibroscan) can be used to assess and monitor individuals with contraindications to MRI (e.g., pacemaker devices, etc.).

Evidence Discussion

Individuals with single-ventricle physiology who have undergone the Fontan Procedure which redirects venous blood flow to the pulmonary circulation invariably develop liver complications, which can include the development of nodules and cirrhosis secondary to the altered vascular anatomy, and thus are at risk for hepatocellular carcinoma. In addition, the congestive hepatopathy associated with the Fontan procedure makes differentiation of focal liver lesions from congestive changes more challenging than other cirrhotic conditions. Thus, most institutions use MRI rather than US for monitoring in the setting of cirrhosis. In addition, the evaluation for HCC is challenging due to the vascular changes associated with the Fontan procedure, because the typical HCC pattern of delayed venous-phase contrast washout may not be appreciated within the background congestive hepatopathy. Thus, biopsy is usually required. Also, distinguishing dysplastic lesions from true HCC based on LiRADS criteria is very challenging as well. There are no current society endorsed guidelines, and institutions may vary in the monitoring of chronic liver disease in this patient population. The above algorithm represents an accepted approach and is consistent with the consensus from the Fontan-Associated Liver Disease proceedings from the American College of Cardiology Shareholders Meeting (2015) as well as the consensus of a multidisciplinary group of American Society of Transplantation members (2020).[2,7,8,15]

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MR Cholangiopancreatography (MRCP) (AB-27)

Guideline

MRCP (AB-27.1)
References (AB-27)

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MRCP (AB-27.1)

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- MRCP (Magnetic Resonance Cholangio Pancreatography) is a non-invasive imaging procedure, which is used to visualize the biliary and pancreatic ductal system. It is used most often in the following circumstances:
  - Suspected gallstone pancreatitis (See: **Pancreatitis (AB-33)**)
  - Suspected biliary pain (See: **Right Upper Quadrant and Epigastric Pain (AB-2.3)**
  - Pancreatic cyst and pseudocyst evaluation (See: **Pancreatic Lesion (AB-31)**, and **Pancreatitis (AB-33)**)
  - Choledochal cyst surveillance (See: **Biliary Disease PEDAB-16.2**)
  - Evaluation of abnormal liver chemistries (See: **Abnormal Liver Chemistries (AB-30.1)**)
  - Evaluation of the pancreas secondary to abdominal trauma with suspected duct injury or pseudocyst
  - Recurrent pancreatitis of unknown etiology (See: **Pancreatitis (AB-33)**)
  - Evaluation and follow-up of Primary Sclerosing Cholangitis (See: **Primary Sclerosing Cholangitis (PSC) (AB-23.4)**)
  - Evaluation of jaundice (See: **Abnormal Liver Chemistries (AB-30.1)**)
  - Evaluation of congenital anomalies of the cystic and hepatic ducts
  - Post-surgical biliary anatomy and complications (See: **Liver Transplant, Post-Transplant Imaging (AB-42.3)**)
  - For further evaluation of ultrasound or CT findings of abnormally dilated biliary duct, dilated pancreatic duct, or enlargement or fullness of the pancreas.
- Code assignment for MRCP
  - In general, there is no specific CPT code to describe MRCP. To report an MRCP, one of the MRI Abdomen codes should be selected, depending on contrast needs (CPT® 74181, CPT® 74182, or CPT® 74183). There is also a level II HCPCS code for MCRP, S8037. Simultaneous billing of any of these codes is redundant and unnecessary.
  - Reporting or billing a *second* MRI code to represent the “MRCP portion” of the study is not supported. When this occurs, it is usually seen as two simultaneous MRI requests, an MRI Abdomen without and with contrast (CPT® 74183) AND an additional MRI Abdomen without contrast (CPT® 74181). This second MRI code, as noted, is not supported. Both the primary MRI Abdomen AND the MRCP portion of the study are covered by the single MRI Abdomen code (CPT® 74183).

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- Requests for 3D rendering (either CPT® 76376 or CPT® 76377) are approvable, if requested, in addition to the primary MRI Abdomen code (CPT® 74181, CPT® 74182, or CPT® 74183).

Evidence Discussion

Magnetic Resonance Cholangiopancreatography (MRCP) is the preferred imaging modality for assessing the biliary and pancreatic systems, offering soft tissue contrast resolution without ionizing radiation exposure. Literature highlights MRCP's high sensitivity and specificity in detecting various hepatobiliary pathologies, including choledocholithiasis, cholangitis, pancreatitis and pancreatic neoplasms. Moreover, MRCP provides detailed visualization of the pancreatic duct and biliary tree, facilitating accurate diagnosis and surgical planning. While ERCP is the gold standard for visualization of pancreaticobiliary ducts and provides opportunity for therapeutic intervention, MRCP is a non-invasive method that has gained wide acceptance for diagnostic evaluation.[1,2,4]

Limitations around MRCP include its slower acquisition time with associated higher sensitivity to motion artifact, potential need for sedation, contraindications related to ferrous magnetic implants or foreign bodies, and relatively higher cost compared to alternate options, such as ultrasound or CT. Accessibility could also be an issue, potentially leading to diagnostic delays in some healthcare settings. Safety concerns mainly revolve around gadolinium-based contrast agents, particularly in individuals with compromised renal function.[1,2,4]

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Gallbladder (AB-28)

Guideline

Gallbladder (AB-28.1)
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Gallbladder (AB-28.1)

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- Findings on ultrasound or EUS suspicious for malignancy:
  - CT Abdomen with or without and with contrast (CPT® 74160 or CPT® 74170)
- Findings on ultrasound inconclusive for adenomyomatosis:
  - Contrast-Enhanced US (CEUS, CPT® 76978, CPT® 76979)
    - If US and CEUS are inconclusive for adenomyomatosis:
      - MRI Abdomen without and with contrast (CPT® 74183)
- For confirmed gallbladder malignancy:
  - See **Gallbladder and Biliary Tumors - Initial Work-up/Staging (ONC-14.6)** in the Oncology Imaging Guidelines

Gallbladder Polyps

- Individuals at increased risk for gallbladder malignancy (if surgery not chosen):
  - Age >50
  - Primary Sclerosing Cholangitis
  - Indian ethnicity
  - Sessile polyp or gallbladder wall thickening >4mm
- Increased risk for gallbladder malignancy:
  - Polyp <6 mm
    - Ultrasound at 6 months, then yearly for 5 years
  - Polyp 6-9 mm (If cholecystectomy is not chosen)
    - Ultrasound at 6 months, then yearly for 5 years
- No increased risk for gallbladder malignancy:
  - Polyp <6 mm
    - Ultrasound at 1, 3, and 5 years
  - Polyp 6-9 mm
    - Ultrasound at 6 months, and then yearly for 5 years
- Gallbladder polyp ≥10 mm:
  - Surgery recommended. If surgery not performed, follow guidelines for increased risk of gallbladder malignancy as noted above.
- Alternative Imaging:
  - Endoscopic ultrasound (EUS) may provide additional information in the diagnosis of gallbladder polyps. There is insufficient data that advanced imaging (CT or MRI) should be used ahead of conventional ultrasound in the investigation of gallbladder polyps.

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Evidence Discussion

Transabdominal ultrasound is the preferred modality for surveillance of polyps, aiming for stability at the 5-year mark as an endpoint. There is insufficient data that advanced imaging (CT or MRI) should be used ahead of conventional ultrasound in the investigation of gallbladder polyps.[1]

Cholecystectomy is recommended for symptomatic individuals, lesions that increase by more than 2 mm in size, and polypoid lesions in individuals who are considered high risk.[8]

There is no role for CT, MRI, or endoscopic ultrasound in the surveillance of polypoid lesions of the gallbladder. However, advanced imaging is useful in evaluation of ultrasound findings that are suspicious for malignancy. CT can help to demonstrate any bile duct dilation as well as assist in staging, planning, and management of any found malignancy.[4]

Ultrasound is also the preferred modality for gallbladder adenomyomatosis. Bonatti, et al. state "the use of high-frequency probes and a precise focal depth adjustment enable correct identification and characterization of GA in the majority of cases" (2017). MRI is reserved only for instances of suspected gallbladder adenomyomatosis when ultrasound techniques are inconclusive.[5]

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Liver Lesion Characterization (AB-29)

Guideline

Liver Lesion Characterization (AB-29.1)
Fatty Liver (Metabolic Associated Steatotic Liver Disease (MASLD), formerly known as NAFLD) (AB-29.2)
Polycystic Liver Disease (AB-29.3)
Isolated or Incidental Hepatomegaly (AB-29.4)
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Liver Lesion Characterization (AB-29.1)

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Note:

Advanced imaging approvals in this section refers to MRI Abdomen without and with contrast (CPT® 74183), CT Abdomen with contrast (CPT® 74160), CT Abdomen without and with contrast (CPT® 74170) and Contrast-Enhanced Ultrasound (CPT® 76978-initial lesion, CPT® 76979-additional lesions). In the following section, if only CT Abdomen with contrast (CPT® 74160) is noted as the appropriate study, it is because the American College of Radiology has determined that a prior without contrast study does not provide any added benefit. It should also be noted that a standard "triple-phase CT" liver does not involve a prior without contrast study (See: CT Imaging (AB-1.2))

Indeterminate Liver Lesions

- Indeterminate liver lesion discovered on US:
  - Indeterminate Liver Lesion ≥1cm on initial imaging
    - No suspicion or evidence of extrahepatic malignancy or underlying liver disease:
      - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160) or Contrast-Enhanced US (CEUS, CPT® 76978, CPT® 76979)
    - Known history of an extrahepatic malignancy:
      - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast or without and with contrast (CPT® 74160 or CPT® 74170)
    - Known history of chronic liver disease:
      - See: **Chronic Liver Disease, Cirrhosis, and Screening for HCC (AB-26.1)**
  - Indeterminate Liver Lesion <1cm on initial imaging
    - Known underlying chronic liver disease
      - See: **Chronic Liver Disease, Cirrhosis, and Screening for HCC (AB-26.1)**
    - Known history of an extrahepatic malignancy:
      - MRI Abdomen without and with contrast (CPT® 74183) is the preferred study.
      - Contrast-Enhanced US (CPT® 76978, CPT® 76979) is appropriate.
- Indeterminate liver lesion discovered on non-contrast CT or non-contrast MRI
  - Liver lesion on initial imaging
    - No suspicion or evidence of extrahepatic malignancy or underlying liver disease
      - Multiphase CT Abdomen with contrast (CPT® 74160), MRI Abdomen without and with contrast (CPT® 74183), or CEUS (CPT® 76978 and/or CPT® 76979)

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    - Known history of extrahepatic malignancy:
        - MRI Abdomen without and with contrast (CPT® 74183), CT Abdomen with contrast or without and with contrast (CPT® 74160 or CPT® 74170), or CEUS (CPT® 76978 or CPT® 76979)
    - Known chronic liver disease:
        - See: **Chronic Liver Disease, Cirrhosis, and Screening for HCC (AB-26.1)**
- Additional scenarios and follow-up imaging for an Indeterminate lesion in an individual based on risk factors:
    - Low risk individuals defined as:
        - No known primary malignancy
        - No hepatic dysfunction (abnormal liver tests)
        - No underlying chronic liver disease
        - No history of alcoholism, sclerosing cholangitis, choledochal cysts, hemochromatosis, or anabolic steroid use
    - High risk individuals would have ONE or MORE of the above conditions.
    - Indeterminate lesion <1cm on US, CT, or MRI, **low-risk** individual and no suspicious imaging features noted on the study
        - No further imaging
    - Indeterminate lesion <1cm in **high-risk** individuals on US, CT, or unenhanced MRI not specifically dealt with in the above guidelines:
        - MRI Abdomen without and with contrast (CPT® 74183)
        - If, after MRI, the lesion remains indeterminate or not fully characterized
            - See: **Liver Metastases (ONC-31.2)** or malignancy-specific guidelines in the Oncology Imaging Guidelines.
            - If biopsy cannot be performed, follow-up MRI can be obtained in 3-6 months. Additional imaging in this setting can be considered on an individual basis. This timeframe would also apply if the lesion is indeterminate and an MRI with Eovist is requested for further evaluation in this setting.
    - Most lesions ≥1cm can be categorized by MRI or histology. For lesions which have been categorized, regardless of size, see below.

Focal Liver Lesions

- For the imaging of specific focal liver lesions:
    - Suspected hepatic adenoma:
        - MRI is considered the best technique for characterization. Follow-up imaging can be CT Abdomen (CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) every 6 months for 2 years, and then annually, to establish any growth patterns and assess for malignant transformation.
    - Hepatic Hemangioma (if not completely characterized on initial CT without a liver protocol):

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        - Multiphase CT Abdomen (CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183)
        - Follow-up imaging is medically necessary as follows:
            - In individuals with cirrhosis or chronic hepatitis B, continued imaging with multiphase CT Abdomen (CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) every 3-6 months for one year.
                - For continued HCC surveillance after one year, see also: **Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)**
            - Giant hemangiomas (>4cm) can be followed by limited abdominal US in 6-12 months. If no change in size, no further follow-up is indicated, unless it becomes symptomatic.
            - See below for pre-operative considerations
    - Focal Nodular Hyperplasia (FNH):
        - MRI Abdomen (CPT® 74183) or CT Abdomen (CPT® 74160 or CPT® 74170) to confirm a diagnosis of FNH. The use of Eovist contrast is often diagnostic in differentiating FNH from other lesions seen on MRI or CT.
        - Additional follow-up is annual US for 2 to 3 years in women diagnosed with FNH who are continuing to use oral contraceptives. Follow-up with CT (CPT® 74160 or CPT® 74170) or MRI (CPT® 74183) can be done if the lesion is not adequately visualized on US.
    - Hepatic cysts:
        - Asymptomatic, simple cysts do not require additional follow-up.
        - For complicated cysts (US shows internal septations, fenestrations, calcifications, irregular walls, as well as the presence of daughter cysts):
            - CT Abdomen (CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) can be performed

- **Additional indications for advanced imaging (MRI Abdomen or CT Abdomen)**:
    - If documented that a percutaneous liver biopsy is to be considered if imaging is atypical or inconclusive.
    - Fatty liver (hepatic steatosis) on US with a focal liver lesion.
    - **If there is a technical limitation to US (e.g. marked heterogeneity, or other specifically noted technical limitations of US such as obscuration by intestinal gas, chest wall deformity, etc.)
    - For suspected liver metastases, see: **Liver Metastases (ONC-31.2)** in the Oncology Imaging Guidelines
- Preoperative studies for individuals with large hemangiomas or adenomas considered for resection:
    - MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) can be considered
- For Indeterminate Lesions ≥1cm in categories for which defined guidelines do not exist (i.e., underlying chronic liver disease, **Chronic Liver Disease, Cirrhosis,**

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and Screening for HCC (AB-26.1), underlying malignancy, **Liver Metastases (ONC-31.2)** or the specific malignancy in the Oncology Imaging Guidelines, hepatic adenoma, etc.) a biopsy should be considered when the findings from advanced imaging are inconclusive. In clinical situations when a biopsy cannot be performed (such as a medical contraindication or a liver transplant candidate due to the risk of needle-tract seeding), or is inconclusive, a short-term surveillance MRI can be performed in 3-4 months to monitor lesion stability.

- This can be repeated every 6 months, as necessary in this scenario. This timeframe would also apply if an MRI with Eovist is requested for short-term follow-up of an indeterminate lesion imaged on MRI Abdomen without and with contrast performed with other contrast, such as gadolinium. An exception would be if the differential is between FNH vs. hepatic adenoma or other benign lesions. FNH follow-up is yearly, and hepatic adenoma would require a 6 month follow-up study; if the differential of the lesion is between FNH and hepatic adenoma, then the follow-up study should be 6 months.
- Nuclear Medicine imaging of the Liver (CPT® 78201, CPT® 78202, CPT® 78803, CPT® 78215, CPT® 78216, or CPT® 78830) are rarely performed, but can be considered medically necessary when US, CT, and MRI are unavailable or contraindicated for:
  - evaluation of liver mass, trauma, or suspected focal nodular hyperplasia (FNH)
  - differentiation of hepatic hemangioma from FNH
  - diffuse hepatic disease or elevated liver function tests

Evidence Discussion

For further characterization of a liver lesion seen on other imaging, CT offers high spatial resolution and rapid image acquisition, making it suitable for initial characterization of liver lesions. CT can be highly accurate in establishing whether or not a liver lesion is benign.[1,2] CT Abdomen is generally not the appropriate study in liver lesions < 1 cm with a known history of an extrahepatic malignancy due to the resolution of CT does not allow for definitive characterization of lesions <1cm.

MRI provides superior soft tissue contrast and multi-parametric capabilities, facilitating further tissue characterization when needed (particularly small lesions). Nonetheless, the use of gadolinium-based contrast agents in MRI poses safety concerns, including the risk of nephrogenic systemic fibrosis (NSF) in individuals with impaired renal function. For individuals with a history of malignancy outside the liver, MRI is more accurate at differentiating between benign and malignant lesions. Thus, CT is not recommended over MRI in this scenario.[1,2]

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Fatty Liver (Metabolic Associated Steatotic Liver Disease (MASLD), formerly known as NAFLD) (AB-29.2)

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- Fatty liver (hepatic steatosis) incidentally discovered on imaging (US/CT/MRI) or suspected:
  - Magnetic Resonance Elastography (MRE) (CPT® 76391) may be medically necessary if criteria are met, see: **Liver Elastography (AB-45)**
  - Magnetic Resonance-Protein Density Fat Fraction (MRI-PDFF, usually requested as CPT® 74181 or 74183), MR Spectroscopy (MR-S, CPT® 76390), and the multiparametric MRI referred to as Liver Multiscan (LMS, Category III CPT® code 0648T or 0649T) for evaluation of fatty liver disease:
    - With regards to the above procedures, their main current utility is in assessing response to therapy in clinical trials. Their role in clinical practice, or with what frequency one would image, has not been defined. In view of this, they are experimental and investigational at this time.
  - HCC Screening for Fatty Liver with cirrhosis or advanced fibrosis:
    - See: **Chronic Liver Disease, Cirrhosis, and Screening for HCC (AB-26.1)**
  - MRI or CT for the further evaluation of incidentally discovered fatty liver on US, in the absence of a specific finding needing further characterization such as a nodule, is generally not medically necessary. See: **Liver Lesion Characterization and Additional Indications for Advanced Imaging (AB-29.1)**. In addition, the finding of fatty liver alone on CT with contrast does not require MRI for confirmation.
  - Requests for imaging studies to screen individuals at high-risk for MASLD (formerly known as NAFLD) (e.g., diabetes or obesity) or for screening family members of individuals with MASLD is not approvable at this time.

Evidence Discussion

Fatty liver is often detected incidentally by ultrasound, CT, or MRI performed for other indications. Fat detected in the liver may have many causes including medications, starvation, excessive alcohol intake, other chronic medical illnesses, and metabolic syndrome. Non-Alcoholic Fatty Liver Disease (NAFLD), now known as Metabolic Dysfunction-Associated Steatotic Liver Disease (MASLD), is the most common cause of steatotic (fatty) liver. NALFD (used throughout henceforth) can often lead to serious liver injury (non-alcoholic steatohepatitis: NASH) and complications of cirrhosis. Therefore,

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monitoring using additional imaging modalities may be medically necessary, in addition to other non-invasive tests.[32]

For those individuals where fatty liver is incidentally discovered on imaging (US/CT/MRI) or in conditions where NAFLD is suspected, Magnetic Resonance Elastography (MRE) may be medically necessary.[32]

Other procedures, such as Magnetic Resonance-Protein Density Fat Fraction, MR Spectroscopy, and the multiparametric MRI referred to as Liver Multiscan may be ordered for evaluation of fatty liver disease but their main current utility is in assessing response to therapy in clinical trials and are considered investigational.[32]

Requests for imaging studies to screen individuals at high-risk for NAFLD (e.g., diabetes or obesity) or for screening family members of individuals with NAFLD is not medically necessary at this time.[3, 32]

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Polycystic Liver Disease (AB-29.3)

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- Polycystic Liver Disease
  - Defined as >20 cysts, or the presence of cysts occupying one-half the volume of the hepatic parenchyma.
  - Most commonly seen as an extra-renal manifestation of Autosomal Dominant Polycystic Kidney Disease, though may occur as Autosomal Dominant Polycystic Liver Disease.
  - Imaging:
    - For prognostication purposes MRI Abdomen (CPT® 74183) or CT Abdomen (CPT® 74160 or CPT® 74170) can be performed initially to assess liver volume.
    - Surveillance imaging of asymptomatic individuals is not medically necessary.
    - Suspected complications such as cyst rupture or hemorrhage (manifested by acute pain in the upper abdomen):
      - MRI Abdomen (CPT® 74183) or CT Abdomen (CPT® 74160 or CPT® 74170)

Evidence Discussion

Ultrasonography is the first step in diagnosing polycystic liver disease (PLD). Abdominal ultrasound to screen for PLD should be offered to all individuals diagnosed with autosomal dominant polycystic kidney disease (ADPKD). Imaging follow up is not routinely medically necessary in asymptomatic individuals. CT Abdomen or MRI Abdomen may be medically necessary in symptomatic individuals to assess the extent of PLD/cyst burden and to assess the liver volume. MRI or CT can be used in PLD to evaluate the distribution of cysts within the liver parenchyma and the relation to hepatic vasculature. Ultrasound or MRI Abdomen may medically necessary to diagnose cyst hemorrhage, when suspected. CT Abdomen is not medically necessary to diagnose cyst hemorrhage. CT may detect gas or calcification but is less accurate for assessing cyst contents. There is no need to screen family members of individuals with PLD for the presence of hepatic cysts unless symptoms are present. Screening for intracranial aneurysms is not medically necessary for individuals with PCLD. Routine post treatment imaging is not medically necessary.[33-38]

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Isolated or Incidental Hepatomegaly (AB-29.4)

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- Initial imaging of hepatomegaly discovered or suspected on physical examination:
  - US Abdomen (CPT® 76700 or CPT® 76705) and Duplex (CPT® 93975 or CPT® 93976)
- Further evaluation of abnormalities on initial ultrasound that require further characterization:
  - Refer to specific guidelines for the abnormality detected on US
    - Fatty liver (liver steatosis), see: **Fatty Liver (Metabolic Associated Steatotic Liver Disease (MASLD), formerly known as NAFLD) (AB-29.2)**
    - Hepatic lesion, see: **Liver Lesion Characterization (AB-29.1)**
- Hepatomegaly discovered on ultrasound and no indeterminate abnormalities:
  - Medical workup, including lab studies such as liver tests, and history and physical should be performed to assess for suspected underlying disease (e.g. infiltrative disease such as amyloid, lymphoma, etc.)
    - Lab abnormalities and/or symptoms of a specific disease process should follow imaging studies outlined in the guideline for that disease process.
    - Advanced imaging in the absence of symptoms or lab abnormalities indicative of an underlying disorder is not medically necessary.

Background and Supporting Information

As noted by the AASLD "…imaging tests, such as ultrasound, computed tomography (CT), and MR, do not reliably reflect the spectrum of liver histology in individuals with NAFLD." In addition, "MR imaging, either by spectroscopy or by proton density fat fraction is an excellent noninvasive modality for quantifying hepatic fat and is being widely used in NAFLD clinical trials…..However, the utility of noninvasively quantifying HS (hepatic steatosis) in individuals with NAFLD in routine clinical care is limited."[3]

Evidence Discussion

Hepatomegaly (enlarged liver) can be detected by physical exam and imaging studies, such as ultrasound, CT, MRI and nuclear medicine studies. An enlarged or palpable liver does not always indicate primary liver disease, so advanced imaging should be directed by history, other physical findings and laboratory results.[1,3,14,21]

An enlarged liver can be caused by[1,3,14,21]:

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- Primary liver disease (hepatitis, alcoholic liver disease, NAFLD (non-alcoholic fatty liver disease), other causes of liver inflammation
- Metastatic or primary liver tumors
- Infiltrative disease (such as amyloidosis, infiltrative lymphoma)
- Impaired venous outflow (such as right heart failure, Budd-Chiari syndrome)
- Storage disorders (such as Gaucher Disease, Alpha-1 antitrypsin deficiency)
- Polycystic liver disease
- Other less common causes

Initial imaging studies should be chosen based on history, physical exam, laboratory studies and prior imaging studies. Usually, ultrasound of the abdomen and/or duplex scan would be the initial tests. Advanced imaging, such as CT or MRI are likely to be considered medically necessary based on findings based on specific guidelines based on the abnormality detected on ultrasound.[1,3,14,21]

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References (AB-29)

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Abnormal Liver Chemistries (AB-30)

Guideline

Abnormal Liver Chemistries (AB-30.1)
References (AB-30)

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Abnormal Liver Chemistries (AB-30.1)

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Elevated AST and/or ALT (>33 IU/l for males [assigned at birth], >25 IU/l for females [assigned at birth]) and other LFTs are normal or Hepatocellular pattern of elevation (AST and ALT disproportionately elevated to ALKP):

- <2X normal:
  - Repeat lab after 3 weeks and discontinuation of medications associated with elevated LFTs (such as statins, niacin, sulfa, rifampin, tetracycline, estrogen) if applicable.
  - If LFTs remain elevated: Abdominal US (CPT® 76700 or CPT® 76705)
    - Above studies do not explain the cause of the elevated transaminases AND HAV IgG, HBsAg, HBcAb, HBsAb, HCV Ab, iron panel (may include ferritin, serum iron, iron-binding capacity, or transferrin saturation) have been performed and are inconclusive:
      - CT Abdomen with contrast (CPT® 74160)
- 2 to 15X normal:
  - Abdominal US (CPT® 76700 or CPT® 76705)
    - Above studies do not explain the cause of the elevated transaminases AND HAV IgG, HBsAg, HBcAb, HBsAb, HCV Ab, iron panel (may include ferritin, serum iron, iron-binding capacity, or transferrin saturation) have been performed and are inconclusive:
      - CT Abdomen with contrast (CPT® 74160)
- >15X normal:
  - Abdominal US with Doppler (CPT® 76700 or CPT® 76705 and CPT® 93975) OR
  - CT Abdomen with contrast (CPT® 74160) OR
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
    - Above studies do not explain the cause of the elevated transaminases AND HAV IgG, HBsAg, HBcAb, HBsAb, HCV Ab, iron panel (may include ferritin, serum iron, iron-binding capacity, or transferrin saturation) have been performed and are inconclusive:
      - MRI Abdomen without and with contrast (CPT® 74183) and/or MRCP (CPT® 74181)

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- If the findings suggest chronic liver disease, see: **Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)**
- If the findings suggest hemochromatosis, see: **Hereditary (Primary) Hemochromatosis (HH) and Other Iron Storage Disease (AB-11.2)**

Elevated alkaline phosphatase level (or GGT), and other LFTs are normal or Cholestatic pattern of elevation (ALKP elevated disproportionately to AST and ALT)

- If isolated ALKP elevation, GGT should be obtained for confirmation of hepatic etiology, prior to imaging.
- If ALKP is elevated with other LFTs, no confirmatory test is necessary.
    - Confirmed hepatic etiology of elevated ALKP:
        - Abdominal or RUQ ultrasound (CPT® 76700 or CPT® 76705)
            - Dilated biliary ducts on US:
                - MRCP
        - No dilated biliary ducts on US:
            - Anti-mitochondrial antibody (AMA) should be checked prior to advanced imaging.
                - If AMA is negative, and ALKP >2X ULN:
                    - MRCP
                - If AMA is negative, and ALKP 1 to 2X ULN:
                    - observe for 6 months
                    - if ALKP remains elevated after 6 months: MRCP
- CT Abdomen with contrast (CPT® 74160) if the above studies are unrevealing or individual cannot undergo MRCP.

Isolated elevated bilirubin (no other LFTs elevated)

- Elevation is unconjugated (elevated indirect bilirubin), and no other LFT elevations:
    - No advanced imaging
- Elevation is conjugated (elevated direct bilirubin)
    - RUQ ultrasound
        - Dilated biliary ducts on ultrasound:
            - MRCP
        - No dilated biliary ducts on US:

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- Anti-mitochondrial antibody (AMA) should be checked prior to advanced imaging
  - AMA negative and elevation persists or is unexplained:
    - MRCP or liver biopsy

- CT Abdomen with contrast (CPT® 74160) if the above studies are unremarkable or the individual cannot undergo MRCP.

Clinical jaundice, no known predisposing condition

- Abdominal ultrasound (CPT® 76700 or CPT® 76705)
  - For further imaging, follow guideline for elevated bilirubin
- Clinical jaundice, suspected mechanical obstruction based on clinical condition or laboratory values (e.g., known choledocholithiasis, acute and chronic pancreatitis, suspected stricture from a recent invasive procedure, previous biliary surgery, suspected tumor):
  - CT Abdomen with contrast (CPT® 74160) or MRI and/or MRCP (CPT® 74183 or CPT® 74181)
- US findings suggesting mechanical biliary obstruction, non- diagnostic or technically limited US (e.g., large amounts of intestinal gas, obesity with BMI >35):
  - CT Abdomen with contrast (CPT® 74160) or MRI and/or MRCP (CPT® 74183 or CPT® 74181)

Additional considerations

- For individuals with elevated LFTs and suspicion of sclerosing cholangitis, such as those with IBD, see: **Primary Sclerosing Cholangitis (PSC) (AB-23.4)**.
- For individuals with elevated LFTs and history of underlying malignancy, please refer to the specific oncology guidelines, when appropriate.
- Requests for additional advanced imaging (CT, MRI, etc.) are based on the prior imaging results, as appropriate to the finding (for example, if a lesion is identified that needs further characterization, refer to liver lesion imaging as per **Liver Lesion Characterization (AB-29.1)**)
- For MRE indications, see: **Liver Elastography (AB-45)**

Background and Supporting Information

- The standard laboratory tests commonly referred to as “LFTs” include bilirubin, alkaline phosphatase (alkphos or ALKP), aspartate transaminase (AST), alanine transaminase (ALT), and gamma-glutamyl transferase (GGT).
- The major patterns of elevation which affect work-up are:
- Hepatocellular (AST and ALT disproportionately elevated to ALKP)

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- Cholestatic (ALKP elevated disproportionately to AST and ALT)
- Mixed pattern (ALKP, AST, and ALT all elevated)
- Isolated hyperbilirubinemia (elevated bilirubin and normal ALKP, ALT and AST)
- "R" Ratio
  - "R" Ratio: The so-called "R" ratio can be used to determine whether a pattern of multiple elevated liver chemistries is predominately cholestatic or hepatocellular in origin
  - R=(ALT/Upper limit of normal (ULN))/(ALKPH/ULN ALKPH)
    - If the "R" ratio:
      - >5 = hepatocellular
      - <2 = cholestatic
      - 2-5 = mixed pattern
  - For hepatocellular, use AST or ALT elevation guidelines
  - For cholestatic, use ALKPH elevation guidelines
  - Use ULN for ALT as noted above, and ULN for alkphos based on the individual lab report

Evidence Discussion

Liver blood tests look at how well the liver is functioning and can indicate whether there is any damage or inflammation inside the liver. Obtaining liver chemistries for both screening and diagnostic purposes are essential. When abnormalities are found they will frequently direct the provider to obtain further diagnostic testing including advanced imaging.[1,11]

A liver blood test looks at the chemicals (enzymes), proteins and other substances made by the liver to assess whether levels of any of these are abnormal. The major initial tests are for alanine transaminase, aspartate transaminase, alkaline phosphatase, and gamma-glutamyl transpeptidase.[1,11]

Repeating abnormal tests helps to confirm damage to the liver.[1,11]

The synthetic function of the liver can be assessed by evaluating levels of albumin and vitamin-dependent clotting factors.[1,11]

Iron storage, autoimmune, infectious, cholestatic, hepatocellular, drug induced, and other liver diseases are identified, followed, and diagnosed with the help of abnormal liver chemistries.[1,11]

Liver chemistries are an essential part of the non-invasive diagnosis and management of liver disease.[1,11]

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Pancreatic Lesion (AB-31)

Guideline

Pancreatic Cystic Lesions (AB-31.1)
Incidental Pancreatic Mass or Suspected Metastatic Disease to Pancreas (AB-31.2)
References (AB-31)

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Pancreatic Cystic Lesions (AB-31.1)

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- Pancreatic Cyst seen on Imaging-Initial Management:
  - MRI Abdomen (CPT® 74183) and/or MRCP are the tests of choice for initial evaluation.
    - Both MRI Abdomen and MRCP may be medically necessary, but only one CPT® 74183 should be used, not two.
  - CT Pancreatic protocol (CPT® 74160) or EUS are alternatives in individuals who are unable to undergo MRI.
  - Indeterminate cysts may benefit from a second imaging modality or EUS prior to proceeding with surveillance. MRI/MRCP can be approved to better characterize the lesion, without reference to the timeframe for follow-up imaging, if a previous US or CT Abdomen has been performed.
  - Radiographic diagnosis of a non-neoplastic cyst or classic features of a serous cystadenoma
    - No further imaging
  - If ANY of the following are present the individual should proceed to EUS + FNA and depending on findings, surgical consultation:
    - Main duct >5mm
    - Cyst ≥3cm
    - Change in main duct caliber with upstream atrophy
      - If EUS does not reveal findings of main duct involvement, patulous ampulla, cytology with high-grade dysplasia or pancreatic malignancy, or a mural nodule, then follow up MRI should performed in 6 months.
- Pancreatic Cyst Follow up Imaging
  - If high risk features (See below High Risk Considerations and Features) are not present, then the next follow-up imaging proceeds as follows:
    - Cyst <1cm: MRI in 2 years
    - Cyst 1-<2cm: MRI in 1 year
    - Cyst 2-3cm: if cyst is not clearly an Intraductal papillary mucinous neoplasms (IPMN) or Mucinous cystic neoplasm (MCN) then proceed with EUS. If it is an IPMN or MCN, then MRI at 6-12 months.
    - If the cyst is determined to be a serous cystadenoma, then no further evaluation unless symptomatic.
- Additional Surveillance for a presumed Intraductal papillary mucinous neoplasms (IPMN) or Mucinous cystic neoplasms (MCN) (imaging from time of presentation):
  - Cyst <1cm

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- MRI every 2 years for 4 years
- If stable after 4 years consider lengthening of interval imaging
- If increase in cyst size, then MRI or EUS in 6 months
- If stable, repeat again in 1 year and if stable return to MRI every 2 years

◦ Cyst 1-<2cm
- MRI yearly for 3 years
- If stable for 3 years, then change to MRI every 2 years for 4 years
- If stable after the additional 4 years, consider lengthening of interval for surveillance
- If increase in cyst size, repeat MRI in 6 months. If stable, repeat MRI in 1 year and if remains stable, resume original surveillance schedule.

◦ Cyst 2-<3cm
- MRI every 6-12 months for 3 years
- If stable after 3 years, change to MRI every year for 4 years
- If remains stable, consider lengthening of surveillance interval

◦ Cyst ≥3cm
- MRI alternating with EUS every 6 months for 3 years
- If stable for 3 years, increase interval to MRI alternating with EUS yearly for 4 years
- If remains stable, consider lengthening of surveillance interval
- If increase in cyst size, EUS + FNA

◦ Additional considerations
- Individuals with asymptomatic cysts that are diagnosed as pseudocysts on initial imaging and clinical history, or are determined to be serous cystadenomas, do not require further evaluation.
- Individuals who are not medically fit for surgery should not undergo further surveillance of incidentally found pancreatic cysts, irrespective of size.
- Surveillance should be discontinued if an individual is no longer a surgical candidate. However, follow-up imaging can be performed if requested for a symptomatic cyst (such as the development of jaundice secondary to cyst), in which palliative treatment might be available.
- High-Risk Considerations and Features
  - Individuals with IPMNs or MCNs with new onset or worsening diabetes
  - Rapid increase in cyst size (>3mm/year) during surveillance may have an increased risk of malignancy and should undergo a short-interval MRI or EUS.
  - Additional high-risk features which may prompt early evaluation are:
    - jaundice secondary to the cyst
    - acute pancreatitis secondary to the cyst

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- significantly elevated CA 19-9
- presence of a mural nodule or solid component either within the cyst or in the pancreatic parenchyma
- dilation of the main pancreatic duct >5mm
- focal dilation of the pancreatic duct concerning for main duct IPMN or an obstructing lesion
- IPMNs or MCNs measuring ≥3cm in diameter
- presence of high-grade dysplasia or pancreatic cancer on cytology. In this circumstance, imaging should be at the discretion of the provider.

- Post-op surveillance
  - Surgically resected serous cystadenomas, pseudocyst, or other benign cyst:
    - No additional imaging after resection
  - Surgically resected mucinous cystic neoplasms (MCNs) without an associated pancreatic malignancy (can have low, intermediate, or high-grade dysplasia):
    - No additional post-op surveillance
  - Surgically resected MCNs with invasive cancer:
    - Standard surveillance-based pancreatic cancer guidelines (See: **Pancreatic Cancer-Surveillance/Follow-up (ONC-13.5)** in the Oncology Imaging Guidelines) for 5 years. No surveillance required after 5 years.
  - Surgically resected IPMNs
    - IPMN with cancer
      - Pancreatic cancer surveillance guidelines (See: **Pancreatic Cancer-Surveillance/Follow-up (ONC-13.5)** in the Oncology Imaging Guidelines)
    - IPMN with high-grade dysplasia
      - MRI Abdomen (CPT® 74183) or EUS every 6 months
    - IPMN with low- or intermediate-grade dysplasia
      - MRI Abdomen (CPT® 74183) every 2 years
  - Surgically resected solid-pseudopapillary neoplasm with negative margins:
    - MRI Abdomen (CPT® 74183) yearly for 5 years
- See: **MR Cholangiopancreatography (MRCP) (AB-27)** for coding guidelines for MRCP.

Evidence Discussion

- Some pancreatic cystic lesions have malignant potential and need to be followed by either advanced imaging, endoscopic ultrasound, or both.[1,2,6]
- Advanced imaging includes MRI, MRCP, and CT imaging as these modalities are most effective in characterizing these lesions. MRI abdomen or MRCP are the initial studies of choice. The American Gastroenterological Association states, "MRI is the preferred surveillance imaging modality over computed tomography because MRI does not expose the individual to radiation and better demonstrates the structural

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relationship between the pancreatic duct and associated cyst. Also, MRI is less invasive than EUS". Thus, CT is reserved as an alternative for individuals who are unable to undergo MRI.[1]

- Follow-up imaging may or may not be medically necessary based on the nature of the cystic lesion, the size, or change in size of the lesion and how rapidly the size of the lesion changes. Smaller lesions with no concerning characteristics or changes undergo less surveillance due to the small absolute risk of malignancy. Concerning features such as rapid increase in size have increased risk of malignancy and therefore undergo more frequent or longer-term surveillance intervals.[1,2,6]

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Incidental Pancreatic Mass or Suspected Metastatic Disease to Pancreas (AB-31.2)

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- For the evaluation of incidental pancreatic mass or suspected metastatic disease to the pancreas, CT Abdomen with contrast with dual phase imaging (CPT® 74160), or MRI Abdomen without and with contrast (CPT® 74183).

Evidence Discussion

A pancreatic protocol CT involves scan acquisition during a parenchymal and portal venous phase, each of which are post-contrast administration. Dual phase, MDCT (multidetector CT) scans play a critical role in diagnosing and staging pancreatic cancers. MR and EUS can be used in groups of individuals where CT scan results are inconclusive in tumor localization and/or staging, particularly in vascular involvement.[7,8]

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Pancreatic Pseudocysts (AB-32)

Guideline

Pancreatic Pseudocysts (AB-32.1)

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Pancreatic Pseudocysts (AB-32.1)

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See: **Acute Pancreatitis (AB-33.1)** or **Chronic Pancreatitis (AB-33.2)**

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Pancreatitis (AB-33)

Guideline

Acute Pancreatitis (AB-33.1)
Chronic Pancreatitis (AB-33.2)
Exocrine Pancreatic Insufficiency (AB-33.3)
Asymptomatic Elevation of Pancreatic Enzymes (AB-33.4)
References (AB-33)

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Acute Pancreatitis (AB-33.1)

AB.PX.0033.1.A
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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- Imaging:
  - Initial imaging for suspicion of pancreatitis (typical symptoms, <48 to 72 hours, first-time presentation)
    - Abdominal ultrasound (CPT® 76700 or CPT® 76705)
      - Purpose is to establish the presence/absence of gallstones and biliary ductal dilation.
      - Doppler ultrasound (CPT® 93975) can be approved to assess vasculature, if requested.
    - If ultrasound or CT is performed and is nondiagnostic due to technical limitation (obesity, overlying gas, etc.):
      - MRI/MRCP (CPT® 74183 or CPT® 74181)
      - CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen with contrast (CPT® 74160) if ultrasound is nondiagnostic and MRI/MRCP cannot be performed.
    - In suspected acute biliary pancreatitis and/or cholangitis (dilated ducts or choledocholithiasis on ultrasound, elevated liver chemistries with a negative ultrasound, suspicion of cholangitis (classic triad is RUQ pain, fever, and jaundice))
      - MRI/MRCP (CPT® 74183 or CPT® 74181)
  - Initial imaging with atypical signs and symptoms when diagnoses other than pancreatitis are being considered (e.g., bowel perforation, bowel ischemia):
    - CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen with contrast (CPT® 74160)
    - MRI/MRCP* (CPT® 74181 or CPT® 74183) can be considered medically necessary for pregnant individuals (non-contrast), or those with renal insufficiency (without or without and with depending on request).
  - Follow-up imaging (late phase and thereafter):
    - Continued or worsening symptoms:
      - CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen with contrast (CPT® 74160) or MRI and/or MRCP (CPT® 74183 or CPT® 74181)
    - Follow-up of known pancreatic or peri-pancreatic fluid collections (including pseudocysts), to follow-up symptomatic collections, or for interventional planning:

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- MRI/MRCP (CPT® 74183 or CPT® 74181) or CT Abdomen and Pelvis (CPT® 74177)
  - If requested, CT Abdomen with contrast (CPT® 74160) or Abdominal ultrasound (CPT® 76705 or CPT® 76700) can be approved.

Note:

Frequency or intervals for additional follow-up is not defined and depends on clinical circumstances, response to therapy, etc.

- If, despite initial imaging, the etiology of the pancreatitis is still in doubt:
  - MRI/MRCP (CPT® 74183 or CPT® 74181) or CT Abdomen and Pelvis with (CPT® 74177)
    - If requested, CT Abdomen with contrast (CPT® 74160) can be approved.

- Acute recurrent pancreatitis
  - Abdominal ultrasound (CPT® 76705 or CPT® 76700)
  - MRI/MRCP (CPT® 74183 or CPT® 74181)
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
  - See: **Chronic Pancreatitis (AB-33.2)**

Background and Supporting Information

- While MRI/MRCP will give better evaluation of the pancreatic parenchyma as well as biliary and pancreatic ducts, it does NOT provide coverage and adequate evaluation of the bowel to assess alternative diagnoses such as bowel ischemia or perforation.
- Knowledge base:
  - Acute pancreatitis (2 of 3 of the following criteria):
    - Characteristic abdominal pain (typically epigastric or left upper quadrant pain with radiation to the back, chest, or flank)
    - Amylase or lipase > 3 times the upper limit of normal
    - Radiographic evidence of pancreatitis on cross-sectional imaging
  - Early Phase takes place in the first week
    - Goals of imaging:
      - Establish the correct diagnosis or provide an alternative diagnosis
      - Establish the etiology
      - Stage the morphologic severity
      - Assess for complications in individuals who deteriorate or fail to improve
  - Late phase can last weeks to months thereafter
    - Goals of imaging:
      - Monitor established pancreatic collections
      - Delineate the presence of symptomatic and asymptomatic complications
      - Guide interventional procedures

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- Etiologies of pancreatitis:
  - Gallstones and alcohol account for 75-80% of all cases
  - Hypercalcemia, hypertriglyceridemia, medications, a benign or malignant obstruction, pancreatic mass, genetic causes (hereditary pancreatitis), autoimmune pancreatitis (IgG4), infectious etiologies, ischemia secondary to vascular disease, anatomic abnormalities (e.g., pancreas divisum), physiologic abnormalities (Sphincter of Oddi dysfunction), idiopathic causes.
- Complications:
  - Early Phase
    - Generally manifests as a systemic inflammatory response
    - In the first week, imaging findings correlate poorly with clinical severity
    - Advanced imaging is most useful when performed 5-7 days after admission, when local complications have developed and pancreatic necrosis can be clearly defined
    - IEP=acute interstitial edematous pancreatitis
    - Necrotizing Pancreatitis
  - Late Phase:
    - APFC (Acute peripancreatic fluid collection) occurs during the first 4 weeks. If it does not resolve within 4 weeks, it can become organized and develop into a pseudocyst, which contains only fluid with no nonliquefied components.
    - Walled-off necrosis (sequelae of necrotizing pancreatitis): inhomogenous nonliquefied components, encapsulated with a wall.

Note:

Most cases of pancreatitis are mild. More severe cases are usually hospitalized and imaging is performed in that setting. The majority of imaging requests are for the initial evaluation of suspected pancreatitis in individuals with epigastric pain, and then the follow-up imaging of discharged individuals with respect to complications experienced during the hospitalization, to further elucidate the etiology of the pancreatitis if this was not previously established, or to evaluate continued post-discharge symptoms.

Evidence Discussion

Abdominal imaging is useful to confirm the diagnosis of acute pancreatitis (AP). As per 2024 ACG Guidelines, abdominal ultrasound should be performed as the initial imaging study in individuals with AP to evaluate for biliary pancreatitis. Advanced imaging should be reserved for individuals in whom the diagnosis is unclear. When ultrasound results are inconclusive due to overlying bowel gas or other patient factors, or when amylase and/or lipase levels remain elevated, CT or MRI should be considered as the next step. Although contrast-enhanced CT offers over 90% sensitivity and specificity in diagnosing

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acute pancreatitis, its routine use is not recommended since the diagnosis is clear in many individuals who typically experience a mild, uncomplicated course.[1-5,21]

In individuals who fail to improve after 48–72 hours, exhibiting persistent symptoms such as pain, fever, nausea/vomiting, and inability to tolerate oral feeding, imaging studies like CT or MRI/MRCP are recommended. These are used to assess local complications, including necrotizing pancreatitis or pancreatic or peri-pancreatic fluid collections. Although MRI takes more time and can be challenging for claustrophobic individuals, it offers advantages for those with contrast allergies or renal insufficiency. Additionally, MRI can more accurately detect stones in the common bile duct (CBD) and diagnose pancreatic duct disease or follow up on symptomatic fluid collections.[1-5,21]

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Chronic Pancreatitis (AB-33.2)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- If chronic pancreatitis is suspected:
  - Initial imaging:
    - CT Abdomen with contrast or without and with contrast (CPT® 74160 or CPT® 74170) OR MRI Abdomen without and with contrast (CPT® 74183)
      - If diagnostic criteria are met (pancreatic calcification in combination with pancreatic atrophy and/or dilated pancreatic duct):
        - No further imaging indicated (See below regarding worsening symptoms)
    - If initial CT is inconclusive or nondiagnostic of chronic pancreatitis:
      - MRI/MRCP with secretin enhancement (CPT® 74183 or CPT® 74181), OR
      - Endoscopic ultrasound (EUS)
    - If EUS is inconclusive, pancreatic function testing and/or ERCP is medically necessary
    - If abdominal ultrasound is requested at any stage for evaluation of chronic pancreatitis, this can be approved in lieu of advanced imaging
  - If initial imaging fails to confirm chronic pancreatitis, but the clinical suspicion remains, the above testing can be repeated in 6 months.
- Known chronic pancreatitis with worsening symptoms or pain:
  - CT Abdomen with or without and with contrast (CPT® 74160 or CPT® 74170), MRI/MRCP (CPT® 74183 OR CPT® 74181) OR Abdominal ultrasound (CPT® 76700 or CPT® 76705) is medically necessary
  - Possible etiologies of worsening pain include:
    - peptic ulcer disease
    - GI cancers
    - pseudocysts
    - duodenal or common bile duct obstruction
    - pancreatic duct stone or strictures
    - inflammatory masses at the head of the pancreas
- For pre-surgical planning or post-surgical evaluation for treatment of complications of chronic pancreatitis
  - CT Abdomen with or without and with contrast (CPT® 74160 or CPT® 74170), OR MRI/MRCP (CPT® 74183 or CPT® 74181) OR Abdominal ultrasound (CPT® 76700 or CPT® 76705)
- Routine screening for pancreatic cancer in chronic pancreatitis

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- Routine surveillance to monitor for the occurrence of pancreatic cancer in individuals with chronic pancreatitis is not supported at this time. For other indications for imaging in chronic pancreatitis, see the above. For pancreatic cancer screening guidelines in inherited syndromes, including hereditary pancreatitis, see: **Screening Studies for Pancreatic Cancer (ONC-13.1)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Clinical signs of chronic pancreatitis include history of alcohol use, abdominal pain, weight loss, steatorrhea, malabsorption, recurrent pancreatitis, fatty food intolerance, low fecal elastase.

Evidence Discussion

CT or MRI is used as first-line diagnostic imaging for chronic pancreatitis (CP) as they are both universally available, reproducible, and valid when compared to other imaging modalities. While ultrasound has been used for many years as a non-invasive and inexpensive method to evaluate the pancreas, there are considerable limitations that limit its diagnostic utility.[8,13]

Due to its discrepancy in cost, availability, invasiveness, and objectivity, as well as its low specificity, endoscopic ultrasound (EUS) should be used only if the diagnosis is still in question after cross-sectional imaging is performed.[8,13]

Individuals with early CP may have completely normal conventional MRCP/MRI studies, and only the secretin stimulation will depict the mildly abnormal pancreatic duct compliance.[8,13]

When the diagnosis of CP cannot be made following standard cross-sectional imaging or EUS, secretin-enhanced MRCP is suggested as it allows for better visualization of the main- and side-branch ducts by stimulating release of bicarbonate from the pancreatic duct cells and allows for quantification of the degree of filling into the duodenum which may correlate with the severity of CP and also help quantify the degree of exocrine pancreatic function. It does carry a high cost, which is why it is recommended to be used only when diagnosis is not confirmed with first-line testing. However, EUS does carry poor interobserver agreement, and definitive diagnosis is felt to also require advanced radiologic imaging. It is also a more invasive procedure. For this reason, there are also practice guidelines that advocate for the use of MRI/MRCP with secretin enhancement prior to EUS.[8,13]

While multiple other imaging modalities, such as contrast-enhanced EUS, ERCP, transcutaneous ultrasonography, and pancreatic elastography have been used to establish the diagnosis of CP, high-quality RCT evidence is not available to warrant their inclusion as first-line diagnostic tests for CP.[8,13]

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As noted in the American College of Gastroenterology Clinical Guideline for Chronic Pancreatitis, "There is a lack of evidence to suggest that performing screening examinations on individuals with CP (chronic pancreatitis) to detect malignancy is beneficial. Although the overall prevalence of pancreatic malignancy is increased in individuals with CP, there are no RCTs (randomized controlled trials), systematic reviews, or meta-analyses to support screening this individual population for pancreatic malignancy." As such, the ACG Guideline concludes "at this time there is no definitive benefit to screen individuals with CP for pancreatic ductal adenocarcinoma. This is based on the invasive and costly nature of testing, the inherent difficulty in screening given the structural changes of CP, and the inability to alter in many cases the natural history of the disease even if malignancy is detected at an early stage."[13]

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Exocrine Pancreatic Insufficiency (AB-33.3)

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- The presence of any red flag findings per **General Guidelines (AB-1.0)** precludes adjudication based on any other criteria.
- Pancreatic Insufficiency
  - The initial evaluation for pancreatic insufficiency should include ONE of the following laboratory results:
    - Elevation in fecal fat
    - Fecal elastase <200 mcg/g
    - Serum trypsinogen <20ng/mL
  - CT Abdomen with (CPT® 74160) or without and with contrast (CPT® 74170) OR MRI/MRCP (CPT® 74183 or 74181) for the evaluation of suspected pancreatic insufficiency:
    - for suspected pancreatic insufficiency with any one of the above laboratory findings
  - For suspected pancreatic insufficiency due to known chronic pancreatitis, see: **Chronic Pancreatitis (AB-33.2)**
  - For suspected pancreatic insufficiency due to known cystic fibrosis, see: **(PEDAB-16)** and **(PEDCH-5.1)**
  - For suspected pancreatic cancer, see: **Pancreatic Cancer – Suspected/ Diagnosis (ONC-13.2)**

Background and Supporting Information

- Exocrine pancreatic insufficiency (EPI) reflects reduced pancreatic enzymes with resulting maldigestion/malabsorption. When intraduodenal levels of lipase fall below 5-10% of normal output, individuals may manifest with abdominal pain, bloating/ cramping, flatulence, and progressive steatorrhea.

Evidence Discussion

Fecal elastase is the most appropriate initial test for exocrine pancreatic insufficiency (EPI) with a level <100 ug/g of stool providing good evidence of EPI, and levels of 100-200 ug/g being indeterminate for EPI. It is an indirect measurement that is simple, noninvasive, and relatively non-expensive. While direct measurements of pancreatic secretions in to the duodenum are accurate, they are invasive, time-consuming and a more significant burden to the patient than this indirect test.[24]

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Quantitative fecal fat testing is generally not practical for routine clinical use.[24]

While cross-sectional imaging methods such as CT and MRI/MRCP cannot be used to solely identify EPI, they play an important role in the diagnosis of both benign and malignant pancreatic disease, and can also identify gross pancreatic structural changes. Cross-sectional imaging is thus useful for diagnosing underlying pancreatic disease as well as abnormalities that may support an EPI diagnosis.[24]

EPI develops in more than half of individuals with chronic pancreatitis, 27-62% of individuals with relapsing acute pancreatitis, 85% of individuals with cystic fibrosis, and 50-92% of individuals with unresectable pancreatic ductal adenocarcinoma. It is seen in 40-50% of individuals with resectable pancreatic ductal adenocarcinoma before treatment and 65% after treatment. It should thus be suspected in these individuals.[24]

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Asymptomatic Elevation of Pancreatic Enzymes (AB-33.4)

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- If there is the incidental elevation of amylase or lipase:
  - If isolated amylase elevation, prior to imaging, the source of the elevation should be confirmed as pancreatic by the performance of amylase isoenzymes demonstrating that the source is not salivary, or the absence of macroamylase should be ascertained by blood test.
  - If the lipase is elevated alone or in combination with an elevated amylase, or If the amylase is confirmed as pancreatic in origin:
    - Abdominal Ultrasound is medically necessary initially.
    - If US is inconclusive, nondiagnostic, or the elevated pancreatic enzymes persist:
      - MRI/MRCP is medically necessary (CPT® 74183).
        - It is best performed as a secretin-stimulation test in this setting.
      - CT Abdomen (pancreatic protocol, CPT® 74160) is medically necessary if there is a contraindication to MRI.
  - If the pancreatic enzyme elevation persists at one year, either of the above studies can be repeated.

Evidence Discussion

Abdominal imaging is required for the differential evaluation of elevated serum amylase and/or lipase levels and can confirm the diagnosis of acute pancreatitis. Biliary duct dilation and stone disease are readily apparent on an ultrasound, which should be performed as the initial imaging study.[21-23]

When ultrasound results are inconclusive due to overlying bowel gas or other patient factors, or when amylase and/or lipase levels remain elevated, CT or MRI should be considered as the next step. Although contrast-enhanced CT offers over 90% sensitivity and specificity in diagnosing acute pancreatitis, its routine use is not recommended since the diagnosis is clear in many individuals who typically experience a mild, uncomplicated course.[21-23]

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Spleen (AB-34)

Guideline

Spleen (AB-34.1)
Splenic Trauma (AB-34.2)
References (AB-34)

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Spleen (AB-34.1)

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- Incidental splenic findings on US:
  - CT Abdomen (CPT® 74170) or MRI Abdomen (CPT® 74183) is medically necessary.
- Incidental splenic findings on CT or MRI:
  - Imaging is diagnostic of a benign lesion (simple cyst, hemangioma) or characteristics are benign-appearing (homogeneous, low attenuation, no enhancement, smooth margins):
    - No follow-up imaging
  - Imaging characteristics are not diagnostic:
    - Prior imaging available:
      - One year stability: no follow up imaging
      - Lack of stability: consider MRI if not done, biopsy, or PET/CT (CPT® 78815).
    - No prior imaging:
      - No known malignancy:
        - Suspicious imaging features: (suggesting possible malignancy)
          - MRI Abdomen (CPT® 74183) if not already done or biopsy
          - If MRI still inconclusive and biopsy is not feasible then PET/CT (CPT® 78815) is medically necessary.
        - Indeterminate imaging features: (equivocal but not suspicious for malignancy)
          - Follow up MRI Abdomen (CPT® 74183) in 6 and 12 months.
      - Known malignancy:
        - <1 cm: follow up MRI Abdomen (CPT® 74183) in 6 and 12 months.
        - ≥1 cm: consider MRI Abdomen (CPT® 74183) if not done, biopsy
          - If MRI still inconclusive and biopsy is not feasible then PET/CT (CPT® 78815) is medically necessary.
        - (See diagnosis-specific in the *Oncology Imaging Guidelines*).
- Clinically detected splenomegaly
  - Abdominal US (CPT® 76700 or CPT® 76705) should be the first imaging study to evaluate splenic size.
  - If splenomegaly is confirmed, the following evaluation is medically necessary prior to advanced imaging:
    - CBC, evaluation of the peripheral blood smear, LFTs, UA, chest x-ray, HIV testing.

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- CT Abdomen without and with contrast or with (CPT® 74170 or CPT® 74160) is medically necessary if the etiology of the splenomegaly remains unexplained.
- MRI Abdomen (CPT® 74183) is medically necessary for pregnant individuals, or individuals with iodinated contrast allergy.

◦ Nuclear medicine imaging of the liver/spleen (CPT® 78201, CPT® 78202, CPT® 78803, CPT® 78215, CPT® 78216, or CPT® 78830) is rarely performed, but is medically necessary if CT and MRI are contraindicated, as well as for evaluation of an accessory spleen.

Background and Supporting Information

These guidelines are consistent with ACR recommendations for the follow-up of incidental splenic masses. It is noteworthy, however, that a recent study from Beth Israel Deaconess Medical Center in which the authors retrospectively reviewed 379 individuals who were found to have an incidental splenic mass on CT found that in individuals without a history of malignancy, constitutional symptoms of fever or weight loss, or left upper quadrant or epigastric pain (205/379) there were 2 incidences of malignancy. However, in both of these cases the splenic masses were neither isolated nor indeterminate findings as the CTs demonstrated disease in other locations. An isolated splenic malignancy (which can occur but is very rare) was found only in 2 individuals and both of these had constitutional symptoms. Thus, the authors claim that “the isolated and incidentally found splenic mass is of unlikely clinical significance, regardless of its appearance.” They concluded that “in individuals with an incidental splenic mass identified at imaging and with the absence of a history of malignancy, fever, weight loss, or pain in the left upper quadrant or epigastrium, such masses are highly likely to be benign regardless of their appearance. Additional imaging or follow-up is not medically necessary, even if the mass does not show the appearance of simple cyst. Further work-up is only needed if the splenic mass is seen in conjunction with other findings worrisome for malignancy.” These authors challenge the use of the ACR guidelines.

Evidence Discussion

- Splenomegaly is usually the result of systemic disease, and diagnostic studies should be directed toward identifying the etiology. Ultrasound is the preferred modality for documentation of splenomegaly found on physical exam. If the etiology of the splenomegaly is determined (benign or malignant), follow-up imaging would be addressed relative to that disease process.[10]
- The accuracy, cost-effectiveness, and lack of radiation make abdominal ultrasonography a first-line step for confirmation of size.[10]
- Both CT and MRI are valid studies for initial evaluation and follow-up of indeterminate splenic lesions due to the non-specific hypoechogenicity found on ultrasound. These

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should be performed both with and without contrast to improve diagnosis of a solid organ lesion. Nuclear medicine imaging is rarely needed but has a role in detection of accessory splenic lesions.[11]

- There is no evidence-based data supporting the use of serial CT or MRI scans to monitor individuals with incidental splenic lesions that have benign characteristics or lesions that are stable after one year.

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Splenic Trauma (AB-34.2)

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- Ultrasound Abdomen (CPT® 76700 or CPT® 76705) and Pelvis (CPT® 76856 or CPT® 76857) or CT Abdomen and Pelvis without and with contrast (CPT® 74178) or CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for ANY of the following:
  - Blunt abdominal trauma with suspected splenic rupture, OR
  - Suspected post-procedural splenic injury, OR
  - Individuals with penetrating trauma to the left upper quadrant. See: **Blunt Abdominal Trauma (AB-10)**

Background and Supporting Information

Splenomegaly is usually the result of systemic disease, and diagnostic studies are directed toward identifying the causative disease. Complete blood count with differential, LFT's, and peripheral blood smear examination are often performed prior to considering advanced imaging. There is no evidence-based data to support performing serial CT or MRI to follow individuals with incidental splenic lesions.

Evidence Discussion

Spleen being a vascular organ, prompt diagnosis and management of potentially life-threatening bleeding is the primary goal. Emergency splenectomy remains a life-saving procedure; hence, the goal of imaging is to utilize abdominal imaging to determine injury to organs and vasculature with speed and accuracy. Thus, CT and ultrasound (US) are the primary imaging methods to determine splenic injury.[4,6]

US is useful in trauma individuals as it is able to rapidly determine the presence of fluid in peritoneal space. However, it cannot rule out injury to organs with accuracy.[4,6]

CT scan has increased sensitivity and specificity for organ and vascular injury and for identifying individuals a surgical approach. CT is highly sensitive for identifying significant intra-abdominal pathology (97 to 98 percent sensitivity and 97 to 99 percent specificity).[4,6]

Although a noncontrast CT scan may demonstrate sub-capsular hematoma or hemoperitoneum, a contrast-enhanced CT is better able to demonstrate parenchymal and vascular injuries.[4,6]

MRI is not recommended as an imaging study of choice because it is time-consuming to perform and is not as readily accessible as the imaging methods mentioned above (especially in hemodynamically unstable individuals).[4,6]

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Indeterminate Renal Lesion (AB-35)

Guideline

Indeterminate Renal Lesion – General Information (AB-35.0)
Indeterminate Renal Lesion (AB-35.1)
Pre-operative Assessment for Robotic Kidney Surgery (AB-35.2)
References (AB-35)

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Indeterminate Renal Lesion – General Information (AB-35.0)

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For acute flank pain, rule out renal stone, see: **Flank Pain, Rule Out or Known Renal/ Ureteral Stone (AB-4)**

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Indeterminate Renal Lesion (AB-35.1)

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- Incidental Renal Mass on Ultrasound
  - If categorized as simple cyst or Bosniak I or II, no further imaging is medically necessary.
  - Otherwise ONE of the following is medically necessary:
    - CT Abdomen without and with contrast (CPT® 74170)
    - MRI Abdomen without and with contrast (CPT® 74183)
    - Contrast-Enhanced Ultrasound (CPT® 76978) for one lesion
    - Targeted Dynamic Ultrasound (CPT ® 76979) if more than one lesion
- CT Abdomen without and with contrast (CPT® 74170) or MRI Abdomen without and with contrast (CPT® 74183) is medically necessary for further characterization if the original study reveals incomplete visualization of a renal lesion (for example, if only partially visualized on a CT Chest).
- Incidental Renal Mass on CT
  - If characterized as heterogeneous (thick or irregular wall, mural nodule, septa, or calcification):
    - Considered indeterminate. MRI Abdomen without and with contrast (CPT® 74183) OR CT Abdomen without and with contrast (CPT® 74170) is medically necessary
  - If characterized as homogeneous (thin or imperceptible wall, NO mural nodule, septa, or calcification):
    - 10 to 20 HU (Hounsfield units)
      - Likely benign, not fully characterized: no further work-up
    - 21 to 69 HU
      - Indeterminate: MRI Abdomen without and with contrast (CPT® 74183) OR CT Abdomen without and with contrast (CPT® 74170) is medically necessary
    - ≥70 HU
      - Hemorrhagic or proteinaceous cyst, unlikely to be neoplastic: no further work-up
  - If characterized as TSTC (too small to characterize) and homogeneous:
    - If labeled likely benign cyst, not fully characterized:
      - No further work-up
    - If labeled inconclusive based on subjective evaluation:
      - Considered indeterminate. MRI Abdomen without and with contrast (CPT® 74183) (preferred) OR CT Abdomen without and with contrast (CPT® 74170)

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is medically necessary ideally within 6-12 months but no sooner than 6 months.

- Incidental cystic renal mass on CT or MRI without and with contrast (completely characterized, and does NOT contain fat)
  - Bosniak I (benign simple) or II (minimally complicated)
    - No further work-up
  - Bosniak IIF
    - CT Abdomen without and with contrast (CPT® 74170) or MRI Abdomen without and with contrast (CPT® 74183) is medically necessary at 6 and 12 months, then yearly for 5 years
    - If no changes for 5 years, cyst is considered benign and of no clinical significance
  - Bosniak III or IV should be referred for additional management or if chosen, active surveillance see: **Surveillance (ONC-17.4)** in the Oncology Imaging Guidelines
- Incidental solid renal mass or incidental mass too small to characterize evaluated on CT or MRI without and with contrast and does NOT contain fat
  - TSTC (too small to characterize)
    - If labeled likely benign cyst:
      - No further work-up
    - If labeled inconclusive based on subjective evaluation:
      - MRI Abdomen without and with contrast (CPT® 74183) (preferred), OR CT Abdomen without and with contrast (CPT® 74170) is medically necessary ideally within 6-12 months but no sooner than 6 months.
  - If solid mass <1.0cm
    - MRI Abdomen without and with contrast (CPT® 74183) (preferred), OR CT Abdomen without and with contrast (CPT® 74170) is medically necessary beginning at 6 months, then yearly for 5 years
    - If stable at 5 years (average growth ≤3mm per year): No further work-up
    - If mass shows growth (≥4mm per year) or morphologic change: refer for management, consider renal biopsy. If biopsy is technically challenging or relatively contraindicated, a T2 weighted image MRI Abdomen without and with contrast (CPT® 74183) is medically necessary
  - Solid mass 1.0-4.0cm:
    - Considered a small renal neoplasm: refer for management, consider biopsy. If biopsy is technically challenging or relatively contraindicated, a T2 weighted imaging MRI Abdomen without and with contrast (CPT® 74183) is medically necessary. If active surveillance chosen due to limited life expectancy or co-morbidities, see: **Surveillance (ONC-17.4)** in the Oncology Imaging Guidelines
  - Solid renal mass >4.0cm

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        - Considered a renal neoplasm: refer for management, or biopsy. If biopsy is technically challenging or relatively contraindicated, a T2 weighted image MRI Abdomen without and with contrast (CPT® 74183) is medically necessary. If active surveillance chosen due to limited life expectancy or co-morbidities, see: **Surveillance (ONC-17.4)** in the Oncology Imaging Guidelines
- Incidental renal mass containing fat (contains a region of interest measuring <-10 HU on CT)
  - No calcification angiomyolipoma (AML)
    - Solitary and without documentation of growth:
      - <4cm: no further work-up
        - If no prior imaging study for comparison, one follow-up MRI Abdomen (CPT® 74183) OR CT Abdomen (CPT® 74170) can be repeated in 6-12 months to assess for any growth.
      - ≥4cm, and considered an AML with potential for clinical symptoms: refer for management.
    - Multiple lesions or growth documented based on old studies:
      - Refer for management. If active surveillance chosen due to limited life expectancy or co-morbidities, see: **Surveillance (ONC-17.4)** in the Oncology Imaging Guidelines.
  - With calcification (suspected renal cell carcinoma):
    - CT Abdomen without and with contrast (CPT® 74170) OR MRI Abdomen without and with contrast (CPT® 74183) is medically necessary if only a non-contrast CT has been performed. If active surveillance is chosen due to limited life expectancy or co-morbidities, see: **Surveillance (ONC-17.4)** in the Oncology Imaging Guidelines.
- Active Surveillance: For all Active Surveillance indications, see: **Surveillance (ONC-17.4)** in the Oncology Imaging Guidelines

Note: PET/CT or PET/MRI are not considered medically necessary because their role evaluating the incidental renal mass is limited.

Bosniak Classification:

I- Benign simple cyst with a hairline thin wall without septa, calcification, or solid component. Homogeneous near-water attenuation density (10 to 20 HU) without enhancement.

II- Benign minimally complicated cyst that may contain a few hairline thin septa that may have “perceived” but not measurable enhancement. Fine calcification or a segment of slightly thickened calcification may be present in the wall or septa. Also, a well-marginated nonenhancing homogeneous mass <3cm with density above simple fluid attenuation (hyperdense cyst).

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IIF- Usually benign complicated renal cyst with multiple hairline thin septa or minimal smooth thickening of the wall or septa. Wall or septa may contain thick and nodular calcification and may have “perceived” but not measurable enhancement. Also, a well-marginated intrarenal nonenhancing mass >3cm with density above simple fluid.

III -Indeterminate complicated cystic renal mass with thickened irregular walls or septa that have measurable enhancement.

IV-Malignant cystic renal mass with enhancing soft tissue components (cystic renal cell carcinoma).

Evidence Discussion

Advantages of ultrasound includes universal availability, portability, and lack of ionizing radiation. Doppler ultrasound can distinguish between cystic and solid lesions, as well as characterize the quality, presence, and velocity of flow. Therefore, ultrasound can classify a lesion as either a simple cyst or a Bosniak I or II, eliminating the need for further imaging.[1,9,10]

The American Urological Association recommends that individuals with a solid or complex cystic renal mass obtain high quality, multiphase, cross-sectional abdominal imaging to optimally characterize any renal lesion seen on ultrasound, or found incidentally on other imaging studies or non-contrast enhanced abdominal imaging.[1,9,10]

Advanced imaging techniques such as computer tomography (CT) and magnetic resonance imaging (MRI) offer excellent 3-dimensional resolution. CT scans expose individuals to a significant dose of ionizing radiation; however, their rapid image acquisition reduces the potential for motion artifacts. In contrast, MRI provides better soft tissue contrast resolution than CT and does not involve ionizing radiation exposure. Yet, its longer imaging times make it prone to motion artifacts and may necessitate sedation. Additionally, MRIs are contraindicated for individuals with non-MRI compliant implants or ferromagnetic foreign bodies.[1,9,10]

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Pre-operative Assessment for Robotic Kidney Surgery (AB-35.2)

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- Pre-operative assessment for robotic kidney surgery is medically necessary as follows:
  - If not previously performed:
    - CT Abdomen without and with contrast (CPT® 74170) OR
    - MRI Abdomen without and with contrast (CPT® 74183)
  - CTA Abdomen (CPT® 74175) or CTA Abdomen and Pelvis (CPT® 74174) OR
  - MRA Abdomen (CPT® 74185), or MRA Abdomen and Pelvis (CPT® 74185 and CPT® 72198)

Evidence Discussion

Robotic kidney surgery is typically performed for renal masses. Preoperative imaging is essential for:[10]

- Characterizing a renal mass
- Assessing local extension
- Evaluation vascular anatomy
- Planning surgical approach.

CT Abdomen and pelvis without and with contrast is appropriate and provides detailed anatomical and vascular information.[10]

- CT scans expose individuals to a significant dose of ionizing radiation; however, their rapid image acquisition reduces the potential for motion artifacts.[10]

MRI Abdomen without and with contrast is also medically necessary and can be used as an alternative to CT in individuals with contrast allergy or renal insufficiency.[10]

- MRI provides better soft tissue contrast resolution than CT and does not involve ionizing radiation exposure.[10]

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Renal Failure (AB-36)

Guideline

Renal Failure (AB-36.1)
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Renal Failure (AB-36.1)

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- Initial evaluation of acute or chronic renal failure with EITHER of the following:
  - Ultrasound kidney and bladder (CPT® 76770 or CPT® 76775)
  - Doppler Ultrasound (CPT® 93975 or CPT® 93976)
- MRA Abdomen (CPT® 74185) is medically necessary when there is suspected:
  - renal vein/caval thrombosis OR
  - renal artery stenosis as cause of renal failure
- CT Abdomen without contrast (CPT® 74150) is not medically necessary except to rule out ureteral obstruction or retroperitoneal mass.
- Nuclear renal imaging (CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709) is medically necessary for ANY of the following:
  - Renal transplant follow-up
  - Kidney salvage vs. nephrectomy surgical decisions
  - Acute renal failure with no evidence of obstruction on recent ultrasound.
  - Chronic renal failure to estimate prognosis for recovery.
- Nuclear medicine studies of the kidney (CPT® 78700 or CPT® 78701) are medically necessary for evaluation of the following anatomic renal anomalies:
  - Suspected horseshoe kidney OR
  - Suspected solitary or ectopic kidney

Evidence Discussion

The main role of imaging is to detect treatable causes of renal failure such as ureteral obstruction or renovascular disease and to evaluate renal size and morphology. Ultrasound is the modality of choice for initial imaging, with duplex Doppler reserved for suspected renal artery stenosis or thrombosis. ACR appropriateness criteria states that ultrasound contrast media are not nephrotoxic, ultrasound has the greatest diagnostic value in the detection of hydronephrosis, and ultrasound is highly sensitive for hydronephrosis and bladder distention. It also allows for evaluation of general information about the kidney such as size and shape. CT may be appropriate, particularly for urinary tract obstruction. CT is useful in determining the cause of hydronephrosis by demonstrating if mass or obstruction is present and at what level in the urinary tract. MRA is useful when renovascular causes of failure are suspected. MRA has shown to be able to detect renal artery stenosis. However, the use of iodinated and gadolinium-based contrast should be evaluated critically depending on specific patient factors and cost-benefit ratio.[1,3]

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Tc-99m dimercaptosuccinic acid (DMSA) scintigraphy is ideal for functional renal cortical imaging and is most useful for detection of focal renal parenchymal abnormalities and scars in the setting of acute or chronic pyelonephritis or for differential renal function.[1,3]

Tc-99m mercaptoacetyltriglycine (MAG3) is the most frequently used renal tubular agent, specifically to quantify renal tubular extraction.[1,3]

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Renovascular Hypertension (AB-37)

Guideline

Renovascular Hypertension (AB-37.1)

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Renovascular Hypertension (AB-37.1)

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- For consideration of Renovascular Hypertension/Renal Artery Stenosis, see: **Renovascular Hypertension/Renal Artery Stenosis (PVD-6.6)** in the Peripheral Vascular Disease Imaging Guidelines

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Polycystic Kidney Disease (AB-38)

Guideline

Polycystic Kidney Disease (AB-38.1)
References (AB-38)

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Polycystic Kidney Disease (AB-38.1)

AB.PK.0038.1.A

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- Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) is medically necessary for:
  - suspected polycystic kidney disease OR
  - screening individuals at risk for autosomal dominant polycystic disease (ADPKD)
    - In the absence of any clinical change, follow-up screening is not medically necessary if a screening ultrasound was performed at age 40 or later and was negative for any cysts. (The negative predictive value of an ultrasound in this age group is 100% for both PKD1 and PKD2, if no cysts are identified.)
    - If an initial ultrasound is negative for any cysts, a follow-up ultrasound is medically necessary at the discretion of the ordering provider for individuals <40 years of age.
- MRI Abdomen without contrast (CPT® 74181) is medically necessary:
  - if a cystic renal lesion is detected in an individual at-risk of PKD, for prognostic purposes
  - for volume averaging (Total Kidney Volume – TKV) prior to medical treatment and for post treatment follow-up for PKD.

Background and Supporting Information

- Ultrasound is very effective in establishing a diagnosis of ADPKD, though may miss early small cysts. However, the negative predictive value in the various age groups of a negative ultrasound is as follows:
  - ≥40: 100% for PKD1 and PKD2
  - 30-39: 100% for PKD1 and 96.8% for PKD2
  - 5-29: 99.1% for PKD1 and 83.5% for PKD2
- In addition, the preferable advanced imaging study is MRI Abdomen without contrast (CPT® 74181). This is because of the increased risk of gadolinium-induced nephrogenic fibrosis in individuals with PKD.

Evidence Discussion

Screening studies are important for individuals at risk for polycystic kidney disease, as well as imaging protocols to assess and monitor renal parenchyma and evolving cysts, which can predict patient outcomes.[1]

Screening protocols that utilize ultrasonography, a readily available and safe imaging modality, can reliably quantify and characterize renal cysts, aiding in the diagnosis of ADPKD. A negative ultrasound result has a high negative predictive value for excluding ADPKD.[1]

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After diagnosis, advanced imaging may be medically necessary to assess total kidney volume, and to characterize cystic renal lesions, such as before treatment/procedures Optimal follow-up imaging intervals in this setting have not yet been established. Requests for follow-up imaging can be considered on a case-by-case basis. [5]

Given the significant association with CKD, contrast (both gadolinium and iodine-based) would preferentially be avoided for both CT and MR. The choice of advanced imaging would typically be magnetic resonance imaging without contrast unless the benefits outweigh the risks.[1]

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Hematuria and Hydronephrosis (AB-39)

Guideline

Hematuria with Urinary Tract Infection (UTI) (AB-39.1)
Asymptomatic Hematuria (AB-39.2)
Hematuria and Flank Pain (Suspicion for Renal/ureteral Stones) (AB-39.3)
Hydronephrosis of Unexplained or Indeterminate Cause (AB-39.4)
References (AB-39)

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Hematuria with Urinary Tract Infection (UTI) (AB-39.1)

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- Individuals suspected to have a UTI as the etiology of microscopic hematuria should be treated for the UTI and should then undergo repeat urinalysis to confirm resolution of the hematuria.
- If the hematuria persists following treatment of UTI, proceed with the risk-based evaluation as per **Asymptomatic Hematuria (AB-39.2)**.

Background and Supporting Information

- Signs and symptoms of UTI: urinary frequency, burning on urination, urgency, dysuria, positive urine leukocyte esterase, presence of WBCs in the urine, fever, elevated WBC as per the testing laboratory's range

Evidence Discussion

An individual who is diagnosed with microscopic hematuria, defined by the American Urological Association guidelines as 3 or more RBC/HPF, and is found to have a concomitant urinary tract infection should have a repeat urinalysis to confirm resolution of the hematuria based on the AUA guidelines.[6]

If microscopic hematuria persists after treatment of the infection, the individual should undergo risk assessment based on the AUA guidelines which provide guidance on the use of advanced imaging.[6]

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Asymptomatic Hematuria (AB-39.2)

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- A positive dipstick should prompt a microscopic examination.
- Microscopic hematuria is defined as ≥3 red blood cells per high power field.
- Prior to imaging for the evaluation of microscopic hematuria, individuals should be stratified into low, intermediate, or high risk, based on the following criteria:
  - Low risk (individual meets ALL criteria listed)
    - Women <50 years of age or Men <40 years of age
    - Never smoker or <10 pack years
    - 3-10 RBC/HPF on a single urinalysis
    - No additional risk factors for urothelial cancer:
      - Irritative lower urinary tract symptoms
      - Prior pelvic radiation therapy
      - Prior cyclophosphamide/ifosfamide chemotherapy
      - Family history of urothelial cancer or Lynch Syndrome
      - Occupational exposures to benzene chemicals or aromatic amines (e.g. rubber, petrochemicals, dyes)
      - Chronic indwelling foreign body in the urinary tract
  - Intermediate risk (individual meets ANY ONE of these criteria)
    - Women age 50-59 years, Men age 40-59 years
      - 10-30 pack years of smoking
      - 11-25 RBC.HPF on a single urinalysis
      - Low-risk individual with no prior evaluation and 3-10 RBC/HPF on repeat urinalysis
      - Any one of the additional risk factors for urothelial cancer (see above)
  - High-risk (individual meets ANY ONE of these criteria)
    - Women or Men ≥60 years
    - >30 pack-years of smoking
    - >25 RBC/HPF on a single urinalysis
    - Gross hematuria
- Low- or intermediate-risk individuals:
  - Renal ultrasound (combined with cystoscopy)
    - Low-risk individuals may opt for observation with repeat urinalysis within 6 months. If no imaging was performed initially, and follow-up urinalysis reveals persistent hematuria with 3-10 RBC/HPF the individual may be imaged according to Intermediate-Risk criteria. If >10 RBC/HPF, they should be imaged according to High-risk guidelines.

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- High-risk individuals
  - CT Urogram (CPT® 74178) is medically necessary (3D imaging is appropriate if requested)
    - If CT is contraindicated, MR Urography is medically necessary (CPT® 74183 and 72197)
      - If both CT and MR are contraindicated due to contrast, non-contrast CT Urography or renal ultrasound should be performed. See also: **Pregnancy Considerations for Imaging (AB-1.12)**.
- Persistent microscopic hematuria if previously evaluated by renal ultrasound
  - Imaging as per High-risk individuals above
- For the evaluation of microscopic hematuria in individuals with inherited risk factors for renal cortical tumors, ONE of the following imaging studies is medically necessary:
  - Renal ultrasound OR
  - CT Abdomen without and with contrast (CPT® 74170) OR
  - MRI Abdomen without and with contrast (CPT® 74183)

Note:

Inherited risk factors for renal cortical tumors include:

- Von Hippel Lindau
- Birt-Hogg-Dube
- Hereditary Papillary Renal Cell Cancer
- Hereditary Leiomyomatosis Renal Cell Cancer
- Tuberous Sclerosis

- Follow-up
  - Individuals with a negative hematuria evaluation who undergo repeat urinalysis
    - If repeat urinalysis is negative:
      - No further workup
    - If repeat urinalysis demonstrates persistent hematuria
      - Repeat imaging as requested (Renal Ultrasound or CT Urography)
- **Note:** 3-D Reconstruction enhances a CT Urogram. Requests for 3-D reconstruction (CPT® 76377 or 76376) for a CT Urogram can be considered medically necessary.

Evidence Discussion

- Microscopic hematuria is defined as ≥3 red blood cells per high power field. Hematuria is NOT defined as a positive dipstick. A positive dipstick should prompt a microscopic examination. A positive dipstick is not considered as defining microhematuria.[6]

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- Low-risk individuals with microscopic hematuria may opt for a repeat urinalysis prior to proceeding to a workup. Intermediate-risk and high-risk individuals should undergo a workup with upper and lower tract imaging.[5,6]
  - Upper tract imaging with renal ultrasound is the standard for low and intermediate individuals given the overall low rate of malignancy detected in individuals with microscopic hematuria. Renal ultrasound is noninvasive, readily available, and carries no risk of ionizing radiation while demonstrating a high sensitivity for renal masses and hydronephrosis.
  - Upper tract imaging for high risk individuals should include advanced imaging with urography (CT with/without contrast is preferred with associated 3D rendering if requested). MR Urogram (MR Abdomen and Pelvis with/without contrast) can be performed if CT is contraindicated.
  - Individuals with severe renal dysfunction, dye allergy, or other reasons where both CT and MRI are contraindicated should undergo renal ultrasound or non-contrast CT paired with retrograde pyelography.
- Individuals with microhematuria with family history of renal cell carcinoma or known genetic renal tumor syndrome should undergo upper tract imaging (renal ultrasound, CT or MR Urography) regardless of risk category.[1,6]
- An individual with previous negative workup with persistent microscopic hematuria may undergo repeat upper tract imaging.[1,6]

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Hematuria and Flank Pain (Suspicion for Renal/ureteral Stones) (AB-39.3)

AB.HH.0039.3.A

v1.0.2026

- CT Abdomen and Pelvis without contrast (CPT® 74176) OR CT Urogram (CPT® 74178)
  - Request for 3-D reconstruction (CPT® 76377 or CPT® 76375) to be performed with a CT Urogram is considered medically necessary.
  - US abdomen or retroperitoneum is medically necessary in lieu of CT for any of the above indications.

Evidence Discussion

- Individuals with flank pain presenting with either microscopic or gross hematuria should undergo advanced imaging with CT of the abdomen and pelvis.
  - The choice of contrast is at the discretion of the provider and may differ for individuals with previous history or high risk of nephrolithiasis and individuals with a higher risk of malignancy.
  - 3D reconstruction of CT Urography may be performed as requested.
  - Alternatively, the provider may request abdominal or retroperitoneal ultrasound in lieu of a CT initially.

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Hydronephrosis of Unexplained or Indeterminate Cause (AB-39.4)

AB.HH.0039.4.A
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- CT Urogram (CPT® 74178)
  - Requests for 3-D reconstruction (CPT® 76377 or CPT® 76375) to be performed with a CT Urogram is considered medically necessary.
  - US abdomen or retroperitoneum is medically necessary in lieu of a CT for any of the above indications.
- Retroperitoneal Ultrasound (CPT® 76770) is medically necessary every 6 to 12 months for the following:
  - uncomplicated hydronephrosis OR
  - neurogenic bladder OR
  - myelomeningocele (open spinal dysraphism) OR
  - spina bifida

Evidence Discussion

- A new diagnosis of hydronephrosis without a known cause should undergo further workup. Advanced imaging with CT Urography with 3D reconstruction may be performed if requested to evaluate the course of the urinary tract for obstruction.[7]
- Alternatively, the provider may request abdominal or retroperitoneal ultrasound in lieu of a CT initially.[7]
- Individuals with known chronic, uncomplicated hydronephrosis or individuals with neurogenic bladder (spina bifida or other neurologic conditions) may undergo surveillance imaging with retroperitoneal ultrasound every 6-12 months to monitor for progression or development of hydronephrosis to prevent renal deterioration.[7]

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Urinary Tract Infection (UTI) (AB-40)

Guideline

Urinary Tract Infection (AB-40.0)
Pyelonephritis (AB-40.1)
Lower Urinary Tract Infection (AB-40.2)
References (AB-40)

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Urinary Tract Infection (AB-40.0)

AB.UT.0040.0.A
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These guidelines refer to UTI without Hematuria.

For UTI with Hematuria, see: **Hematuria and Hydronephrosis (AB-39)**

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Pyelonephritis (AB-40.1)

AB.UT.0040.1.A

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- CT Abdomen and Pelvis without and with contrast (CPT® 74178) or CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if:
  - suspected complicated: diabetes, immunocompromised, history of stones, prior renal surgery, or fever ≥101 F (≥38.5 C) OR
  - not responding to therapy after 3 days OR
  - recurrent pyelonephritis (at least 1 prior pyelonephritis) OR
  - males with first time UTI or recurrent UTI without etiology

  Note:

  3-D Reconstruction enhances a CT Urogram (CPT® 74178).

  Requests for 3-D reconstruction (CPT® 76377 or 76376) for a CT Urogram can be considered medically necessary.

- MRI Abdomen without or with and without contrast (CPT® 74181 or CPT® 74183) is medically necessary if:
  - Elevated creatinine
- Pregnant individuals should be evaluated initially by renal ultrasound (CPT® 76770 or CPT® 76775) and if further imaging is medically necessary, MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195).

Evidence Discussion

- Pyelonephritis is a clinical diagnosis and advanced imaging is often not beneficial according to guidance from the American College of Radiology and the American Urological Association, as a majority of individuals will clinically improve with appropriate antibiotic therapy.[1,9,10]
- Advanced imaging may be medically necessary with contrasted CT (urography if requested) in individuals with complicated clinical pictures which may include immunocompromised individuals or those with diabetes mellitus, history of nephrolithiasis, prior renal surgery, or those with fever. All males with urinary tract infection are considered to have a complicated urinary tract infection and thus advanced imaging may be considered medically necessary[1,9,10]
- Alternative imaging with MRI of the abdomen and pelvis with and without contrast may be medically necessary if renal dysfunction is present.[1,9,10]
- If an individual is unresponsive to therapy after 3 days, or if there is at least one prior episode of pyelonephritis, advanced imaging may be medically necessary. [1,9,10]

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- Pregnant individuals are considered high risk for complications from pyelonephritis, however first line imaging should be with renal ultrasound to avoid ionizing radiation exposure. If further imaging is felt medically necessary, MRI of the abdomen and pelvis without contrast may be performed.[1,3,9,10]

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Lower Urinary Tract Infection (AB-40.2)

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- CT Abdomen and Pelvis without and with contrast (CPT® 74178) is medically necessary if:
  - suspected complicated: diabetes,immunocompromised,history of stones, prior renal surgery, or fever ≥101 F (≥38.5 C) OR
  - not responding to therapy after 3 days OR
  - males with first time UTI or recurrent UTI without etiology OR
  - recurrent UTI ≥3 per year OR
  - recommendation by or in consultation with a urologist or specialist

  Note: 3-D Reconstruction enhances a CT Urogram (CPT® 74178). Requests for 3-D reconstruction (CPT® 76377 or 76376) for a CT Urogram can be considered medically necessary.
- MRI Abdomen and MRI Pelvis without or with and without contrast (CPT® 74181 and CPT® 72195 or CPT® 74183 and CPT® 72197) is medically necessary if requested when ALL of the following apply:
  - Criteria (as above) for CT Abdomen and Pelvis without and with contrast are met, AND
  - Elevated creatinine
- See: **Periurethral Cysts and Urethral Diverticula (PV-13)** in the Pelvis Imaging Guidelines

Evidence Discussion

- Advanced imaging for a lower urinary tract infection is not beneficial in most clinical scenarios according to guidance from the American College of Radiology and the American Urological Association, as few individuals with cystitis will progress to an upper urinary tract infection.[1,2,8,9]
- CT of the abdomen and pelvis with and without contrast may be medically necessary in the context of a complicated urinary tract infection, recurrent urinary tract infections (greater than 3 episodes in one year), or if recommended by a urologist or specialist.[1,2,8,9]
- Complicated urinary tract infections may include immunocompromised individuals or those with diabetes mellitus, history of nephrolithiasis, prior renal surgery, or those with fever. All males with urinary tract infection are considered to have a complicated urinary tract infection and thus advanced imaging may be considered medically necessary[1,2,8,9]

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- Alternative imaging with MRI of the abdomen and pelvis with and without contrast may be medically necessary if renal dysfunction is present. [1,2,8,9]

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Patent Urachus (AB-41)

Guideline

Patent Urachus (AB-41.1)

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Patent Urachus (AB-41.1)

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For evaluation of suspected patent urachus, see: **Patent Urachus (PV-23.1)** in the Pelvis Imaging Guidelines.

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Transplant (AB-42)

Guideline

Liver Transplant, Pre-Transplant (AB-42.1)
Liver Transplant, Living Donor Pre-Transplant Imaging (Donor Imaging) (AB-42.2)
Liver Transplant, Post-Transplant Imaging (AB-42.3)
Post-Transplant Lymphoproliferative Disorder (PTLD) (AB-42.4)
Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)
Kidney Transplant, Post-Transplant (AB-42.6)
Heart Transplant (AB-42.7)
References (AB-42)

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Liver Transplant, Pre-Transplant (AB-42.1)

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- Individuals **WITHOUT hepatocellular carcinoma (HCC)** referred to a transplant center for liver transplant evaluation can undergo advanced imaging as follows:
  - Per the transplant institution's protocol, OR
  - Per the studies and intervals listed below:

Imaging Study	Interval	Comments
Both of the following US studies: ◦ Abdominal US (CPT® 76700 or CPT® 76705) and ◦ Doppler (CPT® 93975)	◦ Every 6 months	
ONE of the following abdomen/pelvis advanced imaging studies: ◦ CT Abdomen (CPT® 74160 or CPT® 74170) ◦ MRI Abdomen (CPT® 74183)	◦ Annually ▪ Individuals with known cholangiocarcinoma may have more frequent repeat of studies at left per institution's protocol	
Additional abdomen/ pelvis advanced imaging, for individuals on the transplant list with known Primary Sclerosing Cholangitis (PSC): ◦ MRCP (See: **MRCP (AB-27.1)** for acceptable CPT® codes)	◦ Per the transplant institution's protocol	

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Imaging Study	Interval	Comments
◦ CT Chest with or without contrast (CPT® 71260 or CPT® 71250)	◦ One-time ▪ Individuals with known cholangiocarcinoma may have more frequent repeat of studies at left per institution's protocol	◦ Repeat studies based on clinical indications per **Chest Imaging Guidelines**
ONE of the following: ◦ MRI Bone Marrow Blood Supply (CPT® 77084) or ◦ Bone scan (CPT® 78306)	◦ One-time	
Echocardiography with ONE of the following: ◦ CPT® 93306 (preferred) ◦ CPT® 93307 ◦ CPT® 93308	◦ Annually	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
CT Coronary angiography (CCTA) (CPT® 75574)	◦ Annually	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
Stress imaging in place of but not in addition to CT Coronary angiography (CCTA) - ONE of the following: ◦ CPT® 93350 ◦ CPT® 93351 ◦ CPT® 78452 ◦ CPT® 75563 ◦ CPT® 78492 ◦ CPT® 78431	◦ Annually	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications

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Imaging Study	Interval	Comments
For individuals with systemic amyloidosis: ◦ Cardiac MRI – ONE of the following: ▪ CPT® 75557 ▪ CPT® 75561 ◦ If Cardiac MRI is contraindicated or indeterminate, ONE of the following SPECT studies may be performed: ▪ CPT® 78803 ▪ CPT® 78830	◦ One-time	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
If required to further assess CAD seen on a recent CCTA that is of uncertain physiologic significance, CT-FFR (Noninvasive estimated coronary fractional flow reserve derived from coronary computed tomography angiography) with ONE of the following: ◦ CPT® 0501T ◦ CPT® 75580	◦ One-time	See: **Cardiac Imaging in Transplants CD-16** for descriptions of CPT codes or further indications

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Imaging Study	Interval	Comments
In place of CT Coronary angiography or stress imaging for initial pre-transplant evaluation, OR If CT Coronary angiography and/or CT-FFR or stress imaging is abnormal WITH addition of right heart catheterization if requested for evaluation of pulmonary hypertension: ◦ Left heart catheterization or left and right heart catheterization with ONE of the following: ▪ CPT® 93458 ▪ CPT® 93454 ▪ CPT® 93460 ▪ CPT® 93456 ◦ Or if prior CABG, with ONE of the following: ▪ CPT® 93459 ▪ CPT® 93455 ▪ CPT® 93461 ▪ CPT® 93457	◦ One-time	Repeat studies as per **Cardiac Imaging in Transplant CD-16**
ONE of the following, for vascular evaluation in anticipation of transplant: ◦ CTA (CPT® 74175) ◦ MRA Abdomen (CPT® 74185)	◦ One-time	

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Imaging Study	Interval	Comments
ANY of the following may be performed immediately prior to transplant: ◦ Abdominal US (CPT® 76700 or CPT® 76705) AND Doppler (CPT® 93975) ◦ CT Abdomen (CPT® 74160 or CPT® 74170) OR MRI Abdomen (CPT® 74183) ◦ CT Abdomen and Pelvis (CPT® 74177) or CT Pelvis (CPT® 72193) ◦ CTA (CPT® 74175) OR MRA Abdomen (CPT® 74185)	◦ Once, immediately prior to transplant	

- Individuals **WITH hepatocellular carcinoma (HCC)** referred to a transplant center for liver transplant evaluation can undergo advanced imaging as follows:
  - Per the transplant institution's protocol, OR
  - Per the studies and intervals listed below:

Imaging Study	Interval	Comments
Both of the following US studies: ◦ Abdominal US (CPT® 76700 or CPT® 76705) and ◦ Doppler (CPT® 93975)	◦ Every 6 months	

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Imaging Study	Interval	Comments
ONE of the following abdomen/pelvis advanced imaging studies: ◦ CT Abdomen (CPT® 74160 or CPT® 74170) ◦ MRI Abdomen (CPT® 74183)	◦ Every 3 months ▪ Can be approved at interval as requested according to the transplant center's protocol for waitlisted individuals under active locoregional therapy to control tumor growth (i.e., tumor ablation)	
◦ CT Chest with contrast (CPT® 71260)	◦ Every 6 months ▪ Can be approved at interval as requested according to the transplant center's protocol for waitlisted individuals under active locoregional therapy to control tumor growth (i.e., tumor ablation)	
◦ Bone Scan (CPT® 78306)	◦ Every 6 months	
Echocardiography with ONE of the following: ◦ CPT® 93306 (preferred) ◦ CPT® 93307 ◦ CPT® 93308	◦ Annually	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
CT Coronary angiography (CCTA) (CPT® 75574)	◦ Once in 3 years	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications

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Imaging Study	Interval	Comments
Stress imaging in place of but not in addition to CT Coronary angiography (CCTA) - ONE of the following: ◦ CPT® 93350 ◦ CPT® 93351 ◦ CPT® 78452 ◦ CPT® 75563 ◦ CPT® 78492 ◦ CPT® 78431	◦ Annually	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
For individuals with systemic amyloidosis: ◦ Cardiac MRI – ONE of the following: ▪ CPT® 75557 ▪ CPT® 75561 ◦ If Cardiac MRI is contraindicated or indeterminate, ONE of the following SPECT studies may be performed: ▪ CPT® 78803 ▪ CPT® 78830	◦ One-time	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications

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Imaging Study	Interval	Comments
If required to further assess CAD seen on a recent CCTA that is of uncertain physiologic significance, CT-FFR (Noninvasive estimated coronary fractional flow reserve derived from coronary computed tomography angiography) with ONE of the following: ◦ CPT® 0501T ◦ CPT® 75580	◦ One-time	See: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications

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Imaging Study	Interval	Comments
In place of CT Coronary angiography or stress imaging for initial pre-transplant evaluation, OR If CT Coronary angiography and/or CT-FFR or stress imaging is abnormal WITH addition of right heart catheterization if requested for evaluation of pulmonary hypertension: ◦ Left heart catheterization or left and right heart catheterization with ONE of the following: ▪ CPT® 93458 ▪ CPT® 93454 ▪ CPT® 93460 ▪ CPT® 93456 ◦ Or if prior CABG, with ONE of the following: ▪ CPT® 93459 ▪ CPT® 93455 ▪ CPT® 93461 ▪ CPT® 93457	◦ One-time	Repeat studies as per **Cardiac Imaging in Transplant CD-16**

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Imaging Study	Interval	Comments
ANY of the following may be performed immediately prior to transplant: ◦ Abdominal US (CPT® 76700 or CPT® 76705) AND Doppler (CPT® 93975) ◦ CT Abdomen (CPT® 74160 or CPT® 74170) OR MRI Abdomen (CPT® 74183) ◦ CT Abdomen and Pelvis (CPT® 74177) or CT Pelvis (CPT® 72193) ◦ CTA (CPT® 74175) OR MRA Abdomen (CPT® 74185) ◦ MRI Bone Marrow Blood Supply (CPT® 77084)	◦ Once, immediately prior to transplant	

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Liver Transplant, Living Donor Pre-Transplant Imaging (Donor Imaging) (AB-42.2)

AB.TX.0042.2.A

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- CT Abdomen with contrast (CPT® 74160) OR CT Abdomen without and with contrast (CPT® 74170) OR MRI Abdomen without and with contrast (CPT® 74183) is medically necessary to assess liver anatomy and volumetrics.
- MRCP is medically necessary to assess biliary anatomy (See: **MRCP (AB-27.1)** for proper coding)
- CTA Abdomen without and with contrast (CPT® 74175) OR MRA Abdomen without and with contrast (CPT® 74185) is medically necessary to assess vascular anatomy.
- For donor imaging post-transplant, imaging is medically necessary per transplant center protocol. If no transplant center protocol exists, see condition-specific guideline appropriate to the individual's signs and symptoms.

Evidence Discussion

Living donor liver transplantation (LDLT) has become a widely accepted solution to alleviate the ongoing shortage of cadaveric livers for deceased donor liver transplantation (DDLT). Radiologic evaluation plays a crucial role in assessing both donor candidates and recipients to confirm their eligibility and determine the most suitable surgical approach.[54]

A comprehensive pre-operative assessment of the vascular, liver volume, and biliary anatomy is vital for the safe and successful harvesting, transplantation, and long-term success of the graft. Computed tomography (CT) and magnetic resonance imaging (MRI) are the preferred imaging techniques for this purpose. These cross-sectional methods offer detailed views of the vascular and biliary structures, assess the hepatic parenchyma, and enable volumetric analysis.[16,55]

LDLT evaluation typically combine MRI/MRCP and CT to leverage the higher spatial resolution of CT for arterial evaluation and the superior soft tissue, parenchymal and biliary analysis provided by MRI. Besides examining the liver parenchyma for abnormalities such as steatosis, a detailed evaluation of the hepatic volume, vascular and biliary system for significant anatomic variants is essential, as these variants can influence surgical techniques and outcomes for both recipients and donors.[56]

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Liver Transplant, Post-Transplant Imaging (AB-42.3)

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- Cardiac Imaging:
  - See: **Cardiac Imaging in Transplants CD-16** in the Cardiac Imaging Guidelines
- Suspected post-operative complications:
  - Vascular thrombosis (suspected hepatic artery thrombosis) imaging is medically necessary as follows:
    - Doppler ultrasound (CPT® 93975)
    - CTA Abdomen without and with contrast (CPT® 74175) OR MRA Abdomen without and with contrast (CPT® 74185)
  - Suspicion of biliary anastomotic strictures:
    - MRCP (See: **MRCP (AB-27.1)** for appropriate CPT codes)
    - Vascular imaging as above for vascular thrombosis may also be requested and considered medically necessary for this indication
  - Other suspected post-operative complications (e.g., infection, etc.)
    - Imaging as requested by the transplant institution or team
- Transplant individuals without prior HCC or cholangiocarcinoma:
  - Routine post-transplant imaging is not medically necessary.
  - If cirrhosis develops post-transplant:
    - See: **Cirrhosis and Liver Screening for Hepatocellular Carcinoma (HCC) (AB-26.1)**, **Ascites (AB-26.2)**, and **Portal Hypertension (AB-26.3)** for HCC screening guidelines.
  - Fibrosis assessment post-liver transplant:
    - Transient elastography (CPT® 91200) is medically necessary, which is the most studied modality in this setting.
- Surveillance after transplant for HCC is medically necessary as follows:
  - Based on RETREAT score
    - 0 points: No additional screening needed
    - 1-3 points: CT or MRI Abdomen (CPT® 74160, or CPT® 74170, or CPT® 74183) and CT Chest (CPT® 71260 or CPT® 71250) every 6 months for 2 years.
    - 4 points: CT or MRI Abdomen (CPT® 74160, or CPT® 74170, or CPT® 74183) and CT Chest (CPT® 71260 or CPT® 71250) every 6 months for 5 years
    - ≥5 points: CT or MRI Abdomen (CPT® 74160, or CPT® 74170, or CPT® 74183) and CT Chest (CPT® 71260 or CPT® 71250) every 3 months for 2 years, then every 6 months between the 2nd and 5th years.

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- If there is a suspicion of recurrent tumor based on clinical findings and/or sequentially increasing AFP:
  - CT Abdomen (CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) is medically necessary
- Imaging after transplant for primary sclerosing cholangitis (PSC):
  - Suspected recurrence of PSC;
    - MRCP (See: **MRCP (AB-27.1)** for proper coding)
- Imaging after transplant for cholangiocarcinoma is medically necessary as follows:
  - Liver ultrasound (CPT® 76705 or CPT® 76700) or MRI Abdomen and MRCP (CPT® 74183) every 6 months for 5 years post-transplantation.
  - CT Chest (CPT® 71250 or CPT® 71260) every 6 months for 5 years post-transplantation

Background and Supporting Information

Consensus guidelines regarding post-transplant surveillance imaging have not yet been established. There have been recent attempts to establish evidence-based guidelines, including the development of the RETREAT score, validated recently in a study conducted at University of California, San Francisco, Mayo Clinic-Rochester, and Mayo Clinic-Jacksonville. This scoring system has been adopted for use by UCSF and guides post-transplant imaging for individuals who have undergone transplant for HCC.

The RETREAT score is a protocol used to estimate the risk of tumor recurrence after liver transplantation in individuals who have been transplanted for the treatment of hepatocellular carcinoma. It is comprised of three factors which are assessed before and after transplant. Points are assigned based on criteria which include the alpha-fetoprotein level before liver transplantation, the presence or absence of microvascular invasion, and the sum of the diameter of the largest viable tumor and the number of viable nodules on pathologic examination of the explant liver.

The RETREAT score is obtained by adding the total number of points scored in each of the 3 variables (range 0-8). For example: RETREAT score=0 if the individual has AFP 0-20 ng/mL at LT, no vascular invasion and no viable tumor in the explant.

The RETREAT score is calculated as follows:

Risk Factor	Score
Alpha-fetoprotein level before LT	
0-20	0
21-99	1

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Risk Factor	Score
100-999	2
≥1000	3
Microvascular invasion present	2
Sum of the diameter of the largest viable tumor and the number of viable nodules	
0	0
1.1-4.9	1
5.0-9.9	2
≥10	3

Evidence Discussion

Clinical manifestations of liver transplant complications can be subtle and non-specific and medical imaging plays an important role. Often, a rise in liver enzymes is the earliest sign of graft problems, allowing for timely clinical intervention to protect allograft function.[20-22,24-30,32,33]

Throughout the lifetime of a post liver transplant individual, complications affecting the liver allograft could be caused by vascular and biliary complications, immune-mediated injury, drug-related issues, infectious complications, and recurrence of the primary liver disease.[20-22,24-30,32,33]

Thus, managing these individuals depends on a thorough clinical history, symptoms, laboratory data, and imaging studies; at times multiple imaging modalities are required.

There is no specific consensus of what type, or when a post liver transplant individual will need or require an imaging test and it typically depends on post liver transplant imaging protocols specific to a transplant centre, or abnormal laboratory tests.[20-22,24-30,32,33]

However, as standard practice, ultrasound sonography plus colour-Doppler ultrasound examination is routinely performed at 24–48 h, on the 7th day and 21st day (Mayo Clinic protocol), and on the first and third month after transplantation to evaluate the

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liver parenchyma and vascular structures integrity. The frequency and indication vary between transplant centres, and post-transplant protocols.[20-22,24-30,32,33]

In addition, testing is performed anytime there is an unexpected change in liver enzymes potentially including additional testing such as CT imaging and MR imaging techniques, including contrast-enhanced CT or MR angiography and MR cholangiography to further evaluate the transplanted liver. These tests can reveal abnormalities in vascular structures, bile ducts, liver parenchyma, and extrahepatic tissues.[20-22,24-30,32-33]

In the case of a history of pre-liver transplant hepatocellular carcinoma (HCC), even with adherence to Milan criteria, HCC recurs post-LT in 10%–15% and is the most common cause of death in this population. [20-22,24-30,32,33]

A multicenter analysis has proposed and validated a risk stratification score, Risk Estimation of Tumor Recurrence After Transplant (RETREAT), which incorporates AFP at LT, vascular invasion, the sum of the largest viable tumor diameter, and number of viable tumors on explant.[20-22,24-30,32,33]

RETREAT stratifies 5-year recurrence risk from <3% in individuals without viable tumor on explant or microvascular invasion and AFP <20ng/ml (i.e., RETREAT 0) up to 75% in the highest-risk individuals (RETREAT≥5).[6]

In this population, because the two most common sites of post-transplant recurrence are the lung (~40%) followed by the liver (33%), surveillance is advised. The AASLD advises surveillance for detection of post-transplant HCC recurrence using multiphasic contrast-enhanced abdominal CT or MRI and chest CT scan. The optimal timing and duration of post-transplant surveillance is uncertain; however, risk scores may be considered to guide decisions. [20-22,24-30,32,33]

Beyond allograft-related complications, metabolic syndrome, cardiovascular disease, renal dysfunction, and malignancies are leading causes of morbidity and mortality in this individual population. These individuals will require cardiovascular evaluation, breast cancer, and lung cancer surveillance per individual risk and transplant centre expert team recommendations as some individuals could carry a slightly higher risk than the non-transplant population.[20-22,24-30,32,33]

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Post-Transplant Lymphoproliferative Disorder (PTLD) (AB-42.4)

AB.TX.0042.4.A

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- CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for known or suspected PTLD.
- Additional evaluation of suspected PTLD is the same as the evaluation of lymphoma. See: **Post-Transplant Lymphoproliferative Disorders (ONC-27.9)** in the Oncology Imaging Guidelines for further recommendations
- Routine use of imaging to screen for PTLD is not medically necessary as there is insufficient supporting evidence.

Background and Supporting Information

- Post-transplant lymphoproliferative disease (PTLD) is a major complication of solid organ transplantation and the spectrum ranges from benign hyperplasia to malignant lymphoma. It has an incidence of 1-20%, and is usually related to Epstein-Barr virus infection in the setting of immunosuppression.

Evidence Discussion

For suspected PTLD advanced imaging studies are extremely helpful. CT Chest/ Abdomen/Pelvis with contrast are the mainstay for known or suspected PTLD. PTLD generally is rapid growing and small ill-defined masses of lymphoid tissue cannot be initially identified on sonography. Since PTLD has the potential of being reversed by decreasing immunosuppression, early detection with more advanced imaging can very beneficial.[37]

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Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)

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Pre-Transplant Evaluation (Per Institution Protocol)

- Individuals referred to a transplant center for kidney or kidney-pancreas transplant evaluation can undergo advanced imaging as follows:
  - Per the transplant institution's protocol, OR
  - Per the studies and intervals listed below:

Imaging Study	Interval	Comments
ONE of the following abdomen/pelvis imaging studies is medically necessary: • CT Abdomen and Pelvis without contrast (CPT® 74176) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CTA Abdomen without and with (CPT® 74175) • CTA Abdomen and Pelvis (CPT® 74174) • CTA Pelvis without and with (CPT® 72191)	One-time	
ONE of the following echocardiography studies is medically necessary: • CPT® 93306 (preferred) • CPT® 93307 • CPT® 93308	Annual	See also: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
ONE of the following stress imaging studies is medically necessary: • CPT® 93350 • CPT® 93351 • CPT® 78452 • CPT® 75563 • CPT® 78492 • CPT® 78431	Annual	See also: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications

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Additional Pre-Transplant Evaluation (Per Indication)

Individuals referred to a transplant center for kidney or kidney-pancreas transplant evaluation can undergo the following additional advanced imaging when the listed indications are met:

Indication	Imaging Study	Interval	Comments
• 20 pack-year history of smoking	ONE of the following is medically necessary: • CT Chest without contrast (CPT® 71250) • CT Chest with contrast (CPT® 71260)	One-time	For lung cancer screening with Low Dose Computed Tomography (LDCT), see: **U.S. Preventative Services Task Force: Lung Cancer Screening (Commercial and Medicaid) (CH-33.1)**
• Autosomal dominant polycystic kidney disease	ONE of the following is medically necessary: • MRA Head (CPT® 70544, 70545, or 70546) • CTA Head (CPT® 70496)	One-time	Repeat imaging as per **Intracranial Aneurysms (HD-12.1)**
• History of stroke, or • History of TIA, or • Carotid bruit on exam	ONE of the following is medically necessary: • Carotid duplex bilateral study (CPT® 93880 or CPT® 73882)	One-time	Repeat imaging as per **Initial Imaging (PVD-3.1)**
• Presence of systemic amyloidosis	ONE of the following cardiac MRI studies is medically necessary: • CPT® 75557 • CPT® 75561	One-time	See also: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications

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Indication	Imaging Study	Interval	Comments
BOTH of the following: • Presence of systemic amyloidosis AND • Cardiac MRI is either contraindicated or indeterminate	ONE of the following nuclear medicine studies is medically necessary: • CPT® 78803 • CPT® 78830	One-time	See also: **Cardiac Imaging in Transplant CD-16** for descriptions of CPT codes or further indications
• In place of stress imaging for initial pre-transplant evaluation, or • Stress imaging is positive for ischemia	ONE of the following heart catheterization is medically necessary: • CPT® 93458 • CPT® 93454 • If prior CABG: ◦ CPT® 93459 ◦ CPT® 93455	One-time	Repeat imaging as per **Cardiac Imaging in Transplant CD-16**

Kidney Donor Nephrectomy or Pre-Transplant Nephrectomy

Indication	Imaging Study	Comments
• Individuals being evaluated for living kidney donation, or • Individual is planning removal of one or both kidneys	ONE of the following is medically necessary: • CTA Abdomen (CPT® 74175) • MRA Abdomen (CPT® 74185) • MRI Abdomen without and with contrast (CPT® 74183)	For CTA and MRA, 3D rendering is included with the original study

Evidence Discussion

Individuals being assessed for kidney or kidney-pancreas transplant require advanced imaging of the abdomen and/or pelvis either with or without contrast (to include angiography). This allows assessment of any intra-abdominal pathology, which may complicate transplantation. MR angiography may be indicated for assessment of the

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native kidneys when considering pre-transplant nephrectomy. Individuals may also be assessed according to the standardized imaging protocol of the transplant center.[14-18]

Although there is some debate regarding coronary artery disease (CAD) screening and transplant outcomes, a preoperative cardiac workup is essential for prognostication given the significant association with chronic kidney disease (CKD) and CAD. This may include a transthoracic echocardiogram as well as a stress echocardiogram and/or cardiac catheterization.[14-18]

Cardiac MRI can be performed in individuals with systemic amyloidosis, as cardiac involvement is the leading cause of morbidity and mortality. If the MRI is indeterminate or contraindicated, myocardial Tc-99m pyrophosphate imaging may be performed.[14-18]

Individuals with an extensive smoking history of greater than 20 pack-years may undergo CT of the chest (either with or without contrast), which is guided by evidence of the National Lung Screening Trial to reduce risk of mortality.[14-18]

Any individual with a history of transient ischemic attack (TIA) or stroke may undergo a carotid duplex study for preoperative assessment. Individuals with autosomal dominant polycystic kidney disease (ADPKD) may undergo MR or CT angiography of the head to screen for aneurysms.[14-18]

Individuals being assessed for kidney donation should have advanced abdominal imaging with CT or MR angiography to assess kidney size and vasculature.[14-18]

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Kidney Transplant, Post-Transplant (AB-42.6)

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- Ultrasound of transplanted kidney:
  - Current ultrasound imaging protocols of the transplanted kidney commonly include a Doppler study and are coded as CPT® 76776.
    - Do not report non-invasive vascular codes CPT® 93975 and CPT® 93976 in conjunction with CPT® 76776.
  - Ultrasound of the transplanted kidney performed without duplex Doppler should be reported as a limited retroperitoneal ultrasound (CPT® 76775).

Evidence Discussion

- Imaging evaluation of the transplanted kidney may be necessary for routine surveillance or to allow for early diagnosis of post-transplant complications or graft dysfunction.[14-18]
- The preferred initial imaging is duplex ultrasound with Doppler as this provides readily-available, reliable imaging which is non-invasive and does not require the use of ionizing radiation nor intravenous contrast.[14-18]

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Heart Transplant (AB-42.7)

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- For cardiac transplant imaging indications, see: **Cardiac Imaging in Transplants (CD-16)**

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Hepatic and Abdominal Arteries (AB-43)

Guideline

Hepatic Arteries and Veins (AB-43.1)
Abdominal Veins Other than Hepatic and Portal Veins (AB-43.2)
Renal Vein Thrombosis (AB-43.3)
References (AB-43)

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Hepatic Arteries and Veins (AB-43.1)

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- Portal Vein Thrombosis (PVT):
  - Doppler US (CPT® 93975) is the initial noninvasive modality for the diagnosis of Portal Vein Thrombosis
  - CT Abdomen with contrast (CPT® 74160 or 74170 – 4 phase CT), MRI Abdomen without and with contrast (CPT® 74183) or CTA Abdomen (CPT® 74175) is medically necessary for ANY of the following:
    - to assess the extension of thrombus into the mesenteric veins when Doppler US (or other imaging, such as abdominal US) is positive for PVT
    - to exclude tumor thrombus among individuals with cirrhosis who develop new portal and/or mesenteric vein thrombosis
    - for continued concern for PVT (for example in an individual with a hypercoagulable state or abdominal malignancy) if Doppler US is negative or inconclusive
  - To assess for development of intestinal ischemia among individuals with known portal and/or mesenteric vein thrombosis (MVT) (e.g., development of fever, rebound, leukocytosis, elevated serum lactate levels):
    - In lieu of the above imaging modalities, if requested: CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary.
  - For suspicion of portal hypertensive or portal cavernoma cholangiopathy in individuals with known PVT or MVT (cholestatic liver chemistry profile (See **Abnormal Liver Chemistries (AB-30.1)**), known portal cavernoma, extrahepatic biliary abnormalities on imaging):
    - MRCP (CPT® 74183 or CPT® 74181) is medically necessary

Note: Portosystemic collaterals in the region surrounding the common bile duct in individuals with chronic PVT can be associated with common bile duct obstruction.

- For routine follow-up of PVT:
  - US/Doppler every 6 months. If these are reported as not providing adequate visualization, CT Abdomen (CPT® 74160), MRI Abdomen (CPT® 74183), or CTA Abdomen (CPT® 74175), is medically necessary.
- For follow-up of PVT being treated with anticoagulation:
  - US/Doppler, CT Abdomen (CPT® 74160), MRI Abdomen (CPT® 74183), or CTA Abdomen (CPT® 74175) is medically necessary in 3-6 months.
  - Further follow-up every 6 months with US/Doppler unless these are reported as not providing adequate visualization, in which case any of the above studies can be approved.

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- TIPS (transjugular intrahepatic portosystemic shunt)
  - Pre-procedure evaluation:
    - Abdominal US, including Doppler (CPT® 76700 and/or CPT® 93975), Multiphase CT Abdomen (CPT® 74160 or CPT® 74170), Multiphase CTA Abdomen (CPT® 74175), Multiphase MRA Abdomen (CPT® 74185), or MRI Abdomen liver protocol (CPT® 74183) is medically necessary
    - Echocardiogram (CPT® 93306) (see: **Transthoracic Echocardiography (TTE) – Indications/Initial Evaluation (CD-2.2)**)
  - For routine follow-up to monitor stent patency:
    - US with Doppler (CPT® 93975) is medically necessary 7-14 days after shunt creation, and then at 3 months, 6 months, and then every 6 months thereafter.
      - If requested earlier than the above intervals because of a clinical deterioration or suspicion of stent occlusion, the Doppler is medically necessary.
  - If Doppler imaging is indeterminate or if there is a negative Doppler with clinical signs of worsening portal hypertension the following is medically necessary:
    - Multiphase CT Abdomen (CPT® 74160 or CPT® 74170), Multiphase CTA Abdomen (CPT® 74175), Multiphase MRA Abdomen (CPT® 74185), or MRI Abdomen liver protocol (CPT® 74183)
  - Echocardiogram (CPT® 93306) is medically necessary for the following:
    - One time post-procedure for routine follow up
    - Any time post-procedure:
      - for new signs or symptoms
      - for concern for new or worsening pulmonary hypertension
      - See also: **Frequency of Echocardiography Testing (CD-2.3)** in the Cardiac Imaging Guidelines
- For the evaluation of Budd-Chiari Syndrome
  - Doppler US (CPT® 93975) is the initial diagnostic test for the evaluation of BCS.
  - CT Abdomen with contrast (CPT® 74160), or MRI Abdomen without and with contrast (CPT® 74183) or CTA Abdomen (CPT® 74175) is medically necessary for ANY of the following:
    - to assess thrombus extension
    - to rule out tumor thrombus
    - to assess response to anticoagulation therapy
    - if there is high suspicion of BCS despite a negative or inconclusive Doppler US
    - to additionally assess indeterminate hepatic nodules detected on the prior US (any of the above studies or CT Abdomen without and with contrast CPT® 74170)
  - For pre-operative evaluation of anticipated interventional vascular therapies or TIPS the following is medically necessary:

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        - Abdominal US, including Doppler (CPT® 76700 and/or CPT® 93975), Multiphase CT Abdomen (CPT® 74160 or CPT® 74170), Multiphase CTA Abdomen (CPT® 74175), Multiphase MRA Abdomen (CPT® 74185), or MRI Abdomen liver protocol (CPT® 74183)
    - For HCC Surveillance in individuals with chronic BCS the following is medically necessary:
        - Abdominal US (CPT® 76700 or CPT® 76705) and serum alpha-fetoprotein every 6 months
        - Triphasic CT Abdomen (CPT® 74160 or CPT® 74170), or MRI Abdomen (CPT® 74183) for the evaluation of hepatic nodules seen on US or AFP ≥15 ng/ml.
        - The LiRADS reporting system does not apply to HCC surveillance in this population, due to the vascular origin of many of the hepatic imaging abnormalities.
- Hereditary Hemorrhagic Telangiectasia (HHT)
    - The liver may be involved in individuals with HHT, and artery-to-vein or vein-to-vein shunting may occur resulting in liver vascular malformations (LVMs).
    - Screening the liver for LVMs is not medically necessary.
    - For symptoms suggestive of liver vascular malformations (LVMs) (including an audible bruit or palpable thrill over the hepatic region on physical examination, abnormal liver tests) or for the development of signs or symptoms of heart failure, biliary ischemia, hepatic encephalopathy, mesenteric ischemia, or portal hypertension the following is medically necessary:
        - CT Abdomen (CPT® 74160), CTA Abdomen (CPT® 74175), MRI Abdomen with and without (CPT® 74183), MRCP (CPT® 74183), or MRA Abdomen (CPT® 74185)
- CTA Abdomen and Pelvis (CPT® 74174), or CTA Abdomen (CPT® 74175) or MRA Abdomen (CPT® 74185) is medically necessary additionally for:
    - Evaluation of portal and hepatic veins prior to or following surgical intervention for the treatment of portal hypertension (See: **Portal Hypertension (AB-26.3)**)
    - Evaluation of hepatic vasculature prior to and following embolization procedure (See: **Hepatocellular Carcinoma (HCC) – Restaging/Recurrence (ONC-14.4)** and **Hepatocellular Carcinoma (HCC) – Surveillance/Follow-up (ONC-14.5)** and **Liver Metastases (ONC-31.2)** in the Oncology Imaging Guideline)
    - Evaluation of hepatic vasculature prior to planned hepatectomy (See: **Liver Transplant, Pre-Transplant (AB-42.1)**)
    - Evaluation of liver donor (See: Liver Transplant, **Living Donor Pre-Transplant Imaging (Donor Imaging) (AB-42.2)** for specific guidance)
- Hepatic arterial aneurysms:
    - See: **Visceral Artery Aneurysm (PVD-6.5)** in the Peripheral Vascular Disease Imaging Guidelines

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Background and Supporting Information

Primary Budd-Chiari Syndrome is due to thrombotic occlusion of the hepatic venous outflow tract. Most individuals have an underlying prothrombotic condition such as a myeloproliferative disease, an inherited thrombophilia (e.g. Factor V Leiden), a systemic disease such as vasculitis, or hormonal factors, such as recent oral contraceptive use. Secondary Budd-Chiari Syndrome is caused by malignant tumors or extrinsic compression of the hepatic veins.

LI-RADS assessment should not be applied to individuals <18 years old or those with cirrhosis from congenital hepatic fibrosis secondary to vascular disorders (e.g, Budd-Chiari syndrome, chronic portal vein occlusion, cardiac congestion, hereditary hemorrhagic telangiectasia).

Evidence Discussion

- In cases of Primary Budd-Chiari syndrome, Doppler ultrasound is widely used to evaluate hepatic/portal vasculature. Ultrasonographic evaluation is associated with advantages such as high sensitivity and specificity, and also high positive and negative predictive values.[5]
- Advantages of Doppler ultrasound include low cost, wide availability, and lack of radiation exposure.[5]
- One disadvantage of Doppler ultrasound is its limited ability to evaluate certain anatomies. For instance, it may not be able to detect the extension of portal vein thrombus into splanchnic vessels.[5]
- CT scan is highly accurate in evaluating hepatic vasculature, with sensitivity, specificity, PPV and NPV in the range of 90-99%.[5]
- Advantages of CT scan include better visualization of structures, such as thrombus extension. Another advantage of CT is that it allows for concomitant evaluation of bowel.[5]
- CT scan has drawbacks such as higher cost, radiation exposure, and potential complications from the use of contrast, when compared to ultrasound.[5]
- MRI and MRA may be more appropriate as alternative to CT. Advantages include lack of radiation and a "better safety profile." Disadvantages include longer image acquisition time, higher cost, and various technical limitations., including signal loss, overestimation of stenoses, and contraindications/complications related to implanted metallic devices.[5]
- Pre-TIPS (Transjugular Intrahepatic Portosystemic Shunt), endovascular variceal obliteration or embolization, should ideally include cross-sectional imaging to have an adequate anatomical map of the portal vein and hepatic veins.[17]
- Screening the liver for LVMs is not medically necessary. As per recent ACG Guidelines, "there is no evidence to suggest that making a diagnosis in an asymptomatic individual has clinical benefits or prevents death.[5]

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Abdominal Veins Other than Hepatic and Portal Veins (AB-43.2)

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- CTA Abdomen and Pelvis (CPT® 74174), or CTA Abdomen (CPT® 74175) or MRA Abdomen (CPT® 74185) is medically necessary if ONE of the following:
  - Nephrotic syndrome
  - Renal vein thrombosis
  - Mesenteric vein thrombosis
- Suspicion of iliac vein thrombus when a lower extremity duplex or abdominal duplex is inconclusive or equivocal, see: **Acute Deep Venous Thrombosis (DVT) (PVD 12.2)**
- Suspicion of inferior vena cava thrombus when a lower extremity duplex or abdominal duplex is inconclusive or equivocal, see: **Acute Deep Venous Thrombosis (DVT) (PVD 12.2)**

Evidence Discussion

- Computed Tomography Angiography (CTA) is a diagnostic imaging test that can assess both arterial and venous structures, as well as nonvascular structures in cases of venous thrombosis.[18,19] By combining the evaluation of both vascular and nonvascular findings, it is possible to achieve a sensitivity of 96% and a specificity of 90-94% when assessing for mesenteric venous obstruction.[18]
- In cases of chronic mesenteric venous thrombosis, duplex ultrasound can be a helpful tool for diagnosis. However, due to potential technical difficulties such as overlying bowel gas or limited acoustic windows, imaging may not always be possible.[18,19] In such cases, a CTA scan may be a better option as it allows for a more comprehensive evaluation of both vascular and intestinal structures.[18]
- Contrast-enhanced Magnetic Resonance Angiography (MRA) has been shown to provide a vascular assessment that is comparable to catheter angiography.[1]
- Compared to catheter angiography, MRA is less invasive, cheaper, and does not expose individuals to ionizing radiation.[1]
- Various MRA techniques allow for quantification of blood flow as well as evaluation of oxygen saturation, which are not possible with CTA.[1]
- MRA is less dependent on the operator compared to vascular ultrasound and is less prone to limitations related to individual body habitus or overlying bowel gas.[1]
- Disadvantages of MRA are motion artifact and risk of nephrogenic systemic fibrosis with gadolinium exposure in individuals with severe renal insufficiency.[1]

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Renal Vein Thrombosis (AB-43.3)

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- MRA Abdomen (CPT® 74185) is medically necessary if ONE of the following:
  - Nephrotic syndrome
  - Proteinuria – 3 grams or more in 24 hours
  - Lupus nephritis
  - Hypercoagulable state, ONE of the following:
    - Antiphospholipid antibodies
    - Behçet's syndrome
    - Protein C deficiency
    - Protein S deficiency

Evidence Discussion

- Computed Tomography Angiography (CTA) is a diagnostic imaging test that can assess both arterial and venous structures, as well as nonvascular structures in cases of venous thrombosis.[18,19] By combining the evaluation of both vascular and nonvascular findings, it is possible to achieve a sensitivity of 96% and a specificity of 90-94% when assessing for mesenteric venous obstruction[18]
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- Compared to catheter angiography, MRA is less invasive, cheaper, and does not expose patients to ionizing radiation.[1]
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- MRA is less dependent on the operator compared to vascular ultrasound and is less prone to limitations related to individual body habitus or overlying bowel gas.[1]
- Disadvantages of MRA are motion artifact and risk of nephrogenic systemic fibrosis with gadolinium exposure in individuals with severe renal insufficiency.[1]

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Suspected Neuroendocrine Tumors of the Abdomen (AB-44)

Guideline

Suspected Neuroendocrine Tumors of the Abdomen (AB-44)

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Suspected Neuroendocrine Tumors of the Abdomen (AB-44)

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For the evaluation of a suspected neuroendocrine tumor of the abdomen: See **Gastrointestinal/Pancreatic Neuroendocrine Cancers - Suspected/Diagnosis (ONC-15.2)** in the Oncology Imaging Guidelines.

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Liver Elastography (AB-45)

Guideline

Liver Elastography (AB-45)
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Liver Elastography (AB-45)

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- Initial staging of liver fibrosis in suspected fatty liver disease (hepatic steatosis):
  - Transient Elastography or Vibration-Controlled Transient Elastography (VCTE, e.g. Fibroscan) (CPT® 91200) is medically necessary as the initial imaging modality.
    - Typically repeated within a 3-year period. If repeat transient elastography fails, see MRE criteria below
  - Magnetic Resonance Elastography (MRE, CPT® 76391) is medically necessary for ANY of the following:
    - Transient Elastography failure despite use of an XL-probe, OR BMI ≥35
    - Conflict between clinical picture and transient elastography results (e.g., individual with portal hypertension but VCTE suggests no fibrosis)
    - VCTE liver stiffness measurement of ≥8 kPa
    - FIB 4 score of >2.67
    - Liver biopsy demonstrates fibrosis stage F2-F4
- Special considerations for MRE:
  - For MRE requests in the setting of hemochromatosis, see: **Hereditary (Primary) Hemochromatosis (HH) and Other Iron Storage Diseases (AB-11.2)**)
  - For individuals for whom drug treatment is a consideration and EITHER of the following:
    - Evaluation for liver stiffness as part of required assessment for drug candidacy
    - Evaluation for liver stiffness to assess treatment efficacy as required by the drug manufacturer and/or FDA
  - Note: The correct CPT code for MR Elastography is CPT® 76391. It is a stand-alone code and it does not require an additional CPT code such as MRI Abdomen (CPT® 74183).
  - An additional MRI Abdomen code should only be approved if there is another appropriate indication for it, other than the Elastography study (for example, MRE for fibrosis scoring in MASLD (formerly known as NAFLD) due to a BMI ≥35, AND further evaluation of an indeterminate hepatic lesion).
- The use of other ultrasound elastographic codes (CPT® 76981, CPT® 76982, and CPT® 76983) is not medically necessary.

Background and Supporting Information

- For the assessment of fibrosis in chronic liver disease, the American Gastroenterological Association recommends Vibration Controlled Transient Elastography (VCTE) as the preferred imaging modality over MR Elastography

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(MRE), except in the case of MASLD (formerly known as NAFLD) in high-risk populations.

- Transient Elastography (VCTE) is the most studied elastography technique and informs multiple evidence-based guidelines with respect to fibrosis scoring. No national evidence-based guideline recommends the use of either Acoustic Radiation Force Impulse Imaging (ARFI) or real-time tissue elastography (RTTE) over the use of VCTE for any clinical protocol, nor is there direct evidence that ARFI or RTTE improves health outcomes over and above VCTE.
- Vibration-Controlled Transient Elastography (VCTE) (e.g. Fibroscan, CPT® 91200) may be considered medically necessary to assess for advanced fibrosis and cirrhosis in conditions including:
  - Hepatitis C
  - Hepatitis B
  - Chronic alcoholic liver disease
  - All other chronic liver diseases
- Liver fibrosis staging: The most common staging system is METAVIR, which uses a scale of F0 to F4 (F0 indicates no fibrosis, while F4 represents cirrhosis). Other staging systems include Ishak and Batts-Ludwig, which also assess fibrosis based on histological examination of liver tissue. For Ishak Scoring System: Stages 0-5: Progressively more severe fibrosis, with stage 5 being the most advanced before cirrhosis; Stage 6: Cirrhosis. For Batts-Ludwig Scoring System: Grades 0-4: Progressively more severe fibrosis, with grade 4 being cirrhosis.
- FIB-4 index is calculated as follows[22]:
  - FIB-4 = (Age in years x AST level)/(Platelet count x √ of ALT)

Evidence Discussion

Targeted screening of populations at increased risk for advanced liver disease is advised to identify and manage those with clinically significant fibrosis.[10,22,23,26]

Although liver biopsy remains the reference standard for the grading and staging of nonalcoholic steatohepatitis (NASH), it has important limitations related to risk, cost, and sampling error. Noninvasive biomarkers are emerging as valuable tools for predicting adverse liver-related outcomes.[10,22,23,26]

The most validated laboratory-based fibrosis biomarker is FIB-4, which outperforms other calculations in its ability to identify individuals with a low probability of advanced fibrosis. A FIB-4 score > 2.67 is associated with a high risk of advanced fibrosis.[10,22,23,26]

Liver stiffness is a physical characteristic of the liver that increases with fibrosis severity. Vibration Controlled Transient Elastography (VCTE), e.g., Fibroscan, is the most commonly used method to assess liver stiffness. Transient elastography (VCTE) is the

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most studied elastography technique and informs multiple evidence-based guidelines with respect to fibrosis scoring. No national evidence-based guideline recommends the use of either ARFI or real-time tissue elastography (RTTE) over the use of VCTE for any clinical protocol, nor is there direct evidence that ARFI or RTTE improves health outcomes over and above VCTE. VCTE-derived liver stiffness measurement (LSM) of < 8 kPa can be used to rule out advanced fibrosis, especially if used with FIB-4. An LSM between 8 and 12kPa may be associated with fibrotic NASH, and a value > 12 kPa is associated with a high likelihood of advanced fibrosis.[10,22,23,26]

For the assessment of cirrhosis in individuals with hepatitis C, the American Gastroenterological Association (AGA) noted that VCTE was recommended over MRE with the exception of NAFLD in high-risk populations, in which MRE resulted in a lower rate of false positives compared to VCTE.[10,22,23,26]

Magnetic Resonance Elastography (MRE) is not the first-line approach for risk stratification, however it becomes an important tool when clinical uncertainty exists, concomitant cross-sectional imaging is needed, there is a discrepancy between the clinical picture and VCTE results, or when VCTE is unavailable or limited by individual factors.[10,22,23,26]

Liver stiffness measurements can provide important context when evaluating individuals for potential drug therapy. While not a formal requirement, imaging-based fibrosis assessment is often part of a comprehensive clinical evaluation.[27]

Liver stiffness can also be used to monitor response to therapy over time. For individuals undergoing treatment with Resmetirom, a follow-up elastography assessment at 12 months may help determine whether the therapy is effective or if adjustments are needed.[27]

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Hiccups (AB-46)

Guideline

Hiccups (AB-46.0)
References (AB-46)

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Hiccups (AB-46.0)

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- Hiccups may be associated with cerebrovascular disease, brain tumors, and intracranial injury, though it would be very rare for hiccups to be the only presenting symptom of serious neurologic disease. If concern is expressed for neurologic involvement, please see the appropriate guideline in HD imaging (e.g., **Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)** and **Anti-MOG syndromes (HD-16.3)**)
- Hiccups <48 hours without any localizing or specific symptoms:
  - No advanced imaging is medically necessary
- Hiccups ≥48 hours:
  - History and physical examination, laboratory and CMP and baseline chest x-ray
  - Abnormal or negative chest x-ray with symptoms referable to the chest:
    - CT Chest with contrast (CPT® 71260) is medically necessary
  - Lab or history/physical findings suggest a gastrointestinal etiology:
    - CT Abdomen with contrast (CPT® 74160) is medically necessary

Evidence Discussion

If there are additional signs or symptoms to evaluate, further testing is medically necessary. CT Chest and/or bronchoscopy is the study of choice for evaluation wheezing, dyspnea, abnormal chest radiography, or abnormal pulmonary function tests. MRI Brain and/or lumbar puncture are indication for potential central nervous system causes. Evaluation of esophageal and other symptoms is performed with upper endoscopy, esophageal manometry, and/or CT Abdomen. Cardiac etiologies may be evaluated with EKG & Echo.[3-6]

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Retroperitoneal Fibrosis (AB-47)

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Retroperitoneal Fibrosis (AB-47.0)
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Retroperitoneal Fibrosis (AB-47.0)

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- Individuals diagnosed with retroperitoneal fibrosis:
  - ONE of the following every 3 months is medically necessary until stability demonstrated:
    - CT Abdomen and Pelvis with contrast (CPT® 74177)
    - MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195)
    - MRI Abdomen and Pelvis with and without contrast (CPT® 74183 and CPT® 72197)
    - Retroperitoneal or Abdominal ultrasound (CPT® 76770 or CPT® 76700) can be considered medically necessary if requested.
  - After stability established repeat imaging is medically necessary every 6 months.
  - Requests for non-contrasted studies in individuals with renal insufficiency is medically necessary. Gadolinium may induce nephrogenic systemic fibrosis in individuals with moderate or severe renal insufficiency, especially if the GFR is <30 ml/min.
  - Additional imaging:
    - CT Chest (CPT® 71260) is also medically necessary upon initial diagnosis if requested, to further evaluate for the possibility of malignancy as an underlying etiology.
- PET/CT (CPT® 78815)
  - Can be considered medically necessary initially, after diagnosis, to establish avidity patterns to assess for the likelihood of malignancy and for stratification for the likelihood of response to steroids.
  - Follow-up is considered medically necessary if there is documentation of an anticipated therapeutic change based on the results (such as a change in immunosuppression therapy or stent removal).
- Methysergide-induced retroperitoneal fibrosis:
  - Methysergide for migraine treatment is generally no longer available but is rarely being used at some centers. It has a known complication of retroperitoneal fibrosis.
  - Individual screening is medically necessary at baseline and then every 6 months with ONE of the following:
    - CT Abdomen and Pelvis with contrast (CPT® 74177)
    - CT Abdomen and Pelvis without contrast (CPT® 74176)
    - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
    - MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195)

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- Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775)

Background and Supporting Information

Retroperitoneal fibrosis is a rare disease, and may be idiopathic (IgG4 or non-IgG-4 related) or secondary. Secondary causes include malignancy, infections, previous radiation therapy, previous abdominal surgery, drugs such as methysergide, and biologic agents.

Evidence Discussion

- Ultrasound may be used as a screening tool, but has low sensitivity and is often insufficient to distinguish retroperitoneal fibrosis from other abdominal masses.[2]
- CT and MR allow for characterizing morphology and extent of retroperitoneal fibrosis both at initial diagnosis and in treatment monitoring. It also helps to define the involved vascular structures, and can visualize disease in other abdominal viscera that may be associated with retroperitoneal fibrosis. CT may have advantages in imaging availability and imaging time. MR may have advantages in avoiding ionizing radiation and improved soft tissue characterization.[1]
- PET may be used to evaluate metabolic activity and may be of value after diagnosis to characterize active inflammation versus malignancy and to document response to treatment. The role of PET scan in establishing a diagnosis is limited due to the potential for nonspecific uptake.[1,6,7]
- Follow-up may be appropriate every 3-12 months to access disease status and response to therapy.[1]

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Fistulae (AB-48)

Guideline

Fistulae (AB-48)
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Fistulae (AB-48)

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- Suspected enteric fistulae
  - ONE of the following is medically necessary:
    - MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195), OR
    - CT Enterography or CT Abdomen and Pelvis with contrast (CPT® 74177), OR
    - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
- Suspected colovesical fistulae
  - ONE of the following is medically necessary:
    - CT Abdomen and Pelvis without contrast (CPT® 74176), OR
    - MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195), OR
    - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
- Enterocutaneous fistulae
  - Suspected enterocutaneous fistulae or surgical planning of known complex fistulae:
    - ONE of the following is medically necessary:
      - CT Abdomen and Pelvis with contrast (CPT® 74177), OR
      - MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195), OR
      - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT®72197)
- Complicated diverticulitis with fistula, see: **Lower Abdominal Pain (AB-2.2)**
- Perianal/perirectal fistulae and abscess related to Crohn's disease, see: **Perirectal/ Perianal Crohn's Disease (AB-23.3)**
- Other fistulae related to Crohn's disease, see: **Known IBD (AB-23.2)**
- Perianal/perirectal fistulae NOT related to Crohn's disease, see: **Fistula in Ano (PV-21.1)** in the Pelvis Imaging Guidelines
- For colovaginal, rectovesicular, rectovaginal, or urinary-vaginal communicating fistulae, see: **Pelvic Fistula (PV-21.3)** in the Pelvis Imaging Guidelines
- For pilonidal cyst, see: **Pilonidal Cyst (PV-21.4)** in the Pelvis Imaging Guidelines

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Background and Supporting Information

- Examples of gastrointestinal fistulae include tracheo- and broncho-esophageal, entero-cutaneous, entero-enteric, entero-colic, entero-vesical, colo-vesical, recto-vaginal, perianal, and aorto-enteric.
- Etiologies of fistulae include: complication of inflammatory disease (e.g., Diverticulitis, Crohn's disease), complication of surgical procedures (which are the most common cause of intestinal fistula, comprising more than half of all fistulae), obstetric injury (e.g., recto-vaginal, ano-vaginal), malignancy, radiation, non-surgical injuries, and foreign bodies.

Evidence Discussion

Magnetic resonance imaging (MRI) and small intestine contrast enhanced ultrasonography (SICUS) have now emerged as suitable radiation-free alternatives to CT imaging, with comparable diagnostic accuracy. MRI is often considered the imaging modality of choice for evaluation of fistulae owing to its superior soft-tissue contrast and ability to provide surgeons with the highest quality information derived from just one study, including anatomic location of fistulae and associated pelvic pathology.[1,2,11,12,13]

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General Considerations (BR-Preface 1)

Guideline

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Abbreviations for Breast Guidelines

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Abbreviations for Breast Guidelines	
BI-RADS™	Breast Imaging Reporting and Database System
BRCA	breast cancer gene
CAD	computer-aided detection
CT	computed tomography
CTA	computed tomography angiography
CTV	computed tomography venography
DCIS	ductal carcinoma in situ
FDA	Food and Drug Administration
FDG	fluorodeoxyglucose
FNA	fine needle aspiration
HRCT	high-resolution computed tomography
LCIS	lobular carcinoma in situ
MRA	magnetic resonance angiography
MRI	magnetic resonance imaging
NSM	nipple-sparing mastectomy
PEM	positron-emission mammography
PET	positron-emission tomography

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General Guidelines (BR-Preface 1.0)

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- A current clinical evaluation since the onset or change in symptoms is usually required prior to considering advanced imaging.
  - A clinical evaluation should include the following:
    - A relevant history and physical examination since the onset or change in symptoms
    - Appropriate laboratory studies and non-advanced imaging modalities, such as mammogram and/or ultrasound
    - Other meaningful contact (telephone call, electronic mail or messaging) since the onset or change in symptoms by an established individual can substitute for a face-to-face clinical evaluation
- Current clinical evaluation is not required prior to screening studies.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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BI-RADS™ Categories Chart (BR-Preface 1.1)

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BI-RADS™ Categories Chart	
Category	**Description**
Category 0: Incomplete	Need additional imaging evaluation or prior mammograms for comparison. Category 0 classification requires that additional imaging study be specified, e.g., ultrasound, additional mammogram view, MRI.
Category 1: Negative	There is nothing to comment on. The breasts are symmetrical and no masses, architectural disturbances, or suspicious calcifications are present.
Category 2: Benign Finding	This is also a negative mammogram, but the interpreter may wish to describe a finding. Involuting, calcified fibroadenomas, multiple secretory calcifications, fat-containing lesions (such as oil cysts, lipomas, galactoceles, and mixed density hamartomas) all have characteristic appearances, and may be labeled with confidence. The interpreter might wish to describe intramammary lymph nodes, implants, etc. while still concluding that there is no mammographic evidence of malignancy.

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BI-RADS™ Categories Chart	
Category	**Description**
Category 3: Probably Benign Finding – Short Interval Follow-up Suggested	A finding placed in this category should have a very high probability of being benign. It is not expected to change over the follow-up interval, but the radiologist would prefer to establish its stability. Data is becoming available that sheds light on the efficacy of short interval follow-up. At the present time, most approaches are intuitive. These will likely undergo future modification as more data accrue as to the validity of an approach, the interval required, and the type of findings that should be followed.
Category 4: Suspicious Abnormality – Biopsy Should Be Considered	There are lesions that do not have the characteristic morphologies of breast cancer but have a definite probability of being malignant. The radiologist has sufficient concern to urge a biopsy. If possible, the relevant possibilities should be cited so that the individual and their physician can make the decision on the ultimate course of action.
Category 5: Highly Suggestive of Malignancy – Appropriate Action Should Be Taken	These lesions have a high probability of being cancer and should be biopsied or treated surgically.
Category 6: Known Biopsy-Proven Malignancy – Appropriate Action Should Be Taken	These lesions have been biopsied and are known to be malignant.

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BI-RADS™ Breast Density Categories (BR-Preface 1.2)

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BI-RADS™ Breast Density Categories
Category A: Almost entirely fatty
Category B: Scattered fibroglandular densities
Category C: Heterogeneously dense
Category D: Extremely dense

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Breast Ultrasound (BR-1)

Guideline

Breast Ultrasound (BR-1.1)

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Breast Ultrasound (BR-1.1)

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- Routine performance of breast ultrasound (US) as stand-alone screening or with screening mammography is not medically necessary.
  - Breast ultrasound is a supplemental screening alternative for high-risk females (as described in **MRI Breast Indications [BR-5]**) with dense breasts on mammography, when MRI Breast without and with contrast cannot be performed. The inability to perform MRI Breast may be because it cannot be tolerated (i.e., insurmountable claustrophobia or body habitus), or there exists a contraindication (i.e., non-MRI compatible implantable devices or an inability to receive MRI contrast).
  - Breast ultrasound (CPT® 76641 or CPT® 76642) is medically necessary when requested by the treating provider to complete the screening process, OR recommended by the radiologist report, OR to address a finding on the mammogram.
- Breast ultrasound (CPT® 76641: unilateral, complete; or, CPT® 76642: unilateral, limited) can be used to further evaluate abnormalities found on mammogram, especially in differentiating cysts from solid lesions.
  - A clinical office visit is not necessary prior to breast ultrasound when an abnormality has been identified on a mammogram.
- BI-RADS™ Cat 3 ultrasound follow-up imaging for stable findings at 6 months:
  - if repeat imaging remains BI-RADS™ 3, repeat at 12 months, 18 months, and 24 months from the date of the initial imaging.
    - After 2 years of stability, the finding should be assessed as benign (Cat 2).
  - if repeat imaging is BI-RADS™ 1 or 2, then imaging reverts to routine per individual's risk profile.
- For breast implant imaging, please see **Breast Implant Evaluation (BR-5.2)**.
- Axilla ultrasound (CPT® 76882)
  - For individuals with clinically suspicious lymph nodes, pre-operative axillary ultrasound with a FNA or biopsy can help identify individuals who have positive nodes.
    - See **Axillary Lymphadenopathy (and Mass) (CH-2.2)** in the Chest Imaging Guidelines.
  - Bilateral should be coded CPT® 76882 x 2.
- US-guided breast biopsy (CPT® 19083) includes the imaging component.
  - Additional lesions should be billed using CPT® 19084.

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- Breast ultrasound (CPT® 76641 or CPT® 76642) can be repeated at least 6 months after an US-directed breast biopsy to document successful lesion sampling if histology is benign and non-specific, equivocal or uncertain.
- 3D Reconstruction (CPT® 76376 or CPT® 76377) is **NOT** medically necessary for breast ultrasound. It is commonly requested in conjunction with automated breast ultrasound (ABUS); there is no evidence to support its clinical usefulness.

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MRI Breast Coding (BR-2)

Guideline

MRI Breast Coding (BR-2.1)

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MRI Breast Coding (BR-2.1)

BR.MR.0002.1.A

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- The use of gadolinium contrast is required for the evaluation of breast parenchyma.
- The use of gadolinium contrast is **NOT** necessary for the evaluation of implant integrity in asymptomatic, average-risk individuals.
- Throughout this guideline, when MRI Breast is medically necessary, any **ONE** of the following codes is supported:
  - CPT® 77049 MRI Breast Bilateral, including CAD, without and with contrast
  - HCPCS C8908 MRI Breast Bilateral, without and with contrast
- If the individual meets medical necessity for advanced imaging to assess implant integrity, the appropriate code is CPT® 77047 MRI Breast Bilateral, without contrast.
- Computer-aided detection (CAD) is included with the MRI Breast CPT® 77049 and CPT® 77048 procedures.
  - The use of CAD has little influence on the sensitivity and specificity of MRI Breast interpretation.
  - Since the CAD software automatically performs 3D imaging, CPT® 76376 or CPT® 76377 should **NOT** be used in conjunction with MRI Breast.
- MRI-guided breast biopsy (CPT® 19085) includes the imaging component and the needle placement under MR guidance; CPT® 77021 MR guidance for needle placement is **NOT** an appropriate code to bill for a breast biopsy.
  - Additional lesions should be billed using CPT® 19086.
  - This program does not manage CPT® codes 19085 or 19086.

Background and Supporting Information

- Although MRI Breast has superior sensitivity in identifying new unknown malignancies, it carries a significant false positive risk when compared to mammogram and ultrasound. Incidental lesions are seen on 15% of MRI Breast and increase with younger age. The percentage of incidental lesions that turn out to be malignant varies from 3% to 20% depending on the individual population. Cancer is identified by MRI Breast in only 0.7% of those with “inconclusive mammographic lesions.”

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Breast Reconstruction (BR-3)

Guideline

Breast Reconstruction (BR-3.1)

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Breast Reconstruction (BR-3.1)

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- CTA or MRA of the body part **from which the free-tissue transfer flap is being taken**, can be performed for breast reconstruction pre-operative planning.
  - Examples include:
    - CTA Abdomen and/or Pelvis (CPT® 74175 or CPT® 72191 or CPT® 74174) or MRA Abdomen and/or Pelvis (CPT® 74185 and/or CPT® 72198) for Deep Inferior Epigastric Perforators (DIEP) flap
    - CTA Chest (CPT® 71275) for Thoracodorsal Artery Perforator (TDAP) flap
- Routine use of CTA Chest (CPT® 71275) to evaluate **recipient** vessels is **NOT** medically necessary.
  - **Criteria exception**: In circumstances where there has been previous cardiac/vascular surgery and/or known vascular anomalies in the chest, it may be warranted.
- There is currently insufficient evidence-based data to support the need for routine advanced imaging for TRAM flaps or other flaps performed on a vascular pedicle.

Evidence Discussion

The American College of Radiology (ACR) Appropriateness Criteria® stated that either MRA abdomen and pelvis with and without IV contrast or CTA abdomen and pelvis with IV contrast are usually appropriate for pre-operative planning in individuals undergoing DIEP flap breast reconstruction.[2] Studies have found CTA mapping results in a shorter operative time when compared with no mapping in cases of breast reconstruction with free-tissue flap transfer (e.g., with Deep Inferior Epigastric Perforator (DIEP) flaps).[1]

In contrast, routine use of CTA chest to evaluate for recipient vessels (often the internal mammary vessels) is not medically necessary. This is because a number of studies have found that the anatomy and course of these vessels is largely consistent, and that there is good concordance between surgical and radiological findings – either with ultrasound or CTA.[3] CTA, however, carries with it significant risks, including contrast nephrotoxicity and allergic reactions, and the significantly higher risk of radiation exposure in the chest than in the abdomen.[4] As such, many surgeons will use hand-held Doppler ultrasound either pre- and/or intra-operatively to evaluate recipient vessels. In certain circumstances, such as with previous surgery and/or radiation that would be expected affect the candidacy of potential recipient vessels, pre-operative CTA of the chest may be considered.

As pedicled flaps, by definition, do not require a microvascular anastomosis and are not disconnected from their blood supply, there is no current evidence to support routine pre-operative imaging in these individuals. A recent study evaluating the use of pre-

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operative CTA in individuals undergoing pedicled TRAM flap reconstruction found that there was no significant difference in terms of operative time nor flap loss in individuals who had a pre-operative CTA compared those who did not.[5]

Per the National Comprehensive Cancer Network (NCCN), "common donor sites for autologous tissue include the abdomen (ie, DIEP, MS TRAM [Muscle-Sparing Transverse Rectus Abdominis Myocutaneous], SIEA [Superficial Inferior Epigastric Artery], free TRAM, pedicled TRAM), gluteal region (ie, SGAP [Superior Gluteal Artery Perforator], IGAP [Inferior Gluteal Artery Perforator]), thigh (ie, TUG [Transverse Upper Gracilis], VUG [Vertical Upper Gracilis], DUG [Diagonal Upper Gracilis], PAP [Profunda Artery Perforator]), or the back (ie, LD [Latissimus Dorsi], TDAP, LAP [Lumbar Artery Perforator])".[46]

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MRI Breast Indications (BR-5)

Guideline

MRI Breast Indications (BR-5.1)
Breast Implant Evaluation (BR-5.2)

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MRI Breast Indications (BR-5.1)

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MRI Breast Considerations

- When MRI Breast imaging is medically necessary (per the criteria listed in the sections below), an MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary.
- MRI Breast Unilateral is **NOT** clinically supported.
- See **Breast Ultrasound (BR-1)** when there is a contraindication to MRI contrast.
- See **MRI Breast Coding (BR-2)** for MRI-guided breast biopsy.
- See **Breast Cancer (ONC-11)** in the Oncology Imaging Guidelines for imaging indications related to breast cancer as follows:
  - Breast Cancer - Initial work-up/Staging
  - Breast Cancer - Restaging/Recurrence
  - Breast Cancer - Surveillance/Follow-up
  - Annual screening with prior history of breast cancer

MRI Following a Screening Mammogram and/or US in Asymptomatic Individuals

- MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary for **EITHER** of the following:
  - When requested by the treating provider to complete the screening process, OR recommended by the radiologist report, OR to address a finding on the mammogram.
  - Documented histopathologic discordance between core-needle biopsy findings and imaging findings. MRI Breast is medically necessary for further evaluation **after** the discordant biopsy (before consideration for surgical management vs. observation).
    - Discordance exists when the biopsy result does not adequately explain the abnormal (BI-RADS™ 4 or 5) findings on mammogram and/or ultrasound.
- For symptomatic individuals, please refer to the following appropriate condition-based guideline:
  - **Nipple Discharge/Galactorrhea (BR-6.1)**
  - **Breast Pain (Mastodynia) (BR-7.1)**
  - **Breast Imaging in Males (BR-9.1)**
  - **Breast Mass (BR-14.1)**
  - **Skin Changes (BR-15.1)**
  - **Nipple Inversion/Retraction (BR-16.1)**
  - **Malignant Phyllodes Tumor/Cystosarcoma Phyllodes (BR-17.1)**

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- See **MRI BI-RADS™ 3** section for lesions categorized as BI-RADS™ 3 on MRI.

MRI BI-RADS™ 3

- A probably benign lesion on **MRI** (MRI BI-RADS™ 3) should undergo repeat MRI in 6 months.
  - If repeat imaging remains MRI BI-RADS™ 3, then repeat at 12 months, 18 months, and 24 months from the date of the initial imaging.
    - After 2 years of stability, the finding should be assessed as benign (Cat 2)
  - If repeat imaging is BI-RADS™ 1 or 2, then imaging reverts to routine per individual's risk profile. See **Risk Factors** section.

Post-Biopsy or Attempted Biopsy Imaging

- For lesions initially seen on MRI Breast and that have benign and non-specific, equivocal or uncertain histology (based on a stereotactic, MRI-guided, or US-directed breast biopsy), an MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary at least 6 months after the biopsy to document successful lesion sampling.
- MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary 6 months after attempted MRI-guided breast biopsy, when recommended by a radiologist, due to targeted lesion not visualized at the time of the procedure.

Risk Factors

- Routine MRI Breast following bilateral mastectomy is **NOT** medically necessary (even if high-risk screening criteria may otherwise be met and/or nipple-sparing mastectomy was done).
- Annual MRI Breast screening with MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary for individuals meeting the high-risk criteria in the table below (for male breast imaging, please see **Breast Imaging in Males (BR-9.1)**):

High-Risk Indications	Age at which screening can start**
Genetic Mutations:*	
Li Fraumeni (p53)	20
BRCA 1 or 2	25
STK11, Peutz-Jeghers syndrome (PJS), PTEN Mutation (Cowden Syndrome), CDH1, NF1, PALB2, ATM, CHEK2	30**

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High-Risk Indications	Age at which screening can start**
BARD1, RAD51C, RAD51D	40**
Personal history of atypia/LCIS/breast cancer:	
ADH, ALH, LCIS	At diagnosis but not prior to age 25
Personal history of breast cancer at or before the age of 50	At diagnosis
Family history:	
If the individual has **NOT** been tested for BRCA mutation **and** there is a first-degree relative (parent, sibling, child; half siblings are considered second-degree relatives) with BRCA 1 or BRCA 2 mutation. Annual screening is **NOT** medically necessary if the individual has been tested and is negative for BRCA 1 or BRCA 2 mutation unless they meet other criteria.	40**
Two or more first-degree relatives with breast or ovarian cancer	40**
One first-degree relative with breast cancer or ovarian cancer that was diagnosed ≤ age 50	40**
One first-degree relative with bilateral breast cancer, or both breast and ovarian cancer	40**
A first- or second-degree male relative (father, brother/half-brother, uncle, grandfather) diagnosed with breast cancer	40**
Elevated clinical lifetime-risk:	
Clinical lifetime-risk estimated at greater than or equal to 20% as calculated by one of the following models: • Gail (National Cancer Institute (NCI)) • Tyrer-Cuzick (International Breast Cancer Intervention Study (IBIS)) • The Breast Cancer Surveillance Consortium (BCSC) • Breast and Ovarian Analysis of Disease Incidence and Carrier Estimation Algorithm (BOADICEA)/CanRisk • BRCAPRO Model	40**
Personal history of radiation therapy when younger than age 30:	

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High-Risk Indications	Age at which screening can start**
Radiation to chest, whole lung, mediastinum, axilla, mantle (including mini mantle or extended mantle), total or subtotal lymphoid irradiation or total body irradiation (TBI)	25 or 8 years after completion of radiation therapy *whichever comes later*
Breast Density:	
Heterogeneously Dense Breasts (Category C) or Extremely Dense Breasts (Category D) with no additional risk factors	40

*The following have unknown or insufficient evidence of breast cancer risk and additional MRI screening is NOT medically necessary at this time: MSH2, MLH1, MSH6, PMS2, EPCAM, NBN, genetic variants of unknown significance, genetic variants favoring polymorphism, and genetic variants of intermediate penetrance. Any gene mutation not specified in the table above has not currently been found to have sufficient evidence to support surveillance with MRI.

OR 10 years prior to the age of diagnosis of the earliest relative with breast cancer (first-, second-, and third-degree relatives) *whichever comes first*, **but not before age 25

Background and Supporting Information

- myRisk® Hereditary Cancer (Myriad Genetics, Inc.) is not accepted as a risk calculator to determine high-risk for breast cancer.

Evidence Discussion

High-Risk Indications

Li-Fraumeni Syndrome is associated with an increased incidence of premenopausal breast cancer, with the median age of diagnosis being in the early 30s.[10] Accordingly, the National Institute for Health and Care Excellence (NICE)[9] recommended annual MRI screening beginning at age 20.[9]

While the American Cancer Society (ACS) found that there is not enough evidence to make a recommendation for or against screening MRI in these populations[6], the NCCN has recommended annual breast MRI for those with ADH, ALH or LCIS who have at least a 20% residual lifetime risk of developing breast cancer. Screening could begin at the age of diagnosis of ADH or lobular neoplasia, but not before the age of 25. They further noted that the residual lifetime risk calculation depends on the age at diagnosis.[7]

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BRCA1 and *2* are associated with a risk of developing breast cancer >60%.[8] The NCCN guidelines recommended starting MRI screening at the age of 25.[8]

STK11 mutations are associated with a 32%-54% risk of developing primary breast cancer. *CDH1* and *PALB2* mutations each confer a risk of 41%-60% of developing breast cancer. NCCN guidelines recommended starting MRI screening in these individuals at age 30. For individuals with NF1, the risk of developing breast cancer is 20%-40%. NCCN guidelines recommended considering annual MRI screening from ages 30-50. *ATM* mutations are associated with a 20%-30% risk of developing breast cancer, and *CHEK2* mutations similarly are associated with a 20%-40% risk. NCCN guidelines suggested consideration of annual breast MRI starting at age 30-35 in both of these groups. *PTEN* mutations are associated with a 40%-60% risk of developing breast cancer. While NCCN guidelines are silent on breast cancer screening for this population, ESMO guidelines recommended starting annual MRI at the age of 30.[8,11]

BARD1, *RAD51C* and *RAD51D* are each associated with a 17%-30% risk of developing breast cancer. The NCCN guidelines recommended considering an annual breast MRI starting at age 40.[8]

However, mutations and variants with a <15% absolute risk of developing breast cancer lack sufficient evidence to suggest that screening MRI would be beneficial. Therefore, the NCCN did not recommend screening MRI for these individuals unless other risks are present.[8]

The ACR Appropriateness Criteria® for "Female Breast Cancer Screening" had noted that "some females with a personal history of breast cancer may also fit into the high-risk category, particularly those diagnosed before 50 year of age...".[42] They also went on to state that these women may have a greater than 20% estimated lifetime risk of another breast cancer diagnosis.[42] The NCCN also noted that MRI Breast for screening is recommended annually for individuals diagnosed with breast cancer at or before age 50[46] who have not undergone bilateral mastectomy (see the Postmastectomy Imaging section below).

The ACS considered individuals who have a first-degree relative with a BRCA 1 or 2 gene mutation and who have not been tested themselves to be at high risk. They recommended an annual MRI screening starting at age 30.[6] On the other hand, NCCN guidelines suggested that untested individuals with a first-degree relative with a BRCA 1 or 2 mutation should start screening either 10 years before the youngest family member was diagnosed with breast cancer, but not before age 25, or at age 40, whichever comes first.[7]

Per NCCN recommendations, BRCAPRO, Tyrer- Cuzick, Breast Cancer Surveillance Consortium (BCSC), and Breast and Ovarian Analysis of Disease Incidence and Carrier Estimation Algorithm (BOADICEA)/CanRisk are appropriate models used to calculate clinical lifetime-risk.[46]

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The NCCN has issued guidance that recommended individuals with extremely dense breast tissue on mammogram begin screening with MRI Breast at age 50, but also notes that "consideration can be given to start at age 40 based on individual risk factors".[7] The Updated Recommendations from ACR also addressed the use of MRI Breast in individuals with dense breast tissue for supplemental screening. They did not differentiate between heterogeneously and extremely dense breasts in their recommendation and instead, recommended screening for those with dense breasts starting at age forty.[84] ACR considers dense breasts to be heterogeneously dense (Category C) and extremely dense (Category D).[85]

MRI utilizes a magnetic field and radio waves with computer processing to produce detailed images whereas CT uses ionizing radiation. Radiation dosages vary based on many factors and can be harmful to tissues. Thus, from a radiation safety perspective, MRI should be utilized when appropriate and supported by existing literature; however, the NCCN also acknowledged potential harms of MRI use, such as increased false positives, increased recall, and increased benign biopsies.[7]

Post-Biopsy or Attempted Biopsy Imaging

A study conducted by Pinnamaneni et al showed that of the 89 biopsies that were canceled secondary to nonvisulaization, 74% of lesions resolved by 6 month follow-up, however 1.9% yielded carcinoma at the 6 month follow-up. Pinnamaneni et al. concluded that, "the majority of canceled MRI-guided biopsy lesions resolved on later follow-up; however, because of the small possibility of a missed malignancy, follow-up MRI imaging at 6 months is recommended".[90]

The initial evaluation of individuals who present with a suspicious finding on breast imaging or a palpable mass upon examination involves a biopsy (percutaneous or surgical if percutaneous is not feasible). If the biopsy results are discordant with the imaging findings, an MRI for further evaluation is supported.[16]

Postmastectomy Imaging

According to the ACR Appropriateness Criteria® for "Imaging after Mastectomy and Breast Reconstruction", there is not enough evidence to support MRI imaging for breast cancer screening following a bilateral mastectomy.[73] In addition, in a study by Weed et al, it was found that "the use of surveillance MRI after NSM [nipple-sparing mastectomy] lead to increased rates of biopsy without improvement in overall survival in our study".[88]

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Breast Implant Evaluation (BR-5.2)

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Breast Implant Imaging

- Breast MRI is **NOT** medically necessary for evaluation of capsular contracture.
- Imaging for routine surveillance and/or suspected rupture of breast implants is dependent upon the type of implant. Please see below.

SALINE

Asymptomatic Screening in Females or Transfeminine Individuals

- For all ages, routine imaging is not medically necessary.

Exam Equivocal for Rupture in Females or Transfeminine Individuals

- If less than 30 years old, breast ultrasound (CPT® 76641 or CPT® 76642) is medically necessary.
- If 30 years old or older, breast ultrasound (CPT® 76641 or CPT® 76642) or diagnostic mammogram is medically necessary.
- If breast ultrasound or diagnostic mammogram results are indeterminate for saline implant rupture, additional imaging with MRI Breast Bilateral without contrast (CPT® 77047) is medically necessary for further evaluation.

SILICONE

Asymptomatic Screening in Females or Transfeminine Individuals

- For all ages, if it is less than 5 years since the implants were placed, routine advanced imaging is not medically necessary.
- For all ages, if it has been 5 years or more since the implants were placed, breast ultrasound (CPT® 76641 or CPT® 76642) is considered medically necessary.
  - Further evaluation with MRI Breast Bilateral without contrast (CPT® 774047) is medically necessary if the breast ultrasound is indeterminate.
  - Repeat breast ultrasounds (CPT® 76641 or CPT® 76642) can be done every 2 to 3 years after initial negative imaging.

Exam Equivocal for Rupture in Females or Transfeminine Individuals

- For all ages, breast ultrasound (CPT® 76641 or CPT® 76642) or diagnostic mammogram or MRI Breast Bilateral without contrast (CPT® 77047) is medically necessary.
- If breast ultrasound (CPT® 76641 or CPT® 76642) or diagnostic mammogram results are indeterminate for silicone implant rupture, additional imaging with MRI Breast Bilateral without contrast (CPT® 77047) is medically necessary for further evaluation.

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Evidence Discussion

Breast Implant Evaluation

The two types of breast implants include saline and silicone. Saline implant rupture is more clinically apparent, since the body readily resorbs the leaking saline and the implant shell appears deflated on exam.[13] Thus, there is no role for MRI Breast(s) in asymptomatic women with saline implants.[14] However, if the exam is equivocal for rupture, initial imaging supported by the ACR includes diagnostic mammogram and/or ultrasound in individuals >30 years old. In those <30 years of age, diagnostic mammogram is not typically performed and ultrasound is the initial imaging of choice.[14]

An exam is not as reliable for detecting the rupture of silicone implants as it is for saline implants. Therefore, if an exam is equivocal for rupture, imaging with a combination of ultrasound, mammogram, and/or MRI of the breast (with the choice of mammogram depending upon age) is appropriate.[15]

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Nipple Discharge/ Galactorrhea (BR-6)

Guideline

Nipple Discharge/Galactorrhea (BR-6.1)

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Nipple Discharge/Galactorrhea (BR-6.1)

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Physiologic nipple discharge (non-spontaneous or multi-duct, no suspicious findings on clinical exam)

- For individuals less than 40 years old, imaging is not medically necessary.
- For individuals 40 years old and older, a screening mammogram is medically necessary.
- If there is concern for a prolactinoma, please refer to **Pituitary, Sella, Hypothalamus (HD-19.1)**.

Pathologic nipple discharge (spontaneous, unilateral, single duct, clear or bloody, persistent and reproducible)

- For individuals less than 30 years old, breast ultrasound (CPT® 76641 or CPT® 76642) with or without a diagnostic mammogram is the medically necessary initial imaging.
  - If the breast ultrasound or diagnostic mammogram (if performed) is a BI-RADS™ category 1-3, a MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary.
  - If the breast ultrasound or diagnostic mammogram (if performed) are a BI-RADS™ category 4 or 5, a MRI Breast is **NOT** medically necessary. Biopsy is recommended in these circumstances.
- For individuals 30 years old and older, breast ultrasound (CPT® 76641 or CPT® 76642) and diagnostic mammogram are the medically necessary initial imaging.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 1-3, a MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary.
  - If the breast ultrasound or diagnostic mammogram are a BI-RADS™ category 4 or 5, a Breast MRI is **NOT** medically necessary. Biopsy is recommended in these circumstances.

Background and Supporting Information

- Physiologic nipple discharge is predominantly bilateral but may be unilateral. It is commonly multi-duct. It is predominantly milky but may be white or a variety of colors including serous, yellow, green, brown, or gray. Evaluation for hyperprolactinemia can be considered.
- For milky discharge, prolactin and TSH levels are recommended to diagnose prolactinoma; pituitary imaging is not needed if normal serum Prolactin.

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- Pathologic nipple discharge is defined as unilateral, bloody or serous, arising from a single duct, persistent, and spontaneous.

Evidence Discussion

No specific breast imaging is used for evaluation of physiologic discharge, other than usual screening mammogram in the appropriate age group. Otherwise, the evaluation is medical, including lab studies to rule out endocrine etiology. In a study of 13,443 women with nipple discharge, 316 (2.3%) had nonspontaneous discharge, only 1 (0.3%) of whom had carcinoma.[19] Similarly, a retrospective review of 273 women who underwent diagnostic and therapeutic surgery for nipple discharge found no malignancies in those presenting with physiologic nipple discharge.[20]

The evaluation of pathologic nipple discharge is aimed at determining if there is an underlying intraductal papilloma, high-risk lesion, or a malignancy. Larger studies estimate the rate of malignancy or high-risk histopathologic lesions to be 11% to 16% of individuals with pathologic nipple discharge.[22] Initial radiographic evaluation includes both diagnostic mammography and targeted breast ultrasound. If both are non-diagnostic, then MRI is the next imaging modality used for evaluation. Contrast-enhanced MRI has demonstrated sensitivities of 93 to 100 percent for invasive cancers as well as benign papillary lesions.[23]

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Breast Pain (Mastodynia) (BR-7)

Guideline

Breast Pain (Mastodynia) (BR-7.1)

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Breast Pain (Mastodynia) (BR-7.1)

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- Evaluation of breast pain requires a history and physical exam.
  - When breast pain is present with another breast symptom such as nipple discharge, skin change(s), or palpable mass, the imaging should be done in accordance with the accompanying symptom's guideline rather than this guideline for breast pain.
- If pain is cyclical and/or generalized across more than one quadrant of the breast, an up-to-date screening mammogram is medically necessary.
- If pain is focal and the individual is 30 years old or older, diagnostic mammogram and breast ultrasound (CPT® 76641 or CPT® 76642) are medically necessary as the initial imaging.
- If pain is focal and the individual is less than 30 years old, breast ultrasound (CPT® 76641 or CPT® 76642) is medically necessary as the initial imaging.
- Advanced imaging is **NOT** medically necessary in individuals with breast pain or breast abscesses.

Background and Supporting Information

- The risk of malignancy following a negative clinical examination (clinical breast exam, mammogram, ultrasound) has been estimated to be only 0.5%.

Evidence Discussion

In a study of 2820 individuals presenting with breast pain, the cancer detection rate in those who underwent breast imaging was found to be 0.09%, 1% and 1.4% in individuals under the age of 40, 40-49 and 50 years of age or older, respectively.[24] Similarly, in a case control study comparing 987 women with painful breasts and 987 controls, the prevalence of breast cancer was similar between the two groups (0.8% vs. 0.7%, respectively).[25] Given these data, in the absence of other factors, the ACR recommends against the use of MRI in individuals with breast pain.[26]

Breast abscesses can present with a variety of etiologies. In a review of various inflammatory diseases of the breast, Scott et al points to ultrasound as the appropriate initial imaging. It is also noted that while diagnostic mammogram can be done, it may not be very beneficial in all etiologies.[92]

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Alternative Breast Imaging Approaches (BR-8)

Guideline

Alternative Breast Imaging Approaches (BR-8.1)

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Alternative Breast Imaging Approaches (BR-8.1)

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Molecular Breast Imaging (MBI)

- Molecular Breast Imaging (CPT® 78800) is supported in individuals who meet criteria for breast cancer screening with MRI (per **BR-5**) but for whom MRI is contraindicated.
  - See **Risk Factors** below.

Risk Factors

- Routine MRI Breast following bilateral mastectomy is **NOT** medically necessary (even if high-risk screening criteria may otherwise be met and/or nipple-sparing mastectomy was done).
- Annual MRI Breast screening with MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary for individuals meeting the high-risk criteria in the table below (for male breast imaging, please see **Breast Imaging in Males (BR-9.1)**):

High-Risk Indications	Age at which screening can start**
Genetic Mutations:*	
Li Fraumeni (p53)	20
BRCA 1 or 2	25
STK11, Peutz-Jeghers syndrome (PJS), PTEN Mutation (Cowden Syndrome), CDH1, NF1, PALB2, ATM, CHEK2	30**
BARD1, RAD51C, RAD51D	40**
Personal history of atypia/LCIS/breast cancer:	
ADH, ALH, LCIS	At diagnosis but not prior to age 25
Personal history of breast cancer at or before the age of 50	At diagnosis
Family history:	

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High-Risk Indications	Age at which screening can start**
If the individual has **NOT** been tested for BRCA mutation **and** there is a first-degree relative (parent, sibling, child; half siblings are considered second-degree relatives) with BRCA 1 or BRCA 2 mutation. Annual screening is **NOT** medically necessary if the individual has been tested and is negative for BRCA 1 or BRCA 2 mutation unless they meet other criteria.	40**
Two or more first-degree relatives with breast or ovarian cancer	40**
One first-degree relative with breast cancer or ovarian cancer that was diagnosed ≤ age 50	40**
One first-degree relative with bilateral breast cancer, or both breast and ovarian cancer	40**
A first- or second-degree male relative (father, brother/half-brother, uncle, grandfather) diagnosed with breast cancer	40**
Elevated clinical lifetime-risk:	
Clinical lifetime-risk estimated at greater than or equal to 20% as calculated by one of the following models: • Gail (National Cancer Institute (NCI)) • Tyrer-Cuzick (International Breast Cancer Intervention Study (IBIS)) • The Breast Cancer Surveillance Consortium (BCSC) • Breast and Ovarian Analysis of Disease Incidence and Carrier Estimation Algorithm (BOADICEA)/CanRisk • BRCAPRO Model	40**
Personal history of radiation therapy when younger than age 30:	
Radiation to chest, whole lung, mediastinum, axilla, mantle (including mini mantle or extended mantle), total or subtotal lymphoid irradiation or total body irradiation (TBI)	25 or 8 years after completion of radiation therapy *whichever comes later*
Breast Density:	

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High-Risk Indications	Age at which screening can start**
Heterogeneously Dense Breasts (Category C) or Extremely Dense Breasts (Category D) with no additional risk factors	40

*The following have unknown or insufficient evidence of breast cancer risk and additional MRI screening is NOT medically necessary at this time: MSH2, MLH1, MSH6, PMS2, EPCAM, NBN, genetic variants of unknown significance, genetic variants favoring polymorphism, and genetic variants of intermediate penetrance. Any gene mutation not specified in the table above has not currently been found to have sufficient evidence to support surveillance with MRI.

OR 10 years prior to the age of diagnosis of the earliest relative with breast cancer (first-, second-, and third-degree relatives) *whichever comes first*, **but not before age 25

Other Alternative Breast Imaging Techniques

Other alternative breast imaging techniques may have FDA approval, but they remain investigational with respect to **BOTH** screening and diagnosis of breast cancer. These include the following:

- Nuclear breast imaging, including:
  - Scintimammography
  - Breast specific gamma imaging (BSGI)
- PET Mammography (PEM)
- Thermography
- Impedance Mammography
- Other techniques to detect oxygen consumption, light absorption, microwave transmission, nitrous oxide production
- CT Breast (CPT® 0633T, CPT® 0634T, CPT® 0635T, CPT® 0636T, CPT® 0637T, or CPT® 0638T)
- Cone Beam CT Breast

Background and Supporting Information

- CT Breast
  - CT Breast is evolving and currently being studied as a mode of breast cancer detection. It remains under investigation, and is not to be used in lieu of conventional breast imaging modalities.
- Positron-Emission Mammography

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- There is currently insufficient data available to generate appropriateness criteria for this modality, and this procedure should be considered investigational at this time.
  - High-resolution positron-emission mammography (PEM) by Naviscan™ PET Systems, also referred to as Naviscan™ or PET mammography, performs high-resolution metabolic imaging for breast cancer using an FDG tracer. The PEM detectors are integrated into a conventional mammography system, allowing acquisition of the emission images immediately after the mammogram.
  - Requesting providers often ask for PEM as CPT® 78811 or "PET scan of the breast."
  - The spatial resolution of this technique is at the individual duct level (1.5 mm) and allows visualization of intraductal as well as invasive breast cancers. This technique is especially adept at detecting ductal carcinoma in situ.
  - Early clinical trials have shown high clinical accuracy in characterizing lesions identified as suspicious on conventional imaging or physical examination, as well as in detecting incidental breast cancers not seen on other imaging modalities.
  - A prospective multi-center clinical trial for females with newly diagnosed breast cancer anticipating breast-conservation surgery was performed. These females underwent both high-resolution PEM imaging and breast MRI. Results showed that PEM and MRI had comparable breast-level sensitivity, although MRI had greater lesion-level sensitivity and more accurately depicted the need for mastectomy. PEM had greater specificity at the breast and lesion levels. Of these, 3.6% of the females had tumors seen only with PEM.
  - The radiation exposure from a PEM study is 23 times higher than for digital mammography.

Evidence Discussion

There is limited data regarding the use of MBI in individuals of average breast cancer risk. However, in those classified as high risk (lifetime risk ≥ 20%), the NCCN guideline supported MBI for those who met criteria for supplemental breast MRI, but who could not undergo MRI.[7]

There is no data to support other alternative breast imaging techniques. They are not supported for screening by the ACR, NCCN, or other breast society guidelines. As more data becomes available, the guidelines will be updated accordingly.

The ACS considers individuals who have a first-degree relative with a BRCA 1 or 2 gene mutation and who have not been tested themselves to be at high risk. They recommended an annual MRI screening starting at age 30.[6] On the other hand, NCCN guidelines suggested that untested individuals with a first-degree relative with a BRCA 1 or 2 mutation should start screening either 10 years before the youngest family member

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was diagnosed with breast cancer, but not before age 25, or at age 40, whichever comes first.[7]

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Breast Imaging in Males (BR-9)

Guideline

Breast Imaging in Males (BR-9.1)

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Breast Imaging in Males (BR-9.1)

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See **Breast Ultrasound (BR-1)**

Screening for Males at Increased Risk for Breast Cancer

- A clinical breast exam every 12 months is medically necessary.
- Annual mammogram, especially for those with *BRCA2* P/LP variants in whom the lifetime risk of breast cancer is up to 7%, starting at age 50 or 10 years before the earliest known male breast cancer in the family, is medically necessary.
  - MRI of the male breast is not medically necessary given the paucity of evidence supporting its efficacy in male breast disease.

Symptomatic Male Breast Imaging

- Diagnostic Mammogram and/or breast ultrasound (CPT® 76641 or CPT® 76642) is medically necessary for evaluation of the symptomatic male breast and preferred method depends on age and the suspected etiology of disease.
  - MRI of the male breast is not medically necessary given the paucity of evidence supporting its efficacy in male breast disease.

Background and Supporting Information

- Breast cancer in males presents as a mass, skin/nipple change, or pathologic nipple discharge.

Evidence Discussion

Breast cancer management in cis-gender males is similar to females. NCCN guidelines recommended that, for males presenting with bilateral breast enlargement consistent with gynecomastia or pseudogynecomastia, reassurance with clinical management of the presumed cause (e.g., drug induced, hypogonadism, hyperthyroidism, etc) is all that was needed. For males presenting with palpable symptoms not explained by gynecomastia, or for those presenting with bloody nipple discharge, work up should include mammography and ultrasound, followed by core needle biopsy if these studies should be found to be BIRADS™ category 4-5.[7] Mammography has been found to be accurate in distinguishing benign from malignant lesions in men, and has a sensitivity and specificity of 92% and 90%, respectively, such that more advanced imaging is generally not required.[27]

The NCCN noted support of annual mammogram for cis-gender males, noting it is especially recommended in those "with *BRCA2* P/LP variants in whom the lifetime risk of

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breast cancer is up to 7%, starting at age 50 or 10 years before the earliest known male breast cancer in the family (whichever comes first)".[8]

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Breast Evaluation in Pregnant or Lactating Females (BR-10)

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Breast Evaluation in Pregnant or Lactating Females (BR-10.1)

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Breast Evaluation in Pregnant or Lactating Females (BR-10.1)

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- Breast ultrasound (CPT® 76641 or CPT® 76642) is first-line imaging in pregnant and lactating females.
- If pregnant/lactating female has a palpable mass **OR** has persistent unilateral bloody nipple discharge and the ultrasound is negative or suspicious, follow with diagnostic mammogram (with lead abdominal shielding).
- IV Gadolinium is required with MRI to evaluate breast parenchyma but is contraindicated in pregnancy. Biopsy, rather than advanced imaging, is recommended after inconclusive mammogram and ultrasound.
- MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is supported for evaluation in lactating women if criteria are met otherwise (see **BR-5.1**).
- For imaging requests related to a breast abscess, please see **Breast Pain (Mastodynia) (BR-7.1)**.

Evidence Discussion

Pregnancy-associated breast cancer (PABC) is defined as breast cancer diagnosed during pregnancy, throughout the first postpartum year, or during lactation.

The most common presentation of PABC is a palpable mass, but >80% of palpable masses that are biopsied in pregnant and breastfeeding women are benign.[80]

Given the difficulty examining the pregnant and lactating individual, diagnostic breast imaging is crucial in characterizing the features of a palpable mass. In up to 20% of lactating women, isolated bloody nipple discharge without an associated mass can occur, most commonly due to benign etiologies. However, if persistent, bloody nipple discharge can also be a sign of breast cancer. Diagnostic imaging is also recommended in these women.

Ultrasound has the highest sensitivity for the diagnosis of PABC.[81,82] Additionally, both pregnant and lactating woman are predominantly young and have dense breast tissue. Therefore the sensitivity of mammography decreases in these women. For that reason, ultrasound is the first-line imaging in pregnant and lactating women.[82]

Advanced imaging with breast MRI has a limited role in pregnant women. The IV administration of gadolinium is contraindicated. If there is clinical suspicion of malignancy, a biopsy is the next step in evaluation.[61,83]

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Transgender Breast Cancer Supplemental Screening (BR-12)

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Transgender Breast Cancer Supplemental Screening (BR-12.1)

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Transgender Breast Cancer Supplemental Screening (BR-12.1)

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- Annual supplemental breast ultrasound (CPT® 76641 or CPT® 76642) **AND/OR** MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) screening is medically necessary for the following:
  - Transmasculine (female-to-male) with **ALL** of the following risk factors:
    - Reduction mammoplasty or no chest surgery
    - Age ≥25
    - High-risk (≥20% lifetime risk)
- Annual breast ultrasound and/or MRI Breast, in addition to mammogram, for breast cancer screening is **NOT** medically necessary in any other scenarios, including **ANY** of the following:
  - Transfeminine (male-to-female)
  - Transmasculine (female-to-male), who have had bilateral mastectomies
  - Transmasculine (female-to-male), who have **NOT** had mastectomies **AND** are at average risk or intermediate risk
- Acceptable models of calculating clinical lifetime-risk include the following: Gail (NCI), Tyrer-Cuzick (IBIS), The Breast Cancer Surveillance Consortium (BCSC), Breast and Ovarian Analysis of Disease Incidence and Carrier Estimation Algorithm (BOADICEA)/CanRisk, or BRCAPRO.

Evidence Discussion

A number of studies have found that transgender individuals who have transitioned from female to male have the same risk of developing breast cancer as their cis-gendered female counterparts.[28-30] As such, those who still have breast tissue (i.e., have only undergone reduction mammoplasty or no chest surgery), should be screened similarly to cis-gendered women.

The ACR Appropriateness Criteria® recommend the use of ultrasound and/or MRI for individuals who are at intermediate to high risk based on either having a lifetime risk ≥ 20%, a personal history of breast cancer, lobular neoplasia or atypia, chest wall irradiation, or have a genetic predisposition to developing breast cancer.[30] The ACR, does however, recommend transmasculine (female-to-male) individuals start screening earlier than their cis-gendered counterparts (starting at 25-30 years of age).[30]

For transmasculine individuals who are at low to average risk, mammography alone is sufficient.[28-30] Individuals who have had bilateral mastectomies have minimal

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residual breast tissue, such that breast cancer screening using imaging is not medically necessary.[28-30]

The ACR found insufficient evidence to support the use of routine MRI screening in transfeminine (male-to-female) individuals, regardless of duration of hormone use and/or genetic factors. Transfeminine individuals who would otherwise be considered "high risk" based on personal or family history may consider annual mammography. Similaraly, mammography may be appropriate in transfeminine individuals who have taken feminizing hormones for more than 5 years.[30]

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3D Rendering (BR-13)

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3D Rendering (BR-13.1)

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3D Rendering (BR-13.1)

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- 3D rendering (CPT® 76376 or CPT® 76377) should **NOT** be used in conjunction with **ANY** 3D mammography code.
- 3D rendering (CPT® 76376 or CPT® 76377) is **NOT** indicated for breast ultrasound. It is commonly requested in conjunction with automated breast ultrasound (ABUS); there is no evidence to support its clinical usefulness.
- 3D rendering (CPT® 76376 or CPT® 76377) should **NOT** be used in conjunction with MRI Breast.

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Breast Mass (BR-14)

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Breast Mass (BR-14.1)

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Breast Mass (BR-14.1)

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- MRI Breast is **NOT** medically necessary to determine biopsy recommendations for suspicious or indeterminate lesion(s) that can be readily biopsied on physical exam, such as palpable masses.
- For individuals 30 years old and older, diagnostic mammogram and breast ultrasound (CPT® 76641 or CPT® 76642) are medically necessary as the initial imaging.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 4 or 5, a Breast MRI is NOT medically necessary. Biopsy is recommended in these circumstances.
- For individuals less than 30 years old, breast ultrasound (CPT® 76641 or CPT® 76642) is medically necessary as the initial imaging.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 4 or 5, a Breast MRI is NOT medically necessary. Biopsy is recommended in these circumstances.

Evidence Discussion

According to the ACR Appropriateness Criteria® for "Palpable Breast Masses" there is a paucity of evidence to support the use of MRI Breast in the evaluation of a palpable mass regardless of what the BI-RADS™ is on mammogram.[93]

NCCN guidance for imaging of a palpable breast mass supports the use of diagnostic mammogram and/or ultrasound (preferred modality is dependent on age).[7]

Imaging with BI-RADS™ assessment of category 4 require biopsy. MRI is not supported prior to biopsy.[17]

Imaging with BI-RADS™ assessment of category 3 require short-term follow up imaging: at 6, 12, and 24 months.[18]

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Skin Changes (BR-15)

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Skin Changes (BR-15.1)

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Skin Changes (BR-15.1)

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- Diagnostic mammogram with or without breast ultrasound (CPT® 76641 or CPT® 76642) is the medically necessary initial imaging.
  - If the diagnostic mammogram or breast ultrasound (if performed) is a BI-RADS™ category 1-3, a MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary.
  - If the diagnostic mammogram or breast ultrasound (if performed) is a BI-RADS™ category 4 or 5, a MRI Breast is NOT medically necessary. Biopsy is recommended in these circumstances.
    - If a core needle biopsy is performed and is benign, a MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary.
- Advanced imaging is **NOT** medically necessary in individuals with breast abscesses.

Evidence Discussion

NCCN guidance for imaging of skin changes of the breast supports the use of diagnostic mammogram with or without breast ultrasound as the initial imaging. Additional imaging with MRI Breast is appropriate for BI-RADS™ 1, 2, or 3 on the initial imaging.[7]

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Nipple Inversion/ Retraction (BR-16)

Guideline

Nipple Inversion/Retraction (BR-16.1)

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Nipple Inversion/Retraction (BR-16.1)

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This guideline is only to be used when there is no palpable mass. If there is an associated palpable mass, please see **Palpable Mass (BR-14.1)**.

Congenital/Life-Long

- If there are no recent changes, only standard screening is recommended.
- If there are recent changes, see Acquired/New Onset below.

Acquired/New Onset

- If nipple discharge is present, please see **Nipple Discharge/Galactorrhea (BR-6.1)**.
- If skin changes are present, please see **Skin Changes (BR-15.1)**.
- For individuals 30 years old and older, diagnostic mammogram and breast ultrasound (CPT® 76641 or CPT® 76642) are medically necessary for the initial imaging.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 1, 2, or 3, but is clinically suspicious, a MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 4 or 5, a Breast MRI is NOT medically necessary. Biopsy is recommended in these circumstances.
- For individuals less than 30 years old, breast ultrasound (CPT® 76641 or CPT® 76642) with or without diagnostic mammogram is medically necessary for the initial imaging.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 1, 2, or 3, but is clinically suspicious, a MRI Breast Bilateral without and with (CPT® 77049 or HCPCS C8908) is medically necessary.
  - If the breast ultrasound or diagnostic mammogram is a BI-RADS™ category 4 or 5, a Breast MRI is NOT medically necessary. Biopsy is recommended in these circumstances.

Evidence Discussion

NCCN guidance for imaging of nipple inversion/retraction supports the use of diagnostic mammogram and/or breast ultrasound as the initial imaging (preferred modality is dependent on age). Additional imaging with MRI Breast is dependent on the BI-RADS™ category of the initial imaging as well as level of clinical suspicion.[7]

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Malignant Phyllodes Tumor/Cystosarcoma Phyllodes (BR-17)

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Malignant Phyllodes Tumor/Cystosarcoma Phyllodes (BR-17.1)

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Malignant Phyllodes Tumor/ Cystosarcoma Phyllodes (BR-17.1)

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- MRI Breast Bilateral without and with contrast (CPT® 77049 or HCPCS C8908) is medically necessary pre-operatively to establish extent of disease where a diagnosis of malignant phyllodes tumor has previously been established by tissue diagnosis.

Background and Supporting Information

- Phyllodes tumor is usually benign and has clinical characteristics of fibroadenoma, although they may exhibit rapid growth. MRI Breast has not been shown to be of value in distinguishing fibroadenoma from phyllodes tumor.
- Diagnosis is made by tissue diagnosis (percutaneous core biopsy or excisional biopsy). FNA biopsy is inaccurate in phyllodes tumor diagnosis and is not recommended.
- Treatment is wide local excision. Axillary lymph node dissection is not necessary. It has a predilection for local recurrence following local excision.
- If biopsy establishes a diagnosis of **malignant phyllodes** (cystosarcoma phyllodes), it should be treated as a soft tissue sarcoma. See **Sarcomas – Bone, Soft Tissue, and GIST (ONC-12)** in the Oncology Imaging Guidelines.

Evidence Discussion

Phyllodes tumors of the breast are usually benign, fibroepithelial lesions that have a range of biologic behaviors. Diagnosis is made by percutaneous core biopsy or excisional biopsy. MRI Breast has not been shown to be of value in distinguishing phyllodes tumor from fibroadenoma. However, malignant phyllodes have the propensity to metastasize. Thus, MRI is supported in malignant phyllodes to determine the extent of disease and resectability.[12]

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General Information

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Abbreviations for the Cardiac Imaging Guidelines

Abbreviation	Description
ACC	American College of Cardiology
ACS	Acute coronary syndrome
AHA	American Heart Association
ANOCA	Angina with no obstructive coronary artery disease
ASCOT	Anglo-Scandinavian Cardiac Outcomes Trial
ASD	Atrial septal defect
BMI	Body mass index
CABG	Coronary artery bypass grafting
CAC	Coronary artery calcium
CAD	Coronary artery disease
CAD-RADS	The Coronary Artery Disease Reporting and Data System
CHF	Congestive heart failure
CMD	Coronary microvascular dysfunction
COPD	Chronic obstructive pulmonary disease
CT	Computed tomography
CCTA	Coronary computed tomography angiography
CTA	Computed tomography angiography

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Abbreviation	Description
CTV	Computed tomography venography
CV	Cardiovascular
EBCT	Electron beam computed tomography
ECP	External counterpulsation (also known as EECP)
ECG	Electrocardiogram
ECP	External counterpulsation
ETT	Exercise treadmill stress test
FDG	Fluorodeoxyglucose, a radiopharmaceutical used to measure myocardial metabolism
FFR	Fractional flow reserve
HCM	Hypertrophic cardiomyopathy
INOCA	Ischemia with no obstructive coronary artery disease
IV	Intravenous
LAD	Left anterior descending coronary artery
LDL-C	Low density lipoprotein cholesterol
LHC	Left heart catheterization
LV	Left ventricle
LVEF	Left ventricular ejection fraction
MI	Myocardial infarction
MINOCA	Myocardial infarction with no obstructive coronary artery disease
MPI	Myocardial perfusion imaging (SPECT study, nuclear cardiac study)

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Abbreviation	Description
MRA	Magnetic resonance angiography
MRI	Magnetic resonance imaging
mSv	millisievert (a unit of radiation exposure) equal to an effective dose of a joule of energy per kilogram of recipient mass
MUGA	Multi gated acquisition scan of the cardiac blood pool
PCI	Percutaneous coronary intervention (includes percutaneous coronary angioplasty (PTCA) and coronary artery stenting)
PET	Positron emission tomography
PTCA	Percutaneous coronary angioplasty
RHC	Right heart catheterization
SCAD	Spontaneous coronary artery dissection
SPECT	Single photon emission computed tomography
TEE	Transesophageal echocardiogram
TAVI	Transcatheter aortic valve implantation
TAVR	Transcatheter aortic valve replacement
TIA	Transient Ischemic Attack
VSD	Ventricular septal defect

Glossary

Acute Chest pain chest pain is new onset (<2 mo) or involves a change in pattern, intensity, or duration compared with previous episodes in an individual with recurrent symptoms

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Acute Coronary Syndrome	sudden imbalance between myocardial oxygen consumption and demand, usually related to coronary artery obstruction, but includes other non-coronary artery origins such as acute coronary insufficiency (e.g., coronary embolism), hypotension, non-ischemic myocardial injury (e.g., myocarditis) and multifactorial causes that are not mutually exclusive (e.g., stress cardiomyopathy)
Agatston Score	a nationally recognized calcium score for the coronary arteries based on Hounsfield units and size (area) of the coronary calcium
Angina pectoris	Retrosternal chest discomfort that builds gradually in intensity (over several minutes), is usually precipitated by stress (physical or emotional) or occurring at rest (as in the case of an ACS) with characteristic radiation (e.g., left arm, neck, jaw) and its associated symptoms (e.g., dyspnea, nausea, lightheadedness). When actively treated (e.g., nitroglycerin) or spontaneously resolving (e.g., with rest), it dissipates over a few minutes
Anginal variants or equivalents	Symptoms or pain at a site other than the chest occurring in an individual at high cardiac risk (i.e., known CAD, diabetes). Anginal equivalents have the same importance as angina pectoris and can present as dyspnea, diaphoresis, nausea, extreme fatigue, pain, pressure, tightness, or discomfort in shoulders, arms, neck, back, upper abdomen, or jaw not otherwise explained by another process.
ARVD/ARVC – Arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy	a potentially lethal inherited disease with syncope and rhythm disturbances, including sudden death, as presenting manifestations
BNP	B-type natriuretic peptide, blood test used to diagnose and track heart failure (n-T-pro-BNP is a variant of this test)
Brugada Syndrome	an electrocardiographic pattern that is unique and might be a marker for significant life-threatening dysrhythmias

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Chronic Coronary Syndrome	CAD that is chronic, often progressive, and is treatable with lifestyle adjustments, medications, and invasive interventions instituted to achieve disease stabilization or regression
Double Product (Rate Pressure Product)	an index of cardiac oxygen consumption, is the systolic blood pressure times heart rate, generally calculated at peak exercise; over 25000 means an adequate stress load was performed
Fabry Disease	an infiltrative cardiomyopathy, can cause heart failure and arrhythmias
Fatigue	a subjective feeling of weakness, tiredness or exhaustion. Exertional fatigue is acute in nature, with rapid onset, short duration, and short recovery period.
Hibernating myocardium	viable but poorly functioning or non-functioning myocardium which likely could benefit from intervention to improve myocardial blood supply
Optimized Medical Therapy	should include (where tolerated) antiplatelet agents, calcium channel antagonists, partial fatty acid oxidase inhibitors (e.g., ranolazine), statins, short-acting nitrates as needed, long-acting nitrates up to 6 months after an acute coronary syndrome episode, beta blocker drugs (optional), angiotensin-converting enzyme (ACE) inhibitors/angiotensin receptor blocking (ARB) agents (optional)
Platypnea	shortness of breath when upright or seated (the opposite of orthopnea) and can indicate cardiac malformations, shunt or tumor

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Risk factors for Cardiac Chest pain	Comorbidities that increase the risk of developing coronary artery disease such as age ≥65 y, diabetes, hypertension (i.e., stage 1 hypertension (130–139 mm Hg systolic or 80–89 mm Hg diastolic), or stage 2 hypertension (≥140 mm Hg systolic or ≥90 mm Hg diastolic)), adverse pregnancy outcomes (includes hypertensive disorders of pregnancy, preterm delivery, gestational diabetes, placental abruption), family history of premature CAD (history of any direct blood relatives (parents, siblings, children) who have had any of the following conditions at age 55 or younger for male relatives or age 65 or younger for female relatives: AMI, sudden cardiac death without obvious cause, CABG surgery, PCI), familial/genetic factors (includes genetic abnormalities leading to vasculopathies, aneurysmal disorders, cardiomyopathies, coagulopathies, and inherited dysrhythmia syndromes)
Silent ischemia	absence of ischemic symptoms or signs prior to objective demonstration of ischemia by stress testing and/or demonstration of obstructive CAD
Stable Chest Pain	chest pain symptoms are chronic (≥2 mo) and associated with consistent precipitants such as exertion or emotional stress
Syncope	loss of consciousness; near-syncope is not syncope
Takotsubo cardiomyopathy	apical dyskinesis oftentimes associated with extreme stress and usually thought to be reversible
Troponin	a marker for ischemic injury, primarily cardiac
TAVR (TAVI)	A minimally invasive procedure to treat aortic valve disease

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Practice Estimate of Effective Radiation Dose chart for Selected Imaging Studies

Imaging Study	Estimate of Effective Radiation Dose
PET myocardial perfusion study: Rubidium-82 Nitrogen-13 ammonia	4 mSv 2 mSv
Thallium myocardial perfusion study (MPI) Dual isotope: 3.5 mCi 201Tl rest/ 30 mCi 99mTc sestamibi stress	23 mSv
Diagnostic conventional coronary angiogram (catheterization)	2mSv to 22 mSv
Coronary computed tomography angiography (CCTA)	8.7 mSv
Cardiac CT for calcium scoring	1mSv to 5mSv
CT Abdomen and Pelvis	7.7 mSv
Chest x-ray	0.1 mSv

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General Guidelines

- A pertinent clinical evaluation since the onset or change in symptoms is required prior to considering advanced imaging, which includes:
  - Relevant history and physical examination and appropriate laboratory studies and non-advanced imaging modalities, such as recent ECG, chest x-ray or ECHO/ ultrasound, performed after symptoms started or worsened
    - Effort should be made to obtain copies of reported abnormal ECG studies in order to determine whether the ECG is uninterpretable for ischemia on ETT.
    - Most recent previous stress testing and its findings should be obtained.
  - A recent clinical evaluation documenting any subjective findings (complaints, changes in behavior) or objective findings (clinical exam findings)
    - Other meaningful contact (telephone call, electronic mail or messaging) by an established individual can substitute for a face-to-face clinical evaluation such as requests based on new increased or worsening symptoms.
    - Some conditions may require a face-to-face evaluation as discussed in the applicable condition-specific guideline sections (such as requests based on new physical exam findings).
    - A recent clinical evaluation may be medically unnecessary if the individual is undergoing a guideline-supported, scheduled follow-up imaging or other designated procedural evaluation.
    - Exceptions due to routine surveillance indications are addressed in the applicable condition-specific guideline sections.
  - Vital signs, height and weight, BMI, or description of general habitus is needed.
  - Clinical question to be answered by advanced imaging that will affect management of the individual's clinical condition
- Cardiac imaging is not medically necessary if the results will not affect clinical management decisions. If a decision to perform cardiac catheterization or other angiography has already been made, there is often no need for imaging stress testing.
- Assessment of ischemic symptoms (if present) based on the descriptions below following this section

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Cardiac evaluation

Cardiac Chest Pain

For the purpose of this guideline, when diagnostic testing is requested to evaluate cardiac symptoms for suspected ischemic origin, chest pain is defined as follows:

- Cardiac chest pain (stable angina pectoris, typical angina, likely angina) is defined as **one** of the following:
  - Retrosternal chest discomfort described as pain, pressure, squeezing, heaviness, tightness, or burning, and may radiate to the left arm, neck, jaw, or back that is precipitated by exercise or stress and resolved with rest or nitroglycerin
  - In an individual with known CAD (history of myocardial infarction, prior coronary revascularization with PCI or CABG, imaging or testing showing obstructive CAD) or SCAD (spontaneous coronary artery dissection) with any of the following symptoms:
    - chest discomfort
    - unexplained exertional fatigue
    - unexplained dyspnea on exertion
    - an individual's known anginal equivalent (symptoms with prior known cardiac event)
  - Retrosternal chest discomfort described as pain, pressure, squeezing, heaviness, tightness, or burning, and may radiate to the left arm, neck, jaw, or back that is precipitated by exercise or stress is considered cardiac chest pain in the setting of any of the following:
    - age ≥65
    - diabetes mellitus
    - hypertension
    - history of adverse pregnancy outcomes (hypertensive disorders of pregnancy, preterm delivery, gestational diabetes, small-for-gestational-age delivery, placental abruption, and pregnancy loss)
    - chest pain associated with syncope
    - family history of premature CAD (occurring before age 56 in males or before age 66 in females)
- Possible Cardiac Chest Pain:
  - Symptoms suggestive of a cardiac origin but lacking definitive features or occurring in a time course not suggestive of angina (such as atypical pain location or quality, pain resolves spontaneously within seconds or lasts for an extended period and is unrelated to exertion)
  - May occur in the presence of mild associated symptoms, or in the presence of some risk factors without diagnostic confirmation
- Non-cardiac explanation:

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  - Pain likely due to non-cardiac causes, especially when persistent or recurring despite negative cardiac evaluations
  - Pain qualities include sharp, stabbing, fleeting, or positional. Associated symptoms may include heartburn unrelated to meals or abdominal discomfort.
- Anginal equivalents (individuals with previously documented CAD only):
  - Symptoms consistent with individual's known angina pattern in an individual with a history of CABG or PCI
  - Dyspnea on exertion
  - Fatigue (overwhelming sense of unexplained exhaustion causing a decreased capacity for physical activity or mental work)

Other signs and symptoms suggestive of potential cardiac etiology:
- dyspnea
- orthopnea
- paroxysmal nocturnal dyspnea
- heartburn unrelated to meals/nausea and vomiting
- palpitations
- syncope
- heart failure

Note: Chest pain remains the predominant symptom reported by females among those diagnosed with an acute coronary syndrome.

For the purpose of this guideline, evidence documenting the presence of obstructive CAD includes any of the following:
- Prior heart catheterization or CCTA revealing any of the following:
  - ≥40% stenosis of the left main coronary artery
  - ≥50% stenosis for other major epicardial vessels
  - Significant stenosis defined by an FFR of ≤0.80
  - History of a prior PCI or CABG

For the purpose of this guideline, evidence documenting the presence of non-obstructive CAD includes prior heart catheterization or CCTA revealing any of the following:
- <40% stenosis of the left main coronary artery
- <50% stenosis for other major epicardial vessels
- FFR >0.8

The Coronary Artery Disease Reporting and Data System (CAD-RADS) classification of percentage luminal diameter coronary artery stenosis on coronary CT angiography (CCTA) is as follows:
- CAD-RADS 0: 0%
- CAD-RADS 1: 1% to 24%

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- CAD-RADS 2: 25% to 49%
- CAD-RADS 3: 50% to 69%
- CAD-RADS 4: 70% to 99% or ≥50% left main coronary artery stenosis
- CAD-RADS 5: 100% (total occlusion)

For the purposes of this guideline, evidence documenting a prior MI includes any of the following:

- presence of diagnostic Q waves on an ECG
- a fixed perfusion defect on MPI
- akinetic or dyskinetic wall motion on echocardiogram
- area of Late Gadolinium Enhancement (LGE) on cardiac MRI suggesting scar

Findings that may alter the ECG changes during exercise or are uninterpretable for ischemia on a stress test:

- Complete Left Bundle Branch Block (bifasicular block involving right bundle branch and left anterior hemiblock does not render ECG uninterpretable for ischemia)
- Ventricular paced rhythm
- Pre-excitation pattern such as Wolff-Parkinson-White
- ≥1.0 mm ST segment depression (NOT nonspecific ST/T wave changes)
- LVH with repolarization abnormalities, also called LVH with strain (NOT without repolarization abnormalities or by voltage criteria)
- T wave inversion in at least two contiguous inferior and/or lateral leads. This includes leads II, AVF, V5 or V6. (T wave inversion isolated in lead III or T wave inversion in lead V1 and V2 are not included).
- Individual on digitalis preparation

The Exercise Treadmill Test (ETT)

- Requirements for a diagnostic ETT include:
  - ECG that can be interpreted for ischemia.
  - Individual capable of exercise to achieve target heart rate on a treadmill or similar device (5 METs or greater; see functional capacity below). Target heart rate is calculated as 85% of the maximum age predicted heart rate (MPHR). MPHR is estimated as 220 minus the individual's age.
- An abnormal ETT (exercise treadmill test) includes at least one of the following:
  - ST segment depression (horizontal or downsloping, ≥1.0 mm below baseline)
  - Development of chest pain
  - Drop in systolic blood pressure >10 mmHg during exercise
  - Non-sustained ventricular tachycardia ≥3 consecutive ventricular beats at a rate of >100 beats per minute

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- Sustained ventricular tachycardia (ventricular rhythm at rate >100 beats/minute lasting >30 seconds or requiring termination due to hemodynamic compromise in <30 seconds)

- Functional capacity ≥5 METs equates to the following:
  - Can walk four blocks without stopping
  - Can walk up a hill
  - Can climb one flight of stairs without stopping
  - Can perform heavy work around the house
  - Can walk 4 mph at a brisk pace

Evidence Discussion

Chest pain

Many factors are taken into consideration when an individual presents chest pain or symptoms suspect for an underlying cardiac etiology, such as coronary artery disease or congestive heart failure. Details such as precipitating and alleviating factors, duration of the symptoms, chronicity, progression, and prior medical history or risk factors that may escalate the concern for ischemia. Symptoms associated with cardiac etiology can include chest pain, pressure, heaviness, tightness, dyspnea, diaphoresis, nausea/vomiting, lightheadedness, syncope, and palpitations. Cardiac pain related to ischemia is characterized by pressure, squeezing, heaviness, tightness, or burning in the chest, often retrosternal, and radiating to the left arm, neck, jaw, or back. In general, ischemic chest pain has a gradual onset over minutes, is relieved by rest or nitroglycerin, and is precipitated by exertion or emotional stress.[1] Risk factors increasing the likelihood of an ischemic etiology of chest pain include age ≥65, diabetes, hypertension, history of adverse pregnancy outcomes, chest pain associated with syncope, family history of premature CAD, and genetic predispositions.[2,3]

CAD RADS 2

Coronary Artery Disease – Reporting and Data System (CAD-RADS)[4] is a standardized reporting framework for coronary CT angiography (CCTA) results designed to improve communication and guide clinical management. Classification includes stenosis severity (0–5), plaque burden (P1–P4), and modifiers (e.g., HRP,

[1] Gulati M, Levy PD, Mukherjee D, et al. 2021 AHA/ACC/ASE/CHEST/SAEM/SCCT/SCMR guideline for the evaluation and diagnosis of chest pain: executive summary: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2021;144:e336–e367. doi:10.1161/CIR.0000000000001030 .

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[3] Bays HE, Taub PR, Epstein E, et al. Ten things to know about ten cardiovascular disease risk factors. *Am J Prev Cardiol*. 2021;5:100149. doi: 10.1016/j.ajpc.2021.100149.

[4] Fletcher GF, Ades PA, Kligfield P, et al. Exercise standards for testing and training: a scientific statement from the American Heart Association. *Circulation*. 2013;128(8):873-934. doi:10.1161/CIR.0b013e31829b5b44.

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I, S, G, E) for CT-FFR and myocardial CT perfusion for ischemia assessment and is applicable to both stable and acute chest pain evaluations.[5] [6]

Background and Supporting Information

An observational study found that, compared with the Duke Activity Status Index, subjective assessment by clinicians generally underestimated exercise capacity

Upsloping ST segment depression is not considered to be an abnormal ETT finding because of its low specificity.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

[5] Cury RC, Leipsic J, Abbara S, et al. CAD-RADS™ 2.0 – 2022 Coronary Artery Disease – Reporting and Data System: An Expert Consensus Document of SCCT, ACC, ACR, and NASCI. *Radiol Cardiothorac Imaging*. 2022;4(5):e220183. doi:10.1148/ryct.220183.

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Stress Testing

Guideline

Stress Testing with Imaging (CD-1.4)
Stress Testing with Imaging-Preoperative (CD-1.5)
References (CD-1)

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Stress Testing with Imaging (CD-1.4)

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General information

- Imaging Stress Tests include any one of the following:
  - Stress Echocardiography see **Stress Echocardiography (Stress Echo) – Coding (CD-2.7)**
  - SPECT MPI see **Myocardial Perfusion Imaging (MPI) – Coding (CD-3.1)**
  - Stress perfusion MRI see **Cardiac MRI – Indications for Stress MRI (CD-5.3)**
  - PET Perfusion see **Cardiac PET-Perfusion-Indications (CD-6.2)**
- Stress testing with imaging can be performed with maximal exercise or chemical stress (adenosine, dipyridamole, dobutamine, or regadenoson) and does not alter the CPT® codes used to report these studies.

Stress Testing with Imaging-Coding

Stress echo, SPECT MPI or stress MRI

Codes Addressed	CPT®
Cardiac MRI for morphology and function without contrast, with stress imaging	75559
Cardiac MRI for morphology and function without and with contrast, with stress imaging	75563
MPI, tomographic (SPECT) including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78451
MPI, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78452

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Codes Addressed	CPT®
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78453
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78454
Echocardiography (TTE), (2D), with or without m-mode, during rest and cardiovascular stress, with interpretation and report	93350
Echocardiography (TTE), (2D), m-mode, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation	93351

Cardiac perfusion PET

Codes addressed	CPT®
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); single study, at rest or stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78430
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); multiple studies at rest and stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78431
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); single study at rest or stress (exercise or pharmacologic)	78491
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); multiple studies at rest and stress (exercise or pharmacologic)	78492

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Stress Testing with Imaging-Indications

Stress test with imaging (Stress echo, SPECT MPI, PET MPI, or stress MRI) is considered medically necessary in **any** of the following clinical settings:

Cardiac Chest Pain

New, recurrent or worsening cardiac chest pain as defined in **General Guidelines (CD-1.0)**

Symptomatic with known CAD

Prior history of PCI (percutaneous coronary intervention) or CABG (coronary artery bypass graft surgery) or a history of obstructive CAD as defined in **General Guidelines (CD-1.0)** and **any** of the following:

- Cardiac chest pain as defined in **General Guidelines (CD-1.0)**
- Anginal equivalents such as dyspnea or exertional fatigue
- Symptoms similar to prior ischemic episode

Newly elevated troponin with known CAD

Prior history of PCI (percutaneous coronary intervention) or CABG (coronary artery bypass graft surgery) or a history of obstructive CAD as defined in **General Guidelines (CD-1.0)** and a new Troponin elevation after last evaluation with stress test, CCTA, or diagnostic cardiac catheterization

Symptomatic and does not meet the requirements for an ETT without imaging

New, recurrent, or worsening symptoms of chest pain, or new, recurrent, or worsening symptoms of exertional dyspnea or exertional fatigue that is not explained by another process, **and** the individual does not meet the medically necessary components needed for an ETT due to a history of **any** of the following:

- Findings that may alter the ECG changes during exercise or are uninterpretable for ischemia on a stress test:
  - Complete Left Bundle Branch Block (bifasicular block involving right bundle branch and left anterior hemiblock does not render ECG uninterpretable for ischemia)
  - Ventricular paced rhythm
  - Pre-excitation pattern such as Wolff-Parkinson-White
  - ≥1.0 mm ST segment depression (NOT nonspecific ST/T wave changes)
  - LVH with repolarization abnormalities, also called LVH with strain (NOT without repolarization abnormalities or by voltage criteria)

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  - T wave inversion in at least two contiguous inferior and/or lateral leads. This includes leads II, AVF, V5 or V6. (T wave inversion isolated in lead III or T wave inversion in lead V1 and V2 are not included).
  - Individual on digitalis preparation
- Inability to perform the exercise needed to achieve target heart rate (85% of the maximum age predicted heart rate or 220 minus the individual's age) on a treadmill or similar device (5 METs or greater) due to any of the following limitations:
  - Unable to safely perform ETT due to gait disturbances, use of assistive devices, or other documented neuromuscular abnormalities
  - Use of home oxygen for daily activity
  - Documented activity-limiting dyspnea or heart failure
  - Provider assessment that performing an ETT would be unsafe
  - Unstable angina with concern for active acute coronary syndrome, or recent elevated troponin
  - Severe aortic stenosis or other significant aortic valve disease
  - Severe hypertension that can not be controlled prior to stress testing

Symptomatic with suspected or known non-obstructive CAD

Persistent symptoms of chest pain, exertional dyspnea or exertional fatigue despite normal or equivocal ETT

Symptomatic with inconclusive or abnormal ETT or elevated CAC

New, recurrent or worsening symptoms of chest pain, or new, recurrent, or worsening symptoms of exertional dyspnea or exertional fatigue that is not explained by another process, and any of the following:

- Inconclusive ETT (exercise treadmill test) due to any of the following:
  - <85% maximum predicted heart rate achieved
  - Exercise ECG is uninterpretable for ischemia (for example due to development of rate-related left bundle branch block during exercise)
- Abnormal ETT with any of the following results:
  - ST segment depression (horizontal or downsloping, ≥1.0 mm below baseline)
  - Development of chest pain
  - Drop in systolic blood pressure >10 mmHg during exercise
  - Non-sustained ventricular tachycardia ≥3 consecutive ventricular beats at a rate of >100 beats per minute
  - Sustained ventricular tachycardia (ventricular rhythm at rate >100 beats/minute lasting >30 seconds or requiring termination due to hemodynamic compromise in <30 seconds)
- Coronary artery calcium (CAC) score ≥100

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Symptomatic with Risk Factors

New, recurrent or worsening symptoms of chest pain, or new, recurrent, or worsening symptoms of exertional dyspnea or exertional fatigue that is not explained by another process, in individuals 50 years of age or above with 2 or more of the following CV risk factors (diabetes, smoking history, FH of premature CAD, hypertension or hyperlipidemia)

Heart failure or left ventricular systolic dysfunction

Stress test with imaging is medically necessary to evaluate heart failure or left ventricular systolic dysfunction when there is documentation of **any** of the following:

- New or worsening heart failure
- New left ventricular systolic dysfunction (left ventricular ejection fraction <50%)
- Worsening left ventricular systolic dysfunction (decline in left ventricular ejection fraction ≥10%)
- Significant ischemic ventricular dysfunction (suspected hibernating myocardium) to assess myocardial viability when there are persistent symptoms or heart failure and revascularization is being considered

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Note:

MRI, cardiac PET, SPECT MPI, or Dobutamine stress echo can be used to assess myocardial viability depending on physician preference. See also **Cardiac PET – Metabolic – Indications (CD-6.4)**

Syncope or arrhythmia

Stress test with imaging is medically necessary for **any** of the following:

- Syncope of suspected ischemic etiology not otherwise explained
- Sustained ventricular tachycardia (ventricular rhythm at rate >100 beats/minute lasting >30 seconds or requiring termination due to hemodynamic compromise in <30 seconds).
- Non-sustained ventricular tachycardia ≥3 consecutive ventricular beats at rate >100 beats/minute
- Frequent PVCs (premature ventricular complexes) >30 PVCs per hour
- Prior to starting a Class IC antiarrhythmic agent (flecainide or propafenone) to assess for CAD and annually while taking the medication

With or without symptoms for moderate coronary artery stenosis

Stress test with imaging is medically necessary to evaluate the functional significance of moderate stenosis when there is documentation of **either** of the following:

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- CCTA (coronary computed tomography angiography) with moderate stenosis (50 to 69%-CAD-RADS 3 as defined in **General Guidelines CD-1.0**)
- Invasive coronary angiography with intermediate severity stenosis and invasive physiological testing has not been done

Without symptoms

Stress test with imaging is medically necessary for any of the following:

- Prior to starting Interleukin-2
- An uninterpretable ECG as described in **General Guidelines (CD 1.0)** that has not been previously evaluated
- Every 2 years if there is a history of silent ischemia (absence of ischemic symptoms or signs prior to objective demonstration of ischemia by stress testing and/or demonstration of obstructive CAD as defined in **General Guidelines (CD-1.0)**
- Prior to starting a Class IC antiarrhythmic agent (flecainide or propafenone) to assess for CAD and annually while taking the medication
- Asymptomatic individual who has an ischemic EKG response on ETT (horizontal or downsloping ST depression ≥1.0 mm below baseline)

Evidence Discussion

Appropriate Use Criteria from major professional societies including the American College of Cardiology and the American Heart Association support the use of myocardial perfusion imaging (MPI) with single photon emission computed tomography (SPECT) for individuals with signs or symptoms consistent with coronary artery disease (CAD) such as typical angina. In addition, they support the use of SPECT perfusion imaging in symptomatic individuals with known CAD and those with prior interventions such as coronary artery bypass surgery, coronary stenting, and preoperative risk stratification. These guidelines balance the appropriate testing of individuals with SPECT versus unnecessary and potentially harmful testing and downstream procedures.

Also, as supported by society guidelines, use of SPECT is medically necessary in detection of cardiac transthyretin amyloidosis (ATTR) after light chain amyloidosis (AL) is ruled out by appropriate blood and urine testing. Positron emission tomography (PET) is an advanced imaging approach in that it allows for more accurate attenuation correction than SPECT, which can be helpful in individuals with higher BMIs or other sources of attenuation. In addition, due to accurate attenuation correction and the unique attributes of PET radionuclides, PET allows the quantitation of myocardial blood flow and flow reserve which has been shown to be helpful in the diagnosis and prognosis of obstructive and no obstructive CAD.

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Stress Testing with Imaging-Preoperative (CD-1.5)

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- There are **2** steps that determine the need for imaging stress testing in (stable) pre-operative individuals:
  - Step 1: Would the individual qualify for imaging stress testing independent of planned surgery?
    - If yes, proceed to stress testing guidelines **Stress Testing with Imaging – Indications (CD-1.4)**.
    - If no, go to step 2.
  - Step 2: Is the surgery considered high-, moderate-, or low-risk? (see **Table-2**)
    If high- or moderate-risk, proceed below. If low-risk, there is no evidence to determine a need for pre-operative cardiac testing.
    - **High-risk Surgery:** All individuals in this category should receive an imaging stress test if there has not been an imaging stress test within 1 year unless the individual has developed new cardiac symptoms or a new change in the EKG since the last stress test.
    - **Intermediate-risk Surgery:** One or more clinical risk factors and unable to perform an ETT per guidelines if there has not been an imaging stress test within 1 year unless the individual has developed new cardiac symptoms or a new change in the EKG since the last stress test.
    - **Low-risk:** Pre-operative imaging stress testing is not medically necessary.
  - Clinical Risk Factors (for cardiac death and non-fatal MI at time of non-cardiac surgery)
    - History of ischemic heart disease (previous MI, previous positive stress test, use of nitroglycerin, typical angina, ECG Q waves, previous PCI or CABG)
    - History of compensated previous congestive heart failure (history of heart failure, previous pulmonary edema, third heart sound, bilateral fine crackles, chest x-ray showing heart failure)
    - History of previous TIA or stroke
    - Diabetes Mellitus
    - Creatinine level >2 mg/dL

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Table-2 Cardiac Risk Stratification List

High-risk (> 5%)	Intermediate-risk (1-5%)	Low-risk (<1%)
• Open aortic and other major open vascular surgery • Open peripheral vascular surgery • Esophagectomy • Pneumonectomy • Open intraperitoneal and/or intrathoracic surgery with organ resection	• Open intraperitoneal and/or intrathoracic surgery without major organ resection • Open carotid endarterectomy • Head and neck surgery • Open orthopedic surgery • Open prostate surgery	• Endoscopic procedures • Superficial procedures • Cataract surgery • Breast surgery • Ambulatory surgery • Laparoscopic and endovascular procedures that are unlikely to require further extensive surgical intervention

Evidence Discussion

Appropriate Use Criteria from major professional societies including the American College of Cardiology and the American Heart Association support the use of myocardial perfusion imaging (MPI) with single photon emission computed tomography (SPECT) for individuals with signs or symptoms consistent with coronary artery disease (CAD) such as typical angina. In addition, they support the use of SPECT perfusion imaging in symptomatic individuals with known CAD and those with prior interventions such as coronary artery bypass surgery, coronary stenting, and preoperative risk stratification. These guidelines balance the appropriate testing of individuals with SPECT versus medically unnecessary and potentially harmful testing and downstream procedures.

Also, as supported by society guidelines, use of SPECT is medically necessary in detection of cardiac transthyretin amyloidosis (ATTR) after light chain amyloidosis (AL) is ruled out by appropriate blood and urine testing.

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Echocardiography (ECHO)

Guideline

Transthoracic Echocardiogram (TTE)-Coding (CD-2.1)
Transthoracic Echocardiography (TTE) Indications/Initial Evaluation (CD-2.2)
Frequency of Echocardiography Testing (CD-2.3)
References (CD-2)
Transesophageal Echocardiography (TEE) (CD-2.4) (CD-2.5)
Stress Echocardiography (Stress Echo) (CD-2.7)
3D Echocardiography (CD-2.9)
Myocardial Strain Imaging (CD-2.12)
References (CD-2)

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Transthoracic Echocardiogram (TTE)-Coding (CD-2.1)

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Transthoracic Echocardiography (TTE)-Coding

Transthoracic Echocardiography

Description	CPT®
TTE for congenital cardiac anomalies, complete	93303
TTE for congenital cardiac anomalies, follow-up or limited	93304
TTE with 2-D, M-mode, Doppler and color flow, complete	93306
TTE with 2-D, M-mode, without Doppler or color flow	93307
TTE with 2-D, M-mode, follow-up or limited	93308

3D Echocardiography

Description	CPT®
3D rendering with interpretation and reporting of computed tomography, magnetic resonance imaging, ultrasound, or other tomographic modality with image post-processing under concurrent supervision; not requiring image post-processing on an independent workstation	76376
3D rendering with interpretation and reporting of computed tomography, magnetic resonance imaging, ultrasound, or other tomographic modality with image post-processing under concurrent supervision; requiring image post-processing on an independent workstation	76377

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Description	CPT®
3D echocardiographic imaging and post-processing during transesophageal echocardiography, or during transthoracic echocardiography for congenital cardiac anomalies, for the assessment of cardiac structure(s) (e.g., cardiac chambers and valves, left atrial appendage, interatrial septum, interventricular septum) and function, when performed (List separately in addition to code for echocardiographic imaging) Code with (93303-93304, 93312, 93314, 93315, 93317, 93350-93351)	+93319

Doppler Echocardiography

Description	CPT®
Doppler echo, pulsed wave and/or spectral display	+93320
Doppler echo, pulsed wave and/or spectral display, follow-up or limited study	+93321
Doppler echo, color flow velocity mapping	+93325
CPT® 93320 and CPT® 93321 should not be requested or billed together	

C Codes

C codes are unique temporary codes established by CMS. C codes were established for contrast echocardiography. Each echocardiography C code corresponds to a standard echo code (Class I CPT® code) The C code and the matching CPT code should not both be approved.

C Code	Transthoracic Echocardiography	CPT®
C8921	TTE for congenital cardiac anomalies, complete	93303
C8922	TTE for congenital cardiac anomalies, follow-up or limited	93304
C8929	TTE with 2-D, M-mode, Doppler and color flow, complete	93306
C8923	TTE with 2-D, M-mode, without Doppler or color flow	93307
C8924	TTE with 2-D, M-mode, follow-up or limited	93308

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Myocardial strain imaging

Description	CPT®
Myocardial strain imaging using speckle tracking-derived assessment of myocardial mechanics (List separately in addition to codes for echocardiography imaging)	+93356

Investigational codes

Description	CPT®
Myocardial contrast perfusion echocardiography, at rest or with stress, for assessment of myocardial ischemia or viability	0439T
Noninvasive detection of heart failure derived from augmentative analysis of an echocardiogram that demonstrated preserved ejection fraction, with interpretation and report by a physician or other qualified health care professional	0932T

Transthoracic Echocardiography (TTE) – Coding General Information (CD-2.1.1)

- Complete transthoracic echocardiogram with spectral and color flow Doppler (CPT® 93306).
  - 93306 includes the Doppler exams, so CPT® codes 93320-93325 should not be assigned together with CPT® 93306.
  - Doppler codes (CPT® 93320, CPT® 93321, and CPT® 93325) are 'add-on codes' (as denoted by the + sign) and are assigned in addition to code for the primary procedure.
- For a 2D transthoracic echocardiogram without Doppler, report CPT® 93307.
- Limited transthoracic echocardiogram (CPT® 93308) should be billed if the report does not "evaluate or document the attempt to evaluate" all of the required structures.
  - A limited transthoracic echocardiogram is reported with CPT® 93308.
  - CPT® 93321 (not CPT® 93308 if Doppler is included in the study. CPT® 93325 can be reported with CPT® 93308 if color flow Doppler is included in the study.
  - A limited congenital transthoracic echocardiogram is reported with CPT® 93304.
- Doppler echo may be used for evaluation of the following:
  - shortness of breath
  - known or suspected valvular disease
  - known or suspected hypertrophic obstructive cardiomyopathy
  - shunt detection

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Background and Supporting Information

- Providers performing echocardiography on a pediatric individual may not know what procedure codes they will be reporting until the initial study is completed.
- If a congenital issue is found on the initial echocardiogram, a complete echocardiogram is reported with codes CPT® 93303, CPT® 93320, and CPT® 93325 because CPT® 93303 does NOT include Doppler and color flow mapping.
- If no congenital issue is discovered, then CPT® 93306 is reported alone and includes 2D, Doppler and color flow mapping.
- Since providers may not know the appropriate code(s) that will be reported at the time of the pre-authorization request, they may request all 4 codes (CPT® 93303, CPT® 93320, CPT® 93325, and CPT® 93306).
- CPT® 76376 and CPT® 76377 are not unique to 3D Echocardiography. These codes also apply to 3D rendering of MRI and CT studies. See **3D Echocardiography – Coding (CD-2.9)**.
- CPT® 93325 may also be used with fetal echocardiography.
- CPT® 93319 3D echocardiography imaging post-processing of TEE or TTE to evaluate congenital cardiac abnormalities. See **3D Echocardiography – Coding (CD-2.9)**.

Myocardial Contrast Perfusion Echocardiography (CD-2.11)

(CPT® 0439T)

- Investigational see **Transthoracic Echocardiography (TTE) – Coding (CD-2.1)**

Detection of Heart Failure with Preserved Ejection Fraction

Noninvasive detection of heart failure derived from augmentative analysis of an echocardiogram

Artificial intelligence (AI) applications are being used to extract patterns from medical images in an effort to add clinically relevant information to that obtained by physician interpretation of images. AI algorithms have been employed in the software analysis of echocardiograms in an effort to aid detection of heart failure. Further studies are needed to evaluate the accuracy, reliability, and clinical efficacy of these applications.

- CPT® 0932T Noninvasive detection of heart failure derived from augmentative analysis of an echocardiogram is considered experimental, investigational, or unproven at this time

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Transthoracic Echocardiography (TTE) Indications/Initial Evaluation (CD-2.2)

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Transthoracic Echocardiography (TTE) is medically necessary for the initial evaluation of **any** of the following:

Symptoms or signs suggesting cardiovascular disease

- chest pain/discomfort
- dyspnea/shortness of breath, or hypoxemia
- palpitations
- pre-syncope/syncope
- headache with transcranial Doppler evidence of a shunt or high-degree of suspicion for embolic process
- dependent lower extremity edema
- abnormal precordial or peripheral pulse
- new or changing heart murmur or click
- suspected hypertensive heart disease (initial evaluation)
- initial evaluation of known/suspected heart failure based on symptoms and/or signs
- suspected endocarditis with any:
  - fever
  - peripheral stigmata of endocarditis
  - new murmur
- history of rheumatic heart disease
- suspected pericardial diseases
- suspected cardiac injury due to blunt chest trauma

Diagnostic tests suggesting cardiovascular disease

- newly diagnosed RBBB or LBBB
- presence of pathological Q waves in 2 or more contiguous leads
- frequent VPCs defined as occurring more frequently than 30 times per hour or occurring in a pattern of bigeminy, trigeminy, or runs of ventricular tachycardia
- non sustained or sustained ventricular tachycardia (VT)
- ventricular fibrillation (VF)
- newly diagnosed atrial fibrillation/flutter
- cardiomegaly on a Chest x-ray or other imaging

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- elevated level of cardiac biomarkers (Creatinine Kinase isoforms, Troponin I or T, Brain Natriuretic Peptide (BNP), N-Terminal pro b-type natriuretic peptide (NT-proBNP) above the normal reference range as defined by the local laboratory assays)
- initial evaluation of known/suspected heart failure based on diagnostic tests
- suspected inherited or acquired cardiomyopathy (e.g., restrictive, infiltrative, dilated, hypertrophic)
- suspected pulmonary hypertension. See **Pulmonary Hypertension CD-8.1.**
  - ECG changes of right ventricular hypertrophy
  - right ventricular hypertrophy or pulmonary artery dilation on other imaging
  - pulmonary embolism with persistent or new symptoms
- dilated aortic root and/or ascending aorta seen on other imaging
- suspected endocarditis with positive blood cultures indicating bacteremia
- suspected pericardial diseases
- cardiac mass suspected on other imaging
- to rule out intra-cardiac thrombus in individuals with left ventricular systolic dysfunction prior to undergoing catheter ablation of ventricular arrhythmia.
- ≥6 weeks post-myocardial infarction

Extra-cardiac conditions and therapies associated with cardiovascular disease or risk

- CONDITIONS
  - Initial cardiac evaluation of a known systemic, congenital, or acquired disease that could be associated with structural heart disease
  - At risk for developing iron-overload cardiomyopathy (hereditary hemochromatosis or hereditary or acquired hematologic conditions requiring multiple transfusions)
  - Known or suspected connective tissue disease or a genetic condition that predisposes to an aortic aneurysm or dissection (may repeat every 2 years if negative). See **Screening for Vascular related genetic connective tissue Disorders PVD-2.2**.
  - At risk for pulmonary hypertension. See **Pulmonary Hypertension CD-8.1.**
    - scleroderma
    - lupus
    - mixed connective tissue disease
    - Hereditary Hemorrhagic Telangiectasia
    - individuals with pulmonary hypertension mutations (e.g., BMPR2)
  - Suspected pulmonary hypertension in the presence of:
    - liver disease
    - lung disease

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    - HIV
  - Newly diagnosed or strongly suspected cerebral ischemia or peripheral embolic event
- THERAPIES
  - Use of anorectic drugs, ergot derivatives, or other agents associated with valvular heart disease
  - Pre-procedure evaluation for TIPS (transjugular intrahepatic portosystemic shunt). See **Hepatic Arteries and Veins AB-43.1.**
  - Prior to solid organ transplant or hematopoietic stem cell transplant
  - Prior to exposure to cardiotoxic medications or radiation therapy. See **Cardiotoxic agent/Cancer Therapeutics-related Cardiac Dysfunction CD-12.1.**

Inherited cardiovascular conditions

- Individual has first-degree relative diagnosed with thoracic aortic aneurysm or dissection (may repeat every 2 years if negative). See **Screening for Vascular related genetic connective tissue Disorders PVD-2.2**, **Thoracic Aortic Aneurysm PVD-6.2.**
- Individual has first-degree relative diagnosed with bicuspid aortic valve. See **Screening for TAA in individuals with bicuspid aortic valves PVD-2.3.**
- Individual has diagnosed first-degree relative or member is genotype positive for an inherited cardiomyopathy including any of the following:
  - hypertrophic cardiomyopathy
  - non compaction cardiomyopathy
  - familial Dilated Cardiomyopathy
  - arrhythmogenic Cardiomyopathy (e.g., ARVC)

Additional indications

- One repeat echocardiogram is medically necessary if requested for agitated saline (bubble study) or contrast study (for evaluation of shunts or for left ventricular cavity opacification) when agitated saline or contrast were not administered at the time of the initial study and the results of the initial study indicate the need for agitated saline or contrast.
- Evaluation of congenital heart disease: see **Adult Congenital Heart Disease CD-11** and **Congenital Heart Disease PEDCD-2** in the Pediatric Cardiology imaging guidelines.

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Frequency of Echocardiography Testing (CD-2.3)

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Repeat Testing by Interval

Repeat Transthoracic Echocardiography (TTE) is medically necessary for **any** of the following:

Every 3 years

Valvular heart disease (See also below indications for Valve surgery or intervention)

- bicuspid aortic valve
- mild aortic stenosis or aortic valve sclerosis without stenosis
- mild aortic or mitral regurgitation
- any mitral stenosis that is not severe, mitral valve area >1.5 cm^2
- rheumatic valve changes with commissural fusion
- valve surgery including **any** of the following:
  - surgical valve repair
  - mechanical valve replacement
  - bioprosthetic valve replacement when <10 years since implant

Cardiomyopathy

- First-degree relative with a diagnosis of inherited cardiomyopathy including:
  - Hypertrophic Cardiomyopathy
  - Familial Dilated Cardiomyopathy
  - Idiopathic Dilated Cardiomyopathy
- Individuals genotype-positive for:
  - Familial Dilated Cardiomyopathy
  - Arrhythmogenic Cardiomyopathy (e.g., ARVC)

Every 2 years

Vascular disease

- First-degree relative with known thoracic aortic aneurysm or dissection a repeat echocardiogram is allowed every 2 years when both:

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- Prior aortic imaging (echo, CT, MR) is negative.
- Last aortic imaging was ≥2 years. See **Screening for Vascular related genetic connective tissue Disorders (PVD-2.2).**

Every year

Valvular heart disease

(See also below indications for valve surgery or intervention)

- moderate or severe regurgitation
- moderate or severe stenosis
- significant valve deformity (regardless of extent of regurgitation or stenosis) when there is documentation of either:
  - thickened myxomatous valve
  - bileaflet prolapse
- surgical bioprosthetic valve replacement when ≥10 years since implant
- post-transcatheter valve repair or replacement

Cardiomyopathy/heart failure

- history of left ventricular systolic dysfunction to evaluate the effectiveness of ongoing therapy
- hypertrophic cardiomyopathy see **Hypertrophic Cardiomyopathy CD-14**
- frequent right ventricular pacing >40%
- chronic LBBB
- frequent PVCs defined as >10,000 PVCs within 24 hours or >10% PVC burden over 24 hours
- left ventricular non-compaction cardiomyopathy
- at risk for developing iron-overload cardiomyopathy (hereditary hemochromatosis or hereditary or acquired hematologic conditions requiring multiple transfusions)
- inherited neuromuscular, metabolic, hematologic or cutaneous syndromes that are known to be associated with the development of cardiomyopathy

Pericardial disease

- Chronic pericardial effusions when findings would potentially alter therapy

Vascular disease

- Aortic root dilatation that has not yet been repaired. See **Thoracic aortic aneurysm PVD 6.2** and

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- For post-repair, see **Post-Aortic Endovascular/Open Surgery Surveillance Studies PVD-6.8**.

At risk for pulmonary hypertension

See **Pulmonary Hypertension CD-8.1**.

- Systemic Sclerosis or Scleroderma
- individuals with pulmonary hypertension mutations (e.g., BMPR2)
- first-degree relatives of individuals with pulmonary hypertension
- connective tissue disorder with symptoms consistent with pulmonary hypertension
- individuals with TR velocity ≥2.8 m/s, with no other findings on additional testing
- individuals being treated with medications associated with pulmonary hypertension
- individuals who have a concern documented for pulmonary hypertension and had a negative echocardiogram but still show signs or symptoms of pulmonary hypertension

Every 6 months or twice a year

Valvular heart disease

- Asymptomatic, severe mitral regurgitation if valve surgery is being considered

Pulmonary Hypertension

- See below indications for Pulmonary Hypertension (See **Pulmonary Hypertension CD-8.1**).
- Surveillance of stable individuals with moderate or severe pulmonary hypertension (pulmonary artery systolic pressure ≥50 mmHg)

Anytime

Repeat transthoracic echocardiogram is medically necessary **anytime** (without regard for the number or timing of previous echo studies) if there is a change in clinical status, or new signs and symptoms with documentation of **any** of the following:

- cardiac murmurs
- myocardial infarction or acute coronary syndrome
- congestive heart failure (new or worsening):
  - new symptoms of dyspnea
  - orthopnea
  - paroxysmal nocturnal dyspnea
  - elevated BNP
- known pericardial disease with clinical concern for cardiac tamponade or pericardial constriction
- infective endocarditis for any of the following:

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  - repeat imaging within 5-7 days for initially negative or inconclusive imaging when clinical suspicion of endocarditis remains high
  - new or worsening symptoms or signs of endocarditis
  - as needed to guide changes in antibiotic therapy
  - at completion of antibiotic therapy
- stroke/transient ischemic attack
- decompression illness
- prosthetic valve dysfunction or thrombosis
- cardiac transplant
- individuals with Left Ventricular Assist Device (LVAD)
- See also section on **Repeat testing per condition** below and **Left ventricular assist devices (LVAD) (CD-9.4)**.

Repeat Testing Per Condition

Repeat Transthoracic Echocardiography (TTE) is medically necessary for **any** of the following:

Valve surgery or intervention

Repeat Transthoracic Echocardiography (TTE) is medically necessary for any of the following:

Surgical valve repair or mechanical valve replacement
- 6 weeks post-surgery to establish baseline
- surveillance every 3 years after surgery

Surgical bioprosthetic valve replacement
- 6 weeks post-surgery to establish baseline
- surveillance every 3 years after surgery until 10 years
- then annually

TAVR follow-up
- 1 week after procedure to establish baseline
- 1 month post-procedure
- 1 year post-procedure
- then annually

Mitral Valve Repair (mitral valve clip) follow-up
- 1 month post-procedure
- 6 months post-procedure
- 1 year post-procedure

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- then annually

Transcatheter Tricuspid Valve Replacement follow-up

- TTE (CPT® 93306) is medically necessary post-procedure at the following intervals:
  - 1 month
  - 6 months
  - 1 year
  - then annually

See also **Transcatheter Aortic Valve Replacement (TAVR) (CD-4.8).**

See also **Percutaneous Mitral Valve Repair (mitral valve clip) (CD-13.5)**.

PFO closure, TIPS, Cardiac device therapy, LVAD

Repeat Transthoracic Echocardiography (TTE) is medically necessary for any of the following:

- **PFO Closure**
  - pre-operative evaluation for closure of PFO
  - post-procedural evaluation of PFO repair
  - 6 month follow-up after PFO repair
  - annually if there is a residual shunt on post-operative imaging

  For ASD closure, see **ASD-Atrial septal defects (CD 11.2.1)**.
- **TIPS (transjugular intrahepatic portosystemic shunt).** See **Hepatic Arteries and Veins (AB 43.1)**.
  - 1 time post-procedure for routine follow-up
  - **any time** post-procedure (either):
    - for new signs or symptoms
    - for concern for new or worsening pulmonary hypertension or heart failure
- **Cardiac device therapy**
  - Re-evaluation of EF is medically necessary 40 days after MI, 3 months after revascularization, or 3 months after maximally tolerated optimal medical therapy to determine either:
    - candidacy for device therapy
    - optimal choice of device
  - 1 time follow up within 12 months of implantation of a CRT-D device
- **Left ventricular assist device (LVAD).** See **Left ventricular assist devices (LVAD) (CD-9.4)**.
  - Prior to implant
  - Routine Post-implant at the following intervals:

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- 2 weeks
- 1 month
- 3 months
- 6 months
- 12 months
- every 6 months thereafter

Pulmonary hypertension

Follow-up echocardiogram (TTE) on individuals with PH is medically necessary for any of the following:

- every 6 months for surveillance of stable individuals with moderate or severe pulmonary hypertension (pulmonary artery systolic pressure ≥50 mm Hg)
- 3 months after any change in PH therapy
- prior to planned intubation (e.g., for elective surgery)
- prior to planned pregnancy
- during pregnancy as often as requested by provider
- anytime, without regard for the number or timing of previous echo studies to evaluate change in clinical signs or symptoms

See also **Pulmonary Hypertension CD-8.1**.

Hypertrophic Cardiomyopathy (HCM)

Repeat Transthoracic Echocardiography (TTE) is medically necessary for individuals with hypertrophic cardiomyopathy for any of the following:

- **Surveillance imaging**
  - Every year
- **Mavacamten: Initiation of treatment**
  - Baseline at the beginning of treatment
  - 4 weeks after treatment initiation
  - 8 weeks after treatment initiation
  - 12 weeks after treatment initiation
  - Then every 12 weeks while on mavacamten
- **Mavacamten: Changes in treatment**
  - 4 weeks after any interruption of treatment (any missed dose)
  - After any dosage change (including restart of treatment):
    - 4 weeks after dosage change
    - 12 weeks after dosage change
  - After initiating a weak CYP2C19 inhibitor (e.g., omeprazole) or moderate CYP3A4 inhibitor (e.g., ciprofloxacin):

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    - 4 weeks after start of medication
    - 12 weeks after start of medication
  - At any time regardless of timing of prior echo when there are new cardiac signs or symptoms, or worsening of clinical status
- **Post-Septal Reduction Therapy (SRT)**
  - Within 3 to 6 months after surgical myectomy or alcohol septal ablation

See also **Hypertrophic Cardiomyopathy CD-14**.

Cardiac Transplant

Anytime (without regard for the number or timing of previous ECHO studies) when there is a history of cardiac transplant, per transplant center protocol

Cardiotoxic Agents

For re-evaluation in an individual previously or currently undergoing therapy with cardiotoxic agents or radiation therapy, follow **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**.

Evidence Discussion

Transthoracic Echocardiogram

- Transthoracic echocardiogram (TTE) is an ultrasonic examination of the heart through the chest wall. Given that sound waves are used (sonography), there is no exposure to ionizing radiation or possible complications related to contrast induced nephropathy.
- It is readily accessible and transportable allowing for the test to be performed at multiple different locations with no need for blood work.
- TTE is widely accepted as initial imaging modality of choice for the general evaluation of cardiac and pericardial structure and function.
- Multiple evidence-based professional society guidelines indicate that TTE can be appropriately used for the evaluation of multiple cardiac issues including but not limited to evaluation of cardiac symptoms, dyspnea, and pulmonary hypertension.
- A complete comprehensive TTE will utilize multiple acoustic windows and incorporate 2D imaging with appropriate data; including measurements with color and spectral Doppler imaging.
- The test is performed and interpreted by qualified individuals in a facility that is accredited in performing echocardiograms.
- TTE is the primary imaging tool for screening of family members with thoracic aortic disease as well as genetic syndromes that are associated with thoracic aortic aneurysms and dissection. It is not medically necessary for screening of

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asymptomatic individuals in the general public in the absence of family history of cardiac, vascular, or associated connective tissue disorder.

- TTE offers real-time hemodynamic assessment which may be used in the guidance of therapeutic interventions.
- Repeat echocardiograms may be done for the surveillance of known valvular heart disease, cardiomyopathies, pulmonary hypertension, and cardiotoxic agents. The need for surveillance echocardiograms are dependent on factors such as morphology, severity, family history of cardiomyopathies, timing of surgery, left ventricular function, symptoms, and for follow-up of post cardiac structural interventions. Timing of surveillance echocardiograms are in alignment with recommendations from various national and international medical specialty organization guidelines and Appropriate Use Criteria based on studies which analyzed progression of valvular disease and timing of invasive intervention.
- Decisions regarding routine echocardiographic follow-up should not be based on the degree of regurgitation alone but should take into account associated structural valvular and cardiac abnormalities. For example, a structurally normal mitral valve with moderate mitral regurgitation by color flow Doppler and normal left atrial size, does not generally require routine echocardiographic follow-up. However, a thickened, myxomatous appearing mitral valve with bi-leaflet prolapse and only trivial or mild mitral regurgitation should be followed echocardiographically at routine intervals.

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Transesophageal Echocardiography (TEE) (CD-2.4) (CD-2.5)

CD.EC.0002.5.A
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Transesophageal Echocardiography (TEE)-Coding (CD-2.4)

TEE coding

Transesophageal Echocardiography	CPT®
TEE with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93312
TEE probe placement only	93313
TEE image acquisition, interpretation, and report only	93314
TEE for congenital anomalies with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93315
TEE for congenital anomalies, probe placement only	93316
TEE for congenital anomalies, image acquisition, interpretation and report only	93317

Doppler Echocardiography

Description	CPT®
Doppler echo, pulsed wave and/or spectral display	+93320
Doppler echo, pulsed wave and/or spectral display, follow-up or limited study	+93321
Doppler echo, color flow velocity mapping	+93325
Doppler echo, if performed, may be reported separately in addition to the primary TEE codes: CPT® 93312, CPT® 93314, CPT® 93315, and CPT® 93317	

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C codes

HCPCS	Description	CPT®
C8925	TEE with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93312
C8926	TEE for congenital anomalies with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93315

- The complete transesophageal echocardiogram service, including both (1) probe (transducer) placement and (2) image acquisition/interpretation, is reported with CPT® 93312.
  - Probe placement only is reported with CPT® 93313.
  - The image acquisition/interpretation only is reported with CPT® 93314.
- Physicians assign codes CPT® 93312, CPT® 93313, and/or CPT® 93314 to report professional services if the test is performed in a hospital or other facility where the physician cannot bill globally.
  - Modifier-26 (professional component) is appended to the appropriate code.
  - CPT® 93313 and CPT® 93314 should never be used together. If both services are provided, CPT® 93312 is reported.
- Hospitals should report TEE procedures using CPT® 93312 (the complete service). CPT® 93313 and CPT® 93314 are not used for hospital billing.

Transesophageal Echocardiography (TEE)- Indications (CD-2.5)

TEE (CPT® 93312, 93320, and 93325) is medically necessary when there is documentation of **any** of the following:

- limited transthoracic echo window when further information is needed to guide management (e.g., suspected or confirmed endocarditis, suspected intracardiac mass, etc.)
- assessing valvular dysfunction, especially mitral regurgitation when TTE is inadequate and intervention is being considered to repair/replace valve
- evaluation of cardiac mass, suspected tumor, or thrombus
- pre-procedural assessment of PFO/ASD
- pre-operative evaluation prior to planned LVAD implant
- embolic source or intracardiac shunting when TTE is inconclusive
  - Examples: atrial septal defect, ventricular septal defect, patent foramen ovale, aortic cholesterol plaques, thrombus in cardiac chambers, valve vegetation, tumor
- embolic events when there is an abnormal TTE or a history of atrial fibrillation

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  - clarify atria/atrial appendage, aorta, mitral/aortic valve beyond the information that other imaging studies have provided
- cardiac valve dysfunction
  - differentiation of tricuspid from bicuspid aortic valve in setting of aortic enlargement or significant stenosis or significant regurgitation
  - congenital abnormalities
- assessing for left atrial thrombus prior to cardioversion of atrial fibrillation or atrial flutter
- assessing for left atrial thrombus prior to planned atrial fibrillation ablation/pulmonary vein isolation procedure
- for initial imaging of ascending and descending thoracic aortic aneurysms
- For repeat imaging or established thoracic aneurysms, TEE is medically necessary **only** when imaging with CT or MR is contraindicated.
- TEE is medically necessary for the evaluation of individuals with hypertrophic cardiomyopathy if TTE is inconclusive for **any** of the following:
  - mitral regurgitation secondary to structural abnormalities of the mitral valve
  - subaortic membrane or aortic valve stenosis
- TEE is medically necessary for the pre-procedure planning for surgical myectomy or alcohol septal ablation evaluation of individuals with hypertrophic cardiomyopathy.
- Left atrial appendage (LAA) Closure device (e.g., WATCHMAN®) for any of the following:
  - pre-procedural evaluation
  - repeat imaging 45 days post-procedure
    - Another follow-up study is medically necessary before the 1-year surveillance when the imaging study at 45 days shows a peri-device gap ≥5 mm or Device Related Thrombus usually at 3-6 months.
  - 1 year post-procedure
  - See also **Percutaneous Mitral Valve Repair (mitral valve clip) (CD-13.5).**

Evidence Discussion

- Transesophageal echocardiogram (TEE) is a semi-invasive ultrasonic examination of the heart through the esophagus. Due to the proximity of the esophagus to the heart and great vessels, it allows for additional and more accurate information than transthoracic echocardiography for several specific diagnoses, catheter-based cardiac interventions, and cardiac surgery.
- TEE is utilized not only for diagnostic purposes but also for dynamic decision-making with cardioversion, surgical intervention, and assessment of surgical repair.
- TEE has become an essential imaging tool for pediatric and adult cardiologists, cardiac surgeons, and anesthesiologists and is used in outpatient and inpatient settings as well as operating rooms.

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- The test is performed and interpreted by physicians that have demonstrated both cognitive and technical competence in TEE.
- TEE is readily accessible, has no ionizing radiation exposure, and does not require use of IV contrast.
- There are relative and absolute contraindications which are reviewed prior to insertion of the TEE probe which include and are not limited to esophageal pathology, coagulopathy, and cervical spine injury.

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Stress Echocardiography (Stress Echo) (CD-2.7)

CD.EC.0002.7.A
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Stress Echo – Coding

Associated codes

Stress Echocardiography	CPT®
Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report;	93350
Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report: including performance of continuous electrocardiographic monitoring, with physician supervision	93351
Doppler Echocardiography	
Doppler echo, pulsed wave and/or spectral display	+93320
Doppler echo, pulsed wave and/or spectral display, follow-up/limited study	+93321
Doppler echo, color flow velocity mapping	+93325

Associated HCPCS codes

CPT®	Stress Echocardiography	HCPCS
93350	Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report;	C8928
93351	Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report: including performance of continuous electrocardiographic monitoring, with physician supervision	C8930

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CPT® 93350 and CPT® 93351 do not include Doppler studies.

Doppler echo (CPT® +93320 and CPT® +93325), if performed, may be reported separately in addition to the primary SE codes: CPT® 93350 or CPT® 93351.

Stress Echocardiogram-Indications Other Than Ruling-out CAD (CD-2.7)

CPT® 93350 or 93351

- See: **Stress Testing with Imaging – Indications (CD-1.4)**
- In addition to the evaluation of CAD, stress echo can be used to evaluate the following conditions:
  - dyspnea on exertion (specifically to evaluate pulmonary hypertension)
  - right heart dysfunction
  - valvular heart disease when the outcome would affect a therapeutic or interventional decision
  - pulmonary hypertension when the outcome will measure response to therapy and/or prognostic information
  - hypertrophic cardiomyopathy (as defined in **Obstructive Hypertrophic Cardiomyopathy (HCM) (CD-12.3)** for **either** of the following:
    - Exercise stress echo (CPT® 93351 or 93350) is medically necessary for the detection and quantification of dynamic left ventricular outflow tract obstruction in symptomatic individuals with HCM who do **not** have a resting or provocable outflow tract gradient ≥50 mm Hg on TTE.
    - Stress echo can be repeated when there is documentation of **any** of the following:
      - in 1 to 2 years if the resting or provocable outflow tract gradient is <30 mm Hg on prior stress echo
      - worsening symptoms
      - There has been a therapeutic change (i.e., change in medication, surgical procedure performed).
- In general, spectral Doppler (CPT® 93320 or 93321) and color-flow Doppler (CPT® 93325) are medically necessary in the evaluation of the above conditions and can be added to the stress echo code.

Evidence Discussion

- Stress echocardiography is a ultrasonic examination of the heart through the chest wall during rest and stress. Given the ultrasound modality, there is no exposure to ionizing radiation or concern for contrast induced renal failure.

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- Stress echocardiography is used to assess for global and regional systolic function and evaluation of valvular function at rest and during stress. This allows for non-invasive diagnosis and management of coronary artery disease, various cardiomyopathies, pulmonary hypertension, and valvular heart disease.
- A complete comprehensive stress echocardiogram will incorporate 2D imaging with multiple acquisition windows during rest and stress and may also incorporate color and spectral Doppler imaging. This provides a dynamic evaluation of myocardial and valvular structure and function under physiological (exercise) or pharmacological stress.
- The test is performed and interpreted by qualified individuals in a facility that is proficient in echocardiograms in compliance with published criteria for quality cardiac diagnostic testing.

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3D Echocardiography (CD-2.9)

CD.EC.0002.9.A
v1.0.2026

3D Echocardiography – Coding

Description	CPT®
3D rendering with interpretation and reporting of computed tomography, magnetic resonance imaging, ultrasound, or other tomographic modality with image post-processing under concurrent supervision; not requiring image post-processing on an independent workstation	76376
3D rendering with interpretation and reporting of computed tomography, magnetic resonance imaging, ultrasound, or other tomographic modality with image post-processing under concurrent supervision; requiring image post-processing on an independent workstation	76377
3D echocardiographic imaging and post-processing during transesophageal echocardiography, or during transthoracic echocardiography for congenital cardiac anomalies, for the assessment of cardiac structure(s) (e.g., cardiac chambers and valves, left atrial appendage, interatrial septum, interventricular septum) and function, when performed (List separately in addition to code for echocardiographic imaging) Code with (93303-93304, 93312, 93314, 93315, 93317, 93350-93351)	+93319

3D Echocardiography – Indications (CD-2.9)

Echocardiography with 3-dimensional (3D) rendering is becoming universally available, yet its utility remains limited based on the current literature.

- CPT ® 93319 with one of the following (CPT® 93303, 93304, 93312, 93314, 93315, or 93317) for congenital cardiac abnormalities
- 3D Echo (CPT® 76376 or CPT® 76377) may be medically necessary when a primary echocardiogram is approved and **one** of the following is needed:
  - left ventricular volume and ejection fraction assessment when measurements are needed for treatment decision (e.g., implantation of ICD, alteration in cardiotoxic chemotherapy, cardiac transplant)

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- mitral valve anatomy specifically related to mitral valve stenosis
- pre-procedural evaluation of left atrial appendage occlusion (e.g., WATCHMAN®)
- pre-operative evaluation for LVAD implant
- guidance of transcatheter procedures such as:
  - mitral valve clipping
  - TAVR
  - left atrial appendage closure device (e.g., WATCHMAN®)

Evidence Discussion

- 3D echocardiography (3DE) is a newer modality of ultrasonic examination of the heart through the chest wall or through the esophagus that is added on to either a transthoracic or transesophageal echocardiogram.
- This allows for both real-time and post-processed 3D analysis of the cardiac structures and function. This produces images that are less constrained than that of 2D echocardiograms.
- 3DE is readily accessible and transportable allowing for the test to be done at multiple different locations with no need for blood work or IV line insertion.
- 3DE has become important in pre-surgical planning, guidance of catheter intervention, and functional assessment of the heart in various cardiac conditions including but not limited to congenital heart disease, valvular disease, and structural heart disease.

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Myocardial Strain Imaging (CD-2.12)

CD.EC.0002.12.A
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CPT® 93356

- Myocardial strain imaging (CPT® 93356, speckle tracking longitudinal strain) is medically necessary for the initial evaluation of LVH, in addition to the primary echocardiogram, when there is documentation of **both**:
  - unclear etiology of LVH not due to hypertensive heart disease
  - concern for infiltrative cardiomyopathy
- Myocardial strain imaging is medically necessary in heart transplant evaluation and surveillance per transplant protocol.
- Myocardial strain imaging (CPT® 93356) in addition to the primary echocardiogram in individuals receiving therapy with cardiotoxic agents for **any** of the following:
  - Initial evaluation-prior to treatment with **either**:
    - medications that could result in cardiotoxicity/heart failure
    - radiation that could result in cardiotoxicity/heart failure
  - re-evaluation of an individual previously or currently undergoing therapy as per echocardiogram parameters (see **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1))**
  - re-evaluation of an individual undergoing therapy with worsening symptoms

Evidence Discussion

- Myocardial strain imaging or speckle-tracking echocardiography (STE) is a modern ultrasound technique that is an adjunct to traditional transthoracic echocardiography to evaluate myocardial deformation.
- Given the ultrasound modality, there is no exposure to ionizing radiation or concern for contrast-induced renal failure.
- The main areas of application of this technique have been in the assessment of myocardial mechanics, ischemic heart disease, cardiomyopathies, LV diastolic dysfunction, and in detecting sub-clinical myocardial dysfunction in individuals undergoing chemotherapy for cancer or in those affected by heart valve diseases.
- STE is considered the optimal deformation parameter for the detection of subclinical LV dysfunction which allows for clinical intervention prior to reduction in LVEF particularly useful in the field of cardio-oncology.

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CD.EC.0002.A
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Nuclear Cardiac Imaging

Guideline

Myocardial Perfusion Imaging (MPI)(CD-3.2)
Multi-gated Acquisition (MUGA) Coding (CD-3.3)
MUGA Study – Cardiac Indications (CD-3.4)
Myocardial Sympathetic Innervation Imaging in Heart Failure (CD-3.6)
Non-imaging Heart Function and Cardiac Shunt Imaging (CD-3.7)
References (CD-3)

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Myocardial Perfusion Imaging (MPI) (CD-3.2)

CD.NC.0003.1.A
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MPI – Coding

Nuclear Cardiac Imaging Procedure Codes

Myocardial Perfusion Imaging (MPI)	CPT®
MPI, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78451
MPI, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78452
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78453
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78454
Absolute quantitation of myocardial blood flow (AQMBF), single-photon emission computed tomography (SPECT), with exercise or pharmacologic stress, and at rest, when performed (List separately in addition to code for primary procedure)	+0742T

- The most commonly performed myocardial perfusion imaging are single (at rest or stress, CPT® 78451) and multiple (at rest and stress, CPT® 78452) SPECT studies.

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Less commonly ordered are planar studies (CPT 78453 and 78454) see **Stress Testing with Imaging – Indications (CD-1.4)**.

- Evaluation of the individual's left ventricular wall motion and ejection fraction are routinely performed during MPI and are included in the code's definition.
- First pass studies, (CPT® 78481 and CPT® 78483), MUGA, (CPT® 78472 and CPT® 78473) and SPECT MUGA (CPT® 78494) should not be reported in conjunction with MPI codes.
- Attenuation correction, when performed, is included in the MPI service by code definition. No additional code should be assigned for the billing of attenuation correction.

- **Multi-day Studies:** In the absence of written payer guidelines to the contrary, it is not appropriate to bill separately for the rest and stress segments of MPI even if performed on separate calendar dates. A single code is assigned to define the entire procedure on the date all portions of the study are completed.

Note:

3D rendering should not be billed in conjunction with MPI.

MPI – Indications (CD-3.2)

See: **Stress Testing with Imaging – Indications (CD-1.4)**

Absolute quantitation of myocardial blood flow (AQMBF)(SPECT)

AQMBF obtained by CZT-SPECT is considered experimental, investigational, or unproven at this time.

Evidence Discussion

Appropriate Use Criteria from major professional societies including the American College of Cardiology and the American Heart Association support the use of myocardial perfusion imaging (MPI) with single photon emission computed tomography (SPECT) for individuals with signs or symptoms consistent with coronary artery disease (CAD) such as typical angina. In addition, they support the use of SPECT perfusion imaging in symptomatic individuals with known CAD and those with prior interventions such as coronary artery bypass surgery, coronary stenting, and preoperative risk stratification. These guidelines balance the appropriate testing of individuals with SPECT versus medically unnecessary and potentially harmful testing and downstream procedures.

Also, as supported by society guidelines, use of SPECT is medically necessary in detection of cardiac transthyretin amyloidosis (ATTR) after light chain amyloidosis (AL) is ruled out by appropriate blood and urine testing.

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Multi-gated Acquisition (MUGA) Coding (CD-3.3)

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Cardiac blood pool imaging, or radionuclide ventriculography, can be used to evaluate ventricular function. Cardiac blood pool imaging includes first pass studies (CPT® 78481 and 78483) as well as gated equilibrium studies (CPT® 78472, 78473, 78494, and +78496).

Gated equilibrium studies can also be referred to as multi-gated acquisition (MUGA) scan or equilibrium radionuclide angiography (ERNA). Imaging for gated equilibrium studies can be planar or 3D (single photon emission computed tomography, SPECT).

Of note, all cardiac blood pool imaging is synchronized with electrocardiographic RR interval (EKG-gated); thus, regular rhythm is required for accurate LV assessment.

Gated Equilibrium Studies – Planar	CPT®
Cardiac blood pool imaging, gated equilibrium; planar, single study at rest or stress, wall motion study plus ejection fraction, with or without quantitative processing	78472
Cardiac blood pool imaging, gated equilibrium; planar, multiple studies, wall motion study plus ejection fraction, at rest and stress, with or without additional quantification	78473
Gated Equilibrium Studies-SPECT	
Cardiac blood pool imaging, gated equilibrium, SPECT, at rest, wall motion study plus ejection fraction, with or without quantitative processing	78494
First Pass studies	
Cardiac blood pool imaging (planar), first pass technique; single study, at rest or with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78481

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Gated Equilibrium Studies – Planar	CPT®
Cardiac blood pool imaging (planar), first pass technique; multiple studies, at rest and with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78483
Cardiac blood pool imaging, gated equilibrium, single study, at rest, with right ventricular ejection fraction by first pass technique (List separately in addition to code for primary procedure) This CPT code is an add-on code to 78472.	+78496

- The technique employed for a MUGA service guides the code assignment.
  - CPT® 78472 is used for a planar MUGA scan at rest or stress.
  - CPT® 78473 for planar MUGA scans, multiple studies at rest and stress.
- Planar MUGA studies (CPT® 78472 and CPT® 78473) should not be reported in conjunction with:
  - SPECT MPI (CPT® 78451-CPT® 78454)
  - First pass studies (CPT® 78481-CPT® 78483)
  - SPECT MUGA (CPT® 78494).
- CPT® +78496 is assigned only in conjunction with CPT® 78472.

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MUGA Study – Cardiac Indications (CD-3.4)

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MUGA (Multi Gated Acquisition) – Blood Pool Imaging Indications

- Echocardiography is the preferred method of following left ventricular systolic function.
- MUGA may be medically necessary when a recent ECHO, as indicated in **Transthoracic Echocardiography (TTE) – Indications (CD-2.2)** and/or **Frequency of Echocardiography Testing (CD 2.3)**, was technically limited and prevented accurate assessment of left ventricular function.
- MUGA is medically necessary when there is a significant discrepancy between LVEF assessment by ECHO and another modality (i.e., one study reports normal LVEF and the other, a reduced LVEF) AND there is clear documentation as to how quantitative measurement of LVEF will affect individual management (e.g., implantation of an ICD, alteration in cardiotoxic chemotherapy, etc.).
- MUGA may be performed in place of an ECHO in the following circumstances:
  - to determine candidacy for ICD/CRT and/or to determine optimal choice of device in individuals who meet criteria for ICD based on ejection fraction and other criteria
  - when previously or currently undergoing therapy with potentially cardiotoxic agents, including chemotherapy and radiation, AND a history of previous low LV ejection fraction (LVEF <50%). See **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**.
- MUGA is not medically necessary when requested simply to compare LVEF by the same modality. A prior MUGA is not a reason to approve another MUGA.

Right ventricular first pass study

- (CPT® 78472 and 78496) may be performed when ECHO is technically limited and prevents accurate assessment of RV function AND when further information about RV function is needed to guide management (e.g. established/diagnosed pulmonary hypertension, suspected or confirmed pulmonary embolus).

First pass studies

- First pass studies (CPT® 78481 and CPT® 78483) may be approved in place of MUGA when indications are met for MUGA and/or there is need for information that cannot be obtained by MUGA.

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- First pass studies, (CPT® 78481 and CPT® 78483), MUGA (CPT® 78472 and CPT® 78473), and SPECT MUGA (CPT® 78494) should not be reported in conjunction with MPI codes.

Evidence Discussion

Multi-gated acquisition (MUGA) imaging (also referred to as radionuclide angiography, gated blood pool scan, equilibrium radionuclide angiography) is a method to measure ejection fraction and wall motion of the heart.

Appropriate Use Criteria from major professional societies including the American College of Cardiology and the American Heart Association support the use of planar imaging and/or single photon emission computed tomography (SPECT) of labeled red blood cells for individuals who require accurate assessment of ejection fraction and/or wall motion when echocardiography or other imaging approaches are inadequate or disparate. This includes individuals with cardiomyopathy and those exposed to cardio-toxic agents.

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Myocardial Sympathetic Innervation Imaging in Heart Failure (CD-3.6)

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- Nuclear imaging using I-123-meta-iodobenzylguanidine (I-123-mIBG) in an attempt to image increased myocardial sympathetic activity is considered to be experimental and investigational.
- The AMA has established the following set of Category III codes to report these studies:
  - CPT® 0331T Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment
  - CPT® 0332T Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment; with tomographic SPECT

Background and Supporting Information

In heart failure, the sympathetic nervous system is activated in order to compensate for the decreased myocardial function. Initially, this is beneficial. However, long-term this compensatory mechanism is detrimental and causes further damage.

Markers have been developed using radioactive iodine in an attempt to image this increased myocardial sympathetic activity. Currently, AdreView™ (Iodine-123 meta-iodobenzylguanidine) is the only FDA-approved imaging agent available for this purpose.

Evidence Discussion

I-123-meta-iodobenzylguanidine (MIBG) imaging of the sympathetic nerve activity of the heart has been proposed and approved for the identification of individuals with heart failure. However, its clinical utility has not found widespread acceptance, and its clinical usefulness remains in question. There are no societal guidelines for its routine use. The guidelines contain many other imaging platforms such as echocardiography, magnetic resonance imaging, perfusion, and metabolic imaging that have proven superior for the diagnosis and management of individuals with heart failure.

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Non-imaging Heart Function and Cardiac Shunt Imaging (CD-3.7)

CD.NC.0001.7.A
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- Procedures reported with CPT® 78414 and CPT® 78428 are essentially obsolete but are medically necessary when other preferred modalities are unavailable or non-diagnostic.
- Echocardiogram is the preferred method for cardiac shunt detection, rather than the cardiac shunt imaging study described by CPT® 78428.
- Ejection fraction can be obtained by echocardiogram, SPECT MPI, MUGA study, cardiac MRI, cardiac CT, or cardiac PET depending on the clinical situation, rather than by the non-imaging heart function study described by CPT® 78414.

Evidence Discussion

Non-imaging Heart Function and Cardiac Shunt Imaging radionuclide techniques are no longer in use in current clinical practice. These techniques have been rendered obsolete and have been replaced by other cardiac imaging modalities that provide far superior structural and functional information to guide clinical management decisions. The preferred cardiac imaging modalities in current clinical use that have replaced the obsolete techniques include echocardiography, SPECT MPI, MUGA study, cardiac MRI, cardiac CT, and cardiac PET.

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Cardiac CT

Guideline

Cardiac CT and CTA-General information and Coding (CD-4.1)
CT for Coronary Calcium Scoring (CD-4.2)
CCTA – Indications for CCTA (CD-4.3)
CCTA – Regardless of Symptoms (CD-4.4)
Anatomical and Functional Analysis of CCTA Results (CD-4.5)
CT Heart for Evaluation of Cardiac Structure and Morphology (CD-4.6)
CT Heart for Congenital Heart Disease (CD-4.7)
Transcatheter Aortic Valve Replacement (TAVR) (CD-4.8)
3D Predictive Model Generation for Pre-planning of Cardiac Procedure (CD-4.9)
References (CD-4)

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Cardiac CT and CTA-General information and Coding (CD-4.1)

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Associated Codes

Cardiac Imaging Procedure Codes

Cardiac CT and CCTA	CPT®
CT, heart, without contrast, with quantitative evaluation of coronary calcium • The code set for Cardiac CT and CCTA (CPT® 75572-CPT® 75574), include quantitative and functional assessment (for example, calcium scoring) if performed • CPT® 75571 describes a non-contrast CT of the heart with calcium scoring and should be reported only when calcium scoring is performed as a stand-alone procedure. ◦ Can be used to report a preliminary non-contrast scan which indicates an excessive amount of calcium such that the original scheduled study must be discontinued. ◦ CPT® 75571 should not be reported in conjunction with any of the contrast CT/CTA codes (CPT® 75572-CPT® 75574).	75571
CT, heart, with contrast, for evaluation of cardiac structure and morphology (including 3D image post-processing, assessment of cardiac function, and evaluation of venous structures, if performed).	75572
Computed tomography, heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease (including 3D image post-processing, assessment of left ventricular [LV] cardiac function, right ventricular [RV] structure and function and evaluation of vascular structures, if performed).	75573
CTA, heart, coronary arteries and bypass grafts (when present), with contrast, including 3D image post-processing (including 3D image post-processing, assessment of cardiac function, and evaluation of venous structures, if performed).	75574

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Cardiac CT and CCTA	CPT®
Quantification and characterization of coronary atherosclerotic plaque to assess severity of coronary disease, derived from augmentative software analysis of the data set from a coronary computed tomographic angiography, with interpretation and report by a physician or other qualified health care professional	75577
Non-invasive estimate of coronary fractional flow reserve (FFR) derived from augmentative software analysis of the data set from a coronary computed tomography angiography, with interpretation and report by a physician or other qualified health care professional	75580

Cardiac CT and CTA-General information

- Only 1 code from the set: CPT® 75572-CPT® 75574 can be reported per encounter.
- CPT® 75574 includes evaluation of cardiac structure and morphology when performed; therefore, additional cardiac CT code/s should not be assigned.
- Automated quantification and characterization of coronary atherosclerotic plaque (ordered as CPT® 0623T, or 0624T, 0625T, 0626T) is a service in which coronary computed tomographic angiography (CCTA) data are analyzed using computerized algorithms to assess the extent and severity of coronary artery disease.

Background and Supporting Information

The high negative predictive value (98%-99%) of CCTA in ruling out significant coronary artery disease has been confirmed in multiple studies.

3D rendering should not be billed in conjunction with Cardiac CT and CCTA.

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CT for Coronary Calcium Scoring (CD-4.2)

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Coronary artery calcium score (CPT® 75571) is **not** supported for evaluation of CAD in symptomatic individuals.

CT Calcium Scoring-Asymptomatic and for CAD Screening

- Coronary artery calcium score (CPT® 75571) is medically necessary when there is documentation of **all** of the following:
  - Results will impact risk-based decisions for preventive interventions.
  - an LDL-C level ≥70 mg/dL (1.8 mmol/L) AND <190 mg/dL (4.9 mmol/L)
  - Individual is an adult age 40-75.
  - 10-year ASCVD risk including pooled cohort equation is between 5.0% to 19.9%
  - There is no documented CAD.
  - Individual is not currently on a statin.
  - Individual is not a smoker.
  - There is no history of diabetes.
  - There is no family history of premature CAD (occurring before age 56 in males or before age 66 in females).
  - There has been no calcium score performed in the previous 5 years.
  - There has been no prior calcium score >0.
- Coronary calcium scoring is not medically necessary in someone with known CAD.

Suspected or Known Low Gradient Aortic Stenosis

- Coronary artery calcium score (CPT® 75571) is medically necessary in low gradient aortic stenosis when symptomatic, severe aortic stenosis is suspected. Low gradient aortic stenosis is defined as an AVA <1 and a mean gradient <40 mmHg.

Individuals with a history of chest radiation therapy

CT calcium scoring is considered medically necessary to evaluate for radiation induced CAD when all of the following conditions exist:

- The individual has no symptoms suspicious for ischemia such as chest discomfort located retrosternally, epigastric, or referred pain (pain that is felt in a place different

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than the source) in the neck, shoulder, arm, or jaw described as pain, pressure, constricting, or tightness occurring with exertion or emotional stress and relieved by rest, nitroglycerin, or both.
- The individual has no clinical history or prior cardiac imaging documenting CAD.
- There has been no prior calcium score >0.
- There has been ≥5 years since the last RT (radiotherapy/radiation) treatment for the initial study or at least 5 years since the last CT calcium scoring for subsequent testing.

Evidence Discussion

The identification of coronary artery calcium (CAC) on a non-contrast computed tomography signifies the presence of coronary atherosclerosis and predicts major cardiac events independent of clinical risk factors. Measuring CAC score has been widely adopted to assist in the risk reclassification of coronary heart disease and to serve as an arbitrator for statin and aspirin therapy initiation. Whilst this general screening strategy brings precision to risk assessment, the most cost-effective implementation algorithm has not been studied systematically. This is reflected in the variation of major global and inter-societal guidelines for CAC scoring with unknown public health and economic ramification. Based on moderate quality, non-randomized evidence, the American College of Cardiology and the American Heart Association have specified the following clinical scenarios where CAC scoring is most likely to impact risk-based decision for preventive interventions:

- an LDL-C level ≥70 mg/dL (1.8 mmol/L) AND <190 mg/dL (4.9 mmol/L)
- Individual is an adult age 40-75.
- 10-year Atherosclerotic Cardiovascular Disease (ASCVD) risk including pooled cohort equation is between 5.0% to 19.9%.
- There is no documented coronary artery disease (CAD).
- Individual is not currently on a statin.
- Individual is not a smoker.
- There is no history of diabetes.
- There is no family history of premature CAD.
- There has been no calcium score performed in the previous 5 years.
- There has been no prior calcium score >0.

In addition to CAC, the degree of calcification of the aortic valve has been demonstrated to correlate with the degree of aortic stenosis and prognosis. The utility of calcium scoring has been extended to the aortic valve in the setting of symptomatic, low gradient aortic stenosis (defined as an aortic valve area of less than 1 cm^2 and mean gradient of less than 40 mmHg), when severe aortic stenosis is suspected.

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CCTA – Indications for CCTA (CD-4.3)

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CCTA (CPT® 75574) is medically necessary for any of the following:

- new, recurrent, or worsening cardiac chest pain as defined in **General Guidelines (CD-1.0)**
- new, recurrent, or worsening symptoms of chest pain, or unexplained exertional dyspnea, or unexplained exertional fatigue, and any of the following:
  - Symptoms persist after a normal stress test.
  - equivocal, borderline, abnormal or discordant prior noninvasive evaluation since the onset of symptoms where obstructive coronary artery disease remains a concern
  - new abnormal rest ECG findings since the onset of symptoms, such as a new LBBB or T-wave inversions (not including isolated T wave inversion in III or leads v1-2), when ischemia is a concern
  - a history of prior CABG when the **only** concern is for graft patency
  - Individual has undergone an urgent evaluation (ER or urgent care) of the symptoms since the onset.
  - history of cardiac transplant (e.g., concern for transplant vasculopathy)
  - history of chest radiation
  - Chest pain occurring with syncope
  - Coronary artery calcium score (CAC) score ≥100
  - Individuals 50 years of age or above with 2 or more of the following CV risk factors:
    - diabetes
    - current cigarette smoking
    - family history of premature CAD
    - hypertension
    - hyperlipidemia
    - history of adverse pregnancy outcomes
- Evaluation of an individual under the age of 40 for suspected anomalous coronary artery(ies) or for treatment planning when there is a history of one or more of the following:
  - syncopal episodes during strenuous activities
  - persistent chest pain brought on by exertion or emotional stress and normal stress test
  - full sibling(s) with history of sudden death syndrome before age 40 or with documented anomalous coronary artery

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  - resuscitated sudden death and contraindications for conventional coronary angiography
  - prior nondiagnostic coronary angiography in determining the course of the anomalous coronary artery in relation to the great vessels, origin of a coronary artery or bypass graft location (any):
    - Anomalies of origin:
      - LCA or the RCA arising from the pulmonary artery
      - interarterial course between the pulmonary artery and the aorta of either the RCA arising from the left sinus of Valsalva or the LCA arising from the right sinus of Valsalva
    - Anomalies of course:
      - myocardial bridging
    - Anomalies of termination:
      - coronary artery fistula
- Initial imaging study in individuals with hypertrophic cardiomyopathy and stable anginal symptoms
  - Chest discomfort is common in individuals with hypertrophic cardiomyopathy. The incidence of false positive myocardial perfusion imaging abnormalities is higher in these individuals, whereas the incidence of severe coronary artery stenosis is low.
- Individuals who have recovered from unexplained sudden cardiac arrest in lieu of invasive coronary angiography **(both)**:
  - confirm the presence or absence of ischemic heart disease
  - exclude the presence of an anomalous coronary artery

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CCTA – Regardless of Symptoms (CD-4.4)

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- Evaluation of newly diagnosed congestive heart failure or cardiomyopathy (all):
  - No prior history of coronary artery disease, the ejection fraction is <50%
  - No contraindications to cardiac CT angiography
  - No cardiac catheterization, SPECT, cardiac PET, or stress echocardiogram has been performed since the diagnosis of congestive heart failure or cardiomyopathy.
- Unclear coronary artery anatomy despite conventional cardiac catheterization
- Evaluation of structural heart disease for use of class IC antiarrhythmic agent (flecainide or propafenone) in lieu of stress test with imaging
- Ventricular tachycardia (3-beat runs or greater), exercise induced ventricular tachycardia or ventricular fibrillation
- Re-do CABG **(either)**:
  - assess bypass graft patency
  - evaluate the location of the left internal mammary artery (LIMA) and or right internal mammary artery (RIMA) prior to repeat bypass surgery
- Follow-up Left main stent 1 time at 6-12 months
- Pre-procedural planning for Percutaneous Coronary Intervention (PCI) of Chronic Total Occlusion (CTO)
- Evaluate coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels:
  - To evaluate the great vessels, CTA Chest (CPT® 71275) can be performed instead of CCTA or in addition to CCTA.
  - For anomalous pulmonary venous return, can add CT Abdomen and Pelvis with contrast (CPT® 74177).
  - See **Adult Congenital Heart Disease CD-11** for lesion specific imaging.
- When CCTA will replace conventional invasive coronary angiography for any of the following:
  - Delayed presentation or retrospective evaluation of suspected Takotsubo syndrome (stress cardiomyopathy)
  - Preoperative assessment of the coronary arteries in planned surgery for any of the following:
    - Aortic dissection

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    - Aortic aneurysm
    - Valvular surgery
    - Liver transplant (for initial pre-transplant evaluation and may be repeated once in 3 years)
  - To assess for coronary involvement in individuals with systemic vasculitis (e.g. Giant Cell Arteritis, Takayasu's, Kawasaki's disease) when there are clinical features suggestive of underlying vasculitis including:
    - unexplained elevated cardiac markers (erythrocyte sedimentation rate, C-reactive protein)
    - constitutional symptoms (fever, chills, night sweats, weight loss)
    - multiple visceral infarcts in the absence of embolic etiology
- For **Cardiac Trauma**, see also **Cardiac Trauma – Imaging (CD-10.1)**.
- Preoperative assessment for planned liver or kidney transplant

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Anatomical and Functional Analysis of CCTA Results (CD-4.5)

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Fractional flow reserve by computed tomography (CPT® 75580)

Fractional flow reserve (FFR) is typically measured using invasive techniques. FFR can be obtained non-invasively from coronary computed tomography angiography data (FFR-CT).

- Indications:
  - FFR-CT is medically necessary to further assess CAD seen on a recent CCTA that is of uncertain physiologic significance

Coronary CTA plaque quantification

CPT® 75577

Coronary CT plaque quantification or coronary plaque analysis using data from CCTA is medically necessary when **all** of the following conditions exist:

- The individual has acute or stable chest pain with no known coronary artery disease (CAD).
- Results from the current completed and interpreted CCTA indicate any of the following risk categories:
  - intermediate risk
  - CAD-RADS 1
  - CAD-RADS 2
  - CAD-RADS 3
- Cardiac evaluation is negative or inconclusive for acute coronary syndrome.

Non-indications

- Coronary CTA plaque quantification or coronary plaque analysis using data from CCTA is not medically necessary in any of the following clinical scenarios:
  - unstable coronary syndromes
  - in conjunction with invasive coronary catheterization
  - for the purpose of screening or disease surveillance
  - within 30 days of a myocardial infarction
  - Current completed and interpreted CCTA documents any of the following results:
    - Normal CCTA
    - CAD-RADS-0

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- High-grade stenosis (>70%)
- CAD-RADS 4
- CAD-RADS 5

Note:

See **General Guidelines CD 1.0** for **CAD-RADS** classifications

Evidence Discussion

Coronary Computed Tomography Angiography (CCTA) provides non-invasive anatomic coronary imaging with excellent sensitivity for detecting coronary plaques and stenosis. However, artifacts can significantly lower the specificity, which potentially leads to higher healthcare resource utilization. Notwithstanding this concern, the ability to characterize coronary plaques and stenosis on a CCTA proved to be advantageous over functional stress testing, with respect to diagnostic characteristics and prognostication, as demonstrated by high-quality evidence, including multiple randomized trials.

To ensure a clinically meaningful study, in addition to paying meticulous attention to the scanner functionality and protocol selection, careful individual selection is imperative. Based on the known performance characteristics of CCTA and high-quality clinical trials from Europe and North America, there is general agreement between the American and European guidelines to focus CCTA testing on the following general categories:

- Cardiac chest pain with no known coronary artery disease (CAD)
- Absence of symptoms or possible cardiac chest pain with no known CAD, when objective evidence of cardiac structure or function abnormality is present
- Information about specific coronary artery abnormalities, such as left main coronary stent and bypass graft patency, congenital coronary anomaly, non-atherosclerotic coronary disease may potentially alter clinical management decision.
- CCTA will replace conventional invasive coronary angiography when the likelihood of coronary intervention is not high based on clinical assessment.

CTA derived fractional flow reserve (FFRct) is one of the value-added technologies that have been shown to improve the accuracy of CAD diagnosis over and above CCTA alone. While it modestly adds negative predictive value to CCTA by excluding critical coronary lesion with a FFRct value >0.8, the positive predictive value is inadequate to confer confidence to locate lesions with significant stenosis. Effective adoption of this technology, therefore, is contingent upon ensuring optimal CCTA image quality and a mindful individual selection process; for example, avoid applying FFRct to those with left main coronary disease ≥50% or those with critical triple vessel disease.

Coronary Plaque Analysis (AI-QCT/AI-CPA) using coronary computed tomography angiography (CCTA) is an adjunctive tool used to assess intermediate risk plaque or CAD-RADS1 to CAD-RADS 3 category on CCTA done on individuals with chest pain

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and no known coronary artery disease. Multiple software have been developed as an adjunct to CCTA to aid in the visualization, characterization, and to improve accuracy of this assessment. This is intended to improve clinical diagnosis and management of CAD.

AI-CPA is currently being used to help stage medical treatment based on severity of plaque and to help predict those who are increased risk for ACS. It has also been shown to be consistent with consistent with invasive fractional flow reserve and intravascular ultrasound (IVUS). It is best used to target those most susceptible to acute coronary syndrome and to guide preventative measures of coronary artery disease.

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CT Heart for Evaluation of Cardiac Structure and Morphology (CD-4.6)

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CPT® 75572- Indications

- Cardiac vein identification for lead placement in left ventricular pacing
- To evaluate the anatomy of the pulmonary veins prior to a pulmonary vein isolation (ablation) procedure for atrial fibrillation
  - Post-procedure between 3-6 months after ablation

  Note:

  See also **Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)**

- If echocardiogram is inconclusive for:
  - cardiac or pericardial tumor or mass
  - cardiac thrombus
  - pericarditis/constrictive pericarditis
  - complications of cardiac surgery
- In place of MRI when there is clinical suspicion of arrhythmogenic right ventricular dysplasia or arrhythmogenic cardiomyopathy (ARVD/ARVC) if the clinical suspicion is supported by established criteria. For ARVD, see **Cardiac MRI – Indications (excluding Stress MRI) (CD-5.2).**
- Recurrent laryngeal nerve palsy due to cardiac chamber enlargement
- Prior to planned valvular interventions (TAVR, TMVR and TTVR) in individuals with radiation-induced valve disease
- CT Cardiac (CPT® 75572) can be performed instead of TEE for assessment of left atrial appendage (LAA) occlusion device or to assess for thrombus. Ssee: **Transesophageal Echocardiography (TEE) – Indications (CD-2.5).**
  - pre-procedural evaluation
  - repeat imaging 45 days post-procedure
    - Another follow-up study is medically necessary before the 1-year surveillance when the imaging study at 45 days shows a peri-device gap ≥5 mm or Device Related Thrombus usually at 3-6 months.
  - 1 year post-procedure
  - See also **Percutaneous Mitral Valve Repair (mitral valve clip) (CD-13.5).**

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Background and Supporting Information

Coronary imaging is not included in the code definition for CPT® 71275.

Repeat testing is medically necessary post pulmonary vein isolation procedure because of a 1%-2% incidence of asymptomatic pulmonary vein stenosis.

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CT Heart for Congenital Heart Disease (CD-4.7)

CD.CT.0004.7.A
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CPT® 75573

- Coronary artery anomaly evaluation
  - A cardiac catheterization was performed, and not all coronary arteries were identified.
- Thoracic arteriovenous anomaly evaluation
  - A MRI Cardiac or CT angiogram Chest was performed and suggested congenital heart disease.
- Complex adult congenital heart disease evaluation
  - No CT Cardiac or MRI Cardiac has been performed, and there is a contraindication to MRI Cardiac.
  - A CT Cardiac or MRI Cardiac was performed 1 year ago or more.
- See also section **Adult Congenital Heart Disease (CD-11)**.

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Transcatheter Aortic Valve Replacement (TAVR) (CD-4.8)

CD.CT.0004.8.A
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CT Imaging in Transcatheter Aortic Valve Replacement (TAVR)

Pre-TAVR imaging

Pre-aortic valve replacement

- Once the decision has been made for aortic valve replacement, the following may be used to determine if an individual is a candidate for TAVR:
  - CTA Chest (CPT® 71275), Abdomen and Pelvis (combination code CPT® 74174) are medically necessary; and
  - CT Cardiac (CPT® 75572) is medically necessary to measure the aortic annulus; or
  - Coronary CTA (CCTA CPT® 75574) is medically necessary to both measure the aortic annulus and assess the coronary arteries in lieu of heart catheterization.
- A repeat diagnostic left heart catheterization is **not** medically necessary when the individual is undergoing a transcatheter aortic valve replacement (TAVR).

Transfemoral access not feasible

Alternative imaging can be obtained to evaluate vascular access for TAVR in individuals for whom it is documented either via the office note or prior imaging that transfemoral access would not be feasible due to **any** of the following exclusion criteria:

- small vessels
- highly calcified vessels
- stenosed or occluded vessels
- prior aortoiliac vascular intervention

Imaging is medically necessary based on the documented intended access site (transaxillary or transcarotid) and should be of the involved body areas. The following studies are medically necessary based on the documented planned access site:

- CTA of the Head (CPT® 70496) and/or Neck (CPT® 70498) for transcarotid access
- CTA of the Chest (CPT® 71275) and/or Upper Extremity (CPT® 73206) for transaxillary access

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Post-TAVR imaging

CT Cardiac (CPT® 75572) is medically necessary:

- if any of the post-TAVR TTEs are indeterminate or raises a concern about any of the following:
  - valve thrombosis
  - infective endocarditis
  - structural degeneration
- when a Valve-in-Valve implantation or surgical re-do AVR is being contemplated
- Routine CT surveillance or follow up for incidental Hypoattenuated Leaflet Thickening (HALT) with or without restricted leaflet motion, also referred to as Hypoattenuation Affecting Motion (HAM), is NOT medically necessary.

Evidence Discussion

The ability of the cardiac CT technology to provide a tomographic view of the cardiovascular system has resulted in its ubiquitous adoption in the pre-procedure planning for almost all cardiac structural interventions. Specifically, cardiac CT circumvents the image window limitation of echocardiography, allows high definition visualization of the posterior structures, and facilitates pre-procedural planning for pulmonary vein isolation, coronary sinus pacer leads insertion, and left atrial appendage occlusion device implantation, among other trans-catheter structural interventions.

In non-interventional settings, cardiac CT provides an alternative to cardiac MRI when structural information cannot be adequately obtained by an echocardiography. Most notably, the evaluation of a cardiac mass, extent of pericardial disease, complex congenital heart disease and cardiomyopathy, can be performed by a cardiac CT when cardiac MRI is not available or contraindicated.

Trans-catheter Aortic Valve Replacement (TAVR)

The success of a Trans-catheter Aortic Valve Replacement (TAVR) procedure is contingent upon a meticulous pre-TAVR planning imaging study where cardiac CT allows accurate annulus sizing, coronary heights measurement, and calcification distribution evaluation, in addition to access site planning. Post-operatively, clinically suspected complications such as thrombus formation, infective endocarditis, or structural degeneration can be confirmed on a cardiac CT; a routine surveillance strategy, however, is not supported because of unclear or even potentially harmful outcome of treating incidental findings.

Transcatheter Mitral Valve Repair (mitral valve clip)

In non-interventional settings, cardiac CT provides an alternative to cardiac MRI when structural information cannot be adequately obtained by an echocardiography. Most notably, the evaluation of a cardiac mass, extent of pericardial disease, complex

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congenital heart disease and cardiomyopathy, can be performed by a cardiac CT when cardiac MRI is not available or contraindicated. Percutaneous treatment of mitral regurgitation can be accomplished using venous access to apply a clip device (e.g., MitraClip currently FDA approved) to provide edge-to-edge mitral leaflet coaptation, approximating opposing sections of the anterior and posterior mitral valve leaflets. FDA approved indications include treatment for individuals with symptomatic, moderate to severe, or severe primary mitral regurgitation whose surgical risks are prohibitive, as well as symptomatic moderate to severe or severe secondary mitral regurgitation who have failed optimal medical therapy. This therapy should include, if medically necessary, cardiac resynchronization therapy.

Transcatheter Mitral & Tricuspid Valve Replacement

Transcatheter Tricuspid Valve Replacement device (EVOQUE) is currently FDA approved. In individuals being considered for transcatheter tricuspid or mitral valve replacement, Cardiac CT CPT® 75572 provides detailed imaging of the valve annulus including measurement of valve annulus to guide selection of the appropriate-sized valve. CTV Chest (CPT® 71275) and CTV Abdomen and Pelvis (CPT® 74174) provide imaging of the iliac veins and inferior vena cava to exclude stenosis or significant tortuosity and aid in determining eligibility and procedure planning.

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3D Predictive Model Generation for Pre-planning of Cardiac Procedure (CD-4.9)

CD.CT.0004.9.A
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Coding

Description	HCPCS
3D predictive model generation for pre-planning of a cardiac procedure, using data from cardiac computed tomographic angiography with report	C9793

Criteria

3D predictive model generation for pre-planning of a cardiac procedure, using data from cardiac computed tomographic angiography is considered to be experimental, investigational, or unproven.

Evidence Discussion

Cardiac Computed Tomography Angiography is a 3D imaging acquisition, viewing, and reporting system. Standard Cardiac Computed Tomography Angiography includes the viewing of 3D images. Currently, there is not enough data to support the use of 3D predictive model generation for pre-planning of cardiac procedures. It has not been shown to improve outcomes when compared with standard Cardiac Computed Tomography Angiography.

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Cardiac MRI

Guideline

Cardiac MRI and Cardiac Indications for MRA Chest (CD-5.2)
Cardiac Stress MRI- (CD-5.3)
References (CD-5)

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Cardiac MRI and Cardiac Indications for MRA Chest (CD-5.2)

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Cardiac MRI – Coding

Cardiac Imaging Procedure Codes

Cardiac MRI	CPT®/HCPCS
Cardiac magnetic resonance imaging for morphology and function without contrast	75557
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences	75561
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure)	+75565
Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with strain imaging	C9762

- Only 1 procedure code from the set (CPT® 75557-CPT® 75563) should be reported per session.
- Only 1 flow velocity measurement (CPT® +75565) should be reported per session when medically necessary.

Cardiac MRI and Cardiac Indications for MRA Chest

Indications (excluding Stress MRI)

- Assess myocardial viability (to differentiate hibernating myocardium from scar) when necessary to determine if revascularization should be performed (CPT® 75561)
- Assessment of global ventricular function, myocardial composition, and mass if a specific clinical question is left unanswered by a recent echocardiogram and results will affect individual management (CPT® 75557 or CPT® 75561). Particularly useful in evaluating:

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  - cardiomyopathy (ischemic, diabetic, hypertrophic, or muscular dystrophy)
  - non-compaction
  - infiltrative heart disease such as amyloidosis and sarcoidosis (see also **Cardiac Sarcoidosis (CD-3.9)**)
  - post cardiac transplant
  - hypertrophic cardiomyopathy
  - suspected acute myocarditis, cardiac aneurysm, trauma, and contusions
  - monitoring cancer chemotherapy effect on the heart (especially if an accurate assessment of right ventricular function is documented as necessary)
- Pre- and post-operative congenital heart disease assessment. See **Adult Congenital Heart Disease (CD-11)** for defect specific indications (CPT® 75557 or 75561).
  - MRA Chest (CPT® 71555) may be added if the aorta or pulmonary artery need to be visualized beyond the root.
  - May add CPT® +75565 in conjunction with CPT® 75557 or 75561, only if there is a need to clarify findings on a recent echocardiogram and cardiac Doppler study when there is documentation of **either** of the following:
    - significant valvular disease that may require intervention
    - intracardiac flow disturbances (e.g., ASD, VSD)
- MRA Chest (CPT® 71555) is medically necessary for the following:
  - Thoracic aortic dissection. See **Aortic Dissection and Other Aortic Conditions (PVD-6.7)** in the Peripheral Vascular Disease Imaging Guidelines.
  - Coarctation of the aorta. See:
    - **Coarctation of the Aorta (CD-11.3.2)** for adults
    - **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)** for infants and children in the Pediatric Cardiac Imaging Guideline
  - Thoracic aortic aneurysm. See **Thoracic Aortic Aneurysm (TAA) (PVD-6.2)** in the Peripheral Vascular Disease Imaging Guidelines.
- Coarctation of the aorta
  - For adults, see **Coarctation of the Aorta (CD-11.3.2)**.
    - For infants and children, see **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)** in the Pediatric Cardiac Imaging Guideline.
- Assessment of ventricular arrhythmia for ablation planning (CPT® 75557 or 75561)
- Assessment of arrhythmogenic left ventricular cardiomyopathy (ALVC) (CPT® 75557 or 75561)
- Arrhythmogenic right ventricular dysplasia or arrhythmogenic right ventricular cardiomyopathy (ARVD/ARVC) suspicion (CPT® 75557 or 75561) must have one of the following:
  - non-sustained or sustained VT of LBBB morphology OR >500 PVCs over 24 hours on event recorder or Holter monitor

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  - ARVD/ARVC confirmed in a first-degree relative either by criteria, autopsy, pathogenic genetic mutation or sudden death <35 years of age with suspected ARVD/ARVC
  - inverted T waves in right precordial leads (V1, V2 and V3) or beyond in individuals >14 years of age in the absence of complete RBBB
  - right ventricular akinesis, dyskinesis, or aneurysm noted on echo or RV angiography
- Differentiate constrictive pericarditis from restrictive cardiomyopathy (CPT® 75561)
- Evaluate cardiac tumor or mass when echocardiogram is inconclusive (CPT® 75557 or 75561)
- Evaluate valvular heart disease when echocardiogram is inconclusive:
  - CPT® 75557 **or** CPT® 75561
  - May add CPT® 75565 when there is documentation of either of the following:
    - significant valvular disease that may require intervention
    - intracardiac flow disturbances (e.g., ASD, VSD)
- MRI Cardiac (CPT® 75557 or 75561) **or** chest MRV (CPT® 71555), but not both, for pulmonary vein anatomy for planned ablation procedures in individuals with atrial fibrillation. See **Pulmonary Vein Imaging – Indications (CD-8.2)** for guidelines on follow-up imaging after ablation procedure.
- Suspected cardiac thrombus when echocardiogram is inconclusive (CPT® 75557).
- Right ventricular function evaluation (CPT® 75557 in conjunction with CPT® +75565) if there has been a recent ECHO and there is documented need to perform Cardiac MRI in order to resolve an unanswered question about flow dynamics
- Cardiac MRI (CPT® 75557 or 75561) and CMR velocity flow mapping (CPT® 75565) for preoperative evaluation prior to planned LVAD implant
- Shunting through a VSD (CPT® 75557 in conjunction with CPT® +75565) if a recent ECHO has been done, including a bubble study, and there is documented need to perform Cardiac MRI in order to resolve an unanswered question about flow dynamics
- Conditions that would **not** require an ECHO prior to an MRI:
  - detect anomalous coronary arteries (CPT® 75561)
  - assess coronary arteries in Kawasaki disease
  - Fabry disease (CPT 75561)
  - initial evaluation for cardiac sarcoidosis or iron overload cardiomyopathy (hemosiderosis, hemochromatosis)

Non-indications

C9762-Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with strain imaging. The use of CMR strain imaging for the

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quantification of segmental dysfunction is considered investigational and experimental at this time.

Quantitative analysis of myocardial tissue composition by MRI (CPT® codes 0648T, 0649T, 0697T and 0698T) are considered experimental, investigational, or unproven at this time. There is insufficient clinical data to support their use.

Cardiac MRI – Aortic Root and Proximal Ascending Aorta

- See **Thoracic Aortic Aneurysm (TAA) (PVD-6.2)** in the Peripheral Vascular Disease imaging guidelines.

Cardiac MRI – Duchenne Muscular Dystrophy (DMD)

Cardiac MRI (CPT® 75557 or 75561-does not include CPT® 75565 or 71555 unless otherwise indicated)

- Asymptomatic individual with documented DMD can have annual surveillance cardiac MRI starting at 6 years old (yearly echo is recommended prior to age 6)
- Asymptomatic, documented carrier of DMD can have cardiac MRI every 3 years starting at 18 years

Cardiac MRI – Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade

- Contrast-enhanced cardiac MRI (CPT® 75561) is useful for evaluating pericarditis, neoplastic, and other effusion, tamponade, or myocardial infiltration if a specific clinical question is left unanswered by echocardiogram or another recent imaging study.

Cardiac MRI – Myocarditis

Clinical evaluation of suspected myocarditis

Initial testing for suspected myocarditis should consist of an electrocardiogram, measurement of cardiac troponin, and an echocardiogram.

Cardiac MRI (CPT® 75561) is medically necessary for suspected myocarditis in the presence of **all** of the following:

- New onset or persisting symptoms suggestive of myocarditis documented by **any** of the following:

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  - dyspnea
  - chest pain
  - palpitations
  - syncope
  - effort intolerance
- Evidence for recent or ongoing myocardial injury documented by **any** of the following results on initial screening:
  - ventricular dysfunction noted on any cardiac imaging study, or
  - new or persisting ECG abnormalities suspicious for myocarditis
    - ST changes, T wave changes, Q waves, or
    - conduction abnormalities, such as LBBB or AV block, or
    - VT or VF
  - elevated troponin
- Strong suspicion for viral etiology of myocardial injury with documentation of **both**:
  - recent systemic viral disease, recent mRNA COVID-19 vaccination, or prior myocarditis
  - no evidence of coronary ischemia as documented by **any** of the following:
    - lack of risk factors for CAD
    - age < 35 years
    - negative cardiac imaging study, such as MPI, CCTA, catheterization

Return to Play Screening for athletes at risk for myocarditis

Cardiac MRI (CPT® 75561) is medically necessary for **Return to Play Screening** for athletes when there is documentation of **both** of the following:

- Individual has a history of a clinical condition associated with myocarditis (i.e., COVID-19 infection or recent mRNA COVID-19 vaccination).
- Initial screening has been performed with documentation of **either** of the following:
  - Initial screening (ECG, troponin, and TTE) showed evidence for recent or ongoing myocardial injury (as defined above in Clinical Evaluation of Suspected Myocarditis) with ongoing symptoms concerning for myocarditis (dyspnea, chest pain, palpitations, syncope, or effort intolerance).
  - Normal results of initial screening with persistent or new onset symptoms concerning for myocarditis.

Background and Supporting Information

Evidence Discussion

- Guidelines and appropriate use criteria support the use of transthoracic echocardiogram (TTE) as the initial study for a broad range of cardiac conditions.

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TTE can be used to evaluate cardiac morphology and function and provides information necessary to diagnose and guide treatment in conditions including heart failure and cardiomyopathy, ischemic heart disease, and valvular dysfunction. TTE is also used to initially assess intracardiac and extracardiac structures, including suspected cardiac masses, sources of emboli, and pericardial conditions. TEE visualizes portions of the great vessels and provides initial imaging of these structures.

- TTE has significant advantages over other imaging modalities; it is easily accessible and even portable, provides no exposure to radiation, is inexpensive, and has minimal, if any, risks.
- Like TTE, MRI assesses cardiac morphology and function; additionally, MRI provides information regarding myocardial perfusion, metabolism, and tissue composition.
- When questions remain unanswered after TTE, MRI can provide additional information due to its ability to assess myocardial composition and mass; these characteristics make cardiac MRI especially useful in the settings of cardiomyopathy, non-compaction, infiltrative heart disease, post cardiac transplant, cardiac masses, pericardial disease, and myocarditis.
- Beyond imaging with TTE, MRI can provide improved visualization of valvular pathology, because images are not limited by body habitus, is of high spatial resolution, can more accurately quantify the magnitude of valve insufficiency and stenosis, and can assess the effects of valve dysfunction on ventricular mass or volume. Qualities of MRI that augment evaluation of valvular disease may also improve visualization of intracardiac shunts over TTE alone.
- Advantages of MRI assessment of the great vessels include avoidance of ionizing radiation, provision of excellent spatial resolution of structures, and is not limited by body habitus.
- Conditions that are better evaluated initially by MRI, include coronary artery anomalies, and diseases of abnormal myocardial composition: Arrhythmogenic right ventricular dysplasia, Fabry disease-(Late enhancement MRI may predict the effect of enzyme replacement therapy on myocardial changes that occur with this disease), cardiac sarcoidosis, and Duchenne Muscular Dystrophy.
- When TTE and other less advanced imaging provide the information necessary to diagnose and treat cardiovascular conditions, MRI is not medically necessary.
- Disadvantages of MRI include being less accessible and possibly being incompatible for use in those with implanted devices. The gadolinium contrast agents used in MRI require precautions in those with advanced renal disease and severe liver disease.
- The decision to use MR imaging is made in the context of other testing; duplication of information should be avoided, and more readily available modalities with lower risk, initially considered. An assessment should be made as to whether additional testing will provide complementary diagnostic, therapeutic, or prognostic information that will optimize care.

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- As noted in the "2022 ACC expert consensus decision pathway on cardiovascular sequelae of COVID-19 in adults" and the 2017 "Sports cardiology: core curriculum for providing cardiovascular care to competitive athletes and highly active people," an athlete is defined as an individual who places a high premium on exercise training, competition, and sports achievement.

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Cardiac Stress MRI- (CD-5.3)

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Cardiac Stress MRI – Coding

Cardiac Imaging Procedure Codes

Cardiac MRI	CPT®/HCPCS
Cardiac magnetic resonance imaging for morphology and function without contrast; with stress imaging	75559
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences; with stress imaging	75563
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure)	+75565
Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with stress imaging	C9763
Non-invasive determination of absolute quantitation of myocardial blood flow (AQMBF), derived from augmentative algorithmic analysis of the dataset acquired via contrast cardiac magnetic resonance (CMR), pharmacologic stress, with interpretation and report by a physician or other qualified health care professional (List separately in addition to code for primary procedure)	+0899T
Non-invasive estimate of absolute quantitation of myocardial blood flow (AQMBF), derived from assistive algorithmic analysis of the dataset acquired via contrast cardiac magnetic resonance (CMR), pharmacologic stress, with interpretation and report by a physician or other qualified health care professional (List separately in addition to code for primary procedure)	+0900T

- Only 1 procedure code from the set (CPT® 75557- CPT® 75563) should be reported per session.
- Only 1 flow velocity measurement (CPT® +75565) should be reported per session when medically necessary.

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- C9763-Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with stress imaging. The use of stress CMR for the quantification of segmental dysfunction is considered investigational and experimental at this time.

Indications for Cardiac Stress MRI

Indications

- For Indications for Stress MRI, see **Stress Testing with Imaging – Indications (CD-1.4)**.
- If a nuclear perfusion (MPI) stress test was performed and was equivocal, a stress MRI is medically necessary.
- To further evaluate the etiology of myocardial infarction with no obstructive coronary arteries (MINOCA) in the setting of a normal coronary angiogram

Absolute quantitation of myocardial blood flow (AQMBF)

Non-invasive determination of absolute quantitation of myocardial blood flow (AQMBF) using AI-analysis of data obtained from cardiac magnetic resonance imaging (CMR) with pharmacological stress (CPT® 0899T or 0900T) is medically necessary to further evaluate cardiac function when performed with a cardiac MRI stress (CPT® 75559 or 75563).

Non-indications for Cardiac Stress MRI

Quantification of segmental dysfunction

C9763-Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with stress imaging. The use of stress CMR for the quantification of segmental dysfunction is considered investigational and experimental at this time.

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Cardiac PET

Guideline

Cardiac PET – Coding (CD-6.1)
Cardiac PET – Perfusion – Indications (CD-6.2)
Cardiac PET – Absolute Quantitation of Myocardial Blood Flow (AQMBF) (CD-6.3)
References
Cardiac PET – Metabolic – Indications (CD-6.4)
FDG PET/CT for Infections (CD-6.5)
References (CD-6)

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Cardiac PET – Coding (CD-6.1)

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Cardiac Imaging Procedure Codes

Cardiac PET	CPT®
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), single study	78459
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); single study at rest or stress (exercise or pharmacologic)	78491
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); multiple studies at rest and stress (exercise or pharmacologic)	78492
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), single study; with concurrently acquired computed tomography transmission scan	78429
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); single study, at rest or stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78430
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); multiple studies at rest and stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78431

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Cardiac PET	CPT®
Myocardial imaging, positron emission tomography (PET), combined perfusion with metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), dual radiotracer (e.g., myocardial viability);	78432
Myocardial imaging, positron emission tomography (PET), combined perfusion with metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), dual radiotracer (e.g., myocardial viability); with concurrently acquired computed tomography transmission scan	78433
Absolute quantitation of myocardial blood flow (AQMBF), positron emission tomography (PET), rest and pharmacologic stress (List separately in addition to code for primary procedure)	+78434
Positron emission tomography (PET) with concurrently acquired computed tomography (CT) for attenuation correction and anatomical localization imaging; skull base to mid-thigh	78815

- 3D rendering should not be billed in conjunction with PET.
- Separate codes for such related services as treadmill testing (CPT® 93015-CPT® 93018) and radiopharmaceuticals should be assigned in addition to perfusion PET. These services are paid according to each individual payer.

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Cardiac PET – Perfusion – Indications (CD-6.2)

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CPT® 78430, CPT® 78431, CPT® 78491 and CPT® 78492

- See **Stress Testing with Imaging (CD-1.4)** for stress imaging indications.
- Cardiac PET is considered medically necessary when requested after an equivocal nuclear perfusion (SPECT MPI) stress test.
- Routine use in post heart transplant assessment of transplant CAD

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Cardiac PET – Absolute Quantitation of Myocardial Blood Flow (AQMBF) (CD-6.3)

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Absolute Quantitation of Myocardial Blood Flow (CPT® 78434)

Quantitation of myocardial blood flow at rest and with stress in mL/g/min and the calculation of myocardial perfusion reserve (the ratio of stress to rest flow) can be used for diagnosis and prognosis of coronary artery disease and cardiac endothelial dysfunction that can be seen in diabetes, left ventricular hypertrophy, and heart transplantation vasculopathy, among other conditions.

- AQMBF with PET (CPT® 78434) is an add-on procedure that is medically necessary when one of the following apply:
  - Primary study Myocardial PET rest/stress perfusion (CPT® 78492 or 78431 only) has been approved.
  - Primary study Myocardial PET rest/stress perfusion (CPT® 78492 or 78431 only) has been ordered and is being approved at the same time (see: **Cardiac PET- Perfusion- Indications (CD-6.2)** or **Stress Testing with Imaging- Indications (CD-1.4)**).

Evidence Discussion

Despite its utility, AQMBF is a technically challenging measurement. Variables include:
- Different tracers (N-13 ammonia vs Rb-82 Cl) give different values.
- Different mathematical models are used (static vs dynamic).
- Different stressors are used that give different hyperemic flow results (adenosine vs dipyridamole vs regadenoson).
- Data can be collected in 2D vs 3D modes. Saturation of crystals is more problematic in 3D.
- Cardiac, respiratory, and individual motion can degrade measurement accuracy.
- Different vendor software is used by different reading labs.
- Resting blood flow can be elevated due to pain, anxiety, lack of vagal tone, hypertension, etc. and can be normalized by using the rate pressure product (RPP) for calculation of myocardial perfusion reserve (MBF) the ratio of myocardial hyperemic flow/rest flow.

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Evidence Discussion

Myocardial perfusion imaging (MPI) is used for the diagnosis of coronary artery disease (CAD). Results of MPI have a high level of sensitivity and specificity. Positron emission tomography (PET) is one imaging modality that allows the performance of MPI and also has the ability to measure absolute myocardial blood flow (AQMBF) (CPT 78434) in ml/g/min. This has been shown to add to the diagnosis of CAD and aid in treatment decisions.

As supported by the ACC/AHA Appropriate Use Criteria, the use of MPI with PET is medically necessary for individuals with signs or symptoms that would be consistent with CAD such as typical angina or other typical or atypical symptoms in individuals with known CAD or prior interventions such as coronary artery bypass surgery or coronary stenting. This balances the appropriate testing of individuals with PET versus medically unnecessary and potentially harmful testing and downstream procedures.

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Cardiac PET – Metabolic – Indications (CD-6.4)

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- Cardiac PET Metabolic and cardiac SPECT or PET perfusion (CPT® 78429 or CPT® 78459 and CPT® 78451, or CPT® 78432, or CPT® 78433)
  - to determine myocardial viability when a previous study has shown significant left ventricular dysfunction when under consideration for revascularization
- To diagnose strongly suspected cardiac sarcoid or monitor response to therapy for established cardiac sarcoid. See **Cardiac Sarcoidosis (CD-3.9)**.

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FDG PET/CT for Infections (CD-6.5)

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- FDG PET/CT (CPT® 78815 or CPT® 78429) is medically necessary in the assessment of suspected prosthetic heart valve endocarditis when there is documentation of **both** of the following:
  - TTE and/or TEE are equivocal or non-diagnostic
  - Suspicion for prosthetic heart valve endocarditis remains high (all):
    - C-reactive protein ≥40 mg/L
    - no evidence of prolonged antibiotic therapy
    - The implantation was ≥3 months ago and there is no evidence of surgical adhesives used during the valve implantation.
- FDG PET/CT for LVAD driveline infection (CPT® 78815 or 78429)
  - Early infection detection for LVAD drivelines is desirable, since once the infection extends to the cannula and pump pocket, eradication becomes difficult. CT findings are nonspecific, and metal device artifacts of the driveline itself affects sensitivity.
  - FDG PET/CT is medically necessary for suspected LVAD infection if other studies and examination remain inconclusive.

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Diagnostic Heart Catheterization

Guideline

LHC – Unstable/Active Coronary Artery Syndromes (CD-7.3.1)
Diagnostic Heart Catheterization – Coding (CD-7.2)
Diagnostic Left Heart Catheterization (LHC) (CD-7.3)
Right Heart Catheterization and Right and Left Heart Catheterization without Coronary Angiography (CD-7.4)
Combined Right and Left Heart Catheterization Indications (CD-7.5)
Planned (Staged) Coronary Interventions or Post-catheterization Physiologic Evaluation (CD-7.6)
Evaluation of Conditions Other than Coronary Artery Disease (CD-7.7)
References (CD-7)

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LHC – Unstable/Active Coronary Artery Syndromes (CD-7.3.1)

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Diagnostic Left Heart Catheterization (LHC) is medically necessary for individuals in acute settings or with **active** unstable angina and should be handled as medical emergencies.

- LHC may be medically necessary for new onset, accelerating, or worsening ischemic symptoms suggestive of acute coronary syndrome (ACS) occurring at rest, or with minimal exertion resolving with rest, including:
  - cardiac chest pain (typical angina) with or without new onset, evolving ischemic EKG changes
  - symptoms consistent with the known angina pattern in an individual with a history of CABG or PCI
- Left and right heart catheterization (CPT® 93460 or 93461) is considered medically necessary in place of a left heart catheterization when criteria has been met for a left heart catheterization and there is documentation of **any** of the following conditions:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy

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Diagnostic Heart Catheterization – Coding (CD-7.2)

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Diagnostic Heart Catheterization – Code Sets

Cardiac Catheterization Procedure Codes

Cardiac catheterization Procedure	CPT®
Congenital Heart Disease Code "Set"	93593-93597
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; normal native connections	93593
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; abnormal native connections	93594
Left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone, normal or abnormal native connections	93595
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); normal native connections	93596
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); abnormal native connections	93597
Anomalous coronary arteries, patent foramen ovale, mitral valve prolapse, and bicuspid aortic valve	93451-93464, 93566-93568

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Cardiac catheterization Procedure	CPT®
RHC without LHC or coronaries	93451
LHC without RHC or coronaries	93452
RHC and retrograde LHC without coronaries	93453
Native coronary artery catheterization;	93454
with bypass grafts	93455
with RHC	93456
with RHC and bypass grafts	93457
with LHC	93458
with LHC and bypass grafts	93459
with RHC and LHC	93460
with RHC and LHC and bypass grafts	93461
LHC by trans-septal or apical puncture	+93462
Angiography of non-coronary arteries and veins performed as a distinct service	Select appropriate codes from the Radiology and Vascular Injection Procedures sections.

- CPT® 93593 to 93597 are medically necessary for invasive evaluation of congenital heart disease. See specific conditions in **Congenital heart disease – General Information (CD-11.1).**

Diagnostic Heart Catheterization – Coding Notes (CD-7.2)

- Cardiac catheterization (CPT® 93451-CPT® 93461) includes all "road mapping" angiography necessary to place the catheters, including any injections and imaging supervision, interpretation, and report.

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- Cardiac catheterization (CPT® 93452-CPT® 93461) (for all conditions other than congenital heart disease) includes contrast injections, imaging supervision, interpretation, and report for imaging typically performed.
- Catheter placements in native coronaries or bypass grafts (CPT® 93454-CPT® 93461) include intraprocedural injections for bypass graft angiography, imaging supervision, and interpretation.
- Injection codes CPT® 93563-CPT® 93565 should not be used in conjunction with CPT® 93452-CPT® 93461.
- Codes CPT® 93451-CPT® 93461 do not include contrast injections and imaging supervision, interpretation, and report for imaging that is separately identified by the following specific procedure codes: CPT® 93566, CPT® 93567 and CPT® 93568.
- Separate diagnostic cardiac catheterization codes should only be assigned in conjunction with interventional procedures in the following circumstances:
  - No prior or recent diagnostic catheterization is available to guide therapy.
  - Individual's condition has significantly changed since the last diagnostic catheterization.
  - The treatment plan may be affected.
  - Other vessels may be identified for treatment.
  - Further establishment of a diagnosis from a non-invasive study is necessary.

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Diagnostic Left Heart Catheterization (LHC) (CD-7.3)

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Stable Established CAD Post Revascularization with CABG or PCI (CD-7.3.2)

CPT® 93454, 93455, 93458, 93459

Note: These guidelines apply to individuals with stable conditions and who are not in the acute setting (acute coronary syndrome or unstable angina).

- Diagnostic Left Heart Catheterization (LHC) is medically necessary in individuals with established Coronary Artery Disease (CAD) post revascularization by percutaneous coronary intervention (PCI) or coronary artery bypass graft surgery (CABG) when there is documentation of **one** of the following:
  - new, recurrent, or worsening ischemic symptoms similar to prior ischemic episode
  - new, recurrent, or worsening cardiac chest pain as defined in **General Guidelines (CD-1.0)**
  - new, recurrent, or worsening symptoms of chest pain, exertional dyspnea, or exertional fatigue AND intermediate or high-risk findings on non-invasive stress testing as documented by one of the following:
    - cardiac chest pain induced by exercise treadmill testing or dobutamine stress testing
    - exercise treadmill testing inducing any of the following:
      - at least 1 mm downsloping ST-depression
      - 2 mm horizontal ST-depression
      - at least 1 mm ST-elevation in two leads
      - ventricular tachycardia of at least 3 consecutive beats
    - myocardial perfusion imaging (SPECT or PET) with ≥5% reversible ischemic burden
    - stress echo with at least 2 segments of inducible ischemia
    - severe stress induced left ventricular dysfunction (drop in left ventricular ejection fraction with stress ≥10%)
  - new left ventricular systolic dysfunction (left ventricular ejection fraction <50%)
  - worsening left ventricular systolic dysfunction (decline in left ventricular ejection fraction ≥10%)

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  - new or worsened congestive heart failure
  - ventricular fibrillation
  - sustained ventricular tachycardia
- Left and right heart catheterization (CPT® 93460 or 93461) is considered medically necessary in place of a left heart catheterization when criteria has been met for a left heart catheterization and there is documentation of **any** of the following conditions:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy

Stable Symptomatic Suspected or Established Coronary Artery Disease (CD-7.3.3)

CPT® 93454, 93455, 93458, 93459

For the purpose of this guideline, cardiac chest pain is defined in **General Guidelines (CD-1.0)**.

- Diagnostic left heart catheterization to screen for coronary artery disease (CAD) in asymptomatic individuals who are not anticipating other cardiac procedures is **not** considered medically necessary.
- LHC with coronary arteriography (CPT® 93454, 93455, 93458, or 93459) is medically necessary when there is documentation of one of the following:
  - New onset, persistent, or worsening of cardiac chest pain symptoms and either:
    - Symptomatic failure of a 12 week trial of OMT including as tolerated all of the following:
      - anti-platelet therapy
      - statin and/or other lipid-lowering therapy
      - anti-anginal therapy implemented to pursue a goal heart rate of 60 beats per minute or less
      - anti-hypertensive therapy as may be indicated to pursue a goal systolic blood pressure (SBP) of <140 mmHg and a goal diastolic blood pressure (DBP) of <90 mmHg
    - Worsening of cardiac chest pain symptoms during 12 week trial of OMT
  - New onset, persistent, or worsening of cardiac chest pain symptoms and documentation of **either** of the following:
    - Established CAD per **General Guidelines (CD-1.0)** or age ≥50 years and/or ≥2 CAD risk factors: diabetes mellitus, smoking, family history of premature CAD, hypertension, dyslipidemia, or
    - Cardiac chest pain at a low level of exercise or at rest despite optimal medical therapy

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- LHC may be medically necessary irrespective of OMT for symptomatic (chest pain or dyspnea on exertion) individuals who also have high-risk findings on Coronary CT Angiography (See **CCTA-Indications for CCTA (CD-4.3))**to include any of the following:
  - left main coronary artery stenosis ≥40%
  - proximal or mid left anterior descending coronary artery stenosis ≥70%
  - proximal or mid double-vessel major epicardial (not branch) coronary artery stenosis ≥60%
  - proximal or mid triple-vessel major epicardial (not branch) coronary artery stenosis ≥50%
  - CT-FFR measured to be ≤0.8 in the proximal or mid segment of any coronary artery or major epicardial branch irrespective of degree of stenosis
- LHC may be medically necessary irrespective of OMT for symptomatic (chest pain or dyspnea on exertion) individuals who have high-risk findings on non-invasive stress testing including any of the following:
  - cardiac chest pain induced by exercise treadmill testing or dobutamine stress testing
  - myocardial perfusion imaging with ≥10% reversible ischemic burden
  - stress echo with at least 3 segments of inducible ischemia
  - exercise treadmill testing inducing at least 2.5 mm downsloping ST-depression or 3 mm horizontal ST-depression in two leads
  - ventricular tachycardia of at least 3 consecutive beats induced by an exercise treadmill test
- LHC is medically necessary if coronary artery disease is the suspected etiology for **any** of the following findings:
  - new or worsened congestive heart failure
  - new left ventricular systolic dysfunction (left ventricular ejection fraction <50%)
  - worsening left ventricular systolic dysfunction (decline in left ventricular ejection fraction ≥10%)
  - ventricular fibrillation or sustained ventricular tachycardia

- Left and right heart catheterization (CPT® 93460 or 93461) is considered medically necessary in place of a left heart catheterization when criteria has been met for a left heart catheterization and there is documentation of **any** of the following conditions:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy

Background and Supporting Information

In addition to OMT, physician-guided behavioral modification therapy (BMT) is medically necessary including all of the following:

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- Mediterranean diet
- Moderate intensity physical activity for at least thirty minutes per day at least five times per week as possible
- Attempts at smoking cessation to include at least one of the following:
  - Cognitive behavioral therapy
  - Nicotine withdrawal replacement therapy

Exclusion of Significant Coronary Artery Disease Involvement in Other Cardiac Pathology (CD-7.3.4)

CPT® 93454, 93455, 93458, 93459

- LHC is medically necessary when the etiology is unclear for **any** of the following conditions when coronary artery disease is suspected:
  - new or worsened congestive heart failure
  - new left ventricular systolic dysfunction (left ventricular ejection fraction <50%)
  - worsening left ventricular systolic dysfunction (decline in left ventricular ejection fraction ≥10%)
  - ventricular fibrillation or sustained ventricular tachycardia
  - unheralded syncope (not near syncope)
  - suspected myocarditis
  - CCTA findings with high risk findings such as:
    - left main disease with ≥50% stenosis
    - three-vessel disease with ≥70 stenosis
    - two-vessel disease with ≥70% stenosis, including the proximal LAD
    - one-vessel disease of the proximal LAD with ≥70% stenosis and FFR-CT ≤0.8
- Left and right heart catheterization (CPT® 93460 or 93461) is considered medically necessary in place of a left heart catheterization when criteria has been met for a left heart catheterization and there is documentation of **any** of the following conditions:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy

Evaluation of Structural Heart Disease (CD-7.3.5)

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Left heart catheterization with coronary arteriography is medically necessary for any of the following:

- Evaluation prior to planned invasive procedure or surgery

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  - Ruling out coronary artery disease prior to planned non-coronary cardiac or great vessel surgery including any of the following:
    - cardiac valve surgery
    - surgical myectomy for hypertrophic cardiomyopathy
    - aortic dissection
    - aortic aneurysm
    - congenital disease repair such as atrial septal defect
  - Pre-organ transplant (non-cardiac)-in place of stress imaging for initial pre-transplant evaluation (per the transplant center's protocol) **or** if stress imaging is positive for ischemia. Repeat periodic screening while on a transplant waiting list (in the absence of other clinical indications) is not medically necessary. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.
  - Prior to catheter ablation of ventricular arrhythmia at one of the following sites:
    - sinus of Valsalva
    - coronary venous system
    - epicardium
- Valvular heart disease when either:
  - There is a discrepancy between the clinical findings (history, physical exam, and non-invasive test results).
  - Valvular surgery is being considered.
- Suspected pericardial disease
- Previous cardiac transplant:
  - per transplant center protocol
  - to assess for accelerated coronary artery disease associated with cardiac transplantation

Left and right heart catheterization (CPT® 93460 or 93461) is considered medically necessary in place of a left heart catheterization when criteria has been met for a left heart catheterization and there is documentation of **any** of the following conditions:

- The major component of the individual's symptoms is dyspnea
- Newly diagnosed or worsening cardiomyopathy

Evidence Discussion

Medical specialty organization guidelines recommend guideline directed medical therapy as the cornerstone of management of chronic coronary artery disease. These recommendations are based on a large body of evidence; including multiple large randomized controlled clinical trials (COURAGE, ISCHEMIA, and BARI-2D), and meta-analyses that demonstrate a significant improvement in angina symptoms, and cardiac events with guideline directed medical therapy. These trials further demonstrate that

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there is no improvement in survival or cardiac events with routine revascularization compared to guideline directed medical therapy.

Management options for coronary artery disease should account for the risks of invasive coronary angiography weighed against the benefits. Invasive coronary angiography is associated with cardiac (e.g., coronary artery injury and myocardial infarction) and extra-cardiac (e.g., vascular access site bleeding, ischemic stroke, and contrast-mediated acute kidney injury) risks. Invasive coronary angiography should be utilized as the treatment option when there is a favorable benefit-to-risk assessment.

Based on medical specialty organization guidelines and Appropriate Use Criteria, indications for invasive coronary angiography include the following:

- angina despite guideline directed medical therapy
- angina with high-risk findings on stress testing
- new left ventricular systolic dysfunction or heart failure of suspected ischemic etiology
- known coronary artery disease with prior revascularization with recurrent angina
- prior to non-coronary cardiac surgery, major vascular surgery, or organ transplant

  In addition to OMT, physician-guided behavioral modification therapy (BMT) is recommended including all of the following:
  - Mediterranean diet
  - Moderate intensity physical activity for at least thirty minutes per day at least five times per week as possible
  - Attempts at smoking cessation to include at least either cognitive behavioral therapy or nicotine withdrawal replacement therapy

Angiography-Derived Fractional Flow Reserve (CD-7.3.6)

Coding

Description	HCPCS
Catheter placement in coronary artery(s) for coronary angiography, including intraprocedural injection(s) for coronary angiography, imaging supervision and interpretation with left heart catheterization including intraprocedural injection(s) for left ventriculography, when performed and intraprocedural coronary fractional flow reserve (FFR) with 3D functional mapping of color-coded FFR values for the coronary tree, derived from coronary angiogram data, for real-time review and interpretation of possible atherosclerotic stenosis(es) intervention	C7557

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Criteria

Angiography-Derived Fractional Flow Reserve based on three-dimensional reconstruction of angiographic images is considered to be experimental, investigational, or unproven.

Background and Supporting Information

Fractional flow reserve (FFR) is an index of the physiological significance of a coronary artery stenosis. It is derived by using a coronary artery pressure guidewire to measure the distal coronary artery pressure. Angiography-Derived Fractional Flow Reserve is an alternative approach to derive FFR without the use of a coronary artery pressure guidewire, based on 3D reconstruction of angiographic images. Mathematical modeling based on hemodynamic laws is then applied to the 3D reconstruction to derive the pressure dynamics along the coronary artery, with results displayed in a color-coded map. Further research is needed to evaluate the accuracy and applicability of Angiography-Derived Fractional Flow Reserve.

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Right Heart Catheterization and Right and Left Heart Catheterization without Coronary Angiography (CD-7.4)

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- Diagnostic right heart catheterization (CPT® 93451) OR Diagnostic right and left heart catheterization without coronary angiography (CPT® 93453) is medically necessary when results will impact the diagnosis and management of **any** of the following:
  - atrial septal defect (ASD) including shunt detection and quantification
  - ventricular septal defect (VSD) including shunt detection and quantification
  - patent foramen ovale (PFO)
  - anomalous pulmonary venous return
  - congenital defects including persistent left vena cava
  - pulmonary hypertension
  - pericardial diseases
  - valvular disease
  - right heart failure
  - left heart failure
  - newly diagnosed or worsening cardiomyopathy
  - preoperative evaluation for valve surgery
  - during a left heart catheterization where the etiology of the symptoms remains unclear
  - pre-lung transplant to assess pulmonary pressures
  - pre-liver transplant to assess pulmonary pressures
  - uncertain intravascular volume status with an unclear etiology
  - prior to LVAD implant and post LVAD implant as needed for hemodynamic assessment to guide changes to therapy
  - assessment post-cardiac transplant
    - for routine endomyocardial biopsy
    - assess for rejection
    - assess pulmonary artery pressure
    - can be done per the institution protocol or anytime organ rejection is suspected and biopsy is needed for assessment
  - evaluation of right ventricular morphology.
  - suspected arrhythmogenic right ventricular dysplasia.

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Evidence Discussion

General information RHC (CPT® 93451)

- It is performed most commonly from the femoral vein, less often through the subclavian, brachial, or internal jugular vein and inter-atrial septal puncture approach.
- It includes a full oximetry for detection and quantification of shunts.
- Cardiac outputs are calculated by several techniques including the Fick thermodilution.
- Diagnostic right heart catheterization or right and left heart catheterization is performed for functional hemodynamic evaluation and may be combined with cardiac angiography for structural cardiac imaging and coronary angiography for anatomical evaluation of the coronary arteries. Diagnostic right heart catheterization or right and left heart catheterization is medically necessary in selected situations to obtain specific clinical information required for management decisions if it was not provided by non-invasive evaluation. This may be medically necessary in selected situations for management of valvular heart disease, myocardial disease, pericardial disease, congenital heart disease, pulmonary hypertension, heart failure, and post-cardiac transplantation. These indications are addressed in established evidence-based medical specialty organization guidelines pertaining to these conditions. If the clinical information required for management decisions was already provided by non-invasive evaluation, diagnostic heart catheterization does not add to management and, moreover, carries cardiac and extra-cardiac risks including coronary artery injury, myocardial infarction, vascular access site bleeding, ischemic stroke, and contrast-mediated acute kidney injury.

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Combined Right and Left Heart Catheterization Indications (CD-7.5)

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Combined Right and Left Heart Catheterization (CPT® 93460 or 93461) is medically necessary for pre-operative surgical planning for any of the following procedures:

- heart valve surgery
- congenital heart defect repair
- lung transplant
- liver transplant

Background and Supporting Information

- Left and right heart catheterization (CPT® 93460 or 93461) is considered medically necessary in place of a left heart catheterization when criteria has been met for a left heart catheterization and there is documentation of **any** of the following conditions:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy

Note:

See the individual diagnostic heart catheterization sections for more information.

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Planned (Staged) Coronary Interventions or Post-catheterization Physiologic Evaluation (CD-7.6)

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- Planned (Staged) Coronary Interventions applies to individuals with clinically stable symptoms if there is documentation of a recent diagnostic catheterization finding of a significant lesion that was not intervened as part of the initial procedure and the documentation explicitly states that a subsequent procedure will be performed for planned/staged PCI of that lesion.
- The CPT® codes for percutaneous coronary interventions (PCI) include the following imaging services necessary for the procedure(s):
  - contrast injection, angiography, 'road-mapping', and fluoroscopic guidance
  - vessel measurement
  - angiography following coronary angioplasty, stent placement, and atherectomy
- Separate codes for these services should not be assigned in addition to the PCI code(s) because the services are already included.
- A repeat diagnostic left heart catheterization is not medically necessary when the individual is undergoing a planned staged percutaneous coronary intervention.

Evidence Discussion

Diagnostic cardiac catheterization with coronary angiography is performed prior to an interventional percutaneous coronary intervention (PCI) procedure. PCI may be performed on the same day as the diagnostic coronary angiogram (ad-hoc PCI) or on a later day (planned staged PCI). Diagnostic coronary angiogram is required only once prior to the interventional procedure. A repeat diagnostic coronary angiogram is not medically necessary prior to a planned staged percutaneous coronary intervention. A repeat diagnostic coronary angiogram does not add to the findings of the initial diagnostic coronary angiogram and, moreover, carries cardiac and extra-cardiac risks including coronary artery injury and myocardial infarction, vascular access site bleeding, ischemic stroke, and contrast-mediated acute kidney injury.

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Evaluation of Conditions Other than Coronary Artery Disease (CD-7.7)

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- Right and left heart catheterization (CPT® 93453) is medically necessary for any of the following:
  - preoperative assessment prior to planned valvular surgery
  - evaluation of pulmonary hypertension out of proportion to or unexplained by the severity of valvular disease documented by other non-invasive imaging modalities (i.e., echo, CMR)
  - left ventricular dysfunction out of proportion to the severity of valvular disease documented by other non-invasive imaging modalities
  - suspected pericardial tamponade as documented by clinical findings or other non-invasive imaging modalities
  - suspected, or clinical uncertainty, between constrictive pericarditis versus restrictive cardiomyopathy physiology when there are questions left unanswered by other cardiac non-invasive imaging modalities
  - known or suspected cardiomyopathy with or without heart failure documented by prior advanced imaging
  - Re-evaluation of known cardiomyopathy for any of the following:
    - change in clinical status
    - change in cardiac exam
    - when required to guide therapy
  - hypertrophic cardiomyopathy
  - subvalvular aortic stenosis
- Right and left heart catheterization (CPT® 93453) is medically necessary when there is uncertainty between clinical impression and other non-invasive imaging modalities to evaluate the following valvular diseases:
  - mitral stenosis
  - mitral regurgitation
  - aortic stenosis
  - aortic regurgitation
- Left heart catheterization (CPT® 93452) for hemodynamic evaluation of the left ventricle and aorta is medically necessary to evaluate aortic stenosis when there is uncertainty between the clinical impression and non-invasive imaging modality findings.

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Evidence Discussion

Diagnostic right heart catheterization or right and left heart catheterization is performed for functional hemodynamic evaluation and may be combined with cardiac angiography for structural cardiac imaging and coronary angiography for anatomical evaluation of the coronary arteries. Diagnostic right heart catheterization or right and left heart catheterization is medically necessary in selected situations to obtain specific clinical information required for management decisions if it was not provided by non-invasive evaluation. This may be medically necessary in selected situations for management of valvular heart disease, myocardial disease, pericardial disease, congenital heart disease, pulmonary hypertension, heart failure, and post-cardiac transplantation. These indications are addressed in established evidence-based medical specialty organization guidelines pertaining to these conditions. If the clinical information required for management decisions was already provided by non-invasive evaluation, diagnostic heart catheterization does not add to management and, moreover, carries cardiac and extra-cardiac risks including coronary artery injury, myocardial infarction, vascular access site bleeding, ischemic stroke, and contrast-mediated acute kidney injury.

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Surgery, American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, and Society of Thoracic Surgeons [published correction appears in *J Am Coll Cardio*l. 2018 Apr 13]. *J Am Coll Cardiol.* 2017;69(17):2212-2241. doi:10.1016/j.jacc.2017.02.001.

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Adult Congenital Heart Disease

Guideline

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Atrioventricular Septal Defect (AV Canal, AVSD, Endocardial Cushion Defect) (CD-11.2.4)
Patent Ductus Arteriosus (PDA) (CD-11.2.5)
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Congenital Mitral Stenosis (CD-11.2.7)
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Right Ventricle-to-Pulmonary Artery Conduit (CD-11.3.8)
Transposition of the Great Arteries (TGA) (CD-11.3.9)
Congenitally Corrected TGA (CD-11.3.10)
Fontan Palliation of Single Ventricle Physiology (CD-11.3.11)
Severe Pulmonary Hypertension (PH) and Eisenmenger Syndrome (CD-11.3.12)
Coronary Artery Anomalies (CD-11.3.13)
References (CD-11)

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Congenital Heart Disease – General Information (CD-11.1)

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- This section covers adult congenital heart disease (CHD). For other associated disorders, please see the condition specific sections.
  - Marfan Syndrome
  - Hypertrophic cardiomyopathy (HCM)
  - Bicuspid aortic valve (BAV)

Definitions (CD-11.1.1)

Physiological stages (A, B, C, D)

Each congenital heart lesion is divided into 4 physiological stages (A, B, C, D).

Characteristics	Physiological stage			
	A	B	C	D
NYHA functional class	I	II	III	IV
Hemodynamic or anatomic sequelae	None	Mild ventricular enlargement of dysfunction, small shunt	Moderate or greater, ventricular dysfunction. Any venous or arterial stenosis	Moderate or greater, ventricular dysfunction. Any venous or arterial stenosis
Valvular	None	Mild	Moderate or greater	None
Aortic enlargement	None	Mild	Moderate	Severe

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Characteristics	Physiological stage			
	A	B	C	D
Exercise capacity limitation	Normal	Abnormal objective cardiac limitation	Moderate	Severe
Renal hepatic pulmonary dysfunction	None	None	Mild but responsive to medication	Refractory to treatment
Cyanosis/ hypoxemia	None	None	Mild	Severe
Arrhythmias	None	Arrhythmia not requiring treatment	Needs medical therapy	Refractory to medical therapy
Pulmonary hypertension	None	None	Mild to moderate	Severe or Eisenmenger

- CHD Anatomic classification
  - Class I-Simple
    - Native disease
      - isolated small ASD
      - isolated small VSD
      - mild isolated pulmonic stenosis
    - Repaired conditions
      - previously ligated or occluded ductus arteriosus
      - repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement
      - repaired VSD without significant residual shunt or chamber enlargement
  - Class II-Moderate Complexity
    - Repaired or unrepaired conditions
      - aorto-left ventricular fistula
      - anomalous pulmonary venous connection, partial or total
      - anomalous coronary artery arising from the pulmonary artery
      - anomalous aortic origin of a coronary artery from the opposite sinus

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- AVSD (partial or complete, including primum ASD)
- congenital aortic valve disease
- congenital mitral valve disease
- coarctation of the aorta
- Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
- infundibular right ventricular outflow obstruction
- ostium primum ASD
- moderate and large unrepaired secundum ASD
- moderate and large persistently patent ductus arteriosus
- pulmonary valve regurgitation (moderate or greater)
- pulmonary valve stenosis (moderate or greater)
- peripheral pulmonary stenosis
- sinus of Valsalva fistula/aneurysm
- sinus venosus defect
- subvalvular aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
- supravalvular aortic stenosis
- straddling atrioventricular valve
- repaired tetralogy of Fallot
- VSD with associated abnormality and/or moderate or greater shunt

◦ Class III-Great Complexity (or Complex)
  ▪ cyanotic congenital heart defect (unrepaired or palliated, all forms)
  ▪ double-outlet ventricle
  ▪ Fontan procedure
  ▪ interrupted aortic arch
  ▪ mitral atresia
  ▪ single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)
  ▪ pulmonary atresia (all forms)
  ▪ TGA (classic or d-TGA; CCTGA or l-TGA)
  ▪ truncus arteriosus
  ▪ other abnormalities of atrioventricular and ventriculoarterial connection (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Modalities (CD-11.1.2)

• Echocardiogram-transthoracic (TTE) or transesophageal (TEE)

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  - Transthoracic echocardiography (TTE) is an indispensable tool in the initial and serial follow-up evaluation to identify abnormalities and changes that commonly influence management decisions.
- Cardiac MRI (CMR)
  - CMR plays a valuable role in assessment of RV size and function, because it provides data that are reproducible and more reliable than data obtained with alternative imaging techniques.
  - For intracardiac congenital heart disease, CMR will typically include flow velocity mapping for shunts and flow assessment.
  - Imaging that only requires aortic arch imaging, does not require intracardiac CMR, only MRA Chest.
- Cardiac Computed Tomography (CCT) and Cardiac Computed Tomography Angiography (CCTA)
  - The most important disadvantage of CCT (including CT angiography) as an imaging technique is the associated exposure to ionizing radiation.
- Cardiac catheterization
  - (hemodynamic and/or angiographic) in individuals with adult CHD AP classification II and III, or interventional cardiac catheterization in individuals with adult CHD AP classification I to III should be performed by, or in collaboration with, cardiologists with expertise in adult CHD.
- Exercise Testing
  - Exercise test does not imply stress imaging.
- Stress Imaging
  - includes MPI, stress echo, stress MRI
  - PET stress may be included as per **Cardiac PET (CD-6)**.
- Circumstances where CMR, CCT, TEE, and/or Cardiac Catheterization may be Superior to TTE
  - assessment of RV size and function in repaired Tetralogy of Fallot (TOF), systemic right ventricles, and other conditions associated with right ventricular (RV) volume and pressure overload
  - identification of anomalous pulmonary venous connections
  - serial assessment of thoracic aortic aneurysms, especially when the dilation might extend beyond the echocardiographic windows
  - accurate assessment of pulmonary artery (PA) pressure and pulmonary vascular resistance
  - assessment for re-coarctation of the aorta
  - sinus venosus defects
  - vascular rings
  - evaluation of coronary anomalies
  - quantification of valvular regurgitation

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Coding (CD-11.1.3)

Modality	
Echocardiogram	
Transthoracic echocardiogram (TTE)	CPT®
TTE for congenital cardiac anomalies; complete	93303
TTE for congenital cardiac anomalies; limited study	93304
TTE (2D) m-mode recording, complete, with spectral and color flow doppler echocardiography	93306
TTE (2D) with or without m-mode recording; complete	93307
TTE (2D) with or without m-mode recording; limited study	93308
Transesophageal echocardiogram (TEE)	
TEE (2D) including probe placement, imaging, interpretation, and report	93312
TEE for congenital cardiac anomalies; including probe placement, imaging, interpretation, and report	93315
MRI	
Cardiac (CMR)	CPT®
Cardiac MRI for morphology and function without contrast	75557
Cardiac MRI for morphology and function without and with contrast	75561
MRI Chest	
MRI Chest without contrast	71550
MRI Chest with contrast	71551

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Modality	
MRI Chest with and without contrast	71552
MRI Angiography (MRA) / MRA Chest	
MRA Chest (excluding myocardium) with or without contrast	71555
CT	
Cardiac (CCT)	CPT®
CT Heart, with contrast material, for evaluation of cardiac structure and morphology	75572
CT Heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease	75573
CT Angiography-cardiac (CCTA)	CPT®
CTA Heart, coronary arteries and bypass grafts (when present), with contrast, including 3D image post-processing	75574
CT Chest	CPT®
CT Thorax without contrast	71250
CT Thorax with contrast	71260
CT Thorax without and with contrast	71270
CT Angiography-Chest (CTA Chest)	CPT®
CTA Chest without and with contrast	71275
Stress Imaging (echo, MRI, MPI)	
Stress echo	CPT®

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Modality	
Echocardiography (TTE), (2D), with or without m-mode, during rest and cardiovascular stress, with interpretation and report	93350
Echocardiography (TTE), (2D), m-mode, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation	93351
Stress MRI	CPT®
Cardiac MRI for morphology and function without contrast, with stress imaging	75559
Cardiac MRI for morphology and function without and with contrast, with stress imaging	75563
Myocardial perfusion imaging (MPI)	CPT®
MPI, tomographic (SPECT) including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78451
MPI, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78452
Pulmonary perfusion imaging	CPT®
Pulmonary perfusion imaging (e.g., particulate)	78580
Pulmonary ventilation (e.g., aerosol or gas) and perfusion imaging	78582
Quantitative differential pulmonary perfusion, including imaging when performed	78597

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Modality	
Quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed	78598

Congenital Heart Disease Imaging Indications

- The following sections are based on the congenital heart lesion. Requests for imaging based on other cardiac conditions, such as CAD, HCM, and acquired valvular lesions, should follow the adult cardiac guidelines for those conditions.

Aortopathies with CHD

- Dilated aortic arches are not uncommon with several congenital heart diseases and postoperative procedures including Aortic stenosis, Ross repair, Tetralogy of Fallot, Transposition of the great arteries (TGA), Pulmonary atresia, hypoplastic left heart syndrome (HLHS), Truncus Arteriosis, and single ventricle.

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ASD-Atrial Septal Defects (CD-11.2.1)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - CMR, CCT (CPT® 75573), and/or TEE are medically necessary when TTE is suboptimal and either:
    - ASD is suspected
    - to evaluate pulmonary venous connections in known ASD
  - MRA Chest or CTA Chest are medically necessary when TTE shows pulmonary venous anomalies
    - If normal, repeat pulmonary vein imaging is not required.
- Diagnostic catheterization is medically necessary when there is either:
  - evidence of pulmonary hypertension
  - unanswered questions on CMR/CCT for venous drainage
- Stress imaging and coronary artery imaging is based on **Stress Testing with Imaging – Indications (CD-1.4)**.

Consideration of surgery

- Transesophageal echocardiogram (TEE) is recommended to guide percutaneous ASD closure.

Post-procedure imaging

- TTE is medically necessary post ASD device placement:
  - 1 week (if Amplatzer)
  - 1 month
  - 6 months (evaluate for erosion)
  - 12 months
  - then every 1-2 years
- Due to low-risk of erosion in PFO devices, PFO device closure requires follow-up at 6-12 months. No additional evaluation unless PFO not closed (see **Frequency of Echocardiography Testing (CD-2.3) PFO closure, TIPS, Cardiac device therapy, LVAD).**

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Surveillance imaging ASD surgically closed or without surgical interventions

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	36	24	12	12

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Anomalous Pulmonary Venous Connections (CD-11.2.2)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - CMR and/or MRA Chest, or CT Cardiac and/or CTA Chest at time of diagnosis if any issues with pulmonary veins or RV volume
  - Cardiac catheterization at time of diagnosis for hemodynamic data and issues not answered on other imaging
- Routine stress imaging or coronary artery imaging not required
- Echocardiogram, CMR, CT, per cardiology request for clinical changes
  - Diagnostic heart catheterization if questions unanswered on imaging

Consideration of surgery

- Echocardiogram, CMR, CT, per cardiology request
- Diagnostic heart catheterization if unanswered questions on other imaging is needed for surgical management

Surveillance imaging anomalous pulmonary venous connections

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echo (TTE)	36	24	12	12

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Ventricular Septal Defect (VSD) (CD-11.2.3)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - CMR or CCT can be performed if questions are unanswered on echocardiogram
  - Catheterization at time of diagnosis for hemodynamics if pulmonary hypertension (PH) or shunt size is a question

Consideration of surgery

- TTE
  - CMR or CCT can be performed if questions are unanswered on echocardiogram.
  - Catheterization at time of diagnosis for hemodynamics if pulmonary hypertension (PH) or shunt size is a question

Surveillance imaging VSD

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echo (TTE)	36	24	12	12

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Atrioventricular Septal Defect (AV Canal, AVSD, Endocardial Cushion Defect) (CD-11.2.4)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - CMR or CT Cardiac at time of diagnosis if there are unanswered questions on echocardiogram
  - Cardiac catheterization at time of diagnosis when CMR and TTE leave questions unanswered that affect individual management
- Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**

Consideration of surgery

- TTE
  - CMR or CT Cardiac if there are unanswered questions on TTE and information is needed for surgical management
  - Cardiac catheterization if CMR and TTE leave questions unanswered that affect surgical management

Surveillance imaging AVSD

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echo (TTE)	24	24	12	12

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Patent Ductus Arteriosus (PDA) (CD-11.2.5)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - MRI or MRA Chest or CT or CTA Chest if there are questions left unanswered by echocardiogram
  - Cardiac catheterization for hemodynamics (if planned device closure, diagnostic cardiac catheterization is not medically necessary as it is included in the procedure code)
- Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**

Consideration of surgery

- TTE
- MRI or MRA Chest or CT or CTA Chest if there are unanswered questions on echo and information is needed for surgical management

Surveillance imaging PDA

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echo (TTE)	36	24	12	12

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Cor Triatriatum (CD-11.2.6)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - CMR and/or MRA Chest or CT Cardiac and/or CTA Chest medically necessary as baseline, with clinical changes, and prior to surgery
  - Diagnostic catheterization may be approved if additional information is required for medical management

Consideration of surgery

- TTE
- CMR and/or MRA Chest or CT Cardiac and/or CTA Chest

Surveillance imaging

- Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**

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Congenital Mitral Stenosis (CD-11.2.7)

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Imaging at baseline and for cardiac clinical changes

- TTE

Consideration of surgery

- TTE

Surveillance imaging congenital mitral stenosis

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echo (TTE)	24	24	12	12

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Subaortic Stenosis (SAS) (CD-11.2.8)

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This section relates to subaortic stenosis caused by a discrete membrane or tunnel-like obstruction.

Imaging at baseline and for cardiac clinical changes

- Echocardiogram (TTE) (CPT® 93303 or 93304 or 93306 or 93308)
- Stress echo (CPT® 93350 or 93351) or SPECT MPI (CPT® 78452) or Stress MRI (CPT® 75559 or 75563)
  - once at the time of diagnosis
  - new or changed signs or symptoms of ischemia
  - changes in cardiac function
  - any signs or symptoms allowed in **Stress Testing with Imaging – Indications (CD-1.4)**

Consideration of surgery

- Echocardiogram (TTE) (CPT® 93303 or 93304 or 93306 or 93308)
- Stress echo (CPT® 93350 or 93351) or SPECT MPI (CPT® 78452) or Stress MRI (CPT® 75559 or 75563)

Surveillance imaging SAS

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echo (TTE)	24	24	12	12
Stress imaging		24	24	12

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Congenital Valvular Aortic Stenosis (CD-11.2.9)

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Imaging at baseline and for cardiac clinical changes

- TTE
- TEE is medically necessary when TTE is limited or equivocal.
- MRA Chest or CTA Chest if one of the following:
  - suspicion of Coarctation based on exam and echocardiogram
  - proximal ascending aorta not well visualized on TTE

Consideration of surgery

- TTE
- TEE is medically necessary when TTE is limited or equivocal.

Surveillance imaging congenital valvular aortic stenosis

Modality	Physiological stage / intervals for routine imaging			
Stage (valvular AS)	**Progressive (stage B) Mild Vmax 2.0-2.9 m/s**	**Progressive (stage B) Moderate Vmax 3.0-3.9 m/s**	**Severe (stage C) ≥4.0 m/s**	**Aortic root dilation >4.5 cm**
TTE	3 years	1 years	6 months	12 months
MRA Chest or CTA				if ascending allowed yearly

Degree of aortic stenosis (AS) severity			
	Mild AS	**Moderate AS**	**Severe AS**
Vmax (m/s)[a] maximum Doppler velocity	2.0-2.9	3.0-3.9	≥4.0

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Degree of aortic stenosis (AS) severity			
	Mild AS	**Moderate AS**	**Severe AS**
Mean gradient (mmHg)[a]	<30	30-49	≥50
AVA (cm^2) aortic valve area	>1.5	1.0-1.5	<1.0
AVAi (cm^2/m^2 BSA) indexed aortic valve area	≥1.0	0.6-0.9	<0.6
[a]At normal transvalvular flow, BSA= body surface area			

Adapted from: ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC).

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Aortic Disease in Turner Syndrome (CD-11.2.10)

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Dissection more common for a given aortic diameter. Mid-ascending aortic disease more common and my not be reliably seen on echocardiogram

Imaging at baseline and for cardiac clinical changes

- TTE
- MRA Chest or CTA Chest to rule out mid ascending aortic aneurysm if mid aorta was not seen on echocardiogram

Consideration of surgery

- TTE
- MRA Chest or CTA Chest if mid aorta was not seen on echocardiogram

Surveillance imaging aortic disease in Turner Syndrome

- Echocardiogram (TTE) yearly
  - MRA Chest or CTA if mid ascending aorta not visualized
- For documented thoracic aortic aneurysm (TAA) ≤ 4cm
  - Routine MRA Chest or CTA yearly
- For documented thoracic aortic aneurysm (TAA) > 4cm
  - MRA Chest or CTA every 6 months.

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Supravalvular Aortic Stenosis (CD-11.3.1)

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Supravalvular aortic stenosis is a relatively rare condition overall but is seen commonly in individuals with Williams syndrome or homozygous familial hypercholesterolemia.

Imaging at baseline and for cardiac clinical changes

- TTE
- MRA Chest or CTA Chest
- Cardiac MRI or CTA Cardiac to assess coronary ostia
- New cardiac symptoms-any of the following:
  - CT Cardiac or cardiac MR
  - CTA Chest or MRA Chest
  - Stress imaging as per **Stress Testing with Imaging – Indications (CD-1.4)**

Consideration of surgery

- Cardiac catheterization for any individuals pre-cardiac intervention for coronary arteries

Surveillance imaging- Supravalvular AS

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	24	24	12	12
CMR or CCT	36	36	36	36

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Coarctation of the Aorta (CD-11.3.2)

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Coarctation is suspected based on clinical findings:
- BP higher in upper extremities than in the lower extremities
- absent femoral pulses
- continuous murmur
- abdominal bruit
- berry aneurysm with hemorrhage
- rib notching on x-ray
- abnormal thoracic aortic imaging and blood pressures

Imaging at baseline and for cardiac clinical changes
- TTE
  - No further imaging is required if echocardiogram (TTE), blood pressure, and exam rule out Coarctation.
  - If echocardiogram and exam are equivocal or positive, one of the following is medically necessary:
    - CTA Chest
    - MRA Chest
  - Individuals with Coarctation of the aorta do not require intra-cardiac MRI unless issue cannot be resolved on echocardiogram.
  - Screening for intracranial aneurysm by MRA (CPT®70544, 70545, 70546) or CTA (CTP® 70496) of head is medically necessary.
- ETT for diagnosis of exercise-induced hypertension does not require imaging.
- Cardiac MRI not required unless issues unresolved by echo for intracardiac anatomy.
- Diagnostic catheterization is medically necessary prior to stenting of the coarctation.
- Stress imaging, TEE, Cardiac MRI or CT, Coronary imaging are not routine.

Symptomatic
- Individuals with Coarctation are at risk for dissection. When individual has new or worsening symptoms any of the following:
  - echocardiogram (TTE)
  - MRA Chest or CTA
- For exertional symptoms, one of the following:
  - Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**
  - Cardiac MRI or CT Cardiac

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Consideration of surgery

- TTE
- MRA Chest or CTA Chest if TTE is equivocal or positive
- Diagnostic catheterization is medically necessary prior to stenting of the coarctation.

Surveillance imaging Coarctation of the Aorta

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	24	24	12	12
MRA Chest or CTA Chest	36	36	12	12

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Valvular Pulmonary Stenosis (CD-11.3.3)

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Imaging at baseline and for cardiac clinical changes

- TTE
- For issues affecting management not well visualized on TTE
  - Cardiac MRI or CT Cardiac
  - MRA Chest or CTA Chest

Consideration of surgery

- TTE
- Cardiac MRI or CT Cardiac and/or MRA Chest or CTA Chest for issues affecting surgical management not well visualized on TTE

Valvular PS imaging and testing

- Echocardiogram-stages
  - Mild PS-peak gradient <36 mmHg (peak velocity < 3m/s)
  - Moderate PS-peak gradient 36-64 mmHg (peak velocity 3-4 m/s)
  - Severe PS-peak gradient >64 mmHg (peak velocity > 4 m/s); or mean gradient >35 mmHg.
- Routine stress imaging is not required.
- Routine chest or cardiac or ischemia workup not required.

Surveillance imaging Valvular PS

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	36	24	12	12

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Isolated Pulmonary regurgitation after PS repair-Echo and CMR at same interval as TOF

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	24	12	12	12
CMR	36	24	12	12

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Branch and Peripheral pulmonary stenosis (CD-11.3.4)

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Overview
- Can be seen in newborns as a normal variant in the first 6 months of life
- Can be seen in surgeries of right ventricular outflow (TOF)
  - Noonan
  - Alagille
  - Williams
  - Maternal rubella exposure
  - Keutel syndrome

Imaging at baseline and for cardiac clinical changes
- TTE
- MRA Chest or CTA Chest
- Cardiac CT or Cardiac MR
- Catheterization may be considered if other advanced imaging is not adequate for management.
- VQ scan or MRA Chest for differential blood flow

Consideration of surgery
- TTE
- MRA Chest or CTA Chest
- Cardiac CT or Cardiac MR
- Catheterization is medically necessary when other advanced imaging does not provide necessary information for surgical management.

Surveillance imaging branch and peripheral pulmonary stenosis

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	24	24	12	12

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Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Cardiac MRI or CT Cardiac	36	36	24	24
MRA Chest or CTA Chest	36	36	24	24

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Double Chambered RV (CD-11.3.5)

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Imaging at baseline and for cardiac clinical changes

- TTE

Consideration of surgery

- TTE

Surveillance imaging double chambered right ventricle (RV)

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echocardiogram (TTE)	24	24	12	12

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Ebstein Anomaly (CD-11.3.6)

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Imaging at baseline and for cardiac clinical changes

- TTE
- TEE if TTE is not adequate
- Cardiac MRI or CT Cardiac

Consideration of surgery

- TEE
- Cardiac MRI or CT Cardiac

Surveillance imaging Ebstein Anomaly

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echocardiogram (TTE)	12	12	12	12
Cardiac MRI or CT Cardiac	60	36	24	12

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Tetralogy of Fallot (TOF, VSD with PS) (CD-11.3.7)

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Includes TOF with pulmonary atresia, VSD PA

Imaging at baseline and for cardiac clinical changes

Baseline

- TTE
- Cardiac MR or CCT/CCTA Cardiac
- MRA Chest or CTA Chest
- Cardiac catheterization if other advanced imaging leaves unanswered questions

New or worsening symptoms

- Repeat baseline advanced imaging
  - new or worsening symptoms
  - new EKG changes
- Stress imaging (stress echo, stress MRI, or MPI) allowed for typical chest pain, even if intermediate pretest probability at atypical symptoms in individuals with known or undefined coronary artery (CA) anatomy or CA pathology
- VQ scan or MRA chest for left/right perfusion abnormality

Prior to cardiac intervention or surgery

- Repeat baseline imaging (TTE/MR/CT)
- Catheterization prior to surgery or intervention
  - If planned Catheter Pulmonary Valve replacement, procedure includes diagnostic catheterization and hemodynamics, and diagnostic catheterization is not billed separately.

Surveillance imaging Tetralogy of Fallot (TOF)

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	24	12	12	12

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Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Cardiac MRI or CCT/CCTA	36	24	12	12
CTA Chest or MRA	36	24	12	12

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Right Ventricle-to-Pulmonary Artery Conduit (CD-11.3.8)

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Imaging at baseline and for cardiac clinical changes

- TTE
- Cardiac MRI or CCT/CCTA Cardiac
- MRA Chest or CTA Chest
- Catheterization allowed for new symptoms or with new imaging findings as needed for management
- Stress imaging (stress echo, stress MRI or MPI) as requested for cardiac symptoms

Consideration of surgery- Surgical repair for many lesions such as TOF/ Truncus / Pulmonary atresia

- TTE
- Cardiac MRI or CCT/CCTA Cardiac
- MRA Chest or CTA Chest

Surveillance imaging Right Ventricle-to-Pulmonary Artery Conduit

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	12	12	12	12
CMR or CCT/ CCTA	36	36	12	12
MRA Chest or CTA Chest	36	36	12	12

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Transposition of the Great Arteries (TGA) (CD-11.3.9)

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Imaging at baseline and for cardiac clinical changes

- TTE
- Cardiac MRI or CCT/CCTA
- MRA Chest or CTA

In addition to repeat imaging with the above baseline studies, the following studies are medically necessary as follows:

- stress imaging as requested for symptoms or signs of ischemia
- V/Q scan for left to right PA perfusion or MRA Chest
- Symptomatic individuals should be offered stress physiological imaging and repeat anatomic imaging considered if symptoms are suggestive of coronary ischemia (regardless of Diamond Forester pretest probability category).
- catheterization right and left heart when above advanced imaging does not explain clinical issues

Consideration of surgery

- TTE
- Cardiac MRI or CCT/CCTA
- MRA Chest or CTA

Surveillance imaging TGA

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE	12	12	12	12
CMR or CCT/ CCTA	36	24	12	12

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Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
MRA Chest or CTA Chest	36	24	12	12

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Congenitally Corrected TGA (CD-11.3.10)

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Imaging at baseline and for cardiac clinical changes

- TTE
- CMR or CCT/CCTA and MRA or CTA Chest
- Repeat CMR and/or Echocardiogram for changes in clinical status

Consideration of surgery

- TTE
- CMR or CCT/CCTA and MRA or CTA Chest

Surveillance imaging congenitally corrected TGA

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echocardiogram (TTE)	12	12	12	12
CMR or CCT/ CCTA	36	36	12	12
CTA Chest or MRA Chest	36	36	12	12

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Fontan Palliation of Single Ventricle Physiology (CD-11.3.11)

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Including Tricuspid Atresia and Double Inlet Left Ventricle, HLHS, HRHS, PA, Mitral atresia, AVC unbalanced, single ventricle, DIRV, pulmonary atresia, HLHS, Glen procedure, TA, double outlet right ventricle (DORV), and single ventricle physiology

Imaging at baseline and for cardiac clinical changes

- TTE
- CMR or CCT/CCTA for issues that are equivocal on TTE (can be done annually versus based on below chart for individuals who have prior issues that were equivocal on echo, and the data is required for management)
- Cardiac catheterization prior to surgical interventions
- Echocardiogram/CMR/CCT or CCTA/MRA Chest or CTA Chest/catheterization with any new signs or symptoms
- V/Q scan or MRA for lung perfusion left vs. right

Consideration of surgery

- Cardiac catheterization prior to surgical interventions

Surveillance imaging Fontan Palliation of Single Ventricle Physiology

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Echocardiogram (TTE)	12	12	12	12
CMR or CCT/ CCTA Cardiac	36	24	24	24
CTA Chest or MRA	36	24	24	24

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Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
Cardiac catheterization	120 (10 years)	120 (10 years)	120 (10 years)	120 (10 years)

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Severe Pulmonary Hypertension (PH) and Eisenmenger Syndrome (CD-11.3.12)

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Imaging at baseline or for cardiac clinical changes

- TTE
  - initial diagnosis
  - with new signs or symptoms
- CMR/CCT or CCTA or CTA/MRA Chest or Cardiac catheterization
  - echocardiogram (TTE) results suggest PH
  - new signs or symptoms with PH

Consideration of surgery or for treatment planning

- TTE
- Cardiac catheterization if TTE suggests PH
- CMR/CCT or CCTA or CTA/MRA Chest if additional information is needed for treatment planning

Surveillance imaging Severe Pulmonary artery hypertension (PH) and Eisenmenger syndrome

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	**A**	**B**	**C**	**D**
TTE			6	6
CMR or CCT/ CCTA			As needed	As needed
MRA Chest or CTA Chest			As needed	As needed
Catheterization			As needed	As needed

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Coronary Artery Anomalies (CD-11.3.13)

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Imaging at baseline and for cardiac clinical changes

- TTE
  - at baseline
  - any signs or symptoms
- CCT/CCTA or CMR or catheterization for initial evaluation
- Coronary artery from wrong sinus-baseline stress imaging regardless of symptoms
- Stress imaging for any cardiac signs or symptoms
- For Kawasaki guideline regarding echo, Stress imaging, coronary imaging, see pediatric guideline **Kawasaki Disease (PEDCD-6)**.

Consideration of surgery

- TTE
- CCT/CCTA or CMR or catheterization

Evidence Discussion (CD-11)

Adult Congenital Heart Disease Imaging

- Management of Adult Congenital heart (ACHD) disease involves a multimodal imaging approach. Multimodal imaging helps account for patient-specific considerations, strengths and weaknesses of each modality, institutional resources, and available expertise. Approaches to limit and monitor radiation exposure are recommended during imaging of individuals with ACHD, and studies that do not involve ionization radiation should be prioritized whenever appropriate.
- Echocardiogram, MRI, CT, catheterization, and nuclear stress testing all have strengths and weaknesses, including varying radiation levels, temporal resolution, spatial resolution, and ability to image blood flow. Advantages of the various modalities are as follows: MRI has the ability to image the right ventricle in 3D; Echocardiograms include high temporal resolution and availability, and the ability to measure pressures with catheterizations; Cardiac CTs have the ability to rapidly acquire 3D imaging; and nuclear imaging allows the ability to measure myocardial perfusion concerns.

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Perinatal Imaging in Cardiovascular Disease

Guideline

Diagnostic Cardiovascular Imaging Pre-Pregnancy to Post-Partum (CD-15.1)
Maternal Imaging in Cardiovascular Disease (CD-15.2)
Maternal Imaging in Individuals with Aortopathy (CD-15.3)
Imaging in Pregnancy with Congenital Heart Disease (CHD) (CD-15.4)
References (CD-15)

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Diagnostic Cardiovascular Imaging Pre-Pregnancy to Post-Partum (CD-15.1)

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Peripartum Red Flag Signs and Symptoms

Imaging as requested, as listed within **Imaging Modalities in Maternal Imaging** (Echocardiogram, Exercise stress echo, Coronary angiography, Cardiac MRI), is medically necessary for peripartum individuals for any of the following **red flag** signs or symptoms:

- chest pain
- dyspnea
- orthopnea
- cough
- lower extremity edema when there is a concern for heart disease
- tachycardia
- unheralded syncope
- headache
- acute visual changes
- new onset hypotension
- hypertension

Imaging Modalities

General information

Ultrasound and magnetic resonance are the preferred imaging modalities to minimize radiation exposure in pregnancy. When imaging using ionizing radiation is necessary, radiation exposure should follow the ALARA principle (As Low As Reasonably Achievable). Shared decision-making regarding diagnostic testing should occur in consultation with the individual, cardiologist, and obstetrical team when weighing the risk of fetal exposure to radiation against the need to diagnose or plan treatment for serious illness.

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Transthoracic echocardiography (TTE)

- TTE (CPT® 93306) is the primary cardiac imaging modality in pregnancy. Baseline and surveillance echocardiography is medically necessary for several conditions as noted in **Maternal imaging in cardiovascular disease (CD-15.2)** Table 1.
- A repeat echocardiogram is medically necessary when there are new or worsening cardiovascular signs or symptoms, as described in **Peripartum Red Flags**, **Transthoracic Echocardiography (TTE)– Indications/initial evaluation (CD-2.2)** and **Frequency of Echocardiography Testing (CD-2.3)**, during and after pregnancy.

Exercise Stress Echo

- Exercise stress echo (CPT® 93350, 93351) is medically necessary pre-conception to assist with risk stratification in individuals with a documented history of **any** of the following:
  - current left ventricular dysfunction
  - previous history of left ventricular dysfunction
  - valvular heart disease of any severity
  - There is a concern for myocardial reserve.
- See Stress echocardiogram in **Stress Testing with Imaging – Indications (CD-1.4)** and **Stress echo–indications other than ruling out CAD (CD-2.7)**.
- See **Hypertrophic Cardiomyopathy (CD-14).**

Coronary Angiography

- Fetal risk from ionizing radiation is highest before 20 weeks gestational age. When coronary angiography is medically necessary, the ALARA principle should be followed. Invasive management of acute coronary syndrome is associated with lower in-hospital mortality and should be considered. See **Diagnostic Heart Catheterization**.

Cardiac MRI

- Cardiac MRI (CPT® 75557) is utilized in pregnant individuals to measure aortic dimensions, wall motion, and ventricular function when the echocardiogram is non-diagnostic. Gadolinium-based contrast agents are not necessary in aortic imaging or most other indications in pregnancy. See **Cardiac MRI**.

Evidence Discussion

- Cardiovascular disease (CVD) in pregnancy has become increasingly prevalent in recent years.
- The increase in plasma volume during pregnancy requires significant physiological adaptation.
- Maternal mortality has increased in the last two decades with CVD accounting for 33% of all deaths.

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- Invasive management of myocardial infarction (MI) is associated with lower in-hospital mortality.
- Research has underrepresented females of childbearing age leading to significant deficits in our knowledge of cardiovascular care of these individuals.
- Cardiac Imaging using ionizing radiation
  - Multiple imaging modalities expose the pregnant individual and fetus to ionizing radiation.
  - This exposure causes concern for an elevated risk of childhood cancer.
  - Shared decision-making should be employed when weighing the fetal exposure to radiation against the need to diagnose serious illness.

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Maternal Imaging in Cardiovascular Disease (CD-15.2)

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Table 1: Suggested frequency of echo monitoring during pregnancy

Cardiovascular disease	Monitoring frequency
Hypertensive disorders of pregnancy (BP ≥130/90)	• An echocardiogram (TTE) (CPT® 93303, 93304, 93306, 93307, 93308) is medically necessary once during pregnancy in all hypertensive disorders of pregnancy. • A repeat TTE is medically necessary when there are new or worsening cardiovascular signs or symptoms.
Valvular disorders/ Native and Prosthetic	• 1 TTE is medically necessary during the first trimester (weeks 1-12 of pregnancy) for individuals with known or suspected valvular heart disease. • A repeat TTE is medically necessary when there are new or worsening cardiovascular signs or symptoms.
Severe Aortic stenosis (AS)	• A repeat TTE is medically necessary every 1-2 months or when there are new or worsening cardiovascular signs or symptoms.
Mitral stenosis (MS)	• TTE is medically necessary each trimester (12 weeks) and prior to delivery in individuals with mild MS. • TTE is medically necessary every 1–2 months in individuals with moderate to severe MS. • A repeat TTE is medically necessary when there are new or worsening cardiovascular signs or symptoms.
Dilated cardiomyopathy	• TTE is medically necessary during each trimester (12 weeks). • A repeat TTE is medically necessary when there are new or worsening cardiovascular signs or symptoms.

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Cardiovascular disease	Monitoring frequency
Hypertrophic cardiomyopathy	• TTE is medically necessary in **asymptomatic** individuals each trimester. • TTE is medically necessary in **symptomatic** individuals every 1-2 months. • A repeat TTE is medically necessary when there are new or worsening cardiovascular signs or symptoms.
Peripartum cardiomyopathy	• TTE is medically necessary in individuals with signs and symptoms of heart failure. • A repeat TTE is medically necessary when there are new or worsening cardiovascular signs or symptoms. ◦ TTE is medically necessary in subsequent pregnancies: ▪ at the time of the first prenatal visit ▪ at the end of the first and second trimesters ▪ 1 month prior to delivery ▪ after delivery prior to discharge ▪ 1 month postpartum ▪ at any time when there are worsening signs or symptoms of heart failure • TTE is medically necessary as requested when the individual is being treated with Cabergoline for peripartum cardiomyopathy.
Pulmonary hypertension	• TTE is medically necessary in individuals with signs and symptoms of pulmonary hypertension. • A repeat TTE is medically necessary at the discretion of the health care provider.

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Maternal Imaging in Individuals with Aortopathy (CD-15.3)

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Pre-pregnancy imaging

Individuals at risk for aortic aneurysms **(Table 15-3-1)** should be evaluated with echocardiogram (TTE) **and** Computed Tomography (CT)/Magnetic Resonance Imaging (MRI) of the Chest/Abdomen/Pelvis **(Table 15-3-2)** within 1 year prior to conception to evaluate for aortic valve disease and aortic dimensions.

Table 15-3-1

Individuals at risk for aortic aneurysm, aortic dissection, limb-threatening ischemia
Bicuspid Aortic Valve
Turner Syndrome
Coarctation of the Aorta
Marfan Syndrome
Type IV Ehlers-Danlos
Loeys-Dietz
Familial Thoracic Aortic Disease and Aortic Dissection (defined as first-degree relative with history of aortic dissection or thoracic aortic aneurysm or two second-degree relatives with history of aortic dissection or thoracic aortic aneurysm)

Table 15-3-2

Imaging for Aortic conditions	CPT®
CT Chest and/or Abdomen and/or Pelvis	71260 74177 74160 72193

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Imaging for Aortic conditions	CPT®
CTA Chest and/or Abdomen and/or Pelvis	71275 74175 72191 74174
MRA Chest and/or Abdomen and/or Pelvis	71555 74185 72198
Transthoracic Echocardiogram (TTE)	93303 93304 93306 93307 93308
Transesophageal Echocardiogram (TEE)	93312 93313 93314 93315 93316 93317
Doppler echocardiography is medically necessary as add-on codes for TEE	+93320 +93321 +93325

Surveillance imaging during pregnancy and postpartum

Follow-up imaging with Echocardiogram (TTE) and CTA/MRA. TEE can be substituted if TTE is equivocal.

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Suggested Frequency of Aortic Imaging during pregnancy and postpartum in individuals known to be high-risk for aortic aneurysm

Table 15-3-3

Condition	WHO Class	Imaging frequency	Postpartum (up to 42 days after birth)
Turner Syndrome	• II-III: Aortic root <20mm/m^2 with associated risk factors or <25mm/m without associated risk factors	• Once during pregnancy if normal aortic dimension, or every 2 months if repaired coarctation	• Once during the postpartum period
	• IV: Aortic root ≥20mm/m2 with associated risk factors or ≥25mm/m without associated risk factors	• Every 6 weeks if aorta diameter dilated >30mm	• Once during the postpartum period
	• Any individual with Turner who has severe coarctation	• At discretion of provider	• Once during the postpartum period
Marfan Syndrome	• III: Aortic root <45mm, mod-severe Aortic Insufficiency	• Every trimester if <40mm	• Once during the postpartum period
	• IV: Aortic root ≥45mm, history of dissection	• Every 6 weeks if aorta is ≥40mm	• Once during the postpartum period
Vascular Ehlers-Danlos	• Type IV	• Every 6 weeks	• Once during the postpartum period
Loeys-Dietz	• III: Aortic diameter <40mm	• Every 6 weeks	• Once during the postpartum period
	• IV: Aortic diameter ≥40mm	• Every 6 weeks	• Once during the postpartum period

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Condition	WHO Class	Imaging frequency	Postpartum (up to 42 days after birth)
Familial thoracic aortic aneurysms and dissections	• III: Aortic diameter <40mm	• Every trimester if <40mm aortic diameter	• Once during the postpartum period
	• IV: Aortic diameter ≥40mm	• Every 6 weeks if ≥40mm aortic diameter	• Once during the postpartum period

Background and Supporting Information

First-degree relative (sibling, parent, child)

Second-degree relative (aunt/uncles, grandparent, niece, nephew, cousin, or half-sibling of an individual)

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Imaging in Pregnancy with Congenital Heart Disease (CHD) (CD-15.4)

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Pre-pregnancy imaging based on the World Health Organization (WHO) chart for imaging in pregnancy with CHD

Imaging modality and indication	CPT®
Echocardiogram (TTE) when planning pregnancy	93306
TEE if TTE equivocal	93312
CMR (cardiac MRI) can be performed prior to pregnancy in those lesions where CMR would be routinely performed at some later date	75557
CTA or MRA of chest if known aortic disease, Pulmonary artery disease, anomalous pulmonary veins, anomalous systemic veins (also see peripartum aortopathy table)	71275, 71555
Echocardiogram with new signs or symptoms	93303, 93304, 93306, 93308
Postpartum imaging per provider requested frequency	imaging as noted above
Stress imaging pre/during pregnancy when known coronary artery anomaly, pulmonary hypertension, LVOT obstruction, cardiac dysfunction, single ventricle	93350, 93351, 93320, 93325
WHO II, III, IV, can have echo/MR/CT/stress imaging prior to pregnancy	imaging as noted above

- Congenital heart disease imaging in pregnancy
  - Echocardiogram (TTE) when planning pregnancy
  - TEE if TTE equivocal
  - CMR can be performed prior to planning pregnancy in those lesions where CMR would be routinely performed at some later date.

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- CTA Chest or MRA Chest of arch if known disease with aortic involvement or if known dilation
- Repeat echocardiogram and MR (can be without gad) can be performed based on the WHO classification II, III, IV, or other risk factors.
- Severe complex CHD TTE (CPT® 93306)
  - Every 2-4 weeks for major physiological changes
  - As often as needed for any of the following:
    - pulmonary hypertension
    - changes in function
    - to guide delivery after 24 weeks
- Echocardiogram can be performed if there are new signs or symptoms during pregnancy.
- Postpartum first year can have more frequent imaging.
- Stress imaging pre/during pregnancy for individuals with known coronary artery anomaly, pulmonary hypertension, LVOT obstruction, cardiac dysfunction, single ventricle.
- WHO II, III, IV can have echocardiogram/MR/CT/stress imaging prior to pregnancy
- WHO I-one echocardiogram during pregnancy
- WHO II-one echocardiogram per trimester during pregnancy
- WHO II/III-echocardiogram every 2 months during pregnancy
- WHO III/IV-echocardiogram monthly during pregnancy
  - Individuals may require more (even weekly) if treatment decision, delivery is considered.

- Syndromes that allow cardiac imaging at the time of diagnosis if not previously done. This list is not exhaustive.
  - DiGeorge/velocardiofacial (22q11.2)
  - Down syndrome (trisomy 21)
  - Holt Oram (TBX5)
  - Klinefelter syndrome (47 XXY)
  - Noonan (PTPN11, KRAS, SOS1 RAF1, NRAS, BRAF, MAP2K1)
  - Turner (45X)
  - Williams (7q11.23 deletion)
  - any syndrome associated with congenital heart disease
- Echocardiogram at time of diagnosis (either genetic testing or clinical features)
- CMR or CCTA if arch involved in disease.

Surveillance Imaging

Surveillance imaging after baseline studies

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After initial imaging, TTE should be performed at regular intervals. For individuals with aortopathy, Chest MRA may be added at the same frequency. If echocardiographic imaging is known to be poor or inadequate, CMR may be used instead of TTE.

The World Health Organization modified classification of maternal cardiovascular risk

The World Health Organization established a modified classification of maternal cardiovascular risk used as a tool to evaluate risk status for pregnant individuals with various cardiovascular conditions.

Class	Risk	Sample Lesions
WHO Class I	No detectable increased risk of maternal mortality and no or mild increase in morbidity.	• Mild Pulmonary stenosis • Small PDA • Mild MVP • Successfully repaired simple lesions (ASD, VSD, PDA, anomalous pulmonary venous drainage) • Isolated PACs or PVCs
WHO Class II	Small increased risk of maternal mortality or moderate increase in morbidity.	• Un-operated ASD or VSD • Repaired TOF (uncomplicated) • Most arrhythmias
WHO Class II-III	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity.	• Mild left ventricular impairment • Hypertrophic cardiomyopathy • Native or tissue valvular heart disease not considered WHO I or IV • Aorta <45 mm in aortic disease associated with bicuspid aortic valve • Repaired coarctation

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Class	Risk	Sample Lesions
WHO Class III	Significantly increased risk of maternal mortality or severe morbidity. • Expert counseling required. • If pregnancy is decided upon, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth, and the postpartum period.	• Mechanical valve • Systemic right ventricle • Fontan circulation • Unrepaired cyanotic heart disease • Other complex congenital heart disease
WHO Class IV	Extremely high-risk of maternal mortality or severe morbidity. • Pregnancy contraindicated. • If pregnancy occurs, termination should be discussed. • If pregnancy continues, care for as WHO class III.	• Pulmonary arterial hypertension from any cause • Severe systemic ventricular dysfunction (LVEF <30%, NYHA functional class III-IV) • Severe mitral stenosis; severe symptomatic aortic stenosis • Aortic dilation ≥50 mm in aortic disease associated with bicuspid aortic valve • Native severe coarctation of the aorta

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Cardiotoxic Agent-Related Cardiac Dysfunction

Guideline

Cardiotoxic Agent-Related Cardiac Dysfunction (CD-12)
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Cardiotoxic Agent-Related Cardiac Dysfunction (CD-12)

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Cardiotoxic Agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)

Transthoracic Echocardiogram (TTE)

Transthoracic Echocardiogram (TTE) is medically necessary to determine Left Ventricular (LV) function in individuals on cardiotoxic chemotherapeutic drugs.

- 3D echocardiography (CPT® 93319) is medically necessary in addition to the primary TTE and is the preferred echocardiography modality for the assessment of left ventricular ejection fraction (LVEF) and cardiac volumes. See **3D Echocardiography (CD 2.9)**.
- Myocardial strain imaging (CPT® 93356) to obtain a Global Longitudinal Strain (GLS) is medically necessary as an important adjunct in screening for CTRCD. See **Myocardial Strain Imaging (CD 12.2).**

Multimodality imaging

- Guidelines support using echocardiography rather than MUGA for the determination of LVEF and/or wall motion EXCEPT in one of the circumstances described previously in **MUGA Study – Cardiac Indications (CD-3.4)** (see **Background and supporting Information** below).
- CT coronary calcium scoring (CPT® 75571) is medically necessary to screen for radiation induced coronary artery disease as follows:
  - The individual has no symptoms suspicious for ischemia such as chest discomfort located retrosternally, epigastric, or referred pain (pain that is felt in a place different than the source) in the neck, shoulder, arm, or jaw described as pain, pressure, constricting, or tightness occurring with exertion or emotional stress and relieved by rest, nitroglycerin, or both.
  - The individual has no clinical history or prior cardiac imaging documenting CAD.
  - There has been no prior calcium score >0.
  - There has been ≥5 years since the last RT (radiotherapy/radiation) treatment for the initial study or at least 5 years since the last CT calcium scoring for subsequent testing.

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- Cardiac MRI (CMR) is medically necessary in the evaluation of CTRCD for the following:
  - TTE is not diagnostic
  - tissue characterization for cardiomyopathy or myocarditis, particularly when Immune Checkpoint Inhibitor Myocarditis is being questioned
  - See also **Cardiac MRI and MRA Chest – Indications (CD 5.2)**.

Frequency of CTRCD screening

- Baseline
  - All individuals can have a baseline TTE prior to initiation of cardiotoxic agents.
- During treatment with cardiotoxic agents
  - The frequency of monitoring depends on the agent administered and the individual's baseline cardiovascular toxicity risk. (see **Background and Supporting Information** below)
- Post treatment with cardiotoxic agents surveillance
  - One TTE is medically necessary 3 to 12 months after completion of therapy.
- Adult cancer survivors
  - Long term surveillance TTE is medically necessary every 5 years, except in those with low risk.
  - Additional surveillance TTE at 1 and 3 years after cancer therapy completion is medically necessary in those deemed to be high risk for CTRCD.
- Adult survivors of childhood and adolescent cancer
  - Surveillance TTE is medically necessary every 2 years for survivors with indeterminate risk due to unknown cancer therapy history.
  - Surveillance TTE is medically necessary every 2 years for high risk survivors, defined as a history of any of the following:
    - Radiation dose ≥30 Gy
    - Anthracycline dose of ≥100 mg/m^2 and radiation dose ≥15 Gy
    - Anthracycline dose ≥250 mg/m^2
  - Surveillance TTE is medically necessary every 5 years for moderate risk survivors, defined as a history of any of the following:
    - Radiation dose ≥15 to <30 Gy
    - Anthracycline dose of < 250 mg/m^2 and radiation dose <15 Gy
  - No screening is medically necessary in low risk survivors, defined as a history of any of the following:
    - Anthracycline dose of >0 to <100 mg/m^2
    - Radiation dose >0 to <15 Gy

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- TTE is medically necessary for cancer survivors with a history of chest radiotherapy or anthracycline exposure who are pregnant or planning to become pregnant as follows:
  - Baseline exam
  - 12 weeks of pregnancy if missed the baseline TTE or in high risk survivors
  - 20 weeks of pregnancy for high risk survivors
  - TTE can be repeated for any cardiac symptoms at any other time as needed during or immediately following pregnancy
- For adults who received anthracyclines in childhood, see **Cardiotoxicity and Echocardiography (PEDONC-19.2).**

Background and Supporting Information

- High value screening protocol to detect CTRCD using TTE (CPT® 93306 or 93308) is contingent upon careful baseline cardiotoxicity risk assessment and stratification into low, moderate, and high risk. Currently, there is no universally accepted risk score. Documentation of risk by the provider is sufficient to guide screening strategy.
- Advantages of Echocardiography in comparison to MUGA in individuals on cardiotoxic chemotherapy:
  - no ionizing radiation
  - no IV access required when echo contrast is not used
  - allows view of the pericardium to look for effusion
  - allows estimate of pulmonary pressure
  - may allow visualization of a clot or tumor in the Inferior Vena Cava (IVC) and/or the right heart

Myocardial Strain Imaging (CD-12.2)

- Myocardial strain imaging (CPT® 93356) in addition to the primary echocardiogram in individuals receiving therapy with cardiotoxic agents for **any** of the following:
  - Initial evaluation-prior to treatment with **either**:
    - medications that could result in cardiotoxicity/heart failure
    - radiation that could result in cardiotoxicity/heart failure
  - re-evaluation of an individual previously or currently undergoing therapy as per echocardiogram parameters (see **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1))**
  - re-evaluation of an individual undergoing therapy with worsening symptoms

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Mavacamten for Obstructive Hypertrophic Cardiomyopathy (HCM) (CD-12.3)

Echocardiogram (CPT® 93306) is medically necessary for individuals treated with mavacamten for class II-III obstructive HCM as follows:

Initiation of treatment

- Baseline-at the beginning of treatment
- 4 weeks after treatment initiation
- 8 weeks after treatment initiation
- 12 weeks after treatment initiation
- Then every 12 weeks while on mavacamten

Changes in treatment

- 4 weeks after any interruption of treatment (any missed dose)
- After any dosage change (including restart of treatment):
  - 4 weeks after dosage change
  - 12 weeks after dosage change
- After initiating a weak CYP2C19 inhibitor (e.g., omeprazole) or moderate CYP3A4 inhibitor (e.g., ciprofloxacin):
  - 4 weeks after start of medication
  - 12 weeks after start of medication
- At any time regardless of timing of prior echo when there are new cardiac signs or symptoms, or worsening of clinical status

See also **Hypertrophic Cardiomyopathy (HCM) (CD-14)**

Background and Supporting Information

Hypertrophic Cardiomyopathy (HCM) is a clinical diagnosis, established by imaging with 2D echocardiography or cardiovascular magnetic resonance (CMR) showing a maximal end-diastolic wall thickness of ≥15 mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults. More limited hypertrophy (13-14 mm) can be diagnostic, particularly when present in family members of an individual with HCM or in conjunction with a positive genetic test, and/or associated with typical dynamic outflow obstruction, or distinctly abnormal ECG patterns.

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Cardiac Sarcoidosis

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Cardiac Sarcoidosis (CD-3.9)
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Cardiac Sarcoidosis (CD-3.9)

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Imaging indications

Suspected cardiac sarcoidosis

- Cardiac MRI with gadolinium (CPT® 75561) is medically necessary initial imaging for identification of suspected cardiac sarcoidosis.
- Cardiac PET (CPT® 78432 or 78433) **or** PET metabolic study (CPT® 78459 or 78429) and SPECT perfusion image (CPT® 78451) is medically necessary for suspected cardiac sarcoidosis for any of the following indications:
  - initial imaging when there is a contraindication to MRI imaging (non MRI-safe pacemaker, renal failure)
  - initial Cardiac MRI is equivocal or additional information is needed based on results from the MRI
  - to confirm diagnosis if suggested by MRI
  - initial imaging to evaluate for suspected cardiac sarcoidosis when there is documented sarcoidosis outside of the heart

Monitoring of treatment of established cardiac sarcoidosis

- PET-Cardiac PET metabolic is medically necessary to monitor therapy in cardiac sarcoidosis. Requires PET with F-18 FDG metabolic study combined with a PET perfusion study (CPT® 78432 or 78433) OR PET metabolic study (CPT® 78459 or 78429) and SPECT perfusion image (CPT® 78451).
  - prior to treatment of cardiac sarcoidosis
  - PET (heart FDG metabolic with perfusion study as above) can be repeated at 3-6 month intervals if there is active disease or to make therapeutic decisions.

Evidence Discussion

- Cardiac imaging is reasonable to detect cardiac sarcoidosis in the following:
  - any individual with extra cardiac sarcoidosis even if no cardiac symptoms
  - echo with basal thinning of the intraventricular septum, depressed EF <50 or regional wall motion abnormality not associated with CAD
  - young individuals with unexplained ventricular tachycardia, especially monomorphic VT

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  - individuals with unexplained cardiomyopathy or heart failure (i.e., CAD has been ruled out)
  - individuals with unexplained arrhythmia especially advanced AV block or VT
- Initial imaging should be cardiac MRI with late gadolinium enhancement (LGE) protocol unless there is a contraindication to MRI imaging (non MRI-safe pacemaker, renal failure). Absence of LGE is a strong negative predictor for low rates of cardiac morbidity and mortality from cardiac sarcoidosis and further testing is not usually medically necessary.
- PET – Metabolic imaging with F-18 FDG for diagnosis if there is a contraindication to MRI and cardiac sarcoidosis is suspected. This requires PET with F-18 FDG metabolic study combined with a PET perfusion study.
- Full body PET/CT (CPT® 78815) is not medically necessary for the diagnosis or monitoring response to therapy of cardiac sarcoidosis. It may be considered to assist in diagnosis and/or treatment options in some instances of pulmonary sarcoidosis. See **Sarcoid (CH-15.1)** in the Chest Imaging Guidelines.

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Cardiac Amyloidosis

Guideline

Myocardial Tc-99m Pyrophosphate Imaging in Cardiac Amyloidosis (CD-3.8)

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Myocardial Tc-99m Pyrophosphate Imaging in Cardiac Amyloidosis (CD-3.8)

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Coding

MUGA (Multi Gated Acquisition) – Blood Pool Imaging	CPT®
Myocardial Imaging, infarct avid, planar, qualitative or quantitative	78466
Myocardial Imaging, infarct avid, planar, qualitative or quantitative with ejection fraction by first pass technique	78468
Myocardial Imaging, infarct avid, planar, qualitative or quantitative tomographic SPECT with or without quantification	78469
Radiopharmaceutical Localization Imaging Limited area	78800
Radiopharmaceutical Localization Imaging SPECT	78803
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (e.g., head, neck, chest, pelvis), single-day imaging	78830

- Historically this method of imaging the myocardium was used to identify recent infarction, hence, the term "infarct-avid scan." Although still available, the sensitivity and specificity for identifying infarcted myocardial tissue are variable and the current use for this indication is limited. See **Cardiac MRI (CD-5)**.
- Tc-99m pyrophosphate imaging (CPT® 78469, 78803, or 78830) is medically necessary to identify cardiac amyloidosis.
- Chest SPECT and planar imaging may be used, as well as whole-body imaging for identification of systemic ATTR (transthyretin) amyloidosis.

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- For a single planar imaging session alone (without a SPECT study), report CPT® 78800 Radiopharmaceutical Localization Imaging Limited area
- Note: When reporting CPT 78803, planar imaging of a limited area or multiple areas should be included with the SPECT

Indications

Tc-99m pyrophosphate imaging (CPT® 78469, 78803, or 78830) is medically necessary for **any** of the following:

- For diagnosis of ATTR amyloidosis in an individual undergoing evaluation for kidney transplant when **both**:
  - There is known systemic amyloidosis.
  - Cardiac MRI (CMR) is either contraindicated or indeterminate. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.
- For diagnosis of ATTR amyloidosis after negative screening for presence of a monoclonal light chain to exclude AL amyloidosis :
  - Serum kappa/lambda free light chain ratio (not SPEP)
    - Abnormal if ratio is <0.26 or >1.65
  - Serum or urine immunofixation electrophoresis (IFE)
    - Abnormal if monoclonal protein detected
- Diagnosis of cardiac ATTR in an individual with cardiac MRI or echocardiography findings consistent with or suggestive of cardiac amyloidosis
- Diagnosis of cardiac ATTR in an individual with known systemic amyloidosis or ATTR gene positive
- Diagnosis of an individual with suspected cardiac ATTR amyloidosis when there is a contraindication to CMR such as renal insufficiency or an implantable cardiac device

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Note:

Cardiac follow-up should be based on Echocardiogram, Tn, NT-proBNP, clinical exam and symptom

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Background and Supporting Information

- The following conditions would raise high index of suspicion:
  - left ventricular hypertrophy but low voltage on ECG
  - heart failure with preserved ejection fraction and an increase in left ventricular wall thickness
  - unexplained heart failure with preserved ejection fraction and concomitant right heart failure in an individual over the age of 60

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- individuals, especially elderly males, with signs/symptoms of heart failure and any of the following:
  - lumbar spinal stenosis
  - spontaneous biceps tendon rupture
  - bilateral carpal tunnel syndrome
  - atrial arrhythmia in the absence of usual risk factors
- known or suspected familial amyloidosis
- low flow, low gradient aortic stenosis

Evidence Discussion

Appropriate Use Criteria from major professional societies including the American College of Cardiology and the American Heart Association support the use of myocardial perfusion imaging (MPI) with single photon emission computed tomography (SPECT) for individuals with signs or symptoms consistent with coronary artery disease (CAD) such as typical angina. In addition, they support the use of SPECT perfusion imaging in symptomatic individuals with known CAD and those with prior interventions such as coronary artery bypass surgery, coronary stenting, and preoperative risk stratification. These guidelines balance the appropriate testing of individuals with SPECT versus unnecessary and potentially harmful testing and downstream procedures.

Also, as supported by society guidelines, use of SPECT is medically necessary in detection of cardiac transthyretin amyloidosis (ATTR) after light chain amyloidosis (AL) is ruled out by appropriate blood and urine testing.

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Cardiac Trauma

Guideline

Cardiac Trauma Imaging (CD-10.1)
References (CD-10)

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Cardiac Trauma Imaging (CD-10.1)

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Cardiac Trauma Imaging (CD-10.1)

- One of the following can be used to evaluate cardiac or aortic trauma:
  - Echocardiogram (TTE, TEE)
  - Cardiac MRI Cardiac (CPT® 75557 or 75561, and 75565)
  - Cardiac CT Cardiac (CPT® 75572)
  - CCTA (CPT® 75574)
  - Chest CTA Chest (CPT® 71275)
  - Chest CT Chest (CPT® 71260, CPT 71270)

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Congestive Heart Failure

Guideline

Congestive Heart Failure (CD-9)
References (CD-9)

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Congestive Heart Failure (CD-9)

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CHF – Imaging (CD-9.1)

- Congestive heart failure (CHF), including post-cardiac transplant failure:
  - Echocardiogram is the first study after the clinical evaluation for suspected CHF.
  - MUGA, cardiac MRI, or cardiac CT may be medically necessary if the ECHO is limited or does not completely answer the question.
  - Stress test to assess for CAD may be medically necessary. Follow stress testing guideline: **Stress Testing with Imaging – Indications (CD-1.4).**
- Arteriovenous fistula with "high output" heart failure:
  - CT Chest with contrast (CPT® 71260) and/or CT Abdomen and/or CT Pelvis with contrast (CPT® 74160 or 72193 or 74177) **or**
  - CTA Chest (CPT® 71275) and/or CTA Abdomen and/or CTA Pelvis (CPT® 74175 or CPT® 72191 or CPT® 74174) **or**
  - MRI Chest and/or MRI Abdomen and/or MRI Pelvis without and with contrast (CPT® 71552 and/or 74183 and/or 72197) **or**
  - MRA Chest and/or MRA Abdomen and/or MRA Pelvis (CPT® 71555 and/or CPT® 74185 and/or CPT® 72198)
- Right-sided congestive heart failure can be a manifestation of pulmonary hypertension or serious lung disease.
  - CT Chest (CPT® 71260) or CTA Chest (CPT® 71275) to evaluate for recurrent pulmonary embolism

Evidence Discussion

- Congestive heart failure is a complex clinical syndrome with signs and symptoms that are a result of structural and/or functional impairment of ventricular ejection or filling which results in objective evidence of pulmonary or systemic congestion.
- Identification of the etiology of the cardiac dysfunction is crucial in the diagnosis of heart failure (HF) to determine subsequent management.
- Evaluation of HF usually entails multiple different diagnostic tools incorporating tools such as echocardiogram, stress testing, genetic testing, nuclear imaging, CT/ MRI imaging, endomyocardial biopsy and cardiac catheterization.
- Treatment of HF is targeted towards treating the underlying cause, improving the function and structure of the heart, and reducing mortality and morbidity. This not only includes medications but may also entail the use of cardiac resynchronization

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devices, monitoring devices, ventricular assist devices, cardiothoracic surgery, and heart transplant.

Myocardial Sympathetic Innervation Imaging in Heart Failure (CD-3.6)

- Nuclear imaging using I-123-meta-iodobenzylguanidine (I-123-mIBG) in an attempt to image increased myocardial sympathetic activity is considered to be experimental and investigational.
- The AMA has established the following set of Category III codes to report these studies:
  - CPT® 0331T Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment
  - CPT® 0332T Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment; with tomographic SPECT

Background and Supporting Information

In heart failure, the sympathetic nervous system is activated in order to compensate for the decreased myocardial function. Initially, this is beneficial. However, long-term this compensatory mechanism is detrimental and causes further damage.

Markers have been developed using radioactive iodine in an attempt to image this increased myocardial sympathetic activity. Currently, AdreView™ (Iodine-123 meta-iodobenzylguanidine) is the only FDA-approved imaging agent available for this purpose.

Evidence Discussion

I-123-meta-iodobenzylguanidine (MIBG) imaging of the sympathetic nerve activity of the heart has been proposed and approved for the identification of individuals with heart failure. However, its clinical utility has not found widespread acceptance, and its clinical usefulness remains in question. There are no societal guidelines for its routine use. The guidelines contain many other imaging platforms such as echocardiography, magnetic resonance imaging, perfusion, and metabolic imaging that have proven superior for the diagnosis and management of individuals with heart failure.

Left Ventricular Assist Devices (LVAD) (CD-9.4)

Left ventricular assist devices (LVAD) are implantable devices used in individuals with advanced heart failure refractory to medical therapy as a bridge to transplantation or as a destination therapy.

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Pre-LVAD implant

The following imaging studies are medically necessary for preoperative evaluation prior to planned LVAD implant:

- TTE (CPT® 93306)
- 3D rendering for echocardiography (CPT® 76376 or 76377)
- Cardiac MRI (CPT® 75557 or 75561) and CMR velocity flow mapping (CPT® 75565)
- CT Chest (CPT® 71250 or 71260) or CTA Chest (CPT® 71275) or MRI Chest (CPT® 71552) or MRA Chest (CPT® 71555)
- CTA Abdomen and Pelvis (CPT® 74174) or MRA Abdomen and Pelvis (CPT® 74185 and 72198)
- CT Coronary Angiography (CPT® 75574) in individuals post-coronary artery bypass grafting to assess the location and course of the bypass grafts to guide the surgical approach
- Transesophageal echocardiography (CPT® 93312, 93320, 93325)
- Right heart catheterization (CPT® 93451) or Right and left heart catheterization (CPT® 93453)

Post LVAD implant

TTE (CPT® 93306) is medically necessary as follows:

- Post-implant at the following intervals:
  - 2 weeks
  - 1 month
  - 3 months
  - 6 months
  - 12 months
  - Every 6 months thereafter

CT

- CTA Chest (CPT® 71275) or CT Chest with contrast (CPT® 71260) is medically necessary for evaluation of LVAD malfunction.
- CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) is medically necessary for the evaluation of LVAD-related infections.

FDG PET/CT for LVAD driveline infection (CPT® 78815 orCPT® 78429)

- Early infection detection for LVAD drivelines is desirable, since once the infection extends to the cannula and pump pocket, eradication becomes difficult. CT findings are nonspecific and metal device artifacts of the driveline itself affects sensitivity.
- FDG PET/CT is medically necessary for suspected LVAD infection if other studies and examination remain inconclusive.
- See **FDG PET/CT for Infections (CD-6.5)**.

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Right heart catheterization (RHC)

- RHC (CPT® 93451) or Right and Left heart catheterization (CPT® 93453) as needed for hemodynamic assessment to guide changes to therapy.

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Pre-surgical Cardiac Testing

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Cardiac Surgery Imaging (CD-13)

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Pre-surgical Cardiac Testing – General Information (CD-13.1)

- It is important to differentiate requests for preoperative CT imaging before cardiac surgery according to type of procedure planned:
  - Primary cardiac operation – individuals who have not had prior heart surgery
  - Redo procedures – individuals who have had a prior procedure (it is important to determine the type of procedure as this may impact which modality is most appropriate for the pre-operative assessment)
  - Minimally invasive procedures, such as minimally invasive aortic valve operations, minimally invasive or robotic mitral operations, TAVR, MitraClip™ or other percutaneous valve procedures (such as valve in valve aortic or mitral, percutaneous tricuspid and TMVR which will be increasing in the future)
- In re-operative cardiac surgery, the benefit of preoperative CT is to assess for aortic calcifications, to evaluate the anatomic relationships in the mediastinum, such as the location of the various cardiac chambers and great vessels and proximity to the sternum, and to assess for the location of prior bypass grafts. Information can then be used to change the operative strategy including non-midline approach, peripheral vascular exposure, and alternative cannulation sites and for establishing cardiopulmonary bypass before re-sternotomy. These techniques can result in decreased incidence of intraoperative injury to heart, great vessels and prior bypass grafts and lower rates of postoperative stroke. IV contrast is necessary with these studies to delineate the anatomic structures. However, in individuals with renal insufficiency, the provider might choose to forgo the contrast if they do not want to contrast load the individual prior to placing them on the heart-lung machine.
- Aortic atherosclerosis is recognized as the single most important determinant of postoperative stroke. There is evidence to support that preoperative CT is associated with lower postoperative stroke rates and mortality after primary cardiac surgery.
  - CT Chest without contrast (CPT® 71250) can be performed pre-operatively to allow the surgeon to:
    - visualize the extent and location of aortic atherosclerosis
    - change the operative strategy such as those problematic areas are avoided

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Primary Cardiac Surgery – No Previous Cardiac Surgery (CD-13.2)

- CT Chest without contrast (CPT® 71250) to evaluate for the presence of ascending aortic calcification may be medically necessary prior to primary cardiac surgery when there is documented high risk for aortic calcification including any of the following:
  - aortic calcification on chest x-ray or other diagnostic test (TEE, fluoroscopy, etc.)
  - calcific aortic stenosis
  - end stage renal disease (dialysis)

Re-operative Cardiac Surgery (CD-13.3)

- Individuals undergoing re-operative cardiac surgery may undergo **one** of the following tests for preoperative assessment:
  - CT Chest with contrast
  - CTA Chest
  - CCTA only if prior CABG (this might be in addition to CT with contrast as CCTA will not show the extent of the thoracic aorta that needs to be visualized)
  - CT Heart usually does not provide the necessary information and is not medically necessary routinely.

Minimally Invasive or Robotic Cardiac Surgery (CD-13.4)

- CTA Chest CPT® 71275 (or CT Chest with contrast CPT® 71260) and CTA Abdomen and Pelvis CPT® 74174 (or CT Abdomen and Pelvis with contrast CPT® 74177) are medically necessary for pre-operative assessment of suitability for the approach and for procedural planning of arterial and venous cannulation and cardiopulmonary bypass for individuals being considered for minimally invasive or robotic cardiac surgical procedures including the following:
  - valve repair or replacement
  - coronary artery bypass graft surgery
  - aortic root or ascending aorta repair
  - resection of intracardiac tumor, or thrombus or vegetation
  - open lead extraction
  - atrial septal defect repair

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Transcatheter Valve Interventions (CD-13.5)

Transcatheter Mitral Valve Repair (mitral valve clip)

- The following imaging may be used to determine if an individual is eligible for the procedure:
  - Transthoracic echo (TTE) (CPT® 93306) with or without 3D rendering
  - Transesophageal echo with or without 3D rendering
  - Heart catheterization, including right heart catheterization if requested
  - CCT (CPT® 75572) is recommended in individuals with radiation-induced valve disease
- CTA/CTV of Chest, Abdomen, and Pelvis is not medically necessary prior to planned transcatheter mitral valve repair.
- Post-procedure transthoracic echo (TTE) can be performed at the following intervals:
  - 1 month
  - 6 months
  - 1 year
  - then annually

Transcatheter Tricuspid Valve Replacement

Pre-procedure imaging

- The following imaging studies are medically necessary to determine eligibility and for procedure planning Pre-Transcatheter Tricuspid Valve Replacement (TTVR):
  - Cardiac CT CPT® 75572
  - CTV Chest CPT® 71275
  - CTV Abdomen and Pelvis CPT® 74174

Note:

Cardiac CT (CPT® 75572) is recommended in individuals with radiation-induced valve disease.

Post-procedure follow up

- TTE (CPT® 93306) is medically necessary post-procedure at the following intervals:
  - 1 month
  - 6 months
  - 1 year
  - then annually

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CT Imaging in Transcatheter Aortic Valve Replacement (TAVR)

Pre-TAVR imaging

Pre-aortic valve replacement

- Once the decision has been made for aortic valve replacement, the following may be used to determine if an individual is a candidate for TAVR:
  - CTA Chest (CPT® 71275), Abdomen and Pelvis (combination code CPT® 74174) are medically necessary; and
  - CT Cardiac (CPT® 75572) is medically necessary to measure the aortic annulus; or
  - Coronary CTA (CCTA CPT® 75574) is medically necessary to both measure the aortic annulus and assess the coronary arteries in lieu of heart catheterization.
- A repeat diagnostic left heart catheterization is **not** medically necessary when the individual is undergoing a transcatheter aortic valve replacement (TAVR).

Transfemoral access not feasible

Alternative imaging can be obtained to evaluate vascular access for TAVR in individuals for whom it is documented either via the office note or prior imaging that transfemoral access would not be feasible due to **any** of the following exclusion criteria:

- small vessels
- highly calcified vessels
- stenosed or occluded vessels
- prior aortoiliac vascular intervention

Imaging is medically necessary based on the documented intended access site (transaxillary or transcarotid) and should be of the involved body areas. The following studies are medically necessary based on the documented planned access site:

- CTA of the Head (CPT® 70496) and/or Neck (CPT® 70498) for transcarotid access
- CTA of the Chest (CPT® 71275) and/or Upper Extremity (CPT® 73206) for transaxillary access

Post-TAVR imaging

CT Cardiac (CPT® 75572) is medically necessary:

- if any of the post-TAVR TTEs are indeterminate or raises a concern about any of the following:
  - valve thrombosis
  - infective endocarditis
  - structural degeneration
- when a Valve-in-Valve implantation or surgical re-do AVR is being contemplated

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- Routine CT surveillance or follow up for incidental Hypoattenuated Leaflet Thickening (HALT) with or without restricted leaflet motion, also referred to as Hypoattenuation Affecting Motion (HAM), is NOT medically necessary.

Evidence Discussion

The ability of the cardiac CT technology to provide a tomographic view of the cardiovascular system has resulted in its ubiquitous adoption in the pre-procedure planning for almost all cardiac structural interventions. Specifically, cardiac CT circumvents the image window limitation of echocardiography, allows high definition visualization of the posterior structures, and facilitates pre-procedural planning for pulmonary vein isolation, coronary sinus pacer leads insertion, and left atrial appendage occlusion device implantation, among other trans-catheter structural interventions.

In non-interventional settings, cardiac CT provides an alternative to cardiac MRI when structural information cannot be adequately obtained by an echocardiography. Most notably, the evaluation of a cardiac mass, extent of pericardial disease, complex congenital heart disease and cardiomyopathy, can be performed by a cardiac CT when cardiac MRI is not available or contraindicated.

Trans-catheter Aortic Valve Replacement (TAVR)

The success of a Trans-catheter Aortic Valve Replacement (TAVR) procedure is contingent upon a meticulous pre-TAVR planning imaging study where cardiac CT allows accurate annulus sizing, coronary heights measurement, and calcification distribution evaluation, in addition to access site planning. Post-operatively, clinically suspected complications such as thrombus formation, infective endocarditis, or structural degeneration can be confirmed on a cardiac CT; a routine surveillance strategy, however, is not supported because of unclear or even potentially harmful outcome of treating incidental findings.

Transcatheter Mitral Valve Repair (mitral valve clip)

In non-interventional settings, cardiac CT provides an alternative to cardiac MRI when structural information cannot be adequately obtained by an echocardiography. Most notably, the evaluation of a cardiac mass, extent of pericardial disease, complex congenital heart disease and cardiomyopathy, can be performed by a cardiac CT when cardiac MRI is not available or contraindicated. Percutaneous treatment of mitral regurgitation can be accomplished using venous access to apply a clip device (e.g., MitraClip currently FDA approved) to provide edge-to-edge mitral leaflet coaptation, approximating opposing sections of the anterior and posterior mitral valve leaflets. FDA approved indications include treatment for individuals with symptomatic, moderate to severe, or severe primary mitral regurgitation whose surgical risks are prohibitive, as well as symptomatic moderate to severe or severe secondary mitral

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regurgitation who have failed optimal medical therapy. This therapy should include, if medically necessary, cardiac resynchronization therapy.

Transcatheter Mitral & Tricuspid Valve Replacement

Transcatheter Tricuspid Valve Replacement device (EVOQUE) is currently FDA approved. In individuals being considered for transcatheter tricuspid or mitral valve replacement, Cardiac CT CPT® 75572 provides detailed imaging of the valve annulus including measurement of valve annulus to guide selection of the appropriate-sized valve. CTV Chest (CPT® 71275) and CTV Abdomen and Pelvis (CPT® 74174) provide imaging of the iliac veins and inferior vena cava to exclude stenosis or significant tortuosity and aid in determining eligibility and procedure planning.

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Pulmonary Hypertension

Guideline

Pulmonary Hypertension (PH) (CD-8.1)
References (CD-8)

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Pulmonary Hypertension (PH) (CD-8.1)

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Pulmonary Hypertension Imaging indications

Transthoracic echocardiogram (TTE) (CPT® 93306) should be performed initially as it can help determine the probability of pulmonary hypertension.

Screening

A screening echocardiogram (TTE) for PH is medically necessary in individuals with documented history of **any** of the following:

- individuals pre-operatively for planned liver transplant
- individuals evaluated for transjugular portosystemic shunt
- portal hypertension
- liver disease with signs and symptoms of PH
- bronchopulmonary dysplasia

Annual screening echocardiogram (TTE) is medically necessary in individuals with a documented history of any of the following:

- systemic sclerosis (SSc)
- individuals with PH mutations (e.g., BMPR2)
- first-degree relatives of individuals with PH
- connective tissue disorder with symptoms consistent with PH
- individuals with TR velocity ≥2.8 m/s, with no other findings on additional testing
- individuals being treated with medications associated with PH
- individuals who have a concern documented for PH and had a negative echocardiogram but still show signs or symptoms of PH

Follow-up testing is not medically necessary in individuals with TR velocity <2.8 m/s and no other signs, symptoms, or risk factors of PH.

Initial imaging

Transthoracic echocardiogram (TTE) (CPT® 93306) is medically necessary for symptoms and signs of PH including **any** of the following:

- notes documenting clinical concern for pulmonary hypertension
- EKG findings concerning for PH such as **any** of the following:
  - right ventricular hypertrophy (RVH)

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  - right axis deviation
  - right atrial enlargement
- right ventricular hypertrophy or pulmonary artery dilation on other images
- history of pulmonary embolism with persistent or new onset DOE, or exercise limitation
- suspected PH in individuals with lung disease
- DOE in individuals with connective tissue disorder, HIV, portal hypertension, SSc.
- Symptoms of PH (**any** of the following):
  - breathlessness
  - SOB
  - decreased exercise tolerance
  - fatigue and rapid exhaustion
  - palpitations
  - dyspnea on bending forward
  - hemoptysis
  - exercise induced abdominal distention and nausea
  - weight gain due to fluid retention
  - syncope during or shortly after physical activity
  - exertional chest pain

Stress echo (CPT® 93350 or 93351) is medically necessary for **any** of the following:

- to assess for treatment in the setting of concomitant valvular disease
- as indicated by **Stress Testing with Imaging-Indications (CD-1.4)** or **Stress Echo-Indications Other than Ruling out CAD (CD-2.7)**
- There is documented concern for chronic thrombo-embolic pulmonary hypertension.

Cardiac MRI (CPT® 75557) is medically necessary when there is documentation of **any** of the following:

- TTE is equivocal or unclear (e.g., for RV function), and the information is needed for management.
- MRI and TTE may both be required for individuals who need RV pressure and function assessed, and prior RV function cannot be assessed by echocardiogram.
- MRI can replace TTE when the issue that makes the imaging by echo unclear is likely to be seen in future echocardiograms.

Other advanced imaging is medically necessary after TTE for the following:

- High-resolution CT Chest (CPT® 71250) is medically necessary in the setting of hypoxemia to rule out restrictive lung disorders such as pulmonary fibrosis.
- CTA Chest (CPT® 71275) or MRA Chest (CPT® 71555) is medically necessary to evaluate for suspected acute and/or chronic pulmonary embolism.

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- V/Q scan (CPT® 78580-Pulmonary Perfusion Imaging or CPT® 78582-Pulmonary Ventilation (e.g., Aerosol or Gas) and Perfusion Imaging) is medically necessary to evaluate for any of the following:
  - suspected acute pulmonary embolism
  - to evaluate for chronic thromboembolic pulmonary hypertension at 3 to 6 months post pulmonary embolism if both of the following apply:
    - persistent or new onset dyspnea on exertion or exercise limitation
    - evidence of pulmonary hypertension on follow up echocardiogram
  - to evaluate for chronic thromboembolic pulmonary hypertension in individuals with pulmonary hypertension of uncertain etiology
- SPECT imaging (CPT® 78803) or SPECT/CT imaging (CPT® 78830) can be added to V/Q scan if requested.
- Transesophageal (TEE) contrast echocardiography or other imaging techniques (e.g., CT angiography, cardiac MRI) may be medically necessary, in addition to 2D Doppler and contrast examinations, to identify CHD to detect or exclude **any** of the following:
  - sinus venosus
  - atrial septal defects
  - patent ductus arteriosus
  - anomalous pulmonary venous connections

Indications for initial catheterization

Right heart catheterization (RHC) is medically necessary for **any** of the following:

- Echocardiogram findings
  - TR velocity ≥3.4 m/second
  - TR velocity ≥2.9 m/second and presence of other PH signs on echo or other testing, or risk factors or associated indications
- individuals with SSc where breathlessness remains unexplained (RHC is recommended despite normal echocardiogram)
- individuals with connective tissue disorder who have symptoms or concerns for PH with a negative or equivocal echocardiogram
- when recommended to determine if shunt closure is recommended due to congenital heart disease
- RHC if **moderate** to **severe** PH on echocardiogram
- RHC is medically necessary prior to starting PH medical therapy.
- Individuals with **low**-risk profile (based on an evidence based PAH Risk Score Calculator such as REVEAL 2.0, ESC/ERS & Compera 2.0) only need catheterization if medically necessary for another reason or equivocal studies.
- RHC is allowed when LHC indicated for separate indication.
- RHC preoperative for surgical intervention treating the cause of PH (MV, TV, AV, PV)
- Eisenmenger syndrome – RHC is medically necessary when requested by provider.

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Left heart catheterization (LHC) or Right and left heart catheterization as per the following guidelines:

- **Evaluation of Conditions Other than Coronary Artery Disease (CD-7.7)**
- **Diagnostic Left Heart Catheterization (LHC) (CD-7.3)**

Repeat Testing

Follow-up echocardiogram (TTE) on individuals with PH is medically necessary for any of the following:

- every 6 months for surveillance of stable individuals with moderate or severe pulmonary hypertension (pulmonary artery systolic pressure ≥50 mm Hg)
- 3 months after any change in PH therapy
- prior to planned intubation (e.g., for elective surgery)
- prior to planned pregnancy
- during pregnancy as often as requested by provider
- anytime, without regard for the number or timing of previous echo studies to evaluate change in clinical signs or symptoms

Pulmonary embolism (PE)

- TTE is medically necessary 3 to 6 months post pulmonary embolism if **any** of the following apply:
  - persistent or new onset dyspnea on exertion, or exercise limitation
  - pulmonary hypertension or right ventricular dysfunction on initial echo at PE diagnosis
  - history of recurrent pulmonary embolism

RHC is medically necessary for known PH as follows:

- at baseline
- then every 6 months
- anytime for clinical changes or with treatment changes

Other related sections

- **Frequency of Echocardiography Testing (CD-2.3)** in the Cardiac Imaging Guidelines
- **Right Heart Catheterization (RHC) (CD-7.4)** in the Cardiac Imaging Guidelines
- Severe **Severe Pulmonary Hypertension (PH) and Eisenmenger Syndrome (CD-11.3.12)** in the Cardiac Imaging Guidelines
- **Congenital Heart Disease Modality Considerations (PEDCD-2.3)** in the Pediatric Cardiac Imaging Guidelines
- **Pediatric Pulmonary Hypertension-General (PEDCD-7)** in the Pediatric Cardiac Imaging Guidelines

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- **Pulmonary Embolism (PE) (CH-25.1)** in the Chest Imaging Guidelines

Evidence Discussion

Pulmonary hypertension (PH) is a complex, chronic disease with multiple etiologies that requires extensive evaluation, including ECG (right ventricular hypertrophy with/without strain, right atrial dilatation); chest x-ray; arterial blood gas, pulmonary function testing, CT angiography based on the etiology.

PH can be subdivided into the following 5 groups based on the underlying cause:
- Pulmonary arterial hypertension (PAH)
- PH due to left heart disease
- PH due to lung disease
- Chronic thromboembolic PH (CTEPH)
- PH with unclear and/or multifactorial mechanisms

Probability of PH is assessed at initial evaluation:
- High probability:
  - TR velocity ≥3.4 m/s
  - TR velocity between 2.9 m/s to 3.4 m/s and **one** of the following:
    - Right ventricle or pulmonary artery enlargement
    - Interventricular septum flattening
    - Right ventricular systolic dysfunction
- Intermediate probability:
  - TR velocity between 2.9 m/s to 3.4 m/s in the absence of other signs of PH
- Low probability:
  - TR velocity <2.8 m/s

Peak TR velocity ≥2.8 m/s may suggest PH; however, the presence or absence of PH cannot be reliably determined by TR velocity alone.

In addition to the tricuspid regurgitation velocity, other findings on echo can increase the probability of PH. Examples of these findings include:
- abnormal tricuspid annular plane systolic excursion (TAPSE)
- abnormal RV fractional area change (RV-FAC)
- abnormal RV free-wall strain
- abnormal tricuspid annulus velocity (S wave) derived from tissue Doppler imaging
- abnormal RV ejection fraction (RVEF) derived from 3D echocardiography

MRI can be a useful test especially with respect to RV function.

Right heart catheterization is the gold standard for diagnosing PH.

See **Severe Pulmonary Artery Hypertension (PH) and Eisenmenger Syndrome (CD-11.3.12)** for additional information regarding Eisenmenger Syndrome.

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Pulmonary Vein Imaging

Guideline

Pulmonary Vein Imaging – Indications (CD-8.2)
References (CD-8)

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Pulmonary Vein Imaging – Indications (CD-8.2)

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Indications

- MRI Cardiac (CPT® 75557 or 75561), MRV Chest (CPT® 71555), CTV Chest (CPT® 71275), or CT Cardiac (CPT® 75572) to evaluate anatomy of the pulmonary veins:
  - prior to planned atrial fibrillation ablation/pulmonary vein isolation procedure
  - post-procedure between 3-6 months after ablation because of a 1% to 2% incidence of asymptomatic pulmonary vein stenosis
    - If no pulmonary vein stenosis is present, no further follow-up imaging is required.
    - If pulmonary vein stenosis is present on imaging following ablation and symptoms of pulmonary vein stenosis (usually shortness of breath) are present, imaging is considered medically necessary at 1, 3, 6, and 12 months.

Background and Supporting Information

The majority (81%) of pulmonary vein stenosis remain stable over 1 year. Progression occurs in 8.8%, and regression occurs in a small percentage.

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Cardiac Imaging in Individuals with Known or Planned Transplants

Guideline

Cardiac Imaging in Transplants (CD-16)

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Cardiac Imaging in Transplants (CD-16)

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Imaging Criteria

Codes Included

- SPECT MPI (CPT® 78451, 78452)
- Stress echocardiogram (CPT® 93350, 93351)
- Stress MRI (CPT® 75559, 75563)
- PET MPI (CPT® 78430, CPT® 78431, CPT® 78491, 78492)
- CT chest (CPT® 71250 or 71260)
- Ultrasound abdomen (CPT® 76700 or 76705)
- CT abdomen (CPT® 74150 or 74160)
- MRI abdomen (CPT® 74181 or 74183)
- Right heart catheterization (CPT® 93451)
- Right and left heart catheterization (CPT® 93453)
- Left heart catheterization (CPT® 93458)
- Echocardiogram (CPT® 93306)
- 3D echocardiogram (CPT® 93319, 76376, 76377)
- Strain imaging (CPT® 93356)

Non-cardiac transplants

- Stress Testing in Individuals for Non-Cardiac Transplant
  - Imaging stress testing every year (stress echocardiogram, SPECT MPI, stress MRI, or PET MPI per the transplant center's protocol) is medically necessary prior to transplant for candidates for any type of organ, bone marrow, or stem cell transplant. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.
  - An imaging stress test can be repeated annually after transplant for at least 2 years or within 1 year of a prior cardiac imaging study if there is evidence of progressive vasculopathy.
  - After 2 consecutive normal imaging stress tests, repeated testing is not medically necessary more often than every other year without evidence for progressive vasculopathy or new symptoms.
  - Stress testing after 5 years may proceed according to normal patterns of consideration.

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Cardiac transplant

- Pre-Cardiac Transplant evaluation
  - One of the following is medically necessary for candidates being evaluated for heart transplant as suggested by the transplant center protocol:
    - SPECT MPI
    - Stress MRI
    - Stress echocardiogram
    - PET MPI
  - The following modalities are medically necessary for an individual being evaluated for cardiac transplant:
    - CT Chest (CPT® 71250 or 71260) **and**
    - Abdominal imaging with: Ultrasound Abdomen (CPT® 76700 or 76705) or CT Abdomen (CPT® 74150 or 74160) and/or MRI Abdomen (CPT® 74181 or 74183) **and**
    - Right heart catheterization (CPT® 93451) or Right and left heart catheterization (CPT® 93453) or left heart catheterization (CPT® 93458)
- Post-Cardiac transplant assessment of transplant CAD:
  - Echocardiogram, 3D echocardiogram (CPT® 93319, CPT® 76376, CPT® 76377,) and strain imaging (CPT® 93356) may be performed as needed per transplant protocol
  - **One** of the following imaging studies may be performed annually:
    - SPECT MPI (CPT® 78451, CPT® 78452)
    - Stress echocardiogram (CPT® 93350, CPT® 93351)
    - Stress MRI (CPT® 75559, CPT® 75563)
    - PET MPI (CPT® 78430, CPT® 78431, CPT® 78491, CPT® 78492)

Background and Supporting Information

Individuals who have undergone organ transplant are at increased risk for ischemic heart disease secondary to their medication. Risk of vasculopathy is 7% at 1-year, 32% at 5 years, and 53% at 10 years.

For individuals being evaluated for cardiac transplant, MRI may be performed as the initial abdominal imaging modality or for further evaluation after initial abdominal imaging with Ultrasound or CT.

Evidence Discussion

Stress testing with imaging is medically necessary to exclude the presence of significant coronary artery disease as part of evaluating candidacy for any type of organ, bone marrow, or stem cell transplant. While on the transplant waiting list, stress testing with imaging is medically necessary annually to exclude progression of coronary artery

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disease. Stress testing with any imaging modality (stress echo, SPECT MPI, stress MRI, or stress cardiac PET perfusion) is supported per the transplant center's protocol. Recommendations regarding stress testing with imaging for pre-transplant evaluation are provided in established evidence-based medical specialty organization guidelines.

Stress testing with imaging also has a role in surveillance following cardiac transplant to evaluate for development of obstructive coronary artery disease due to cardiac allograft vasculopathy. Stress testing with any imaging modality (stress echo, SPECT MPI, stress MRI, or stress cardiac PET perfusion) is medically necessary annually post cardiac transplant. Recommendations regarding stress testing with imaging for post cardiac transplant surveillance are provided in established evidence-based medical specialty organization guidelines.

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Hypertrophic Cardiomyopathy

Guideline

Hypertrophic Cardiomyopathy (HCM) (CD-14)
References (CD-14)

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Hypertrophic Cardiomyopathy (HCM) (CD-14)

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HCM Imaging Indications

Hypertrophic Cardiomyopathy (HCM) is a clinical diagnosis, established by imaging with 2D echocardiography or cardiovascular magnetic resonance (CMR) showing a maximal end-diastolic wall thickness of ≥15 mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults. More limited hypertrophy (13-14 mm) can be diagnostic, particularly when present in family members of an individual with HCM or in conjunction with a positive genetic test, and/or associated with typical dynamic outflow obstruction, or distinctly abnormal ECG patterns.

Screening

- For screening for inherited hypertrophic cardiomyopathy, see **Transthoracic Echocardiography (TTE-Indications (CD-2.2)** and **Frequency of Echocardiography Testing (CD-2.3).**

Initial imaging, new or changed symptoms

Transthoracic echocardiography (TTE)

- TTE is medically necessary for the initial evaluation of a genotype positive individual with inherited hypertrophic cardiomyopathy.

Transesophageal echocardiography (TEE) (CPT® 93312, 93320, 93325)

- TEE is medically necessary for the evaluation of individuals with hypertrophic cardiomyopathy if TTE is inconclusive for **any** of the following:
  - mitral regurgitation secondary to structural abnormalities of the mitral valve
  - subaortic membrane or aortic valve stenosis
- TEE is medically necessary for the pre-procedure planning for surgical myectomy or alcohol septal ablation evaluation of individuals with hypertrophic cardiomyopathy.

Stress echocardiogram

- Exercise stress echo (CPT® 93351 or 93350) is medically necessary for the detection and quantification of dynamic left ventricular outflow tract obstruction in symptomatic individuals with HCM who do **not** have a resting or provocable outflow tract gradient ≥50 mm Hg on TTE.

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- Stress echo can be repeated when there is documentation of **any** of the following:
  - In 1 to 2 years if the resting or provocable outflow tract gradient is <30 mm Hg on prior stress echo
  - Worsening symptoms
  - There has been a therapeutic change (e.g., change in medication, surgical procedure performed).

Cardiac CTA (CPT® 75574)

- Initial imaging study in individuals with hypertrophic cardiomyopathy and stable anginal symptoms
  - Chest discomfort is common in individuals with hypertrophic cardiomyopathy. The incidence of false positive myocardial perfusion imaging abnormalities is higher in these individuals, whereas the incidence of severe coronary artery stenosis is low.

Cardiac MRI (CMR)

- Cardiac MRI (CPT® 75557 or CPT® 75561) for assessment of global ventricular function, myocardial composition, and mass if a specific clinical question is left unanswered by a recent echocardiogram and results will affect patient management.

Left heart catheterization with coronary arteriography

- Left heart catheterization with coronary arteriography (CPT® 93458 or 93454) is medically necessary to rule out coronary artery disease prior to planned surgical myectomy for hypertrophic cardiomyopathy.

Surveillance imaging

- TTE is medically necessary every year when there is no change in clinical status or treatment.

Monitoring treatment

Repeat TTE (CPT® 93306) is medically necessary in individuals with Obstructive Hypertrophic Cardiomyopathy (HCM) for the following:

Mavacamten for obstructive hypertrophic cardiomyopathy

Initiation of treatment

- Baseline-at the beginning of treatment
- 4 weeks after treatment initiation
- 8 weeks after treatment initiation
- 12 weeks after treatment initiation
- Then every 12 weeks while on mavacamten

Changes in treatment

- 4 weeks after any interruption of treatment (any missed dose)

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- After any dosage change (including restart of treatment):
  - 4 weeks after dosage change
  - 12 weeks after dosage change
- After initiating a weak CYP2C19 inhibitor (e.g., omeprazole) or moderate CYP3A4 inhibitor (e.g., ciprofloxacin):
  - 4 weeks after start of medication
  - 12 weeks after start of medication
- At any time regardless of timing of prior echo when there are new cardiac signs or symptoms, or worsening of clinical status

Post- Septal Reduction Therapy (SRT)

TTE is medically necessary within 3 to 6 months after SRT (surgical myectomy or alcohol septal ablation) to evaluate the procedural results in individuals with hypertrophic cardiomyopathy

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General Information

Guideline

General Guidelines (CRID-1.0)
Procedure Codes Addressed in this Guideline

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General Guidelines (CRID-1.0)

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Terms used in this guideline

Abbreviations

ACE inhibitor	Angiotensin-converting enzyme inhibitor
AMI	Acute myocardial infarction
ARVC	Arrhythmogenic right ventricular cardiomyopathy
AV	Atrioventricular
CC	Complications/comorbid conditions
CHF	Congestive heart failure
CM	Cardiomyopathy
CRT	Cardiac resynchronization therapy
EP	Electrophysiology
GDMT	Guideline-directed medical therapy
HCM	Hypertrophic cardiomyopathy
HFpEF	Heart Failure with Preserved Ejection Fraction
HFrEF	Heart Failure with Reduced Ejection Fraction
ICD	Implantable cardioverter defibrillator
LBBB	Left bundle branch block

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LQT	Long QT syndrome
LV	Left ventricle
LVEF	Left ventricular ejection fraction
MCC	Major complications/comorbid conditions
MI	Myocardial infarction
NCCM	Non-compaction cardiomyopathy
NYHA	New York Heart Association functional classification
OMT	Optimal medical therapy
RBBB	Right bundle branch block
RV	Right ventricle
SCA/SCD	Sudden cardiac arrest/Sudden cardiac death
TAVI	Transcatheter aortic valve implantation
TAVR	Transcatheter aortic valve replacement
VF	Ventricular fibrillation
VT	Ventricular tachycardia

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Definitions

NYHA Heart Failure Definitions	**class I** - No limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, or dyspnea. **class II** - Slight limitation of physical activity. Comfortable at rest. Ordinary physical activity results in fatigue, palpitation, or dyspnea. **class III** - Marked limitation of physical activity. Comfortable at rest. Less than ordinary activity causes fatigue, palpitation, or dyspnea. **class IV** - Unable to carry on any physical activity without discomfort. Symptoms of heart failure may be present even at rest. If any physical activity is undertaken, discomfort is increased.
Abnormal blood pressure response to exercise	Flat response/failure to augment; rise then fall during exercise; vasoactive cardiovascular drugs may result in an abnormal blood pressure response to exercise
Ambulatory class IV CHF	Class IV heart failure with: 1) no active acute coronary syndrome; 2) no inotropes; and 3) on OMT
Cardiomyopathy	A disease that affects the heart muscle, making it harder for the heart to pump blood to the rest of the body. It is further defined based on the type of change in the heart muscle including: • hypertrophic - muscle becomes thicker • dilated -muscle becomes weaker and stretched out • nondilated - muscle develops scarring or abnormal tissue • restrictive - muscle becomes stiff and less flexible

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Hypertrophic cardiomyopathy	Hypertrophic Cardiomyopathy (HCM) is a clinical diagnosis, established by imaging with 2D echocardiography or cardiovascular magnetic resonance (CMR) showing a maximal end-diastolic wall thickness of ≥15 mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults. More limited hypertrophy (13–14 mm) can be diagnostic, particularly when present in family members of a patient with HCM or in conjunction with a positive genetic test, and/or associated with typical dynamic outflow obstruction, or distinctly abnormal ECG patterns.
Incessant VT	Frequent recurrences of ongoing hemodynamically stable VT
Ischemic Cardiomyopathy	Decreased ability of the heart to pump blood properly caused by damage or scarring of the heart muscle from reduced blood supply to the heart (i.e., myocardial infarction or coronary artery disease)
Long QT Syndrome (LQTS)	A congenital disorder characterized by a prolongation of the QT interval on ECG and a propensity to ventricular tachyarrhythmias, which may lead to syncope, cardiac arrest, or sudden death. The QT interval on the ECG, measured from the beginning of the QRS complex to the end of the T wave, represents the duration of activation and recovery of the ventricular myocardium. QT intervals corrected for heart rate (QTc) longer than 0.44 seconds are generally considered abnormal, though a normal QTc can be more prolonged in females (up to 0.46 sec).The Bazett formula is the formula most commonly used to calculate the QTc, as follows: QTc = AT/square root of the R-R interval (in seconds).
Non-Compaction Cardiomyopathy	A rare congenital cardiomyopathy that affects children and adults. It results from the failure of myocardial development during embryogenesis. It is also called spongiform cardiomyopathy. Symptoms are often a result of a poor pumping performance by the heart. The disease can be associated with other problems with the heart and the body.

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Non-Sustained Ventricular Tachycardia (NSVT)	Three or more consecutive ventricular beats at a rate of greater than 120 beats/min with a duration of less than 30 seconds
Nonischemic Cardiomyopathy	Decreased ability of the heart to pump blood properly not caused by coronary artery disease or ischemic injury
Optimal Medical Therapy (OMT)	Optimal medical therapy for heart failure includes 3 months of **all** of the following: • Beta-blocker • Mineralocorticoid antagonist (MRA) • One of the following: ◦ ACE inhibitor ◦ angiotensin II receptor blocker ◦ angiotensin receptor-neprilysin inhibitor **Note:** SGLT2i treatment improves prognosis in heart failure. Due to concerns over patient access, cost, and patient tolerance, although recommended for heart failure, the use is not mandatory to meet OMT. If OMT is not tolerated due to documented adverse effects (hypotension, renal dysfunction, or electrolyte imbalances) this does not preclude ICD eligibility.
Structural Heart Disease	A structural or functional abnormality of the heart, or of the blood vessels supplying the heart, that impairs its normal functioning.
Sudden Cardiac Arrest (SCA)	The abrupt loss of all heart activity
TAVR (TAVI)	A minimally invasive procedure to treat aortic valve stenosis
Wolff-Parkinson-White (WPW) syndrome	A cardiac condition where an accessory pathway allows electrical conduction between the heart chambers outside of the normal AV node route and creates a re-entry circuit, which can lead to supraventricular tachycardia (SVT), atrial fibrillation (AF), ventricular fibrillation (VF), and sudden cardiac death.

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Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

General requirements

Current clinical information, which may include history, physical examination, symptoms, laboratory results, and imaging reports, are necessary for determining the medical necessity of implantable cardiac devices.

- The information provided should have clinical relevance to the request.
- If the information provided makes no reference to the potential indication for the request, then the medical necessity for the procedure(s) cannot be supported.
- Requests for a device when a same or similar device has already been placed is not supported without clear documentation that fulfills guideline criteria.

Procedures should be requested after initial consultation and physician treatment planning that includes full counseling of the individual with shared decision-making.

Removal and replacement (CRID-1.2)

- Generator replacement (CPT® 33212, 33213, 33221, 33227, 33228, 33229, 33230, 33231, 33240, 33262, 33263, 33264, 0614T, 0801T, 0802T, 0803T) with a same or similar device is medically necessary when:
  - Interrogation shows device is nearing Elective Replacement Indicator (ERI) or End of Life (EOL).
  - Interrogation report documents the device is not functioning correctly and requires replacement.

Evidence Discussion

The decision to replace a cardiac implantable device at elective replacement indicator is shared between the individual with the implanted device and the treating physician. Because clinical situations may change, indications should be reconsidered based on the most recent guidelines to evaluate for continuance of therapy or changing the type

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of device. For example, currently, there insufficient data regarding ICD replacement in individuals who have received a primary prevention ICD, but have not experienced a clinically relevant arrhythmia since the device was placed.[1] Clinical situations and an individuals understanding of their options may change. It is essential in the shared-decision process to ensure the individual is aware of any change in clinical status or change in evidence that may impact the decision to replace a cardiac device.

[1] Russo, A, Desai, M, Do, M. et al. ACC/AHA/ASE/HFSA/HRS/SCAI/SCCT/SCMR 2025 Appropriate Use Criteria for Implantable Cardioverter-Defibrillators, Cardiac Resynchronization Therapy, and Pacing. *JACC.* 2025 Mar, 85 (11) 1213–1285.

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Procedure Codes Addressed in this Guideline

CID.AD.101.A
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Procedure description	CPT®
Insertion of new or replacement of permanent pacemaker with transvenous electrode(s); atrial	33206
Insertion of new or replacement of permanent pacemaker with transvenous electrode(s); ventricular	33207
Insertion of new or replacement of permanent pacemaker with transvenous electrode(s); atrial and ventricular	33208
Insertion of pacemaker pulse generator only; single existing single lead	33212
Insertion of pacemaker pulse generator only; with existing dual leads	33213
Upgrade of implanted pacemaker system, conversion of single chamber system to dual chamber system (includes removal of previously placed pulse generator, testing of existing lead, insertion of new lead, insertion of new pulse generator)	33214
Insertion of pacemaker pulse generator only; with existing multiple leads	33221
Insertion of pacing electrode, cardiac venous system, for left ventricular pacing, with attachment to previously placed pacemaker or pacing cardioverter-defibrillator pulse generator	33224
Insertion of pacing electrode, cardiac venous system, for left ventricular pacing, at time of insertion of pacing cardioverter-defibrillator pulse generator (including upgrade to dual chamber system and pocket revision)	33225
Removal of permanent pacemaker pulse generator with replacement of pacemaker pulse generator; single lead system	33227
Removal of permanent pacemaker pulse generator with replacement of pacemaker pulse generator; dual lead system	33228
Removal of permanent pacemaker pulse generator with replacement of pacemaker pulse generator; multiple lead system	33229

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Procedure description	CPT®
Insertion of pacing cardioverter-defibrillator pulse generator only; with existing dual leads	33230
Insertion of pacing cardioverter-defibrillator pulse generator only; with existing multiple leads	33231
Insertion of pacing cardioverter-defibrillator pulse generator only; with existing single leads	33240
Insertion or replacement of permanent pacing cardioverter-defibrillator system with transvenous lead(s), single or dual chamber	33249
Removal of pacing cardioverter-defibrillator pulse generator with replacement of pacing cardioverter-defibrillator pulse generator; single lead system	33262
Removal of pacing cardioverter-defibrillator pulse generator with replacement of pacing cardioverter-defibrillator pulse generator; dual lead system	33263
Removal of pacing cardioverter-defibrillator pulse generator with replacement of pacing cardioverter-defibrillator pulse generator; multiple lead system	33264
Insertion or replacement of permanent subcutaneous implantable defibrillator system, with subcutaneous electrode, including defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters when performed	33270
Transcatheter insertion or replacement of permanent leadless pacemaker, right ventricular, including imaging guidance (e.g., fluoroscopy, venous ultrasound, ventriculography, femoral venography) and device evaluation (e.g., interrogation or programming), when performed	33274
Transcatheter implantation of wireless pulmonary artery pressure sensor for long-term hemodynamic monitoring, including deployment and calibration of the sensor, right heart catheterization, selective pulmonary catheterization, radiological supervision and interpretation, and pulmonary artery angiography, when performed	33289

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Procedure description	CPT®
Initial open implantation of baroreflex activation therapy (BAT) modulation system, including lead placement onto the carotid sinus, lead tunneling, connection to a pulse generator placed in a distant subcutaneous pocket (ie, total system), and intraoperative interrogation and programming	64654
Insertion or replacement of permanent cardiac contractility modulation system, including contractility evaluation when performed, and programming of sensing and therapeutic parameters; pulse generator with transvenous electrodes	0408T
Insertion or replacement of permanent cardiac contractility modulation system, including contractility evaluation when performed, and programming of sensing and therapeutic parameters; pulse generator only	0409T
Insertion of wireless cardiac stimulator for left ventricular pacing, including device interrogation and programming, and imaging supervision and interpretation, when performed; complete system (includes electrode and generator [transmitter and battery])	0515T
Insertion of wireless cardiac stimulator for left ventricular pacing, including device interrogation and programming, and imaging supervision and interpretation, when performed; electrode only	0516T
Insertion of wireless cardiac stimulator for left ventricular pacing, including device interrogation and programming, and imaging supervision and interpretation, when performed; pulse generator component(s) (battery and/or transmitter) only	0517T
Removal and replacement of wireless cardiac stimulator for left ventricular pacing; pulse generator component(s) (battery and/or transmitter)	0519T
Removal and replacement of wireless cardiac stimulator for left ventricular pacing; pulse generator component(s) (battery and/or transmitter), including placement of a new electrode	0520T
Insertion or replacement of implantable cardioverter-defibrillator system with substernal electrode(s), including all imaging guidance and electrophysiological evaluation (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters), when performed	0571T

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Procedure description	CPT®
Removal and replacement of substernal implantable defibrillator pulse generator	0614T
Transcatheter insertion of permanent dual-chamber leadless pacemaker, including imaging guidance (e.g., fluoroscopy, venous ultrasound, right atrial angiography, right ventriculography, femoral venography) and device evaluation (e.g., interrogation or programming), when performed; complete system (i.e., right atrial and right ventricular pacemaker components)	0795T
Transcatheter insertion of right atrial pacemaker component (when an existing right ventricular single leadless pacemaker exists to create a dual-chamber leadless pacemaker system)	0796T
Transcatheter insertion of right ventricular pacemaker component (when part of a dual-chamber leadless pacemaker system)	0797T
Transcatheter removal and replacement of permanent dual-chamber leadless pacemaker, including imaging#guidance (e.g., fluoroscopy, venous ultrasound, right atrial angiography, right ventriculography, femoral venography) and device evaluation (e.g., interrogation or programming), when performed; dual-chamber system (i.e., right atrial and right ventricular pacemaker components)	0801T
Transcatheter removal and replacement of right atrial pacemaker component	0802T
Transcatheter removal and replacement of right ventricular pacemaker component (when part of a dual-chamber leadless pacemaker system)	0803T
Transcatheter insertion of permanent single-chamber leadless pacemaker, right atrial, including imaging guidance (e.g., fluoroscopy, venous ultrasound, right atrial angiography and/or right ventriculography, femoral venography, cavography) and device evaluation (e.g., interrogation or programming), when performed	0823T
Transcatheter removal and replacement of permanent single-chamber leadless pacemaker, right atrial, including imaging guidance (e.g., fluoroscopy, venous ultrasound, right atrial angiography and/or right ventriculography, femoral venography, cavography) and device evaluation (e.g., interrogation or programming), when performed	0825T

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Procedure description	CPT®
Insertion of permanent cardiac contractility modulation-defibrillation system component(s), including fluoroscopic guidance, and evaluation and programming of sensing and therapeutic parameters; pulse generator and dual transvenous electrodes/leads (pacing and defibrillation)	0915T
Insertion of permanent cardiac contractility modulation-defibrillation system component pulse generator only	0916T
Removal and replacement of permanent cardiac contractility modulation defibrillation pulse generator only	0923T
Transcatheter implantation of wireless left atrial pressure sensor for long-term left atrial pressure monitoring, including sensor calibration and deployment, right heart catheterization, transseptal puncture, imaging guidance, and radiological supervision and interpretation	0933T

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Pacemaker Devices

Guideline

Permanent Pacemaker Implantation (CRID-7)
References
Leadless Pacemaker (CRID-11.1)
References

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Permanent Pacemaker Implantation (CRID-7)

CID.PM.107.A
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Codes included

Description	CPT®
Insertion of new or replacement of permanent pacemaker with transvenous electrode(s); atrial	33206
Insertion of new or replacement of permanent pacemaker with transvenous electrode(s); ventricular	33207
Insertion of new or replacement of permanent pacemaker with transvenous electrode(s); atrial and ventricular	33208
Upgrade of implanted pacemaker system, conversion of single chamber system to dual chamber system (includes removal of previously placed pulse generator, testing of existing lead, insertion of new lead, insertion of new pulse generator)	33214

Indications for Permanent Pacemaker (CRID-7.1)

Sinus node dysfunction

Permanent pacemaker implantation is medically necessary for any of the following:

- Symptomatic non-reversible sinus node dysfunction as evidenced by **both** of the following:
  - Documented sinus node dysfunction including one of the below:
    - Sinus bradycardia at rate less than 50 beats per minute
    - Sinus pauses greater than 3 seconds
  - Symptoms attributable to non-reversible sinus node dysfunction including **one** of the below:
    - Syncope or pre-syncope
    - Heart failure symptoms
    - Exertional fatigue and impaired exercise tolerance

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- Sinus bradycardia at rate less than 40 beats per minute and symptoms possibly related to bradycardia
- Symptomatic sinus bradycardia (as defined above) is a consequence of essential medical management and continued treatment is clinically necessary
- Symptoms attributable to bradycardia as listed above and evidence of tachy-brady syndrome (sinus bradycardia, ectopic atrial bradycardia, or sinus pause alternating with periods of atrial flutter or atrial fibrillation)
- Symptomatic chronotropic incompetence defined as limitations in activity due to the inability of the heart to increase its rate during exercise to achieve 80% of maximum predicted heart rate (220-age).

Atrioventricular block (AVB)

Permanent pacemaker implantation is medically necessary for any of the following:

- AVB including one of the below, with or without symptoms:
  - Second-degree Mobitz type II
  - High-grade (greater than or equal to 2 consecutive P waves at a constant physiologic rate that do not conduct to the ventricles)
  - Third-degree (complete heart block)
- Any degree of AVB with one of the following symptoms that are clearly attributable to the AVB:
  - Syncope or pre-syncope
  - Heart failure symptoms
  - Exertional fatigue and impaired exercise tolerance
- Third-degree and advanced second-degree AV block at any anatomic level associated with sustained or non-sustained ventricular tachycardia (ventricular rhythm at rate greater than 100 bpm lasting greater than or equal to 3 consecutive beats) presumed due to AV block
- Marked first-degree AVB (PR interval greater than or equal to 0.3 seconds) or second-degree AVB with symptoms similar to those of pacemaker syndrome
- Symptomatic AVB as a consequence of guideline directed management and continued treatment is clinically necessary
- Persistent or permanent atrial fibrillation and symptomatic bradycardia including one of the following:
  - Rate less than 50 beats per minute
  - Regular QRS intervals indicating complete atrioventricular block (AVB)
- Second degree AV block with a documented pause of greater than or equal to 5 seconds during waking in the presence of atrial fibrillation, with or without symptoms
- Second degree AV block with documented periods of asystole greater than or equal to 3.0 seconds in the presence of sinus rhythm, with or without symptoms

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- Second-degree AVB noted to be located at intra-His or infra-His levels at electrophysiology study (EPS), with or without symptoms
- Any AVB indication listed above occurring after acute myocardial infarction that does not resolve within 5 days
- Congenital complete or high-degree AVB in the presence of any of the following:
  - Symptoms related to bradycardia such as syncope, pre-syncope, heart failure symptoms, exertional fatigue, or impaired exercise tolerance
  - Wide QRS escape rhythm
  - Mean daytime heart rate below 50 bpm
  - Pauses greater than 3 times the cycle length of the ventricular escape rhythm
  - Complex ventricular ectopy
  - Prolonged QT interval
  - Ventricular dysfunction, ventricular dilatation or significant mitral regurgitation

Conduction Disorders with 1:1 Atrioventricular Conduction

Permanent pacemaker implantation is medically necessary for any of the following:

- Individuals with syncope and bundle branch block and one of the following at electrophysiology study (EPS):
  - Baseline HV interval greater than or equal to 70 ms
  - Second- or third-degree intra- or infra-Hisian block during incremental atrial pacing
- Alternating bundle branch block, with or without symptoms
- HV interval greater than or equal to 100 milliseconds noted at EPS, with or without symptoms
- Intra-Hisian- or infra-Hisian block noted at EPS, with or without symptoms

Recurrent syncope

Permanent pacemaker implantation is medically necessary for individuals with recurrent syncope and any of the following:

- Spontaneous documented symptomatic asystolic pause greater than 3 seconds due to sinus arrest or atrioventricular block (AVB)
- Spontaneous documented asymptomatic asystolic pause greater than 6 seconds due to sinus arrest or AVB
- Cardioinhibitory carotid sinus syndrome as documented by one of the below:
  - Syncope caused by spontaneously occurring carotid sinus stimulation
  - Carotid sinus pressure that induces syncope and/or ventricular asystole of greater than or equal to 3 seconds
- Syncope associated with asystole of greater than or equal to 3 seconds during tilt testing

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- Bundle branch block and one of the following at electrophysiology study (EPS):
  - Baseline HV interval greater than or equal to 70 ms
  - Second- or third-degree intra- or infra-Hisian block during incremental atrial pacing
- Syncope after cardiac transplantation with or without documentation of bradyarrhythmia
- Recurrent Adam-Stokes syndrome

Peri-procedural and post-operative indications

Permanent pacemaker implantation is medically necessary for any of the following:

- Prior to a planned catheter ablation of the atrioventricular (AV) junction for one of the following:
  - Rate control strategy for management of atrial fibrillation
  - Supraventricular tachycardia resulting in tachycardia induced cardiomyopathy that is not controlled with ablation or medical therapy
- Post Transcatheter Aortic Valve Implantation (TAVI) for any of the following:
  - Complete or high-degree atrioventricular block (AVB) that persists for 24 to 48 hours after TAVI
  - New-onset alternating bundle branch block after TAVI
  - Pre-existing right bundle branch block (RBBB) and new conduction abnormality onset during or after (TAVI) such as:
    - Transient high-degree AVB
    - PR prolongation
    - QRS axis change
- Sinus node dysfunction or AVB associated with symptoms or hemodynamic instability occurring after cardiac surgery that does not resolve within 5 days
- Post cardiac transplant for any of the following:
  - Relative bradycardia that is prolonged or recurrent, which limits rehabilitation or discharge after postoperative recovery
  - Syncope with or without documentation of bradyarrhythmia

Neuromuscular diseases known to involve the heart

Permanent pacemaker implantation is considered medically necessary for progressive neuromuscular diseases known to involve the heart with any degree of atrioventricular (AV) block including first degree AV block or any fascicular block, with or without symptoms, because there may be unpredictable progression of AV conduction disease. Progressive neuromuscular diseases known to involve the heart include:

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- Myotonic muscular dystrophy
- Kearns-Sayre syndrome
- Erb dystrophy (limb-girdle muscular dystrophy)
- Peroneal muscular atrophy

Single Chamber Pacemaker Upgrade to Dual Chamber Pacemaker

Upgrade of a single-chamber ventricular pacemaker (VVI) to dual-chamber pacemaker (DDD) is medically necessary when **any** of the following conditions are present:

- Sinus node dysfunction, where atrial pacing is required to maintain chronotropic competence
- Atrioventricular (AV) conduction abnormalities, including first-degree, second-degree (Mobitz I or II), high-grade AV block, or complete heart block, where synchronized atrioventricular pacing is anticipated to improve hemodynamic performance
- Symptoms consistent with pacemaker syndrome (fatigue, hypotension, or syncope resulting from the loss of AV synchrony)

Permanent Pacemaker Implantation - Non-indications (CRID-7.2)

- Permanent pacemaker implantation is **not** medically necessary in any of the following settings:
  - Sinus node dysfunction when there is documentation of any of the following
    - Individual is asymptomatic
    - The symptoms suggestive of bradycardia have been clearly documented to occur in the absence of bradycardia
    - Sinus node dysfunction is due to nonessential drug therapy
  - Fascicular block without AV block or without symptoms concerning for AV block
  - Incidentally noted hypersensitive cardioinhibitory response to carotid sinus stimulation when the individual remains asymptomatic or has vague symptoms
  - Asymptomatic first-degree AV block
  - Asymptomatic type-1 second-degree AV block at the supra-His (AV node) level or that which is not known to be intra- or infra-Hisian
  - Asymptomatic transient AV block in the absence of intraventricular conduction defects or in isolated single fascicular block
  - Situational vasovagal syncope when avoidance behavior is effectively preventing syncopal episodes
  - Prior to Transcatheter Aortic Valve Replacement (TAVR) as a prophylactic measure in individuals with right bundle branch block (RBBB) when there is no indication for permanent pacing
  - For the purpose of cardiac contractility modulation (CCM)

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- Upgrade of a single chamber pacemaker (VVI) to a dual chamber pacemaker (DDD) is **not** medically necessary for the indication of persistent atrial fibrillation when it is **not** part of a planned rhythm control strategy.

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Leadless Pacemaker (CRID-11.1)

CID.PM.111.A
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Codes included

Description	CPT®
Transcatheter insertion of permanent dual-chamber leadless pacemaker, including imaging guidance (e.g., fluoroscopy, venous ultrasound, right atrial angiography, right ventriculography, femoral venography) and device evaluation (e.g., interrogation or programming), when performed; complete system (i.e., right atrial and right ventricular pacemaker components)	0795T
Transcatheter insertion of right atrial pacemaker component (when an existing right ventricular single leadless pacemaker exists to create a dual-chamber leadless pacemaker system)	0796T
Transcatheter insertion of right ventricular pacemaker component (when part of a dual-chamber leadless pacemaker system)	0797T
Transcatheter insertion of permanent single-chamber leadless pacemaker, right atrial, including imaging guidance (e.g., fluoroscopy, venous ultrasound, right atrial angiography and/or right ventriculography, femoral venography, cavography) and device evaluation (e.g., interrogation or programming), when performed	0823T
Transcatheter insertion or replacement of permanent leadless pacemaker, right ventricular, including imaging guidance (e.g., fluoroscopy, venous ultrasound, ventriculography, femoral venography) and device evaluation (e.g., interrogation or programming), when performed	33274

Indications for Leadless Pacemaker

Leadless right ventricular pacemaker (CRID-11.1.1)

CPT® 33274

Permanent right ventricular leadless pacemaker implant is medically necessary when **both** of the following are met:

- The individual meets **any** of the following indications for leadless right ventricular pacemaker:

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  - Symptomatic paroxysmal or permanent high-grade AV block in the presence of Atrial Fibrillation (AF)
  - Symptomatic paroxysmal or permanent high-grade AV block in the absence of AF, as an alternative to dual chamber pacing, when atrial lead placement is considered difficult, high risk, or not deemed necessary for effective therapy
  - Symptomatic bradycardia-tachycardia syndrome or sinus node dysfunction (sinus bradycardia or sinus pauses), as an alternative to atrial or dual chamber pacing, when atrial lead placement is considered difficult, high risk, or not deemed necessary for effective therapy
- The following contraindications for leadless pacemaker are **not** present:
  - An implanted inferior vena cava filter
  - A mechanical tricuspid valve

Leadless dual chamber pacemaker system (CRID-11.1.2)

CPT® 0795T

A leadless dual chamber pacemaker is considered not medically necessary when either of the following **contraindications** is present:

- An implanted inferior vena cava filter
- A mechanical tricuspid valve

Permanent dual chamber leadless pacemaker implant is medically necessary in an individual with no contraindications to a leadless pacemaker, when one of the following indications for a permanent dual chamber pacemaker is documented:

Sinus node dysfunction

Permanent pacemaker implantation is medically necessary for any of the following:

- Symptomatic non-reversible sinus node dysfunction as evidenced by **both** of the following:
  - Documented sinus node dysfunction including one of the below:
    - Sinus bradycardia at rate less than 50 beats per minute
    - Sinus pauses greater than 3 seconds
  - Symptoms attributable to non-reversible sinus node dysfunction including **one** of the below:
    - Syncope or pre-syncope
    - Heart failure symptoms
    - Exertional fatigue and impaired exercise tolerance
- Sinus bradycardia at rate less than 40 beats per minute and symptoms possibly related to bradycardia
- Symptomatic sinus bradycardia (as defined above) is a consequence of essential medical management and continued treatment is clinically necessary

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- Symptoms attributable to bradycardia as listed above and evidence of tachy-brady syndrome (sinus bradycardia, ectopic atrial bradycardia, or sinus pause alternating with periods of atrial flutter or atrial fibrillation)
- Symptomatic chronotropic incompetence defined as limitations in activity due to the inability of the heart to increase its rate during exercise to achieve 80% of maximum predicted heart rate (220-age).

Atrioventricular block (AVB)

Permanent pacemaker implantation is medically necessary for any of the following:

- AVB including one of the below, with or without symptoms:
  - Second-degree Mobitz type II
  - High-grade (greater than or equal to 2 consecutive P waves at a constant physiologic rate that do not conduct to the ventricles)
  - Third-degree (complete heart block)
- Any degree of AVB with one of the following symptoms that are clearly attributable to the AVB:
  - Syncope or pre-syncope
  - Heart failure symptoms
  - Exertional fatigue and impaired exercise tolerance
- Third-degree and advanced second-degree AV block at any anatomic level associated with sustained or non-sustained ventricular tachycardia (ventricular rhythm at rate greater than 100 bpm lasting greater than or equal to 3 consecutive beats) presumed due to AV block
- Marked first-degree AVB (PR interval greater than or equal to 0.3 seconds) or second-degree AVB with symptoms similar to those of pacemaker syndrome
- Symptomatic AVB as a consequence of guideline directed management and continued treatment is clinically necessary
- When part of a planned rhythm control strategy, persistent atrial fibrillation with symptomatic bradycardia including **either** of the following:
  - Rate less than 50 beats per minute
  - Regular QRS intervals indicating complete atrioventricular block (AVB)
- Second degree AV block with a documented pause of greater than or equal to 5 seconds during waking in the presence of atrial fibrillation, with or without symptoms
- Second degree AV block with documented periods of asystole greater than or equal to 3.0 seconds in the presence of sinus rhythm, with or without symptoms
- Second-degree AVB noted to be located at intra- or infra-His levels at electrophysiology study (EPS), with or without symptoms
- Any AVB indication listed above occurring after acute myocardial infarction that does not resolve within 5 days

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- Congenital complete or high-degree AVB in the presence of any of the following:
  - Symptoms related to bradycardia such as syncope, pre-syncope, heart failure symptoms, exertional fatigue, or impaired exercise tolerance
  - Wide QRS escape rhythm
  - Mean daytime heart rate below 50 bpm
  - Pauses greater than 3 times the cycle length of the ventricular escape rhythm
  - Complex ventricular ectopy
  - Prolonged QT interval
  - Ventricular dysfunction, ventricular dilatation or significant mitral regurgitation

Conduction Disorders with 1:1 Atrioventricular Conduction

Permanent pacemaker implantation is medically necessary for any of the following:

- Individuals with syncope and bundle branch block and one of the following at electrophysiology study (EPS):
  - Baseline HV interval greater than or equal to 70 ms
  - Second- or third-degree intra- or infra-Hisian block during incremental atrial pacing
- Alternating bundle branch block, with or without symptoms
- HV interval greater than or equal to 100 milliseconds noted at EPS, with or without symptoms
- Intra-Hisian- or infra-Hisian block noted at EPS, with or without symptoms

Recurrent syncope

Permanent pacemaker implantation is medically necessary for individuals with recurrent syncope and any of the following:

- Spontaneous documented symptomatic asystolic pause greater than 3 seconds due to sinus arrest or atrioventricular block (AVB)
- Spontaneous documented asymptomatic asystolic pause greater than 6 seconds due to sinus arrest or AVB
- Cardioinhibitory carotid sinus syndrome as documented by one of the below:
  - Syncope caused by spontaneously occurring carotid sinus stimulation
  - Carotid sinus pressure that induces syncope and/or ventricular asystole of greater than or equal to 3 seconds
- Syncope associated with asystole of greater than or equal to 3 seconds during tilt testing
- Bundle branch block and one of the following at electrophysiology study (EPS):
  - Baseline HV interval greater than or equal to 70 ms

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  - Second- or third-degree intra- or infra-Hisian block during incremental atrial pacing
- Syncope after cardiac transplantation with or without documentation of bradyarrhythmia
- Recurrent Adam-Stokes syndrome

Peri-procedural and post-operative indications

Permanent pacemaker implantation is medically necessary for any of the following:

- Prior to a planned catheter ablation of the atrioventricular (AV) junction for supraventricular tachycardia resulting in tachycardia induced cardiomyopathy that is not controlled with ablation or medical therapy
- Post Transcatheter Aortic Valve Implantation (TAVI) for any of the following:
  - Complete or high-degree atrioventricular block (AVB) that persists for 24 to 48 hours after TAVI
  - New-onset alternating bundle branch block after TAVI
  - Pre-existing right bundle branch block (RBBB) and new conduction abnormality onset during or after (TAVI) such as:
    - Transient high-degree AVB
    - PR prolongation
    - QRS axis change
- Sinus node dysfunction or AVB associated with symptoms or hemodynamic instability occurring after cardiac surgery that does not resolve within 5 days
- Post cardiac transplant for any of the following:
  - Relative bradycardia that is prolonged or recurrent, which limits rehabilitation or discharge after postoperative recovery
  - Syncope with or without documentation of bradyarrhythmia

Neuromuscular diseases known to involve the heart

Permanent pacemaker implantation is considered medically necessary for progressive neuromuscular diseases known to involve the heart with any degree of atrioventricular (AV) block including first degree AV block or any fascicular block, with or without symptoms, because there may be unpredictable progression of AV conduction disease. Progressive neuromuscular diseases known to involve the heart include:

- Myotonic muscular dystrophy
- Kearns-Sayre syndrome
- Erb dystrophy (limb-girdle muscular dystrophy)
- Peroneal muscular atrophy

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Leadless right atrial pacemaker (CRID-11.1.3)

CPT® 0823T

Permanent leadless right atrial pacemaker implant is medically necessary when **both** of the following are met:

- Symptomatic sinus node dysfunction (dizziness, syncope, shortness of breath) with normal AV and intraventricular conduction systems
- The following contraindications for leadless pacemaker are not present:
  - An implanted inferior vena cava filter
  - A mechanical tricuspid valve

General information

Right ventricular leadless pacemaker

The permanent right ventricular leadless pacemakers (CPT® 33274) consist of a single leadless device implanted directly into the right ventricle. The Medtronic Micra™ VR and Abbott Aveir™ VR right ventricular leadless pacemakers are capable only of VVI and VVIR pacing. The Medtronic Micra™ AV right ventricular leadless pacemaker is also capable of VDD pacing. The right ventricular leadless pacemakers do not have capability for atrial pacing. The estimated battery life is about 10 years

Dual chamber leadless pacemaker

In contrast to the right ventricular leadless pacemakers referred to above, the dual chamber leadless pacemaker (i.e., Abbott Aveir™ DR leadless pacemaker system) has dual-chamber sensing and pacing functionality. The Abbott Aveir™ DR leadless pacemaker system consists of two separate components: one implanted in the right atrium and the other in the right ventricle.

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Implantable Cardioverter-Defibrillator (ICD) Devices

Guideline

Indications for ICD implantation (CRID-2)
References
Pediatric Implantable Cardioverter Defibrillator
References

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Indications for ICD implantation (CRID-2)

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Codes included

Description	CPT®
Insertion or replacement of permanent pacing cardioverter-defibrillator system with transvenous lead(s), single or dual chamber	33249
Insertion or replacement of permanent subcutaneous implantable defibrillator system, with subcutaneous electrode, including defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters when performed	33270

Secondary Prevention ICD indications (CRID-2.1)

Cardiac Arrest Survivors (CRID-2.1.1)

ICD implantation is medically necessary in a survivor of cardiac arrest in the setting of any of the following conditions:

- Cardiac arrest from hemodynamically unstable or sustained VT, polymorphic VT, or VF associated with an acute (less than 48 Hours) MI with **both** of the following conditions:
  - Obstructive CAD with coronary anatomy not amenable to revascularization
  - Ejection fraction less than or equal to 35%
- Cardiac arrest from hemodynamically unstable or sustained VT, polymorphic VT, or VF associated with acute (<48 hours) post-coronary revascularization when **both** of the following conditions exist:
  - No evidence for acute coronary occlusion, restenosis, acute infarct, or other clearly reversible cause
  - Ejection fraction is less than or equal to 35%
- Cardiac arrest from hemodynamically unstable or sustained VT, polymorphic VT, or VF associated with coronary artery disease with **both** of the following conditions:
  - No recent MI (less than or equal to 40 days) prior to arrest
  - No recent coronary revascularization (less than or equal to 3 months) prior to arrest

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- Cardiac arrest from hemodynamically unstable or sustained VT, polymorphic VT, or VF during exercise testing when cardiac catheterization performed after the arrest results in **any** of the following:
  - Significant CAD not amenable to revascularization
  - Significant CAD identified and complete revascularization performed
  - Significant CAD identified and incomplete revascularization performed
- Cardiac arrest from hemodynamically unstable or sustained VT, polymorphic VT, or VF not associated with coronary artery disease in the setting of **any** of the following conditions:
  - Non-ischemic dilated cardiomyopathy
  - Myocardial sarcoidosis
  - Giant-cell myocarditis
- Cardiac arrest with idiopathic VF and a normal Ejection Fraction (greater than or equal to 50%) when no reversible causes have been identified including **all** of the following:
  - Pharmacologically induced sustained VT/VF
  - Sustained VT/VF with hypokalemia
  - Wolff-Parkinson-White (WPW) syndrome with VT/VF and successfully ablated pathway

Sustained VT/VF (CRID-2.1.2)

ICD implantation is medically necessary to treat an individual with sustained VT or VF and either of the following conditions:

- Genetic diseases associated with sustained VT/VF and a normal Ejection Fraction (greater than or equal to 50%) including any of the following:
  - Congenital long QT syndrome
  - Short QT syndrome
  - Catecholaminergic polymorphic VT
  - Brugada syndrome
  - Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)
  - Hypertrophic cardiomyopathy
- Sustained hemodynamically stable monomorphic VT associated with structural heart disease (coronary artery disease with prior MI or non-ischemic dilated cardiomyopathy) and all of the following are met:
  - All inducible VT's are successfully ablated
  - Ejection fraction is less than or equal to 35%
  - Any identified bundle branch re-entry has been successfully ablated

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Syncope (CRID-2.1.3)

ICD implantation is medically necessary to treat unexplained syncope in an individual with any of the following conditions:

- Long QT syndrome in an individual on beta blocker therapy
- Brugada pattern on ECG
- Catecholaminergic polymorphic VT in an individual on a beta blocker
- Coronary artery disease with a prior MI in the setting of **either** of the following:
  - LVEF 36%-49% and both of the following:
    - No Acute MI (less than or equal to 40 days)
    - Inducible sustained VT/VF on EP study
  - Ejection fraction less than or equal to 35%
- Non-ischemic structural heart disease and any of the following:
  - Left ventricular hypertrophy (LVH) with EF less than or equal to 35%
  - Non-ischemic dilated cardiomyopathy with ejection fraction less than or equal to 35%
  - Left ventricular non-compaction with ejection fraction less than 50%
  - Tetralogy of Fallot with prior corrective surgery
  - Hypertrophic cardiomyopathy
  - Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

Evidence Discussion

Secondary prevention refers to an indication for an ICD exclusively for patients who have survived >1 cardiac arrest(s) or episode(s) of sustained VT or VF. A ventricular arrhythmia is considered sustained if it lasts >30 seconds or result in hemodynamically significant symptoms prior to that time (i.e., requiring cardioversion or defibrillation). Patients with cardiac conditions associated with a high risk of sudden cardiac death (SCD) who have unexplained syncope presumed to be the result of a self-terminating ventricular arrhythmia are also considered to have a secondary prevention indication.

Primary Prevention ICD Indications (CRID-2.2)

Ischemic Cardiomyopathy (CRID-2.2.1)

Primary prevention is an indication for an ICD to prevent SCD. It refers to use of ICDs in individuals who are at risk for, but have not yet had, an episode of sustained VT, VF, or cardiac arrest.

ICD implantation is medically necessary for primary prevention of sudden cardiac death (SCD) associated with ischemic cardiomyopathy in the setting of **any** of the following:

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- Post-acute MI (less than or equal to 40 Days) with LVEF less than or equal to 30% in the setting of either of the following:
  - Complete revascularization after recent MI with PCI or CABG and **all** of the following:
    - Asymptomatic NSVT (greater than or equal to 4 days post-MI)
    - EPS with inducible VT
    - EPS performed after revascularization and less than 40 day post MI
  - Obstructive CAD not amenable to revascularization with **both** of the following:
    - Asymptomatic NSVT (greater than or equal to 4 days post-MI)
    - EPS with inducible sustained VT (EPS performed within 40 days of MI)
- Post-acute MI (less than or equal to 40 Days) LVEF 31% to 40% when all of the following apply:
  - Asymptomatic NSVT (greater than or equal to 4 days post-MI)
  - EPS with inducible sustained VT (if revascularized, EPS performed after revascularization, within 40 days of MI)
- Post-acute MI (less than or equal to 40 Days) and Pre-Existing Chronic Cardiomyopathy (greater than or equal to 3 Months) with any of the following:
  - LVEF less than or equal to 30% due to old infarction with functional class I-III
  - LVEF less than or equal to 35% with NYHA functional class II-III
- Post MI (greater than 40 Days) with any of the following conditions:
  - LVEF less than or equal to 35% and both:
    - NYHA functional class I, II, or III
    - No PCI/CABG within 3 months
  - LVEF 36%-40% with all of the following:
    - No PCI/CABG within 3 months
    - Asymptomatic NSVT
    - EPS with inducible sustained VT
  - PCI or CABG (less than or equal to 3 Months) with LVEF less than or equal to 35% on OMT for greater than or equal to 3 months prior to PCI/CABG
- Ischemic cardiomyopathy with LVEF less than or equal to 35% and either of the following:
  - Individual has completed greater than or equal to 3 months of **OMT**
  - Individual has completed less than 3 months of **OMT** with either of the following findings:
    - NSVT
    - EPS with inducible sustained VT

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Post Myocardial Infarction (MI) Need for Pacemaker Therapy (CRID-2.2.2)

ICD implantation is considered medically necessary in individuals with ischemic cardiomyopathy and a need for post-MI pacemaker therapy when **both** of the following clinical conditions exist:

- LVEF less than or equal to 35% in the setting of **either** of the following:
  - Individual is post-acute MI (less than or equal to 40 Days)
  - Individual is post MI (greater than 40 Days) with PCI or CABG (greater than or equal to 3 Months)
- **Any** of the following indications for pacemaker exist:

Sinus node dysfunction

Permanent pacemaker implantation is medically necessary for any of the following:

- Symptomatic non-reversible sinus node dysfunction as evidenced by **both** of the following:
  - Documented sinus node dysfunction including one of the below:
    - Sinus bradycardia at rate less than 50 beats per minute
    - Sinus pauses greater than 3 seconds
  - Symptoms attributable to non-reversible sinus node dysfunction including **one** of the below:
    - Syncope or pre-syncope
    - Heart failure symptoms
    - Exertional fatigue and impaired exercise tolerance
- Sinus bradycardia at rate less than 40 beats per minute and symptoms possibly related to bradycardia
- Symptomatic sinus bradycardia (as defined above) is a consequence of essential medical management and continued treatment is clinically necessary
- Symptoms attributable to bradycardia as listed above and evidence of tachy-brady syndrome (sinus bradycardia, ectopic atrial bradycardia, or sinus pause alternating with periods of atrial flutter or atrial fibrillation)
- Symptomatic chronotropic incompetence defined as limitations in activity due to the inability of the heart to increase its rate during exercise to achieve 80% of maximum predicted heart rate (220-age).

Atrioventricular block (AVB)

Permanent pacemaker implantation is medically necessary for any of the following:

- AVB including one of the below, with or without symptoms:
  - Second-degree Mobitz type II
  - High-grade (greater than or equal to 2 consecutive P waves at a constant physiologic rate that do not conduct to the ventricles)
  - Third-degree (complete heart block)

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- Any degree of AVB with one of the following symptoms that are clearly attributable to the AVB:
  - Syncope or pre-syncope
  - Heart failure symptoms
  - Exertional fatigue and impaired exercise tolerance
- Third-degree and advanced second-degree AV block at any anatomic level associated with sustained or non-sustained ventricular tachycardia (ventricular rhythm at rate greater than 100 bpm lasting greater than or equal to 3 consecutive beats) presumed due to AV block
- Marked first-degree AVB (PR interval greater than or equal to 0.3 seconds) or second-degree AVB with symptoms similar to those of pacemaker syndrome
- Symptomatic AVB as a consequence of guideline directed management and continued treatment is clinically necessary
- Persistent or permanent atrial fibrillation and symptomatic bradycardia including one of the following:
  - Rate less than 50 beats per minute
  - Regular QRS intervals indicating complete atrioventricular block (AVB)
- Second degree AV block with a documented pause of greater than or equal to 5 seconds during waking in the presence of atrial fibrillation, with or without symptoms
- Second degree AV block with documented periods of asystole greater than or equal to 3.0 seconds in the presence of sinus rhythm, with or without symptoms
- Second-degree AVB noted to be located at intra-His or infra-His levels at electrophysiology study (EPS), with or without symptoms
- Any AVB indication listed above occurring after acute myocardial infarction that does not resolve within 5 days
- Congenital complete or high-degree AVB in the presence of any of the following:
  - Symptoms related to bradycardia such as syncope, pre-syncope, heart failure symptoms, exertional fatigue, or impaired exercise tolerance
  - Wide QRS escape rhythm
  - Mean daytime heart rate below 50 bpm
  - Pauses greater than 3 times the cycle length of the ventricular escape rhythm
  - Complex ventricular ectopy
  - Prolonged QT interval
  - Ventricular dysfunction, ventricular dilatation or significant mitral regurgitation

Conduction Disorders with 1:1 Atrioventricular Conduction

Permanent pacemaker implantation is medically necessary for any of the following:

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- Individuals with syncope and bundle branch block and one of the following at electrophysiology study (EPS):
  - Baseline HV interval greater than or equal to 70 ms
  - Second- or third-degree intra- or infra-Hisian block during incremental atrial pacing
- Alternating bundle branch block, with or without symptoms
- HV interval greater than or equal to 100 milliseconds noted at EPS, with or without symptoms
- Intra-Hisian- or infra-Hisian block noted at EPS, with or without symptoms

Recurrent syncope

Permanent pacemaker implantation is medically necessary for individuals with recurrent syncope and any of the following:

- Spontaneous documented symptomatic asystolic pause greater than 3 seconds due to sinus arrest or atrioventricular block (AVB)
- Spontaneous documented asymptomatic asystolic pause greater than 6 seconds due to sinus arrest or AVB
- Cardioinhibitory carotid sinus syndrome as documented by one of the below:
  - Syncope caused by spontaneously occurring carotid sinus stimulation
  - Carotid sinus pressure that induces syncope and/or ventricular asystole of greater than or equal to 3 seconds
- Syncope associated with asystole of greater than or equal to 3 seconds during tilt testing
- Bundle branch block and one of the following at electrophysiology study (EPS):
  - Baseline HV interval greater than or equal to 70 ms
  - Second- or third-degree intra- or infra-Hisian block during incremental atrial pacing
- Syncope after cardiac transplantation with or without documentation of bradyarrhythmia
- Recurrent Adam-Stokes syndrome

Peri-procedural and post-operative indications

Permanent pacemaker implantation is medically necessary for any of the following:

- Prior to a planned catheter ablation of the atrioventricular (AV) junction for one of the following:
  - Rate control strategy for management of atrial fibrillation
  - Supraventricular tachycardia resulting in tachycardia induced cardiomyopathy that is not controlled with ablation or medical therapy
- Post Transcatheter Aortic Valve Implantation (TAVI) for any of the following:

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  - Complete or high-degree atrioventricular block (AVB) that persists for 24 to 48 hours after TAVI
  - New-onset alternating bundle branch block after TAVI
  - Pre-existing right bundle branch block (RBBB) and new conduction abnormality onset during or after (TAVI) such as:
    - Transient high-degree AVB
    - PR prolongation
    - QRS axis change
- Sinus node dysfunction or AVB associated with symptoms or hemodynamic instability occurring after cardiac surgery that does not resolve within 5 days
- Post cardiac transplant for any of the following:
  - Relative bradycardia that is prolonged or recurrent, which limits rehabilitation or discharge after postoperative recovery
  - Syncope with or without documentation of bradyarrhythmia

Neuromuscular diseases known to involve the heart

Permanent pacemaker implantation is considered medically necessary for progressive neuromuscular diseases known to involve the heart with any degree of atrioventricular (AV) block including first degree AV block or any fascicular block, with or without symptoms, because there may be unpredictable progression of AV conduction disease. Progressive neuromuscular diseases known to involve the heart include:

- Myotonic muscular dystrophy
- Kearns-Sayre syndrome
- Erb dystrophy (limb-girdle muscular dystrophy)
- Peroneal muscular atrophy

Non-ischemic Cardiomyopathy (CRID-2.2.3)

ICD implantation is medically necessary for primary prevention of SCD associated with non-ischemic cardiomyopathy in the setting of **any** of the following conditions:

- Idiopathic non-ischemic cardiomyopathy when **all** of the following apply:
  - LVEF less than or equal to 35% after greater than or equal to 3 months of **OMT**
  - Age less than or equal to 85 years
  - Functional NYHA class I-III
- Non-ischemic cardiomyopathy with recent valve surgery or TAVR with **all** of the following:
  - Recent Valve surgery or TAVR (within 3 months)
  - LVEF less than or equal to 35%

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- Need for pacemaker and LV function felt not likely to improve
- Non-ischemic cardiomyopathy from other specific etiologies in the setting of **any** of the following conditions in an individual on greater than or equal to 3 months of **OMT**:
  - Giant-cell myocarditis regardless of the LVEF
  - LVEF less than or equal to 35% with any of the following conditions
    - Sarcoid heart disease, no MRI performed
    - Myotonic dystrophy
    - Chagas disease
    - Peripartum cardiomyopathy with all of the following:
      - less than 3 months postpartum
      - LVEF persists less than or equal to 35% postpartum
  - Genetic conditions with structural heart disease **any** of the following:
    - Hypertrophic cardiomyopathy with **1 or more** of the following risk factor(s):
      - LV wall thickness greater than or equal to 30 mm
      - SCD in 1 or more first-degree relatives presumably caused by HCM
      - 1 or more episodes of unexplained syncope within the preceding 6 months
      - Spontaneous NSVT
      - Abnormal blood pressure response with exercise
    - Arrhythmogenic right ventricular dysplasia/CM
    - Evidence of structural cardiac disease with Lamin A/C mutation or other genetic ACM with LVEF less than or equal to 45%
  - In individuals without structural heart disease with **either** of the following genetic conditions:
    - Catecholaminergic Polymorphic VT with **both** of the following:
      - Non-sustained VT
      - one of the following:
        - Not tolerating medical therapy (beta-blockers, flecainide, or propafenone)
        - Breakthrough non-sustained ventricular arrhythmia on medical therapy (beta-blockers, flecainide, or propafenone)
    - Brugada by ECG (Type I ECG Pattern) with inducible VT or VF at EPS

Pre or Post Heart Transplant (CRID-2.2.4)

ICD implantation is medically necessary for primary prevention of SCD for **either** of the following indications:

- Individual is on waiting list for heart transplant (outpatient status)
- Individual is post heart transplant with multiple episodes of cellular or antibody-mediated rejection

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ICD Non-indications (CRID-2.7)

Non-indications

ICD implantation is considered **not medically necessary** in any of the following settings:

- Incessant VT or VF (hemodynamically stable VT or VF continuing for hours)
- VF or VT is due to a reversible cause such as:
  - Severe electrolyte disturbance
  - Drug-induced Torsades de Pointes (TdP)
  - Acute, reperfused myocardial infarction with preserved ejection fraction
- In an ablation candidate without structural heart disease

Substernal implantable cardioverter-defibrillator (CRID-2.8)

CPT® 0571T

- Substernal implantable cardioverter-defibrillator systems involve inserting a defibrillator lead directly beneath the sternum anterior to the heart and is intended to provide anti-tachycardia pacing as well as post-shock pacing without intravenous leads.
- At this time, substernal implantable cardioverter-defibrillator systems are considered experimental, investigational, and unproven.

Evidence Discussion

ICD implantation is **not** medically necessary for individuals who do not have a reasonable expectation of survival with an acceptable functional status for at least one year, even if they meet ICD implantation criteria listed in this guideline.

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Pediatric Implantable Cardioverter Defibrillator

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Pediatric ICD Indications (CRID-12)

General Information

The decision to manage a pediatric condition with an implantable cardioverter defibrillator should be shared between a pediatric electrophysiologist and an informed individual/guardian/parent. This includes education of the individual/guardian/parent on all available options/potential side-effects, including inaction. This is of particular importance in individuals with limited health literacy at risk for poor health outcomes. These guidelines have been developed based on the 2021 PACES Expert Consensus Statement on the Indications and Management of Cardiovascular Implantable Electronic Devices in Pediatric individuals and the 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy.

These guidelines are not considered to be all-inclusive but to provide a path to the current indications supported by the societies and evidence.

Coding

Description	CPT®
Insertion or replacement of permanent pacing cardioverter-defibrillator system with transvenous lead(s), single or dual chamber	33249
Insertion or replacement of permanent subcutaneous implantable defibrillator system, with subcutaneous electrode, including defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters when performed	33270

Implantable Cardioverter-defibrillator General Indications (CRID-12.0)

ICD implantation is considered medically necessary for survivors of pediatric sudden cardiac arrest (SCA) from ventricular tachycardia (VT) or ventricular fibrillation (VF) when both of the following have been addressed:

- completely reversible causes have been excluded

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- ICD is decided in consultation with an informed individual/guardian/parent to be more beneficial than alternative treatments that may significantly reduce the risk of SCA

ICD implantation is considered medically necessary for sustained VT that cannot be adequately controlled with medication and/or catheter ablation.

ICD therapy is considered medically necessary for primary prevention of SCA when all of the following conditions exist:

- decision has been made in consultation with an informed individual/guardian/parent
- the individual has a history of either of the following:
  - a genetic cardiovascular disease and risk factors for SCA
  - a pathogenic mutation and a family history of recurrent SCA

ICD replacement is considered medically necessary in a pediatric individual when indications have been reconsidered based on the most recent guidelines to evaluate for discontinuance of therapy.

Long QT Syndrome (LQTS) (CRID-12.1)

ICD implantation is considered medically necessary in individuals with LQTS in any of the following settings:

- survivors of SCA along with the use of beta-blockade
- individuals with symptoms (arrhythmic syncope or VT) when beta-blockade is either ineffective or not tolerated and cardiac sympathetic denervation or other medications are not considered effective alternatives
- primary prevention in individuals with established clinical risk factors and/or pathogenic mutation

Catecholaminergic Polymorphic Ventricular Tachycardia (CRID-12.2)

ICD implantation is considered medically necessary in individuals with a diagnosis of CPVT in any of the following settings:

- persistent cardiac arrest or arrhythmic syncope despite treatment with beta-blocker and flecainide or cardiac sympathetic denervation
- in combination with pharmacologic therapy with or without cardiac sympathetic denervation when aborted SCA is the initial presentation of CPVT
- documented polymorphic or bidirectional VT despite pharmacologic therapy with or without cardiac sympathetic denervation

Brugada Syndrome (BrS) (CRID-12.3)

ICD implantation is considered medically necessary in individuals with a diagnosis of BrS when there is a history of any of the following conditions:

- survivors of SCA

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- documented spontaneous sustained VT
- a spontaneous type I Brugada ECG pattern with a history of recent syncope presumed due to a ventricular arrhythmia
- syncope presumed due to a ventricular arrhythmia with a type I Brugada ECG pattern only with provocative medications

Hypertrophic Cardiomyopathy (HCM) (CRID-12.4)

ICD implantation is considered medically necessary in individuals with HCM in the setting of any of the following conditions:

- survivors of SCA
- spontaneous sustained VT
- ICD is decided after considering the potential complications of long-term ICD placement in consultation with an informed individual/guardian/parent to be more beneficial than alternative treatments in an individual with ≥1 primary risk factors including any of the following:
  - unexplained syncope
  - massive left ventricular hypertrophy
  - non-sustained VT
  - family history of early HCM-related SCD
- in individuals with HCM without the above risk factors but with secondary risk factors for SCA such as extensive LGE on cardiac MRI or systolic dysfunction

Arrhythmogenic Cardiomyopathies (ACM) (CRID-12.5)

ICD implantation is considered medically necessary in individuals in any of the following settings:

- individual has been resuscitated from SCA or sustained VT that is not hemodynamically tolerated
- in individuals with hemodynamically tolerated sustained VT, syncope presumed due to ventricular arrhythmia, or an LVEF less than or equal to 35%.
- in individuals with inherited ACM associated with increased risk of SCD based on an assessment of additional risk factors

Non-ischemic Dilated Cardiomyopathy (NIDCM) (CRID-12.6)

ICD implantation is considered medically necessary in individuals with NIDCM who either survive SCA or experience sustained VT not due to completely reversible causes.

ICD implantation is considered medically necessary in individuals with NIDCM and syncope or an LVEF ≤35% after less than or equal to 3 months of **OMT**.

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Congenital Heart Disease (CHD) (CRID-12.7)

ICD implantation is considered medically necessary for individuals with CHD for any of the following reasons:

- survivors of SCA when evaluation to define the cause of the event does not reveal any completely reversible causes
- individuals with hemodynamically unstable sustained VT who have undergone hemodynamic and electrophysiologic evaluation
- individuals with systemic LVEF ≤35% and sustained VT or presumed arrhythmogenic syncope
- individuals with spontaneous hemodynamically stable sustained VT who have undergone hemodynamic and electrophysiologic evaluation
- individuals with a single or systemic right ventricular ejection fraction less than or equal to 35%, particularly in the presence of additional risk factors such as VT, arrhythmic syncope, or severe systemic AV valve insufficiency.

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Cardiac Resynchronization Therapy (CRT) Devices

Guideline

Indications for Cardiac Resynchronization Therapy (CRT-D) Implantation (CRID-5)
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Indications for Cardiac Resynchronization Therapy (CRT-D) Implantation (CRID-5)

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Procedures included

CPT® 33224, 33225, 33208, 33229, 33249, 33264

Ischemic Cardiomyopathy (CRID-5.1)

CRT-D implantation is medically necessary for the treatment of ischemic cardiomyopathy when LVEF is less than or equal to 35% after greater than or equal to 3 month **OMT** with **any** of the following conditions:

- Sinus Rhythm with LBBB and **both** of the following:
  - QRS from 120 msec to 149 msec
  - NYHA Functional Class II, III, or ambulatory IV
- Sinus Rhythm with LBBB and **both** of the following:
  - QRS greater than or equal to 150 msec
  - NYHA functional class I, II, III, or ambulatory IV
- Sinus rhythm with non-LBBB and **both** of the following:
  - QRS greater than or equal to 150 msec
  - NYHA functional class III, or ambulatory IV
- Persistent or permanent atrial fibrillation and LBBB with QRS greater than or equal to 150 msec

CRT-P is considered medically necessary when all of the requirements of CRT-D have been met and the individual in consultation with the providing physician declines the ICD function.

Non-ischemic Cardiomyopathy (CRID-5.3)

CRT-D is medically necessary for the treatment of non-ischemic cardiomyopathy when LVEF is less than or equal to 35% after greater than or equal to 3 months of **OMT** with **any** of the following conditions:

- Sinus rhythm with LBBB and **both** of the following:

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  - QRS 120 msec to 149 msec
  - NYHA Functional Class II, III, or ambulatory IV
- Sinus rhythm with LBBB and **both** of the following:
  - QRS greater than or equal to 150 msec
  - NYHA Functional Class I, II, III, or ambulatory IV
- Sinus rhythm with Non-LBBB and **both** of the following:
  - QRS greater than or equal to 150 msec
  - NYHA Functional class III, or ambulatory class IV
- Persistent or Permanent atrial fibrillation and **both** of the following:
  - QRS greater than or equal to 150 msec
  - LBBB

CRT-P is considered medically necessary when all of the requirements of CRT-D have been met and the individual in consultation with the providing physician declines the ICD function.

Current or Anticipated RV Pacing with an Indication for an ICD (CRID-5.6)

CRT-D implantation is considered medically necessary for treatment of individuals who meet **all** of the following conditions:

- Current or anticipated RV pacing over 40%
- LVEF less than or equal to 35% after greater than or equal to 3 months of **OMT**
- NYHA Functional Class I, II, III, or ambulatory IV

CRT-P is considered medically necessary when all of the requirements of CRT-D have been met and the individual in consultation with the providing physician declines the ICD function.

Indications for upgrade to CRT-D (CRID-5.7)

Upgrade to CRT-D is medically necessary in individuals who have an implanted non-CRT-D ICD when **either** of the following conditions are documented:

- LVEF greater than or equal to 50% prior to ICD implant and **both** of the following are documented:
  - RV pacing burden greater than or equal to 40%
  - **One** of the following changes has occurred after implantation of non-CRT ICD:
    - Decline in LVEF greater than or equal to 10%
    - New or worsening heart failure symptoms (NYHA Class II or III)

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- LV ejection fraction less than or equal to 35% after greater than or equal to 3 months of **OMT** and **both** of the following are documented:
  - New or worsening symptomatic heart failure (NYHA functional Class II, III, or ambulatory class IV) following implantation of a non-CRT pacemaker or cardioverter-defibrillator (ICD)
  - Ventricular pacing greater than or equal to 40%

CRT-P is considered medically necessary in an individual with a non CRT pacemaker when all of the other requirements of CRT-D have been met and the individual in consultation with the providing physician declines the ICD function.

Indications for CRT-P (CRID-5.8)

Procedures included

CPT® 33224, 33225, 33208, 33229

CRT-P is medically necessary for any of the following:

- High grade AV block and NYHA Class I, II or III Congestive Heart Failure:
  - Initial CRT-P implantation is medically necessary in individuals who have **all** of the following:
    - LV ejection fraction less than 50%
    - NYHA Class I, II, or III heart failure
    - High grade AV block, including AV nodal ablation, requiring more than 40% ventricular pacing CRT-P
- Pacing-induced cardiomyopathy
  - Upgrade from non-CRT pacemaker to CRT-P is medically necessary in individuals who have **all** of the following:
    - LVEF greater than or equal to 50% prior to implantation of non-CRT pacemaker
    - Right ventricular pacing burden greater than or equal to 40%
    - One of the following occurring after implantation of non-CRT pacemaker:
      - Decline in LVEF greater than or equal to 10%
      - New or worsening heart failure symptoms NYHA Class II or III
- See also **Indications for Cardiac Resynchronization Therapy (CRT)-D Implantation** for individuals who have met requirements for CRT-D, but decline the ICD function

Indications for conduction system pacing

His bundle pacing or left bundle branch area pacing (CPT® 33207 or CPT® 33208) is considered medically necessary when **Indications for CRT)** are met and one of the following applies:

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- LV lead placement was attempted and was unsuccessful or suboptimal
- His bundle pacing or left bundle branch area pacing is planned in place of biventricular pacing

CRT Implantation - Non-indications (CRID-5.9)

Ischemic cardiomyopathy

CRT-D or CRT-P implantation is **not** medically necessary in individuals with ischemic cardiomyopathy who have had a myocardial infarction within the past 40 days or who have had coronary revascularization within the past 90 days **unless** a separate indication for permanent pacemaker or ICD implantation exists (thus preventing a likely repeat procedure for an upgraded device in the near future)

Reversible causes of cardiomyopathy

CRT-D implantation is **not** medically necessary in the setting of a reversible cardiomyopathy such as: toxic, metabolic, or tachycardia induced cardiomyopathy

Note: Once the reversible aberration is corrected, clinical reassessment is indicated

Wireless cardiac resynchronization - Criteria (CRID-5.10)

- Permanent LV leadless pacemakers (CPT® 0515T) are implanted directly in the left ventricle for synchronization with RV leads in the setting of cardiac resynchronization therapy. At this time, they are considered experimental, investigational, or unproven.

Evidence Discussion CRT

Evidence Discussion

Cardiac Resynchronization Pacing Therapy (CRT-P)

Asynchrony of electrical activation of different portions of the ventricles results in dyssynchrony of mechanical contraction leading to decreased efficiency of pump function. The cardiac resynchronization pacing device synchronizes electrical activation through a right ventricular lead and a lead inserted into a branch of the coronary venous system for left ventricular activation. Clinical decision-making for cardiac resynchronization pacing therapy includes consideration of an individual's risk of worsening symptomatic heart failure and the anticipated likelihood of favorable clinical response to cardiac resynchronization therapy balanced against short-term procedural risks (e.g., cardiac tamponade and contrast-mediated acute kidney injury) as well as

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long-term device-related risks (e.g., thrombosis and infection). Randomized controlled clinical trials and systematic reviews of these trials provide valuable information to guide optimal selection of candidates for cardiac resynchronization pacing therapy. For instance, the benefit-risk assessment for cardiac resynchronization pacing therapy is favorable for individuals with left ventricular ejection fraction less than 50% who require frequent ventricular pacing greater than 40% of the time. Conversely, the benefit-risk assessment for cardiac resynchronization pacing therapy is NOT favorable for individuals who have better left ventricular ejection fraction or who do not require ventricular pacing or require ventricular pacing only infrequently. Indications for cardiac resynchronization pacing therapy are based on well-established clinical medical specialty organization evidence-based guideline recommendations and Appropriate Use Criteria.

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Other Cardiac Implantable Devices

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Cardiac Devices for Other Conditions

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Wireless pulmonary artery pressure sensor - Criteria (CRID-13)

Codes addressed

Description	CPT®
Transcatheter implantation of wireless pulmonary artery pressure sensor for long-term hemodynamic monitoring, including deployment and calibration of the sensor, right heart catheterization, selective pulmonary catheterization, radiological supervision and interpretation, and pulmonary artery angiography, when performed	33289

Indications

Although FDA approved, the wireless pulmonary artery pressure sensor device (e.g., CardioMEMS™ HF System) has yet to be incorporated into the standard of care and is considered experimental, investigational, or unproven at this time.

Evidence Discussion

Wireless Pulmonary Artery Pressure Sensor devices (CPT® 33289) such as CardioMEMS™ HF System, are implanted into a branch of the pulmonary artery during right heart catheterization and require a specialized delivery system. These devices monitor constant pulmonary artery pressures over time, utilizing the concept that as pulmonary artery pressures increase, outpatient medical therapy can be adjusted. This can potentially reduce inpatient admissions and treatment.

- Although FDA approved, these devices have yet to be incorporated into the standard of care and remain investigational and experimental at this time.

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Wireless Left Atrial Pressure Sensor (CID-14)

Codes included

Description	CPT®
Transcatheter implantation of wireless left atrial pressure sensor for long-term left atrial pressure monitoring, including sensor calibration and deployment, right heart catheterization, transseptal puncture, imaging guidance, and radiological supervision and interpretation	0933T

Indications

Further studies are needed to evaluate the safety, clinical efficacy, and long-term stability of the wireless left atrial pressure sensor device. Wireless left atrial pressure sensor is considered experimental, investigational, or unproven at this time.

Evidence Discussion

Wireless left atrial pressure sensor (e.g., V-LAP™ system) implantation is performed percutaneously via femoral vein access and trans-atrial septal approach. It is proposed that the availability of hemodynamic parameters might allow clinicians to tailor heart failure treatment.

Cardiac Contractility Modulation (CRID-15)

Codes included

Description	CPT®
Insertion or replacement of permanent cardiac contractility modulation system, including contractility evaluation when performed, and programming of sensing and therapeutic parameters; pulse generator with transvenous electrodes	0408T
Insertion of permanent cardiac contractility modulation-defibrillation system component(s), including fluoroscopic guidance, and evaluation and programming of sensing and therapeutic parameters; pulse generator and dual transvenous electrodes/leads (pacing and defibrillation)	0915T

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Cardiac contractility modulation (CCM)

CPT® 0408T

Currently, there is insufficient evidence of benefit of cardiac contractility modulation (CCM) on clinical outcomes. CCM devices (e.g.,Optimizer Smart®) are considered experimental, investigational, or unproven at this time.

Combined Cardiac Contractility Modulation and Implantable Cardioverter-Defibrillator Device (CCM-D)

CPT® 0915T

Currently there is insufficient evidence of safety, efficacy, and benefit of combined cardiac contractility modulation and implantable cardioverter defibrillator devices (CCM-D) devices on clinical outcomes. CCM-D devices (e.g., Optimizer® Integra™ CCM-D System) are considered experimental, investigational, or unproven at this time.

Evidence Discussion

Cardiac contractility modulation (CCM) delivers electrical signals during the ventricular absolute refractory period. This does not generate an action potential or mechanical contraction but is proposed to enhance ventricular contractile strength. CCM has been proposed as a potential treatment option for individuals with symptomatic heart failure with reduced left ventricular ejection fraction who are not candidates for cardiac resynchronization therapy.

CCM has previously been used along with a separate implantable cardioverter-defibrillator device (ICD). The Combined Cardiac Contractility Modulation and Implantable Cardioverter-Defibrillator Device (CCM-D) combines CCM and ICD functionality in one device.

Baroreflex Activation Therapy (CRID-16)

Description	CPT®
Initial open implantation of baroreflex activation therapy (BAT) modulation system, including lead placement onto the carotid sinus, lead tunneling, connection to a pulse generator placed in a distant subcutaneous pocket (ie, total system), and intraoperative interrogation and programming	64654

Currently there is insufficient evidence of benefit of baroreflex activation therapy on clinical outcomes. Carotid baroreflex activation therapy (e.g., Barostim™ device) is considered experimental, investigational, or unproven at this time.

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Evidence Discussion

Carotid baroreflex activation therapy (e.g., Barostim™ device) uses a subcutaneously implanted pulse generator and electrode system to deliver electrical impulses to the carotid artery baroreceptors. Carotid baroreflex activation is aimed at decreasing sympathetic activity and increasing parasympathetic activity and has been proposed as a potential treatment option for individuals with symptomatic heart failure with reduced left ventricular ejection fraction.

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General Guidelines (CH-1)

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Abbreviations for Chest Guidelines

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Abbreviations for Chest Guidelines	
AAA	abdominal aortic aneurysm
ACE	angiotensin-converting enzyme
AVM	arteriovenous malformation
BP	blood pressure
CAD	computer-aided detection
CBC	complete blood count
COPD	chronic obstructive pulmonary disease
CT	computed tomography
CTA	computed tomography angiography
CTV	computed tomography venography
DVT	deep venous thrombosis
ECG	electrocardiogram
EM	electromagnetic
EMG	electromyogram
FDA	Food and Drug Administration
FDG	fluorodeoxyglucose
FNA	fine needle aspiration
GERD	gastroesophageal reflux disease

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Abbreviations for Chest Guidelines	
GI	gastrointestinal
HRCT	high resolution computed tomography
IPF	idiopathic pulmonary fibrosis
LFTP	localized fibrous tumor of the pleura
MRA	magnetic resonance angiography
MRI	magnetic resonance imaging
MRV	magnetic resonance venography
NCV	nerve conduction velocity
PE	pulmonary embolus
PET	positron emission tomography
PFT	pulmonary function tests
PPD	purified protein derivative of tuberculin
RODEO	Rotating Delivery of Excitation Off-resonance MRI
SPN	solitary pulmonary nodule
SVC	superior vena cava

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General Guidelines (CH-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms is required prior to considering advanced imaging.
  - A pertinent clinical evaluation should include the following:
    - a detailed history and physical examination
    - appropriate laboratory studies and basic imaging, such as plain radiography or ultrasound
      - A chest x-ray since the onset or change in symptoms that has been over read by a radiologist would be performed in many of these cases prior to considering advanced imaging.
        - Identify and compare with previous chest films to determine presence and stability.
  - For an established individual a meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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General Guidelines – Chest X-Ray (CH-1.1)

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- Chest x-ray can help identify previously unidentified disease and direct proper advanced imaging for such conditions as:
  - mediastinal lymphadenopathy (See **Mediastinal Lymphadenopathy (CH-2.3)**)
  - cough (See **Cough (CH-3.1)**)
  - non-cardiac chest pain (See **Non-Cardiac Chest Pain - Imaging (CH-4.1)**)
  - dyspnea (See **Dyspnea/Shortness of Breath (CH-5.1)**)
  - suspected post endobronchial valve complication (See **Post Endobronchial Valve (EBV) Placement (CH-5.3)**)
  - hemoptysis (See **Hemoptysis (CH-6.1)**)
  - bronchitis (See **Bronchitis (CH-8.1)**)
  - asbestos exposure (See **Asbestos Exposure (CH-9.1)**)
  - chronic obstructive pulmonary disease (See **COPD (CH-10.1)**)
  - e-cigarette or vaping product use-associated lung injury (See **E-cigarette, or Vaping, Product-Use Associated Lung Injury (EVALI) (CH-11.2)**)
  - pleural effusion (See **Pleural Effusion (CH-18.1)**)
  - pneumothorax or hemothorax (See **Pneumothorax/Hemothorax (CH-19.1)**)
  - pneumomediastinum (See **Pneumomediastinum; Subcutaneous Emphysema (CH-19.2)**)
  - fractured ribs, sternum, or clavicle (See **Chest Trauma (CH-21.1)**)
  - chest wall mass (See **Chest Wall Mass (CH-22.1)**)
  - acute and chronic infections (See **Pneumonia and Coronavirus Disease 2019 (COVID-19) (CH-13)** and **Other Chest Infections (CH-14)**)
  - malignancies
- Exceptions to preliminary chest x-ray may include such conditions as:
  - supraclavicular lymphadenopathy (See **Supraclavicular Region (CH-2.1)**)
  - axillary lymphadenopathy (See **Axillary Lymphadenopathy (and Mass) (CH-2.2)**)
  - costochondritis (See **Costochondritis/Other Musculoskeletal Chest Wall Syndrome (CH-4.2)**)
  - pre-operative assessment (See **Pre-Operative Assessment CH-5.2)**)
  - known bronchiectasis (See **Bronchiectasis (CH-7.1)**)
  - cystic fibrosis (See **Adult Cystic Fibrosis (CH-7.2)**)
  - suspected interstitial lung disease (See **Interstitial Lung Disease (ILD)/Diffuse Lung Disease (DLD) (CH-11.1)**)

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- positive PPD or tuberculosis (See **Other Chest Infections (CH-14)**)
- sarcoidosis (See **Sarcoid (CH-15.1)**)
- pulmonary nodules (See **Solitary Pulmonary Nodule (SPN) (CH-16)** and **Pleural-Based Nodules and Other Abnormalities (CH-17.1)**)
- mediastinal mass (See **Mediastinal Mass (CH-20.1)**)
- pectus excavatum and carinatum (See **Pectus Excavatum and Carinatum (CH-23.1)**)
- pulmonary arteriovenous fistula (See **Pulmonary AVM (CH-24.1)**)
- pulmonary embolism (See **Pulmonary Embolism (CH-25.1)**)
- elevated hemidiaphragm (See **Elevated Hemidiaphragm (CH-30.1)**)
- pre- or post-lung transplant (See **Lung Transplantation (CH-32)**)
- lung cancer screening (See **Lung Cancer Screening (CH-33)**)

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General Guidelines – Chest Ultrasound (CH-1.2)

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- Chest ultrasound (CPT® 76604) includes transverse, longitudinal, and oblique images of the chest wall with measurements of chest wall thickness, and also includes imaging of the mediastinum. It may be helpful in identifying pleural effusions (See **Pleural Effusion (CH-18)**).
  - Chest ultrasound:
    - CPT® 76604
  - Breast ultrasound:
    - CPT® 76641: unilateral, complete
    - CPT® 76642: unilateral, limited
    - CPT® 76641 and CPT® 76642 be reported only once per breast, per imaging session
  - Axillary ultrasound:
    - CPT® 76882 (unilateral); if bilateral, can be reported as CPT® 76882 x 2

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General Guidelines – CT Chest (CH-1.3)

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- Intrathoracic abnormalities as covered in these guidelines found on chest x-ray, fluoroscopy, CT Abdomen, or other imaging modalities can be further evaluated with CT Chest with contrast (CPT® 71260).
- CT Chest without contrast (CPT® 71250) can be used for the following:
  - individual has contraindication to contrast
  - follow-up of pulmonary nodule(s)
  - High Resolution CT (HRCT)
- For low-dose CT Chest (CPT® 71271 OR CPT® 71250): See **Lung Cancer Screening (CH-33).**
- CT Chest without and with contrast (CPT® 71270) does not add significant diagnostic information above and beyond that provided by CT Chest with contrast, unless a question regarding calcification, most often within a lung nodule, needs to be resolved.

CT Chest Coding Notes:

- High-resolution CT Chest should be reported only with an appropriate code from the set CPT® 71250-CPT® 71270.
  - No additional CPT® codes should be reported for the “high-resolution” portion of the scan. The “high-resolution” involves additional slices which are not separately billable.
- Low-dose CT chest does not have a specific, separate CPT® code associated with it. It should be reported with the standard CT chest CPT code (CPT® 71250).
  - An exception to this is the low-dose CT used specifically for the annual lung cancer screening scans, for which CPT® 71271 is used.

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General Guidelines – CTA Chest (CPT® 71275) (CH-1.4)

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- CTA Chest (CPT® 71275) is medically necessary for:
  - non-cardiac chest pain (See **Non-Cardiac Chest Pain - Imaging (CH-4.1)**)
  - hemoptysis (See **Hemoptysis (CH-6.1)**)
  - pulmonary arteriovenous fistula (See **Pulmonary AVM (CH-24.1)**)
  - pulmonary embolism (See **Pulmonary Embolism (CH-25.1)**)
  - pre-lung transplantation (See **Pre-Transplant Imaging Studies (CH-32.1)**)
  - prior to minimally invasive or robotic surgery (See **Transcatheter Aortic Valve Replacement (TAVR) (CD-4.8)** in the Cardiac Imaging Guidelines)

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General Guidelines – MRI Chest without and with Contrast (CPT® 71552) (CH-1.5)

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- Indications for MRI Chest are infrequent and may relate to concerns about CT contrast such as renal insufficiency or contrast allergy. MRI is medically necessary for:
  - Clarification of equivocal findings on previous imaging studies (often in the thymic mediastinal region).
    - Certain conditions include:
      - mediastinal lymphadenopathy (See **Mediastinal Lymphadenopathy (CH-2.3)**)
      - chest wall mass (See **Chest Wall Mass (CH-22.1)**)
      - mediastinal mass (See **Mediastinal Mass (CH-20.1)**)
      - chest muscle tendon injuries (See **Muscle/Tendon Unit Injuries/Diseases (MS-11.0)** in the Musculoskeletal Imaging Guidelines)
      - pectoralis tendon rupture (See **Muscle/Tendon Unit Injuries/Diseases (MS-11.0)** in the Musculoskeletal Imaging Guidelines)
      - brachial plexopathy (See **Brachial Plexus (PN-4.1)** in the Peripheral Nerve Disorders Imaging Guidelines)
      - thymoma (See **Thymoma and Thymic Carcinoma - Suspected/Diagnosis (ONC-10.5)** in the Oncology Imaging Guidelines)

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General Guidelines – Nuclear Medicine (CH-1.6)

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CPT	Description
78580	Pulmonary perfusion imaging (eg, particulate)
78582	Pulmonary ventilation (eg, aerosol or gas) and perfusion imaging
78597	Quantitative differential pulmonary perfusion, including imaging when performed
78598	Quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed

- For pulmonary perfusion imaging (eg, particulate) (CPT® 78580) and pulmonary ventilation (eg, aerosol or gas) and perfusion imaging (CPT® 78582): See **Pulmonary Embolism (CH-25.1).**
- For quantitative differential pulmonary perfusion, including imaging when performed (CPT® 78597) and quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed (CPT® 78598): See **Pre-Operative Assessment (CH-5.2).**

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Navigational Bronchoscopy and Biopsy (CH-1.7)

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- CPT® 76497 (Unlisted CT procedure) if:
  - A CT Chest has been performed within the last 6 weeks and study is needed for navigational bronchoscopy.
- CT Chest without contrast (CPT® 71250) if:
  - Previous diagnostic scan was ≥6 weeks ago and study is needed for navigational bronchoscopy.
- Bronchoscopy with computer-assisted, image-guided navigation, includes three-dimensional reconstruction. Do not report in conjunction with 3-D rendering CPT codes (CPT® 76376) or (CPT® 76377).
- Core needle biopsy (CPT® 32408) for:
  - Biopsy of the lung or mediastinum to diagnose malignancy.
- For robotic-assisted lung resection, see **Pre-Operative Assessment (CH-5.2)**.

Background and Supporting Information

- Navigational bronchoscopy: This is a form of guided bronchoscopy. A special sensor inside a bronchoscopy is used to navigate to the desired location within the lung. Computer software generates a virtual bronchial tree which provides a road map to the target lesion. A thin-cut CT Chest with optimized reconstruction parameters is required to generate the virtual map of the lungs. A previous CT Chest may not be usable for navigation if it was not formatted correctly, even if done just a few days prior.
- Names for navigational bronchoscopy systems can include:
  - superDimension or super-D
  - Spin Thoracic Navigation System
  - Archimedes
  - Monarch Platform - robotic
  - Ion - endoluminal robotic bronchoscopy platform
- The diagnostic accuracy of navigational bronchoscopy is 70.9-79%. It has an excellent safety profile, with an adverse event rate (e.g., pneumothorax) of 3.3-5.6%.
- Cone-Beam CT, (CBCT) is a newer technique that helps locate the nodule in real time. Studies have shown comparable results and diagnostic yields to other guided bronchoscopy strategies. CBCT combined with other navigational techniques often reaches a higher navigational success rate and diagnostic rate. However, a recent

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study found no statistically significant differences in diagnostic sensitivity, specificity, and accuracy between those who received CBCT and other methods. Additionally, CBCT can expose the individual and operator to radiation through the repeated acquisition of images. The mean radiation dose was determined to be 48.4 Gy cm^2 per case; this excessive and unnecessary radiation can cause radioactive damage to the individual and the operator. Another study concluded that, "Additional studies are warranted to confirm the safety and efficacy of this technique". Efforts are required to improve diagnostic accuracy and standardized practices before CBCT can be considered mainstream. Large-scale randomized controlled trials that compare CBCT with other navigational techniques are necessary to determine the most optimal navigational strategy, clinical criteria for CBCT usage, and the best way to diagnose peripheral pulmonary lesions.

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Lymphadenopathy (CH-2)

Guideline

Supraclavicular Region (CH-2.1)
Axillary Lymphadenopathy (and Mass) (CH-2.2)
Mediastinal Lymphadenopathy (CH-2.3)
References (CH-2)

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Supraclavicular Region (CH-2.1)

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- Ultrasound (CPT® 76536) is the initial study for palpable or suspected lymphadenopathy.
  - Allows simultaneous ultrasound-guided core needle biopsy (CPT® 76942)
  - CT Neck with contrast (CPT® 70491) or CT Chest with contrast (CPT® 71260) if ultrasound is indeterminate
    - See **General Guidelines (Neck-1.0)** in the Neck Imaging Guidelines

Evidence Discussion

For suspected or palpable supraclavicular lymphadenopathy, ultrasound (US) has an excellent sensitivity rate, up to 100% for the detection of metastases. CT Neck had a lower sensitivity rate of 83% for the same lesion.[6] Ultrasound avoids the ionizing radiation exposure of CT, is readily available, and allows for the use of US-guided fine-needle aspiration cytology for diagnosis.

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Axillary Lymphadenopathy (and Mass) (CH-2.2)

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- There is no evidence-based support for advanced imaging of clinically evidenced axillary lymphadenopathy prior to a biopsy. If axillary node biopsy reveals benign findings, advanced imaging is not medically necessary. If axillary node biopsy reveals findings concerning for malignancy, pathology results will determine the need for further advanced imaging. See **Carcinoma of Unknown Primary Site (ONC-31.7)** in the Oncology Imaging Guidelines for imaging recommendations for carcinoma found in an axillary lymph node.
- Localized axillary lymphadenopathy:
  - Axillary US (CPT® 76882)
    - Initial evaluation of any axillary mass or enlarged node
  - Search for adjacent hand or arm injury or infection, and
  - 3-4 week observation if benign clinical picture (for ipsilateral COVID vaccination-related adenopathy, observation for 12 or more weeks is recommended). Follow-up imaging with ultrasound is medically necessary if there is a significant risk of metastatic adenopathy (e.g., breast, head and neck, upper extremity/trunk melanoma or lymphoma)
    - If axillary adenopathy is unchanged, then consider additional follow up 6 months after initial presentation.
  - Ultrasound directed core needle biopsy or surgical excisional biopsy of the most abnormal lymph node if:
    - Condition persists.
    - Malignancy is suspected.
  - Surgical excisional biopsy if core needle biopsy results are non-diagnostic.
  - Advanced imaging is not medically necessary.
- Generalized axillary lymphadenopathy:
  - Axillary US (CPT® 76882)
    - Initial evaluation of any axillary mass or enlarged node
  - Ultrasound directed core needle biopsy or surgical excisional biopsy of the most abnormal lymph node if condition persists, if malignancy is suspected, or surgical excisional biopsy if core needle biopsy results are non-diagnostic.
  - Diagnostic work-up, including serological tests, for systemic diseases
  - Advanced imaging is not medically necessary.
  - See **Non-Hodgkin Lymphomas (ONC-27)** in the Oncology Imaging Guidelines.

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- Occult primary cancer in axillary lymph node(s):
  - See **Metastatic Cancer, Carcinoma of Unknown Primary Site, and Other Types of Cancer (ONC-31)** in the Oncology Imaging Guidelines.
- For unexplained axillary lymphadenopathy in individuals with silicone breast implants, see **Breast Implant Evaluation (BR-5.2)**

Evidence Discussion

Initial evaluation of an axillary mass or axillary lymphadenopathy (LAN) should be ultrasound (US). US allows for real-time evaluation and immediate image-directed biopsy.[11] CT Chest is usually not appropriate in the evaluation of axillary LAN, especially in the female population with concern for breast cancer.[10]

Ultrasound is a very important initial imaging modality which is easy to obtain, universally available and portable, exposes individuals to no radiation, and is cost effective. It is also excellent in helping to determine next best advanced imaging study including appropriate protocol and contrast level. US not only provides excellent soft tissue resolution, but also provides characterization of cystic lesions (Bosniak classification) whether complex or simple to help guide follow up imaging interval or biopsy.

Background and Supporting Information

- Adenocarcinoma is the most common histology, with breast cancer seen most often; non-palpable breast cancer and axillary metastases accounts for less than 0.5% of all breast cancers. Carcinomas of the lung, thyroid, stomach, colon, rectum, and pancreas have the potential to spread to axillary lymph nodes, but these metastases are rarely the first manifestations of disease.
- COVID-19 vaccine-related unilateral axillary adenopathy has been well documented to occur in 12% of recipients after the first dose and up to 16% after the second dose. In some series the incidence has been as high as 53%. Adenopathy usually develops within the first few days after vaccination and lasts a mean of 10 days. However, 29% had lymphadenopathy which persisted >6 weeks. PET-CT can provide false positive results of unilateral axillary adenopathy up to 7-10 weeks post vaccination. Due to these concerns, in individuals with cancer history, it is recommended that the vaccination be provided in the contralateral arm, especially in case of unilateral breast cancer.
- The Society for Breast Imaging (SBI) recommends that for unilateral axillary adenopathy on screening exams who received a recent COVID-19 vaccination in the ipsilateral upper extremity, a follow up interval of 12 or more weeks is recommended. If axillary adenopathy persists after short term follow up, then consider lymph node sampling to exclude breast and non-breast malignancy. Imaging for urgent cancer related clinical indication should not be delayed in relationship to COVID vaccine timing. For routine surveillance, screening and similar non-urgent

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indications, postponement of imaging for at least 6 weeks after vaccinations should be considered. However, the SBI no longer recommends delaying screening mammograms around COVID-19 vaccinations.

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Mediastinal Lymphadenopathy (CH-2.3)

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- CT Chest with contrast (CPT® 71260) if mediastinal abnormalities are detected on a chest x-ray (over read by a radiologist), other non-dedicated advanced chest imaging, or clarification of mediastinal abnormalities on a non-contrast CT Chest.
  - If contrast cannot be tolerated due to allergy or renal dysfunction, MRI chest without and with contrast (CPT® 71552) OR MRI chest without contrast (CPT® 71550) is medically necessary.
  - Follow-up CT Chest (CPT® 71260) after 3-6 months if:
    - enlarged lymph nodes, ≥15 mm, are in the mediastinum with no other thoracic abnormalities; and
    - thereafter, stability or decreasing size, further advanced imaging is not medically necessary.
  - Further evaluations:
    - Lymph node biopsy (see methods below) is medically necessary for:
      - persistent or increasing lymphadenopathy on follow-up CT Chest; or
      - suspected malignancy.
      - See **Non-Hodgkin Lymphomas (ONC-27)** and/or **Hodgkin Lymphoma (ONC-28)** in the Oncology Imaging Guidelines for suspicion of Lymphoma.
- PET/CT (CPT® 78815) is medically necessary for enlarged lymph nodes, ≥15 mm with no explainable disease or increasing lymph node size on follow-up CT Chest

Evidence Discussion

- CT Chest is medically necessary for mediastinal abnormalities detected on chest x-ray or other non-dedicated advanced imaging. CT allows for further tissue characterization and can distinguish between calcium, macroscopic fat and water attenuation fluid.[1] CT has higher contrast resolution than plain chest radiography. CT does carry with it the risk of both iodinated contrast exposure and ionizing radiation exposure.
- Asymptomatic, incidental mediastinal lymph nodes less than 15mm (in the short axis) do not require follow up. Evison et al found that size was the greatest predictor of lymph node etiology with those less than 15mm always found to be reactive.[8]
- For mediastinal lymph nodes greater than or equal to 15mm, follow-up should be directed by suspected etiology. For those with low or no clinical suspicion for malignancy and no other thoracic abnormalities, follow up CT chest in 3-6 months is medically necessary.[8] If the lymph nodes have increased in size on follow-up imaging, PET/CT or tissue biopsy is medically necessary.[8]

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- For those with no explainable disease and mediastinal lymph nodes greater than or equal to 15mm, PET/CT is medically necessary. However, PET/CT has well-documented false positive results in this setting given the overlap of increased FDG uptake in both oncologic and infection or inflammatory disease processes.[8]

Background and Supporting Information

- Incidentally detected lymph nodes <15mm (in short axis) in individuals with no other findings do not require further evaluation.
- Most benign nodes have smooth and well-defined borders, show uniform and homogeneous attenuation, and demonstrate a central fatty hilum.
- Explainable disease such as emphysema, interstitial lung disease, sarcoidosis, cardiac disease.
- Unexplained causes, consider lymphoma, undiagnosed metastatic disease, including testicular carcinoma in young male, and infection.
- Lymphadenopathy from neoplasms as well as from benign sources of inflammation can result in a positive PET scan. Therefore, the use of PET may not be helpful prior to histologic diagnosis.
- Less invasive methods of mediastinal biopsies are CT or ultrasound directed percutaneous biopsy, transbronchial biopsy, transbronchial biopsy using endobronchial ultrasound, and endoscopic ultrasound-guided FNA.
- More invasive and traditional methods are mediastinoscopy or thoracoscopy/ thoracotomy.

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Cough (CH-3)

Guideline

Cough (CH-3.1)
References (CH-3)

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Cough (CH-3.1)

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- Initial evaluation should include a chest x-ray after the current episode of cough started or changed.
  - In addition all medications known to cause coughing (e.g., ACE inhibitors, Sitagliptin) should be discontinued.
- CT Chest (either with contrast [CPT® 71260] or without contrast [CPT® 71250]), if the initial chest x-ray is without abnormalities and all medications known to cause coughing have been discontinued, for the following:
  - Non-smoker cough after the following sequence for a total 3-week trial and investigation after ALL of the following:
    - Antihistamine and decongestant or intranasal glucocorticoid treatment.
    - Spirometry and/or pulmonary function tests (PFT's).
    - Empiric trial of corticosteroids (oral or inhaled) and/or leukotriene receptor antagonist (e.g., Montelukast).
    - Treatment of gastroesophageal reflux disease (GERD).
      - See **Sinus and Facial Imaging (HD-29.1)** in the Head Imaging Guidelines.
  - Current or past cigarette smokers with either:
    - new cough lasting greater than 2 weeks
    - changed chronic cough in worsening frequency or character
      - See **Hemoptysis (CH-6.1)**
  - See the relevant Chest Imaging Guideline section for advanced chest imaging to evaluate any abnormalities present on the initial chest x-ray.
  - For individuals with high suspicion for lung cancer despite normal chest x-ray:
    - See **Non-Small Cell Lung Cancer Suspected/Diagnosis (ONC-8.2)**
- CT Maxillofacial without contrast (CPT® 70486) or CT Sinus, limited without contrast (CPT® 76380) is medically necessary in those with suspicion of Upper Airway Cough Syndrome (UACS) in the following:
  - Clinical criteria for chronic rhinosinusitis (CRS) or acute/recurrent rhinosinusitis are met, as per **Sinus and Facial Imaging (HD 29.1)**; **OR** ALL of the following:
    - at least a one week trial of daily antihistamine/decongestant
    - initial evaluation with a chest x-ray and/or CT Chest after the current episode of cough started or changed
    - all medications known to cause cough have been discontinued

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Evidence Discussion

CT chest is not recommended routinely in people with a chronic cough, normal chest x-ray, and normal physical exam. There is concern regarding potential cancer risk from CT radiation exposure, especially in women and children.[7] For individuals with cough of unknown etiology or a chronic cough refractory to therapy, a CT chest may identify changes not seen on chest x-ray, such as interstitial lung disease or bronchiectasis.[7,9,12]

Current or former smokers with a new cough or change in chronic cough do not need a trial of therapy for UACS, asthma or GERD prior to a chest CT if an initial chest x-ray is abnormal or non-diagnostic.[3]

CT maxillofacial may be considered for suspected chronic rhinosinusitis (CRS) as the cause of chronic cough after clinical examination and chest x-ray if there is no response to empiric therapy or if the history and nasal endoscopy findings are concerning for CRS.[3,5,9]

Background and Supporting Information

- The resolution of cough usually will occur at a median time of 26 days of stopping use of the angiotensin-converting enzyme (ACE) inhibitor drug. Smoking cessation is “almost always effective” in resolving cough in smoker.
- Cough after URI (Upper Respiratory Infection) can typically last beyond 2-3 weeks.
- Objective evidence of classic asthmatic cough conventionally requires some evidence of variable airflow obstruction such as peak flow variability, or reversibility to bronchodilator of >12%-15%.
- In adults with chronic cough suspected to be due to reflux-cough syndrome, it is recommended that treatment include (1) diet modification to promote weight loss in overweight or obese individuals; (2) head of bed elevation and avoiding meals within 3 hours of bedtime; and (3) in individuals who report heartburn or regurgitation, PPI's, H-2 receptor antagonists, alginate or antacid therapy sufficient to control these symptoms.
- When cough remains refractory to treatment, high-resolution CT scanning of the thorax is recommended to rule out parenchymal lung disease that is not visible on plain chest x-ray.

Cough Health Equity Considerations

Health Equity Considerations for Cough

- Chronic cough is more prevalent in older adults. Younger adults with chronic cough are more likely to be men, and older adults are more likely to be women.

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Non-Cardiac Chest Pain (CH-4)

Guideline

Non-Cardiac Chest Pain – Imaging (CH-4.1)
Costochondritis/Other Musculoskeletal Chest Wall Syndrome (CH-4.2)
References (CH-4)

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Non-Cardiac Chest Pain – Imaging (CH-4.1)

CH.CP.0004.1.A
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- Initial evaluation should include a chest x-ray.
  - CT Chest with contrast (CPT® 71260) or CTA Chest (CPT® 71275) if x-ray is abnormal. See **Pneumonia (CH-13.1)**
- Sub-Sternal Non-Cardiac Chest Pain:
  - If x-ray is normal and the chest pain is substernal, the individual should undergo evaluation of other possible causes of pain prior to advanced imaging (CT Chest with contrast or CTA Chest) including:
    - Cardiac evaluation (See General Guidelines (CD-1) in the Cardiac Imaging Guidelines)
    - GI treatment with any ONE of the following:
      - Trial of anti-reflux medication, or pH probe, or esophageal manometry or
      - Barium swallow or endoscopy
    - Pulmonary Function Test (PFT's) in those with known or suspected respiratory disease
  - CT Chest with contrast (CPT® 71260) if persistent:
    - The initial chest x-ray reveals no abnormalities with known Sickle cell disease
- Non-Cardiac Chest Pain, other than Sub-Sternal:
  - If x-ray is normal and the chest pain is in a location other than substernal:
    - CT Chest with (CPT® 71260) or without (CPT® 71250) contrast and/or bone scan for:
      - known or suspected malignancy, including individuals with chest pain associated with cough and weight loss
    - CT Chest with (CPT® 71260) or without (CPT® 71250) contrast for:
      - suspected infectious or inflammatory condition
      - history of prior chest intervention (surgery, Radiation Therapy)
    - MRI Chest without and with contrast (CPT® 71552) for:
      - necrotizing fasciitis
      - surgical planning prior to debridement procedure
  - For suspected migration of implantable contraceptive devices, see Implantable Contraceptive Devices (PV-10.3)

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Evidence Discussion

It is important to rule out potentially life-threatening causes of acute chest pain, such as an acute coronary syndrome, aortic dissection, and pulmonary embolus. These topics are discussed in other guideline summaries (CD 1.0, CD 1.4, PVD 6.2 and PVD 6.3, CH 25.1). A specialized imaging protocol called the "triple rule-out" is sometimes used to evaluate the pulmonary arteries, aorta and coronary arteries. However, it is associated with higher non-diagnostic imaging quality, radiation and contrast doses.[8] The population for which it may be useful is unknown. It is yet to be proven useful in large clinical trials, and its appropriate use needs to be further defined.[8,9,10]

An evaluation for the cause of non-cardiac "angina-like" chest pain should be done if it persists or recurs despite a negative stress test or anatomic cardiac evaluation, or a low risk designation by a clinical decision pathway.[11] The differential diagnosis of non-cardiac chest pain is broad. The most common causes in a primary care setting are chest wall pain, reflux esophagitis, and costochondritis.[6] Respiratory causes include pneumonia, pleuritis , and pneumothorax. People with COPD or acute asthma exacerbations may experience chest pain.[5,12] A thorough history and physical exam are important to help narrow the differential diagnosis and direct imaging. Musculoskeletal causes are usually diagnosed based on history and physical exam (point tenderness, reproducibility with palpation) without the need for diagnostic imaging. Most individuals should have an ECG and chest x-ray (CR).[11] CR is rapid, non-invasive and is medically necessary in the initial evaluation of acute non-specific chest pain with a low probability of coronary artery disease (CAD).[1,11] In individuals without evidence of cardiac or pulmonary disease, evaluation for a GI cause is reasonable. An empiric trial of acid suppression may be merited. If this is ineffective or there are alarm symptoms, an EGD, pH probe and/or motility study should be considered.[11,13,14]

Individuals with sickle cell disease and acute chest pain should have a CR initially.[11] Acute chest syndrome is defined by a new infiltrate on CR with fever and/or respiratory symptoms. In the presence of unexplained hypoxemia and an unremarkable CR, CT chest may be obtained to evaluate the pulmonary vasculature and lung parenchyma.[15]

CR is medically necessary for non-traumatic chest wall pain and no history of malignancy to evaluate for a specific etiology, such as rib fracture, pneumonia, or pneumothorax.[4,17] Following a normal CR, CT chest is medically necessary to evaluate chest pain in the setting of known or suspected malignancy, suspected infectious or inflammatory condition, or a history of prior chest intervention.[4] CT is more sensitive than CR for characterizing chest wall neoplasms, chest wall infections, and subtle osseous and soft tissue lesions. Chest MRI is useful if there is a high suspicion for necrotizing fasciitis and for surgical planning prior to debridement.[4]

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Costochondritis/Other Musculoskeletal Chest Wall Syndrome (CH-4.2)

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- Initial evaluation should include a chest x-ray.
- CT chest (CPT® 71250 or CPT® 71260) is medically necessary for inconclusive or nondiagnostic findings on chest x-ray AND one of the following:
  - concern for infectious or neoplastic processes
  - persistent symptoms >3 weeks (despite treatment)
- For MRI requests, see the applicable section:
  - **Infection (MS-9.0)**
  - **Muscle and Tendon Injuries (MS-11)**
  - **Joint Instability and Dysfunction (MS-30)**
  - **Chest Wall Mass (CH-22.1)**
- For slipping rib syndrome, dynamic chest ultrasound (CPT® 76604) is medically necessary.
  - CT chest (CPT® 71250 or CPT® 71260) is medically necessary for pre-surgical planning
- For other conditions not mentioned, see **Non-Cardiac Chest Pain - Imaging (CH-4.1)**

Evidence Discussion

Costochondritis is a common cause of chest wall pain in adults presenting to the emergency department and physician's office.[3,16] It is defined as inflammation of costochondral junctions of ribs or costosternal joints, usually at multiple levels and without any swelling or induration.[3] It is a self-limited condition. The diagnosis is largely based on history and physical examination, which reproduces pain on palpation of the chest wall. Upper body movement, deep breathing, and exertional activities often exacerbate the pain.[3,16] Tietze syndrome presents similarly to costochondritis but includes visible edema at the involved joint(s), typically is unilateral involving the second rib, and is often incited by infection or trauma.[16]

There are no laboratory tests or imaging tests findings specifically for the diagnosis of costochondritis.[22] If an individual relates a history of dyspnea or chest wall trauma, a chest radiograph or rib series is medically necessary.[4,16] Chest radiographs can help identify potential sources of previously undifferentiated chest pain such as pneumothorax, pneumomediastinum, fractured ribs, acute and chronic infections, pneumonia, lung mass, and malignancies.[1] A large observational study found that 91%

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of individuals with new-onset costochondritis had resolution of pain after three weeks of treatment with rest and nonsteroidal anti-inflammatory drugs.[18] Thus, chest CT should be reserved for individuals who have persistent symptoms greater than 3-4 weeks, or where there is high suspicion for infection or neoplastic processes.[3] Scintigraphy has been studied to determine its usefulness to diagnose costochondritis, but it is not specific for the diagnosis.[3]

Most treatment recommendations are conservative in nature and have been traditionally accepted, perhaps because of the self-limited nature of the condition.[3,16]

Recalcitrant cases may respond to corticosteroid injections.[16]

Background and Supporting Information

- Chest x-ray could identify pneumothorax, pneumomediastinum, fractured ribs, acute and chronic infections, and malignancies.
- Costochondritis can be readily diagnosed with palpation tenderness and/or hooking maneuver and imaging is non-specific.
- Chest CT, chest MRI, and bone scans are not helpful for the diagnosis of slipping rib syndrome. However, they may be helpful in delineating rib anatomy, which would aid in surgical planning.
- Dynamic chest ultrasound accurately diagnoses slipping rib syndrome in 89% of cases and has a 100% negative predictive value.
- Intercostal nerve blocks could be used for diagnostic and therapeutic purposes.

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Dyspnea/Shortness of Breath (CH-5)

Guideline

Dyspnea/Shortness of Breath (CH-5.1)
Pre-Operative Assessment (CH-5.2)
Post Endobronchial Valve (EBV) Placement (CH-5.3)
References (CH-5)

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Dyspnea/Shortness of Breath (CH-5.1)

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- Initial evaluation should include a chest x-ray since onset or change in symptoms.
  - CT Chest without contrast (CPT® 71250) OR CT Chest with contrast (CPT® 71260) if x-ray is abnormal.
  - CT Chest without contrast (CPT® 71250, including HRCT), or CT Chest with contrast (CPT® 71260) if the initial chest x-ray is indeterminate and the following evaluations have been conducted, since onset or change in symptoms, and are indeterminate:
    - ECG, echocardiogram or stress testing, and
    - Pulse oximetry and pulmonary function studies (PFT's)
- If pulmonary embolus (PE) is suspected, See **Pulmonary Embolism (PE) (CH-25)**.
- For persistent dyspnea post-COVID-19 infection, see Coronavirus Disease 2019 (COVID-19) (CH-13.2).

Evidence Discussion

There is no standard approach for the evaluation of chronic dyspnea, and data that test diagnostic algorithms against standard clinical care are limited; however, clinical practice algorithms have been proposed and found to be effective.[13-16] If the diagnosis is not evident after a history and physical exam, initial diagnostic testing with pulse oximetry, spirometry, chest radiography (CR), ECG, and labs is recommended.[14,15,17] While the individual utility of these tests varies for a specific diagnosis, they are commonly available and easy to perform.[17]

Spirometry can identify obstructive lung disease or suggest restrictive lung disease. The flow-volume loop may suggest intra- or extra-thoracic airway obstruction. Some authors recommend full pulmonary function tests as part of the initial investigation, while others consider spirometry an appropriate initial test.[13,15] Diagnostic accuracy is improved when spirometry is done in addition to a clinical assessment.[17]

ECG has a high negative predictive value for cardiac disease but low specificity. Thus, further testing such as echo is often necessary.[17] The recommended timing of echocardiography differs between algorithms but echo is an important test for cardiac causes of dyspnea.[14,17] The American College of Radiology (ACR) states that for dyspnea of suspected cardiac origin, the initial diagnostic imaging should usually be CR followed by transthoracic echo.[2]

CR remains a valuable first line investigation of dyspnea.[14]The ACR states that CR should generally be the first imaging study.[1] It may reveal abnormalities or guide further imaging decisions. Data on the diagnostic utility of chest CT for chronic dyspnea

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are limited. It is often used following an abnormal CR or if other initial testing is negative. The ACR states that CT may be useful when CR abnormalities require further characterization or clinical findings necessitate additional imaging despite a normal CR.[1] CT without intravenous contrast is usually sufficient unless there is a suspicion for vascular abnormalities. The disadvantage is exposure to ionizing radiation; therefore, CT "requires careful patient selection with consideration given to patient age, risk of diagnostic radiation exposure and estimated diagnostic yield."[17]

Background and Supporting Information

- Dyspnea is the subjective experience of breathing discomfort.

Dyspnea/Shortness of Breath Health Equity Considerations

- Dyspnea disproportionately affects individuals with a low socioeconomic status.

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Pre-Operative Assessment (CH-5.2)

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- For pre-operative assessment prior to a planned segmental, lobar, or lung removal, as well as for pre-interventional assessment prior to a planned endobronchial valve (e.g., Zephyr valve) placement, the following are medically necessary:
  - "Split Function Studies" (CPT® 78597-Quantitative Differential Pulmonary Perfusion, Including Imaging When Performed or CPT® 78598-Quantitative Differential Pulmonary Perfusion and Ventilation (e.g., Aerosol or Gas), Including Imaging When Performed) or SPECT/CT (CPT® 78830)

    AND/OR
  - CT Chest (CPT® 71250, CPT® 71260 or CPT® 71270) for pre-interventional procedure assessment prior to a planned endobronchial valve (e.g., Zephyr Valve) placement.
  - For robotic assisted lung resection, CT chest (CPT® 71250 OR CPT® 71260) is medically necessary if:
    - Previous diagnostic scan was ≥6 weeks ago
    - CPT® 76497 (Unlisted CT procedure) if a CT Chest has been performed < 6 weeks ago

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Post Endobronchial Valve (EBV) Placement (CH-5.3)

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- Suspected Post EBV Complication:
  - Initial evaluation should include a chest x-ray
    - CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) is medically necessary for:
      - acute loss of benefit, lack of initial benefit, increased dyspnea, sudden chest pain, increased cough, suspected valve malposition/migration, or to evaluate target lobe volume reduction

Evidence Discussion

The most common acute complications following EBV placement are pneumothorax, pneumonia, COPD exacerbation and valve migration.[10] Pneumothorax occurs in 20-30% of individuals, the majority within the first 48 hours after the procedure. Individuals who have an acute increase in dyspnea, cough or chest pain, or an acute perceived loss of benefit, should have a chest X-ray (CR) to rule out pneumothorax. If the CR is non-diagnostic, a CT chest should be done to evaluate the valve position, the target lobe and volume reduction more precisely.[9]

Following EBV placement, it may take several days to one month for significant volume reduction and atelectasis of the target lobe to occur. If no significant lung volume reduction is seen on CR at one month, a CT should be done to evaluate valve position.[10] A CT chest is performed routinely at some centers 6-8 weeks after EBV placement.[9] If there has been no clinical benefit and no lobar atelectasis is evident on CT at 6 weeks, a revision bronchoscopy may be necessary.[11] The two most common causes of lack of benefit are the presence of interlobar collateral ventilation or valve misplacement/migration.

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Hemoptysis (CH-6)

Guideline

Hemoptysis (CH-6.1)
Reference (CH-6)

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Hemoptysis (CH-6.1)

CH.HS.0006.1.A

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- Following a chest x-ray performed after hemoptysis started or worsened the following is medically necessary:
  - CT Chest with contrast (CPT® 71260) or CTA Chest (CPT® 71275)
- For recurrent hemoptysis (hemoptysis occurring after medical therapy or embolization), or related to pre-bronchial artery embolization planning, the following is medically necessary:
  - CTA Chest (CPT® 71275) (preferred) or CT chest with contrast (CPT® 71260) if requested

NOTE:

- CT Chest without contrast, (CPT® 71250), is only warranted in individuals with poor renal function or life-threatening contrast allergy.
- There is no data to support the use of CT Chest without and with contrast, (CPT® 71270) in the diagnosis of hemoptysis.

Background and Supporting Information

- The most common causes of hemoptysis are acute respiratory tract infections, respiratory tract neoplasm, bronchiectasis, and tuberculosis. Chest x-ray is the most common initial test because it is easy to obtain, can help lateralize the site of bleeding in 32-80% of cases, and may demonstrate underlying parenchymal abnormalities. If no cause is found on chest x-ray, further evaluation is necessary with CT chest with contrast, CTA chest, and/or bronchoscopy. Angiographic localization of the site of bleeding can be challenging, time-consuming, and requires a significant contrast load; therefore, pre-procedural chest CT and/or bronchoscopy to help localize the bleeding site is valuable. Determination of which arteries to embolize is based on a combination of CT, bronchoscopic localization of bleeding when applicable, and angiographic findings.

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Bronchiectasis (CH-7)

Guideline

Bronchiectasis (CH-7.1)
Adult Cystic Fibrosis (CH-7.2)
References (CH-7)

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Bronchiectasis (CH-7.1)

CH.BR.0007.1.A
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- High resolution CT Chest (HRCT) without contrast (CPT® 71250) for ANY of the following:
  - To confirm suspected diagnosis of bronchiectasis after an initial x-ray.
  - For known bronchiectasis with worsening symptoms or worsening PFT's.
  - For hemoptysis with known or suspected bronchiectasis.

Evidence Discussion

The British Thoracic Society (BTS) recommends performing a baseline chest x ray (CR) in people with suspected bronchiectasis followed by a thin section (< or equal to 1 mm slice thickness) CT scan to confirm the diagnosis.[7] According to the American College of Radiology (ACR), CR is relatively insensitive but is medically necessary and often performed as initial imaging for evaluation of associated conditions and exclusion of diseases that cause similar symptoms.[9] High resolution CT chest (HRCT) is considered the most accurate imaging modality for the diagnosis of bronchiectasis.[8] The ACR states that CT chest without contrast is medically necessary for suspected bronchiectasis to identify and characterize the severity and distribution of bronchiectasis and to evaluate any associated parenchymal lung diseases.[9] CT chest can help identify an etiology, such as allergic bronchopulmonary aspergillosis, Primary Ciliary Dyskinesia, tracheobronchomegaly, or a foreign body.[7,8]

MRI chest for suspected bronchiectasis is not medically necessary because it is inferior to CT for evaluating lung parenchyma, and its use is mainly limited to research settings.[9]

CR is often the initial chest imaging exam to evaluate acute conditions in people with bronchiectasis, such as pneumonia or hemoptysis. CT chest without contrast is medically necessary for the evaluation of complications and assessing changes in clinical status.[9] CT chest with contrast is medically necessary in the setting of a suspected acute infection and associated complication, such as abscess.[9] The BTS recommends a CT chest for people with a deteriorating clinical status, such as worsening symptoms, increased frequency or severity of acute exacerbations, or decreasing lung function.[7] They recommend a CT chest with contrast if PE is suspected. CTA chest with contrast is medically necessary in the setting of hemoptysis to identify dilated bronchial arteries or systemic collateral vessels and for pre-procedure planning.[8,9]

High quality evidence in favor of repeated imaging is lacking.[8]CR may not show structural changes. Repeat HRCT carries the risk of increased radiation. Individuals with diseases associated with bronchiectasis may be evaluated with CT to help guide

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therapy and provide prognostic information.[9]The current indication for repeat HRCT is clinical deterioration.[7])

Bronchiectasis Health Equity Considerations

- Bronchiectasis is more prevalent among females. The most aggressive disease occurs among post-menopausal females.

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Adult Cystic Fibrosis (CH-7.2)

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- CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is medically necessary for the following (without initial chest x-ray):
  - Suspected or initial diagnosis of Cystic Fibrosis
  - Biennially, (every 2 years), for routine surveillance
  - Persistent respiratory symptoms with reduced lung function despite therapy
  - Exacerbations when chest x-ray is indeterminate
  - Hemoptysis
  - Suspected fungal pneumonia
  - Pre and post-lung transplant evaluation
- See **Bronchiectasis (CH-7.1)**

Evidence Discussion

Imaging is an important method of evaluating the lungs in people with cystic fibrosis (CF). It has a stronger correlation with disease severity than pulmonary function tests and facilitates prompt therapy which may help limit irreversible lung damage.[10] Chest x- ray (CR) is less sensitive than CT chest at detecting early structural changes and disease progression.[4] However, CR is still most commonly used as the first line imaging examination for the assessment of acute complications due to its low cost, availability, low radiation and speed of acquisition.[11] CT is increasingly being used to monitor disease progression and make treatment decisions, but the routine use of CT for short term follow up during pulmonary exacerbations is not recommended due to the risk of a high cumulative radiation dose. Low dose chest CT (LDCT) is useful in individuals with persistent respiratory symptoms and decreased lung function despite appropriate therapy.[4] There is little evidence regarding the optimal timing of CT monitoring. The current best clinical practice in several European CF centers is a CT every two years with a radiation dose as low as reasonably achievable (ALARA). Follow up imaging is determined by individual-dependent clinical factors.[4] The CF Foundation guidelines for adult CF clinical care recommend CR every 2-4 years in those with a stable clinical status and state that imaging should be considered if there are symptoms or signs of an acute pulmonary exacerbation, pneumothorax, lobar atelectasis or hemoptysis.[12,13]

Several emerging techniques offer promising means of pulmonary imaging using less ionizing radiation, including ultra-low dose CT (ULDCT) and MRI.[10,14,18,20] The radiation dose with CR is 0.02mSv, 5.4 mSv for standard dose CT, 1 to 2 mSv for LDCT, and 0.05-0.08mSv for ULDCT. While pulmonary MRI has promise as a means of routinely monitoring CF lung disease, it is currently limited by a lack of availability, high cost, lack

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of validation and standardized protocols, and the need for sedation or anesthesia in some individuals.[4,10]

Adult Cystic Fibrosis Health Equity Considerations

- There are disparities in cystic fibrosis outcomes related to race, ethnicity, socioeconomic status, geographic location, sexuality, and gender identity.
  - People with disadvantaged backgrounds have worse cystic fibrosis health outcomes and tend to die younger than those without.
  - Women tend to have worse cystic fibrosis outcomes than men.

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Bronchitis (CH-8)

Guideline

Bronchitis (CH-8.1)
References (CH-8)

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Bronchitis (CH-8.1)

CH.BH.0008.1.A
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- Chest x-ray is medically necessary as initial imaging if symptoms persist or worsen. Advanced imaging is not medically necessary for bronchitis unless directed by condition specific guideline.
  - See the following guidelines for additional information:
    - For Pneumonia, see: Pneumonia (CH-13.1).
    - For Cough, see: Cough (CH-3.1).
    - For Pleural Effusion, see: Pleural Effusion (CH-18.1).
    - For pulmonary mass, see: Pulmonary Nodule (CH-16.1), Pulmonary Nodule (CH-16.1).

Evidence Discussion

Acute bronchitis is a self-limited respiratory infection characterized by cough due to acute inflammation of the trachea and large airways without evidence of pneumonia.[1, 2.] This syndrome should be distinguished from the common cold, an acute exacerbation of chronic bronchitis and acute asthma.[1]

Cough associated with acute bronchitis typically lasts about two to three weeks. Other diagnoses must be considered when cough persists for more than three weeks.[1] Acute bronchitis is mainly caused by viruses, and antibiotics are not typically indicated in individuals without chronic lung disease[1, 2]. Imaging is primarily used to rule out pneumonia. Evidence-based guidelines from the American College of Chest Physicians state that imaging is not needed in individuals with acute bronchitis symptoms who have normal vital signs and normal lung examination findings.[2]

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Asbestos Exposure (CH-9)

Guideline

Asbestos Exposure (CH-9.1)
References (CH-9)

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Asbestos Exposure (CH-9.1)

CH.AE.0009.1.A
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- Chest x-ray as radiographic screening for asbestos exposure.
  - Stable calcified pleural plaques on chest x-ray do not require advanced imaging of the chest.
- CT Chest is not medically necessary to screen populations at risk for asbestos-related diseases.
- High resolution CT Chest (HRCT) (CPT® 71250) for ANY of the following:
  - Any change seen on chest x-ray
  - Progressive respiratory symptoms that may indicate the development or progression of asbestos related interstitial fibrosis

Evidence Discussion

Several well-conducted epidemiologic studies of occupationally exposed workers, family contacts of workers, and persons living near asbestos mines have demonstrated that exposure to asbestos is associated with an increased incidence of asbestosis, lung cancer, mesothelioma, as well as other neoplasms. Asbestosis is a fibrotic lung disease caused by accumulation of asbestos fibers in the lungs. The diagnosis of asbestosis is most commonly made based on a history of exposure to asbestos, the presence of characteristic radiologic abnormalities, end-inspiratory rales, and other clinical features.[1-3]

A chest x-ray of an individual exposed to asbestos may show pleural plaques, pleural calcifications, pleural fibrosis, or small irregular parenchymal opacities. Lung cancer risk is not elevated among individuals with asbestos-related pleural plaques in the absence of asbestosis.[1-3]

Chest x-ray is currently medically necessary to screen for lung changes resulting from asbestos exposure and is recommended for those who have had relatively heavy exposure to asbestos. However, chest x-rays lack specificity. When a chest x-ray abnormality is indeterminate, High Resolution CT Chest (HRCT) is useful in revealing characteristic parenchymal abnormalities. There is a lack of consensus among experts regarding the value of HRCT for screening of asbestos-related pulmonary disease.[1-3]

Background and Supporting Information

- Asbestosis and asbestos-related diseases include: pleural effusion, pleural plaques, lung cancer, and malignant mesothelioma. The risk of developing mesothelioma increases with increasing intensity and duration of exposure.

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Chronic Obstructive Pulmonary Disease (COPD) (CH-10)

Guideline

COPD (CH-10.1)
References (CH-10)

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COPD (CH-10.1)

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- Chest x-ray should be performed initially.
  - CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) can be performed if:
    - Emphysema is known or suspected and a pre-operative study for Lung Volume Reduction Surgery (LVRS) is being requested. OR
    - For known COPD:
      - individuals with persistent exacerbation OR
      - symptoms out of proportion to disease severity OR
      - FEV1 <45% of predicted
    - PFT's, appropriate laboratory studies, and chest x-ray do not reveal a definitive diagnosis and there is suspicion of an additional diagnosis including, but not limited to:
      - Bronchiectasis
      - Sarcoidosis
      - Emphysema
      - Pneumoconiosis
      - Idiopathic pulmonary fibrosis
      - Langerhans cell histiocytosis
      - Hypersensitivity pneumonitis
      - Bronchiolitis obliterans
      - Lipoid pneumonia
      - Drug toxicity
      - Lymphangitic cancer
      - Alpha-1-Antitrypsin Deficiency
- Lung cancer screening is discussed in the following guideline:
  - See "Screening Indications" in **Lung Cancer Screening (CH-33)**
- Pre-interventional lung procedure assessment prior to a planned endobronchial valve (e.g., Zephyr valve) placement
  - See **Pre-Operative Assessment (CH-5.2)**

Evidence Discussion

Chest x-ray (CR) is usually the appropriate initial imaging study for suspected COPD to exclude alternative diagnoses and evaluate for comorbidities and complications.[1-3] CR, pulmonary function tests (PFT's) and selected blood tests lead to a specific diagnosis

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in a significant proportion of people with chronic dyspnea.[3] CT has increased sensitivity and specificity for determining the type, severity, and distribution of emphysema and bronchial abnormalities.[1,3] It is an important part of the pre-procedure evaluation process for lung volume reduction surgery, endobronchial valve placement, and lung transplantation.[3] The GOLD 2023 report recommends that CT be considered for individuals with COPD with persistent exacerbations and concern for another diagnosis, such as bronchiectasis or an atypical infection, or symptoms out of proportion to the disease severity suggested by PFT's.[2] Some authors have proposed broadening the definition of COPD to include CT-detected emphysema, air trapping or airway wall thickening, even in the absence of airflow obstruction on spirometry.[5,6]

CT is helpful if a smoking-related interstitial lung disease, such as pulmonary Langerhans cell histiocytosis or Combined Pulmonary Fibrosis and Emphysema, is suspected.[7,8] It is recommended following a diagnosis of alpha-1 antitrypsin deficiency.[9] CT is also used in the evaluation of central airway abnormalities associated with COPD, such as tracheobronchomalacia and excessive dynamic airways collapse.[3] Annual lung cancer screening CT's should be performed in current or former smokers who meet the USPSTF criteria, but screening CT's are not recommended for those with COPD not due to smoking because there is insufficient evidence to establish benefit over harm.[3]

COPD exacerbations are characterized by dyspnea, cough and/or sputum which worsen over a period of less than two weeks.[3] They are mainly caused by respiratory viral infections. CR is often performed to rule out alternative diagnoses, such as pneumonia, pneumothorax or pulmonary edema.

Background and Supporting Information

- COPD includes asthmatic bronchitis, chronic bronchitis, and emphysema. COPD is airflow reduction (FEV1/FVC ratio <0.7 or FEV1 <80% predicted) in the presence of respiratory symptoms, such as dyspnea. Advanced chest imaging is not typically indicated in COPD exacerbation, which is an acute change in baseline dyspnea, cough, and/or sputum beyond normal day-to-day variations.

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Interstitial Disease (CH-11)

Guideline

Interstitial Lung Disease (ILD)/Diffuse Lung Disease (DLD) (CH-11.1)
E-cigarette, or Vaping, Product Use–Associated Lung Injury (EVALI) (CH-11.2)
References (CH-11)

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Interstitial Lung Disease (ILD)/Diffuse Lung Disease (DLD) (CH-11.1)

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- EITHER high resolution CT Chest (HRCT) without contrast (CPT® 71250) (the diagnostic modality of choice) OR CT Chest with contrast (CPT® 71260) to evaluate for the following:
  - Interstitial changes or diffuse parenchymal changes identified on other imaging (including chest x-ray) (See **Dyspnea/Shortness of Breath (CH-5.1)**
  - In individuals that have pulmonary symptoms with abnormal pulmonary function studies (PFT's) and a normal chest x-ray and there is high clinical suspicion for ILD or DLD, including but not limited to, the following entities: Hypersensitivity Pneumonitis, Cryptogenic Organizing Pneumonia (COP, formally known as BOOP), and Eosinophilic Pneumonia.
  - Initial imaging to identify ILD/DLD in individuals with a high suspicion for connective tissue disease or diagnosis of connective tissue disease (based upon autoantibody testing, serology, etc.), including, but not limited to:
    - rheumatoid arthritis
    - scleroderma
    - idiopathic inflammatory myopathies (polymyositis, dermatomyositis, inclusion body myositis)
    - systemic lupus erythematosus
    - Sjögren's syndrome
    - mixed connective tissue disease
  - Significant exposure to a substance or medication (linked to ILD/DLD) and concern for:
    - Abstestosis
    - Silicosis
    - Coal miner's lung disease
    - Drug induced pulmonary toxicity
      - Follow-up CT chest (CPT® 71250 or CPT® 71260) for worsening symptoms or PFT's, guide for steroid taper, or baseline prior to re-exposure to offending agent
      - Timing at the discretion of the treating provider
  - At any time for detection of Progressive Pulmonary Fibrosis (PPF), in individuals with ILD of known or unknown etiology, defined by at least one of the following:
    - New or worsening respiratory symptoms

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        - Worsening PFT's, defined as decline of either:
            - FVC of 5% or greater within the past year
            - DLCO of 10% or greater within the past year
    - Annually in individuals with known pulmonary fibrosis if needed for:
        - serial examination for improvements in diagnostic accuracy, or
        - evaluation of disease reversibility, stability, or progression.
- Concern for interstitial lung disease post-COVID, See **Coronavirus Disease 2019 (COVID-19) (CH-13.2)**
- HRCT can be done even if a regular CT Chest has been done previously. HRCT is done with a thinner-slice protocol that can provide additional details to help determine ILD subtype.
- HRCT can also be done with inspiratory/expiratory and supine/prone views.

Evidence Discussion

ILD is often suspected in those with chronic dyspnea or non-productive cough, especially in the setting of an inhalational exposure or systemic disease known to be associated with lung involvement.[1,7,11] Chest x-ray (CR) and CT chest without contrast are medically necessary for suspected ILD and provide complementary information.[1,7] A normal CR does not rule out ILD, as CR can be normal in up to 10% of individuals with ILD. Its primary function is to evaluate for an alternative diagnosis. CR remains an important imaging modality to screen for occupational lung disease.[1] High resolution CT (HRCT) has higher sensitivity and specificity for ILD. HRCT may help guide a biopsy site or provide a definitive diagnosis, making a biopsy unnecessary.[1,7] CT can provide prognostic information: individuals with honeycombing or a usual interstitial pneumonia (UIP) pattern on CT have increased mortality.[12]

CR and CT chest without contrast are medically necessary for evaluation of an acute exacerbation of ILD.[7] They can help exclude alternative causes for worsening clinical symptoms and confirm abnormalities consistent with progression of ILD. There are no data to support routine surveillance imaging of ILD, but serial CT's can improve diagnostic accuracy and evaluate disease stability, reversibility or progression.[7] The optimal interval for follow up HRCT to determine disease progression in idiopathic pulmonary fibrosis (IPF) is unknown. Raghu et al recommend consideration of an annual HRCT in people with IPF if there is clinical suspicion of worsening fibrosis or to screen for complications such as lung cancer.[7] In people with an ILD other than IPF and radiologic evidence of fibrosis, disease progression on HRCT is one of three criteria used to define progressive pulmonary fibrosis.[7,13]

Interstitial lung abnormalities (ILA) are abnormalities on CT suggestive of ILD in people without a prior clinical diagnosis.[10,14] They are common incidental findings, especially in older people. ILA are a radiologic observation. Differentiation between ILA and clinical or subclinical ILD must be on the basis of a clinical evaluation. When respiratory

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signs/symptoms or functional impairment is present, ILA likely represent mild ILD. The morphology and distribution of ILA are important: subpleural fibrotic ILA are most likely to progress. There is minimal evidence to support a specific management plan for ILA. Hatabu et al recommend that when ILA are detected, a dedicated HRCT chest can help confirm and characterize the abnormalities, especially if the initial scan was incomplete (i.e., a CT abdomen) or not performed with thin sections.[14] Hunninghake et al recommend that a HRCT should be done in those with ILA.[10] If clinically significant ILD is ruled out, Hatabu et al recommend a repeat CT at 12-24 months for those with subpleural fibrotic ILA or other risk factors for progression to ILD.[14] However, participants in a consensus survey disagreed about repeating a HRCT at the follow up evaluation.[10] People with nonfibrotic nonsubpleural ILA and no symptoms or physiologic impairment do not need reimaging.[14,15]

Background and Supporting Information

- DLD refers to diffuse parenchymal lung diseases or interstitial lung diseases. There are a multitude of pathologies that demonstrate involvement of the alveoli, airways, or both, in addition to the pulmonary interstitium. A single term of ILD would not fully address the entities that are mostly parenchymal in nature, hence the term Diffuse Lung Disease is more technically correct. Both terms are included here for convenience and recognition.
- There is no relevant literature to support the use of CT with IV contrast for initial or follow-up imaging of ILD; however, IV contrast may be of use in evaluation of alternative diagnoses with overlapping clinical features or conditions that also involve the pleura, mediastinum, and pulmonary vessels.
- Progression of fibrosis is typically assessed visually, relying on the percentage of lung volume containing fibrotic features in the upper, mid, and lower lung zones. An increased extent of fibrotic features denotes progression. These may include increased traction bronchiectasis and bronchiolectasis, new ground-glass opacity with traction bronchiectasis, new fine reticulation, increased coarseness of reticular abnormality, new or increased honeycombing, and increased lobar volume loss.

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E-cigarette, or Vaping, Product Use–Associated Lung Injury (EVALI) (CH-11.2)

CH.ID.0011.2.A

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- Chest x-ray is medically necessary as first line imaging for suspected EVALI.
- CT Chest with or without contrast (CPT® 71250 or CPT® 71260) is medically necessary if:
  - Clinical suspicion for EVALI but negative chest x-ray
  - To identify extent of pulmonary involvement identified on chest x-ray
  - Severe (e.g., tachycardia, tachypnea, or hypoxia (O_2 95% or less)) or worsening disease (does not require a repeat chest x-ray)
  - To rule out other diagnoses (infection, pulmonary edema, etc.)
  - Clinical suspicion of pneumothorax or pneumomediastinum (See **Pneumothorax/ Hemothorax (CH-19.1**))

Evidence Discussion

EVALI is a toxic inhalational acute lung injury with imaging and histopathologic patterns of organizing pneumonia and/or diffuse alveolar damage.[16,17] Individuals with EVALI mainly present with respiratory symptoms (e.g., shortness of breath, cough, chest pain), gastrointestinal symptoms (e.g., abdominal pain, diarrhea, nausea, vomiting), and systemic symptoms (e.g., fever, weight loss, fatigue).[22] The diagnostic criteria for EVALI, as defined by the CDC for a "confirmed EVALI" case, is as follows: i) individuals must have vaped within 90 days before the onset of symptoms, ii) bilateral pulmonary opacities on chest imaging, iii) a negative evaluation for infection, iv) no other plausible alternative diagnoses.[22] Chest x-ray (CR) can exclude other diagnoses and is often the first imaging study. CR is not abnormal at initial assessment in all individuals with EVALI. When pulmonary abnormalities are not identified on CR or when further characterization of CR findings are needed to evaluate for another potential cause of symptoms, CT chest can be obtained.[16,17]

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Pneumonia and Coronavirus Disease 2019 (COVID-19) (CH-13)

Guideline

Pneumonia (CH-13.1)
Coronavirus Disease 2019 (COVID-19) (CH-13.2)
References (CH-13)

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Pneumonia (CH-13.1)

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- Chest x-ray should be performed initially in all individuals with suspected pneumonia, prior to considering advanced imaging.
  - CT Chest without or with contrast (CPT® 71250 or CPT® 71260) if initial or repeat chest x-ray findings reveal:
    - complication of pneumonia (e.g., abscess, effusion, necrotizing pneumonia, pneumothorax)
    - possible lung mass associated with the infiltrate or persistent radiographic abnormalities on repeat chest x-ray after 6-8 weeks
  - CT Chest without or with contrast (CPT® 71250 or CPT® 71260) for hypoxia and/ or respiratory distress
  - CT Chest without or with contrast (CPT® 71250 or CPT®71260) after initial chest radiograph is negative or equivocal and one of the following:
    - Abnormal vital signs (including hypoxemia, pulse > 100, respiratory rate > 24, fever > 100)
    - Abnormal exam (including respiratory distress, dyspnea and or abnormal lung auscultation)
    - Advanced age (age >75), or other significant comorbidities
- If pulmonary emboli suspected, see **Pulmonary Embolism (CH-25.1).**
- CT Chest without or with contrast (CPT® 71250 or CPT® 71260) for immunocompromised individuals with any of the following:
  - High suspicion for pneumonia despite equivocal or negative chest x-ray
  - Persistent radiographic abnormalities on repeat imaging after 6-12 weeks
  - Multiple or diffuse opacities or nodules

Evidence Discussion

Chest radiography (CR) is the appropriate first imaging modality in the evaluation of suspected pneumonia.[16] The American Thoracic Society (ATS) and the Infectious Diseases Society of America (IDSA) define a clinical diagnosis of pneumonia as symptoms and signs of pneumonia with radiographic confirmation.[1] Chest radiographs should be performed for individuals with symptoms of dyspnea, tachycardia, tachypnea and fever more than 100°F or lung findings suggestive of pneumonia.[28]

CT is more accurate/sensitive than CR and may be medically necessary when there is a high clinical suspicion for pneumonia (typical or atypical) and a delay in diagnosis could be life threatening.

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CT chest without contrast is medically necessary and CT chest with contrast may be medically necessary in immunocompromised people with an acute respiratory illness and a normal/equivocal/nonspecific CR or a CR that demonstrates multiple, diffuse or confluent opacities.[15]

MRI is not medically necessary for the imaging of pneumonia.[2,15] MRI has a potential role for follow up imaging of parenchymal (Chest wall/mediastinal) disease, but CT is more sensitive and is preferred.[15]

Ultrasound chest may be appropriate for the evaluation of parapneumonic effusions. It can also be used to guide pneumonia-related interventions.[2,29-31]

Routine use of follow up chest imaging in adults who are improving and whose symptoms have resolved within 5-7 days is not recommended by the ATS/IDSA.[1] Repeat CR or CT after the completion of therapy is generally reserved for high risk individuals, suspected complications, disease progression or when the clinical course differs from CR interpretation.[16,27] High risk individuals include individuals over the age of 50, smokers, and those with persistent symptoms.[32,33] For immunocompetent individuals with suspected pneumonia, follow-up imaging within 6-12 weeks is medically necessary to confirm resolution. The follow-up imaging modality should be the same modality in which pneumonia was detected.[2]

Pneumonia Health Equity Considerations

Mortality from pneumonia is higher in non-Hispanic Black individuals than non-Hispanic White individuals. These disparities are the greatest in major metropolitan areas.

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Coronavirus Disease 2019 (COVID-19) (CH-13.2)

CH.PN.0013.2.A

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- CT Chest without contrast (CPT®71250), or with contrast (CPT®71260) is medically necessary in the following clinical situations:
  - Imaging for initial diagnosis:
    - Symptomatic COVID-19 positive individuals with underlying comorbidities (including, but not limited to, age > 65 years, chronic lung disease, current or former smoker, chronic kidney disease, chronic liver disease, dementia, diabetes, Down's syndrome, HIV or other primary, secondary or acquired immunodeficiency, mood disorders, BMI ≥30, pregnancy, solid organ or blood stem cell transplant, cerebrovascular disease, substance use disorder, tuberculosis, cardiovascular disease, malignancy, bronchopulmonary dysplasia, chronic infections, or immunocompromised state).
    - Moderate to severe symptomatic individuals with evidence of significant pulmonary dysfunction or damage (e.g., hypoxemia, moderate-to-severe dyspnea), suspected of having COVID-19, regardless of COVID-19 test results or when viral testing is not available, even with a normal chest x-ray (if done).
    - Thromboembolic complications including pulmonary embolism, stroke and mesenteric ischemia are recognized complications of COVID-19. See **Pulmonary Embolism (CH-25.1)**, **Mesenteric Ischemia (AB-6.1)** in the Abdomen Imaging Guidelines, and **Stroke/TIA (HD-21.1)** in the Head Imaging Guidelines for appropriate imaging guidance.
    - Other systemic complications are being recognized as medical knowledge about this condition evolves. Imaging for possible COVID-19 complications should be managed by the appropriate condition based guidelines.
  - Imaging after initial diagnosis:
    - Imaging in the following clinical circumstances:
      - If there is significant worsening of symptoms in a COVID-19 positive individual and imaging will be used to modify individual management.
      - A recovered COVID-19 positive individual with significant residual functional impairment and/or persistence hypoxemia.
        - Symptomatic post-COVID individuals with concern for interstitial lung disease including organizing pneumonia imaging can be considered pre and post treatment.

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Evidence Discussion

Chest imaging is not medically necessary as a screening test for COVID-19 in asymptomatic people or in people with suspected COVID-19 and mild clinical features unless they are at risk for disease progression.[7]

The American College of Radiology (ACR) states that CT chest should not be used as a screen or first-line test to diagnose COVID-19.[3] Viral testing is the only specific method of diagnosis and confirmation with a viral test is required even if radiologic findings on chest radiography (CR) or CT are suggestive of COVID-19.

Imaging is medically necessary in people with COVID-19 and a worsening respiratory status or in people who have suspected COVID-19, a high pretest probability of disease, and moderate to severe clinical features.[7] Although less sensitive than CT, chest radiography (CR) is typically the first line imaging modality.[2,7,18]

Johnston et al have proposed a management algorithm for individuals with COVID-19 pneumonia which recommends a clinical assessment with CR and PFT's 3-6 months after discharge.[23] Performance of a high resolution CT chest (HRCT) is based on risk factors (ICU admission, noninvasive or mechanical ventilation; male sex; age > 60) and serial assessment of lung function and symptoms.

There is an increased risk of pulmonary embolus (PE) in people with COVID-19, including both microvascular/ in situ thrombosis and macrothrombotic events.[25] It is currently recommended that the same diagnostic strategy and the same D-dimer threshold be used for people with COVID-19 and suspected PE as in those without COVID-19.[25,26]

Background and Supporting Information

- The role of advanced imaging in the diagnosis and management of COVID-19 is very dynamic in this rapidly evolving condition.
- Comorbidities may include: chronic heart, lung, liver, or renal disease; diabetes mellitus; alcoholism; malignancy; asplenia; organic brain disease (dementia, CVA, delirium).
- Findings on both Chest x-ray and CT Chest are non-specific. Chest x-rays may show patchy opacities with lower lung predominance. CT may show peripheral multifocal ground glass opacities with lower lung predominance. However, a significant portion of cases have opacities without a clear or specific distribution. A reverse halo sign or other findings of organizing pneumonia may be seen later during the course of illness. Atypical findings include isolated lobar or segmental consolidation without ground glass opacities, discrete small centrilobular ("tree-in-bud") nodules, pleural effusion.
  - Pediatric individuals may have less pronounced imaging findings than adults.
- CT Chest abnormalities are common 3 months after discharge in adults who have been hospitalized for COVID-19 and are associated with more severe acute disease.

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Fibrosis was seen in a minority of people. Most people re-imaged at one year showed radiologic improvement.

- Major professional society guidelines to date:
  - The American College of Radiology (ACR) recommends that CT Chest should not be used for screening or as a first-line test to diagnose COVID-19.
  - The Centers for Disease Control and Prevention (CDC) recommends viral testing as the only specific method of diagnosis.
  - The CDC has stated that symptoms may appear 2-14 days after exposure to the virus. These symptoms may include:
    - fever or chills
    - cough
    - shortness of breath or difficulty breathing
    - fatigue
    - muscle or body aches
    - headache
    - new loss of taste or smell
    - sore throat
    - congestion or runny nose
    - nausea or vomiting
    - diarrhea
  - The Fleischner Society consensus statement published on April 7, 2020, recommends against the use of imaging in individuals with suspected COVID-19 who are either asymptomatic or have only mild symptoms without evidence of significant pulmonary dysfunction or damage (e.g., absence of hypoxemia, no or mild dyspnea).
  - According to The American Society of Transplantation, screening donors is based on methods below. Screening donors encompasses three different methods.
    - Epidemiologic screening for travel and potential exposures
    - Screening for symptoms suggestive of COVID-19
    - Viral testing (nucleic acid testing of specimens)
    - Screening asymptomatic donors with advanced imaging is not medically necessary.

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Other Chest Infections (CH-14)

Guideline

PPD or TB (Mycobacterium tuberculosis and Nontuberculous Mycobacterial Pulmonary Disease (NTM-PD)) (CH-14.1)
Fungal Infections (Suspected or Known) (CH-14.2)
Wegener's Granulomatosis/Granulomatosis with Polyangiitis and Related Entities (CH-14.3)
Suspected Sternal Dehiscence (CH-14.4)
References (CH-14)

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PPD or TB (Mycobacterium tuberculosis and Nontuberculous Mycobacterial Pulmonary Disease (NTM-PD)) (CH-14.1)

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- CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) with ANY of the following:
  - Normal or equivocal chest x-ray with ONE of the following:
    - Positive PPD skin test or other positive tuberculin skin tests OR
    - Positive QuantiFERON-TB Gold OR
    - Suspected active (or reactivated) tuberculosis
  - Suspected NTM-PD (i.e., atypical mycobacterium infection, Mycobacterium avium complex (MAC))
  - If CT Chest is unremarkable, there is insufficient data to support performing subsequent CT Chest unless symptoms develop or chest x-ray shows a new abnormality.
  - Suspected complications or progression of tuberculosis (e.g., pleural tuberculosis, empyema, and mediastinitis)
  - For those with known TB, follow-up CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) with frequency at the discretion of or in consultation with the pulmonary or infectious disease specialist (not to exceed 3 studies in 3 months).
  - Re-evaluate individuals undergoing active treatment who had abnormalities seen only on CT Chest.

Evidence Discussion

Nontuberculous mycobacterial pulmonary disease (NTM-PD) can be caused by a wide array of pathogens, with the most common including Mycobacterium avium complex (MAC), Mycobacterium kansasii, Mycobacterium xenopi, and Mycobacterium abscessus.

Chest radiography (CR) should be the initial imaging for suspected active M. tuberculosis (MTB) infection based on clinical symptoms and demographics, a newly positive tuberculin skin test (TST), or interferon-gamma release assay (IGRA). CT is medically necessary if CR is equivocal and there is clinical suspicion of active MTB, especially in those with impaired cell-mediated immunity.[11,26] CT may be performed to evaluate suspected complications and monitor response to therapy.[10,12]

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CR is medically necessary to distinguish latent from active MTB in people with evidence of new exposure (a newly positive TST/IGRA or a positive TST/IGRA with unknown prior status) but no clinical symptoms.[1] The yield of CR for active MTB in the absence of clinical symptoms is low. CT is more sensitive than CR for the detection of latent TB.[13,26] CT is recommended when CR is equivocal for active MTB or when a diagnosis of latent MTB may affect future care.[1,14]

When a TST is not available for people who are going to live in a group home, correctional institution or nursing facility, CR is medically necessary as a surrogate screening measure.

Imaging (CR and CT) is an important component in the diagnosis and follow up of nontuberculous mycobacterial pulmonary disease (NTM-PD).[16] The diagnosis and determination of response to therapy are based upon radiologic, clinical, and microbiologic criteria.[17,18] Serial CT imaging is important for monitoring disease progression and response to therapy. Radiologic findings provide prognostic information and may affect treatment recommendations.[18]

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Fungal Infections (Suspected or Known) (CH-14.2)

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- CT Chest with contrast (CPT® 71260) or High resolution CT Chest (HRCT) without contrast (CPT® 71250):
  - Initial diagnosis of any fungal pneumonia or chest infection
  - Suspected complications or progression of the fungal chest infection (e.g., worsening pneumonitis; pleural effusion, empyema, mediastinitis)
  - Suspected Allergic Bronchopulmonary Aspergillosis (ABPA) in asthmatics with atypical presentation or poor response to conventional therapy
- Follow-up CT Chest with contrast (CPT® 71260) or High Resolution CT Chest (HRCT) without contrast (CPT® 71250) with frequency at the discretion of or in consultation with the pulmonary or infectious disease specialist.
- For suspected fibrosing mediastinitis:
  - CT chest with contrast (CPT® 71260) (preferred imaging) OR
  - MRI Chest without and with contrast (CPT® 71552)
  - CTA chest (CPT® 71275) for:
    - Procedural planning
    - Evaluation of vascular stents

Evidence Discussion

Pulmonary fungal infections can be caused by a wide array of pathogens, including the endemic mycoses (Histoplasma species, Blastomyces species, and Coccidioides species), Cryptococcus species, and Aspergillus species.

CT chest is the imaging method of choice for suspected or known pulmonary fungal infections, especially in immunocompromised hosts.[19,20,30] Imaging findings are not specific but can lead to early detection of infection, help direct further diagnostic procedures and narrow the differential diagnosis.[3,20] CT is also used to monitor response to therapy and identify complications.[19] The diagnosis of certain pulmonary fungal infections and determination of response to treatment require a combination of clinical, microbiologic and radiologic criteria.[21-23] Denning et al recommend follow up imaging 3-6 months after starting anti-fungal therapy for chronic pulmonary aspergillosis, then less frequently, or with any major change of clinical status based on the fact that radiologic change is slow and little change is visible on chest X-ray (CR) or CT in less than 3 months.[23]

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CT is not medically necessary in the routine evaluation of suspected asthma without a specific indication but may be of value to identify acute complications following a nondiagnostic CR with suspected alternative diagnoses or associated conditions, such as allergic bronchopulmonary aspergillosis.[7,8,28,29] Serological assays can also play a role in the diagnosis of invasive pulmonary aspergillosis, including the galactomannan and beta-d glucan (Fungitell) assays.

Fibrosing mediastinitis (FM) is a rare benign but potentially life threatening process characterized by progressive proliferation of dense infiltrative fibrous tissue in the mediastinum and/or hila. Granulomatous FM is the more common subtype in the US and is typically associated with prior infection with histoplasmosis. Nongranulomatous FM is less common and is usually associated with an autoimmune disorder, drug exposure, or prior radiation. The most common symptoms are cough, dyspnea, hemoptysis, and pleuritic chest pain. Individuals may develop SVC syndrome or post-obstructive pneumonia. Widening of the mediastinum and mediastinal/hilar lymphadenopathy are the most common abnormal findings on chest x-ray. CT chest with contrast is the modality of choice for suspected FM. MRI is equivalent to CT for evaluating extent of disease, but CT is superior for evaluation of airways and calcification. PET CT is nonspecific and not routinely used. Nonsurgical and surgical procedures for individuals with clinical symptoms are related to compression or obstruction of mediastinal structures. CTA chest may be used for procedural planning or to evaluate the patency of vascular stents. There are no consensus guidelines for follow up imaging. No routine imaging is medically necessary for the granulomatous subtype unless there are progressive symptoms or clinical decompensation. Regular surveillance with CT is recommended for the nongranulomatous subtype to rule out malignancy which may not have been sampled in a biopsy.[32]

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Wegener's Granulomatosis/ Granulomatosis with Polyangiitis and Related Entities (CH-14.3)

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- See **Small Vessel Vasculitis (PVD-6.11)** for concerns of Wegener's Granulomatosis and Related Entities in Peripheral Vascular Disease imaging guidelines.

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Suspected Sternal Dehiscence (CH-14.4)

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- Sternal wound dehiscence is primarily a clinical determination.
- Chest x-ray is performed prior to advanced imaging to identify abnormalities in the sternal wire integrity and/or a midsternal stripe. Other findings include rotated, shifted, or ruptured wires.
- CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) for:
  - differentiating sternal wire migration from sternal dehiscence
  - planned debridement and/or repair

See **Infection (MS-9.0)** for concerns for osteomyelitis or soft tissue infection.

Evidence Discussion

Sternal dehiscence is defined as sternal separation with intact sternal wires migrating with a displaced sternal fragment.[24] The diagnosis is often made clinically; however, early signs may be subtle, and it may be clinically occult.. Early detection of sternal dehiscence on chest x-ray (CR) is important.[24] CT may be used in equivocal cases to assess for sternal separation or for preoperative planning.[24,25,31]

CT provides the best evaluation of sternal non-union when suspected based on pain, clicking and clinical evidence of sternal instability for > 3 months in the absence of infection.[24]

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Sarcoid (CH-15)

Guideline

Sarcoid (CH-15.1)
References (CH-15)

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Sarcoid (CH-15.1)

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- CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) is medically necessary to:
  - Establish or rule out the diagnosis when suspected
- Subsequent CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250), in known sarcoidosis, for ANY of the following:
  - Development of worsening symptoms
  - New symptoms appear after a period of being asymptomatic
  - Treatment change is being considered
- If CT is equivocal, definitive diagnosis can only be made by biopsy.
- PET/CT should not be used in the standard work-up of all sarcoidosis individuals. There is currently no evidence to support the use of PET/CT for screening.
- PET/CT (CPT® 78815) is medically necessary under the following conditions:
  - Help guide biopsy location if:
    - known lesion on CT Chest is difficult to access, to help identify alternative biopsy location
    - no apparent lung involvement and to identify an extrapulmonary biopsy site
  - Differentiation of reversible granulomatous disease from irreversible pulmonary fibrosis and will affect treatment options
  - Help identify treatment failure where either current treatment will be modified or new treatment will be introduced

Evidence Discussion

Sarcoidosis is a multisystem disease of unknown etiology characterized by the formation of noncaseating granulomas in various organs.[4] The diagnosis is based on three major criteria: a compatible clinical presentation, the finding of nonnecrotizing granulomatous inflammation in a tissue sample, and the exclusion of alternative causes of granulomatous disease.[5] Imaging plays an important role in the diagnosis. Although chest x-ray (CR) is often the first imaging test used, high resolution chest CT (HRCT) is more sensitive than CR for the detection of nodules and subtle fibrosis.[4] Histologic examination of tissue remains the gold standard for reaching a definitive diagnosis.[6] However, in the appropriate clinical context, certain patterns of mediastinal and parenchymal involvement on HRCT are virtually diagnostic of sarcoidosis.[6] The American Thoracic Society (ATS) states that if asymptomatic bilateral hilar lymphadenopathy is found on chest imaging, histologic confirmation is not always required.[5]

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The monitoring of individuals with pulmonary sarcoidosis is not standardized. Changes in imaging along with clinical features have been used to assess changes in disease activity.[7] If spirometry and pulmonary symptoms are worsening, additional chest imaging may be useful to detect progression of pulmonary disease or an alternative diagnosis.[8] HRCT can also provide prognostic information by differentiating reversible from irreversible (i.e., fibrotic) lesions and show complications, such as mycetomas or evidence of pulmonary hypertension.[6]

There is interest in the use of FDG PET CT for the diagnosis and monitoring of sarcoidosis. PET CT may reveal a more easily accessible biopsy site which is not clinically evident.[4,7] It may detect multi-organ and/or extra-thoracic involvement and demonstrate active inflammation not easily recognized by physical exam or other methods.[7,9] Studies have shown that FDG uptake in sarcoidosis represents active granulomatous inflammation.[7] The evaluation of disease activity is valuable when there is doubt regarding the activity of lesions and a change in therapy is being considered.[6] Positive scans should be interpreted with caution, however, because FDG uptake can be present in other inflammatory processes and malignancy. A significant correlation between decreased metabolic activity in the lungs, increased pulmonary function tests and improved symptoms in response to immunosuppressive medication has been demonstrated.[7,9] Most of the data regarding PET CT and sarcoidosis come from retrospective studies. Prospective trials are needed to determine the role of PET CT in monitoring the efficacy of therapy and the importance of abnormal PET CT's in asymptomatic individuals.[9] The threshold SUV that distinguishes active disease from fibrosis has not been determined. Few studies have compared the value of HRCT vs PET CT for diagnosis. Data on appropriate time intervals for follow up assessments and the role of PET-guided therapy are scarce.[9]

Sarcoid Health Equity Considerations

- Sarcoidosis occurs 2.5 times more often in Black individuals and females.
- Individuals who are Black, female, and/or have a low socioeconomic status report lower health-related quality of life, as well as increased rates of symptoms, mortality, and hospitalizations related to sarcoidosis.
- The unequal distribution of care and resources between rural/urban locations and different states may impact the quality of care and health outcomes related to sarcoidosis.

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Solitary Pulmonary Nodule (SPN) (CH-16)

Guideline

Solitary Pulmonary Nodule (CH-16.0)
Solitary Pulmonary Nodule – Imaging (CH-16.1)
Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)
Interval Imaging Outcomes (CH-16.3)
PET (CH-16.4)
References (CH-16)

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Solitary Pulmonary Nodule (CH-16.0)

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- For Lung Cancer Screening (LDCT) including incidental findings from LDCT, See **Lung Cancer Screening (CH-33)**

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Solitary Pulmonary Nodule – Imaging (CH-16.1)

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- For these guidelines, manual nodule measurements should be based on the average of long- and short-axis diameters. The size threshold (<6mm) corresponds to a rounded measurement of 5mm or less in these guidelines. Measurements should be rounded to the nearest millimeter. Prediction models used to estimate malignancy yield better results with the average diameter than with the maximum transverse diameter. The dimension of small pulmonary nodules (<10mm) should be expressed as the average of the maximal long-axis and perpendicular maximal short-axis measurements in the same plane. For larger nodules and for masses larger than 10mm, it is generally appropriate to record both long- and short-axis dimensions, with the long-axis dimension being used to determine the T factor in lung cancer staging and being a criterion for tumor response to treatment.
- A pulmonary nodule can be determined to have changed in size when its average diameter has increased or decreased by at least 2mm (rounded to the nearest millimeter). Smaller changes do not reliably indicate change.
- Maximum intensity projection (MIP), and Minimum intensity projection (MinIP) are 2D projections of the volumetric (3D) acquisition data. These projections may be of use in evaluation of pulmonary nodules, but these projections are included in the cross sectional imaging base codes, and is not separately reimbursable.
- EITHER CT Chest with contrast (CPT® 71260) OR CT Chest without contrast (CPT® 71250) is medically necessary initially for discrete nodule(s) in the following scenarios:[1,2,3]
  - Lung nodule(s) seen on an imaging study other than a “dedicated” CT or MRI Chest. Examples of other studies:
    - Chest x-ray
    - CT abdomen
    - MRI spine
    - Coronary CTA (unless full lung field are noted to be included)
  - But NOT in the following which are considered initial dedicated advanced chest imaging:
    - CT Chest without and with contrast (CPT® 71270)
    - CTA Chest (CPT® 71275)
    - MRI Chest without contrast (CPT® 71550)
    - MRI Chest without and with contrast (CPT® 71552)
    - MRA Chest without and with contrast (CPT® 71555)

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- Comparisons should include the earliest available study and the more recent previous CT Chest scans to determine if nodule was present and stable.
  - Similar-sized pleural nodule(s) is treated as a pulmonary nodule(s)
- The size of the lung or pleural nodule(s) is crucial information for decisions making regarding follow-up. The largest of multiple lung and/or pleural nodules will guide the surveillance interval. (See **Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)**, and **Pleural-Based Nodules and Other Abnormalities (CH-17.1)**
- A lung nodule is defined as an approximately rounded opacity (more or less well-defined) measuring up to 3cm in diameter.
- Rounded lesions measuring more than 3cm in diameter are termed lung masses and should be considered indicative of lung cancer until histologically proven otherwise. Approach to lung masses differ from that of nodules and these guidelines are specifically for those abnormalities, occurring within the chest, that meet the definition of a pulmonary nodule(s).

Evidence Discussion

A pulmonary nodule is defined as a well or poorly defined rounded opacity < 3cm in diameter. Focal pulmonary lesions > 3cm are considered masses. Nodule measurement is currently determined by standard linear measurement with electronic calipers. Measurements and averages should be expressed to the nearest whole millimeter. The dimension of small pulmonary nodules (<10mm) should be expressed as the average of the maximal long-axis and perpendicular maximal short-axis measurements in the same plane. For larger nodules and masses, both long- and short-axis dimensions should be recorded.[10] Semi-automated nodule volumetry has superior sensitivity for detecting growth and is recommended as the preferred method by the British Thoracic Society (BTS), but it requires dedicated software and is currently not widely used in clinical practice.[10,12] Nodule growth is defined as an increase of > 1.5mm (> 2 mm^3) by the Lung-RADS criteria, greater than or equal to 2mm change in average diameter by the Fleischner criteria or an increase of at least 25% in volume by the BTS.[1,12,14] A number of studies have established the advantage of post-processing 3D CT techniques, such as maximum intensity projection (MIP), minimum intensity projection (MinIP) and volume rendering (VR) in the detection and assessment of pulmonary nodules.[9,12,15]

If an indeterminate nodule is seen on a CR or CT, prior studies should be reviewed to determine possible growth or stability, including comparison with the earliest available study and more recent ones.[1-3] If stability of a nodule seen on CR cannot be determined, CT chest is appropriate.[2,3] CT is the modality of choice to evaluate pulmonary nodules.[3] Intravenous contrast is not required to identify or characterize nodules.[18] The size of the nodule is crucial for determining the appropriate timing of follow up surveillance imaging.[1]

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For nodules which are detected incidentally on incomplete thoracic CT scans (e.g., cardiac, neck, spine or abdominal CT), the Fleischner society recommends no follow up for most nodules < 6mm based on the estimated low risk of malignancy. The ACR states that an optional follow up CT may be done at 12 months for nodules < 6mm with a suspicious morphology and/or upper lobe location.[3] For nodules 6-8mm, Fleischner and ACR guidelines recommend a CT chest after the appropriate interval (3-12 months, depending on clinical risk). For a nodule > 8mm or a very suspicious nodule, an immediate CT chest is recommended.[1,3]

Background and Supporting Information

Abnormality examples include: mass, opacity, lesion, density, nodule, and calcification.

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Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)

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Solid Pulmonary Nodules

- These time intervals refer to the time from initial detection of the nodule(s).

Incidentally Detected Solid Pulmonary Nodules Follow-up Recommendations				
Nodule Type	**<6mm (<100 mm ^{3})**	**6–8mm**	**>8mm**	**Comments**
Single Nodule	Follow-up (optional) CT at 12 months. No routine follow-up if stable at 12 months	CT at 6–12 months, then CT at 18–24 months if stable *For an 8mm solid nodule PET request, see CH-16.4	CT at 3 months, then CT at 6-12 and then at 18-24 months if stable. Consider PET/CT* or biopsy	Certain individuals at high-risk with suspicious nodule morphology, upper lobe location, or both may warrant 12-month follow-up
Multiple Nodules	Follow-up (optional) CT at 12 months. *No routine follow-up if stable at 12 months	CT at 3–6 months, then at 18–24 months if stable	CT at 3–6 months, then at 18–24 months if stable. Consider PET/CT* or biopsy	Use most suspicious nodule as a guide to management. Follow-up intervals may vary according to size and risk.

- *PET/CT consider for ≥8mm solid lung nodule or solid component of a sub-solid nodule, not for groundglass opacity.
- Follow-up indications after PET/CT:

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  - If a PET/CT was found to be negative, follow-up with CT at 3 months, 9 months, and 21–24 months, if stable.
  - If a PET/CT was found to be positive, a biopsy was negative, non-diagnostic, or biopsy not performed, follow-up with CT at 3 months, 9-12 months, and 24 months, if stable.
- These criteria are not intended for use in the following groups:
  - Individuals aged 35 years or younger
    - Considered to have an overall low risk for pulmonary malignancy
    - In this age group, nodules are most likely to be infectious rather than cancer
    - Management of incidentally-found pulmonary nodules in this group should be individualized
  - Known primary cancer with risks for metastases
  - Immunocompromised individuals at risk for infection

Sub-Solid Pulmonary Nodules

- These time intervals refer to the time from initial detection of the nodule(s).

Incidentally Detected Sub-Solid Pulmonary Nodules Follow-up Recommendations			
Nodule Type	**<6mm (<100mm ^{3})**	**≥6mm (≥100mm^3)**	**Comments**
Single Ground glass opacity (GGO)	Consider follow-up at 2 and 4 years. If solid component(s) or growth develops, consider resection.	CT at 6–12 months to confirm persistence, then follow-up with CT every 2 years until 5 years	In certain suspicious nodules, <6mm, consider follow-up at 2 and 4 years. If solid component(s) or growth develops, consider resection.

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Incidentally Detected Sub-Solid Pulmonary Nodules Follow-up Recommendations			
Single Sub-solid	Consider follow-up at 2 and 4 years. If growth develops, consider resection.	CT at 3–6 months to confirm persistence. If unchanged and solid component remains <6mm, then annual CT should be performed for 5 years. If the solid component has suspicious morphology (i.e., lobulated margins or cystic components), is >8mm or is growing: Consider PET/CT* or biopsy	In practice, part-solid nodules cannot be defined as such until ≥6mm. Persistent part-solid nodules with solid components ≥6mm should be considered highly suspicious.
Multiple Sub-Solid	CT at 3–6 months. If stable, consider CT at 2 and 4 years.	CT at 3–6 months. Subsequent management based on the most suspicious nodule(s).	Multiple <6mm pure ground-glass nodules are usually benign.

• *PET/CT consider for ≥8mm solid lung nodule or solid component of a sub-solid nodule, not for groundglass opacity.

- Follow-up indications after PET/CT:
  - If a PET/CT was found to be negative, follow-up with CT at 3 months, 9 months, and 21–24 months, if stable.
  - If a PET/CT was found to be positive, a biopsy was negative, non-diagnostic, or biopsy not performed, follow-up with CT at 3 months, 9-12 months, and 24 months, if stable.
- These criteria are not intended for use in the following groups:
  - Individuals aged 35 years or younger
    - Considered to have an overall low risk for pulmonary malignancy
    - In this age group, nodules are most likely to be infectious rather than cancer
    - Management of incidentally-found pulmonary nodules in this group should be individualized
  - Known primary cancer with risks for metastases

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  - Immunocompromised individuals at risk for infection
- Sub-solid nodules may either be a part-solid nodule, comprising of both solid and ground glass components or a pure ground glass nodule, the latter may also be referred to as "non-solid".
  - Follow-up after year 5 is medically necessary every 2-4 years for at least 10 years if requested.
- For pulmonary nodule follow-up studies a CT Chest without IV contrast (CPT® 71250) is medically necessary. IV contrast is medically necessary as requested.

Pulmonary Cyst(s)

- May represent a rare form of adenocarcinoma, squamous cell carcinoma, or small cell carcinoma.
- Short-term initial imaging to exclude rapid growth is medically necessary at 3-6 months.
- Further imaging can be managed according to the sub-solid pathway above.

Endobronchial Lesion

- CT chest without contrast (CPT® 71250) at 3 months to confirm resolution or persistence
- If persistent, direct visualization with bronchoscopy is medically necessary

Evidence Discussion

A pulmonary nodule is defined as a well or poorly defined rounded opacity < 3cm in diameter. The Fleischner Society guidelines for the management of incidental pulmonary nodules detected on CT were last updated in 2017.[1] The purpose of the guidelines is to minimize both the number of unnecessary follow up exams and the chance of a malignancy advancing in stage during CT follow up prior to diagnosis. Surveillance is most appropriate if there is a very low probability of cancer or a high risk of complications from surgery or biopsy.[2] It is important to establish the clinical probability of malignancy before ordering imaging.[1,2,12] The Fleischner guidelines are not intended to apply to people younger than 35, people with known primary cancers at risk of metastases or to immunocompromised people at risk of infection. They do not apply to individuals with unexplained fever or respiratory symptoms.[3] For individuals younger than 35, lung cancer is rare. Management should be on a case by case basis, and the use of serial CT's should be minimized.[1]

Solid nodules < 6mm do not require follow up in individuals at low risk of lung cancer or in all individuals at high risk. Nodules which have a suspicious morphology or an upper lobe location may be followed up with a CT in 12 months. Solid nodules 6-8mm may be followed with a CT at 6-12 months in low risk individuals with a further follow up at 18-24 months in high risk individuals. Two follow up CT's should be sufficient to rule out growth in most individuals.

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For solid nodules >8 mm, the options are CT surveillance, an FDG PET/CT, tissue sampling or a combination of these. Surveillance CT scans for solid nodules >8 mm may be done at 3 months, 6-12 months and 18-24 months.[1] The American College of Chest Physicians (ACCP) guidelines recommend a PET/CT or nonsurgical biopsy for solid nodules of at least 8mm when the pretest probability of malignancy is low to moderate.[2] The pretest probability affects the interpretation of PET/ CT results: high risk individuals are at risk of false negative results and low risk individuals are at risk of false positive results.[12] If there is a high pretest probability of cancer, a negative PET/ CT does not reliably rule out cancer and either continued surveillance for at least 2 years or tissue sampling is advised.[2,13] The ACCP states that the optimal interval for surveillance CT's for solid nodules >8mm is not determined, but standard practice is 3-6 months, 9-12 months and 18-24 months. They suggest surveillance with CT if the clinical probability of cancer is very low, the clinical probability of malignancy is low and a PET CT is negative, a PET CT is negative and a needle biopsy is non-diagnostic, or an informed individual prefers a non-aggressive approach. If a solid nodule shows clear growth on serial CT's, a non-surgical biopsy or surgical resection is recommended unless there are specific contraindications.[2] A surgical diagnosis is recommended if there is a high clinical probability of lung cancer, the nodule is intensely hypermetabolic on PET/CT, a non-surgical biopsy is suspicious for cancer or an individual prefers a definitive diagnosis.[2]

Multiple solid nodules <6mm are usually benign, representing granulomas or intrapulmonary lymph nodes. A 12 month follow up CT may be considered in high risk individuals. If there is clinical evidence of infection or the individual is immunocompromised, infection should be considered. A short term follow CT may be appropriate. Multiple solid nodules with at least one nodule greater than or equal to 6 mm can be followed with CT's at 3-6 months and 18-24 months. Management should be based on the largest/most suspicious nodule. Most metastases will grow over 3 months. The risk of cancer increases as the number of nodules increases from 1 to 4 but decreases if the number is greater than 4.

Subsolid nodules (SSN) include pure ground glass nodules (GGN) and part-solid nodules (PSN). SSN are more likely to be malignant than solid nodules but have a better prognosis than lung cancers which present as solid nodules.[12] Many have slow growth rates and may remain stable for years. Pure GGN < 6mm do not require routine follow up. However, this should not preclude the option of follow up CT's at 2 and 4 years in high risk individuals. GGN greater than or equal to 6mm can be followed at 6-12 months and then every two years until 5 years. The Fleischner Society states that these guidelines are not intended to preclude either shorter or longer term follow up in individuals when deemed clinically appropriate. A previous study found growth in 7% of part-solid nodules that had previously been stable for 3 years, and another study found growth >2mm in 13% of nodules after 5 years of stability.[21] The ACCP states there is controversy regarding how long to follow part solid or ground glass nodules and

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that follow up over several years may be appropriate. In appropriate individuals, follow-up after 5 years, every 2-4 years, can be considered for at least 10 years, taking into account comorbidities, life expectancy, and individual preferences.[21]

Solitary PSN < 6mm do not require routine follow up. A follow up CT may be done at 2 and 4 years. For PSN greater than or equal to 6mm with a solid component < 6mm, a follow up CT may be done at 3-6 months and then annually for a minimum of 5 years. The 5 year period is somewhat arbitrary but considered reasonable if the nodule is unequivocally stable during that time period. If the solid component is at least 6mm, follow up at 3-6 months should be done. A persistent PSN with a solid component of at least 6mm or a growing solid component is highly suspicious. If the nodule has suspicious morphology, if the solid component is growing or > 8mm, PET/CT or biopsy should be considered.[1,12] Multiple PSN < 6mm are often infectious. A repeat CT can be done at 3-6 months, then at 2 and 4 years. If at least one of the nodules is greater than or equal to 6mm, a repeat CT can be done at 3-6 months, and management should be based on the most suspicious nodule.

The volume of lung nodules is significantly larger in CT chest with contrast compared to CT chest without contrast. Whichever CT protocol is used initially should also be used in all follow-up scans to avoid the risk of missing any relevant nodule growth.[24,25]

Pulmonary cystic lesions may represent a cyst-related primary lung malignancy. There are no uniform surveillance criteria for these lesions, but some authors recommend a CT at 3-6 months to exclude rapid growth and then follow up CT's according to the SSN nodule guidelines.[8]

NCCN guidelines for the management of incidental pulmonary nodules are consistent with the Fleischner guidelines.[6,7] For pulmonary nodules detected on lung cancer screening CT's, adherence to the American College of Radiology (ACR) Lung-RADS guidelines is recommended.[1,3,6,7,14] The British Thoracic Society and ACCP guidelines do not distinguish the management of screening-detected nodules from nodules detected incidentally.[2,12]

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Interval Imaging Outcomes (CH-16.3)

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- Further advanced imaging is not medically necessary if a nodule has been:
  - Stable for 2 years
    - Nodules(s) stable on chest x-ray
    - Nodule(s) ≥6mm stable on CT Chest
  - Stable for 1 year
    - Nodule(s) <6mm
  - At any time, if:
    - classically benign characteristics by chest x-ray or previous CT (e.g., benign calcification pattern typical for a granuloma or hamartoma)
    - decreasing in size, (≥ 6mm at start, should be followed for a 2 year period as outlined in CH-16.2) disappearing nodule(s)
- For clinically significant lung nodule(s) (lung nodules not meeting the above criteria) which increase in size or number, serial CT screening or surveillance are not medically necessary.
  - In individuals who do not meet PET criteria (per CH-16.4) or in individuals in which biopsy is not being considered, serial CT may be performed per CH-16.2.

Evidence Discussion

If a chest x-ray (CR) or chest CT has demonstrated that a pulmonary nodule has benign characteristics, further imaging is not medically necessary. Benign characteristics include intranodular fat or a diffuse, central, laminated or popcorn pattern of calcification.[2]

If an indeterminate nodule is seen on a CR or CT, prior studies should be reviewed to determine possible growth or stability, including comparison with the earliest available study and more recent ones.[1-3] If stability of a nodule seen on CR cannot be determined, CT chest is appropriate.[2,3] If a solid nodule has been stable for at least 2 years, no additional evaluation is necessary.[2] Two years of radiographic stability is considered strong presumptive evidence of a benign nodule. For solid nodules seen on CT, further follow up is not needed if nodules < 6mm have been stable for one year or if nodules greater than or equal to 6mm have been stable for two years.[1] Malignant nodules show a wide range of growth rates with some demonstrating regression at times. Solid nodules greater than or equal to 6mm that decrease in size but do not completely resolve should be followed to resolution or lack of growth over 2 years.[2,12] For situations where PET, CT, and/or biopsy are not reasonable choices, ongoing

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surveillance would be medically necessary. Clinically significant nodules that are not amendable to PET and/or biopsy may undergo serial CT exams per CH-16.2.

There is controversy regarding how long to follow part solid or ground glass nodules. Follow up over several years may be appropriate.[2] The Fleischner guidelines state that subsolid nodules <6mm may be followed for up to 4 years but that the guidelines are not intended to preclude either shorter or longer term follow up in individuals when deemed clinically appropriate.[1] Subsolid nodules greater than or equal to 6mm may be followed for 5 years. The 5 year period is "somewhat arbitrary but considered reasonable if the nodule is unequivocally stable during that time period."[1]

PET/CT should be considered for solid nodules greater than or equal to 8mm.[1,2] If a solid nodule shows clear growth on serial CT's, a non-surgical biopsy or surgical resection is recommended unless there are specific contraindications.[2] Non-solid nodules which grow or develop solid components are often malignant and further evaluation and/or resection should be considered.[2,14]

If a CT demonstrates multiple solid nodules <6mm and there is clinical evidence of infection or the individual is immunocompromised, infection should be considered and a short term follow up CT may be appropriate.[1] Certain findings on a lung cancer screening CT which suggest an infectious or inflammatory process (e.g., >6 new nodules or solid nodules which are greater than or equal to 8mm appearing in a short interval) are reported as Lung-RADS 0 and may be followed up with a LDCT in 1-3 months.[14] Some findings indicative of an infectious/inflammatory process may not warrant short-term follow-up (e.g., tree-in-bud nodules or new <3 cm ground glass nodules).

Background and Supporting Information:

- Approximately 20% of observed cancers have decreased in size at least at some point during their observation period. Therefore, a decreasing size of a nodule cannot be a reliable indicator of being benign.
- For nodules that increase in number, this is not meant for known stable or benign nodules to be counted.
  - Example, known 4mm nodule stable for 3 years, now presents with a new solid 8mm pulmonary nodule, follow-up will be driven by new nodule size and type.
  - Example #2, known granuloma 5mm from prior CT Chest one year ago and now CT Chest reveals a new 6 mm sub-solid nodule, follow-up would be driven by the new nodule size and type.

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PET (CH-16.4)

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- PET/CT (CPT® 78815) for a solid lung nodule ≥8mm on dedicated advanced chest imaging, as described in **Solitary Pulmonary Nodule – Imaging (CH-16.1).** See **Non-Small Cell Lung Cancer – Suspected/Diagnosis (ONC-8.2)** in the Oncology Imaging Guidelines for lung mass ≥3.1cm.
  - If there is a history of malignancy, refer to the appropriate Oncology restaging/ recurrence guideline for indications for PET imaging.
  - Pleural nodule, See **Pleural-Based Nodules and Other Abnormalities (CH-17.1)**
  - Serial PET studies are not medically necessary
  - Not medically necessary for infiltrate, ground glass opacity, or hilar enlargement
  - Mediastinal lymphadenopathy - See **Mediastinal Lymphadenopathy (CH-2.3)** or Sarcoid concerns – See **Sarcoid (CH-15.1)**
- If a CT finding led to ordering a PET scan, and if that CT was >3 months ago, a repeat CT (CPT® 71250) or (CPT® 72160) is medically necessary prior to considering a PET scan.
  - A change in the status of the original finding may find that a PET scan is no longer medically necessary.

Evidence Discussion

PET/CT may be performed for evaluation of a solid lung nodule greater than or equal to 8mm on chest CT.[1,2,6,7] PET/CT has good sensitivity and moderate specificity for detecting malignancy in individuals with a high risk of cancer and a nodule greater than or equal to 10mm.[12] Consensus opinion is that that nodules <8 to 10mm are not reliably characterized by PET/CT.[2,12] The false negative rate of PET/CT is higher for nodules <8mm and for malignancies with low metabolic activity, such as adenocarcinoma in situ or well differentiated carcinoid tumor. PET/CT has a lower sensitivity and higher false negative rate for ground glass or part solid nodules.[2,12] Infections and inflammatory disorders may cause false positive results.

Repeating a PET/CT is discouraged. If there is a high pretest probability of cancer, a negative PET/CT does not reliably rule out cancer and either continued surveillance for at least 2 years or tissue sampling is advised.[2,13] If a solid nodule shows clear growth on serial CT's, a non-surgical biopsy or surgical resection is recommended unless there are specific contraindications.[2] A surgical diagnosis is recommended if the nodule is hypermetabolic on PET/CT.[2]

PET/CT may be indicated for the pre-treatment staging of individuals with confirmed or strongly suspected lung cancer, as detailed in the oncology guidelines.[2,6,7]

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Background and Supporting Information

- A **nodule** is any pulmonary or pleural lesion that is a discrete, spherical opacity 2-30mm in diameter surrounded by normal lung tissue. A larger nodule is called a mass. Entities that are not nodules, and are considered benign, include non-spherical linear, sheet-like, two-dimensional or scarring opacities.
- **Malignant** nodule features can include spiculation, abnormal calcification, size greater than 7-10mm, interval growth, history of a cancer that tends to metastasize to the lung or mediastinum, and/or smoking history.
  - A nodule that grows at a rate consistent with cancer (doubling time 100 to 400 days) may be sampled for biopsy or resected.
  - Less than 1% of <6mm lung nodules are malignant.
  - Three percent of all 8mm lung nodules are malignant.
  - Only one follow-up at 6-12 months is sufficient for 6-8mm nodules and not all require traditional 2 year follow-up.
  - The larger the solid component of a subsolid nodule, the greater the risk of invasiveness and metastases.
  - Increased risk of primary cancer as the total nodule count increased from 1 to 4 but decreased risk in individuals with 5 or more nodules, most of which likely resulted from prior granulomatous infection.
  - A nodule that does not grow in 6 months has a risk of malignancy at <10%.
- **Benign** features in solid nodules can include benign calcification (80% granuloma, 10% hamartoma), multiple areas of calcification, small size, multiple nodules, negative PET, and stability of size over 2 years.
- **Ground glass** or subsolid opacities, which can harbor indolent adenocarcinoma with average doubling times of 3–5 years.
- **Repeat PET** is discouraged. If the original PET is positive, biopsy may be performed. If the original PET is negative but subsequent CT Chest shows an increase in nodule size, biopsy may be performed.
- **Positive PET** is defined as a standardized uptake value (SUV) in the lung nodule greater than the baseline mediastinal blood pool. A positive PET can occur with infection or inflammation, including absence of lung cancer with localized infection, presence of lung cancer with associated (post-obstructive) infection and/or related inflammation.
- **False negative PET** can be seen in individuals with adenocarcinoma in situ (formally known as bronchoalveolar carcinoma), carcinoid tumors, a small size nodule, non-solid or ground glass opacity. High pre-test likelihood of malignancy with negative findings on PET only reduces the likelihood of malignancy to 14%; while in an individual with a low pre-test likelihood (20%) of malignancy, a negative PET reduces the likelihood of malignancy to 1%.

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Pleural-Based Nodules and Other Abnormalities (CH-17)

Guideline

Pleural-Based Nodules and Other Abnormalities (CH-17.1)
Reference (CH-17)

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Pleural-Based Nodules and Other Abnormalities (CH-17.1)

CH.PB.0017.1.A

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- CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) (with contrast is preferred for initial evaluation) is medically necessary for pleural nodule(s).
  - Pleural nodule(s) seen on an imaging study other than a "dedicated" CT or MRI Chest.
  - Pleural nodule(s) identified incidentally on any of the following dedicated chest studies can replace CT Chest as the initial dedicated study.
    - CT Chest without and with contrast (CPT® 71270).
    - CTA Chest (CPT® 71275).
    - MRI Chest without contrast (CPT® 71550).
    - MRI Chest without and with contrast (CPT® 71552).
    - MRA Chest without and with contrast (CPT® 71555).
  - After preliminary comparison with any available previous chest films to determine presence and stability
  - Using largest measurement of multiple nodule(s). (See **Solitary Pulmonary Nodule – Imaging (CH-16.1)**)
  - Following the Fleischner Society Guidelines for high-risk. (See **Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)**)
- PET/CT (CPT® 78815) is medically necessary if dedicated CT or MRI Chest identifies a pleural nodule/mass or defined area of pleural thickening that is ≥8 mm when there is a likelihood of malignancy including current or previous malignancy, pleural effusion, bone erosion, chest pain.
- For diagnosis or suspicion of pulmonary sequestration:
  - CT chest with contrast (CPT® 71260) OR CTA Chest (CPT® 71275)
  - 3D reconstruction as requested (CPT® 76377) or (CPT® 76376)

Evidence Discussion

- CT Chest is medically necessary for the evaluation of pleural nodules.[1-3] CT scan is widely available and allows for easy access to isotropic 3-D reformatting.[3] A study looking at the utility of CT in investigation for malignancy showed a sensitivity of 68%, a specificity of 78%, a positive predictive value of 80% and a negative predictive value of 65% when CT findings were reported as malignant.[3] CT also carries the risk of exposure to iodinated contrast and ionizing radiation.

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- Follow up for previously detected pleural nodules follows guidelines addressed elsewhere in these guidelines (See **Solitary Pulmonary Nodule – Imaging (CH-16.1)** and **Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)**.[1]
  - Of note, a study looking at over 8,700 LDCT chest scans identified 943 noncalcified nodules attached to the costal pleura, of these 897 were < 10mm in size. There were 603 that were either lentiform, oval, semicircular or triangular in shape and had smooth margins. All of these nodules, that met these qualifications of shape, size and smooth margins, were benign. Follow-up with annual screening, rather than more immediate work-up, was recommended.[2]
- PET/CT may be considered when the identified pleural nodule/mass or thickening is ≥ 8mm and there is a likelihood of malignancy.[1] PET/CT may be useful in differentiating between benign and malignant disease; however, studies have shown a broad range of sensitivities (88-100%) and specifies (35-100%).[3] PET/CT may be complicated by false positives such as infections and prior pleurodesis with talc, or false negatives such as low grade/low metabolic activity epithelioid mesothelioma.[3]

Background and Supporting Information

- Pleural nodule/mass or thickening without suggestion of malignancy would undergo surveillance or biopsy.
- A study looking at over 8,700 LDCT chest scans identified 943 noncalcified nodules attached to the costal pleura, of these 897 were <10mm in size. There were 603 that were either lentiform, oval, semicircular or triangular in shape and had smooth margins. All of these nodules, that met these qualifications of shape, size and smooth margins, were benign. Follow-up with annual screening, rather than more immediate work-up, was recommended.

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Pleural Effusion (CH-18)

Guideline

Pleural Effusion (CH-18.1)
References (CH-18)

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Pleural Effusion (CH-18.1)

CH.EF.0018.1.A
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- CT Chest with contrast (CPT® 71260) is medically necessary after:
- Chest x-ray, (upright posterior/anterior/lateral best), (lateral decubitus films can improve sensitivity); **and**

  one of the following:

  - Thoracentesis, (if possible) to determine if fluid is exudative or transudative and remove as much as possible (this fluid can obscure the underlying lung parenchyma and possibly a mass)
  - Concern for loculated effusion, empyema, paramediastinal location, subpleural lung abscess or cavitation
  - Check position of chest tube(s) or drainage catheters
  - Surgical planning
- Chest ultrasound (CPT® 76604) can be used as an alternative or adjunct to chest x-ray to evaluate for the presence of fluid within the pleural spaces and guide thoracentesis.

Evidence Discussion

The most common initial diagnostic test suggesting a pleural effusion is often chest radiography (CR). CR remains the most accessible form of chest imaging and will often be the initial study for suspected pleural disease.[1,2] Lateral decubitus CR has higher sensitivity and specificity for pleural effusion than other positions.[3] However, complicated effusions are often loculated and may not layer dependently. Lower lobe consolidation may mask the presence of an effusion. The American Association for Thoracic Surgery (AATS) states that CR, although useful as a first step, should be combined with additional imaging if pleural space infection is suspected.[2,9] The American College of Radiology (ACR) states that consensus recommendations endorse CR as the initial imaging modality for suspected parapneumonic or malignant effusion, but there are limited empiric data to support this.

Ultrasound (US) is at least as effective as lateral decubitus views for the detection of pleural fluid and provides a better estimation of fluid volume.[1-3] When standard CR cannot rule out a pleural effusion, US has largely replaced decubitus views due to its speed, portability and greater sensitivity. Identification of a pleural effusion for possible US-guided thoracentesis is the primary reason for chest US. The AATS guidelines state that CR and US are class 1 recommendations (should be done) for suspected pleural space infection.[2] US is more sensitive than CR for detecting pleural effusion.[6,10]

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Although diagnostic imaging plays an important role in the evaluation of pleural effusions, thoracentesis with pleural fluid analysis remains the necessary first invasive step. Pleural fluid analysis is considered mandatory unless the clinical presentation suggests a high pretest probability of a transudative effusion.[1] Initial evaluation should include an ultrasound (US)-guided thoracentesis to categorize the effusion as a transudate or exudate and obtain specimens for microbiology and cytology.[4,5] If a parapneumonic effusion is suspected, diagnostic aspiration must be performed to identify individuals with a complicated effusion that requires drainage.[2,4]

CT is not used routinely as the initial imaging study for pleural effusion unless there is suspicion for loculated fluid in an interlobar fissure or paramediastinal location, or CR demonstrates parenchymal lesions suggestive of cancer, septic emboli or cavitation. CT can better distinguish between a loculated empyema and subpleural lung abscess. CT with IV contrast optimizes imaging of the pleura. CT chest with contrast is a class 2a recommendation (reasonable) for suspected pleural space infection in the AATS guidelines.[2] If the etiology of an exudative effusion cannot be identified, or if it is not safe to perform a thoracentesis, a CT chest with contrast is medically necessary.[4,5] The American College of Radiology (ACR) states that CR or CT chest with IV contrast is medically necessary as initial imaging for people with recent pneumonia and suspected parapneumonic effusion or for people with dyspnea, cough or chest pain with a suspected malignant pleural effusion. Thoracentesis and chest CT cannot rule out malignancy or tuberculosis. Pleural biopsy is indicated for a recurrent undiagnosed exudative effusion.[1,4,5] When a diagnosis cannot be made, monitoring with interval CT scans for up to 2 years is medically necessary.[5] CT is used in the diagnosis and management of late-stage empyema and malignant pleural effusion and can be used to check the position of drains and plan for surgical intervention.[1] CT scans have high specificity but low sensitivity in distinguishing benign from malignant pleural effusion.[6]

Background and Supporting Information

- Bilateral effusions are more often systemic related transudates (congestive heart failure, renal failure, liver insufficiency, etc.), and advanced imaging is rarely needed. Large unilateral effusions can be malignant. Analysis of fluid may include: cytology, culture, cell count, and biochemical studies.
- PA chest x-ray can show a pleural effusion with approximately 200 ml of pleural fluid while a lateral view can reduce this to 50 ml. Ultrasound is even more sensitive with as little as 3-5 ml of fluid being detected. *Thoracentesis can only be safely performed with adequate fluid present. If only a trace effusion or inadequate amount of fluid is seen, a thoracentesis may not be possible.

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Pneumothorax/ Hemothorax (CH-19)

Guideline

Pneumothorax/Hemothorax (CH-19.1)
Pneumomediastinum; Subcutaneous Emphysema (CH-19.2)
References (CH-19)

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Pneumothorax/Hemothorax (CH-19.1)

CH.PT.0019.1.A
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Chest x-ray and CT Chest are the first line tests for detecting pneumothorax/hemothorax and ruling out other lung diseases.

- Chest x-ray is medically necessary initially.
  - CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) if:
    - diagnosis of a small pneumothorax is in doubt, and the presence of a pneumothorax will affect individual treatment decisions
    - preoperative study for treatment of pneumothorax
    - pneumothorax associated with hemothorax
    - suspected complications from hemothorax (e.g., empyema)
    - suspected Alpha-1-Antitrypsin Deficiency (even without pneumothorax)
    - suspected cystic lung disease, including lymphangioleiomyomatosis (LAM), tuberous sclerosis (TS), or Birt-Hogg-Dube (BHD) syndrome
    - to determine the etiology of persistent pneumothorax/air leak, such as chest tube malposition, bronchopleural fistula, loculated pneumothorax, lung parenchymal disease
    - suspected catamenial pneumothorax/thoracic endometriosis
- MRI Chest without and with contrast (CPT® 71552) or MRI Chest without contrast (CPT® 71550) for:
  - detecting diaphragmatic endometriosis
  - pre-surgical planning for thoracic endometriosis

Evidence Discussion

The majority of pneumothorax cases can be confirmed by upright PA chest radiography (CR), which remains the standard initial exam.[16] While US has a high sensitivity/specificity for pneumothorax, it is operator-dependent and is not medically necessary as a primary imaging modality.[19-22]

While CT is more sensitive than CR in detecting pneumothorax, it is generally not medically necessary for diagnosis and should be avoided due to excess radiation.[16] CT may be medically necessary for diagnosing a very small pneumothorax or differentiating between a pneumothorax and a giant bulla in bullous emphysema. Although CT is the best method to measure the size of a pneumothorax, current evidence does not support basing treatment decisions solely on size.[14]

The 2001 ACCP guidelines advise against routine CT use for a first-time primary spontaneous pneumothorax (PSP). However, CT may be medically necessary

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to evaluate suspected pulmonary disorders not apparent on CR. For secondary spontaneous pneumothorax (SSP), CT is acceptable for managing recurrent pneumothorax, persistent air leak, and surgical planning.

In contrast, the 2023 British Thoracic Society recommends CT chest for individuals with symptoms and high-risk characteristics. These include hemodynamic compromise, significant hypoxia, bilateral pneumothorax, underlying lung disease, hemopneumothorax, or age over 49 with a significant smoking history. This recommendation applies if the pneumothorax size on CR is insufficient for safe needle aspiration or chest tube intervention. The European Respiratory Society suggests that CT may be useful in complicated cases, when chest tube misalignment is suspected, when underlying lung disease is suspected, and in individuals requiring surgery.[16]

High-resolution CT (HRCT) has better sensitivity than routine CT in the pre-operative detection of blebs and bullae.[14] However, it is unclear whether HRCT can predict the risk of recurrence or identify which individuals may benefit from surgical intervention. It may help to identify those at lower risk: the positive predictive value of CT bleb scores for ipsilateral recurrence is relatively low at 68%, while the negative predictive value is high at 94%.[15]

Some experts advise considering a CT scan after a first time PSP if there are factors such as a family history of pneumothorax, presence of blebs, cysts, or bullae; female sex; or a family or personal history and/or physical examination findings suggestive of a pneumothorax-associated syndrome.[18]

Although they have low specificity, the most sensitive tests for detecting pneumothorax and hemothorax are CR and CT. For detecting diaphragmatic endometriosis, Magnetic Resonance Imaging (MRI) of the chest is preferable.[5]

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Pneumomediastinum; Subcutaneous Emphysema (CH-19.2)

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- Chest x-ray is medically necessary initially.
  - CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) if:
    - recent vomiting and/or suspected esophageal perforation
    - associated pneumopericardium
    - associated pneumothorax
    - preoperative study for treatment

Evidence Discussion

The diagnosis of Pneumomediastinum (PM) is usually established by a clinical exam and CR. The CR should include a lateral view.[8] CR is the most common diagnostic imaging study.[9-11] The reported sensitivity of CR ranges from 60-90%.[9,12] CT is more sensitive than CR, especially in cases of small amounts of air in the mediastinum.[8]

There is no evidence defining when CT should be used to evaluate pneumomediastinum (PM). CT should be done if the suspicion for PM remains high despite a normal CR or if there is concern for secondary PM due to a specific pathologic event.[9]

CT can be beneficial in detecting injury to the tracheobronchial system, pneumothorax, pneumopericardium or esophageal perforation.[12] Despite the usually benign and self-limiting course of spontaneous PM, additional imaging is often undertaken to rule out esophageal perforation or other underlying disorder. A retrospective review of adolescents and young adults with spontaneous PM demonstrated that no clear criteria were used for obtaining a CT and that the CT's did not impact clinical decisions.[13] The authors concluded that advanced imaging is over-utilized in individuals with suspected spontaneous PM without clinical evidence of necessity.

Background and Supporting Information

- An expiration chest x-ray can enhance the evaluation of equivocal plain x-ray. There is no data supporting the use of serial CT Chest to follow individuals with a known pneumothorax, pneumomediastinum, or hemothorax who are asymptomatic or have stable symptoms. With the exception of the indications above, advanced imaging of the chest is not medically necessary in the diagnosis or management of pneumothorax, or pneumomediastinum. Inspiratory/expiratory chest x-rays are helpful in defining whether a pneumothorax is present.

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Mediastinal Mass (CH-20)

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Mediastinal Mass (CH-20.1)

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- CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) or MRI Chest without and with contrast (CPT® 71552) or MRI Chest without contrast (CPT® 71550) is medically necessary to evaluate mediastinal abnormalities (e.g., mediastinal cyst, including bronchogenic, thymic, pericardial, or esophageal) seen on chest x-ray or other non-dedicated chest imaging.
- MRI Chest without and with contrast (CPT® 71552) or MRI Chest without contrast (CPT® 71550) is medically necessary for indeterminate mediastinal mass on CT Chest.
  - Surveillance imaging is medically necessary no more frequently than 3-month intervals over 2 or more years when the lesion(s) remain indeterminate on MRI and a biopsy has not been performed.
- FDG PET/CT offers limited additional value beyond that of conventional CT in the initial assessment of mediastinal mass(es), with the exception of primary mediastinal lymphoma. See **Non-Hodgkin Lymphomas (ONC-27)** or **Hodgkin Lymphoma (ONC-28)** in the Oncology Imaging Guidelines. A positive FDG PET/CT has little value for discrimination between benign and malignant lesions. A negative FDG PET/CT does not prevent serial CT/MRIs, due to appreciable false negative rate.
  - MRI Chest without and with contrast (CPT® 71552) or MRI Chest without contrast (CPT® 71550) is medically necessary for indeterminate mediastinal mass on FDG PET/CT.
- CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) or MRI Chest without and with contrast (CPT® 71552) or MRI Chest without contrast (CPT® 71550) is medically necessary for subsequent evaluations if:
  - new signs or symptoms, or
  - preoperative assessment
- For Adenopathy; See **Lymphadenopathy (CH-2).**
- For Goiter; See **Thyroid Nodule (NECK-8.1)** in the Neck Imaging Guidelines.
- For Myasthenia Gravis; See **Neuromuscular Junction Disorders (PN-6.1)** in the Peripheral Nerve Disorders Imaging Guidelines.
- For fibrosing mediastinitis: See **Fungal Infections (Suspected or Known) (CH-14.2)**

Evidence Discussion

Mediastinal nodules or masses may present as incidental findings on chest radiographs and cross-sectional imaging. Alternatively, they may be found during the evaluation of symptoms and signs that include chest pain, cough, dyspnea, dysphagia, cardiac tamponade, diaphragmatic paralysis, central venous thrombosis, superior vena cava

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syndrome, B-symptoms (in lymphoma), myasthenia gravis, and other paraneoplastic syndromes. The incidence rate is low with a reported prevalence of 0.73-4%. The most frequent lesions encountered in the mediastinum are thymoma, neurogenic tumours and benign cysts.[2]

CT is superior to chest radiography for detection of invasion of the mass across tissue planes, secondary to its higher contrast resolution. Anterior mediastinal tumors account for 50% of all mediastinal masses. CT has the ability to show the precise location, morphology, and pattern of contrast enhancement of an anterior mediastinal mass as well as its relationship to other mediastinal components or borders.[2,6]

MRI remains superior to CT for detection of invasion of the mass across tissue planes, including the chest wall and diaphragm, and involvement of neurovascular structures secondary to its higher soft-tissue contrast. MRI allows further tissue characterization of mediastinal masses beyond that of CT and FDG-PET/CT. Chemical-shift MRI has been shown to be useful in distinguishing normal thymus and thymic hyperplasia from thymic neoplasms and lymphoma. It can also prove the cystic nature of an indeterminate, non–water attenuation thymic mass on CT, preventing unnecessary biopsy and thymectomy. The frequency of surveillance imaging for indeterminate lesions seen on MRI is usually 3-12 month intervals over 2 years or more, depending upon level of clinical concern.[2,4,6] The diagnostic accuracy of chest MRI for the detection of mediastinal mass is 71-91%.[4]

(FDG)-PET/CT offers limited additional value beyond that of conventional CT in the initial assessment of mediastinal masses, with the exception of its use for primary mediastinal lymphoma staging and surveillance and detection of metastatic lymphadenopathy. A positive FDG-PET/CT has little value for discrimination between benign and malignant lesions.[2,3,5,6]

It is reasonable to perform a chest radiograph as an initial imaging step. Chest radiography can help localize a mass to a specific mediastinal compartment and thereby narrow the differential diagnosis. Chest radiography offers limited assistance regarding tissue characterization of mediastinal masses, with the exception of its occasional demonstration of calcium within a lesion.[2]

There is little relevant literature to support the use of ultrasound (US) in the initial evaluation of a clinically suspected mediastinal mass.[2]

Fibrosing mediastinitis (FM) is a rare benign but potentially life threatening process characterized by progressive proliferation of dense infiltrative fibrous tissue in the mediastinum and/or hila. Granulomatous FM is the more common subtype in the US and is typically associated with prior infection with histoplasmosis. Nongranulomatous FM is less common and is usually associated with an autoimmune disorder, drug exposure, or prior radiation. The most common symptoms are cough, dyspnea, hemoptysis, and pleuritic chest pain. Individuals may develop SVC syndrome or post-obstructive pneumonia. Widening of the mediastinum and mediastinal/hilar

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lymphadenopathy are the most common abnormal findings on chest x-ray. CT chest with contrast is the modality of choice for suspected FM. MRI is equivalent to CT for evaluating extent of disease, but CT is superior for evaluation of airways and calcification. PET CT is nonspecific and not routinely used. Nonsurgical and surgical procedures for individuals with clinical symptoms are related to compression or obstruction of mediastinal structures. CTA chest may be used for procedural planning or to evaluate the patency of vascular stents. There are no consensus guidelines for follow up imaging. No routine imaging is medically necessary for the granulomatous subtype unless there are progressive symptoms or clinical decompensation. Regular surveillance with CT is recommended for the nongranulomatous subtype to rule out malignancy which may not have been sampled in a biopsy.[7]

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Chest Trauma (CH-21)

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Chest Trauma (CH-21.1)

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- Chest x-ray is medically necessary initially.
  - CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is medically necessary for the following situations:
    - Rib or sternal Fracture when clinical findings or other imaging identifies associated complications including pneumothorax, hemothorax, pulmonary contusion, atelectasis, flail chest, cardiovascular injury and/or injuries to solid or hollow abdominal organs.
    - Rib or sternal facture when malignancy is suspected as the etiology for uncomplicated, single fractures, multiple fractures, non-acute fractures, or occult rib fractures that are identified on x-ray.
    - Routine follow-up advanced imaging of rib or sternal fractures is not medically necessary.
- CT Chest without contrast (CPT® 71250) or Tc-99m bone scan whole body (CPT® 78306) is medically necessary for suspected pathological rib fractures, with or without a history of trauma
- For clavicle fractures and sterno-clavicular (SC) dislocation, see **Joint Instability and Dysfunction (MS-30.0)**.
- Advanced imaging of the abdomen or pelvis is not medically necessary for chest trauma when there is no physical examination or laboratory evidence of abdominal and/or pelvic injury.

Evidence Discussion

- Chest x-ray, in combination with physical exam, is the appropriate initial diagnostic modality in those with suspected rib or sternal fracture after chest trauma. Although chest x-ray has low sensitivity (approximately 50%) for detection of rib fracture,[1] it has the benefit of being widely and readily available and able to detect complications that may require additional imaging such as pneumo- or hemothorax and pulmonary contusions.[1,2] In addition, failure to detect rib fractures in uncomplicated cases does not significantly alter the individual management or outcomes. A study by Bansidhar et al. showed no difference in treatment in individual with minor chest trauma who did and did not have rib fractures diagnosed either clinically or radiographically.[1] Therefore, in uncomplicated cases, additional advanced imaging is not medically necessary. Rodriguez et al. demonstrated that yield for CT of thoracic injury with major clinical significance after a normal chest x-ray is 1.5% and would only detect one major injury for every 67 studies.[3]

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- In cases where complications are identified clinically or by other imaging, additional imaging with CT chest is medically necessary. CT does have higher sensitivity for detection of rib fractures[1] and in the detection and extent of pulmonary injuries.[2] It also may be useful in differentiating blunt cardiac injury from acute myocardial infarction. However, CT does carry with it the risk of contrast related renal injury and allergic reactions.[3] It also exposes the individual to a greater amount of ionizing radiation than a chest x-ray and subsequent increased risk of induced cancers.[3] It is estimated that undergoing chest CT will result in one radiation induced cancer per every 720 40-year-old females and 1,538 40-year-old males.[3]
- If a pathological rib fracture is suspected, imaging with either a CT chest or Tc-99m bone scan is medically necessary. CT may be helpful in differentiating primary tumor from metastasis and may aid in detection of the primary malignancy.[1] Tc-99mm bone scan has low specificity but high sensitivity (>95%) for detection of pathologic rib fractures.[1]
- Medial clavicular fractures are rare (<5% of cases) and may necessitate additional imaging with CT or MRI for evaluation.[4] Midshaft and distal clavicular fractures are usually sufficiently evaluated by x-ray.[4]
- Chest x-ray has poor sensitivity for identification of sternoclavicular dislocations. Given the risk for complications such as pneumothorax in posterior displacement, advanced imaging may be medically necessary. CT is advantageous as it has superior image resolution. It also allows for 3D reconstruction to determine exact position of the sternoclavicular joint. MRI can also be utilized but it has poorer resolution than CT. However, it may be advantageous for evaluation of soft tissue conditions or ligamentous injury.[5]
- For isolated chest injury without signs or symptoms of abdominal or pelvic injury, advanced imaging of the abdomen or pelvis is not medically necessary. If abdominal or pelvic injury is suspected, imaging is as dictated elsewhere in these guidelines.

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Chest Wall Mass (CH-22)

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Chest Wall Mass (CH-22.1)

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- Chest x-ray is useful in the workup of a soft-tissue mass and is almost always medically necessary as the initial imaging study.
  - Chest ultrasound (CPT® 76604) is medically necessary as an initial imaging study in the setting of a suspected superficial or subcutaneous lipoma. This modality may also be valuable in differentiating cystic from solid lesions and has also been used to assess the vascularity of lesions.
  - Following a non-diagnostic chest x-ray that does not show an obvious lipoma(s) or clearly benign entity (see **Tissue Growths and Masses (MS-10)** in the Musculoskeletal Imaging Guidelines), ONE of the following is medically necessary:
    - MRI Chest without and with contrast (CPT® 71552) or
    - MRI Chest without contrast (CPT® 71550) or when MRI is contraindicated,
    - CT Chest with contrast (CPT® 71260) or
    - CT Chest without contrast (CPT® 71250)

Evidence Discussion

Radiography is medically necessary as the initial imaging study for both superficial and non-superficial soft tissue masses. Radiography can help identify calcifications, bone involvement, intrinsic fat, and unsuspected skeletal abnormality or deformity. In general, radiographic findings related to a soft tissue mass can provide helpful insight in determining the next most appropriate imaging modality for further characterization.[1-5] Non-contrast enhanced ultrasound is also an excellent triage tool for evaluating superficial soft tissue masses like superficial lipomas.[1,3-5] However, ultrasound is not sufficient to assess for malignancy. When features detected on ultrasound are not clearly benign or when findings are concerning, further advanced imaging is medically necessary.[1]

MRI without and with IV contrast is medically necessary as the next imaging study for a soft tissue mass following non-diagnostic radiographs.[1-4]

MRI helps to define intrinsic tumor characterization, vascular structures, neurovascular involvement, hemorrhage, edema, and tumor necrosis. MRI without IV contrast may be beneficial compared with CT, but the use of MR contrast improves the differentiation of benign from malignant soft tissue masses.[1-4] At least 20% to 25% of soft tissue masses can demonstrate features that allow for confident diagnosis based on MRI alone, many of which are benign and thus would not warrant biopsy.[1,3,4] MRI has a sensitivity of 96%, a specificity of 72%, a positive predictive value of 77%, a negative predictive value of 83%, and a diagnostic accuracy of 88%.[6]

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When MRI is contraindicated, CT with IV contrast is medically necessary following non-diagnostic radiograph.

CT with IV contrast is useful in distinguishing vascularized from potentially necrotic regions of a tumor or calcification.[1-4]

Background and Supporting Information

- Chest x-rays of chest wall masses can detect calcification, ossification, or bone destruction as well as location and size.

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Pectus Excavatum and Pectus Carinatum (CH-23)

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Pectus Excavatum and Carinatum (CH-23.1)
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Pectus Excavatum and Carinatum (CH-23.1)

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- CT Chest without contrast (CPT® 71250) or MRI Chest without and with contrast (CPT® 71552) and 3-D reconstruction (CPT® 76377 or CPT® 76376) if:
  - Candidates for surgical correction.
  - Cardiac or pulmonary dysfunction has been identified
    - ECG and echocardiography if cardiac symptoms or evidence of cardiac function abnormalities.
    - Chest x-ray and PFT's if increasing shortness of breath.

Background and Supporting Information

- Chest measurements derived from CT Chest, such as the Haller Index or the correction index, are helpful to the thoracic surgeon in pre-operative assessment of chest wall deformities to assess for the appropriateness of operative repair prior to the development of symptomatic pectus deformities.
- The Haller index is calculated using the width of the chest divided by the distance between the posterior surface of the sternum and the anterior surface of the spine. A Haller index score is normal at 2.5 to 2.7 and severe at 3.25 or greater. The correction index uses an equation of (b−a)/b×100, in which a is the minimum distance between the anterior spine and the posterior surface of the sternum, and b is the maximum distance between the anterior spine and most anterior internal rib. It yields a percentage that the chest would need to be corrected to achieve normal dimensions, with a normal level being 10% or less.
- Some have suggested that a CXR can replace the CT Chest for Haller Index calculation with a strong correlation and high diagnostic accuracy.
- Expert consensus from The Society of Thoracic Surgeons 2023 recommended that a comprehensive evaluation with spirometry, ECG, and echocardiography be done with any cardio-pulmonary complaint. The Haller index, correction index, pulmonary compression or failed previous repair, in and of itself, was not an indication for surgery. Corrective surgery indications for those with severe pectus excavatum included: progression of deformity, presence of cardio-pulmonary symptoms, mitral valve prolapse, arrhythmia, significant body image disturbances, abnormal PFTs, abnormal cardiac function test, or the presence of cardiac compression on imaging (echo or CT).

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Pulmonary Arteriovenous Fistula (AVM) (CH-24)

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Pulmonary AVM (CH-24.1)

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- CT Chest with contrast (CPT® 71260), CT Chest without contrast (CPT® 71250), CTA Chest (preferred modality for pre-intervention planning) (CPT® 71275), or MRA Chest (CPT® 71555) for evaluation of:
  - suspected pulmonary AVM, including individuals with HHT (Hereditary Hemorrhagic Telangiectasia) or who have a first degree relative with HHT
  - first degree relatives of an individual with a primary pulmonary AVM
  - evaluation of individuals with paradoxical embolus/stroke and no evidence of patent foramen ovale on echocardiogram
  - follow-up of treated AVM's at 6 months post embolization and then every 3-5 years
  - follow-up of untreated AVM's to be determined by treating physician but no more than annually. Usually the interval is 3-5 years due to the slow-growth nature of PAVM's
  - treated or untreated PAVM's with recurrent symptoms

Evidence Discussion

Chest x-ray is the most appropriate initial imaging exam with presentation of hypoxemia or hemoptysis but it does have low sensitivity for pulmonary arteriovenous malformation (PAVM).[3]

CT chest is the test of choice for diagnosing a PAVM. Contrast may be considered for an atypical nodule/soft tissue lesion on CT chest without contrast and suspicion for a PAVM. CTA chest is the gold standard for defining the vascular anatomy of a previously identified PAVM. It is not routinely used for diagnosis except in the setting of concomitant embolization therapy, diagnostic uncertainty, or pre-intervention planning.[3] MRA chest avoids ionizing radiation but is not as sensitive or specific as CT for the diagnosis of PAVM and has limitations detecting PAVM <5mm.[3] It has a potential role in younger people with Hereditary Hemorrhagic Telangiectasia (HHT) who may require lifelong surveillance.[3]

CT chest without contrast may be done to screen for PAVM in people with possible or confirmed Hereditary Hemorrhagic Telangiectasia (HHT).[1] A negative CT chest with or without contrast helps to exclude a clinically significant PAVM.[2]

Background and Supporting Information

- Pulmonary AVMs are abnormal connections between pulmonary arteries and veins, usually found in the lower lobes, that can be either primary (such as in individuals with HHT) or acquired (such as trauma, bronchiectasis). They can be identified in up to

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98% of chest x-rays by a peripheral, circumscribed, non-calcified lesion connected by blood vessels to the hilum of the lung. Treatment is often by surgery or embolization of the feeding artery using platinum coils or detachable balloons.

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Pulmonary Embolism (PE) (CH-25)

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- CT Chest with contrast with PE protocol (CPT® 71260) or CTA Chest (CPT® 71275) is medically necessary if at least one symptom, clinical/laboratory finding, or risk factor from each of the lists below are present.
  - With any ONE of the 3:
    - Dyspnea, new onset and otherwise unexplained;
    - Chest pain, pleuritic;
    - Tachypnea
  - AND, with any ONE of the 3:
    - Abnormal **D-dimer** test;
    - Wells Criteria score* higher than 4 points;
    - One risk factor** or symptom** of new onset demonstrating high clinical probability of PE

RISK FACTORS**	SYMPTOMS ATTRIBUTED TO PE**
Immobilization via hospitalization or institutional care at least 3 days or surgery in last 4 weeks or recent trauma	Signs or symptoms of DVT
Previous history of DVT or PE	Hemoptysis
Cancer actively treated in last 6 months or receiving palliative treatment or myeloproliferative disorder	Right heart strain or failure
Recent history of a long airplane flight	Systolic BP <90
Use of estrogen-based contraceptives (birth control pills), oral estrogen	Syncope
Advanced age (≥70)	Cough
Congestive heart failure, congenital heart disease, hypertension, superficial venous thrombosis, and/or indwelling central vein catheter	Heart Rate >100

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RISK FACTORS**	SYMPTOMS ATTRIBUTED TO PE**
Obesity (BMI ≥30)	Palpitations
COVID-19 for those post-hospitalization or those within 30 days of diagnosis	
Recurrent pregnancy loss	
COPD	
Thrombotic disorders, antiphospholipid antibody syndrome, and/or autoimmune disease	
Inflammatory bowel disease	
Nephrotic syndrome	
In vitro fertilization (IVF) treatment	

Well's Criteria for Clinical Probability of PE*	
Clinical signs/symptoms of DVT (at minimum: leg swelling and pain with palpation of the deep veins)	3
PE is likely or equally likely diagnosis	3
Heart rate >100	1.5
Immobilization at least 3 days or surgery in last 4 weeks	1.5
Previous history of DVT or PE	1.5
Hemoptysis	1
Cancer actively treated in last 6 months or receiving palliative treatment	1
Calculate Probability: Low <2 Moderate 2 to 6 High >6	

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Well's Criteria for Clinical Probability of PE*
Using the above criteria, only 3% of individuals with a low pretest probability had PE versus 63% of those with a high pretest probability.

- Non-urgent cases which do not meet above 2-step criteria, should undergo prior to advanced imaging:
  - Chest x-ray (to rule out other causes of acute chest pain)
  - Primary cardiac and pulmonary etiologies should be eliminated
- Pregnancy is a risk factor for thrombo-embolic events in and of itself. Additional risk factors are not required. Pregnant individuals with suspected PE should proceed with:
  - If signs/symptoms of DVT are present, Doppler studies of the lower extremities (CPT® 93970 bilateral study or CPT® 93971 unilateral study) are medically necessary
  - If no signs/symptoms of DVT, then chest x-ray first.
  - If chest x-ray is normal, then V/Q scan (CPT® 78580 or CPT® 78582) (preferred test), or CTA Chest (CPT® 71275) or CT Chest with contrast with PE protocol (CPT® 71260)
  - If chest x-ray is abnormal or after non-diagnostic V/Q scan or if V/Q scanning is not readily available, then CTA Chest (CPT® 71275) or CT Chest with contrast with PE protocol (CPT® 71260).
- Ventilation-perfusion scans, also called V/Q, scans (CPT® 78580-Pulmonary Perfusion Imaging; CPT® 78582-Pulmonary Ventilation (e.g., Aerosol or Gas) and Perfusion Imaging) or SPECT/CT (CPT® 78830):
  - Is not a replacement for CTA Chest
  - Is medically necessary in any of the following:
    - Suspected pulmonary embolism if there is a contraindication to CT or CTA Chest (ventilation-perfusion scans CPT® 78582)
    - Suspected pulmonary embolism when a chest x-ray is negative and CTA Chest is not diagnostic (CPT® 78580 or CPT® 78582)
    - Follow-up of an equivocal or positive ventilation-perfusion lung scan to evaluate for interval change (CPT® 78580)
    - Suspected Chronic thromboembolic disease or Chronic thromboembolic pulmonary hypertension*, usually after 3 months of effective anticoagulation
- Follow-up imaging in stable or asymptomatic individuals with known PE is not medically necessary
- Follow-up imaging with CT Chest with contrast with PE protocol (CPT® 71260) or CTA Chest (CPT® 71275) for ANY of the following indications:
  - Recurrent or persistent signs or symptoms such as dyspnea, or
  - Elevated D-dimer which is persistent or recurrently elevated, or

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  - Right heart strain or failure identified by EKG, ECHO or heart catheterization
- For Pulmonary Artery Hypertension (PAH), see **Pulmonary Artery Hypertension (PAH) – Indications (CD-8.1)** in the Cardiac Imaging Guidelines.

Evidence Discussion

Symptoms and signs of pulmonary embolus (PE) are nonspecific and common; therefore, knowing whom to test for PE is challenging.[9] In North America, PE is diagnosed in only 5% of people tested for it.[8] Chest x-ray (CR) is nonspecific but may rule out other causes of dyspnea and chest pain. Avoiding the overuse of imaging tests is important, given the potential harms of radiation exposure, high costs and complications. The pretest clinical probability has an important effect on the predictive value of CT pulmonary angiography (CTPA).[8] Determining the clinical pretest probability of PE depends on clinical judgment, which lacks standardization and is subjective, or prediction rules.[8,11] Kahn et al recommend diagnostic imaging in those with a likelihood of PE greater than or equal to 15%, based on the "implicit sense" of the clinician, and either a structured clinical probability score (Wells, Revised or Simplified Geneva score) or a D-dimer above a pre-specified threshold.[9] Other experts recommend that imaging be done in those with a high pretest probability based on empirical clinical judgment or a prediction rule and in those with a low/intermediate pretest probability and a positive D-dimer.[8,10] Imaging can be avoided in people with both a structured clinical probability score at or below the given cutoff and a D-dimer below the given cutoff value.[9] Imaging is likewise not medically necessary in those with a low/intermediate pretest probability based on clinical judgement and a normal D-dimer.[8,10]

CTPA is highly sensitive and specific and is the imaging method of choice for suspected PE.[8-11] It may also demonstrate other potential causes of the presenting symptoms. CTPA is a CT angiogram with intravenous (IV) contrast. The timing of the scan is tailored so that contrast enhances the pulmonary arterial system to identify potential filling defects. CT with contrast is usually not medically necessary. According to the American College of Radiology, when IV contrast is given during CT acquisition for suspected PE, the study should be performed as a CTPA.[10]

Planar V/Q may preferentially be used in outpatients with a low clinical probability of PE and normal CR, in young (especially female) individuals, pregnant women, and individuals with a history of contrast allergy or renal failure.[8] The proportion of diagnostic V/Q scans is higher in individuals with a normal CXR. A normal V/Q scan has a high negative predictive value, but there is a high proportion of non-diagnostic scans and it cannot provide alternative diagnoses.[10] Abnormal regional lung perfusion may suggest PE but is not specific and requires correlation with ventilation studies or other imaging. Investigators have studied single-photon emission CT (SPECT) to improve the sensitivity and specificity of V/Q scans. Kahn et al state that V/Q SPECT is a low radiation option to minimize lung and breast tissue irradiation in younger individuals.[9] Some authors believe that V/Q SPECT should be the preferred study in the evaluation

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of suspected PE.[12] However, large scale prospective trials are needed to validate SPECT techniques before its widespread incorporation into diagnostic algorithms.[8] SPECT has over 90% sensitivity and 72-98% specificity, and V/Q SPECT/CT has the highest diagnostic accuracy.[16-18]

A normal perfusion scan and a negative CTPA appear equally safe for ruling out PE in pregnancy.[8] There is debate regarding which is the first test of choice. CTPA is more expensive and exposes the pregnant woman to more radiation than the fetus; V/Q scans have low radiation and no contrast-related side effects.[13] A compression duplex ultrasound of the lower extremity is medically necessary if there are symptoms/signs of a DVT. If a DVT is diagnosed, anticoagulation can be administered without further imaging. A CXR is medically necessary. An alternative diagnosis may be found, and it can inform the choice between CTPA and a V/Q scan as the next test when there is no clinical evidence of a DVT.[10] If there is no DVT and a CXR is normal, CTPA or a perfusion scan is medically necessary. If the perfusion scan is normal, a ventilation scan may not be needed. If the CXR is abnormal, alternative causes have been considered and PE is still suspected, CTPA is medically necessary.[8]

Acute PE is treated for at least 3 months with anticoagulation. Individuals should have a follow-up visit 2 weeks to 3 months after PE. If symptoms worsen or the diagnosis is complex, follow-up may occur sooner.[6] Whether anticoagulation is stopped after 3 months or continued indefinitely depends on whether the reduced risk of recurrent venous thromboembolism (VTE) outweighs the increased risk of bleeding.[8] The risk of recurrent VTE after stopping anticoagulation is related to the risk factor category for the index PE/VTE event. There are many genetic and acquired risk factors for VTE associated with a low, intermediate or high risk of recurrence.[8,11]

Patency of the pulmonary arterial bed is restored in the majority of people within the first few months, and routine CTPA imaging is not medically necessary.[8] Konstantinides et al recommend a transthoracic echocardiogram in those with dyspnea or functional limitation at follow up.[8] If the probability of pulmonary hypertension is felt to be high, planar V/Q is considered the first line imaging test for suspected chronic thromboembolic pulmonary hypertension (CTEPH). CTPA should not be used as a stand alone test to rule out CTEPH. The diagnosis is based upon measurements made during right heart catheterization and mismatched perfusion defects on V/Q scan more than 3 months after an acute PE.[8]

Background and Supporting Information

- Pulmonary embolism is found in approximately 10% of all those that present with suspicion of PE. Dyspnea, pleuritic chest pain, and tachypnea occur with about 50% incidence with leg swelling or pain just over 50%.
- D-dimer level has a high sensitivity and low specificity for diagnosing PE.
  - A negative D-dimer in combination with low or moderate PE risk classification has a negative predictive value approaching 100%.

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  - D-dimer can be falsely elevated with surgery, injury, malignancy, sepsis, diabetes, pregnancy, or other conditions where fibrin products are likely to be present.
- CT imaging has supplanted V/Q scanning since the latter is difficult to obtain quickly, does not provide a substantial cost savings, and does not diagnose other pulmonary pathology.
- The decision to terminate anticoagulation treatment after previous pulmonary embolism (PE) with absent or stable symptoms is based on clinical evaluation and risk factors.
- Repeat studies do not allow one the ability to distinguish new from residual clot, with luminal diameter and clot character poorly correlated to symptoms and ECHO findings.
- Half to two thirds of individuals with primary thromboembolism have residual pulmonary artery clot at 6 months and 50% remain at one year, depending on modality at follow-up.
- Subsequent persistence or elevation of D-dimer is associated with increased risk of recurrent PE. ECHO and Right Heart Catheterization (RHC) can identify those with pulmonary hypertension. Yet, 1/2 of all have persistent or new pulmonary hypertension after primary thromboembolism and only half of this latter group has dyspnea at rest or exercise intolerance.
- Of note, pregnancy is accompanied by a progressive increase in D-dimer levels, and as such, D-Dimer levels may not be helpful to rule-in or rule-out DVT/PE in pregnancy.

Modality	Fetal radiation exposure in mGy
CXR	0.002-0.1
V/Q	0.32 – 0.74
CTPA	0.03 – 0.66

- Compared with V/Q scan, computed tomography pulmonary angiography (CTPA) is associated with a higher radiation dose to the mother: the calculated doses to breast and lung tissue have been estimated to range from 10mGy to 60mGy and 39.5mGy, respectively, with CTPA ,as compared with 0.98mGy to 1.07mGy and 5.7mGy to 13.5mGy, respectively, with V/Q scan.

Pulmonary Embolism Health Equity Considerations

- Black individuals and female individuals have a higher risk of pulmonary embolism.

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- Underrepresented racial/ethnic groups are less likely to undergo advanced therapies for pulmonary embolism.
- Several risk factors for pulmonary embolism (e.g., hyperestrogenic state due to oral contraceptives, pregnancy, or HRT) are exclusive to female individuals.
- Individuals with a low socioeconomic status have reduced access to high-quality imaging centers and undergo fewer advanced interventions for pulmonary embolism.

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Pulmonary Hypertension (CH-26)

Guideline

Pulmonary Hypertension (CH-26.1)

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Pulmonary Hypertension (CH-26.1)

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- See the **Pulmonary Artery Hypertension (PAH) – Indications (CD-8.1)**

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Subclavian Steal Syndrome (CH-27)

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Subclavian Steal Syndrome (CH-27.1)

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Subclavian Steal Syndrome (CH-27.1)

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- See **Subclavian Steal Syndrome (PVD-4.1)** for concerns of Subclavian Steal Syndrome in Peripheral Vascular Disease imaging guidelines.

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Superior Vena Cava (SVC) Syndrome (CH-28)

Guideline

SVC Syndrome (CH-28.1)

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SVC Syndrome (CH-28.1)

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- For concerns of SVC syndrome, see **Upper Extremity Venous - Imaging (PVD-4.2)** in Peripheral Vascular Disease Imaging guidelines.

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Elevated Hemidiaphragm (CH-30)

Guideline

Elevated Hemidiaphragm (CH-30.1)
References (CH-30)

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Elevated Hemidiaphragm (CH-30.1)

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- CT Chest with contrast (CPT® 71260) and/or CT Neck with contrast (CPT® 70491) with new diaphragmatic paralysis after:
  - previous chest x-rays are available and reviewed to determine if the diaphragmatic elevation is a new finding, and/or
  - fluoroscopic examination ("sniff test") to differentiate true paralysis from weakness
- CT Abdomen with contrast (CPT® 74160) to rule out liver or abdominal process if CT Chest is negative.
- Repeat advanced imaging studies in the absence of new signs or symptoms are not medically necessary.

Evidence Discussion

Diaphragmatic dysfunction includes eventration, weakness and paralysis. Diagnosis is based in part on static and dynamic imaging tests.[1] Unilateral diaphragmatic paralysis is often asymptomatic and suspected when an elevated hemidiaphragm is found incidentally on chest x-ray (CR). CR is a simple and effective test to evaluate the pulmonary parenchyma and the diaphragm.[1] CR has a sensitivity of 66.6% and a specificity of 44%.[4] The positive and negative predictive value of an elevated hemidiaphragm on CR for diaphragmatic dysfunction is 33% and 93%, respectively. The presence of diaphragm elevation is not necessarily a sign of dysfunction, but its absence makes it unlikely.[1]

Flouroscopy has traditionally been the gold standard for diagnosing diaphragmatic paralysis since it can visualize the diaphragm throughout the respiratory cycle and during forced inspiratory maneuvers (i.e., the "sniff test"). Some authors now consider US to be the imaging method of choice for the evaluation of diaphragmatic dysfunction.[1,2] US is non-invasive, portable, quick and does not expose the individual to ionizing radiation. Absence of thickening of the diaphragm during inspiration, absence of caudal movement during normal inspiration or paradoxical movement during the sniff maneuver confirms paralysis.[4]

A common concern is whether there is an underlying serious condition in those individuals with unilateral hemidiaphragm paralysis with no evident etiology after a history, physical exam and CR. Piehler et al concluded that such individuals are unlikely to have an underlying occult malignant or neurologic condition.[3] However, Windisch et al recommended that a one-time CT chest be done if there is clinical suspicion of possible malignancy with damage to the phrenic nerve.[2]

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Additional imaging may be needed to rule out conditions which can cause an elevated hemidiaphragm but are not associated with respiratory muscle weakness. For example, abdominal imaging can be done for suspected hepatic abscess, ascites, or splenomegaly.

Background and Supporting Information

- The right hemidiaphragm sits about 2cm higher than the left.
- “Eventration” is thin membranous replacement of muscle, usually on the right, as the most common cause of elevation.
- Any injury to the phrenic nerve from neck to diaphragm can lead to paralysis.
- Common phrenic causes are traumatic or surgical injury or malignancy involving the mediastinum.
- Any loss of lung volume or increased abdominal pressure can lead to diaphragm elevation.

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Thoracic Outlet Syndrome (TOS) (CH-31)

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Thoracic Outlet Syndrome (CH-31.1)

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Thoracic Outlet Syndrome (CH-31.1)

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- For concerns of Thoracic Outlet Syndrome, see **Thoracic Outlet Syndrome (PVD-4.2)** in Peripheral Vascular Disease Imaging Guidelines.
- For concerns of Neurogenic Thoracic Outlet Syndrome, see **Brachial Plexus (PN-4.1)** in Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines.

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Lung Transplantation (CH-32)

Guideline

Pre-Transplant Imaging Studies (CH-32.1)
Post-Transplant Imaging Studies (CH-32.2)
Reference (CH-32)

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Pre-Transplant Imaging Studies (CH-32.1)

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- Individuals on the waiting list or being considered for the lung transplant can undergo advanced imaging per that institution's protocol, or any of the studies listed below are medically necessary:
  - CT Chest with and without contrast (CPT® 71270), CT Chest with contrast (CPT® 71260), or CT Chest without contrast (CPT® 71250)
  - ECHO
  - Imaging Stress Test (MPI, SE, MRI) or Heart Catheterization (Right and Left); Heart catheterization can also be done after a positive stress test.
  - CTA Chest (CPT® 71275), and/or CTA Abdomen and Pelvis (CPT® 74174) and/or CTA Aorta with bilateral lower extremity run-off (CPT® 75635) is medically necessary (without initial ABI's and/or arterial duplex) for the following individuals:
    - prior abdominal or lower extremity vascular intervention (any timeframe is acceptable)
    - known peripheral artery disease (PAD) from prior imaging
    - current symptoms of claudication, rest pain or gangrene
  - CTA Chest (CPT® 71275) and/or CTA Abdomen and Pelvis (CPT® 74174) and/or CTA Aorta with bilateral lower extremity run-off (CPT® 75635) is medically necessary (after initial ABI's and/or arterial duplex) for the following individuals:
    - Initial ABI's and/or arterial duplex suggest the presence of PAD as evidenced by one of the following:
      - ABI of <0.9
      - presence of plaque
      - presence of vascular calcification, stenosis or occlusion
      - small vessel size on the duplex
  - CT Abdomen and Pelvis with or without contrast (CPT® 74177 or CPT® 74176) for determining extracorporeal membrane oxygenation (ECMO) candidacy
- Other studies that are medically necessary include V/Q scan and Six Minute Walk Test.
- See **Transplant (CD-1.6)** in the Cardiac Imaging Guidelines.

Evidence Discussion

- Computed Tomography (CT) is often performed for evaluation of individuals prior to lung transplantation. CT allows for surgical planning to delineate extent of the disease

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and assess for any contraindications to transplant.[6] CT carries the risk of exposure to iodinated contrast and ionizing radiation.

- Evaluation of donors is commonly performed by chest radiography.[6,8] Occasionally, evaluation of donors is performed by chest CT to assess for malignancy, pneumonia, emphysema, and other diseases.[9]
- Cardiac evaluation with echo and/or ischemic evaluation (image stress testing or heart catheterization) is medically necessary prior to lung transplantation.
- Extracorporeal Membrane Oxygenation (ECMO) has been increasingly utilized for bridging prior to lung transplantation or as an adjunct procedure post-transplant.[4,5] Given the risk of vascular complications,[3] preoperative evaluation of the vasculature is reasonable. For those that are asymptomatic without previously known peripheral artery disease (PAD), initial work up with ankle-brachial index (ABI) and/or arterial duplex is medically necessary.[7] For those that are symptomatic, have a history of known PAD (either from prior imaging or previous vascular intervention) or initial work up has suggested the presence of PAD, advanced imaging is medically necessary for further evaluation.[7]

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Post-Transplant Imaging Studies (CH-32.2)

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- CT Chest with and without contrast (CPT® 71270), CT Chest with contrast (CPT® 71260), or CT Chest without contrast (CPT® 71250) is medically necessary for:
  - initial post-transplant follow-up
  - suspected complication, either surgical, medical or infectious, (See **Background and Supporting Information**)
  - worsening PFT's
  - new finding on other imaging, including chest x-ray
- CTA chest (CPT® 71275) is medically necessary for suspected torsion
- See **Transplant (CD-1.6)** in the Cardiac Imaging Guidelines.

Evidence Discussion

- There are no universally accepted follow-up protocols for routine post-transplant surveillance.[6] CT chest is supported for initial post-transplant follow up.[1,6] CT carries with it the risk of exposure to iodinated contrast and ionizing radiation.[8,9]
- Additional follow-up is based on clinical presentation, suspected complication, or findings on other imaging.[1,6]

Background and Supporting Information

- Complications from lung transplantation are a major cause of morbidity and mortality.
- The three main categories of complications are surgical, medical and infectious.
  - Surgical complications include: anastomotic complications, bronchial dehiscence, bronchial stenosis, pneumothorax, hemothorax, hematoma, wound dehiscence, and infection.
  - Medical complications include: primary graft dysfunction, pulmonary embolism and pulmonary infarction, Tracheobronchomalacia, posttransplant lymphoproliferative disease, primary disease recurrence, and acute and chronic allograft rejection, including bronchiolitis obliterans and restrictive allograft syndrome.
  - Infectious complications include: hospital and community acquired nonmycobacterial pulmonary infections, mycobacterial infections, fungal infections, and viral infections (CMV most common).

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Lung Transplant Health Equity Considerations

- The majority of individuals who receive lung transplants in the United States are of White race and are male. Compared to a matched person of another gender or race, these individuals have a higher likelihood or being allocated a lung transplant.

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Reference (CH-32)

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Lung Cancer Screening (CH-33)

Guideline

U.S. Preventive Services Task Force: Lung Cancer Screening (CH-33.1)
Incidental Pulmonary Nodules Detected on Low Dose CT Chest (LDCT) Images (CH-33.3)
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U.S. Preventive Services Task Force: Lung Cancer Screening (CH-33.1)

CH.CS.0033.1.A
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- Low-dose CT Chest (CPT® 71271) for lung cancer screening in asymptomatic individuals* annually when all of the following criteria have been met:

Screening Indications	Imaging Study
• All criteria below must be met: ◦ Individual has not received a low-dose CT lung screening in less than 12 months; and ◦ Individual has NO health problems that substantially limit life expectancy or the ability or willingness to have curative lung surgery**; and ◦ Individual is between 50 and 80 years of age; and ◦ Individual has at least a 20 pack-year history of cigarette smoking; and ◦ Currently smokes or quit within the past ≤15 years	Low-Dose CT Chest without contrast (CPT® 71271)

For those that no longer qualify for annual LDCT for lung cancer screening but have known lung nodules, follow criteria for follow-up under CH-16. For example, a nodule that is new on the last screening LDCT may warrant continued diagnostic CT evaluation per CH-16.2.

*Symptoms of lung cancer (e.g., hemoptysis, unexplained cough, and/or unexplained weight loss of >15 pounds in the past year) warrant diagnostic evaluation, not screening.

**This is based on a range of chest or other organ signs, symptoms or conditions which would question the member's ability to undergo surgical or non-surgical treatment if a lung cancer was discovered. For example, congestive heart failure, advanced cancer from another site or a member with COPD who uses oxygen when ambulating, would be examples of conditions that would "substantially limit life expectancy." Conversely, stable COPD and its symptoms, including cough, shortness of breath would not "substantially limit life expectancy."

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Lung Cancer Screening Health Equity Considerations

- Black men, individuals with lower levels of education, and individuals with lower household income levels are less likely to be eligible for screening.
- Among individuals who are eligible for lung cancer screenings, Black individuals and those with a lower socioeconomic status are screened at a lower rate and have lower adherence to screening follow-up. This could be attributed to differences in referral rates among providers.
- Barriers to lung cancer screenings include:
  - Individual-level barriers, such as limited knowledge about screening, lack of education and recommendations from providers, distrust of the medical system, and lack of transportation
  - Provider-level barriers, such as lack of knowledge about screening recommendations and lack of time for shared decision-making
  - System-level barriers, such as misconceptions about screening requirements, lack of scanners, limited resources, and low reimbursement rates

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Incidental Pulmonary Nodules Detected on Low Dose CT Chest (LDCT) Images (CH-33.3)

CH.CS.0033.3.A

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- Any Lung-RADS requiring less than 1 year interval follow-up is coded as Low-Dose CT Chest (CPT® 71250) (Not CPT® 71271 which is ONLY the annual screen)
- For lung nodules, including incidental findings from studies other than screening LDCT, or if no longer qualify for screening LDCT, see **Incidental Pulmonary Nodules Detected on CT Images (CH-16.2).**

Lung-RADS Primary Category/ Category Descriptor	Management
0: Incomplete	If findings suggestive of an inflammatory or infectious process, follow-up with LDCT (CPT® 71250) in 1-3 months
1: Negative - very low likelihood of becoming a clinically active cancer	Annual LDCT screening (CPT® 71271) in 12 months
2: Benign appearance or behavior - very low likelihood of becoming a clinically active cancer due to size or lack of growth	Annual LDCT screening (CPT® 71271) in 12 months
3: Probably benign finding(s) - short term follow-up suggested; includes nodules with a low likelihood of becoming a clinically active cancer	6 month LDCT (CPT® 71250) and if unchanged on this CT it is coded as category 2 and returned to annual LDCT screening (CPT® 71271) in 12 months

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Lung-RADS Primary Category/ Category Descriptor	Management
4A: Suspicious - Findings for which additional diagnostic testing and/ or tissue sampling is recommended	PET/CT (CPT® 78815) when there is a ≥8 mm solid nodule or solid-component Follow-up with LDCT (CPT® 71250) in 3 months and if stable or decreased in size on this CT, it is coded as category 3 with follow-up LDCT (CPT® 71250) at 6 months, if stable or decreased in size on this CT, return to annual LDCT screening (CPT® 71271) in 12 months
4B or 4X: Suspicious - Findings for which additional diagnostic testing and/ or tissue sampling is recommended	CT Chest with or without contrast, PET/ CT (CPT® 78815), and/or tissue sampling depending on the probability of malignancy and comorbidities. PET/CT (CPT® 78815) when there is a ≥8 mm solid component. If there is low suspicion of lung cancer, follow-up with LDCT (CPT® 71250) in 3 months with another LDCT (CPT® 71250) in 6 months and if unchanged on this CT return to annual LDCT screening (CPT® 71271) in 12 months

For those that no longer qualify for annual LDCT for lung cancer screening but have known lung nodules, see criteria for follow-up under CH-16. For example, a nodule that is new on the last screening LDCT may warrant continued diagnostic CT evaluation per **CH-16.2**.

Evidence Discussion

- Low-dose computed tomography (LDCT) Chest for lung cancer screening has been shown to have sensitivity ranging from 59% to 100%, a specificity of 26.4% to 99.7%, a negative predictive value of 97.7% to 100% and a positive predictive value from 3.3% to 43.5%.[2] The benefit of lung cancer screening is early detection and treatment. The NLST trial showed a relative risk reduction in lung cancer mortality of 20%.[2] The radiation dose of a LDCT is typically 10% to 30% of a standard-dose CT.[2] The harms of a screening program would include false-positive results and subsequent unnecessary tests and procedures, the exposure to ionizing radiation and ensuing radiation-induced cancer, and increased anxiety and distress.[2]

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- The risk of malignancy associated with a Lung CT Screening Reporting and Data System (Lung-RADS) score is as follows: a score of 2 is <1%; a score of 3 is 1%-2%; a score of 4A is 5%-15%; a score of 4B and 4X is >15%.[3] The American College of Radiology (ACR) recommends follow up imaging of incidental pulmonary nodules detected on low dose lung cancer screening CT's based on the Lung-RADS score;[3,5] however, there is limited data on the impact of screening intervals.[5] The NLST and NELSON studies demonstrating reduction in lung cancer mortality were based on screening intervals of 1 year and 1, 3, and 5.5 years, respectively.[5] Multiple studies have shown that the 3 month follow up recommended for Lung-RADS 4A nodules is optimal, but have raised concerns on stepwise downgrading of a stable 4A nodule to a Lung-RAD 2.[5] Therefore the ACR has modified follow up intervals with stepped management using the following criteria:
  - Nodules that are stable or decreased at follow-up are downgraded to the next lower Lung-RADS category.[5]
  - Nodules that completely resolve or are proven benign after an appropriate diagnostic evaluation are reclassified based on the most concerning finding.[5]
  - Follow-up recommendations are timed from the current examination.[5]

For further specific details concerning Lung-RADS, see v2022 by the ACR.[3]

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General Guidelines (HD-1)

Guideline

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Abbreviations for Head Imaging Guidelines

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Abbreviations for Head Imaging Guidelines	
ACTH	adrenocorticotropic hormone
AD	Alzheimer's Disease
ADH	antidiuretic hormone
AION	arteritic ischemic optic neuritis
AVM	arteriovenous malformation
CBCT	cone-beam computerized tomography
CMV	cytomegalovirus
CSF	cerebrospinal fluid
CT	computed tomography
CTA	computed tomography angiography
DNA	deoxyribonucleic acid
DWI	diffusion weighted imaging (for MRI)
EEG	electroencephalogram
ENT	ear, nose, throat
ESR	erythrocyte sedimentation rate
FDG	fluorodeoxyglucose
FSH	follicle-stimulating hormone

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Abbreviations for Head Imaging Guidelines	
FTD	Frontotemporal Dementia
GCA	giant cell arteritis
GCS	Glasgow Coma Scale
HIV	human immunodeficiency virus
LH	luteinizing hormone
MMSE	mini mental status examination
MRA	magnetic resonance angiography
MRI	magnetic resonance imaging
MRN	magnetic resonance neurography
MS	multiple sclerosis
MSI	magnetic source imaging
NAION	non-arteritic ischemic optic neuritis
NPH	normal pressure hydrocephalus
PET	positron emission tomography
PML	progressive multifocal leukoencephalopathy
PNET	primitive neuro ectodermal tumor
PWI	perfusion weighted imaging (for MRI)
SAH	subarachnoid hemorrhage
SIADH	syndrome of inappropriate antidiuretic hormone secretion

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Abbreviations for Head Imaging Guidelines	
SLE	systemic lupus erythematosus
TIA	transient ischemic attack
TMJ	temporomandibular joint disease
TSH	thyroid-stimulating hormone
VBI	vertebrobasilar insufficiency
VP	ventriculoperitoneal
XRT	radiation therapy

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General Guidelines (HD-1.0)

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- A pertinent clinical evaluation including a detailed history, physical examination including a neurological examination since the onset or change in symptoms, and appropriate laboratory studies should be performed prior to considering the use of an advanced imaging (CT, MR, Nuclear Medicine) procedure.
  - A pertinent clinical evaluation furnished via telehealth, since the onset or change in symptoms, is treated the same as an in-person clinical evaluation.
  - An exception to a pertinent clinical evaluation can be made if the individual is undergoing a guideline-supported, scheduled follow-up imaging evaluation.
    - Scheduled follow-up of known problems such as, multiple sclerosis, tumors, or hydrocephalus, scheduled surveillance with no new symptoms, screening asymptomatic individual due to family history or otherwise meet criteria for repeat imaging, as well as appropriate laboratory studies and non-advanced imaging modalities
  - A detailed neurological exam is required prior to advanced imaging except in the following scenarios:
    - Tinnitus, TMJ, sinus or mastoid disease, ear pain, hearing loss, eye disease, pituitary disease, and epistaxis. (A pertinent clinical evaluation since onset of symptoms is still required)
    - The request is from a neurologist, neurosurgeon, neuro-ophthalmologist, endocrinologist, gynecologist, otolaryngologist, or ophthalmologist who has seen the individual since onset of symptoms, or any provider in consultation with one of the above specialists.
- Other meaningful contact (telephone call, electronic mail, or messaging) since the onset or change in symptoms, with an established individual can substitute for a face-to-face clinical evaluation
- CT Head contrast as requested (CPT® 70450 OR CPT® 70460 OR CPT® 70470) is medically necessary when MRI is contraindicated.

Health Equity Consideration

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence;

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education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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General Guidelines – Anatomic Issues (HD-1.1)

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- If two studies using the same modality both cover the anatomic region of clinical interest, only one is generally needed, with the exception of the following scenarios:
  - CT Maxillofacial (CPT® 70486, CPT® 70487, or CPT® 70488) or CT Orbit/Temporal bone (CPT® 70480, CPT® 70481, or CPT® 70482): both cover the structures of the orbits, sinuses, and face. Two separate imaging studies are only supported if there is suspicion of simultaneous involvement of more posterior lesions, especially of the region involving the middle or inner ear.
  - Pituitary Gland: one study (either MRI Brain [CPT® 70553] or MRI Orbit/Face/Neck [CPT® 70543]) is adequate to report the imaging of the pituitary. If a previous routine MRI Brain was reported to show a possible pituitary tumor, a repeat MRI with dedicated pituitary protocol is supported.
  - Internal Auditory Canal: (IAC) MRI can be reported as a limited study with one code from the set (CPT® 70540, CPT® 70542, or CPT® 70543), but should not be used in conjunction with MRI Brain codes (CPT® 70551, CPT® 70552, or CPT® 70553) if IAC views are performed as part of the brain.
  - Mandible (jaw): CT Maxillofacial (CPT® 70486, CPT® 70487, or CPT® 70488) or CT Neck (CPT® 70490, CPT® 70491, or CPT® 70492) can be used to report imaging of the mandible. CT Neck will also image the submandibular space.
    - If MRI is medically necessary, MRI Orbit/Face/Neck (CPT® 70540, CPT® 70542, or CPT® 70543) can be used to report imaging of the mandible and submandibular space.
    - MRI Temporomandibular Joint(s) (TMJ) is reported as CPT® 70336. This code is inherently bilateral and should not be reported twice on the same date of service.

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General Guidelines – Modality (HD-1.2)

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- MRI is preferable to CT for most indications. For exceptions, See **General Guidelines – CT Head (HD-1.4)**
- MRI is medically necessary for these indications following an initial CT:
  - MRI Brain without and with contrast (CPT® 70553) to follow-up abnormalities seen on CT Head without contrast (CPT® 70450) when a mass, lesion, or infection is found.
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) to follow-up abnormalities seen on CT Head without contrast (CPT® 70450) when there is suspected Multiple Sclerosis or other demyelinating disease.
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) to follow up on stroke or TIA when initial CT Head was done on emergent basis.
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) for evaluation of new onset seizures.

Evidence Discussion (HD-1.2)

- MRI of the brain is the appropriate initial imaging study for diagnosis, characterization and surveillance of a variety of neurologic conditions, including, but not limited to: neoplastic conditions, evaluation of the brain parenchyma, meninges, ischemia and infarction, neurodegenerative disorders, hydrocephalus, demyelinating conditions, post-traumatic brain injury, inflammatory and autoimmune disorders and infectious disorders.[9]
- MRI Brain has some benefit over CT for determining age of intracranial hemorrhage, early stroke (via Diffusion imaging sequences), and detection of micro hemorrhage.[9,35,36,37]
- MRI is also medically necessary for further characterization of abnormalities detected on other imaging tests such as CT or sonography.[9]
- Limitations to MRI include artifacts due to motion and susceptibility effects, contrast complications, contraindication due to ferromagnetic devices or implants. Additionally, severe claustrophobia may require sedation in order to complete the study.[33]

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General Guidelines – MRI Brain (HD-1.3)

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- MRI Brain with contrast (CPT® 70552) should not be ordered except to follow-up on a very recent abnormal or equivocal non-contrast MRI Brain.
- After an MRI Brain without contrast (CPT® 70551), a follow up MRI Brain with contrast (CPT® 70552) may be performed at the discretion of a neurologist, a neurosurgeon, or a neuro-ophthalmologist, or any provider in consultation with a neurologist, neurosurgeon, or neuro-ophthalmologist, and/or at the recommendation of the radiologist.
- Gadolinium is relatively contraindicated in pregnancy, MRI Brain without contrast (CPT® 70551) is supported.
- The AMA CPT manual does not describe nor assign any minimum or maximum number of sequences for any CT or MRI study. Both MRI and CT imaging protocols are often influenced by the individual clinical situation of the individual and additional sequences are not uncommon. There are numerous MRI sequences that are performed to evaluate specific clinical questions, and this technology is constantly undergoing development. Additional sequences, however, are still performed and coded under the routine MRI Brain CPT® 70551, CPT® 70552, or CPT® 70553.

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General Guidelines – CT Head (HD-1.4)

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- Scenarios in which MRI is contraindicated (i.e., pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)
- In urgent cases, CT Head, contrast as requested is medically necessary [CT Head without contrast (CPT® 70450) or CT Head with contrast (CPT® 70460) or CT Head without and with contrast (CPT® 70470)]
- CT Head without contrast (CPT® 70450) is medically necessary for:
  - Mass effect
  - Blood/blood products
  - Urgent/emergent settings due to availability and speed of CT
  - Trauma
  - Recent hemorrhage, whether traumatic or spontaneous
  - Bony structures of the head evaluations including dystrophic calcifications
  - Hydrocephalus evaluation and follow-up (some centers use limited non-contrast "fast or rapid MRI" (CPT® 70551) to minimize radiation exposure in children).
  - Prior to lumbar puncture in individuals
  - Evaluation of optic disc edema and/or papilledema, a non-contrast CT Head is useful to assess for space-occupying processes such as intracranial hemorrhage, mass effect, and hydrocephalus, See **Papilledema/Pseudotumor Cerebri (HD-17.1)** and **Eye Disorders and Visual Loss (HD-32.1)**

Evidence Discussion (HD-1.4)

- CT head is the preferred modality for evaluation of bony structures, acute intracranial hemorrhage, trauma, and detection of abnormalities associated with calcifications.[35,37,38,39]
- This modality is also useful for follow up of intracranial hemorrhage, hydrocephalus shunts, and post-operative follow-up.[35,37,38,39]
- CT head provides more rapid detection of intracranial abnormalities in urgent or emergent situations.[38,39,40]
- CT has less motion artifact than MRI due to its faster acquisition and better spatial resolution than MRI.
- Limitations of CT include lower early detection rates for occult fracture than MRI, ionizing radiation exposure, and lower contrast resolution than MRI.

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General Guidelines – CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5)

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Angiography imaging refers to both arterial and venous imaging. Vascular angiography is not the same imaging as diagnostic MRI.

- MRA Head is performed without contrast (CPT® 70544), with contrast (CPT® 70545), or without and with contrast (CPT® 70546)
- MRA Neck is performed without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549), depending on facility preference and protocols and type of scanner
- CTA Head is performed without and with contrast (CPT® 70496)
- CTA Neck is performed without and with contrast (CPT® 70498)
- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography).
  - If arterial and venous CT or MR studies are both performed in the same session, only **one** CPT® code is used to report both procedures.
  - If an arterial CTA or MRA study has been performed and subsequently a repeat study is needed to evaluate the venous anatomy, then this study is medically necessary.
  - If a venous CTV or MRV study has been performed and subsequently a repeat study is needed to evaluate the arterial anatomy, then this study is medically necessary.
- Medically necessary indications for CTA or MRA Head and Neck vessels include, but are not limited to the following:
  - Pulsatile tinnitus
  - Hemifacial spasm if consideration for surgical decompression
  - Evaluation of stroke or TIA (see **Stroke/TIA (HD-21.1)**) including collateral assessment
  - Trigeminal neuralgia (see **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10.1)**)
  - Cerebral venous sinus thrombosis (see **Cerebral Venous Sinus Thrombosis (HD-21.8)** and **Papilledema/Pseudotumor Cerebri (HD-17.1)**)
    - MRA without and with contrast is medically necessary for venous sinus thrombosis to differentiate total from subtotal occlusion.

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  - Aneurysm suspected with acute “thunderclap” headache syndrome and appropriate screening or evaluation of known subarachnoid hemorrhage and pseudoaneurysms (appropriate to limit CTA to include only the head to avoid unnecessary radiation to the individual)
  - Non-inflammatory vasculopathy, including radiation vasculopathy
  - Traumatic vascular injuries
  - Vascular malformations, vascular anatomic variants, and fistulas
  - Arterial dissections
  - Tumors of vascular origin or involving vascular structures
  - Surgical and radiation therapy localization, planning and neuronavigation
  - Evaluation for vascular intervention and follow-up including post-surgical/post-treatment vascular complications
  - Intra-cranial pre-operative planning if there is concern of possible vascular involvement or risk for vascular complication from procedure
  - Vasculitis and collagen vascular disease
  - Eagle Syndrome - Dynamic/positional CTA to assess for vascular compression (also known as bow-hunter's syndrome) (see **Eagle Syndrome (Neck-10.3)**)
  - NOTE: Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebral-basilar system.
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) is medically necessary for follow-up of aneurysm clipping or coiling procedures (see **Intracranial Cerebral Aneurysms (HD-12.1)**)
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) is medically necessary if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
  - There are high-risk scenarios including, but not exclusive to, fibromuscular dysplasia (FMD), Marfan disease, motor vehicle accident (MVA) with whiplash, or chiropractic manipulation.
- Other vascular imaging indications for headaches require additional information.
  - See **Stroke/TIA (HD-21.1)**, **Sudden Onset of Headache (HD-11.3)**, **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, and **Systemic Infections (HD-11.13)**

Evidence Discussion (HD-1.5)

- Indications for cervicocerebral computed tomography angiography (CTA) of the head and neck vessels include the diagnosis, characterization and/or surveillance of a variety of vascular conditions, including, but not limited to, arterial aneurysms,

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dissections, ischemic stroke and transient ischemic attacks, vasculitis, traumatic vascular injuries, pulsatile tinnitus, tumors of vascular origin, and prior to surgical intervention.[8] CTA may refer to arterial vessels (CTA) or evaluation of venous structures (CTV).[8]

- CTA may be used as the initial imaging modality or as a follow up study for characterizing known disease or assessing changes over time.[8]
- Depending on the indication, CTA may be limited to the head to avoid unnecessary radiation to the individual. Examples include surveillance of intracranial aneurysms (that are not located in the posterior circulation).[8]
- Risks of CTA include exposure to ionizing radiation; thus, magnetic resonance angiography (MRA) is available as an alternative to reduce radiation exposure. In addition, MRA is an alternative for individuals with iodinated contrast allergies or other contraindications to iodinated contrast.[8,41]
- Magnetic resonance angiography (MRA) indications also cover a variety of vascular conditions of the head and neck, for diagnosis, characterization, and surveillance, and may be used to evaluate either arterial (MRA) or venous structures (MRV).[41]
- MRA, as an alternative modality, is noninvasive, and does not require iodinated contrast.[41] Limitations include artifacts due to motion, slow or turbulent flow, and susceptibility effects, and claustrophobia.[41] MRA may be performed without contrast or with gadolinium contrast.[41] Gadolinium contrast administration is limited to those without renal impairment or known gadolinium contrast allergy.[41] Additionally, MRA may not be a feasible option for those with contraindications to MRI such as incompatible pacemakers, cochlear implants, neurostimulators or other devices.[33] In these scenarios, CTA may be the appropriate alternative.[8]

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General Guidelines – PET Coding Notes (HD-1.6)

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- Metabolic Brain PET should be reported as Metabolic Brain PET (CPT® 78608)
- Amyloid Brain PET should be reported as limited PET (CPT® 78811) or limited PET/CT (CPT® 78814)

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General Guidelines – Other Imaging Situations (HD-1.7)

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Persistent Nausea and Vomiting

Screening for Metallic Fragments Before MRI

Gender Affirming Care Head and Neck Surgical Planning

3D Rendering

Eagle Syndrome

CSF Leak with or without Headache

Evidence Discussion (HD-1.7)

Persistent Nausea and Vomiting

- Nausea and vomiting, persistent, unexplained and a negative GI evaluation: MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary

Screening for Metallic Fragments Before MRI

- Screening for metallic fragments before MRI should be done initially with plain x- ray.
  - The use of CT Orbital to rule out orbital metallic fragments prior to MRI is rarely necessary.
  - Plain x-rays are generally sufficient; x-ray detects fragments of 0.12 mm or more, and CT detects those of 0.07 mm or more.
- Plain x-ray is generally sufficient to screen for aneurysm clips

Gender Affirming Care Head and Neck Surgical Planning

- For gender affirming care procedure planning:
  - ANY or ALL the following pre-operative CT requests are medically necessary if the individual has a health plan benefit covering the gender affirming surgeries:
    - CT Maxillofacial without contrast (CPT® 70486)

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        - CT Orbits/Temporal bone without contrast (CPT® 70480)
        - CT Neck with contrast (CPT® 70491)
        - CT Head without (CPT® 70450)
        - 3D rendering (CPT® 76376 or CPT® 76377)
    - Pre-operative imaging is not supported if the gender affirming surgeries are not health plan covered benefits.
    - Requesting providers are encouraged to confirm eligibility with the member's health plan prior to service.

3D Rendering

- CPT® 76376 (3D rendering not requiring imaging post-processing on an independent workstation) or CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT® 76376 (3D rendering not requiring image post-processing on an independent workstation) is medically necessary in the following clinical scenarios:
    - Bony conditions:
        - Evaluation of congenital skull abnormalities in newborns, infants, and toddler (usually for pre-operative planning)
        - Complex joint fractures or pelvis fractures
        - Spine fractures (usually for pre-operative planning)
        - Complex facial fractures
    - Pre-operative planning for other complex surgical cases
    - Cerebral angiography: 3D rendering when performed in conjunction with conventional angiography (i.e., conventional 4 vessel cerebral angiography).
        - See **Cerebral Aneurysms (HD-12.1)**, **Arteriovenous Malformations (AVMs) and Related Lesions (HD-12.2)**, **Stroke/TIA (HD-21.1)**, and **Cerebral Vasculitis (HD-22.1)**[26]
- 3D Rendering (CPT® 76376 or CPT® 76377) for surgical planning and surgical follow up after craniotomy when ordered by surgical specialist or any provider in consultation with a surgical specialist.
- 3D Rendering indications in pediatric head imaging are identical to those in the general imaging guidelines.
- See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines

Eagle Syndrome

- See **Eagle Syndrome (Neck-10.3)** in the Neck Imaging Guidelines.

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- See **General Guidelines - CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5)** for vascular imaging related to Eagle Syndrome.

CSF Leak with or without Headache

- CSF Leak with or without headache, see **Low-Pressure Headache and CSF Leak (HD-11.15)**

Evidence Discussion (HD-1.7)

Neurologic evaluation of Nausea and vomiting

- In the evaluation of persistent, unexplained nausea and vomiting, an MRI Brain is supported after a negative GI evaluation.[2,3,4,43] Nausea and vomiting were reported as the initial symptom of a brain tumor in 5% of brain tumor cases.[44] During the time course until diagnosis, nausea and vomiting is present in 25% of brain tumor cases.[44]

Screening for metallic fragments

- The American College of Radiology White Paper on MR safety advised that all individuals who have a history of orbit trauma by a potential ferromagnetic foreign body for which they sought medical attention are to have their orbits cleared by either a plain x-ray orbit films (2 views) or by a radiologist's review and assessment of a prior CT or MR images obtained since the suspected traumatic event.[42,46] Screening for the presence of a metallic aneurysm clips with plain films of the skull is also recommended.[46] Although CT is more sensitive than plain films, the radiation dose is greatly increased.[42]

3D Rendering

- 3-D/rotational angiography, as part of cerebral angiography, is also useful for radiation dose reduction during diagnostic and interventional neuroradiology procedures.[22]

Gender affirming head and neck surgeries

- As the field has evolved, more centers are using frontal sinus setback as 90% of individuals require frontal bone osteotomy and setback, based on their frontal bone anatomy.[31,32] For individuals requiring bony manipulation, a fine-cut, non-contrast craniofacial CT scan from the vertex to the hyoid bone is used to map the bony framework.[31,32] Virtual surgical planning improves efficiency, safety, and accuracy for frontal sinus setback and mandibular angle reduction surgeries.[45] CT Neck is medically necessary for laryngoplasty surgeries.[6]

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Taste and Smell Disorders (HD-2)

Guideline

Taste and Smell Disorders (HD-2.1)
References (HD-2)

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Taste and Smell Disorders (HD-2.1)

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- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without and with contrast (CPT® 70551) **AND/OR** MRI Orbit/Face/Neck without (CPT® 70540) or MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary with unexplained unilateral or bilateral anosmia (inability to perceive odor) or dysgeusia (complete or partial loss of taste)
- CT Maxillofacial (CPT® 70486, CPT® 70487 or CPT® 70488) is medically necessary initially if sinus or facial bone disorders are suspected
- For individuals who test positive for SARS-CoV-2 (see: **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)** and **Stroke/TIA (HD-21.1)**)

Evidence Discussion (HD-2.1)

- Initial imaging of the olfactory nerve and pathway for unexplained unilateral or bilateral anosmia or for dysgeusia should utilize MRI brain and/or MRI orbits, face and neck.[1,6] These imaging studies are supported by clinical evidence for the identification and characterization of a potential cranial nerve lesion.[1,6]
- CT of the sinuses and face may be superior to identify fractures, inflammatory sinus disease, and other bony lesions in some cases.[1]

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Ataxia (HD-3)

Guideline

Ataxia (HD-3.1)
References (HD-3)

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Ataxia (HD-3.1)

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- Common manifestations include poor coordination, an abnormal (including wide-based) gait, abnormal finger to nose testing, abnormal rapid alternating movements, abnormal eye movements, and/or difficulty with navigation of stairs and around corners.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary in all individuals with ataxia:
  - MRI Cervical without contrast or without and with contrast (CPT® 72141 or CPT® 72156) **AND/OR** MRI Thoracic without contrast or without and with contrast (CPT® 72146 or CPT® 72157) **AND/OR** MRI Lumbar Spine without contrast or without and with contrast (CPT® 72148 or CPT® 72158) may be added if spinal disease is suspected
  - If these symptoms are acute and stroke is suspected, see **Stroke/TIA (HD-21.1)**
  - If MS is suspected, see **Multiple Sclerosis (MS) (HD-16.1)**
  - CT Head without contrast (CPT® 70450) **AND/OR** CT Orbit/Temporal Bone without contrast (CPT® 70480) is medically necessary in addition if these symptoms are acute following head trauma, (see also: **Head Trauma (HD-13.1)**)
- If brain tumor is suspected, see **Primary Central Nervous System Tumors (ONC-2.1)** in the Oncology Imaging Guidelines.
- For suspected Normal Pressure Hydrocephalus, see **Normal Pressure Hydrocephalus (NPH) (HD-8.4)**

Background and Supporting Information

- In general, MRI is preferred over CT, unless there is a history of acute trauma or contraindication to MRI. For all other causes, MRI provides better visualization of the cerebellum and posterior fossa.

Evidence Discussion (HD-3.1)

- MRI Brain is the preferred initial imaging modality for evaluation of ataxia when a central nervous system cause is suspected.[1,4] MRI of the spinal cord, to include the cervical and thoracic spine, may also be added if clinically indicated.[1,5]
- CT head is not recommended for the initial evaluation of non-traumatic ataxia due to inferior soft tissue resolution when compared to MRI Brain. In addition, MRI Brain provides better visualization of the cerebellum and posterior fossa and is more sensitive for the detection of posterior fossa infarcts.[1]

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- In general, MRI is preferred over CT, unless there is a history of acute trauma or a contraindication to MRI.[1]

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Mental Health Disorders and Mental Status Change (HD-4)

Guideline

Autism Spectrum Disorders (HD-4.0)
Mental Health Related Disorders (HD-4.1)
Mental Status Change (HD-4.2)
References (HD-4)

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Autism Spectrum Disorders (HD-4.0)

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- This group of diagnoses, including Asperger syndrome, is classified as pervasive development disorders (PDD). These diagnoses are established on clinical criteria, and no imaging study can confirm the diagnosis.
- Comprehensive evaluation for autism might include history, physical exam, audiology evaluation, speech, language, and communication assessment, cognitive and behavioral assessments, and academic assessment.
  - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for **ANY** of the following:
    - New or worsening cognitive decline or focal neurologic findings documented on a pertinent physical
  - PET imaging is considered not medically necessary in the evaluation of individuals with autism spectrum disorders.

Evidence Discussion (HD-4.0)

- While the diagnosis of Autism Spectrum Disorder is based on behavioral signs and symptoms, MRI Brain without and with contrast is medically necessary for new or worsening focal neurological findings and/or loss of developmental milestones and/or regression. In these clinical situations, advanced imaging may be used to adjust an individual's treatment plan, without which their development may continue to regress.[4]
- PET is considered not currently medically necessary in the evaluation of individuals with autism spectrum disorder PET imaging in this scenario would unnecessarily expose individuals to radiation and provide no clinical utility related to autism spectrum disorder.[5]

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Mental Health Related Disorders (HD-4.1)

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- Mental health diagnoses, to include Attention Deficit Hyperactivity Disorder (ADHD), do not routinely require advanced imaging.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **OR** CT Head without contrast (CPT® 70450) is medically necessary for the exceptions listed below:
  - Acute mental status change, disturbance in consciousness or arousal state
  - Psychotic disorders (including schizophrenia), bipolar disorder and related disorders in the following clinical presentations:
    - Acute psychosis
    - Late onset over age 40
    - Presentation of acute psychiatric symptoms with comorbid serious medical illness
    - Non-auditory hallucinations (e.g., visual, tactile, olfactory) with no known etiology
    - Nonresponse to adequate medication trials
    - Symptoms of an organic brain disorder (e.g., focal deficits, severe headache, or seizures)
- Prior to electroconvulsive therapy (ECT) treatment, the following are medically necessary to screen for intracranial disease: MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450)
- Deep Brain Stimulation Therapy for psychiatric disorders is considered not medically necessary, except for medically refractory Obsessive Compulsive Disorder (OCD).
  - Medically necessary imaging prior to Deep Brain Stimulation (DBS) therapy for medically refractory Obsessive Compulsive Disorder (OCD):
    - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** unlisted CT procedure code (CPT® 76497)

Evidence Discussion (HD-4.1)

- There is no role for advanced imaging in mental health workup (including ADHD). Unnecessary imaging has detrimental effects in that it provides no positive impact on outcomes/management and does expose individuals to unnecessary radiation, contrast, and financial strain.
- It would be appropriate to utilize Advanced imaging (CT or MRI) in the following conditions.
  - Acute mental status change, disturbance in consciousness or arousal state[2]

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  - Psychiatric disorders with the following clinical presentations[2]:
    - Acute psychosis
    - Late onset over age 40
    - Presentation of acute psychiatric symptoms with comorbid serious medical illness
    - Non-auditory hallucinations of unknown etiology
    - Nonresponse to adequate medication trials
    - Symptoms of an organic brain disorder (e.g., focal deficits, severe headache, or seizures)
- Advanced imaging may be medically necessary for electroconvulsive therapy clearance and prior to deep brain stimulation for medically refractory obsessive compulsive disorder.[10]

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Mental Status Change (HD-4.2)

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After a detailed history, which includes onset, duration, and timeframe (i.e., constant vs intermittent) **AND** bedside neurologic exam that includes a mental status evaluation providing a description of the level of alertness, other characteristics and/or cognitive testing, the following are medically necessary:

Indication	Medically Necessary Imaging
Acute or worsening mental status change, initial or repeat imaging	• CT Head without contrast (CPT® 70450) **OR** • If setting is urgent, CT Head contrast as requested (CPT® 70450 OR CPT® 70460 OR CPT® 70470) **OR** • If MRI is contraindicated, CT Head contrast as requested (CPT® 70450 OR CPT® 70460 OR CPT® 70470) • *CT Head permitted even with prior MRI Brain imaging* **OR** • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) • *MRI Brain permitted even with prior head CT imaging*

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Indication	Medically Necessary Imaging
*Presence of any Red Flag, including: • Sudden language, focal motor, or sensory deficit – **Stroke/TIA (HD-21.1)** • Headache – **Headaches with Red Flags (HD-11.2)** • Hypertensive urgency – **Stroke/TIA (HD-21.1)** and **Sudden Onset of Headache (HD-11.3)** • Fever/tachycardia, possible meningitis, or other CNS infection – **CNS and Head Infection (HD-14.1)** • COVID-19 – **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)** • Coagulopathy or anticoagulant use- **Abnormal Blood Clotting (HD-11.9)** • Pregnancy or post-partum – **Pregnancy (HD-11.10)** • Known malignancy – **Low Grade Gliomas (ONC-2.2)**, **High Grade Gliomas (ONC-2.3)** and **Brain Metastases (ONC-31.3)** • Trauma- **Head Trauma (HD-13.1)** • Non-auditory hallucinations – **Mental Health Related Disorders (HD-4.1)** • Suspected increased intracranial pressure – **Papilledema/Pseudotumor Cerebri (HD-17.1)** and **Hydrocephalus Shunts (HD-11.14)** • Seizure/suspected seizure – **Epilepsy/ Seizures (HD-9.1)**	*See relevant guideline

Background and Supporting Information

This section refers to acute/subacute mental status change, which generally involves signs and symptoms which begin over minutes to days, and includes changes in behavior and alertness, agitation, and/or confusion – as opposed to chronic, progressive cognitive decline, as in dementia.

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Acute mental status change or encephalopathy is characterized by changes in behavior or alertness, agitation, confusion, as opposed to chronic, progressive cognitive decline, such as dementia related disorders.

Delirium and psychosis are defined as follows:

- Delirium is a disorder of acute onset involving deficits in attention, awareness, and cognition that fluctuate in severity over time. These are often associated with psychomotor disturbance, altered sleep cycle, and emotional variability. These disturbances may be hyperactive (restlessness, agitation) or hypoactive (psychomotor retardation, lethargy) and there may be accompanying fever and/or autonomic symptoms (tachycardia, sweating), depending on the underlying cause.
- Psychosis is a disorder of impaired reality, characterized by hallucinations, delusions, or both, without insight into their pathologic nature. It may be associated with disorganized behavior, thought blocking, illogicality, tangentiality, perseveration, and/or neologisms.

The purpose of the initial assessment is to characterize the etiology. This may include toxic/metabolic (e.g., hypoglycemic, drug exposures), structural (e.g., trauma, stroke, hypoxic-ischemic, hydrocephalus, tumor), paroxysmal (e.g., seizure, psychiatric), and inflammatory (e.g., infectious, autoimmune).

Of note, even a seemingly mild, reversible brain insult superimposed upon a chronic pathophysiologic process may cause a sudden mental status change, and head imaging may or may not be necessary, depending on the provider's pre-test suspicion of a significant new diagnosis.

Vagal Nerve Stimulators (VNS), which are FDA approved for treatment of depression, are included among potential treatments, which also include medication trials.

Evidence Discussion (HD-4.2)

- Advanced brain imaging is supported for acute onset of mental status change, or worsening symptoms in the setting of a known intracranial process with MRI Brain with or without a previous CT Head.[1]
- Advanced imaging supported for mental status change with precipitating factors including suspected seizure, COVID related symptoms, head trauma, stroke, mass, or known malignancy, suspected increased intracranial pressure, intracranial infection, hypertensive emergency, presence of coagulopathy, pregnancy and postpartum period, associated headache.[2]
- According to the ACR, advantages of MRI for altered mental state include: 1) higher sensitivity for detection of ischemia, encephalitis, subtle cases of SAH; and 2) enhancement of pathology compared with CT. The disadvantages of MRI in this clinical scenario are the same as with MRI in general, including individual

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inconvenience (longer examination time), imaging quality is susceptible to individuals' movements, and implanted devices that are not MRI safe.[2]

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Chiari and Skull-Base Malformations (HD-5)

Guideline

Chiari Malformations (HD-5.1)
Chiari II Malformations (Arnold Chiari Malformation) (HD-5.2)
Chiari III and IV Malformations (HD-5.3)
Basilar Impression/Basilar Invagination (HD-5.4)
Platybasia (HD-5.5)
Evidence Discussion (HD-5)
References (HD-5)

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Chiari Malformations (HD-5.1)

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Indication	Medically Necessary Imaging
Initial evaluation for suspected or known Chiari malformations	• MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) or MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) or MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) or MRI Lumbar Spine without and with contrast (CPT® 72158)
Repeat imaging for one of the following: • New or worsening signs or symptoms • Surgical procedure is actively being considered • At the discretion of or in consultation with a neurologist and/or neurosurgeon coordinating the individual's care	• MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) or MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) or MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) or MRI Lumbar Spine without and with contrast (CPT® 72158)

- Familial screening is NOT medically necessary for Chiari Malformations.
- For CSF flow imaging, see **CSF Flow Imaging (HD-24.4)**

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Background and Supporting Information

Chiari I malformations involve caudal displacement or herniation of the cerebellar tonsils. Chiari I may be associated with syringomyelia and rarely with hydrocephalus. Most cases are asymptomatic and discovered incidentally on a head scan performed for another indication. When symptoms are present, they are usually nonspecific but can include headache, lower cranial nerve palsies, or sleep apnea.

Chiari II malformations are more severe than Chiari I malformations. These individuals usually present at birth. Myelomeningocele is always present, and syringomyelia and hydrocephalus are extremely common.

Chiari III malformations include cerebellar herniation into a high cervical myelomeningocele. Chiari IV malformation refers to complete cerebellar agenesis. Both Chiari III and IV malformations are noted at birth and are rarely compatible with life.

Repeat brain and spine imaging in individuals with Chiari I malformations and known syringomyelia or hydromyelia is highly individualized.

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Chiari II Malformations (Arnold Chiari Malformation) (HD-5.2)

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- See **Chiari Malformations (HD-5.1)**

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Chiari III and IV Malformations (HD-5.3)

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- See **Chiari Malformations (HD-5.1)**

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Basilar Impression/Basilar Invagination (HD-5.4)

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Medically necessary imaging for suspected or known Basilar Impression or Basilar Invagination:

- MRI Brain without contrast (CPT® 70551) **AND/OR** MRI Cervical Spine without contrast (CPT® 72141)
- If surgery is being considered, CT Head (CPT® 70450) **AND/OR** CT Cervical Spine (CPT® 72125) without contrast are also medically necessary **AND/OR** MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** CTA Head (CPT® 70496) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR** CTA Neck (CPT® 70498).
- One-time screening of first-degree relatives with MRI Brain without contrast (CPT® 70551) is medically necessary.

Background and Supporting Information

Basilar impression involves malformation of the occipital bone in relation to C1-2 (cervical vertebrae 1 and 2). The top of the spinal cord is inside the posterior fossa and the foramen magnum is undersized. Over time, this can lead to brain stem and upper spinal cord compression. Basilar impression can also be associated with the Chiari malformation, producing very complex anatomical abnormalities.

Basilar invagination is an abnormality at the craniovertebral junction, either congenital or degenerative, resulting in the odontoid prolapsing into the already limited space of the foramen magnum. It is commonly associated with conditions such as Chiari malformation, syringomyelia, and Klippel-Feil syndrome.[12]

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Platybasia (HD-5.5)

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Medically necessary imaging indications for suspected or known Platybasia:

- MRI Brain without contrast (CPT®70551) or CT Head without contrast (CPT®70450)
- If surgery is being considered,
  - CT Head (CPT®70450) **AND/OR**
  - CT Cervical Spine (CPT®72125) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
  - CTA Neck (CPT® 70498)

Background and Supporting Information

Platybasia is a flattening malformation of the skull base, in which the clivus has a horizontal orientation.

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Evidence Discussion (HD-5)

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- A CT scan of the head is less sensitive than an MRI of the brain for evaluation of intracranial structures, including major structural abnormalities of the posterior fossa.[14,15]
- Neuroimaging in the initial evaluation of Chiari malformation should include the spinal cord due to the common occurrence or increased frequency of associated conditions such as cervical syrinx and tethered cord.[1,2,3,4,9,10]
- For initial evaluation, treatment planning, and follow up, MRI is the preferred modality for malformations of the brain and cervicocranial junction.[7,14,15,16] MRI is ideal for evaluating soft tissues, neural structures, and ligaments.[7,13]
- As congenital brainstem and cerebellar anomalies are associated with spinal anomalies, MRI of the complete spine is helpful for diagnosis, follow up, and treatment planning.[1,2,3,4,14]
- A phase-contrast CSF flow study at the craniocervical junction is a supportive study for evaluation of Chiari malformation.[5,16]
- Evaluation of cervicojunction anomalies, including basilar invagination and platybasia, may require more than one modality for diagnosis and surgical planning.[7,12,13] CT characterizes osseous anatomy and may be helpful for surgical planning.[7,13] MRI is preferred for evaluation of the soft tissues, neural structures, and ligaments for these conditions.[7,13] As craniocervical junction anomalies may lead to compression of adjacent vascular structures, CT- or MR-Angiography of the head and neck are useful for surgical planning.[13]

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Facial Palsy (Bell's Palsy)/Hemifacial Spasm (HD-6)

Guideline

Facial Palsy (HD-6.1)
Hemifacial Spasm (HD-6.2)
Evidence Discussion (HD-6)
References (HD-6)

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Facial Palsy (HD-6.1)

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- MRI Brain without contrast (CPT® 70551) (with attention to posterior fossa and IACs) or MRI Brain without and with contrast (CPT® 70553) (with attention to posterior fossa and IACs) **AND/OR** MRI Orbit/Face/Neck without contrast (CPT® 70540) or MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is/are medically necessary when unexplained facial paresis/paralysis is associated with any of the following "red flag" clinical scenarios:
  - Trauma to the temporal bone
  - History of tumor, cancer, HIV, or Lyme disease
  - No improvement in 8 weeks
  - No full recovery in 3 months
  - Gradual onset over weeks to months
  - Vertigo or hearing loss
  - Bilateral involvement
  - Other atypical or inconsistent features including:
    - Second episode of paralysis on the same side
    - Paralysis of isolated branches of the facial nerve
    - Paralysis associated with other cranial nerves
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for known sarcoidosis with suspected neurosarcoid or CNS involvement is supported, (see also **Autoimmune/ Paraneoplastic Encephalitis & NeuroInflammatory Disorders (HD-14.3)**)
- CT Orbit/Temporal Bone without contrast (CPT® 70480) is medically necessary, in the presence of red flags, to assess osseous integrity of the temporal bone, to characterize fractures, pre-surgical anatomy, inflammatory middle ear disease, bone tumor, facial canal foraminal expansion and/or bone erosion.
- CT Orbit/Temporal Bone with contrast (CPT® 70481) is medically necessary, in the presence of red flags, for suspected tumors and/or infection.
- CT Maxillofacial without contrast (CPT® 70486) is medically necessary to assess bony facial nerve canal **OR** with contrast (CPT® 70487) when infection or tumor are suspected, if requested per institutional protocol.
- MRA Head without contrast (CPT® 70544), with contrast (CPT® 70545), or without and with contrast (CPT® 70546) **AND/OR** MRA Neck without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549) **OR** CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) is/are medically necessary for clinically suspected stroke (see **General Guidelines- CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5)** and **Stroke/TIA (HD-21.1)**).

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Background and Supporting Information

Typical features of Bell's palsy include variable initial ipsilateral temporal and auricular pain before facial weakness, onset over 72 hours, ipsilateral complete facial weakness, and an otherwise normal neurological and systemic examination. There is usually slow improvement over several months. Unless "red flags" are present, imaging is not necessary.

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Hemifacial Spasm (HD-6.2)

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- For hemifacial spasm, facial synkinesis, or blepharospasm, the following are medically necessary:
  - MRI Brain without and with contrast (CPT® 70553)
  - Add CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) for consideration of vascular decompression surgical procedure to clarify the vascular anatomy in individuals who have failed conservative medical management
- For tardive dyskinesia, see **Movement Disorders (HD-15.1)**

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Evidence Discussion (HD-6)

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- Facial nerve palsy/Bell's Palsy, commonly referred to as Bell's Palsy, does not routinely require imaging as recommended by the American Academy of Neurology and the American Academy of Otolaryngology Head and Neck Surgery Foundation.[1,2,3,4,6] Complete recovery typically occurs within 3 to 6 months.[12]
- When imaging is medically necessary, MRI is the preferred modality for evaluating the facial nerve from its origin in the brainstem, through its intracranial and extracranial segments.[2] This would include imaging of the brain, face or both areas concurrently.[2] MRI is useful to exclude structural causes of facial nerve paralysis in the setting of red flags.[2]
- Imaging is reserved for cases with "red flags," which include atypical, recurrent, or persistent cases.[2,4,10,12] Limiting imaging to those with "red flags" avoids unnecessary radiation exposure, identification of incidental findings, contrast reactions, and unnecessary costs.[1,7] The risk of limiting imaging includes missing identifiable and treatable causes of facial paralysis. To mitigate this risk, clinical follow up is recommended at 3 months.[1]
- MRI has sensitivity ranging from 73% to 100% in detecting peripheral spread of tumor.[2]
- As the facial nerve courses through the temporal bone, CT temporal bone is useful to identify temporal bone fractures, bony anatomy, bone erosion and for surgical planning.[2]
- Vascular imaging is helpful if stroke is clinically suspected.[2]
- For evaluation of hemifacial spasm, MRA allows characterization of vascular loops compressing the facial nerve, with sensitivity >95% and correlates well with surgical findings.[2,3]

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References (HD-6)

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Recurrent Laryngeal Palsy/Vocal Cord Palsy (HD-7)

Guideline

Recurrent Laryngeal Palsy/Vocal Cord Palsy (HD-7.1)

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Recurrent Laryngeal Palsy/Vocal Cord Palsy (HD-7.1)

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- See **Recurrent Laryngeal Nerve Palsy in Neck-7.1**

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Mild Cognitive Impairment (MCI) and Dementia (HD-8)

Guideline

Mild Cognitive Impairment (MCI) and Dementia - MRI/CT (HD-8.1)
Mild Cognitive Impairment (MCI) and Dementia - PET (HD-8.2)
Lewy Body Dementia (LBD) - SPECT and PET (HD-8.3)
Normal Pressure Hydrocephalus (NPH) (HD-8.4)
Imaging Related to Alzheimer's Treatment with Amyloid Reduction Medications (HD-8.5)
References (HD-8)

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Mild Cognitive Impairment (MCI) and Dementia - MRI/CT (HD-8.1)

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Mild Cognitive Impairment (MCI) Evaluation

Dementia Evaluation

Imaging Related to Alzheimer's Treatment with Amyloid Reduction Medications

Background and Supporting Information

Evidence Discussion (HD-8.1)

- For subjective cognitive decline with normal cognitive testing, advanced imaging is not medically necessary.

Mild Cognitive Impairment (MCI) Evaluation

For the evaluation of memory loss and/or cognitive impairment due to suspected Mild Cognitive Impairment (MCI) when treatment with amyloid reducing medications is not documented.

- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) or CT Head without contrast (CPT® 70450) is medically necessary when all the following criteria are met:
  - A detailed history of cognitive decline with impairments confirmed by family members or others with knowledge of the individual's status

    OR
  - Abnormal mental status test score or neuropsychological test results consistent with mild cognitive impairment/mild neurocognitive disorder
    - Examples of abnormal mental status test results consistent with MCI include neuropsychological testing demonstrating a decline in cognitive function and/or the following test scores:
      - Montreal Cognitive Assessment (MOCA) 19-25,
      - Saint Louis University Mental Status (SLUMS) score for high school education 21-26, for less than high school education 20-24, or
      - Mini Mental State Exam (MMSE) 22-27

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- See *Background and Supporting Information*for additional information regarding the diagnosis of MCI

Dementia Evaluation

For the evaluation of memory loss and/or cognitive impairment due to suspected dementia when treatment with amyloid reducing medications is not documented.

- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) or CT Head without contrast (CPT® 70450) is medically necessary when all the following criteria are met:
  - Initial clinical diagnosis of dementia has been established
  - A detailed history and neurologic exam including mental status testing should be performed prior to advanced imaging
  - Date of onset of symptoms with documentation of 6 months of cognitive decline
  - Documentation of a decline in cognitive function obtained with either:
    - A detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status

      OR
    - Abnormal mental status test score consistent with dementia or neuropsychological test results consistent with dementia/major neurocognitive disorder
      - Examples of abnormal bedside mental status testing scores include:
        - Mini-Mental State Exam (MMSE) <26,
        - Montreal Cognitive Assessment (MoCA) <26,
        - Memory Impairment Screen (MIS) <5,
        - St. Louis University Mental Status (SLUMS) <21, or
        - Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score >2
  - Presumptive causes of dementia have been excluded
    - Cannot occur exclusively during bouts of delirium
    - Cannot be explained by another mental disorder

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Imaging Related to Alzheimer's Treatment with Amyloid Reduction Medications

The following includes imaging to determine eligibility for treatment with FDA-approved amyloid reducing medications and imaging during treatment:

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Indication	Medically Necessary Imaging
To determine eligibility for treatment	• Baseline (within the last 12 months) MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) • Repeat MRI Brain prior to drug initiation is medically necessary if requested
On donanemab therapy, prior to the 2nd, 3rd, 4th, and 7th infusions	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)
Follow-up while on donanemab therapy with radiographically observed amyloid-related imaging abnormality (ARIA) per the treating neurologist *See Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging
Neurologic signs and/or symptoms occurring while on treatment with donanemab	• CT Head without contrast (CPT® 70450) OR • MRI Brain without contrast (CPT® 70551) OR • MRI Brain without and with contrast (CPT® 70553) • Follow up MRI Brain is medically necessary after CT Head if requested
When transitioning to a different amyloid targeting monoclonal antibody, the individual should meet all initial treatment criteria	

Lecanemab (Leqembi®)

Indication	Medically Necessary Imaging
To determine eligibility for treatment	• Baseline (within the last 12 months) MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) • Repeat MRI Brain prior to drug initiation is medically necessary if requested
On lecanemab therapy, prior to the 3rd, 5th, 7th, and 14th infusions	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)
Follow up while on lecanemab therapy with radiographically observe amyloid-related imaging abnormality (ARIA) per the treating neurologist *See Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging
Neurologic signs and/or symptoms occurring while on treatment with lecanemab	• CT Head without contrast (CPT® 70450) OR • MRI Brain without contrast (CPT® 70551) OR • MRI Brain without and with contrast (CPT® 70553) • Follow up MRI Brain is medically necessary after CT Head if requested
When transitioning to a different amyloid targeting monoclonal antibody, the individual should meet all initial treatment criteria	

- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as (CPT® 0865T or CPT® 0866T).
    - These studies are considered not medically necessary in the evaluation of dementia.
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as 3D rendering (CPT® 76376 and CPT® 76377).
    - These studies are considered not medically necessary in the evaluation of dementia.
- For amyloid PET requests in the treatment with amyloid reducing medications, see **Mild Cognitive Impairment (MCI) and Dementia – PET (HD-8.2)**
- For acute mental status change, see **Mental Status Change (HD-4.2)** and/or **Stroke/ TIA (HD-21.1)**
- For evaluation of Lewy Body Dementia, see **Lewy Body Dementia (LBD) – SPECT and PET (HD-8.3)**
- For the evaluation of Normal Pressure Hydrocephalus, see **Normal Pressure Hydrocephalus (HD-8.4)**
- For evaluation of rapidly progressive memory loss see **Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)**
- For evaluation of suspected vascular dementia see **Cerebrovascular Disease (HD-21.1)**

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Background and Supporting Information

Subjective cognitive decline (SCD) refers to individuals who are concerned about a cognitive decline but perform normally on cognitive testing.

Mild Cognitive Impairment (MCI), also referred to as mild neurocognitive disorder, is marked by focal or multifocal cognitive impairment with minimal impairment of instrumental activities of daily living that do not cross the threshold for dementia.

Cognitive domains affected by MCI may include memory, attention, language, visuospatial processing, executive function, and social behavior. Examples of memory loss include forgetfulness, repeating questions, forgetting recent events or conversations. Attention deficits may include periods of decreased alertness or distractibility. Language impairment may be displayed as word finding difficulties, difficulty communicating and/or understanding. Visuospatial processing impairment may present as getting lost. Executive dysfunction including difficulty completing multi-step commands, planning, or using appliances. Concerning social behaviors include impulsiveness and inappropriate behaviors.

The functional impact of cognitive symptoms should be evaluated. Individuals with mild cognitive impairment may have difficulties completing instrumental activities of daily living such as managing finances, cooking, medications, and using transportation.

On neuropsychological testing, individuals with mild neurocognitive disorder perform at least 1 standard deviation below appropriate norms. Of note, a highly educated individual who scores within the normal range on screening tests, for example, a score of 29/30 on the MMSE, may show objective deficits in cognitive function on formal neuropsychological testing, consistent with a diagnosis of MCI.

Dementia, or major neurocognitive disorder, as defined by the Diagnostic and Statistical Manual of Mental Disorders (DSM-5), refers to significant cognitive decline, with impairment in cognitive performance in domains including complex attention, executive function, learning and memory, language, perceptual-motor skills, or social cognition.

On neuropsychological testing, individuals with a major neurocognitive disorder perform 2 or more standard deviations below appropriate norms.

Impairment in activities of daily living include bathing, dressing, grooming, feeding and toileting.

Presumptive causes of cognitive decline include metabolic abnormalities such as hypothyroidism, uncontrolled diabetes, vitamin deficiencies, and/or untreated disorders such as sleep apnea.

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Evidence Discussion (HD-8.1)

- The primary role of neuroimaging in the work up of individuals diagnosed with mild cognitive impairment or dementia is to exclude other serious differential diagnosis such as tumors, subdural hematomas, and normal pressure hydrocephalus.[2,3] The American Academy of Neurology (AAN) recommended the use of non-contrast CT or Brain MRI for aiding in the diagnosis of dementia.[3,31] Cross sectional imaging may also identify characteristic brain atrophy patterns found in common neurodegenerative diseases and vascular insults.[2] CT imaging may also be used when MRI scans are contraindicated.
- Volumetric MRI Brain for the diagnosis of dementia is not currently recommended for routine clinical use by the AAN.[3,31] There remains a significant evidence gap in the literature regarding clinical validation of volumetric MRI in the diagnosis of dementia. Their use remains limited to research studies.[31,32]

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Mild Cognitive Impairment (MCI) and Dementia - PET (HD-8.2)

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Brain PET CPT® Codes	Definition
78608	Used to report FDG PET metabolic brain studies for dementia, seizure disorders, and brain tumors
78609	Used to report PET brain perfusion studies, primarily used for brain tumors
78811	Limited PET, used for amyloid or tau PET, static imaging to measure amyloid and tau
78814	Limited PET/CT, used for amyloid or tau PET, static images to measure amyloid or tau

Metabolic (FDG) PET Brain CPT® 78608

Amyloid PET Brain CPT® 78814 or 78811

Tau PET Brain CPT® 78814 or 78811

Perfusion PET Brain CPT® 78609

Background and Supporting Information

Evidence Discussion

Metabolic (FDG) PET Brain (CPT® 78608)

Mild Cognitive Impairment

- Metabolic (FDG) PET (CPT® 78608) is medically necessary in the evaluation of for Mild Cognitive Impairment (MCI) when Alzheimer's Disease is suspected and all the following criteria are met:
  - Established diagnosis of mild cognitive impairment based on clinical history, physical examination, and cognitive testing, which may include neuropsychological testing.

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- Documentation of cognitive decline obtained by either:
  - A detailed history of cognitive decline with impairments confirmed by family members or others with knowledge of the individuals status

    OR
  - Abnormal mental status test score or neuropsychological test results consistent with mild cognitive impairment/mild neurocognitive disorder
    - Examples of abnormal mental status test results consistent with MCI include:
      - Montreal Cognitive Assessment (MOCA) score 19-25
      - Saint Louis University Mental Status (SLUMS) score for high school education 21-26, for less than high school education 20-24
      - Mini Mental State Exam (MMSE) score 22-27
- Results of any structural brain imaging (MRI Brain or CT Head) previously performed.

Dementia

- Metabolic (FDG) PET Brain (CPT® 78608) is medically necessary in the evaluation of dementia to distinguish between Alzheimer's disease (AD) and Frontotemporal Dementia (FTD) when all the following criteria are met:
  - Established diagnosis of dementia:
    - Date of onset of symptoms with documentation of 6 months of cognitive decline
    - Documentation of a decline in cognitive function obtained with either:
      - A detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status

        OR
      - Abnormal mental status testing score consistent with dementia
        - Examples of abnormal bedside mental status testing scores include:
          - Mini-Mental State Exam (MMSE) <26,
          - Montreal Cognitive Assessment Survey (MoCA) <26,
          - Memory Impairment Screen (MIS) <5,
          - St. Louis University Mental Status (SLUMS) <21, or
          - Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score >2
  - Results of any structural imaging (MRI Brain or CT Head) previously performed
  - Meets diagnostic criteria for AD and FTD
  - The results are expected to clarify the diagnosis between FTD and AD and help guide future treatment
  - Cause of clinical symptoms is uncertain

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- Evaluation has ruled out specific alternative neurodegenerative disease or causative factors
  - Cannot occur exclusively during bouts of delirium
  - Cannot be explained by another mental disorder
- FDG PET brain (CPT® 78608) is otherwise considered not medically necessary for the purpose of diagnosis and management of other forms of dementia including, but not limited to, Parkinson's disease, normal pressure hydrocephalus, and chronic traumatic encephalopathy.
  - For requests related to Lewy Body Dementia, see **Lewy Body Dementia (LBD) – SPECT and PET (HD-8.3)**
- FDG-PET(CPT® 78608)/MRI Brain without contrast (CPT® 70551) OR MRI Brain without and with contrast (CPT® 70553) imaging may be considered on a case-by-case basis for those imaging centers that will utilize FDG-PET/MRI during an initial evaluation (instead of MRI alone) and who also have a standardization of imaging protocol.

Amyloid PET Brain (CPT® 78814 or 78811)

- Amyloid Brain PET is medically necessary when a diagnosis of MCI or dementia due to Alzheimer's disease is suspected based on cognitive testing, and evaluation including structural brain imaging has been completed, in ANY of the following clinical scenarios:
  - Individuals with MCI or dementia who are <65 years of age and in whom AD is suspected
  - Individuals with MCI or dementia that could be consistent with amnestic AD pathology with onset at ≥65 years of age
  - Individuals with MCI or dementia that could be consistent with AD but has atypical clinical features
  - Individuals with MCI or dementia with equivocal or inconclusive results on CSF biomarkers
  - To inform the prognosis of individuals with MCI due to suspected AD pathology
  - To determine eligibility for treatment with amyloid targeting therapy and to monitor response (as listed in the criteria in the tables below)

Imaging Related to Alzheimer's Treatment with Amyloid Reduction Medications

The following tables include imaging to determine eligibility for treatment with FDA-approved amyloid reducing medications and imaging during treatment.

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Indication	Medically Necessary Imaging
ALL the following criteria must be met to determine eligibility for treatment: • Individual ≥59 years of age and ≤86 years of age • MCI or mild dementia due to AD • MMSE score ≥20 and ≤28 (or equivalent comparison test score) • Progressive change in memory function for at least 6 months • Baseline MRI Brain results are available to the ordering provider • No history of prior intracerebral hemorrhage greater than 1 cm, severe white matter disease, or vasogenic edema • Not currently taking another amyloid reducing drug • Prescribed by a neurologist or in consultation with a neurologist	Amyloid PET Brain (CPT® 78811 or CPT® 78814)
To monitor treatment response: • Follow up imaging during treatment at 6, 12, and 18 months	Amyloid PET Brain (CPT® 78811 or CPT® 78814)
When transitioning to a different amyloid targeting monoclonal antibody, the individual should meet all initial treatment criteria	

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Lecanemab (Leqembi®)

Indication	Medically Necessary Imaging
ALL the following criteria must be met to determine eligibility for treatment: • Individual is ≥50 years of age and ≤90 years of age • MCI or mild dementia due to AD • Qualifying test scores include Mini-Mental Status Exam (MMSE) with score ≥22, Clinical Dementia Rating global score of ≥0.5 or 1.0, Clinical Dementia Rating-Sum of Boxes (CDR-SB) ≥0.5, and/or Memory Box score of 0.5 or greater (or equivalent comparison test score) • Baseline MRI Brain results are available to the ordering provider • No history of brain hemorrhage, bleeding disorder, history of seizures, or recent history (within 12 months) of stroke or transient ischemic attacks • Not currently taking another amyloid reducing drug • Prescribed by a neurologist or in consultation with a neurologist	Amyloid PET Brain (CPT® 78811 or CPT® 78814)
To monitor treatment response: • Post-treatment imaging at 18 months • If treatment is continued after 18 months, a follow up study is medically necessary between 12-18 months	Amyloid PET Brain (CPT® 78811 or CPT® 78814)
When transitioning to a different amyloid targeting monoclonal antibody, the individual should meet all initial treatment criteria	

Tau PET Brain (CPT® 78814 or 78811)

- Tau PET is medically necessary for evaluation of cognitive impairment in individuals with suspected AD with atypical clinical presentation when all the following criteria are met:

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- Established diagnosis of mild cognitive impairment or dementia
- Evaluation has excluded other causes
- Results of structural brain imaging are available to the ordering provider

See *Background and Supporting Information* regarding suspected AD with atypical presentation

Perfusion PET Brain (CPT® 78609)

Perfusion PET Brain imaging (CPT® 78609) is not medically necessary in the evaluation of dementia.

- For cerebral amyloid angiopathy, see **Stroke/TIA (HD-21.1)**

Background and Supporting Information

An atypical clinical presentation of Alzheimer's Disease (AD) is one that meets core clinical criteria of cognitive impairment in multiple domains, which interfere with instrumental activities of daily living and functioning, but has sudden or rapid onset or the onset occurs at a younger age (<65).

Atypical AD variants include:
- Early onset
- Amnestic variant early onset AD
- Visuospatial variant AD (posterior cortical atrophy (PCA))
- Language variant AD (lopogenic variant primary progressive aphasia (PPA))
- Behavioral variant/dys-executive variant AD
- Motor variant AD corticobasal syndrome due to AD

The frontotemporal dementias (FTDs) are a group of neurodegenerative disorders that differ from Alzheimer's disease. The basic pathology involves accumulation of tau proteins in the brain rather than amyloid. Onset tends to be younger (less than 65) and progression usually more rapid than in senile dementia-Alzheimer type (SDAT). There is no treatment, and the medications used to help memory in Alzheimer's disease are not effective.

There are several subtypes of FTD; most common are the behavioral variant with early loss of executive functions, impaired judgment disinhibition and impulsivity, and the semantic variant with primary and progressive loss of language ability (primary progressive aphasia type FTD). Other less common subtypes include progressive supranuclear palsy, corticobasal syndrome, and FTD associated with motor neuron disease.

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Diagnosis of FTD is based on clinical features, neuropsychological testing, and brain imaging (preferably MRI) to rule out other structural disease. Metabolic (FDG) PET Brain is helpful by demonstrating patterns of abnormality more consistent with FTD than Alzheimer's disease.

Recent research has examined the utility of PET/MRI for evaluation of individuals with dementia. Due to the prolonged acquisition time, motion during a PET may lead to artifacts such as blurring of the images. Use of co-registration of PET with MRI can lead to better PET assessment especially with quantitative measurements. Utilization of PET/MRI provides greater confidence in imaging reading by permitting greater structural correlation. A recent study compared FDG-PET/CT and FDG-PET/MRI in a memory disorders clinic. This study identified more individuals with cerebrovascular disease (stroke) and better cortical atrophy characterization. The authors found that PET/MRI provided significant improvement in diagnosis and management of individuals in which dementia is a consideration.

Evidence Discussion (HD-8.2)

The Alzheimer's Association workgroup has revised the criteria for diagnosis and staging of Alzheimer's Disease (AD).[35] The workgroup defined Alzheimer's disease as a biological process, rather than a clinical syndrome, which allows diagnosis in the asymptomatic phase, based on positive biomarkers (Core 1 biomarkers).[35] These Core 1 biomarkers include Amyloid PET and cerebrospinal fluid biomarkers.[35] An abnormal Core 1 biomarker is sufficient to establish a diagnosis of AD.[2,35]

Structural, functional, and molecular brain imaging in individuals with MCI helps to identify potentially treatable structural abnormalities and identify imaging features that may confer an increased risk of transformation to a dementia.[2] In addition, molecular imaging helps inform prognosis, for patients and their families, and allows early treatment initiation.[2,4]

Amyloid PET is positive in 60% of individuals with MCI.[2] Greater cognitive decline is noted in those with MCI and a positive amyloid PET scan, and they are more likely to progress to AD than those with a negative amyloid PET.[2,4] Amyloid PET also results in a change in diagnosis in up to 44% of individuals, therefore increasing diagnostic confidence.[2]

Amyloid PET has a pooled sensitivity of 0.90 and specificity of 0.80 for diagnosis of AD, and a pooled sensitivity of 0.84 and specificity of 0.62 for predicting conversion of MCI to dementia.[36]

Molecular brain imaging is particularly helpful in confirming a diagnosis of AD with atypical clinical presentation.[2] A positive amyloid PET in early onset or atypical AD can lead to a change in diagnosis of 66.8%, improve confidence in diagnosis in 81.5% and

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change management in 80% of cases.[2,37] The use of amyloid PET led to changes in management in 63.5% of Medicare members with dementia.[4,5]

The Society of Nuclear Medicine Imaging and Molecular Imaging released Appropriate Use Criteria (AUC) for amyloid PET which included the following clinical scenarios[2,4]:

- Individuals with MCI or dementia who are <65 years of age and for whom AD is suspected
- Individuals with MCI or dementia consistent with amnestic AD pathology with onset ≥65 years of age
- Individuals with MCI or dementia that could be consistent with AD but has atypical features
- Individuals with MCI or dementia with equivocal or inconclusive results on CSF biomarkers
- To inform the prognosis of patients due to suspected AD pathology
- To determine eligibility for treatment with amyloid-targeting therapy and to monitor treatment response

Tau PET is considered appropriate for the evaluation of individuals presenting with MCI or a dementia syndrome that could be consistent with AD pathology but has atypical features.[2,4] In these clinical scenarios, tau uptake correlates with cognitive function and is helpful to diagnose atypical clinical phenotypes of AD.[2]

The work groups supports tau PET in atypical cases as a tau binding pattern can help establish AD as the cause of impairment and the spatial pattern of tau uptake matches brain regions clinically affected.[4] Furthermore, a high tau burden is associated with more rapid clinical progression.[4]

FDG PET Brain in the evaluation of MCI shows a pattern of hypometabolism consistent with AD in up to 79% of cases.[2] In addition, an abnormal pattern predicts conversion from MCI to AD with an accuracy over 80%.[2] Conversely, a negative FDG PET reduces the likelihood of progression to dementia.[2]

FDG-PET brain accurately discriminates Alzheimer's disease individuals from normal subjects with a sensitivity of 96% and specificity of 100%.[2]

Metabolic (FDG) PET brain is helpful by demonstrating patterns of abnormality more consistent with FTD than Alzheimer's disease. FDG PET brain has a sensitivity of 86% and specificity of 97.6% in evaluating individuals with FTD.[2] The use of FDG PET increases diagnostic accuracy and confidence for both AD and FTD.[2] It is particularly helpful in cases of diagnostic uncertainty.[2]

Recent research has examined the utility of PET/MRI for evaluation of individuals with dementia. Due to the prolonged acquisition time, motion during a PET may lead to artifacts such as blurring of the images. Use of co-registration of PET with MRI can lead to better PET assessment especially with quantitative measurements.[23,25]

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Utilization of PET/MRI provides greater confidence in reading images by permitting greater structural correlation. A recent study compared FDG-PET/CT and FDG-PET/MRI in a memory disorders clinic.[24] The main findings were that FDG PET/MRI revealed more vascular pathology in 35% of individuals, induced a change of the interpretation of FDG PET in 17% of individuals and was considered to influence management in 22% of individuals.[24]

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Lewy Body Dementia (LBD) - SPECT and PET (HD-8.3)

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SPECT Brain Scan

Metabolic (FDG) PET Brain

Background and Supporting Information

Evidence Discussion (HD-8.3)

SPECT Brain Scan

- SPECT Brain Scan (CPT® 78803 or CPT® 78830) is medically necessary for evaluation of suspected Lewy Body Dementia when all the following are met:
  - Established diagnosis of dementia:
    - Date of onset of symptoms with documentation of 6 months of cognitive decline
    - Documentation of decline in cognitive function obtained with either:
      - a detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status

        OR
      - abnormal mental status test score consistent with dementia or neuropsychological test results consistent with dementia/major neurocognitive disorder
        - Examples of abnormal bedside mental status testing scores include:
          - Mini-Mental State Exam (MMSE) <26,
          - Montreal Cognitive Assessment Survey (MoCA) <26,
          - Memory Impairment Screen (MIS) <5,
          - St. Louis University Mental Status (SLUMS) <21, or
          - Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia >2
  - Presumptive causes of dementia have been excluded
    - Cannot occur exclusively during bouts of delirium
    - Cannot be explained by another mental disorder

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- Results of any structural imaging (MRI or CT Head) performed

Metabolic (FDG) PET Brain

- Metabolic (FDG) PET brain (CPT® 78608) is medically necessary to differentiate Lewy Body Dementia (LBD) from Alzheimer's Disease (AD) when all the following criteria are met:
  - Established diagnosis of dementia:
    - Date of onset of symptoms with documentation of 6 months of cognitive decline
    - Documentation of decline in cognitive function obtained with either:
      - a detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status

        OR
      - abnormal mental status test score consistent with dementia or neuropsychological test results consistent with dementia/major neurocognitive disorder
        - Examples of abnormal bedside mental status testing scores include:
          - Mini-Mental State Exam (MMSE)<26,
          - Montreal Cognitive Assessment Survey (MoCA) <26,
          - Memory Impairment Screen (MIS) <5,
          - St. Louis University Mental Status (SLUMS) <21, or
          - Eight- item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score >2
  - Presumptive causes of dementia have been excluded
    - Cannot occur exclusively during bouts of delirium
    - Cannot be explained by another mental disorder
  - Results of any structural imaging (MRI or CT Head) performed

Background and Supporting Information

- Dementia with Lewy bodies is often hard to diagnose because its early symptoms may resemble those of Alzheimer's or a psychiatric illness. Over time people with LBD often develop similar symptoms due to the presence of Lewy bodies in the brain.
  - Clinicians and researchers may use the "1-year rule" to help make a diagnosis. If cognitive, psychiatric, emotional, and/or personality symptoms appear at the same time as or at least a year before movement problems/Parkinsonism, the diagnosis is dementia with Lewy bodies. If cognitive problems develop more than a

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year after the onset of movement problems, Parkinson's disease, the diagnosis is Parkinson's disease dementia (PDD).

- Core Clinical Symptoms
  - Dementia
  - Movement problems/Parkinsonism
  - Cognitive fluctuations
  - Visual hallucinations
  - REM sleep behavior disorder
- Supportive Clinical Symptoms
  - Extreme sensitivity to antipsychotic medications
  - Falls, fainting
  - Severe problems with involuntary functions (maintaining blood pressure, incontinence, constipation, loss of smell)
  - Changes in personality and mood (depression, apathy, anxiety)

Test Results Supporting Diagnosis

- Abnormal ^{123}iodine-MIBG myocardial scintigraphy showing reduced communication of cardiac nerves
- Sleep study confirming REM sleep behavior disorder without loss of muscle tone

Evidence Discussion (HD-8.3)

- For suspected Lewy Body Dementia (LBD), a CT or MRI head is appropriate as the initial imaging study.[14,31]
- To increase diagnostic accuracy of LBD, SPECT modalities are helpful for differentiating LBD from Alzheimer's dementia.[2,6,14,15]
- Functional imaging of the dopamine transporter (DAT) (Iodine-123 Ioflupane) using SPECT shows a deficiency in the nigrostriatal pathway in LBD. This is considered a second line imaging test after cross-sectional imaging has excluded other pathology, such as vascular lesions along the nigrostriatal pathway, which can lead to abnormal DAT images with false positive results.[14]
- An abnormal DAT-SPECT scan has a sensitivity of 78.7% and a specificity of 90.4% for probable LBD.[2,14]
- Metabolic (FDG) PET imaging demonstrates features which would support a diagnosis of LBD. Hypometabolism in the occipital lobe differentiates LBD[2] from AD.[2,38] In addition, the "cingulate island sign", which refers to the preservation of FDG metabolism in the posterior cingulate gyrus, is considered a biomarker for LBD.[2,39]

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Normal Pressure Hydrocephalus (NPH) (HD-8.4)

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- CT Head without contrast (CPT® 70450) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary if the individual has at least two symptoms involving gait abnormality (see *Background and Supporting Information*), urinary incontinence, or dementia **AND**
  - The clinical symptoms cannot be completely explained by other neurological or non-neurological disease, **AND**
  - There is no apparent preceding disorder that would cause hydrocephalus
- The components of Dementia are delineated in **Mild Cognitive Impairment (MCI) and Dementia – MRI/CT (HD-8.1)**, but include:
  - Results of testing and/or neuropsychological testing can be performed when history and mental status examination cannot provide a confident diagnosis.
  - Examples of abnormal mental status testing such as Mini-Mental State Exam (MMSE) with score <26, Montreal Cognitive Assessment Survey (MoCA) with score <26, Memory Impairment Screen (MIS) with score <5, the St. Louis University Mental Status (SLUMS) with score <21, or the Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score >2.
  - Presumptive causes or etiology/ies of dementia
    - Cannot occur exclusively during bouts of delirium
    - Cannot be explained by another mental disorder
- MRI Brain (CPT® 70551, CPT® 70552, or CPT® 70553) is not medically necessary for the diagnosis of NPH if a CT has been performed. However, MRI Brain is medically necessary if needed for pre-surgical planning.
  - After neuro imaging, the next steps are CSF sampling, drainage, and dynamics.
- Follow-up imaging for individuals diagnosed with NPH with a shunt should follow **Hydrocephalus Shunts (HD-11.14)**, **Low-Pressure Headache and CSF Leak (HD-11.15)**, or **Nuclear Medicine (HD-36.1)**

Background and Supporting Information

Normal Pressure Hydrocephalus (NPH) seen typically in the elderly. It comprises a triad of symptoms: cognitive dysfunction, incontinence of urine, and gait disturbance (typically a “magnetic”, small-step, shuffling, or broad-based gait). The reported neuroradiologic marker for this is ventriculomegaly (enlarged ventricles) in the brain. Unfortunately, these symptoms and this neuroradiologic finding is common in the elderly, making the

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diagnosis of NPH in any given individual problematic. It is radiographically common and clinically rare.

Evidence Discussion (HD-8.4)

- Initial neuroimaging for the evaluation of suspected Normal Pressure Hydrocephalus (NPH) includes CT Head or MRI Brain in individuals with clinical symptoms and no explanation for hydrocephalus.[2,21]
- Only a single modality study is medically necessary. The initial best modality is MRI Brain because of its higher intrinsic soft tissue resolution and because it can often be used as a pre-surgical exam.[2,21]
- By using the appropriate single best test, we avoid duplicate imaging and unnecessary radiation to the lens of the eye and other head and neck structures.[2]
- Cine MRI showing hyperdynamic aqueductal CSF flow can also help in identifying shunt-responsive NPH individuals. The benefit of this exam is that it offers us functional information about CSF flow and can help improve individual outcomes.[2,21,34]

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Imaging Related to Alzheimer's Treatment with Amyloid Reduction Medications (HD-8.5)

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For structural brain imaging related to Alzheimer's treatment with amyloid reducing medications, see **Mild Cognitive Impairment (MCI) and Dementia – MRI/CT (HD-8.1)**.

For Amyloid PET brain imaging related to Alzheimer's treatment with amyloid reducing medications, see **Mild Cognitive Impairment (MCI) and Dementia- PET (HD-8.2)**.

Background and Supporting Information

Amyloid reduction medications are medically necessary for the treatment of Mild Cognitive Impairment (MCI) due to Alzheimer's disease and mild, early- stage Alzheimer's disease.[25]

These medications are monoclonal antibodies that selectively bind to aggregated forms of beta amyloid. The accumulation of amyloid plaques in the brain is a defining pathophysiologic feature of Alzheimer's disease. In clinical trials, these medications reduce amyloid beta plaque compared with placebo.[25]

Amyloid related imaging abnormalities (ARIA) have been caused by these medications. ARIA usually occurs early in treatment and may be asymptomatic although serious and life-threatening events may occur. Screening MRI Brain prior to treatment initiation and periodic monitoring during treatment is recommended. For moderate to severe ARIA, treatment may be suspended. Once ARIA is identified on a brain MRI, follow up MRIs are medically necessary to assess for radiographic resolution and/or symptom resolution with the imaging time frame determined by the treating physician. Resumption of dosing is guided by clinical judgment.[25]

ARIA may be further characterized as ARIA with edema (ARIA-E) or ARIA with hemosiderin (ARIA-H). ARIA-E presents on MRI as brain edema or sulcal effusions. ARIA-H includes microhemorrhage and superficial siderosis. ARIA-E and ARIA-H may occur simultaneously.[25]

Although ARIA is usually asymptomatic, symptoms associated with ARIA include headache, confusion, visual changes, dizziness, nausea, aphasia, weakness, gait difficulty and seizures, including status epilepticus. Focal neurologic deficits may also occur.[25] The risk of ARIA is increased in apolipoprotein E ε4 (ApoE ε4) homozygotes.[25]

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Leqembi® dosing after 18 months may be administered every 2 weeks or transitioned to a maintenance regimen every 4 weeks.

Transitioning from one amyloid targeting monoclonal antibody to another should be done when the individual has been off of the prior drug for 5 half-lives prior to initiation of the alternative drug. The half-life of aducanumab is 24.3 days; the half-life of lecanemab is 9.5 days, and the half-life of donanemab is 11.8 days. Individuals should be off of aducanumab for approximately 4 months or off lecanemab or donanemab for approximately 6 weeks prior to initiation of a different amyloid targeting monoclonal antibody.

Evidence Discussion (HD-8.5)

- Structural brain imaging in the work up of individuals diagnosed with dementia is primarily to exclude other significant intracranial abnormalities. A brain MRI will assist with the diagnosis of dementia by excluding structural pathology such as tumors or subdural hematomas.[2]
- Amyloid related imaging abnormalities (ARIA) have been associated with treatment by amyloid reduction medications. ARIA usually occurs early in treatment and may be asymptomatic although serious and life-threatening events may occur. Screening brain MRI prior to treatment initiation and periodic monitoring during treatment is recommended.[17,27] For moderate to severe ARIA, treatment may be suspended. Once ARIA is identified on a brain MRI, follow- up MRIs are medically necessary to assess for radiographic resolution and/or symptom resolution with the imaging time-frame determined by the treating physician.[17,27] Resumption of dosing is guided by clinical judgment.[17,27]
- Amyloid PET brain is a form of molecular imaging, which uses a tracer that binds to amyloid plaques in the brain. At the present time, the use of Amyloid PET brain is limited to confirming the presence of amyloid in the brain, in those with mild cognitive impairment due to Alzheimer's disease or mild dementia due to Alzheimer's disease, prior to treatment with amyloid reducing medications.

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Epilepsy/Seizures (HD-9)

Guideline

Epilepsy/Seizures (HD-9.1)
Perioperative Evaluations for Drug-Resistant Epilepsy (HD-9.2)
References (HD-9)

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Epilepsy/Seizures (HD-9.1)

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for:
  - evaluation of new onset seizures
  - refractory or drug-resistant seizures
  - new neurologic deficit or no return to previous neurologic baseline
  - change in the type of seizure
  - additional evaluation, if CT Head was performed for an initial evaluation for new onset seizure, MRI (as described above) is medically necessary for additional evaluation
  - follow-up, MRI Brain with “Epilepsy Protocol” is medically necessary
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **OR** CT Head without contrast (CPT® 70450) is medically necessary for:
  - repeat imaging at discretion of the neurologist or neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon.
- CT Head without contrast (CPT® 70450) is medically necessary for:
  - evaluation of structural findings in seizure etiologies that contain dystrophic calcifications, such as with oligodendrogliomas and tuberous sclerosis
  - acute setting of seizure evaluation
- CT Head (contrast as requested) (CPT®70450, CPT®70460, **OR** CPT®70470) is medically necessary when:
  - MRI is contraindicated
  - request is urgent
- For seizure and/or altered mental status associated with head trauma, see **Head Trauma (HD-13.1)**
- 3D T1 and/or FLAIR sequences are useful in improving lesion detection for the diagnosis and monitoring of epilepsy. 3D T1 and FLAIR sequences do not require an additional CPT® for 3D rendering (CPT® 76376 and CPT® 76377). [12]
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be requested as (CPT® 0865T or CPT® 0866T)
    - These studies are not medically necessary in the evaluation of epilepsy.

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Evidence Discussion (HD-9.1)

- The use of advanced imaging is medically necessary for the initial evaluation of adults with seizure. Unenhanced CT is more readily available so is usually the initial imaging examination performed for adults presenting with first seizure. In the acute setting this primary exam is utilized to exclude conditions requiring urgent or emergent intervention, such as a bleed.[5] CT is also appropriate if MRI is contraindicated and to evaluate seizure foci that contain dystrophic calcifications, such as oligodendrogliomas and tuberous sclerosis, yet the overall success of CT in detecting focal lesions in epilepsy is low, at approximately 30%.[1,5] In studies where individuals were evaluated with both MRI and CT, CT failed to detect potentially epileptogenic lesions identified on MRI 16-42% of the time1.[5] Therefore, MRI of the brain is the study of choice to evaluate new-onset seizures (when available), refractory or drug-resistant seizures, prior to discontinuation of anti-epileptic therapy, and known seizure with change in semiology.
- If CT is initially performed, it can be followed by an MRI. If an MRI not using the "Epilepsy Protocol" is initially performed, it can be followed by an MRI with the Epilepsy Protocol for greater sensitivity of detection of epileptogenic lesions. The failure rate for detection of lesions improving from 39% to 91% with epilepsy-trained radiologist reading MRI images obtained using a specialized, epilepsy protocol.[1,12,23]

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Perioperative Evaluations for Drug-Resistant Epilepsy (HD-9.2)

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- ANY or ALL the following requests are medically necessary, obtained concurrently or sequentially, for consideration of potential surgery:
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553)
    - Follow-up MRI Brain after a previous routine study if performed with special "Epilepsy Protocol" (typically 3T or 7T magnet, thin sections with angled slices through hippocampus and temporal lobes)
  - Metabolic FDG PET (CPT® 78608)
    - Metabolic PET/MRI is MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without (CPT® 70553) co-registered **WITH** FDG-PET Brain (CPT® 78608) and is medically necessary for pre-surgical evaluation of refractory seizure when requested by neurosurgeon or neurologist or any provider in consultation with a neurosurgeon or neurologist.
  - Metabolic PET/MRI is MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without (CPT® 70553) co-registered **WITH** FDG-PET Brain (CPT® 78608) and is medically necessary for pre-surgical evaluation of refractory seizure when requested by neurosurgeon or neurologist or any provider in consultation with a neurosurgeon or neurologist.
  - Ictal SPECT (CPT® 78803 or 78830)
  - Functional MRI (fMRI) (CPT® 70555 or CPT® 70554)
    - If MRA Head (CPT® 70544) is medically necessary but Functional MRI (CPT® 70554 or CPT® 70555) was erroneously ordered, then CPT® 70544 may be substituted when appropriate, (see **Functional MRI (fMRI) (HD-24.2)**)
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without and with (CPT® 70553)
    - Medically necessary if co-registered with Magnetoencephalography (MEG)
  - Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
    - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be requested as (CPT® 0865T or CPT® 0866T)
      - These studies are considered not medically necessary in the evaluation of epilepsy.
  - 3D rendering CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT® 76376 (3D rendering not requiring image post-processing on an independent workstation) is not medically necessary for

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epilepsy surgery alone, since 3D rendering can be obtained as part of the MRI Brain epilepsy protocol, unless complicated surgical repair considerations involving craniotomy are required.

- When non-invasive EEG monitoring is insufficient, intracranial monitoring with stereo-EEG or grids/strips and depth electrodes is medically necessary with additional imaging for neuronavigation. See **Neurosurgical Imaging (HD-28.1)** and **Neuronavigation (HD-28.2)**
  - Post-operative imaging including after intracranial (EEG) monitoring per neurosurgeon or neurologist or any provider in consultation with neurosurgeon or neurologist.
- See **Primary Central Nervous System Tumors-General Considerations (ONC-2.1)** in the Oncology Imaging Guidelines and/or **Neurosurgical Imaging (HD-28.1)** for additional imaging requests for surgery

Background and Supporting Information

- Magnetoencephalography (MEG) plays an important role in clarifying the significance of abnormalities seen on both structural and functional imaging, for the purpose of epileptogenic zone localization for surgical planning. When used in conjunction with other techniques, MEG plays a major role in the non-invasive epilepsy surgery evaluation. Currently, the guidelines only require review for the MRI co-registered with MEG.
- MEG followed by co-registration with Brain MRI is referred to as Magnetic Source Imaging (MSI)

Below are examples of surgical treatment or an interventional modality that may be under active consideration for individuals with intractable epilepsy (not all inclusive):

- Focal Resection
  - Temporal Lobe Resection
  - Extratemporal Resection
- Lesionectomy
- Multiple Subpial Transections
- Laser Interstitial Thermal Therapy (LITT)
- Anatomical or Functional Hemispherectomy and Hemispherotomy
- Corpus Callosotomy
- Stereotactic Radiosurgery
- Neurostimulation Device Implantations (Neuromodulation) including
  - Vagus Nerve Stimulation (VNS)
  - Responsive Neurostimulation (RNS) system also known as NeuroPace
  - Deep Brain Stimulation (DBS)

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Evidence Discussion (HD-9.2)

- MRI Head for the initial imaging of individuals with known seizure disorder requiring surgical planning to identify the seizure focus including tumor, hippocampal sclerosis, and vascular lesions.[1] Follow-up MRI after a previous standard protocol study if performed with special "Epilepsy Protocol" can provide additional information.[1]
- FDG-PET/CT Brain may be complementary as a functional tool to structural imaging using MRI to localize the focus of refractory seizure activity, with reported sensitivities of PET in the assessment of temporal lobe epilepsy ranging from 87% to 90% and extra-temporal lobe epilepsy ranging from 38% to 55%.[1]
- PET/MRI, performed as MRI Brain without contrast, or without and with contrast, co-registered with FDG-PET brain, increased the sensitivity of brain MRI in 60% of non-lesional individuals and is therefore supported for pre-surgical evaluation of refractory seizures.[27]
- Ictal SPECT, Functional MRI (fMRI) and MRI Brain co-registered with Magnetoencephalography (MEG) are also useful to further identify the seizure focus as well as eloquent areas of the cortex that are essential for language, motor function and memory in surgical candidates when done as a replacement for the higher risk Wada test or direct electrical stimulation mapping.[14,15,16]

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Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10)

Guideline

Trigeminal Neuralgia/Trigeminal Neuropathy (HD-10.1)
Glossopharyngeal Neuralgia/Glossopharyngeal Neuropathy (HD-10.2)
Evidence Discussion (HD-10)
References (HD-10)

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Trigeminal Neuralgia/Trigeminal Neuropathy (HD-10.1)

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- MRI Brain without and with contrast (CPT® 70553) (with special attention to the skull base) or MRI Brain without contrast (CPT® 70551) **AND/OR** facial imaging, MRI Orbit/Face/Neck without contrast (CPT® 70540) or MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary for:
  - symptoms of trigeminal neuropathy
  - trigeminal neuralgia
  - trigeminal neuralgia which involves the ophthalmic nerve, (periorbital or forehead pain), once post-herpetic neuralgia (a complication of shingles) has been excluded by history
- CT Maxillofacial without contrast (CPT® 70486) **OR** CT Maxillofacial with contrast (CPT®70487) is medically necessary for evaluating the skull base and neural foramina
- Contrast-enhanced navigation protocol CT (CPT® 76497) is medically necessary for gamma knife stereotactic radiosurgery for trigeminal neuralgia, (see also, **Neuronavigation (HD-28.2)** and **Post-Operative Imaging (HD-28.3)**) for post-treatment imaging studies
- MRI Cervical spine without contrast (CPT® 72141) **OR** MRI Cervical spine without and with contrast (CPT® 72156) is medically necessary for suspected lesion of the spinal trigeminal tract and nucleus.
- MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) **OR** CTA Head (CPT® 70496) is medically necessary for:
  - trigeminal neuralgia (vascular imaging may be obtained concurrently with structural brain imaging)
  - failed medical treatment
  - surgical planning

Background and Supporting Information

The differential diagnosis of facial pain is extensive, complex, and difficult, and there is considerable case-to-case variation in optimal imaging pathway.

Symptoms of trigeminal neuropathy include facial pain, facial numbness, and/or weakness of the muscles of mastication.

Trigeminal neuralgia, also known as tic douloureux (the involuntary wincing associated with the occurrence of pain), refers to sudden, severe, shooting "electrical" pains along

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one or more sensory divisions of the trigeminal nerve, provoked by movements such as chewing, or by external stimuli, such as wind blowing or touching the face.

The spinal trigeminal tract and nucleus extend from the midpons caudally into the upper cervical cord at the C2-4 levels. For suspected lesions of the spinal trigeminal tract and nucleus, imaging the brain stem and the cervical spinal cord is supported.

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Glossopharyngeal Neuralgia/ Glossopharyngeal Neuropathy (HD-10.2)

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- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) **AND/OR** MRI Orbit/Face/Neck without and with contrast (CPT® 70543) or MRI Orbit/Face/Neck without contrast (CPT® 70540) is medically necessary for suspected glossopharyngeal neuralgia or neuropathy
- CT Neck with contrast (CPT® 70491) is medically necessary to delineate skull-base erosion, deep space neck masses, calcifications, the skull base bony anatomy, and/or the stylohyoid ligament (see also **Eagle Syndrome (Neck-10.3)**)
- MRA Head with contrast (CPT® 70545), or MRA Head without and with contrast (CPT® 70546), **AND/OR** MRA Neck with contrast (CPT® 70548), or MRA Neck without and with contrast (CPT® 70549) is medically necessary to assess for neurovascular compression for the evaluation of glossopharyngeal neuralgia

Background and Supporting Information

- Glossopharyngeal neuralgia presents as severe pain in the throat and neck, classically triggered by swallowing.
- Glossopharyngeal neuropathy may present with pain, dysphagia, loss of gag reflex, impaired taste, and impaired sensation along posterior one-third of the tongue and/or inability to elevate the palate.

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Evidence Discussion (HD-10)

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- The American Academy of Neurology recommends routine use of MRI in the evaluation of individuals with trigeminal neuralgia.[4] Neuroimaging identifies structural causes in up to 15% of individuals. The most commonly identified abnormalities include cerebellopontine angle tumors and multiple sclerosis plaques.[4]
- MRI brain and/or MRI orbits, face and neck are necessary for direct visualization of the entire course of the trigeminal nerve.[5]
- MRA head, when combined with MRI brain for evaluation of vascular compression of the trigeminal nerve, has sensitivity of 97%-100% and specificity of 100%.[5] CTA is less commonly performed concurrently with MRI of the trigeminal nerve.[5]
- CT maxillofacial may be complementary to MRI in characterizing skull-base erosions, calcifications, and skull-base foramina.
- In the evaluation of glossopharyngeal neuralgia, MRI of the brain and/or MRI orbits, face and neck, allows direct visualization of the entire course of the glossopharyngeal nerve. Imaging should include the pharynx and larynx to exclude a neck mass. To further evaluate bony anatomy, calcifications, and the stylohyoid ligament, CT neck is also appropriate. MRA head and neck is helpful to exclude neurovascular compression in individuals with glossopharyngeal neuralgia.[5]

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Headache (HD-11)

Guideline

Headache General Guidelines (HD-11.0)
Headache and Suspected Vascular Dissection (HD-11.1)
Headaches with Red Flags (HD-11.2)
Sudden Onset of Headache (HD-11.3)
Trigeminal Autonomic Cephalgias (HD-11.4)
Skull Base, Orbit, Periorbital or Oromaxillary (HD-11.5)
Suspected Intracranial Extension of Sinusitis or Mastoiditis (HD-11.6)
New Headache Onset Older than Age 50 (HD-11.7)
Cancer or Immunosuppression (HD-11.8)
Abnormal Blood Clotting (HD-11.9)
Pregnancy (HD-11.10)
Physical Exertion (HD-11.11)
Headaches Associated With Head Trauma (HD-11.12)
Systemic Infections (HD-11.13)
Hydrocephalus Shunts (HD-11.14)
Low-Pressure Headache and CSF Leak (HD-11.15)
Cervicogenic Headaches Including Occipital Neuritis/Neuralgia (HD-11.16)
Advanced Imaging Indications Related To Migraines (HD-11.17)
Evidence Discussion (HD-11)
References (HD-11)

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Headache General Guidelines (HD-11.0)

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- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) or CT Head without contrast (CPT® 70450) are medically necessary for any of the following:
  - Headache accompanied by seizures, vomiting, focal neurological complaints including dizziness, visual change, altered mental status, or acute hypertension
  - Abnormal examination findings (including, but not limited to, altered mental status, papilledema, focal signs or symptoms including unilateral weakness or sensory loss, hyperreflexia, clonus, increased tone, Hoffman or Babinski sign, loss of coordination, seizures, gait disturbance, cranial nerve abnormality, vision loss, nystagmus, dysarthria, dysphagia, fever, meningismus)
  - For papilledema, see **Papilledema/Pseudotumor Cerebri (HD-17.1)**)
- Chronic headache with significant change in character, severity, or frequency of headache (for example: progressively worsening headache over a period of days or weeks, transformation of established migraine to chronic daily headaches), the following are medically necessary:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - CT Head without contrast (CPT® 70450)
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) is medically necessary as an addition to evaluate the recent onset of a progressive, severe, daily headache, with or without papilledema and concern for cerebral venous sinus thrombosis.
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only **ONE** CPT® code should be used to report both procedures.
- Advanced imaging of the head is **NOT** medically necessary for any of the following:
  - Primary headache disorder in the absence of focal neurological deficits or "red flags" (see **Headaches with Red Flags (HD-11.2)** and **Advanced Imaging Indications Related to Migraines (HD-11.17)**)
  - Newly diagnosed migraine or tension-type headache with a normal neurologic exam or for chronic stable headache including migraine with no neurologic deficit.
  - Duplex Ultrasound Carotid Arteries (CPT® 93880) does not have a role in the evaluation of headaches (including migraines), except for suspected carotid dissection (see **Initial Imaging (PVD-3.1)** in the Peripheral Vascular Disease

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Imaging Guidelines, **Headache and Suspected Vascular Dissection (HD-11.1),** and **Stroke/TIA (HD-21.1)**)

Background and Supporting Information

- The yield of detecting abnormal, treatable lesions by CT or MRI in individuals with headache but normal neurological exam has been found to be low

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Headache and Suspected Vascular Dissection (HD-11.1)

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- CTA Neck (CPT® 70498) and MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) are medically necessary in the evaluation for headache with suspected carotid or vertebral artery dissection and in certain high-risk scenarios including, but not exclusive to: Fibromuscular dysplasia (FMD), Marfan disease, acute MVA with whiplash, and acute headache and/or neck pain due to chiropractic manipulation.
  - CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) is medically necessary if there is concern for extension of a carotid dissection to the skull base or above.
  - Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebro-basilar system.
- MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) is medically necessary if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon).
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, or CPT® 70498) is medically necessary if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- Other vascular imaging indications for headaches require additional information.
  - See **Stroke/TIA (HD-21.1)**, **Sudden Onset of Headache (HD-11.3)**, **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, and **Systemic Infections (HD-11.13)**

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Headaches with Red Flags (HD-11.2)

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- Headaches with any of the following Red Flags
  - If any of the below unusual symptoms or history are present, advanced imaging studies are medically necessary (see relevant section):
    - Cancer history or immunosuppression (see **Cancer or Immunosuppression (HD-11.8)**)
    - Sudden onset (see **Sudden Onset of Headache (HD-11.3)**)
    - New onset age >50 (see **New Headache Onset Older than Age 50 (HD-11.7)** and **Migraine Exceptions (HD-11.17)**)
    - History of head trauma (see **Headaches Associated with Head Trauma (HD-11.12)**, and **Head and Facial Trauma (HD-13)**)
    - Headache precipitated by cough or Valsalva, physical exertion, or sexual activity (see **Physical Exertion (HD-11.11)**)
    - Currently pregnant (including pregnancy and the immediate postpartum period) (see **Pregnancy (HD-11.10)**)
    - Hypercoagulable state or bleeding disorder (see **Abnormal Blood Clotting (HD-11.9)**)
    - New persistent headache (see **Migraine Exceptions (HD-11.17)**)
    - Headache awakens individual from sleep (see **Sudden Onset of Headache (HD-11.3)**)

Background and Supporting Information

Aura symptoms may accompany or precede a headache within 60 minutes and may include, but are not exclusive to, the following symptoms:

- Visual (flashing lights, loss of vision)
- Sensory (paresthesia)
- Speech and/or language (difficulty speaking)
- Motor (any weakness)
- Brainstem (dizziness, double vision) and retinal (visual complaints)

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Sudden Onset of Headache (HD-11.3)

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- For sudden onset of headache including:
  - worst, most severe headache ever experienced or thunderclap-type (example: awakening from sleep)
  - sudden onset unilateral headache, suspected carotid or vertebral dissection or ipsilateral Horner's syndrome
  - consideration of reversible cerebral vasoconstriction syndrome (RCVS) (typically bilateral headache)
  - high-risk scenarios including Fibromuscular Dysplasia (FMD), Marfan Disease, MVA with whiplash, and chiropractic manipulation
- If any of these onset of headache features are present, the following are medically necessary:
  - CT Head without contrast (CPT® 70450) **OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - CTA Head (CPT® 70496) **or** MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546)
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR** CTA Neck (CPT® 70498) if carotid or vertebral artery dissection is suspected
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only **ONE** CPT® code should be used to report both procedures
- Repeat MRI Brain and/or MRA/CTA Head and Neck imaging is medically necessary in 2-4 weeks if suspicion of Reversible Cerebral Vasoconstriction Syndrome (RCVS) is high and to follow up until resolution of vasospasm, edema and/or hemorrhage, at the discretion of the neurologist, neurosurgeon, hematologist, or physiatrist, or any provider in consultation with a neurologist, neurosurgeon, hematologist, or physiatrist
- MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) is medically necessary if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, CPT® 70498) is medically necessary if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- Other vascular imaging indications for headaches require additional information.

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- See **Stroke/TIA (HD-21.1)**, **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, **Cerebral Aneurysms (HD-12.1)** and **Systemic Infections (HD-11.13)**

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Trigeminal Autonomic Cephalgias (HD-11.4)

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- For trigeminal autonomic cephalgias and cluster headache, the following are medically necessary:
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - MRI Brain without contrast (CPT® 70551)
  - May also include pituitary screening (see **Pituitary, Sella, Hypothalamus (HD-19)**)
- For facial pain (see **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10))**

Background and Supporting Information

Trigeminal autonomic cephalgias includes cluster headache, short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndromes; short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) and hemicrania paroxysmal and continua.

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Skull Base, Orbit, Periorbital or Oromaxillary (HD-11.5)

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- Skull base, orbital, periorbital or oromaxillary imaging is medically necessary for concern of skull base tumors in individuals with head and neck cancers, other skull base abnormalities seen on previous imaging, any invasive sinus infections as well as sinus tumors or orbital tumors with intracranial extension.
- In these clinical scenarios, the following studies are medically necessary:
  - MRI Brain and/or Orbits/Face/Neck without and with contrast (CPT® 70553 and/or CPT® 70543) **OR**
  - MRI Brain and/or Orbits/Face/Neck without contrast (CPT® 70551 and/or CPT® 70540) **OR**
  - CT Head and/or Orbits/Temporal bone without and with contrast (CPT® 70470 and/or CPT® 70482) **OR**
  - CT Head and/or Orbits/Temporal bone with contrast (CPT® 70460 and/or CPT® 70481)

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Suspected Intracranial Extension of Sinusitis or Mastoiditis (HD-11.6)

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- For suspected intracranial extension of sinusitis or mastoiditis, the following is medically necessary:
  - MRI Brain without and with contrast (CPT® 70553)
  - See **Mastoid Disease or Ear Pain (HD-26.1)** and **Skull Base, Orbit, Periorbital or Oromaxillary (HD-11.5)**

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New Headache Onset Older than Age 50 (HD-11.7)

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- For new onset headache in individuals older than 50 years of age, the following are medically necessary:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - CT Head without contrast (CPT® 70450)
  - If Giant Cell Arteritis, also known as Temporal Arteritis, is suspected, MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546), see **Cerebral Vasculitis (HD-22)**

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Cancer or Immunosuppression (HD-11.8)

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- For new headache in individuals with cancer, history of cancer, or those who are immunocompromised, the following are medically necessary:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
- For CT imaging in the setting of individuals with cancer, with history of cancer or who are immunocompromised, see **General Guidelines – CT Head (HD-1.4)**
- See also **Primary Central Nervous System Tumors – General Considerations (ONC-2.1)** and **Brain Metastases (ONC-31.3)**

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Abnormal Blood Clotting (HD-11.9)

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **OR** CT Head without contrast (CPT® 70450) is medically necessary for:
  - new onset headaches in individual with hypercoagulable states or bleeding disorder
    - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) is medically necessary, in addition, for venogram when requested.
      - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only **ONE** CPT® code should be used to report both procedures
  - individuals with potential for bleeding diathesis
    - Taking anticoagulants or two or more anti-aggregants or having a medical condition that predisposes to bleeding (for example, but not limited to: thrombocytopenia, liver failure, idiopathic thrombocytopenic purpura (ITP), etc.).

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Pregnancy (HD-11.10)

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- For new onset headache during pregnancy or immediate post-partum period (within 3 months after delivery), ANY or ALL of the following are medically necessary:
  - MRI Brain without contrast (gadolinium relatively contraindicated in pregnancy) (CPT® 70551)
  - CT Head without contrast (CPT® 70450) if urgent
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) when venogram is requested
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures. (gadolinium relatively contraindicated in pregnancy)
    - Vascular imaging is medically necessary concurrently with brain imaging
- Important causes of secondary headache include vascular disorders, such as pre-eclampsia, reversible cerebral vasoconstriction syndrome, and cerebral venous thrombosis, as well as idiopathic intracranial hypertension
- For post-LP/epidural anesthesia, see **Low-Pressure Headache and CSF Leak (HD-11.15)**

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Physical Exertion (HD-11.11)

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- For onset of headache with Valsalva maneuver, cough, physical exertion, change in position, **or** sexual activity, but not merely a worsening of a pre-existing headache with these activities, the following procedures are medically necessary:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - CT Head without contrast (CPT® 70450) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR**
  - CTA Head without and with contrast (CPT® 70496)
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if carotid or vertebral artery dissection or aneurysm is suspected

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Headaches Associated With Head Trauma (HD-11.12)

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- For new or progressively worsening headache with subacute head trauma, defined as within 7 days to three months post-trauma, with or without unexplained cognitive or neurologic deficits, the following are medically necessary:
  - CT Head without contrast (CPT® 70450) **OR**
  - MRI Brain without contrast (CPT® 70551)
- For persistent headaches attributed to traumatic injury to the head persisting for longer than 3 months following the injury, with or without unexplained cognitive or neurologic deficits, the following are medically necessary:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
- Acute head trauma with headache, (see **Head Trauma (HD-13.1))**
- Acute headache attributed to traumatic injury to the head that developed within 7 days of injury that does not meet criteria under **Head and Facial Trauma (HD-13)**, other subsections may apply including, but not exclusive to: **Headaches with Red Flags (HD-11.2)** and **Sudden Onset of Headache (HD-11.3)**

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Systemic Infections (HD-11.13)

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- For headaches in the setting of acute, subacute, or chronic systemic infections, the following are medically necessary:
  - MRI Brain without contrast (CPT® 70551); or MRI Brain without and with contrast (CPT® 70553)
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546)
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures
  - CT Head without contrast (CPT® 70450) or CT Head without and with contrast (CPT® 70470), when MRI Brain is contraindicated (see **General Guidelines – CT Head (HD-1.4)** for additional CT Head indications)
  - CT Head without (CPT® 70450) prior to performance of lumbar puncture (aka spinal tap)
- See **CNS and Head Infection (HD-14.1)**
- See **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)** for headache related to neuro-COVID-19 or SARS-CoV-2 vaccines

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Hydrocephalus Shunts (HD-11.14)

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Initial Imaging Indications

- MRI Brain without and with contrast (CPT® 70553) is medically necessary

Repeat Imaging Indications including CSF flow shunting and Ventriculostomy

- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is medically necessary for any of the following:
  - New signs or symptoms suggesting shunt malfunction or endoscopic third ventriculostomy (ETV) malfunction
    - Symptoms may include but are not limited to: sepsis after shunt setting adjustments, decreased level of consciousness, protracted vomiting, visual or neurologic deterioration, decline of mentation after initial improvement, or new or changing pattern of seizures
  - Requests ordered by a neurologist, neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon
- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is medically necessary in the post-operative period following shunt placement or ETV, with further follow-up imaging 6-12 months after the procedure and then every 12 months for individuals with stable clinical findings
- Shunting into the peritoneum (VP shunts) can give rise to abdominal complications, but these are generally symptomatic, so surveillance imaging of the abdomen is not medically necessary unless ordered by or in consultation with neurology, neurosurgery, or interventional radiology.
  - Abdominal ultrasound (CPT® 76700) **AND/OR** CT Abdomen or CT Abdomen/Pelvis (contrast as requested), is medically necessary depending on the clinical suspicion
    - Examples include, but are not limited to, suspicion of CSF pseudocyst formation, distal shunt outlet obstruction, or infection.
- See **General Guidelines – Other Imaging Situations (HD-1.7)**

Additional Rarely Used Studies

- Cisternogram (CPT® 78630) is medically necessary for the following:
  - Known hydrocephalus with worsening symptoms.
  - Suspected obstructive hydrocephalus.
  - Suspected normal pressure hydrocephalus with gait disturbance and either dementia or urinary incontinence.

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  - For indications related to CSF Leak, (see **Low-Pressure Headache and CSF Leak (HD-11.15)** and **Nuclear Medicine (HD-36.1)**)
- Cerebrospinal Ventriculography (CPT® 78635) is medically necessary for the following:
  - Evaluation of internal shunt, porencephalic cyst, or posterior fossa cyst.
- Nuclear Medicine Shunt Evaluation (CPT® 78645) and CSF Flow SPECT (CPT® 78803) is medically necessary for the following:
  - Suspected malfunction of ventriculoperitoneal, ventriculopleural, or ventriculovenous shunts.
- For CSF flow imaging, see **CSF Flow Imaging (HD-24.4)**
- See also **General Guidelines - CT Head (HD-1.4)**

Background and Supporting Information

- Ventriculomegaly is the condition where ventricles are enlarged, and this may be due to 1) hydrocephalus, a condition of increased intracranial pressure (ICP) (imaging shows ventricles are disproportionately enlarged compared to sulci), or 2) brain atrophy, most commonly related to age or trauma, which is not associated with increased ICP (imaging shows ventricles and sulci are proportionately enlarged).
- Hydrocephalus is divided into obstructive/non-communicating vs. communicating types, and these usually have different etiologies and radiographic features.
- Obstructive or non-communicating hydrocephalus classically involves an intraventricular obstruction in which CSF flow over the convexities and between the ventricles is reduced, and the proximal ventricle(s) is/are dilated. This is a medical emergency.
- Communicating hydrocephalus involves extraventricular obstruction, poor absorption, or overproduction of CSF. There is normal intracranial CSF flow and absence of disproportionate ventricular dilation, yet there is still a mildly increased CSF pressure. Normal pressure hydrocephalus is an example of this type.
- Distinguishing between ventriculomegaly due to brain atrophy and non-communicating hydrocephalus can be difficult with MRI Brain or CT Head alone, and modalities which visualize CSF flow may be useful such as cisternography or CT cisternography.

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Low-Pressure Headache and CSF Leak (HD-11.15)

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- Evaluation of suspected CSF leak (rhinorrhea/otorrhea) or refractory post-lumbar puncture headache, post-spinal surgery headache, orthostatic headache, or low-pressure headache:

Indication	Medically Necessary Imaging
Intracranial imaging	• MRI Brain without contrast (CPT® 70551) or • MRI Brain without and with contrast (CPT® 70553)
Spinal imaging (MRI)	• MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) or without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158)
Spinal imaging, post-myelogram	• CT Cervical Spine with contrast (CPT® 72126) **AND/OR** • CT Thoracic Spine with contrast (CPT® 72129) **AND/OR** • CT Lumbar Spine with contrast (CPT® 72132)
Spinal imaging, MR myelogram	• MRI Spine, contrast as requested • Requests following initial negative/non-diagnostic MRI Spine and MRI Brain are also medically necessary
Cisternogram, radionucleotide (111 In-DTPA)	• Radionucleotide cisternogram (CPT® 78630)

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Indication	Medically Necessary Imaging
Cisternogram, post-myelogram (iodinated contrast)	• CT Head with contrast (CPT® 70460) **OR** • CT Maxillofacial with contrast (CPT® 70487) **OR** • CT Temporal Bone with contrast (CPT® 70481)
Symptoms of CSF rhinorrhea or otorrhea	• CT Head without contrast (CPT® 70450) **AND/OR** • CT Maxillofacial without contrast (CPT® 70486) **OR** • CT Temporal Bone without contrast (CPT® 70480)

- Additional Cisternogram (CPT® 78630) indications:
  - Known hydrocephalus with worsening symptoms (for example headache)
  - Suspected obstructive hydrocephalus
- Individuals with a Shunt (see **Hydrocephalus Shunts (HD-11.14)**)

Background and Supporting Information

- Common radiological findings of CSF leaks include abnormalities of the cribriform plate or ethmoid sinus, dural dehiscence at the anterior skull base, pneumatization of the sphenoid sinus, and fluid within the middle ear.
- CSF leaks may occur in:
  - CSF shunt over-drainage
  - traumatic CSF leaks
    - Thecal holes and rents from lumbar punctures and epidural catheterizations
    - Spinal and cranial surgeries including skull base and some sinus surgeries
    - Proximal brachial plexus and nerve root avulsion injuries
  - Spontaneous leaks may occur in, but are not exclusive to:
    - Pre-existing weakness of the dural sac including:
      - disorders of connective tissue matrix including Marfan syndrome, Marfanoid features
      - joint hypermobility
    - Trivial trauma in the setting of preexisting dural weakness
    - Spondylotic spurs, herniated discs

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Cervicogenic Headaches Including Occipital Neuritis/Neuralgia (HD-11.16)

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- Brain imaging should follow applicable sections in **Headache (HD-11)**
- MRI Cervical Spine without contrast (CPT® 72141) is medically necessary for:
  - failure of 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed and clinical re-evaluation after treatment period (unless presence of a red flag) as defined in **Red Flag Indications (SP-1.2)**
    - See **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) (SP-3.1)** and **Neck (Cervical Spine) Trauma (SP-3.2)** in the Spine Imaging Guidelines
    - Exemptions to the 6 weeks of conservative care include:
      - high-risk mechanism of cervical spine injury within the last 3 months (see **Neck (Cervical Spine) Trauma (SP-3.2)** in the Spine Imaging Guidelines)
      - **Red Flag Indications (SP-1.2)** in the Spine Imaging Guidelines
      - **ANY** of the following:
        - Bony abnormalities: Atlanto-axial dislocations/instability (including but not limited to: Down's syndrome, Ehlers-Danlos and Marfan syndromes and rheumatoid arthritis), platybasia, osteomas, callous formation of the posterior C1/2 arches
        - Posterior fossa lesions, Chiari malformations, demyelinating disease
        - Myelopathy/myelitis (see **Myelopathy (SP-7.1)** in the Spine Imaging Guidelines)

Background and Supporting Information

- Cervicogenic Headache
  - Headache caused by a disorder of the cervical spine, usually accompanied by neck pain or other signs and symptoms of cervical disease. Typical findings include reduced cervical range of motion, side-locked pain, and symptoms exacerbated by provocative maneuvers such as head movement or digital pressure.
- Occipital Neuralgia/Neuritis - Occipital neuralgia is classified unilateral or bilateral paroxysmal, shooting or stabbing pain in the posterior part of the scalp, in the distribution(s) of the greater, lesser and/or third occipital nerves, sometimes accompanied by diminished sensation or dysaesthesia in the affected area and commonly associated with tenderness over the involved nerve(s).

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- Pain has at least two of the following three characteristics:
  - Recurring in paroxysmal attacks lasting from a few seconds to minutes
  - Severe in intensity
  - Shooting, stabbing or sharp in quality
- Pain is associated with both of the following:
  - Dysaesthesia and/or allodynia apparent during innocuous stimulation of the scalp and/or hair
  - Either or both of the following:
    - Tenderness over the affected nerve branches
    - Trigger points at the emergence of the greater occipital nerve or in the distribution of C2
- Pain is eased temporarily by local anesthetic block of the affected nerve(s)

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Advanced Imaging Indications Related To Migraines (HD-11.17)

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- Advanced imaging of the head is **NOT** medically necessary for newly diagnosed migraine with a normal neurological exam or chronic stable migraine with no neurological deficit and/or no red flags (see **Headaches with Red Flags (HD-11.2)**).
  - See below for advanced imaging indications related to migraines.
- New migraine with age ≥50 (see **New Headache Onset Older than Age 50 (HD-11.7)**)
- Change in frequency or severity of migraine (see **Headaches with Red Flags (HD-11.2)**)
- MRI Brain without (CPT® 70551) or MRI Brain without and with (CPT® 70553) or CT Head without (CPT® 70450) for the following:
  - Unusual, prolonged, or persistent aura (greater than 60 minutes) (See *Background and Supporting Information*)
  - Worst migraine
  - Hemiplegic migraine
    - Migraine with any motor weakness.
  - Migrainous accompaniments
    - Passing neurological symptoms that can affect vision, speech, movement, and behavior-“mimic stroke”
  - Migraine aura without headache
    - Migraine with an aura in which the aura is neither accompanied nor followed by a headache within 60 minutes.
  - Side-locked migraine (unilateral)
    - Unilateral hemicranial pain – includes primary and secondary causes.
      - New daily persistent headache (new daily headache present greater than three months)
      - Trigeminal autonomic cephalgias includes cluster headache short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndromes; short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) and hemicrania paroxysmal and continua are covered in **Trigeminal Autonomic Cephalgias (HD-11.4)**
  - Post-traumatic migraine
    - See **Head Trauma (HD-13.1)** and **Headaches Associated with Head Trauma (HD-11.12)**

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Background and Supporting Information

- Aura symptoms may accompany or precede a headache within 60 minutes and may include, but are not exclusive to, the following symptoms:
  - Visual (flashing lights, loss of vision)
  - Sensory (paresthesia)
  - Speech and/or language (difficulty speaking)
  - Motor (any weakness)
  - Brainstem (double vision) and retinal (visual complaints)

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Evidence Discussion (HD-11)

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- The majority of headaches are due to benign causes and are self-limited. The yield of positive findings on a CT Head for evaluation of headache in the emergency department setting ranged from 7% to 13%.[1]
- The American College of Radiology in the Choosing Wisely campaign recommended against imaging for primary headache syndromes in the absence of red flags and with a normal neurologic exam.[1,32] The American Headache Society and Choosing Wisely Task Force stated that the overuse or misuse of imaging studies for headache was the most commonly mentioned problem.[32] Overuse of CT Head was identified as the main concern.[32] The authors note that a single CT scan of the head exposes individuals to an average of 2 millisievert (mSV) of radiation, the equivalent of 8 months of background radiation.[32]
- Incidental findings are common and can result in anxiety for the individual, additional referrals and specialist consults, and more imaging studies.[30,31] Incidental findings on MRI occur in 2% of the general population.[31]
- The American Headache Society and the American Academy of Neurology recommended neuroimaging in individuals with headaches with atypical features, red flags, and/or abnormal neurologic exam findings.[1,13,29]
- The presence of neurologic or systemic signs, new headaches over age 50, or headaches in the setting of malignancy or immunosuppression, always require further evaluation with advanced imaging, and are considered "red flags," due to the higher likelihood of intracranial pathology.[1,30,36,37] CT Head in the presence of red flags is helpful to exclude intracranial hemorrhage.[1] However, MRI Brain has higher contrast resolution than CT Head and is preferred for evaluation of structural pathologies, particularly in non-urgent settings.[1]
- Subarachnoid hemorrhage due to ruptured cerebral aneurysm accounts for 4%-12% of acute severe headaches.[1] CT Head is medically necessary as initial imaging for thunderclap headache. CT Head had a negative predictive value between 99.9%-100% in detecting aneurysmal subarachnoid hemorrhage within 6 hours of headache onset. The sensitivity is over 90% when CT Head is performed within the first 24 hours. CT Angiography (CTA) Head obtained concurrently or in follow up may identify cerebral aneurysm, dissection, and reversible cerebral vasoconstriction syndrome.[1]
- In selected cases, CT Head is supported for evaluation and follow up of headache caused by subdural or epidural hemorrhage, skull fracture, sinus infection or subarachnoid hemorrhage.[30]
- New headaches in the setting of pregnancy and the postpartum period require special consideration.[1,30,35]

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- Over a third of pregnant women presenting to the hospital with headache have a secondary cause.[1,34,35] Of individuals with headache in the immediate post-partum period, 41% had an abnormal MRI Brain.[35]
- Imaging in this scenario includes MRI Brain, MR Venogram (MRV) Head, and/or MR Angiography (MRA) Head. Gadolinium contrast is relatively contraindicated during pregnancy and should be avoided.[1]
- Trigeminal autonomic cephalgias, including cluster headaches, are required to have MRI brain to exclude pathology in the pituitary region.[1,30] MRI should include the brain and the pituitary region.[1,13]
- Headaches concerning for raised intracranial pressure or intracranial hypotension, required additional evaluation with neuroimaging.[1,30] To exclude hydrocephalus, a mass, or cerebral venous sinus thrombosis, MRI Brain, Orbits, and venogram are medically necessary in the setting of papilledema and/or intracranial hypertension.[1,30] In urgent cases, a CT Head can rapidly diagnose causes such as mass, edema, or hydrocephalus.[1]
- MRI is also useful to evaluate for structural causes of headache due to intracranial hypotension and cerebrospinal (CSF) leaks.[1,30] Depending on the suspected source of the leak, imaging the brain and spinal cord may be required. Spinal imaging may include MRI of the spinal cord, or CT myelogram.[1]
- In the evaluation of headaches from suspected intracranial hypotension with negative initial brain and spine imaging, and high clinical suspicion of a CSF leak, MR myelography is indicated to detect subtle spinal leak sources, such as CSF-venous fistulas and slow meningeal diverticular leaks.[41]
- CTA Head or MRA Head in the evaluation of headache are medically necessary for suspicion of carotid or vertebral arterial dissections, AVMs and cerebral aneurysm, as secondary causes of headache.[1]
- CT Venogram (CTV) head or MRV head in the evaluation of headache are supported for suspicion of cerebral venous sinus thrombosis or stenosis in select cases, included suspected headache associated with pregnancy and the post-partum period, headache with papilledema, intracranial hypertension, and the trigeminal autonomic cephalgias.[1]
- Computerized tomography (CT) is the gold standard for the evaluation of abdominal complications related to ventriculoperitoneal shunts (VPS).[38]

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Cerebral Aneurysm and AVM (HD-12)

Guideline

Cerebral Aneurysms (HD-12.1)
Arteriovenous Malformations (AVMs) and Related Lesions (HD-12.2)
Evidence Discussion (HD-12)
References (HD-12)

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Cerebral Aneurysms (HD-12.1)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Coarctation of the aorta	See **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)**
Screening for high-risk populations as defined by the following criteria (screening usually begins at age 20 unless unusual circumstances as aneurysms are uncommon in children and adolescents): • Positive Family History: Two or more first degree relatives (parent, sibling, or child) with history of cerebral aneurysm or SAH: screening every 5 years beginning at age 20. One first degree relative (parent, sibling, or child) with history of cerebral aneurysm or SAH can have one screening study • Autosomal dominant polycystic kidney disease (screening begins at age 20 to 65 and is repeated at five-year intervals) • Alpha-1-antitrypsin deficiency • Alpha-glucosidase deficiency • Azygos anterior cerebral artery (presence of) • Bicuspid aortic valve • Fibromuscular dysplasia (one screening study after confirmed diagnosis) • Ehlers-Danlos Syndrome Type 4 (Vascular) • Glucocorticoid-remediable aldosteronism (GRA) • Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu Syndrome) See **Screening for Vascular related genetic connective tissue Disorders (PVD-2.2)**	• CTA Head (CPT® 70496) **OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Screening for high-risk populations as defined by the following criteria (screening usually begins at age 20 unless unusual circumstances as aneurysms are uncommon in children and adolescents) - ***CONTINUED***: • Kawasaki disease • Klinefelter syndrome • Klippel-Trenaunay-Weber Syndrome • Loeys-Dietz Syndrome • Marfan Syndrome • Microcephalic osteodysplastic primordial dwarfism • Neurofibromatosis Type 1 • Noonan Syndrome • Pheochromocytoma • Pseudoxanthoma elasticum • Tuberous sclerosis See **Screening for Vascular related genetic connective tissue Disorders (PVD-2.2)**	• CTA Head (CPT® 70496) **OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546)
New or worsening clinical symptoms or signs of cerebral aneurysm, including: • Thunderclap headache (see **Sudden Onset of Headache (HD-11.3)**) • Third nerve palsy with pupillary involvement (pupil-sparing third nerve palsies are not caused by external compression) • Suspicion of aneurysm bleed [CT head or MRI brain or CSF exam showing evidence of subarachnoid hemorrhage (SAH) or intracerebral hemorrhage] • Abnormal CT Head or MRI Brain suggesting possible aneurysm	• CTA Head (CPT® 70496) **OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
Questionable or equivocal findings on an initial MRA Head	CTA Head (CPT® 70496)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
For suspected or confirmed cerebral aneurysm, ruptured or unruptured, for initial evaluation, treatment, intervention, or follow-up OR If initial catheter angiography is negative, repeat imaging is medically necessary.	3D Rendering (CPT® 76377 or CPT® 76376) with cervicocerebral angiography/arteriography and/ or cerebral angiography (See **General Guidelines - Other Imaging Situations (HD-1.7)**)
Follow-up of known cerebral aneurysm: The optimal interval and duration for radiologic follow-up has not been determined. Radiographic follow-up for unruptured or treated intracranial aneurysms upon request by the neurosurgeon or team managing the intracranial aneurysm.	• CTA Head (CPT® 70496) **OR** • MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) **AND/OR** • MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
Additional physical characteristics of a known aneurysm: • To evaluate and treat a giant aneurysm (>2.5 cm) • Posterior fossa aneurysms • Thrombosed or partially thrombosed aneurysms • To evaluate the relationship of the aneurysm to the dura • To evaluate for the presence of calcification • Other surveillance criteria as per the neurosurgeon or team managing the aneurysm repair	• MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
Follow up of cerebral aneurysm located in the vertebro-basilar circulation OR If intracranial etiology of SAH has not been found	• MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR** • CTA Neck (CPT® 70498)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Subacute complications (i.e., vasospasm, delayed cerebral ischemia, and hydrocephalus), beginning days to weeks, arising from a subarachnoid hemorrhage and/or aneurysm treatment, upon request from the neurosurgeon and/or team managing the episode	CT Head **OR** MRI Brain contrast as requested
To evaluate individuals with SAH and negative studies for brain aneurysm in whom spinal abnormalities (i.e., AVM) may be suspected as the cause of hemorrhage	MRI Spine (Cervical without and with contrast CPT® 72156, **AND/OR** Thoracic without and with contrast CPT® 72157, **AND/OR** Lumbar without and with contrast CPT® 72158)
Catheter angiogram negative in SAH individual with remaining suspicion for cerebral aneurysm and these studies have not yet been performed:	• CTA Head (CPT® 70496) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546)

- High-risk scenarios for vascular dissection include, but are not limited to: Fibromuscular dysplasia (FMD), Marfan Disease, MVA with whiplash, and chiropractic manipulation
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- Other vascular imaging indications for headaches require additional information. See **Stroke/TIA (HD-21.1)**,**Sudden Onset of Headache (HD-11.3)**, **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, and **Systematic Infections (HD-11.13)**

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Arteriovenous Malformations (AVMs) and Related Lesions (HD-12.2)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Any aneurysmal and/or AVM disorders listed in this guideline • When MRI contraindicated • Any urgent setting	• CT Head without contrast (CPT® 70450) **AND/OR** • CTA Head (CPT® 70496) **AND/OR** • CTA Neck (CPT® 70498)
Known AVM • When requested by a neurologist, neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRA Head (CPT® 70544, CPT®70545, CPT® 70546) **OR** • CTA Head (CPT® 70496)
Known AVM in the vertebral-basilar system • When requested by a neurologist, neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon	• Imaging as listed above in “known AVM” **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, OR CPT® 70549) **OR** • CTA Neck (CPT® 70498)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Subarachnoid Hemorrhage (SAH) • AVM is suspected based on a history of SAH	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
Hereditary Hemorrhagic Telangiectasia (HHT; Osler-Weber-Rendu Syndrome) • Suspected based on family history with at least one affected first-degree relative (biological parent or sibling) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed HHT • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon, or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Capillary Malformation-Arteriovenous Malformation (CM-AVM) • Suspected based on family history with at least one affected first-degree relative (biological parent or sibling) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed CM-AVM • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon, or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Cerebral Cavernous Malformations (CCM) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed CCM • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon, or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

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Disorders and Indications (Any of the following)	Medically Necessary Imaging
Microcephalic Osteodysplastic Primordial Dwarfism, Type II (MOPD II) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed MOPD II, repeated annually • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon, or geneticist	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, OR CPT® 70549) **OR** • CTA Neck (CPT® 70498)
Sturge-Weber Syndrome • At diagnosis • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** • MRI Orbits/Face/Neck without contrast (CPT® 70540)

- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551), **OR** CT Head without contrast (CPT® 70450) **AND/OR** MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) is medically necessary for symptoms including headache, seizure, and/or focal neurologic deficits

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- For concerns related to stroke, see **Stroke/TIA HD-21.1**
- 3D imaging (CPT® 76376 or CPT® 76377) with MRI Brain without and with contrast (CPT® 70553) OR MRI Brain without contrast (CPT® 70551) is medically necessary
- 3D Rendering (CPT® 76377 or CPT® 76376) with cervicocerebral catheter angiography is medically necessary to define the presence, location, and anatomy of intracranial and cervical vascular malformations at diagnosis and for follow up, including post-treatment, (see **General Guidelines - Other Imaging Situations (HD-1.7)** and **Background and Supporting Information**).
- See **General Guidelines – CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5)**
- Functional MRI (CPT® 70554 OR CPT® 70555) for surgical planning, see **Functional MRI (fMRI) (HD-24.2)**

Background and Supporting Information

- Trauma is the most common reason for subarachnoid hemorrhage. Ruptured berry aneurysm is the most common reason for non-traumatic subarachnoid hemorrhage in adults.
- Small aneurysms are present in about 1% to 2% of adults, but very few ever reach a size for which bleeding is a risk (>5 mm). Small (<3 mm to 4 mm) unruptured aneurysms in those with no personal history of SAH have a 0.1% to 0.5% a year rate of bleeding. The risk of cerebral aneurysm with family history ranges from 2% with one first degree relative to 30% to 35% for identical twin or two parents. The risks and benefits of screening these populations need to be considered before advanced imaging.
- AVMs most often come to clinical notice either by bleeding or by acting as a seizure focus. They are usually congenital, recognized later in life and have an initial risk of bleeding of 2% per year.
- Cerebral angiography is a form of angiography which provides images of blood vessels in and around the brain and/or neck. This is a catheter- based procedure, using x-ray imaging guidance and iodine-based contrast to visualize blood vessels.
- Most intracranial AVMs are congenital, vary widely in their location and type, and are discovered at birth due to associated clinical findings or incidentally later in life. Certain hereditary conditions are associated with an increased risk for AVM development.
- Vascular malformations include arteriovenous, venous, cavernous, and capillary malformations.
- Hereditary AVMs usually have an autosomal dominant pattern of inheritance.

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Evidence Discussion (HD-12)

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- After the initial identification of a subarachnoid hemorrhage, the search for a ruptured cerebral aneurysm begins with imaging of the cerebral vessels with CT Angiography (CTA), MR Angiography (MRA) or diagnostic cervicocerebral catheter angiography.[22]
- CTA Head has over 90% sensitivity and specificity for the diagnosis of cerebral aneurysm in the setting of subarachnoid hemorrhage.[22]
- MRA Head has a sensitivity of 95% and a specificity of 89% for diagnosis of cerebral aneurysm.[22]
- Diagnostic cervicocerebral catheter angiography has the highest spatial and temporal resolution of any vascular imaging study, however, is invasive and requires use of contrast. It has a sensitivity and specificity over 98% for identification of aneurysm and can also diagnose vascular abnormalities in up to 13% with subarachnoid hemorrhage and a negative CTA. In addition, this modality can identify an aneurysm in 25% of previously negative studies and repeat studies are supported for this reason.[22]
- In select cases of subarachnoid hemorrhage when an intracranial aneurysm is not identified, imaging the neck vessels and spinal vessels is appropriate.[12]
- Aneurysm growth ranges from 4% to 14% on follow up imaging. For surveillance of incidentally identified cerebral aneurysms or ruptured and/or treated aneurysms, the less invasive modalities, CTA and/or MRA are supported, over the more invasive diagnostic cervicocerebral angiography.[22]
- Screening for cerebral aneurysms in high-risk individual populations is also recommended with the less invasive modalities, CTA head or MRA head. This includes individuals with autosomal dominant polycystic kidney disease (ADPKD), who had a prevalence of cerebral aneurysm ranging from 10%-11.5%. The American Heart Association recommended screening those with at least 2 family members with cerebral aneurysm or subarachnoid hemorrhage.[22]
- Screening is also recommended for conditions with known increased risk of cerebral aneurysm.[4,22]
- Although vascular imaging is the primary focus of neuroimaging in the diagnosis and follow up of cerebral aneurysms, parenchymal imaging with MRI Brain may be helpful in select clinical scenarios including giant aneurysms, posterior fossa aneurysms, in the setting of cranial neuropathies or focal neurologic findings and suspected stroke.[12]
- Cervicocerebral angiography is the gold standard for imaging arteriovenous malformations (AVM) and arteriovenous fistulas (AVF). CT Angiography head (CTA) has a 90% sensitivity for the overall detection of AVMs and 100% for AVMs >3 cm in size. In the evaluation of pulsatile tinnitus, CTA has a sensitivity of 86% with a

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specificity of 100% in identifying high flow AVFs. MR Angiography (MRA) is an alternative modality in these scenarios.[22]

- MRI Brain for diagnosis and follow up of AVM has an overall sensitivity of 89% and 100% for lesions >3 cm in size.[22]

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Head and Facial Trauma (HD-13)

Guideline

Head Trauma (HD-13.1)
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Head Trauma (HD-13.1)

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For acute head trauma (0 to 7 days post-trauma)

- CT Head without contrast (CPT® 70450) is medically necessary in individuals with **ANY** of the following modified Canadian CT Head Rule/New Orleans Criteria.
  - Regardless of documented or stated head impact, ANY "dangerous mechanism of injury", either direct or indirect, including, but not exclusive to:
    - Fall from height greater than 3 feet
    - Fall greater than 5 steps down stairs
    - Any pedestrian motor vehicle accident
    - High impact motor vehicle accident
  - Individual >60 years old
  - Loss of consciousness, amnesia, or disorientation accompanying blunt head trauma within 24 hours
  - Taking one anticoagulant or two antiaggregants, (e.g., aspirin and Plavix)
  - Known platelet or clotting disorder
  - Glasgow coma scale (GCS) score of less than 15 at 2 hours following injury
  - >30 minutes of amnesia before impact
  - Suspected open skull fracture
  - Signs of basilar skull fracture (Battle's sign, Raccoon eyes, CSF rhinorrhea, cranial nerve palsy, hemotympanum, acute hearing loss)
  - Vomiting
  - Alcohol or drug intoxication
  - Visible trauma above clavicles
  - Deficits in short term memory, altered level of alertness, abnormal behavior, or focal neurological deficit
  - Seizure
  - Headache, see **Headache Associated with Head Trauma (HD-11.12))**

For subacute head trauma (7 days to 3 months post-trauma) and chronic head trauma (greater than 3 months post-trauma) symptoms

- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is medically necessary for the initial imaging of individuals with subacute or chronic head trauma and unexplained cognitive or neurologic deficits.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary if post-traumatic infection is suspected

Repeat and follow-up imaging

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- Follow-up imaging is medically necessary for known subdural hematomas, intracerebral hemorrhage, or contusions can be done at the discretion of the ordering provider with one of the following:
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - MRI Brain without contrast (CPT® 70551) **OR**
  - CT Head without and with contrast (CPT® 70470) **OR**
  - CT Head without contrast (CPT® 70450)
- For short term follow-up imaging of acute traumatic brain injury (TBI) without neurologic deterioration, CT Head without contrast (CPT® 70450) is medically necessary in individuals with ANY of the following risk factors:
  - subfrontal/temporal intraparenchymal contusions
  - anticoagulation
  - age >65 years
  - intracranial hemorrhage
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary as a complementary study when neurological findings or symptoms are not sufficiently explained by CT or in subacute and chronic TBI for new, persistent, or slowly progressive symptoms.

For suspected intracranial venous or arterial injury, the following is medically necessary:

- CTA/CTV Head (CPT® 70496) or MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546)
  - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures (see **General Guidelines - CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5))**

SPECT, PET, CT/MRI perfusion, DTI (diffusion tensor imaging), functional MRI, and MR spectroscopy are considered not medically necessary for the evaluation of traumatic brain injury.

Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain

- Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be requested as (CPT® 0865T or CPT® 0866T)
  - These studies are considered not medically necessary in the evaluation of traumatic brain injury.

See **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3.2)** in the Spine Imaging Guidelines

See **General Guidelines – CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5)** for traumatic vascular injuries

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Background and Supporting Information

Individuals with head trauma are at risk for facial and cervical trauma.

Recent studies have shown that diffusion tensor MRI tractography may be more sensitive in demonstrating abnormalities such as axonal injury in closed head injury than conventional MRI, but these techniques are best described presently as research tools and their use in clinical practice is not determined.

Decisions regarding return to normal activities, including sports, are made based on the clinical status of the individual and repeat imaging is unnecessary.

In cases of post-traumatic infection, contrast-enhanced MRI or CT may be helpful.

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Facial Trauma (HD-13.2)

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- CT Maxillofacial without contrast (CPT® 70486) and/or CT Head without contrast (CPT® 70450) is/are medically necessary for any concern regarding significant injury to facial structures including, but not limited to:
  - Concern for orbital, maxillary, or mandibular fractures
  - Trauma with associated symptoms of anosmia, hearing, vision or speech changes, vertigo, facial numbness
  - Physical exam findings of CSF rhinorrhea (suspected post-traumatic CSF leak), malocclusion, severe focal facial tenderness, focal loss of facial sensation
- CT Orbits/Temporal Bone without contrast (CPT® 70480) and/or CT Head without contrast (CPT® 70450) is/are medically necessary for:
  - concern for orbital injury or orbital wall fracture
  - symptoms of diplopia, blurred vision, vision loss
  - physical exam findings of enophthalmos, entrapment of extraocular muscle(s)
  - suspicion for temporal bone fracture
  - physical exam findings of CSF otorrhea (suspected post-traumatic CSF leak)
- MRI Maxillofacial without contrast (CPT® 70540) **OR** MRI Maxillofacial without and with contrast (CPT® 70543) is medically necessary for evaluation of cranial nerve deficits not explained or incompletely characterized on CT.
- If concern for CSF leak and CT Maxillofacial or Temporal bone is inconclusive, (see **Low-Pressure Headache and CSF Leak (HD-11.15)**)

Background and Supporting Information

Imaging is not necessary in the evaluation of simple nasal fractures if tenderness and swelling is limited to the nasal bridge, the individual can breathe through each naris, and there is no septal hematoma.

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Evidence Discussion (HD-13)

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- For evaluation of acute mild head trauma, less than 10% will have positive findings on CT head, and of this group, less than 1% will require neurosurgical intervention.[7,18]
- The American College of Emergency Physicians and the Choosing Wisely Initiative recommend avoiding CT scans of the head in emergency department individuals with minor head injury who are at low risk based on validated decision rules. This recommendation was based on the concern for individual exposure to ionizing radiation and the increased lifetime risk of cancer with such exposure.[17]
- Selective CT scanning is recommended by validated clinical practice guidelines, including the New Orleans Criteria and the Canadian CT Head Rule.[1,7,18] Both guidelines are 100% sensitive for mild head trauma requiring neurosurgical intervention.[7,18] The New Orleans Criteria has a sensitivity >97% for any traumatic finding on CT, with a specificity less than 6%.[7,18] The Canadian CT Head Rule has a sensitivity between 83.4% - 87.2% with a specificity between 37.2% – 39.7%.[1,7,18] The Canadian CT Head Rule has a 100% sensitivity and 29% specificity in cases of intracranial hemorrhage.[1,7,18]
- When imaging is indicating by a validated clinical decision rule, CT head is the preferred imaging modality for evaluation of acute head trauma.[7,15,17,18]
- If the initial CT head confirms subdural hematoma, follow-up CT head is supported to monitor progression.[7,8]
- For follow-up in individuals with persistent neurologic deficits without etiology identified on initial CT head, MRI brain is more sensitive, and can visualize cortical contusions, subdural hematomas, and white matter lesions in diffuse axonal injury.[7] Up to 27% of individuals with mild traumatic brain injury (TBI) with normal initial CT head show abnormalities on MRI Brain.[7,8]
- When vascular injury is suspected in the setting of head trauma, CT Angiography (CTA) head and neck is a non-invasive, rapid, and useful modality to evaluate for arterial injury.7,1 MR Angiography is an alternative option. For suspected intracranial venous injury, CT Venography (CTV) is medically necessary, with MR Venography (MRV) an alternative option.[7,16]
- There is insufficient evidence to support the use of single-photon emission computed tomography (SPECT), FDG-PET/CT brain, CT/MRI-Perfusion, MR spectroscopy (MRS), functional MRI (fMRI), or diffusion tensor imaging (DTI) in the evaluation of head trauma.[7]
- Individuals with head trauma are also at risk for orbital, facial and temporal bone injuries.[11] CT of the orbit can diagnose fractures, displaced fracture fragments, foreign bodies, traumatic hematoma, and extraocular muscle injury.[11] CT head is also recommended in the evaluation of suspected orbital fractures due to concomitant intracranial injury incidence of 9%.[16]

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- CT maxillofacial is useful in diagnosing maxillofacial injuries including non-displaced fractures. CT provides multiplanar and 3-D image reconstructions, allowing for better characterization of complex fractures, which is useful for surgical planning.[16]
- Over one-third of individuals with frontal sinus fractures are likely to have a concomitant intracranial injury, thus concurrent CT head imaging is recommended in individuals with suspected frontal sinus fractures. In addition, between 8% to 10% of individuals with frontal sinus fractures have subdural or epidural hematomas requiring surgical treatment.[16]
- High resolution CT (HRCT) facial and temporal bone are sensitive modalities for subtle or non-displaced skull base defects, with sensitivity of 92% for identifying cerebrospinal fluid leak.[7,15]

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References (HD-13)

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CNS and Head Infection/ Neuro-COVID-19 (HD-14)

Guideline

CNS and Head Infection (HD-14.1)
Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)
Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)
References (HD-14)

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CNS and Head Infection (HD-14.1)

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INITIAL IMAGING

- Signs of intracranial infection include, but are not limited to
  - headaches, seizures, meningeal signs (neck stiffness)
  - new focal neurological deficits in a setting of fever or elevated white blood cell count (WBC)
  - known infection elsewhere or
  - immunosuppression
- **ONE** of the following studies is medically necessary for suspected intracranial infection if any of these signs of infection are present:
  - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **OR**
  - CT Head (CPT® 70450, CPT® 70460, or CPT® 70470) in cases where MRI is contraindicated, in urgent scenarios, or prior to lumbar puncture, see **General Guidelines-CT Head (HD-1.4)**
  - If vascular involvement is suspected, in addition to MRI Brain, the following are medically necessary:
    - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR**
    - CTA Head (CPT® 70496) **AND/OR**
    - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
    - CTA Neck (CPT® 70498)
    - (CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography)
  - Concern for vasculitis, see **Cerebral Vasculitis (HD-22)**

REPEAT IMAGING

- As requested by an infectious disease specialist, neurologist, neurosurgeon, radiologist, or any provider coordinating care with an infectious disease specialist, neurologist, neurosurgeon, or radiologist
- Repeat imaging would refer to any of the CPT codes listed above as initial imaging.
  - See **General Guidelines – CT Head (HD-1.4)** regarding additional indications for CT Head.
  - See **Skull Base Osteomyelitis (SBO) (HD-20.1)**, **Sinus and Facial Imaging (HD-29.1)**, **Dental/Periodontal/Maxillofacial Imaging (HD-30.2)**, **Mental Status Change (HD-4.2)**, and **Eye Disorders and Visual Loss (HD-32.1)**

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Evidence Discussion (HD-14.1)

- A head CT is recommended for quickly assessing intracranial infections in urgent cases. However, MRI is more effective for examining conditions that affect the cranial nerves, brain tissue, and meninges.[15] Although MRI is superior in detecting minor changes in the brain associated with infections, CT scans can promptly identify pathophysiological changes that may influence the individual's prognosis.[1]
- Acute bacterial meningitis often presents with a normal CT scan. However, due to the high mortality rate of up to 50% when left untreated, a CT scan is necessary to rule out other causes of encephalopathy or neurologic deficits. In certain clinical situations, CT is also required to exclude increased intracranial pressure before performing a lumbar puncture. Venous thromboses increase the risk of hemorrhage and are linked to high mortality. Consequently, both CT and MRI scans, including T1-weighted sequences, are recommended. These scans help detect high signal changes in several venous sinuses and can be complemented with CT- or MR-venography.
- Similar considerations apply when diagnosing non-bacterial central nervous system (CNS) infections. CT scans are advantageous due to quick access to care, faster diagnosis, and earlier treatment initiation, which can reduce morbidity. However, MRI is superior in detecting patterns of vasogenic versus cytotoxic edema, contrast enhancement, and the distribution of involvement, whether multifocal or unifocal/unihemispheric. These distinctions are crucial in differentiating between differential diagnoses, such as systemic infections with hematogenous spread versus head/neck infections with a direct spread pattern.[39]
- Many individuals present with neurologic signs and symptoms that are indicative of either a cortical or subcortical syndrome. Often, these cannot be fully characterized by clinical presentation alone. As a result, radiologic evaluation becomes essential in diagnosing the etiology of the underlying process. The potential causes are varied and include meningoencephalitis, acute cerebrovascular disease, hemorrhagic necrotizing encephalopathy, immune-mediated (Bickerstaff) encephalitis, and demyelinating diseases such as acute disseminated encephalomyelitis (ADEM) and multiple sclerosis (MS).[40] Therefore, prompt and accurate diagnosis is critical to select the most appropriate imaging method (MRI vs. CT) for each clinical scenario.

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Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)

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- The following studies are medically necessary for evaluation of:
  - Acute or chronic Neuro-COVID-19 syndrome
    - MRI Brain without contrast (CPT® 70551) **OR**
    - MRI Brain without and with contrast (CPT® 70553) **OR**
    - CT Head without contrast (CPT® 70450) **OR**
    - CT Head without and with contrast (CPT® 70470) is medically necessary if there is a contraindication to MRI **AND/OR**
    - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) OR CTA Head (CPT® 70496) **AND/OR**
    - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498)
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography):
      - If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code is used to report both procedures
      - If an arterial CTA or MRA study has been performed and subsequently a repeat study is needed to evaluate the venous anatomy, then this study is medically necessary
      - If a venous CTV or MRV has been performed and subsequently a repeat study is needed to evaluate the arterial anatomy, then this study is medically necessary
      - MRA without and with contrast with venous sinus thrombosis to differentiate total from subtotal occlusion is medically necessary
  - For suspected neurologic adverse reactions after SARS- CoV-2 vaccination, the following is medically necessary:
    - MRI Brain without contrast (CPT® 70551) **OR**
    - MRI Brain without and with contrast (CPT® 70553) **OR**
    - CT Head without contrast (CPT® 70450) **OR**
    - CT Head without and with contrast (CPT® 70470) is medically necessary if there is a contraindication to MRI **AND/OR**
    - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) OR CTA Head (CPT® 70496) **AND/OR**

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        - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498)
        - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography):
            - If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code is used to report both procedures
            - If an arterial CTA or MRA study has been performed and subsequently a repeat study is needed to evaluate the venous anatomy, then this study is medically necessary
            - If a venous CTV or MRV has been performed and subsequently a repeat study is needed to evaluate the arterial anatomy, then this study is medically necessary
            - MRA without and with contrast with venous sinus thrombosis to differentiate total from subtotal occlusion is medically necessary
    - If suspected transverse myelitis, acute necrotizing myelitis, and/or COVID infection, then ANY the following are medically necessary:
        - MRI Cervical without and with contrast (CPT® 72156)
        - MRI Thoracic without and with contrast (CPT® 72157)
        - MRI Lumbar Spine without and with contrast (CPT® 72158)
        - See **Stroke/TIA (HD-21.1)** for vascular imaging
        - See **Transverse Myelitis (HD-16.4)** regarding spine imaging to evaluate for post-vaccination neurological syndrome
- Repeat imaging considered on a case-by-case basis for a change in neurological symptoms or signs on the neurological exam and/or change in the treatment.

Background and Supporting Information

- The findings observed in the central nervous system in the acute-phase of COVID- 19 may extend into a prolonged symptomatic phase of Neuro-COVID in long haulers with chronic COVID syndrome. Symptoms may include, but are not inclusive to: "brain fog", dizziness, inability to concentrate, psychiatric symptoms, and confusion.
- Acute-phase neurologic manifestations of COVID-19 include: headache, dizziness, taste and smell dysfunction, impaired consciousness (described as confusion or agitation), cerebrovascular events (ischemic stroke, cerebral venous sinus thrombosis, cerebral hemorrhage), seizures, meningoencephalitis, and immune-mediated neurologic diseases (Guillan-Barre syndrome, Miller-Fisher syndrome, polyneuritis cranialis, transverse myelitis).
- Neurologic adverse reactions in those receiving SARS-CoV-2 vaccines, including mRNA vaccines (Pfizer, Moderna), have been reported, and include, although not limited to: headache, Guillan-Barre syndrome, transverse myelitis, facial nerve palsy, small fiber neuropathy, autoimmune encephalitis, reversible cerebral vasoconstriction syndrome, multiple sclerosis, neuromyelitis optica, intracerebral bleeding, cerebral

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venous sinus thrombosis, hypophysitis, epilepsy, encephalopathy, and acute disseminated encephalomyelitis.
- Cases of Thrombosis with Thrombocytopenia Syndrome (TTS) following administration of the Johnson & Johnson/Janssen COVID-19 Vaccine have been reported in males and females, in a wide age range of individuals 18 years and older, with the highest reporting rate (approximately 8 cases per 1,000,000 doses administered) in females ages 30-49 years; overall, approximately 15% of TTS cases have been fatal. Currently available evidence supports a causal relationship between TTS and the Johnson & Johnson/Janssen COVID-19 Vaccine. The clinical course of these events shares features with autoimmune heparin-induced thrombocytopenia. In individuals with suspected TTS following administration of the Johnson & Johnson/Janssen COVID-19 Vaccine, the use of heparin may be harmful and alternative treatments may be needed. Consultation with hematology specialists is strongly recommended. The American Society of Hematology has published considerations relevant to the diagnosis and treatment of TTS following administration of the Janssen COVID-19 Vaccine.

Evidence Discussion (HD-14.2)
- A head CT is recommended for quickly assessing intracranial infections in urgent cases. However, MRI is more effective for examining conditions that affect the cranial nerves, brain tissue, and meninges.[15] Although MRI is superior in detecting minor changes in the brain associated with infections, CT scans can promptly identify pathophysiological changes that may influence the individual's prognosis.[1]
- Acute bacterial meningitis often presents with a normal CT scan. However, due to the high mortality rate of up to 50% when left untreated, a CT scan is necessary to rule out other causes of encephalopathy or neurologic deficits. In certain clinical situations, CT is also required to exclude increased intracranial pressure before performing a lumbar puncture. Venous thromboses increase the risk of hemorrhage and are linked to high mortality. Consequently, both CT and MRI scans, including T1-weighted sequences, are recommended. These scans help detect high signal changes in several venous sinuses and can be complemented with CT- or MR-venography.
- Similar considerations apply when diagnosing non-bacterial central nervous system (CNS) infections. CT scans are advantageous due to quick access to care, faster diagnosis, and earlier treatment initiation, which can reduce morbidity. However, MRI is superior in detecting patterns of vasogenic versus cytotoxic edema, contrast enhancement, and the distribution of involvement, whether multifocal or unifocal/unihemispheric. These distinctions are crucial in differentiating between differential diagnoses, such as systemic infections with hematogenous spread versus head/neck infections with a direct spread pattern.[39]
- Many individuals present with neurologic signs and symptoms that are indicative of either a cortical or subcortical syndrome. Often, these cannot be fully characterized

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by clinical presentation alone. As a result, radiologic evaluation becomes essential in diagnosing the etiology of the underlying process. The potential causes are varied and include meningoencephalitis, acute cerebrovascular disease, hemorrhagic necrotizing encephalopathy, immune-mediated (Bickerstaff) encephalitis, and demyelinating diseases such as acute disseminated encephalomyelitis (ADEM) and multiple sclerosis (MS).[40] Therefore, prompt and accurate diagnosis is critical to select the most appropriate imaging method (MRI vs. CT) for each clinical scenario.

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Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)

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Indications:

When acute/subacute or rapid progression (<3 months) of altered mental status, focal findings including cranial nerve, motor or sensory symptoms or memory loss or psychiatric symptoms, seizure, and/or focal CNS findings are present.

OR

There is a stated concern for neuro-inflammatory encephalitis from or in consultation with a neurologist, neurosurgeon, psychiatrist, oncologist, rheumatologist, or infectious disease specialist.

Medically Necessary Initial Imaging:

- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **OR**
- CT Head without contrast (CPT® 70450) **OR** CT Head without and with contrast (CPT® 70470) when MRI is unavailable or contraindicated or for bony pathology concerns
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** CTA Head (CPT® 70496) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR** CTA Neck (CPT® 70498) for evaluating large vessel obstructions, aneurysms and vascular malformations, dissection, vasospasm, and vasculopathies such as CNS vasculitis (see **Cerebral Vasculitis (HD-22.1)**, **Cerebral Aneurysms (HD-12.1)**, **Arteriovenous Malformations (AVMs) and Related Lesions (HD-12.2)**, **Stroke/TIA (HD-21.1)**)

Medically Necessary Repeat Imaging:

MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) when specialized sequences are needed such as, but not limited to:

- high T2 contrast sequences (CISS, FIESTA) sequences to identify blood (SWI) or
- identifying acute cytotoxic edema (DWI) or
- when requested by or in consultation with a neurologist, oncologist, rheumatologist, infectious disease specialist, neurosurgeon, or psychiatrist

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Metabolic (FDG) Brain PET (CPT® 78608) is medically necessary to evaluate individuals suspected of having encephalitis, including autoimmune encephalitis, if diagnosis remains unclear after evaluation with MRI Brain, CSF analysis, and/or lab testing including serology.

Neurosarcoidosis:

- The following are medically necessary for known or suspected neurosarcoidosis.
  - MRI Brain without and with contrast (CPT® 70553)

    AND/OR
  - If spinal cord involvement suspected, then
    - MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR**
    - MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR**
    - MRI Lumbar Spine without and with contrast (CPT® 72158)

    AND/OR
  - If peri-orbital involvement suspected, then
    - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
  - Repeat imaging supported if requested by neurologist, rheumatologist, ophthalmologist, oncologist, or radiologist or provider in consultation with a neurologist, rheumatologist, ophthalmologist, oncologist or radiologist.
- For non-neurologic imaging related to sarcoidosis (see **Sarcoid (CH-15.1)**)

Background and Supporting Information

Supportive studies in the evaluation of Autoimmune/Paraneoplastic Encephalopathy include:

- CSF pleocytosis (>5 WBC/μL) or
- EEG changes or
- Supporting labs (including positive CSF antibody positivity and/or serologies)

Potential etiologies:

- Paraneoplastic
  - NMDA Receptor encephalitis
  - LGI1 antibody encephalitis
- Autoimmune
  - Neurosarcoidosis can involve any of the following:
    - Brain, Cranial Nerves, Spinal Cord, and/or Peripheral Nerves
    - Acute Disseminated Encephalomyelitis (ADEM), Anti-MOG Syndrome, Multiple Sclerosis (MS), Neuromyelitis Optica (NMO)
    - IgG4 related disease

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    - CNS histiocytosis
- Neuro-rheumatologic
    - ANCA related disease
    - Behcet's disease
    - Sjogren Syndrome +/- Rheumatoid Arthritis (RA)

FDG-PET imaging of the brain for paraneoplastic and autoimmune encephalitis may be more sensitive than Brain MRI (87% vs. 56%) but is nonspecific. Areas of hypometabolism are seen in neurodegenerative disorders such as dementias. However, topographic patterns of hypometabolism may help characterize the disorder as autoimmune/ paraneoplastic encephalitis, in a way that may help clarify diagnosis and alter management strategies. For example, anterior to posterior gradient of hypometabolism is seen in NMDA Receptor encephalitis. Hemispheric hypometabolism out of proportion to atrophy characterizes Rasmussen encephalitis.

Non-head Imaging

- MRI is helpful in determining the length of spine lesion (short versus longitudinally extensive transverse myelitis), width (partial versus transverse), and location (eccentric, central, hemicord, anterior versus posterior, conus, tracts, or meningeal).
    - See **Myelopathy (SP-7.1)** and **Anti-MOG Syndromes (HD-16.3)**
- The Trident Sign on axial MRI, which has been described in relation to neurosarcoidosis, demonstrates leptomeningeal or dorsal subpial enhancement that may or may not involve the central canal.
    - See **Myelopathy (SP-7.1)**
- Involvement of the conus medullaris is a clue to Anti-MOG (Myelin Oligodendrocyte Glycoprotein-associated disorder) as the cause of longitudinally extensive transverse myelitis.
    - See **Transverse Myelitis (HD-16.4)**
- CT of the chest, abdomen, and pelvis with contrast is a generally accepted first method of screening for occult malignancy or systemic inflammation (e.g., sarcoidosis).
    - See **Paraneoplastic Syndromes (ONC-30.3)** and **Sarcoid (CH-15.1)**

Evidence Discussion (HD-14.3)

- The American College of Radiology (ACR) Appropriateness Criteria® provided guidance on the appropriateness of CT versus MRI Brain in certain clinical scenarios relating to altered mental status. For acute, undifferentiated presentations and with focal symptoms, CT head is an appropriate modality, but with known intracranial process, suspected medical illness or toxic-metabolic etiology, and/or psychosis, MRI Brain, is appropriate and in some cases may be preferable.[45]

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- In the appropriate clinical scenarios, recognition of structural and functional imaging patterns of brain involvement using CT head, MRI brain, and brain PET, in autoimmune encephalitis (including paraneoplastic and non-rheumatologic inflammatory disorders) can facilitate rapid access to appropriate treatment, as well as avoid invasive diagnostic procedures such as brain biopsy.[26]
- FDG-PET/CT brain performed at a median 4 weeks of symptom onset was more often abnormal than initial MRI, EEG, or laboratory cerebrospinal fluid testing, in individuals with suspected autoimmune encephalitis (AE), with focal hypometabolism, the most common PET/CT finding.[2]
- Spine MRI shows abnormalities in up to 45% of cases of paraneoplastic myeloneuropathy.[47]
- The three-dimensional (3D) constructive interference in steady state (CISS) is a gradient-echo MRI or Fast Imaging Employing Steady-state Acquisition Cycled Phases (FIESTA-C) on GE MRI systems are widely employed for over a decade and have been shown to have utility in demonstration of contrast between cerebrospinal fluid and brain parenchymal structure. Therefore, these have particular utility in the examination of cranial nerves, the ventricular system, cavernous sinus, and other structures which are commonly involved in neuro-inflammatory conditions.[48]
- Neuropsychiatric lupus and other neuro-inflammatory conditions have been described to mimic vascular disease, such as vasculitis and small vessel cerebrovascular disease, and in these cases vessel imaging with CT Angiography (CTA) and MR Angiography (MRA) Brain can contribute to meaningful diagnosis.[49]
- In the evaluation of a first episode of psychosis when an autoimmune cause is suspected, up to 4% of cases have abnormalities on MRI brain.[50]
- Sarcoidosis can manifest with neurologic complications in every part of the neural axis, with diagnostic challenges represented by multiple pathophysiologic pathways and frequently lack of specific histopathologic diagnosis. In clinically suspected neurosarcoidosis on the basis of synthesized clinical history and physical examination findings, the demonstration of neuro-inflammation using cerebrospinal fluid testing and contrast-enhanced MRI is useful. In difficult or complicated cases, FDG-PET and Gallium-67 imaging have been useful for identification of targets for biopsy.[51]

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Movement Disorders (HD-15)

Guideline

Movement Disorders (HD-15.1)
References (HD-15)

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Movement Disorders (HD-15.1)

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- The majority of movement disorders are diagnosed based on a clinical diagnosis and imaging is not medically necessary. These include:
  - typical Parkinson's Disease
  - essential tremor or tremors of anxiety or weakness
  - restless leg syndrome
  - tics or spasms which can be duplicated at will
  - Tourette syndrome
  - tardive dyskinesia
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary in the following clinical scenarios:
  - Clinical diagnostic uncertainty
  - Incomplete or uncertain response to medication
  - Anti-psychotic drug-induced Parkinsonism or atypical tardive dyskinesia
  - Atypical Parkinsonism suspected because of unusual clinical features. These may include, but are not limited to:
    - Persistent unilateral signs or symptoms
    - Onset under age 50
    - Rapid progression
    - See *Background and Supporting Information* for further information on atypical parkinsonism and Parkinson's Plus Syndromes
  - Suspected Huntington Disease
- Evaluation for surgical treatment of essential tremor, Parkinson's disease, and/or spasmodic torticollis/dystonia, see **Torticollis and Dystonia (Neck-10.2)** in the Neck Imaging Guidelines
  - Deep Brain Stimulation (DBS) therapy
    - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** unlisted CT procedure code (CPT® 76497) are medically necessary
  - MR guided Focused Ultrasound:
    - CT Head without contrast (CPT® 70450) to evaluate bone density **AND/OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) are medically necessary
  - Repeat imaging studies for pre-surgical evaluation, MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** CT Head without contrast (CPT® 70450), are medically necessary when ordered by a

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Neurosurgeon or Neurologist or any provider in consultation with a Neurosurgeon or Neurologist if greater than 6 months old **and/or** for new symptoms/signs
  - Post-op imaging is medically necessary when ordered by a Neurosurgeon or Neurologist or any provider in consultation with a Neurosurgeon or Neurologist for either procedure, see also **Post-Operative Imaging (HD-28.3)** indications
- Dopamine Transporter Scan [DAT-SPECT Radiopharmaceutical Localization SPECT (Ioflupane I-123 SPECT)] (CPT® 78803 or CPT® 78830) **OR** Fluorodopa F^{18} (F-DOPA) PET Brain (CPT® 78608) is medically necessary to:[2,3,16,17,18]
  - evaluate motor symptoms (tremor, rigidity, bradykinesia, and/or postural instability) in suspected Parkinsonian Syndromes (Parkinson Disease, Multiple System Atrophy, Progressive Supranuclear Palsy, Corticobasal Degeneration) when the diagnosis is unclear, to differentiate from non-neurodegenerative disorders, such as Essential Tremor, drug-induced tremor, vascular parkinsonism, and/or psychogenic tremor
  - Imaging with either modality, **not both**, is medically necessary when the diagnosis remains unclear after evaluation by a neurologist, medication trials and brain imaging.
  - See *Background and Supporting Information* for additional information regarding Parkinson's Plus Syndromes.
  - Neither DAT Scans nor F-DOPA PET scans are medically necessary for differentiation of subtypes of Parkinson's syndromes, to monitor progression of disease, or predict risk of development of disease mainly to exclude other conditions with similar clinical presentations.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for initial imaging for suspected motor neuron disease (see **Motor Neuron Disease/ Amyotrophic Lateral Sclerosis (ALS) (PND-8.1)** in the Peripheral Nerve and Neuromuscular Disorders Imaging Guideline)
- Dementia associated with movement disorder (see **Lewy Body Dementia (LBD) – SPECT Brain Scan and PET (HD-8.3))**

Background and Supporting Information

- Parkinson's Plus Syndromes are a group of disorders characterized by atypical parkinsonism. They are NOT Parkinson's disease. They represent different neurodegenerative diseases with features of PD, and may be confused with PD. These syndromes include, but are not limited to:
  - Multiple system atrophy: orthostatic hypotension (dysautonomia), dysphonia, dysarthria
  - Progressive Supranuclear Palsy: balance difficulties, vertical gaze paresis
  - Corticobasal Syndrome: dysphasia, apraxia, myoclonus, alien-limb phenomenon
- These are distinct entities. Care must be taken to determine if there are unusual features present that will suggest atypical parkinsonian syndrome.

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- Dementia with Lewy bodies (DLB): dementia prior to movement disorder (see **Lewy Body Dementia (LBD) - SPECT Brain Scan and PET (HD-8.3)**)

Evidence Discussion (HD-15)

- The majority of movements disorders are diagnosed based on history and clinical examination findings and do not require imaging.[1,2,3,24] For cases of diagnostic uncertainty, incomplete response to medication, for atypical Parkinsonism or drug-induced parkinsonism, and for suspected Huntington disease, MRI brain is the preferred imaging modality.[1,3,24]
- Structural imaging with MRI Brain is usually normal in individuals with Parkinson's disease but is useful to diagnose causes of secondary parkinsonism, such as stroke, iron deposition, normal pressure hydrocephalus, and neoplasm.[1,2,3]
- CT Head is not preferred due to its limited soft-tissue characterization when compared to MRI.[1]
- Functional imaging studies assessing dopaminergic function in Parkinson's disease include single-photon emission computed tomography (SPECT) and positron emission tomography (PET) imaging.1,2,3 These studies are used as an adjunct diagnostic test.[1,2,3]
- Both Dopamine Transporter single-photon emission computed tomography (DAT-SPECT) and [18F]-fluorodopa (F-DOPA) PET brain are useful to differentiate suspected parkinsonian syndromes from non-neurodegenerative disorders such as Essential Tremor, drug-induced tremors, vascular parkinsonism, and/or psychogenic tremors.[1,2,3,17,18]
- DAT-SPECT has a sensitivity of 91% with a specificity of 100% for essential tremor, a sensitivity of 86.2% with a specificity of 93.8% for drug-induced parkinsonism, and a sensitivity of 86.2% with a specificity of 82.9% for vascular parkinsonism.[18,25,26]
- F-DOPA PET brain has a sensitivity of 73% with a specificity of 91% for evaluation of parkinsonian syndrome vs non-neurodegenerative parkinsonian syndrome.[17]
- Neither DAT-SPECT scans nor F-DOPA PET brain scans are useful for the differentiation between subtypes of Parkinsonian syndromes, to monitor progression of disease nor to predict the risk of development of disease.[1,2,3]
- There is insufficient evidence for the routine use of FDG-PET brain in the diagnosis of parkinsonian syndromes.[1]

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Multiple Sclerosis (MS) and Related Conditions (HD-16)

Guideline

Multiple Sclerosis (MS) (HD-16.1)
Neuromyelitis Optica Spectrum Disorders (HD-16.2)
MOG Antibody-Associated Disease (MOGAD) (HD-16.3)
Transverse Myelitis (HD-16.4)
Evidence Discussion (HD-16)
References (HD-16)

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Multiple Sclerosis (MS) (HD-16.1)

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Establishing a New Diagnosis of Multiple Sclerosis

Repeat Imaging for Unclear Diagnosis

New Neurologic Symptoms in an Individual with Multiple Sclerosis

Baseline Imaging with Disease Modifying Therapy (DMT)

Current Treatment with High-Risk Disease Modifying Therapy (DMT)

Annual Imaging on Low-Risk DMT or No Treatment

History of Clinically Isolated Syndrome (CIS) or Radiologically Isolated Syndrome (RIS)

Prolonged Treatment with Tysabri® (natalizumab)

Progressive Multifocal Leukoencephalopathy (PML) Evaluation

Background and Supporting Information

Multiple Sclerosis General Information Page

- MRI Lumbar Spine is not medically necessary since Cervical and Thoracic studies will usually visualize the entire spinal cord. If the clinical concern is for lumbosacral radiculopathy, see **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)** in the Spine Imaging Guidelines.
- Family members need not be screened, unless they exhibit suspicious signs or symptoms suggestive of MS.
- Computed Tomography (CT) scans of the head and/or spine are **NOT** medically necessary for the evaluation of multiple sclerosis due to inferior soft tissue resolution when compared to MRI.
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as (CPT® 0865T or CPT® 0866T).
    - These studies are not medically necessary in the evaluation of Multiple Sclerosis.
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as 3D rendering (CPT® 76376 and CPT® 76377).

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- These studies are considered not medically necessary in the evaluation of multiple sclerosis.

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Establishing a New Diagnosis of Multiple Sclerosis

Indication	Medically Necessary Imaging
Establishing a new diagnosis of Multiple Sclerosis is based on the following: • Clinical suspicion based on recurrent episodes of variable neurological signs and/or symptoms **AND** • Baseline exclusion of appropriate alternative conditions that can mimic MS **OR** Clinically Isolated Syndrome (CIS)* based on **ALL** the following: • First episode of neurologic symptoms and neurologic deficits concerning for possible demyelinating disease • Symptoms last ≥24 hours • Baseline exclusion of appropriate alternative conditions that can mimic MS **OR** Radiologically Isolated Syndrome (RIS)* based on **ALL** the following: • Individual with brain MRI obtained for unrelated reason with findings conspicuous for demyelinating disease	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) if there is a contraindication to gadolinium If optic neuritis** is suspected the following imaging is **ALSO** medically necessary: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ****For additional information related to optic neuritis see Eye Disorders and Visual Loss (HD-32.1)** **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

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Indication	Medically Necessary Imaging
• Baseline exclusion of appropriate alternative conditions that can mimic MS *For more information about CIS and RIS, see *Background and Supporting Information*	

Repeat Imaging for Unclear Diagnosis

Indication	Medically Necessary Imaging
Diagnosis of Multiple Sclerosis remains unclear or equivocal after initial MRI • Repeat imaging 3-6 months after initial MRI Brain	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

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New Neurologic Symptoms in an Individual with Multiple Sclerosis

Indication	Medically Necessary Imaging
New neurologic signs or symptoms in an individual with multiple sclerosis	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) If optic neuritis is suspected**, the following imaging is **ALSO** medically necessary: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ****For additional information related to optic neuritis, see Eye Disorders and Visual Loss (HD-32.1)** If there are new or worsening symptoms concerning for spinal cord involvement, the following imaging is **ALSO** medically necessary: • MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

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Baseline Imaging with Disease Modifying Therapy (DMT)

Indication	Medically Necessary Imaging
• Before starting **OR** changing disease modifying therapy (DMT) **AND/OR** • 3-6 months after starting or changing DMT to establish a new MRI treatment baseline **AND/OR** • If there are new abnormal MRI Brain findings without clinical symptoms, an additional follow-up MRI Brain is supported after 6 months	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

Current Treatment with High-Risk Disease Modifying Therapy (DMT)

Indication	Medically Necessary Imaging *Every 6 Months*	Medically Necessary Imaging *Annually*
Individuals treated with DMT* associated with either the risk of progressive multifocal leukoencephalopathy (PML) **AND/OR** other CNS opportunistic infections * For list of medications, see *Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

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Annual Imaging on Low-Risk DMT or No Treatment

Indication	Medically Necessary Imaging Annually
Individuals with diagnosed Multiple Sclerosis with **EITHER** of the following: • Not treated with disease modifying therapy (DMT)* **OR** • Treated with low-risk DMT (beta interferon or glatiramer acetate medications) * For list of DMT medications, see *Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

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History of Clinically Isolated Syndrome (CIS) or Radiologically Isolated Syndrome (RIS)

Indication	Medically Necessary Imaging *Annually*
Individual with history of Clinically Isolated Syndrome* (CIS) **OR** Individual with history of Radiologically Isolated Syndrome* (RIS) *For more information about CIS or RIS, see *Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) If there are new or worsening symptoms concerning for spinal cord involvement, the following imaging is **ALSO** medically necessary: • MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) OR • MRI Thoracic Spine without and with contrast (CPT® 72157)

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Prolonged Treatment with Tysabri® (natalizumab)

Indication	Medically Necessary Imaging *Every 3-6 Months*	Medically Necessary Imaging *Annually*
Individuals treated with Tysabri® (natalizumab) with the following medical history: • ≥18 months of treatment ◦ During Tysabri® (natalizumab) treatment and up to 9-12 months after transitioning off Tysabri® (natalizumab) **AND** • JC virus antibody positive	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

Progressive Multifocal Leukoencephalopathy (PML) Evaluation

Indication	Medically Necessary Imaging
Symptoms suggestive of PML* during treatment with Tysabri® (natalizumab) or other medication with similar risk * For more information about PML, see *Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

Background and Supporting Information

- Multiple sclerosis is an autoimmune disease that is associated with inflammation, demyelination, and neurodegenerative changes within the optic nerves, brain, and spinal cord (i.e., central nervous system (CNS)).

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- Multiple sclerosis commonly begins with a relapsing-remitting course with partial or complete neurologic recovery following acute events.
  - An acute demyelinating event lasts at least 24 hours or longer
  - Common types of MS relapses include:
    - Unilateral optic neuritis
    - Brainstem or cerebellar syndrome (i.e. trigeminal neuralgia, diplopia or intranuclear ophthalmoplegia (INO), and/or ataxia)
    - Partial transverse myelitis
- The first event concerning for demyelinating disease without meeting criteria for separation of time is known as a clinically isolated syndrome (CIS).
- Individuals who undergo a brain MRI for other indications (i.e., headaches, trauma, seizure) which incidentally reveals abnormalities that are characteristic for demyelination in the absence of clinical symptoms is known as radiologically isolated syndrome (RIS).
- Progressive Multifocal Leukoencephalopathy (PML) is a progressive multi-focal disease of the central nervous system that can occur in individuals treated with immunosuppressive or immunomodulatory medications.
  - In individuals treated with natalizumab, there is an increased risk of developing PML in individuals who:
    - Received prior immunosuppressive medication, and/or
    - Have a high JC virus antibody index, and/or
    - Received natalizumab for ≥18 months
- Interferon beta medications include (but are not limited to): Avonex®, Betaseron®, Extavia®, Plegridy®, Rebif®
- Glatiramer acetate medications include (but are not limited to): Copaxone, Glatopa®
- High-risk medications are associated with a risk of PML and/or other CNS opportunistic infections (i.e. herpes encephalitis, cryptococcal meningitis). These include (but are not limited to): Tysabri® (natalizumab), Tecfidera® (dimethyl fumarate), Gilenya® (fingolimod), Tascenso® ODT (fingolimod), Aubagio® (teriflunomide), Ocrevus® (ocrelizumab), Kesimpta® (ofatumumab), Mavenclad® (cladribine), Mayzent® (siponimod), Ponvory® (ponesimod), Vumerity® (diroximel fumarate), Zeposia® (ozanimod), Lemtrada® (alemtuzumab), Bafiertam® (monomethyl fumarate), Briumvi® (ublituximab), Rituxan® (rituximab)

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Neuromyelitis Optica Spectrum Disorders (HD-16.2)

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Initial evaluation of Neuromyelitis Optica (NMO) or Neuromyelitis Optica Spectrum Disorders (NMOSD) with any of the following:

Indication	Medically Necessary Imaging
Clinical concern for optic neuritis	• MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540)
Recurrent hiccups or intractable nausea and/or vomiting (clinical concern for area postrema syndrome)	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
Other neurologic signs or symptoms concerning for brain involvement	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

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Indication	Medically Necessary Imaging
Clinical concern for transverse myelitis	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148)

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Indication	Medically Necessary Imaging
Positive NMO antibody test when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

Individual with established diagnosis of (NMOSD) with any of the following:

Indication	Medically Necessary Imaging
Clinical concern for optic neuritis	• MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540)
New neurologic signs or symptoms concerning for brain involvement	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

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Indication	Medically Necessary Imaging
Clinical concern for transverse myelitis	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148)

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Indication	Medically Necessary Imaging
Repeat imaging for **ANY** of the following: • Re-establish baseline after starting treatment (typically 3-6 months after last MRI) • Changing disease modifying therapy (DMT) • As requested when ordered by a neurologist, neuro-ophthalmologist, ophthalmologist, or any provider in consultation with a neurologist, neuro-ophthalmologist, or ophthalmologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

- For Neuromyelitis Optica Spectrum Disorder with concern for occult neoplasm, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.
- Computed Tomography (CT) scans of the head and/or spine are not recommended for the evaluation of NMOSD due to inferior soft tissue resolution when compared to MRI.
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as (CPT® 0865T or CPT® 0866T).
    - These are not medically necessary in the evaluation of NMOSD.
  - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as 3D rendering (CPT® 76376 and CPT® 76377).
    - These studies are considered not medical necessary in the evaluation of NMOSD.

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Background and Supporting Information

- Neuromyelitis optica spectrum disorder (NMOSD, Devic's disease) is a chronic inflammatory autoimmune disease that involves the optic nerves, spinal cord, and brain.
- Core clinical characteristics of NMOSD include
  - Optic neuritis
    - Frequently bilateral optic nerve involvement with severe vision loss
  - Longitudinally extensive transverse myelitis
    - Extends ≥ 3 complete vertebral segments of the spinal cord
  - Area postrema syndrome
    - Otherwise, unexplained episode of recurrent hiccups or intractable nausea and vomiting
  - Brainstem or cerebral syndrome with NMOSD typical brain lesions
  - Rarely paraneoplastic syndromes occur with NMO spectrum disorder
  - Medications used for the treatment of NMO spectrum disorders include (but are not limited to) azathioprine, Enspryng® (satralizumab), mycophenolate, Soliris® (eculizumab), rituximab, Uplizna® (inebilizumab) and Ultomiris® (ravulizumab)[54]
    - Possible adverse reactions associated with treatment include risk of PML and meningococcal infections.

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MOG Antibody-Associated Disease (MOGAD) (HD-16.3)

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Initial evaluation of MOG (myelin oligodendrocyte glycoprotein) antibody-associated diseases (MOGAD) with any of the following:

Indication	Medically Necessary Imaging
Clinical concern for optic neuritis	• MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540)
Neurologic signs or symptoms concerning for brain involvement	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
Clinical concern for transverse myelitis	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148)

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Indication	Medically Necessary Imaging
Positive MOG antibody test when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148)

Individuals with established diagnosis of (MOGAD) with any of the following:

Indication	Medically Necessary Imaging
Clinical concern for optic neuritis	• MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540)
Neurologic signs or symptoms concerning for brain involvement	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

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Indication	Medically Necessary Imaging
Clinical concern for transverse myelitis	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148)
Repeat imaging for **ANY** of the following: • Re-establish baseline after starting treatment (typically 3-6 months after last MRI) • Changing disease modifying therapy (DMT) • As requested when ordered by a neurologist, neuro-ophthalmologist, ophthalmologist, or any provider in consultation with a neurologist, neuro-ophthalmologist, or ophthalmologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** MRI Thoracic Spine without contrast (CPT® 72146)

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- For MOG (myelin oligodendrocyte glycoprotein) Antibody-Associated Disease with concern for occult neoplasm, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.
- Computed Tomography (CT) scans of the head and/or spine are not recommended for the evaluation of MOG (myelin oligodendrocyte glycoprotein) Antibody-Associated Disease due to inferior soft tissue resolution when compared to MRI.

Background and Supporting Information

- MOG (myelin oligodendrocyte glycoprotein)-IgG disorders are CNS inflammatory diseases, distinct from multiple sclerosis and NMO-spectrum disorders.
- Unlike multiple sclerosis and neuromyelitis optica spectrum disorder (NMOSD), individuals with MOG antibody-associated disease (MOGAD) can have a monophasic or relapsing course.
  - Relapses are more common in the first six months after the first episode.
  - An acute relapse is considered when an individual with MOGAD develops new neurologic signs or symptoms at least 30 days following the previous event.
- Diagnosis is based on the clinical presentation, MRI findings, and the presence of auto-antibodies.
- Clinical features of individuals with MOGAD include
  - Optic neuritis
    - Bilateral optic neuritis is common at onset, and seems to be more frequent in individuals with MOGAD than with those with multiple sclerosis or neuromyelitis optica spectrum disorder (NMOSD).
    - Vision improves quickly with return to normal or near normal visual acuity following treatment with intravenous corticosteroids.
  - Transverse myelitis
    - Cauda equina and peripheral nerve root involvement can occur (lumbar spine imaging is medically necessary)
    - Can occur as an isolated episode of transverse myelitis, as a component of ADEM or in conjunction with optic neuritis.
    - T2 spinal cord lesions often are centrally located and can be restricted to the grey matter producing the “H sign” on MRI
    - Most T2 lesions resolve or reduce in size substantially on follow-up MRI
  - Brainstem encephalitis
  - Encephalitis with seizures
  - Acute disseminated encephalomyelitis (ADEM)
    - Occurs mainly in children but can occur in adults.
  - Tumefactive brain lesions
  - Cranial neuropathies

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- Unlike multiple sclerosis, it is rare for individuals with MOGAD to develop asymptomatic lesions within the brain, optic nerves, and/or spinal cord.

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Transverse Myelitis (HD-16.4)

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An initial assessment, to include a pertinent history and neurologic exam, should be performed prior to imaging requests.

Clinical Concern for Transverse Myelitis

Indication	Medically Necessary Imaging
Clinical concern for transverse myelitis	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148) **AND/OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) If optic neuritis is suspected*, the following imaging is **ALSO** medically necessary: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ***For additional information related to optic neuritis see Eye Disorders and Visual Loss (HD-32.1)**

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New Neurologic Signs or Symptoms in an individual with a history of transverse myelitis (TM)

Indication	Medically Necessary Imaging
New neurologic signs or symptoms	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) If optic neuritis is suspected*, the following imaging is **ALSO** medically necessary: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ***For additional information related to optic neuritis, see Eye Disorders and Visual Loss (HD-32.1)**

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Surveillance of Transverse Myelitis

Indication	Medically Necessary Imaging Annually for 5 years
Individual with a history of transverse myelitis • Ordered by a neurologist or any provider in consultation with a neurologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

- For transverse myelitis with concern for occult neoplasm, see **Paraneoplastic Syndromes (ONC-30.3**) in the Oncology Imaging Guidelines.
- Individuals with transverse myelitis present with various symptoms of sensory, motor and/or autonomic dysfunction.
  - Bilateral signs and/or symptoms (although not necessarily symmetrical)
  - Examination findings may include but are not limited to any of the following:
    - Bilateral limb weakness
    - Loss of manual dexterity
    - New or worsening foot drop
    - Sensory abnormalities
    - Sensory level
    - Hyperreflexia (including upgoing toes, positive Babinski, Hoffman's sign, clonus)
    - Gait abnormality (spastic or ataxic gait)
    - See also: *Background and Supporting Information*
  - If inflammation is identified within the spinal cord suggestive of transverse myelitis, a brain MRI is recommended to evaluate for a multifocal inflammatory process
- See **Multiple Sclerosis (MS) (HD-16.1)**, **Neuromyelitis Optica Spectrum Disorders (HD-16.2)**, **MOG Antibody-Associated Diseases (MOGAD) (HD-16.3)**

Background and Supporting Information

- Symptoms may include but are not limited to the following:
  - Motor weakness of a limb or limbs, including paraparesis and/or complete paralysis

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  - Change in sensation in a limb or limbs that may be associated with paresthesias and/or dysthesias.
  - Urinary urgency, incontinence, and/or urinary retention
  - Worsening constipation and/or bowel urgency/incontinence
  - Sexual dysfunction
  - Lhermitte's sign
  - New or worsening spasticity
- Acute transverse myelitis is defined as an acute inflammatory syndrome leading to motor and/or sensory impairment, with or without sphincter dysfunction, secondary to a variety of autoimmune or inflammatory diseases.
- Diagnosed by spinal MRI and/or cerebrospinal fluid.
- Individuals typically progress to maximal neurological deficits within 4 weeks.
- Longitudinally extensive transverse myelitis (≥3 vertebral segments) is more commonly associated with neuromyelitis optica spectrum disorders (NMOSD) and/or MOG antibody-associated diseases (MOGAD)
- Transverse myelitis:
  - May be idiopathic
  - Initial event of multiple sclerosis (see **Multiple Sclerosis (MS) (HD-16.1)**
  - Initial event of neuromyelitis optica spectrum disorder (NMOSD) (see **Neuromyelitis Optica Spectrum Disorders (HD-16.2)**)
  - Initial event of MOG antibody-associated disease (MOGAD) (see **MOG Antibody-Associated Diseases (MOGAD) (HD-16.3)**)
  - May be associated with connective tissue disease
    - Systemic lupus erythematous (SLE)
    - Rheumatoid Arthritis (RA)
    - Sjögren's syndrome
    - Systemic sclerosis
  - Manifestation of neurosarcoidosis (see **Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)**)
  - Post-infectious and/or post-vaccination related
    - COVID-19 and COVID-19 post-vaccination myelitis cases have been reported (see **Neuro-COVID-19 and Sars-COV-2 Vaccines (HD-14.2)**)
    - May have a prodromal syndrome with fever, respiratory and/or gastrointestinal symptoms
    - May be associated with headache, neck stiffness or recurrence of fever

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Evidence Discussion (HD-16)

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- Magnetic resonance imaging (MRI) is the recommended imaging modality for the diagnosis and monitoring of multiple sclerosis (MS) and other inflammatory diseases of the central nervous system (CNS). Its high sensitivity for the evaluation of inflammatory and neurodegenerative processes in the brain and spinal cord has made it the gold standard for the evaluation of individuals with MS.[8,31]
- Computed Tomography Scan (CT) is not recommended for the evaluation of MS due to inferior soft tissue resolution when compared to MRI.[28,41,42]
- MRI plays an important role in the following clinical scenarios:
  - establishing the diagnosis of multiple sclerosis (MS) by establishing evidence for dissemination in space and time.[8,31]
  - diagnostic workup. Approximately 50%-90% of individuals with MS have spinal cord lesions.[8]
  - detecting optic nerve abnormalities in individuals with symptoms concerning for optic neuritis.[7,42]
  - assessment of treatment response and in monitoring for potential treatment related safety concerns. Management and surveillance intervals are primarily consensus based and have been addressed in several evidence and practice-based guidelines.[7,8]
  - evaluation of other central nervous system (CNS) inflammatory diseases, including autoimmune disseminated encephalomyelitis (ADEM), neuromyelitis optica (NMO), myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) and transverse myelitis (TM).[28,31,36,44]
  - detecting conus medullaris involvement in individuals with myelin oligodendrocyte glycoprotein-associated disease (MOGAD) and transverse myelitis.[28,36,44]

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Papilledema/Pseudotumor Cerebri (HD-17)

Guideline

Papilledema/Pseudotumor Cerebri (HD-17.1)
References (HD-17)

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Papilledema/Pseudotumor Cerebri (HD-17.1)

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- See **Eye Disorders and Visual Loss (HD-32.1)**
- Papilledema and Pseudotumor Cerebri (Idiopathic Intracranial Hypertension, Benign Intracranial Hypertension):
  - MRI Orbits/Face/Neck without contrast (CPT® 70540) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** CT Orbits/Temporal bone with contrast (CPT® 70481) **OR** CT Orbit/Temporal bone without contrast (CPT® 70480) **AND/OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is/are medically necessary for:
    - Suspected elevated intracranial pressure **AND/OR** papilledema
    - CT Head without contrast (CPT® 70450) is medically necessary when MRI is contraindicated or for urgent evaluation
    - See **General Guidelines – CT Head (HD-1.4)** regarding required use of CT Head prior to lumbar puncture and/or spinal tap.
    - See **Eye Disorders and Visual Loss (HD-32.1)** regarding concern for orbital pseudotumor or primary orbital disorder.
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) is medically necessary, in addition, for venogram when requested.
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures
    - See **Stroke/TIA (HD-21.1)**
    - See **Cerebral Venous Thrombosis (HD-21.8)**
  - Repeat structural and/or vascular imaging, with any of the above listed studies, for follow-up, including to confirm stent patency, and for evaluation of worsening symptoms, or suspected complications, as requested by a neurologist or a neurosurgeon or any provider in consultation with a neurologist or a neurosurgeon.
    - See also **Hydrocephalus Shunts (HD-11.14)**

Background and Supporting Information

Treatment for idiopathic intracranial hypertension (IIH) includes weight loss and acetazolamide. Surgical interventions include optic nerve sheath fenestration,

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cerebrospinal fluid shunting, and bariatric surgeries. These surgical procedures are invasive and associated with complications.

Venous sinus stenting is a treatment option for those with idiopathic intracranial hypertension who have failed first line therapy. Prior to stenting, individuals should undergo venography. CTV and/or MRV can be performed to confirm stent patency.

Stenting improves tinnitus, papilledema, diplopia/visual disturbances, and headaches.

Post- stent complications include subdural hematoma, subarachnoid hemorrhage, worsening headaches, visual impairment, worsening papilledema, blindness, arterial dissection, and death. Venous sinus stenting has lower complications relative to surgical interventions.

Evidence Discussion (HD-17)

- In the evaluation of suspected or known intracranial hypertension and/or exam findings of papilledema, neuroimaging is helpful for diagnosis, excluding other structural causes, and for the identification of venous outflow obstruction.[2,4,5]
- MRI Brain allows detection of findings supportive of intracranial hypertension and detection of structural abnormalities such as mass, edema, or hydrocephalus.[1,2,3,5,7] MR Venogram allows identification of venous sinus stenosis and thrombosis for treatment planning in these scenarios.[2,3,5]
- Alternatively, CT Head allows exclusion of secondary causes such as hydrocephalus, mass, or edema, particularly in urgent scenarios.[1,2,5,7] CT Venogram allows direct vessel visualization to exclude venous outflow obstruction in these cases.[1,2,5]
- In addition, orbital symptoms may be evaluated with either CT Orbits or MRI Orbits, with CT providing superior bony anatomy evaluation and calcification detection and MRI providing superior soft tissue resolution and evaluation of the optic nerve.[1,5,7]

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Paresthesias and/ or Weakness (HD-18)

Guideline

Sensory/Weakness Complaints (HD-18.1)
References (HD-18)

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Sensory/Weakness Complaints (HD-18.1)

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Advanced imaging for complaints of sensory loss and/or paresthesias (see *Background and Supporting Information)* and/or weakness that are unaccompanied by other symptoms and not preceded by trauma must have the following: a thorough clinical history and a detailed neurological exam (including the symptomatic area).

Imaging for sensory and weakness complaints is medically necessary with the following findings:

Indications	Medically Necessary Imaging
ANY of the following: • Hyperreflexia • Babinski/Hoffman sign* • Increased tone in affected limb • Bladder and/or bowel dysfunction • Motor symptoms in ANY of the following patterns: ◦ Two limbs on same side of body ◦ Face and limb involvement • Sensory symptoms in ANY of the following patterns: ◦ Two limbs on same side of body ◦ Face and limb involvement *See ***Background and Supporting Information***	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

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Findings Specific to the Spinal Cord	Medically Necessary Imaging
ANY of the following: • Decreased pinprick sensation on one side of the body with weakness and decreased proprioception on the other side • Sensory level (also called spinal cord level) on the trunk with sensory loss in both legs • Tight band around the trunk or torso • Pure sensory symptoms with proximal and distal involvement and a symmetric pattern • Decreased or absent reflexes AND noted concern for spinal cord shock or acute spinal cord injury* *See ***Background and Supporting Information***	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

Findings Specific to the Terminal End of the Spinal Cord	Medically Necessary Imaging
Concern for conus medullaris syndrome.* • Symptoms may include, but are not limited to: ◦ Saddle anesthesia ◦ Urinary retention ◦ Bowel incontinence ◦ Lower limb paresthesias ◦ Lower limb weakness *See ***Background and Supporting Information***	• MRI Lumbar Spine without contrast (CPT® 72148) **OR** • MRI Lumbar Spine without and with contrast (CPT® 72158)

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- MRI Lumbar Spine is not typically medically necessary to visualize the spinal cord except in the clinical scenarios noted above. MRI Cervical Spine and MRI Thoracic Spine will image the entire spinal cord.
- Findings NOT consistent with central nervous system localization and NOT supporting brain or spinal cord imaging include:
  - Sensory loss that involves the hands and feet and not the trunk
  - Limb pain
- For symptoms after trauma, refer to **Head Trauma (HD-13.1)** and/or the appropriate level in the Spine Imaging Guidelines
- For generalized weakness, polyneuropathy, and/or other patterns of sensory and/or motor symptoms not referenced above, refer to the following guidelines:
  - Myopathy or myositis, see **Muscle Diseases (PN-8.5)** and **Gaucher Disease (Storage Disorders) (PN-8.6)**
  - Motor Neuron Disease or Amyotrophic Lateral Sclerosis (ALS), see **Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)**
  - Neuromuscular Junction Disorders, see **Neuromuscular Junction Disorders (PN-8.4)**
  - Multifocal Motor Neuropathy (MMN) and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP), see **Polyneuropathy (PN-3.1)**
  - Polyneuropathy, see **Polyneuropathy (PN-3.1)**
  - Neuropathy with concern for malignancy, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines
  - Proximal asymmetric and concern for plexopathy, see **Brachial Plexus (PN-4.1)** and/or **Lumbar and Lumbosacral Plexus (PN-5.1)**
  - Sensory and/or motor symptoms localized to a single nerve, see **Focal Neuropathy (PN-2.1)**
  - Thoracic Outlet Syndrome, see **Thoracic Outlet Syndrome (CH-31.1)** in the Chest Imaging Guidelines
  - Radiculopathy, see appropriate level in the Spine Imaging Guidelines
  - Cauda Equina Syndrome, see **Red Flag Indications (SP-1.2)** in the Spine Imaging Guidelines

Background and Supporting Information

- Paresthesia refers to an abnormal sensation that is associated with nervous system dysfunction and may be described as a tingling, pricking, pins and needles, or a burning sensation. The priority is to determine whether the etiology is due to pathology of the peripheral nervous system (PNS) or central nervous system (CNS).
- A thorough clinical history, including symptom location and time course, can be helpful to differentiate PNS pathologies from CNS. For example, paresthesia affecting one side of the face and/or body (i.e., hemisensory deficit) points strongly towards

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central nervous system dysfunction. Therefore, brain and/or spinal cord imaging may be supported based on the location of symptoms. Typically, lumbar spine imaging is not supported unless there is sphincter involvement, saddle anesthesia, and/or cauda equina syndrome is suspected. In contrast, an insidious course of distal, symmetric limb paresthesia is more commonly associated with peripheral nerve abnormalities. In such cases, NCS/EMG testing results should be completed prior to advanced imaging. (See *Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines)*.

- Upper motor neuron signs (e.g., increased tone, hyperreflexia, presence of Babinski or Hoffman signs) may support a need for central nervous system imaging.
- Lower motor neuron signs (e.g., decreased tone, hypo- or areflexia, muscle atrophy) may support evaluation for peripheral nervous system diseases. Nerve conduction and needle EMG testing should be completed prior to advanced imaging.
- It is important to note that both peripheral and central nervous system disease can co-exist. As a result, if both upper and lower motor neuron signs are observed simultaneously, advanced imaging may be supported regardless of NCS/EMG testing results, (see **Polyneuropathy (PN-3.1)** in the Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines).
- Babinski sign - presence of an upgoing big toe with stimulation of the lateral plantar region of the foot.
- Hoffman sign - involuntary flexion of the fingers, particularly the thumb and index fingers, triggered by flicking the distal segment of the middle finger.
- Spinal cord shock/acute spinal cord injury - occurs after hyperacute or acute injury to the cord and presents with flaccid areflexia below the level of injury. May be associated with hypotension and/or bradycardia if loss of sympathetic tone occurs. Signs may last from days to weeks before upper motor neuron findings develop.
- Conus Medullaris Syndrome - compressive damage to the spinal cord from T12-L2. Symptoms suggestive of conus medullaris syndrome include saddle anesthesia, urinary retention, bowel incontinence, and/or lower extremity motor or sensory changes.

Evidence Discussion (HD-18)

- The imaging modality of choice for the evaluation of signs or symptoms localizing to the spinal cord is with MRI.[3,4,5,10,11] MRI allows visualization of the soft tissues and structures that comprise the neural axis.[3,10,11] Imaging of the cervical and thoracic segments are sufficient to view the entire spinal cord.[4,10]
- MRI of the lumbar spine is reserved for the evaluation of conus and the cauda equina.[4]
- For evaluation of isolated distal symmetric polyneuropathy, MRI of the brain and/or spine rarely change management in these individuals despite being frequently performed. MRI has little role in these scenarios as it evaluates the central nervous system.[7]

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Pituitary, Sella, Hypothalamus (HD-19)

Guideline

Pituitary (HD-19.1)
Post-Operative and Repeat Imaging Indications (HD-19.2)
Empty Sella Turcica (HD-19.3)
Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (HD-19.4)
Evidence Discussion (HD-19)
References (HD-19)

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Pituitary (HD-19.1)

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- Endocrine laboratory studies should be performed prior to considering advanced imaging, except in the cases of stable, non-functioning microadenomas or macroadenomas, cysts, and/or for incidentally found lesions.
- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) with a specific pituitary protocol that includes fine cuts through the sella is medically necessary as the primarily performed pituitary imaging:
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) or CT Head without and with contrast (CPT® 70470) are medically necessary alternatives
  - CT Head without contrast (CPT® 70450) or without and with contrast (CPT® 70470) **AND/OR** CT Maxillofacial without contrast (CPT® 70486) is medically necessary in addition to MRI to visualize perisellar bony structures in the pre-operative evaluation of certain sellar tumors and for pre-operative planning for transphenoidal approaches
    - See **General Guidelines – Anatomic Issues (HD-1.1)** as CT Temporal bone (CPT® 70480) is medically necessary instead of CT Maxillofacial per surgeon's preference and contrast level
  - CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) is medically necessary for surgical planning
  - MRI Brain without and with contrast (CPT® 70553) covers both brain and dedicated pituitary if performed at the same time; no additional CPT® codes are medically necessary
- Medically necessary repeat imaging for incidentally found lesions on other studies:
  - MRI Brain without and with contrast (CPT® 70553) or MRI Orbit/Face/Neck without and with contrast (CPT® 70543) follow-up dedicated pituitary study obtained if a pituitary abnormality is reported incidentally on a MRI Brain or CT Head performed for other reasons (MRI Brain without and with contrast [CPT® 70553] covers both brain and dedicated pituitary if performed at the same time; no additional CPT® codes are needed); further evaluation and subsequent imaging dependent on specific imaging and biochemical laboratory evaluation findings.
- Repeat imaging in the setting of worsening clinical status or new neurologic symptoms
- See in the Pelvic Imaging Guidelines for initial lab and imaging work up to exclude other causes. See Female Hypogonadism or Prolactinoma or other relevant sections in the grid if suspicion for pituitary tumor/disease or **Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (HD-19.4)** for suspicion of hypothalamic causes.

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Pituitary Imaging

Indication	Medically Necessary Initial Imaging	Medically Necessary Repeat Imaging
Microadenoma: Nonfunctioning, unexplained pituitary asymmetries, or incidentally found small tumors (<10 mm)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) at 12 months and then (if stable in size), every 1-2 years for 3 years, and less frequently thereafter based on clinical status
Macroadenoma (≥10 mm): Nonfunctioning and/or not surgically removed including those with a post-operative remnant	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) every 6 months for the first year and then (if stable in size), every year for 3 years, and less frequently thereafter based on clinical status (longer if craniopharyngioma)
Acromegaly* (Elevated IGF-1 confirmed by lack of suppression of growth hormone on glucose suppression testing)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) ◦ At least 12 weeks after surgery to evaluate for residual tumor ◦ If treated with Pegvisomant, 6 to 12 months after treatment initiated, then annually if stable ◦ Long-term follow-up imaging based on clinical and biochemical status at the request of a specialist or any provider in consultation with a specialist

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Indication	Medically Necessary Initial Imaging	Medically Necessary Repeat Imaging
Cushing's Disease** (Pituitary ACTH excess leading to hypercortisolism)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) ◦ At least 12 weeks after surgery as new baseline ◦ Annually after bilateral adrenalectomy for Cushing's disease or ectopic ACTH production ◦ Long-term follow-up imaging based on clinical and biochemical status at the request of a specialist or any provider in consultation with a specialist
Rathke's cleft cyst/ Simple cyst	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) in one year; if stable and without mass effect or invasion into surrounding structures, no further imaging is required.

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Indication	Medically Necessary Imaging
Prolactinomas* **	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) with: ◦ Diagnosis: ▪ Unexplained prolactin level above the normal range ◦ On Dopamine Agonist (DA) therapy with good response: ▪ Macroadenomas 3 months after start of DA therapy ▪ Microadenomas 1 year after start of DA therapy ▪ To decide on stoppage of therapy after ~2 years if in “remission” (normal PRL and no visible tumor on MRI) ◦ On Dopamine Agonist therapy with suboptimal response: ▪ PRL levels rise ▪ New symptoms develop (galactorrhea, vision changes, headaches, pituitary deficiency) ▪ If on high dose maximal DA and no plans for surgery/radiation therapy use guideline for microadenoma or macroadenoma ◦ After Dopamine Agonist therapy: ▪ Rise in PRL level ▪ For DA stoppage at menopause, use guideline for microadenoma or macroadenoma ◦ Not on therapy – refer to recommendations for repeat imaging for microadenoma or macroadenoma ◦ Galactorrhea/nipple discharge with normal prolactin and thyroid function levels: See **Nipple Discharge/ Galactorrhea (BR-6.1)** in the Breast Imaging Guidelines
Medication-induced Prolactinemia**	• To differentiate between medication-induced hyperprolactinemia and hyperprolactinemia due to a pituitary or hypothalamic mass if the medication cannot be discontinued or hyperprolactinemia persists after medication discontinuation

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Indication	Medically Necessary Imaging
TSH, FSH, or LH producing adenomas (inappropriate pituitary hypersecretion of TSH, FSH or LH)***	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) when hormone levels are inappropriately elevated and there is a concern for a pituitary lesion. • Refer to appropriate post-operative, or Microadenoma/ Macroadenoma guidelines based on the size of the lesion and initial management. ◦ Long-term follow-up imaging based on clinical and biochemical status at the request of a specialist or any provider in consultation with a specialist
Male Hypogonadism****	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if **ONE** of the following: ◦ Severe secondary hypogonadism (as indicated by morning serum testosterone level <150 ng/dl and low or normal LH and FSH levels) (See *Background and Supporting Information*) ◦ Below normal testosterone level (serum total testosterone, free testosterone, and/or bioavailable morning testosterone) AND low or normal LH and FSH levels, in an individual with either: ▪ Panhypopituitarism ▪ Hyperprolactinemia ▪ Signs of tumor mass effect (headache, visual impairment, or visual field deficit) ▪ Elevated sex hormone binding globulin (SHBG)
Female Hypogonadism (Secondary Amenorrhea may be a feature)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for normal or low FSH with low estradiol (LH may be normal or low also)

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Indication	Medically Necessary Imaging
Growth Hormone Deficiency (Adult onset)	MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for the following: • Low Growth Hormone (GH) **OR** • Low IGF-1 **AND** • One abnormal provocative test (likely will be Glucagon Stimulation test as GNRH is unavailable and Insulin Tolerance test poses risks) • If 3 or more pituitary hormones are deficient (including GH), then provocative test is not needed.
Secondary (Central) Adrenal Insufficiency	MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for the following: • ACTH is low or normal at 10 or lower **AND** • Low baseline cortisol level <3 µg/dL **OR** • abnormal ACTH stimulation test with suboptimal cortisol stimulation where cortisol does not reach above 18 µg/dL
Central Hypothyroidism	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for the following: ◦ Low free T4 with normal, low, or mildly elevated TSH
Hypopituitarism **(deficiency of one or more pituitary hormones)**	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)

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Indication	Medically Necessary Initial Imaging	Medically Necessary Repeat Imaging for Non-Operative Care
Diabetes Insipidus (DI) - ADH deficiency	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if: ◦ Laboratory testing consistent with DI (serum osmolality should be high and urine osmolality should be low) and etiology uncertain	NA
Syndrome of Inappropriate ADH (SIADH)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if: ◦ Etiology remains uncertain or is thought to be in the nervous system ◦ Urine osmolality should be high and serum osmolality low	NA
Other Pituitary Region Tumors	• Evaluation may require CT in addition to MRI to evaluate for hyperostosis.	

Background and Supporting Information

- ***Acromegaly:** A serum level of growth hormone greater than 1ng/mL when measured two hours following an oral glucose load confirms acromegaly.
- ****Cushing's Disease:** It is important to differentiate Cushing's syndrome (hypercortisolism from any source) from Cushing's disease which is ACTH hypersecretion from the pituitary gland. Hypercortisolism is quantified by 24hour urine cortisol collection, low dose dexamethasone suppression test and/or late-night salivary cortisol measurement. ACTH is elevated or inappropriately normal in Cushing's disease and ectopic sources of ACTH production but suppressed in other causes of hypercortisolism. A high dose dexamethasone suppression test can help determine if the elevated ACTH is from a pituitary or ectopic source. Petrosal sinus sampling may be required for tumor localization pre-operatively in the setting of a normal pituitary MRI or a small adenoma. These tumors may be managed with surgery, medical therapy, radiation, and/or bilateral adrenalectomy.
- ***Prolactinoma:** To establish the diagnosis of hyperprolactinemia, a single measurement of serum prolactin is recommended; a level above the upper limit of normal confirms the diagnosis as long as the serum sample was obtained without excessive venipuncture stress. Pregnancy and primary hypothyroidism should be

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excluded as physiologic causes of prolactin elevation and medications that may be contributing to prolactin elevation should be considered. Dopamine agonist therapy is typically stopped during pregnancy, monitoring of prolactin levels ceases. Routine imaging surveillance during pregnancy is not recommended due to risk to fetus. Repeat imaging with MRI without gadolinium can be performed however for new or worsening symptoms, such as headaches or visual symptoms.

- **** **Medication-induced prolactin elevation:** Medication induced hyperprolactinemia is seen most commonly with antipsychotics/neuroleptics and antidepressants but may also be seen with some anti-emetics and antihypertensive agents. In individuals on prolactin elevating drugs, a prolactin level should be repeated after withdrawal of medications for 72 h, however, this approach may not be safe if this treatment is offered for psychiatric indications. If stopping the drug is not feasible, pituitary MRI is advised to rule out a sellar/parasellar tumor.
- *****TSH, FSH, or LH producing adenomas:** These are the least common of all hormonally active pituitary tumors. Individuals with TSH secreting adenomas have inappropriate TSH elevation in the setting of hyperthyroidism (elevated thyroid hormone levels). Almost all gonadotroph adenomas are clinically non-functioning. The infrequent presentation of a functioning gonadotroph adenoma should be differentiated clinically from appropriate FSH and LH elevation seen in low estrogen states (including menopause) as well as primary hypogonadism (testicular failure). Functioning TSH, FSH or LH pituitary adenomas may be managed with surgical, radiation and/or medical therapies.
- ******Male Hypogonadism:** Alterations in sex hormone-binding globulin (SHBG) can impact testosterone levels. Free or bioavailable testosterone concentrations should be measured when total testosterone concentrations are close to the lower limit of the normal range and when altered SHBG levels are suspected (e.g. moderate obesity, nephrotic syndrome, hypo- and hyperthyroidism, use of glucocorticoids, progestins, estrogens, and androgenic steroids, anticonvulsants, acromegaly, diabetes mellitus, aging, HIV disease, liver cirrhosis, hepatitis). LH and FSH should be obtained to evaluate for secondary (central) hypogonadism once low testosterone level is confirmed. Morning testosterone level is drawn anytime before 10 am for a typical sleep-wake cycle.
- Central hypothyroidism is an anatomic or functional disorder of the pituitary gland or the hypothalamus, resulting in altered TSH secretion. Diagnosis is usually made biochemically with low circulating free T4 (FT4) concentrations associated with low/normal serum TSH levels.

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Post-Operative and Repeat Imaging Indications (HD-19.2)

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- For imaging in the immediate post-operative period or for acute surgical complications
  - See **Primary Central Nervous System Tumors (ONC-2.1)** in the Oncology Imaging Guidelines.
- A routine post-operative MRI is generally done at 3 months and/or at the discretion of, or in consultation with an Endocrinologist, Neurologist, Neurosurgeon, ENT, Ophthalmologist, Neuro-Ophthalmologist or Radiation Oncologist.
- Frequency of follow-up imaging depends on the post-operative size and/or functional status of the pituitary adenoma. Refer to the grid sections for Microadenoma/ Macroadenoma as well as those for disorders of pituitary hormone excess.
- Individuals with hyper-functioning tumors such as acromegaly, Cushing's disease, and excess TSH secretion may be treated with a combination of surgery, medical therapy, and radiation. Long-term monitoring of clinical status and repeat imaging at the discretion of, or in consultation with an Endocrinologist, Neurologist, Neurosurgeon, ENT, Ophthalmologist, Neuro-Ophthalmologist or Radiation Oncologist.

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Empty Sella Turcica (HD-19.3)

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- Enlarged/Empty Sella Turcica: An enlarged sella turcica without evident tumor is an incidental finding on MRI Brain or CT Head from a defect in the dural diaphragm of the sella (especially if there is elevated intracranial pressure from another cause), pituitary surgery, or as a result of a pituitary tumor which has expanded the sella and then infarcted (pituitary apoplexy).
- MRI Brain without and with contrast (pituitary protocol) (CPT® 70553) with thin sections of pituitary or MRI Brain without contrast (CPT® 70551) is medically necessary. CT Head without and with contrast (CPT® 70470) – If MRI is contraindicated.
  - Primary Empty Sella:
    - Incidentally found on other studies, asymptomatic and no related abnormalities: follow-up at 2 years. No further imaging unless clinical symptoms develop (neuro-/ophthalmological symptoms, intracranial hypertension, or endocrine/hormonal abnormalities).
    - Following medical or surgical treatment of related endocrine, neurological, or ophthalmological problems: follow-up imaging every 6 months in the year after treatment and/or at the request of a specialist or any provider in consultation with a specialist (see **Papilledema/Pseudotumor Cerebri (HD-17.1)** for additional imaging recommendations)
  - Secondary Empty Sella
    - Imaging according to the cause or if clinical disease progression (such as adenomas, infiltrative or malignant disorders, hormonal abnormalities, neuro-/ophthalmological symptoms)

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Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (HD-19.4)

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- See **Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (PEDONC-4.10)**

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Evidence Discussion (HD-19)

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- MRI imaging of the Sella region using high-resolution pituitary protocols is the preferred diagnostic imaging modality for evaluation of the pituitary and sellar regions and is considered the gold standard for imaging the pituitary gland when there is suspicion of hypothalamic pituitary disease.[8]
- Both the anatomy and pathology of the pituitary gland and surrounding areas including optic chiasm, infundibulum, and vascular structures, as well as an empty sella, are reliably depicted on MRI.[8]
- MRI is the most sensitive imaging study for evaluating pituitary disease.[8]
- CT of the Sella can be used to detect bone destructive lesions of the skull base, such as craniopharyngiomas, menigiomas, or larger pituitary macroadenomas, but CT is insensitive when compared to MRI for pituitary pathology.[8]
- MRI utilizes a magnetic field and radio waves with computer processing to produce detailed images whereas CT uses ionizing radiation. Radiation dosages vary based on many factors and can be harmful to tissues. Thus, from radiation safety perspective MRI should be utilized when appropriate and supported by existing literature.[27]

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Scalp and Skull (HD-20)

Guideline

Scalp and Skull Lesions (HD-20.1)
Skull Base Osteomyelitis (SBO) (HD-20.2)
References (HD-20)

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Scalp and Skull Lesions (HD-20.1)

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The majority of these are benign soft tissue or bony lesions easily defined by physical examination or with skull x-rays or ultrasound.

- Ultrasound is the initial imaging of scalp lesions
- X-ray is the initial imaging of skull (bony) lesions
- CT Head without or without and with contrast (CPT® 70450 or CPT® 70470) is medically necessary for the following scenarios:
  - Any lesion on physician examination and skull x-ray or ultrasound which is not clearly benign.
  - In cases where surgical planning is in progress, x-rays and/or ultrasound are not required.
  - When bony lesions are detected on physical examination with any of the following:
    - Signs or symptoms of Langerhan's cell histiocytosis
    - Signs or symptoms of multiple myeloma
    - History of a cancer condition with a suspicion of metastasis
    - History of Paget's disease
    - History of radiation therapy to the head region
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary if there is concern for intracranial extension.
- See **Dental/Periodontal/Maxillofacial Imaging (HD-30.2)** for mandibular masses
- The following imaging is medically necessary for children and adults with Pott Puffy Tumor:
  - MRI Brain without and with contrast (CPT®70553) or CT Head without and with contrast (CPT® 70470)
  - Repeat imaging is medically necessary if requested by a neurologist, neurosurgeon, otolaryngologist (ENT) and/or oromaxillofacial surgeon (OMS) or any provider coordinating care with a neurologist, neurosurgeon, otolaryngologist (ENT) and/or oromaxillofacial surgeon (OMS)

Background and Supporting Information

Pott Puffy Tumor is an abscess involving the frontal bone with adjacent osteomyelitis as the result of a frontal sinus infection that spreads contiguously through the wall of the sinus or through hematogenous spread via the veins that drain sinus mucosa.

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Evidence Discussion (HD-20.1)

- The ACR Practice Parameter or the Performance of Computed Tomography (CT) of the head and of the extracranial head and neck gives a broad description of some of the pathologies that would be beneficially imaged using this modality, and points out that the reason for imaging must be for a valid medical reason and should be done with the aim for using only the minimum necessary radiation. This in some cases requires the use of additional modalities.[1,7]
- The majority of skull lesions is benign, but advanced imaging characteristics may aid in defining the lesion as having a relatively high pretest probability of malignancy (prior to histological confirmation). However, clinical contextual information is necessary to help decide which individuals would benefit from advanced imaging including the individual's age and features of the individual's presenting history.[6]
- Ultrasound offers many radiographic advantages for the characterization of scalp masses, which are not visualized by CT or MRI, the primary goal being to differentiate benign vs. malignant scalp masses. Cancers of the scalp represent 2% of all skin cancers. In both squamous and basal cell carcinomas, the US shows hypoechoic solid tumors with increased vascularity, and basal cell pathology can also consist of hyperechoic spots internally.[8]
- In folliculotropic mycosis fungoides (FMF), the most common manifestation of cutaneous T-cell lymphoma, the sonographic features include skin thickening, and hypoechoic upper dermis and hair follicles, with large surrounding hyperechoic deposits.[8]
- Skull (bony) lesions are most often discovered incidentally either clinically or as a result of CT or MRI of the brain performed for another indication, and these skull masses can be either malignant or benign. The individual's history is essential to understand along with the imaging characteristics in order to obtain accurate diagnosis. Radiographic features, both CT- and MRI- specific can be used to differentiate between benign and malignant lesions, identifying whether lesions have well-defined borders, sclerotic margins, and a narrow transition zone.
- The presence of bony destruction is a useful observation, periosteal reaction, soft tissue component, and intracranial or extracranial extension can be identified as malignant features in addition, and patterns such as lytic vs. sclerotic, dingle vs multiple, homogeneous vs varied composition also can give helpful information for diagnosis, and various patterns are recognizable that may support the tissue type of origin such as fibrogenic, chondrogenic, osteogenic, vascular, etc.[6]
- Plain radiograph can identify some of these features as a first diagnostic study, but advanced imaging may be necessary, in conjunction with x-ray, and at times CT and MRI are useful as complementary studies.[9]
- Pott puffy tumor is a rare complication of sinusitis or trauma, and early diagnosis is important since it is treatable with broad spectrum antibiotics, therefore advanced imaging is medically necessary with a clinical suspicion.[10]

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Skull Base Osteomyelitis (SBO) (HD-20.2)

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- Note: SBO may occur from the temporal bones or paranasal sinuses and imaging should be of the region of origin
- Any of the following imaging studies are medically necessary in the diagnosis and treatment of skull base osteomyelitis and/or necrotizing external otitis:
  - MRI Brain without and with contrast (with IAC views) (CPT® 70553)
  - CT Temporal bone without contrast (CPT® 70480)
  - CT Temporal bone with contrast (CPT® 70481)
  - CT Maxillofacial without contrast (CPT® 70486)
  - CT Maxillofacial with contrast (CPT® 70487)
  - CT Neck with contrast (CPT® 70491)
  - CTA/CTV Head (CPT® 70496)
  - MRA/MRV Head, contrast as requested (CPT® 70544, 70545, 70546)
  - Gallium-67 Scan (CPT® 78800 or 78801, and 78803, 78831, 78830 or 78832)
  - Bone Scan (CPT® 78830 or 78832)
    - Skull base osteomyelitis: + Gallium and + Bone scan
    - Necrotizing otitis externa: + Gallium and - Bone scan
  - Indium WBC (CPT® 78800 or 78801, and 78803, 78831, 78830 or 78832) is medically necessary as a substitute for or used in addition to Gallium scanning to evaluate response to therapy and especially in cases that have undergone surgical debridement.
- Treatment response: Gallium-67 Scan every 4-6 weeks till scan is negative
- Surveillance Scanning: Gallium-67 Scan at 4 weeks and 3 months post-treatment
- For CT Head imaging, see General Guidelines - CT Head (HD-1.4) and/or Headache (HD-11)
- Internal Auditory Canal: (IAC) MRI can be reported as a limited study with one code from the set (CPT® 70540, CPT® 70542, or CPT® 70543), but should not be used in conjunction with MRI Brain codes (CPT® 70551, CPT® 70552, or CPT® 70553) if IAC views are performed as part of the brain.

Background and Supporting Information

Skull base osteomyelitis is a rare complication of otitis externa. It occurs most commonly among the immunocompromised, older members (greater than 65 years of age) and members with diabetes.

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MRI Brain will be positive earliest in the disease process. CT scans will best define bony destruction and are positive later in the disease process.

Evidence Discussion (HD-20.2)

- CT, although involves radiation, is more readily available and provides superior information regarding bony erosion and/or demineralization in the individual with suspected osteomyelitis.[5]
- MRI can assist with early detection of bone changes in as early as 3-5 days from onset of osteomyelitis.[5] In cases of diabetic osteomyelitis MRI provides a sensitivity of 90% and a specificity of 79%. MRI provides superior soft tissue detail and intracranial involvement secondary to its superior resolution when compared to CT for evaluation of skull-base osteomyelitis.[5,11,12]
- Both nuclear imaging by means of Technetium 99m (^{99m}Tc) and Gallium 67 (^{67}Ga) scan can assist in localizing infection. The Gallium scan is often used to determine the resolution of the infection and thus the end of antibiotic therapy.[11,12] The Technetium 99m scan can be useful for detecting the infection however often times remains positive prolonged period of time, and thus should not be used to determine resolution of infection.[12]

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Cerebrovascular Disease (HD-21)

Guideline

Stroke/TIA (HD-21.1)
Risk Assessment for Extracranial Carotid Disease (HD-21.2)
Cryptogenic Stroke (HD-21.3)
Transient Global Amnesia (HD-21.4)
Moyamoya Syndrome/Disease (HD-21.5)
Sickle Cell Disease (HD-21.6)
Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations (HD-21.7)
Cerebral Venous Sinus Thrombosis (HD-21.8)
Evidence Discussion (HD-21)
References (HD-21)

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Stroke/TIA (HD-21.1)

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Indications	Medically Necessary Imaging
• Acute ischemic stroke • Transient ischemic attacks (TIA) • Suspected hemorrhagic stroke • Suspected subdural hemorrhage	**ANY** or **ALL** of the following: • CT Head without contrast (CPT® 70450) • CTA Head (CPT® 70496) • CTA Neck (CPT® 70498) • CT Perfusion (CPT® 0042T) • MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549
Repeat imaging for follow-up and resolution of stroke or hemorrhage	• As requested by a neurologist, neurosurgeon, hematologist, or physiatrist (PM&R), or any provider in consultation with a neurologist, neurosurgeon, hematologist, or physiatrist
Contraindication to MRI	• CT Head without contrast (CPT® 70450) **OR** • CT Head without and with contrast (CPT® 70470)
Arterial Vascular Imaging supported for TIA/Stroke evaluation including dissection: • Supported concurrently with brain imaging	• MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR** • CTA Neck (CPT® 70498)

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Indications	Medically Necessary Imaging
Venous vascular imaging for evaluation of venous infarcts	• MRA/MRV ([CPT® 70544, CPT® 70545, or CPT® 70546]) **OR** • CTA/CTV Head (CPT® 70496) **AND/OR** • MRI Brain without contrast (CPT® 70551) • MRI Brain without and with contrast (CPT® 70553) **OR** • CT Head without contrast (CPT® 70450) Also see **Cerebral Venous Sinus Thrombosis (HD-21.8)**
Cervicocerebral Catheter Angiography for stroke evaluation	• 3D Rendering (CPT® 76377 or CPT® 76376)
Stroke in Pregnancy and other hypercoagulable states • See arterial and venous vascular imaging studies above for vascular imaging requests • See *Background and Supporting Information*	• MRI Brain without contrast (CPT® 70551) **OR** • CT Head without contrast (CPT® 70450)
Amaurosis Fugax or Ocular Microembolism • May include optic nerve/retinal arterial or Hollenhorst plaques on exam	See above for TIA or New Stroke brain imaging options and vascular imaging
Reversible Cerebral Vasoconstriction Syndrome	See also **Sudden Onset of Headache (HD-11.3)**

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Indications	Medically Necessary Imaging
Neurologic signs and/or symptoms, including headaches, associated with COVID-19 infection and/or COVID-19 vaccination (Ischemia may be arterial or venous)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) See also **General Guidelines-CT head (HD-1.4)**, **Abnormal Blood Clotting (HD-11.9),** and **Neuro-Covid-19 (HD-14.2)**, and **Cerebral Venous Sinus Thrombosis (HD-21.8)**
Adults with HbSS (Sickle cell disease) or HbSb Thalassemia	One time MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) for screening to detect silent cerebral infarcts Follow-up or repeat testing per neurologist or hematologist or in consultation with a neurologist or hematologist See also **Sickle Cell Disease (HD-21.6)**
Secondary work-up of stroke/TIA	Transcranial Doppler Studies See **Transcranial Doppler CPT® 93886 (HD-24.8)**
Moyamoya Disease when surgery or other vascular intervention is being considered	See **Moyamoya Syndrome/Disease (HD-21.5)**
Evaluation of paradoxical venous thromboembolism in cryptogenic stroke with PFO	See **Acute deep venous thrombosis (DVT) (PVD-12.2)** and **Cryptogenic Stroke (HD-21.3)**

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Indications	Medically Necessary Imaging
Cerebral Amyloid Angiopathy (CAA)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • CT Head without contrast (CPT® 70450) Amyloid-PET Brain (CPT® 78811 or CPT® 78814) is considered not medically necessary for stroke evaluation. See **Mild Cognitive Impairment (MCI) and Dementia - PET (HD-8.2)**
Multisystem Smooth Muscle Syndrome/ Smooth Muscle Dysfunction Syndrome	See **Multisystem Smooth Muscle Syndrome/Smooth Muscle Dysfunction Syndrome (HD-21.7)**

Background and Supporting Information

- Pregnancy is an independent risk factor for stroke. Additional risk factors are not required for assessment of a stroke/TIA with acute focal neurological deficits.
- Additional arterial and venous hypercoagulable states that impose a stroke risk include:
  - Antiphospholipid syndrome
  - Hyperhomocysteinemia
  - Factor V Leiden mutation
  - Prothrombin gene mutation
  - Protein S deficiency
  - Protein C deficiency
  - Anti-thrombin deficiency

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Risk Assessment for Extracranial Carotid Disease (HD-21.2)

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- Duplex Ultrasound Carotid Arteries (CPT® 93880 or CPT® 93882) is medically necessary for the following:
  - Asymptomatic or symptomatic cervical bruits;
  - Clinical suspicion of extracranial carotid occlusion and the rationale is included
  - Pulsatile neck mass
  - Evaluation of blunt or penetrating neck trauma
  - Amaurosis fugax or ocular microembolism (optic nerve/retinal arterial or Hollenhorst plaques seen on exam)
  - Recent history of focal cerebral or ocular transient ischemic attacks
- Follow-up with CTA or contrast enhanced MRA
  - CTA and contrast enhanced MRA are comparable non-invasive imaging alternatives each with their own advantages and disadvantages
- For additional indications for Duplex Ultrasound Carotid Arteries
  - See **Initial Imaging (PVD-3.1)** in the Peripheral Vascular Disease (PVD) Imaging Guidelines
- For repeat (Surveillance) Duplex Ultrasound Carotid Arteries (CPT® 93880 or CPT® 93882)
  - See **Surveillance Imaging with NO History of Carotid Surgery or Intervention (PVD-3.2)** and **Surveillance Imaging WITH History of Carotid Surgery or Intervention (PVD-3.3)** in the Peripheral Vascular Disease (PVD) Imaging Guidelines.

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Cryptogenic Stroke (HD-21.3)

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- 25% of individuals with ischemic stroke have no probable cause and is considered cryptogenic after a standard workup including an echocardiogram, inpatient cardiac telemetry or 24-Holter monitoring, CT or MRI Brain and vessel imaging of the brain or neck arteries and hematologic tests.
- A stroke may also be considered cryptogenic after a standard evaluation fails to yield an etiology in an individual <50 years of age without risk factors with more extensive testing.
- Most cryptogenic sources are embolic in etiology from venous or arterial sources with investigations from disturbances in coagulation and sources of embolism including patent foramen ovale (PFO) and paroxysmal atrial fibrillation.
- Specialized evaluation with the following documentation:
  - MRI/CT Brain with results of stroke
  - Results of MRA/CTA Head and Neck
  - TTE or TEE
  - 24-Hr Holter monitor or Inpatient cardiac telemetry and 12-Lead ECG
- Hematologic testing to include: CBC, Platelet count, INR, PT, PTT, D-Dimer and Arterial and Venous Hypercoagulability tests
  - MRA or CTA Pelvis for the evaluation of paradoxical venous thromboembolism with PFO
    - See **Acute Limb Swelling (PVD-12)** in the Peripheral Vascular Disease (PVD) Imaging Guidelines.
  - Work-up for occult cancer, CT Chest, Abdomen, and/or Pelvis with contrast after the previously indicated tests with results are provided.
    - See **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.

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Transient Global Amnesia (HD-21.4)

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- Transient Global Amnesia (TGA) is a clinical diagnosis with the differential diagnosis including, but not exclusive to: ischemic events, migraine headaches, and transient epileptic amnesia.
- Characteristics of TGA may include the following:
  - Inability to retain new information, lasting for several hours with preservation of alertness and all other cognitive functions with repetitive queries and amnesia
  - Witnessed episode
  - There must be anterograde amnesia during the attack
  - Cognitive impairment is limited to amnesia
  - No clouding of consciousness or loss of personal identity
  - No focal neurological signs/symptoms
  - No epileptic features
  - Attack must resolve within 24 hours
  - No recent head injury or active epilepsy
- Head and vessel imaging for ischemic etiology work-up should follow **Stroke/TIA (HD-21.1)**
- For suspected seizure, see **Epilepsy/Seizures (HD-9.1)**

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Moyamoya Syndrome/Disease (HD-21.5)

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Initial imaging for Moyamoya Syndrome/Disease

- The imaging listed below is medically necessary for initial evaluation of Moyamoya Syndrome/Disease:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548 **OR** CPT® 70549)
    - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) is/are medically necessary

Repeat imaging for Moyamoya Syndrome/Disease

- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548 or CPT® 70549)
  - Follow-up or repeat testing per neurologist, neurosurgeon, hematologist or in consultation with a neurologist, neurosurgeon, or hematologist.
  - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) AND/OR CTA Neck (CPT® 70498) is/are medically necessary
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553)
  - Follow-up or repeat testing per neurologist, neurosurgeon, hematologist or in consultation with a neurologist, neurosurgeon, or hematologist.
- Radiopharmaceutical Localization Imaging SPECT (CPT® 78803, CPT® 78830 or CPT® 78832) with vasodilating agent acetazolamide (Diamox) challenge is supported when surgery or other vascular intervention is considered. Follow-up or repeat testing per neurologist, neurosurgeon, hematologist or in consultation with a neurologist, neurosurgeon, or hematologist.
- 3D Rendering (CPT® 76377 or CPT® 76376) with cerebral angiography is medically necessary to define the presence, location, and anatomy of intracranial and cervical vascular malformations.
  - See **General Guidelines – Other Imaging Situations (HD-1.7)** and in the Preface Imaging Guidelines
- CT Perfusion (CPT® 0042T) **OR** MRI Perfusion (CPT® 70551 OR CPT® 70552 OR CPT® 70553) is medically necessary for any of the following:
  - When requested by neurologist and/or neurosurgeon

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- Prior to change in treatment
- Post-surgical

Screening imaging for Moyamoya Disease

- Screening is not medically necessary for Moyamoya Syndrome.
  - See *Background and Supporting Information*
- Screening for Moyamoya Disease is medically necessary for:
  - First-degree relatives (biological parent, full sibling, or biological child) of individuals with Moyamoya Disease when requested by, or any provider in consultation with, a neurologist, geneticist, or neurosurgeon
- The imaging listed below is medically necessary for screening evaluation of Moyamoya Disease:
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** Transcranial Doppler (TCD) Ultrasound (CPT® 93886 or CPT® 93888) (see **Transcranial Doppler (CPT® 93886) (HD-24.8)** in the Head Imaging Guidelines)
  - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) is medically necessary

CT Perfusion (CPT® 0042T)

- Is medically necessary if requested by a neurologist, neurosurgeon or any provider coordinating care with a neurologist or neurosurgeon.

MRI Perfusion

- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 OR CPT® 70552 OR CPT® 70553)
  - No additional CPT® codes are medically necessary or appropriate to perform MRI perfusion.

Background and Supporting Information

Moyamoya disease (MMD) is a rare cerebrovascular disease characterized by progressive spontaneous bilateral occlusion of the intracranial internal carotid arteries (ICA) and their major branches (middle cerebral artery, MCA, and anterior cerebral artery, ACA) with compensatory capillary collaterals as an expression of pathologically increased angiogenic activity resembling a "puff of smoke" (Japanese: Moyamoya) on cerebral angiography. Moyamoya Disease is most prevalent individuals with East Asian ancestry. Up to 15% of individuals with Moyamoya Disease may have a family history of Moyamoya Disease.

Moyamoya Disease is distinguished from Moyamoya Syndrome (MMS). MMD is a primary disease process. MMS is a secondary process that occurs in response to

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another underlying pathological process that causes stenosis of intracranial blood vessels. There are two peaks of incidence with different clinical presentations, at around 10 years and 30-40 years. The peak appears to occur later in women than men. In children, ischemic symptoms, especially transient ischemic attacks, are predominant. Intellectual decline, seizures, and involuntary movements are also more common in this age group. In contrast, adult individuals present with intracranial hemorrhage more often than pediatric individuals.

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Sickle Cell Disease (HD-21.6)

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for the following:
  - Screening to detect silent cerebral infarcts
  - New symptoms or cognitive impairment occurs or a change in academic performance
  - Prior to any change in therapy
- MRA Head (CPT® 70544, CPT® 70545 OR CPT® 70546) **OR** CTA Head (CPT® 70496) is medically necessary for the following:
  - Any new, indeterminate, or equivocal findings on MRI Brain
  - Prior to any change in therapy

Background and Supporting Information

Individuals with sickle cell disease are at significantly increased risk for stroke and silent infarction, beginning at a very young age. Recent advances allow physicians to identify individuals at high risk for stroke and begin a primary stroke prevention program.

Identification of silent cerebral infarction is important because treatment with prophylactic red cell transfusions to maintain hemoglobin S levels at <30% of total hemoglobin may reduce recurrent stroke and extent of neurologic damage.

- TCD for children aged 17 years old may be medically necessary on a case-by-case basis.
- See **Transcranial Doppler (CPT® 93886)(HD-24.8)** in the Head Imaging Guidelines for other indications for this modality and **Stroke/TIA (HD-21.1)** in the Head Imaging Guidelines.
- After 17 years old, for individuals with a history of abnormal TCDs, TCDs may be repeated every 3 months.
- TCD is not medically necessary for individuals with other phenotypes (Hgb SC, Hgb $S\beta^+$).

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Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations (HD-21.7)

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Indications	Medically Necessary Imaging
Initial evaluation for confirmed ACTA2 mutation	• MRI Brain without and with contrast (CPT® 70553) with OR without MRI perfusion **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549)
Repeat imaging if requested by neurologist and/or neurosurgeon and/or geneticist and/or provider coordinating care with a neurologist and/or neurosurgeon and/or geneticist	• MRI Brain without and with contrast (CPT® 70553) with OR without MRI perfusion **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549)

- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 OR CPT® 70552 OR CPT® 70553)

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  - No additional CPT® codes are medically necessary to perform MRI perfusion.
- Because radiation is a known risk factor for development of moyamoya, MRI/MRA Head is recommended instead of Computed Tomography (CT)/CTA.
  - See *Background and Supporting Information*
- Conventional catheter angiogram 3D rendering (CPT® 76376 or CPT® 76377) should be reserved for individuals with focal neurologic symptoms or evidence on MRA or transcranial Doppler (TCD) of critical or progressive narrowing of the cerebral arteries.
  - See **Screening for Suspected Peripheral Artery Disease/Aneurysmal Disease (PVD-2)**

Background and Supporting Information

Smooth muscle dysfunction syndrome (SMDS)/Multisystemic Smooth Muscle Syndrome (MSMS) presents with a recognizable pattern of complications, including congenital mydriasis, patent ductus arteriosus (PDA), pulmonary arterial hypertension, aortic and other arterial aneurysms, moyamoya-like cerebrovascular disease, intestinal hypoperistalsis and malrotation, and hypotonic bladder.

SMDS/MSMS is caused by heterozygous mutations of the ACTA2 altering arginine 179, most commonly p.Arg179His. With a single exception, all cases are due to de novo mutations.

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Cerebral Venous Sinus Thrombosis (HD-21.8)

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CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography):

- If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code is used to report both procedures.
- If an arterial CTA or MRA study has been performed and subsequently a repeat study is needed to evaluate the venous anatomy, then this study is medically necessary.
- If a venous CTV or MRV study has been performed and subsequently a repeat study is needed to evaluate the arterial anatomy, then this study is medically necessary.

Indications	Medically Necessary Imaging
Venous vascular imaging for evaluation of venous infarcts	MR or CT Venography: • MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** • CTV Head (CPT® 70496)
Suspected cerebral venous thrombosis	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • CT Head with contrast CPT® 70450 **AND/OR** • MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** • CTV Head (CPT® 70496)
Follow up of known cerebral venous sinus thrombosis	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • CT Head without contrast (CPT® 70450) **AND/OR** • MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** • CTV Head (CPT® 70470)

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Indications	Medically Necessary Imaging
To differentiate total from subtotal occlusion	MRA without and with contrast (CPT® 70546)

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Evidence Discussion (HD-21)

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- Guidelines from the American Heart Association (AHA) and American Stroke Association supported the role of neuroimaging in stroke triage and individual selection for endovascular therapy in the management of acute stroke.[1,6,55] Current AHA guidelines also recommended non-invasive imaging of the carotid arteries for individuals with TIA or stroke who may be candidates for carotid endarterectomy or stenting.[1,6] This includes CT Angiography (CTA) of the head and neck or MR Angiography (MRA) of the head and neck.
- For clinically suspected stroke, initial imaging includes CT head to exclude intracranial hemorrhage, exclude other structural causes and assess for early ischemic changes.[1] CT Angiography (CTA) head is medically necessary during the initial evaluation to assess for large vessel occlusion and has high sensitivity of 93% and specificity of 100%.[1,54] CTA neck is also a rapid modality for imaging the extracranial vasculature to identify carotid stenosis, occlusion, and vertebral-basilar disease.[1]
- CT Perfusion (CTP) can identify individuals with large vessel occlusion who may be candidates for endovascular therapy in the acute stroke setting.
- A CT stroke protocol that includes unenhanced CT, CTA head, and CTP has effective radiation doses between 10-15 mSvm, with newer generation scanners and optimized sequences with lower radiation doses closer to 2 mSv.[5]
- MRI Brain with Diffusion Weighted Imaging (DWI) is the most sensitive test to detect an acute ischemic infarct, with a sensitivity of 94% and a specificity of 97%.[1,6,56,57]
- MR Angiography (MRA) Head and Neck is an alternative to CTA for identification of vascular lesions in the setting of a stroke evaluation.[1] Diagnostic cervicocerebral catheter angiography has the highest spatial and temporal resolution of any vascular imaging study, however, is invasive and requires use of contrast.[1]
- Compared with CT, a full stroke protocol with MRI is longer to acquire and susceptible to motion artifacts, in addition to contraindication with metallic devices, and certain implants.[55] MRI, however, does have the advantage of increased sensitivity for acute ischemia, including in transient ischemic attack, and does not require radiation.
- In the delayed stroke evaluation, CT Head may identify complications such as hemorrhagic conversion, mass effect and herniation.[55] MRI Brain in this scenario can confirm the extent of an ischemic stroke, evaluate for underlying pathology, and identify any complications.[55]
- For clinically suspected transient ischemic attack (TIA), CT Head is useful to exclude hemorrhage and other intracranial abnormalities.[1] CT perfusion can identify abnormalities in the setting of TIA in up to one-third of cases.[1] CT Angiography (CTA) head and neck is a rapid modality for evaluating intracranial and extracranial vascular lesions.[1] MRA Head and neck is an alternative modality, preferred in those with renal

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impairment and iodine contrast allergy.[1] MRI Brain is the most sensitive modality for acute ischemic infarct.[1] MRI Brain with Diffusion Weighted Imaging (DWI) sequences can identify ischemic changes in approximately 40% of individuals with TIA.[58]

- For clinically suspected venous sinus thrombosis, imaging is medically necessary to identify the clot and assess for complications, such as venous infarction or hemorrhagic transformation. In addition to imaging previously reviewed for the stroke protocol, CT Venogram (CTV) or MR Venogram (MRV) are appropriate to localize the clot within the venous system.[1]
- For stroke or hemorrhage related to Cerebral amyloid angiopathy (CAA), Amyloid PET Brain has a sensitivity that ranges from 82% to 91%, however, its specificity is poor, ranging from 44% to 55%; therefore, this modality is not recommended.[33]

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Cerebral Vasculitis (HD-22)

Guideline

Cerebral Vasculitis (HD-22.1)
References (HD-22)

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Cerebral Vasculitis (HD-22.1)

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- When CNS vasculitis is suspected, MRI Brain without and with contrast (CPT® 70553) is medically necessary
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549); **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - CTA Neck (CPT® 70498) are medically necessary concurrently with brain imaging
- Primary CNS vasculitis includes Giant Cell Arteritis also known as Temporal Arteritis (see **New Headache Onset Older than Age 50 (HD-11.7)**)
- If initial vascular imaging is suspicious for vasculitis, 3D rendering (CPT® 76377 or CPT® 76376) with cervicocerebral angiography/arteriography (see **General Guidelines- Other Imaging Situations (HD-1.7)**)
- Transcranial Doppler Studies for individuals with documented vasculitis or concern for vasospasm (see **Transcranial Doppler (CPT® 93886) (HD-24.8**))
- FDG-PET/CT Brain (CPT® 78608) is not supported due to lack of peer reviewed literature or expert consensus supporting the study for vasculitis.
- For extra-cranial giant cell arteritis evaluation (see **Giant Cell Arteritis (PVD-6.9.2))**

Background and Supporting Information

The diagnosis of primary central nervous system vasculitis is challenging because of its nonspecific and varied symptoms. Central nervous system vasculitis typically presents with headache, followed by encephalopathy and behavioral changes. Focal neurologic deficits, including but not limited to, visual loss, unilateral weakness, language impairment, sensory loss, incoordination, occurs in 20% to 30% of individuals. Seizures and intracranial hemorrhage may also occur. With a strong clinical suspicion, brain imaging is important for supporting the diagnostic process and directing biopsy.

Classification of vasculitides based on vessel size adapted from Younger. MRA and CTA are useful for the evaluation of the large proximal arteries; evaluation of a possible small vessel vasculitis may be beyond the resolution of routine MRA and CTA Head. However, other abnormalities, such as atherosclerotic disease, arterial dissection, Moyamoya disease, or reversible cerebral vasoconstriction may be demonstrated. Conventional angiogram is superior to MRA and CTA in demonstrating abnormalities in smaller vessels and is considered the "gold standard" in the evaluation of primary small vessel CNS vasculitis.

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Dominant Vessel Involved	Primary	Secondary
Large arteries	• Giant cell arteritis • Takayasu's arteritis	Aortitis with rheumatoid disease; Infection (e.g., syphilis)
Medium arteries	• Classical polyarteritis nodosa • Kawasaki disease	Infection (e.g., hepatitis B)
Small vessels and medium arteries	• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis	Vasculitis with rheumatoid disease, systemic lupus erythematosus (lupus cerebritis), Sjögren's syndrome, drugs, infection (e.g., HIV)
Small vessels	• Henoch-Schönlein purpura • Essential cryoglobulinemia • Cutaneous leukocytoclastic vasculitis	Drugs (e.g., sulphonamides, etc.) Infection (e.g., hepatitis C)

Evidence Discussion (HD-22)

- Non-invasive neuroimaging modalities play a role in the diagnostic evaluation of central nervous vasculitis by providing supportive imaging findings and guiding biopsy.[3,6] The preferred modality for the evaluation of central nervous system vasculitis is MRI, which provides superior soft-tissue resolution.[6] MRI Brain is abnormal in >95% of individuals with CNS vasculitis.[8] MRI Brain shows infarcts in up to 50% of cases and white matter hyperintensities in 42% of cases.[3,6]
- MRA head was found to be abnormal in 81% of individuals with angiographic findings of vasculitis and normal in 100% of individuals with a normal angiogram.[6]
- CT Angiography is an alternative non-invasive modality that also provides visualization of blood vessels.[3,6,8]
- FDG-PET/CT brain is not supported due to the high physiologic FDG uptake in the brain and limited resolution of the camera system. Atherosclerosis may also interfere with the FDG-PET interpretation.[6]

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Dizziness, Vertigo and Syncope (HD-23)

Guideline

Dizziness/Vertigo (HD-23.1)
Syncope (HD-23.2)
References (HD-23)

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Dizziness/Vertigo (HD-23.1)

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Indications	Medically Necessary Imaging
Red Flags: • History of malignancy • Associated symptoms: ◦ Headache ◦ Hearing loss ◦ Unilateral tinnitus ◦ Visual disturbances ◦ Drop attacks ◦ Vestibular migraine ◦ Weakness • Duration of episode: ◦ Episodes lasting hour(s) or continuous • Exam findings: ◦ Inconclusive positional testing or equivocal or unusual nystagmus findings (Negative Dix-Hallpike) ◦ Visual disturbances including loss and diplopia ◦ Hearing loss ◦ Abnormal cranial nerve findings ◦ Ataxia ◦ Positive Romberg sign ◦ Absent head thrust sign ◦ Focal neurologic deficits ◦ Dysarthria ◦ Weakness, including unilateral or hemibody weakness • Failed treatment: ◦ Failure to respond to vestibular therapy or unable to participate due to clinical condition • Abnormal test results: ◦ ENG/VNG results support central cause	• MRI Brain without contrast (CPT 70551) **OR** • MRI Brain without and with contrast (CPT 70553) **OR** • CT Head without contrast (CPT 70450) If MRI contraindicated: • CT Head without contrast (CPT 70450) **OR** • CT Head without and with contrast (CPT 70470) See also: • **Headaches with Red Flags (HD-11.2)** • **Multiple Sclerosis and Related Conditions (HD-16)** • **Brain Metastases (ONC-31.3)**

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Indications	Medically Necessary Imaging
Stroke/TIA	See **Stroke/TIA (HD-21.1)**
Acoustic Neuroma/Vestibular Schwannoma	• MRI Brain without and with contrast (with IAC views) (CPT® 70553) **OR** without contrast (CPT® 70551) • Limited MRI of the internal auditory canals (CPT® 70540, CPT® 70542, OR CPT® 70543) when requested by the provider in place of a complete MRI Brain See also • **Acoustic Neuroma (HD-33.1)** • **Peripheral Nerve Sheath Tumors (PN-9.1)**
Head trauma/Temporal Bone Fracture/ Post-traumatic vertigo	• CT Head without contrast (CPT® 70450) ◦ See **Head Trauma (HD-13.1)** **AND/OR** • CT Orbit/Temporal bone without contrast (CPT® 70480)

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Indications	Medically Necessary Imaging
Vertebrobasilar disease/ Vertebrobasilar Insufficiency/ Dissection	• CTA Head (CPT® 70496 **AND/OR** • CTA Neck (CPT® 70498) **OR** • MRA Head (CPT® 70544, CPT® 70545, OR CPT® 70546) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT®70549) See also: • **General Guidelines - CT and MR Angiography (CTA/ CTV and MRA/MRV) (HD-1.5)** • **Headache and Suspected Vascular Dissection (HD-11.1)** • **Intracranial Cerebral Aneurysms (HD-12.1)**
Semicircular canal dehiscence	• CT Orbit/Temporal bone without contrast (CPT® 70480)

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Indications	Medically Necessary Imaging
Meniere's Disease	• MRI Brain without and with contrast (with IAC views) (CPT® 70553) **OR** without contrast (CPT® 70551) • Limited MRI of the internal auditory canals (CPT® 70540, CPT® 70542, OR CPT® 70543) when requested by the provider in place of a complete MRI Brain

Background and Supporting Information

- Dizziness, a common complaint, with benign and dangerous causes, may be continuous, triggered, or spontaneous.
- For the continuously dizzy individual with nystagmus at the time of evaluation, a head impulse test and a test of skew should be performed to determine if dizziness is due to a peripheral cause or a posterior circulation stroke. Abnormalities on exam may be indications for imaging as detailed in the guideline.
- For triggered dizziness, positional testing such as the Dix-Hallpike maneuver, and/or orthostatic blood pressure measurements, should be performed. If symptoms are reproduced on examination, triggered dizziness is confirmed.
- Spontaneous dizziness may be due to vestibular migraine, TIA, or Meniere's disease, among other causes. A detailed neurologic examination should be performed, and imaging as detailed in the guideline.
- The Dix-Hallpike maneuver should be performed or the individual should be referred to a clinician who could perform the procedure if Benign Paroxysmal Positional Vertigo (BPPV) is suspected.
- The Head Impulse Test (HIT) is also known as the Head thrust test. It is designed to evaluate the vestibular-ocular reflex in an individual with concern for a peripheral vestibulopathy due to ACUTE spontaneous vertigo. The individual is instructed to look at the examiner during the entire test. The individual's head is then quickly turned or rotated to one side and then the other. If normal, the individual's eyes should remain locked on the examiner. If abnormal, the eyes will move in the direction of the head rotation and then quickly correct. This saccade indicates peripheral vestibular hypofunction on the side of the direction that the head is turned. The HIT test is

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abnormal in individuals with vestibular neuronitis, and normal in individuals with a posterior circulation stroke.

- Posterior Canal BPPV (85%-95% of BPPV cases) is defined as:
  - Individual reports repeated episodes of vertigo with changes in head position relative to gravity.
  - Each of the following criteria is fulfilled on physical exam:
    - Vertigo associated with torsional (rotatory), upbeating (toward the forehead) nystagmus is provoked by the Dix-Hallpike test.
    - There is a latency period between the completion of the Dix-Hallpike maneuver and the onset of vertigo and nystagmus.
    - The provoked vertigo and nystagmus increase and then resolve within 60 seconds from the onset of the nystagmus.
- Lateral or Horizontal Canal BPPV (5%-15% of BPPV cases) will have horizontal or no nystagmus to which a supine roll test assesses for this condition.
- Exclusions for Dix-Hallpike maneuver
  - Individual previously diagnosed with BPPV and who on date of encounter in calendar year does not have positional dizziness or vertigo consistent with active BPPV
  - Individual has declined Dix-Hallpike maneuver
  - Individual has cervical spinal disease (i.e., cervical stenosis, severe kyphoscoliosis, limited cervical range of motion, Down's syndrome, severe rheumatoid arthritis, cervical radiculopathies, Paget's disease, ankylosing spondylitis, low back dysfunction, spinal cord injuries, spinal fractures)
  - Individual unable to lay flat (i.e., severe heart disease)
  - Individual has severe atherosclerotic disease or recent dissection involving the anterior or posterior cerebral circulation
  - Unable to be seated in exam chair (i.e., morbidly obese), or maneuver cannot be safely performed given morbid obesity
  - Ehlers Danlos/Marfans/Connective tissue disorder due to risk of cranio spinal instability/dissection
- Triggered episodic vestibular syndrome (t-EVS) usually last seconds to minutes with the most common triggers (vs. exacerbating factors) are head motion or change in body position. In the Emergency Department, benign paroxysmal positional vertigo (BPPV) is the second most common cause of t-EVS after orthostatic hypotension. Far lateral rotation of the neck leads to mechanical occlusion of one or both vertebral arteries causing temporary symptoms of vertigo and nystagmus when this position is maintained and may occur with the individual upright.
- Diagnoses or conditions associated with orthostatic hypotension (OH) or nOH include: Parkinson Disease (PD), Multiple System Atrophy (MSA), Pure Autonomic

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Failure (PAF) or Dementia with Lewy Bodies (DLB), unexplained fall or syncope, peripheral neuropathies secondary to diabetes, amyloidosis and HIV), individuals ≥70 years of age and frail and on multiple medications and individuals with postural (orthostatic) dizziness or nonspecific symptoms that occur when standing. Symptoms may include: lightheadedness or dizziness, the sensation of blacking out, cognitive dysfunction, mental dulling, generalized weakness, neck pain or discomfort in the suboccipital and paracervical region (coat hanger) or platypnea (dyspnea while standing).

- Secondary or advanced laboratory testing is considered for use in select individuals for paraneoplastic syndromes (paraneoplastic panel) and serum and urine protein electrophoresis for monoclonal gammopathy for peripheral neuropathy.
  - See **Polyneuropathy (PN-3.1)** in the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines, **Multiple Myeloma and Plasmacytomas (ONC-25)** in the Oncology Imaging Guidelines, and **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.
- Semicircular canal dehiscence (SCD) is a rare syndrome caused by dehiscence in the boney covering of the affected superior, posterior or lateral semicircular canal. When present, it can result in vestibular symptoms of vertigo associated with auditory symptoms including oscillopsia evoked by noise and conductive hearing loss. The vestibular symptoms in SCD can be debilitating. Individuals may note that loud noises cause them to see things moving or that they experience a similar sensation when they cough, sneeze, or strain to lift something heavy. The signs of vestibular abnormalities in SCD relate directly to the effect of the dehiscence which has created a third mobile window of the inner ear. Some individuals have a conductive hearing loss for low-frequency sounds that can resemble the pattern in otosclerosis.
- Occlusive carotid artery disease does not cause fainting but rather causes focal neurologic deficits such as unilateral weakness. Thus, carotid imaging will not identify the cause of the fainting and increases cost. Fainting is a frequent complaint, affecting 40% of people during their lifetime.

Evidence Discussion (HD-23.1)

- MRI Brain is the preferred initial imaging modality for evaluation of persistent vertigo, vertigo associated with an abnormal neurologic exam, and vertigo due to a suspected central cause. [24,25]
- CT Head is not recommended for the initial evaluation of dizziness due to inferior soft tissue resolution when compared to MRI Brain. In addition, MRI Brain provides better visualization of the cerebellum and posterior fossa and is more sensitive for the detection of posterior fossa infarcts. For suspected superior semicircular canal dehiscence, CT Temporal bone is the appropriate initial imaging study.[24]

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- In the evaluation of dizziness or vertigo in the emergency department, the positivity rate of CT Head was 2%, for MRI Brain 4%, with the diagnostic yield increasing to 12% for MRI Brain if neurologic findings were present.[24]
- For dizziness due to suspected vertebral-basilar insufficiency, MRA sensitivity reaches 97% when performed with contrast-enhancement.[24]
- For suspected vertebral artery dissection, CTA had the highest sensitivity 100%, followed by MRA 77%, and Doppler ultrasound at 71%.[24]
- Vascular imaging should include the entire vertebral artery from the origin at the aortic arch to the basilar artery.[24]

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Syncope (HD-23.2)

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Indications	Medically Necessary Imaging
Syncope with focal signs of a neurologic deficit **OR** Syncope without focal signs of a neurological deficit AND negative or inconclusive Electrocardiogram (EKG)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • CT Head without contrast (CPT® 70450) **AND/OR** • CTA Head (CPT® 70496) **OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • CTA Neck (CPT® 70498) **OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549)
Recurrent syncope with risk of head injury or head trauma related to syncope	See **Head Trauma (HD-13.1)**
Situational syncope, including precipitating factors to syncope such as coughing, defecation, eating, laughing, or urination Myoclonic jerks without symptoms or signs associated with seizure, including but not limited to prolonged amnesia/confusion, tongue biting.	Advanced imaging is not medically necessary
Loss of consciousness with other symptoms or signs of seizure, including but not limited to, prolonged amnesia/ confusion, tongue biting, and/or urinary incontinence.	See **Epilepsy/Seizure (HD-9.1)**

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Evidence Discussion (HD-23.2)

- The Choosing Wisely Campaign Best Practices, supported by the American College of Emergency Physicians, American College of Physicians, and the American Academy of Neurology recommended against neuroimaging in the evaluation of simple syncope and a normal neurologic evaluation.[1,15]
- The initial evaluation for individuals with syncope includes a detailed history, physical exam, and electrocardiography.[1,6,15,26,29] Neuroimaging has a low diagnostic yield of 5% to 6.4% of an acute abnormality on CT Head.[1,15,27] Clinical factors associated with abnormal scans include head trauma or a focal neurologic deficit on exam.[1,15,27]
- In select cases when neuroimaging is medically necessary, structural brain imaging with either CT Head or MRI Brain may be useful, along with vascular imaging, depending on the suspected underlying pathology.[15,29]
- Inappropriate imaging studies may identify incidental findings, incorrectly assumed to be the cause of syncope, leading to further delay in the identification of the true cause and risk additional unnecessary procedures.[26]
- Situational syncope does not require advanced imaging.[3,6,28]

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Other Imaging Studies (HD-24)

Guideline

Transcranial Magnetic Stimulation (TMS) (HD-24.1)
Functional MRI (fMRI) (HD-24.2)
Magnetic Resonance Spectroscopy (MRS) (HD-24.3)
CSF Flow Imaging (HD-24.4)
CT or MRI Perfusion (HD-24.5)
Magnetic Resonance Neurography (MRN) (HD-24.6)
Cone Beam Computed Tomography (CBCT) (HD-24.7)
Transcranial Doppler (CPT® 93886) (HD-24.8)
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Transcranial Magnetic Stimulation (TMS) (HD-24.1)

HD.OI.0024.1.A

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In TMS, an electromagnetic coil placed on the surface of the skull overlying the motor cortex depolarizes the motor axons, creating a motor evoked potential (MEP), which is recorded via superficial skin electrodes as it passes through the upper and lower motor pathways to an innervated muscle.

TMS is typically utilized for behavioral health purposes.

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Functional MRI (fMRI) (HD-24.2)

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- fMRI is useful in pre-operative scenarios to define the “eloquent” areas of brain
  - The ordering physician must be a neurologist, neurosurgeon or radiation oncologist or any provider in consultation with one of these specialists.
- Primary medically necessary indications for fMRI include, but are not limited to, the following:
  - Assessment of intracranial neoplasm and other targeted lesions
  - Pre-surgical planning and operative risk assessment
  - Assessment of eloquent cortex (e.g., language, sensory, motor, visual centers) in relation to a tumor or another focal lesion
  - Surgical planning (biopsy or resection)
  - Therapeutic follow-up, as a one-time, post-operative, follow- up study
  - Evaluation of preserved eloquent cortex
  - Assessment of eloquent cortex for epilepsy surgery
  - Assessment of radiation treatment planning and post-treatment evaluation of eloquent cortex
- fMRI is medically necessary with PET Brain in epilepsy surgery planning
- Procedure codes for functional MRI:
  - CPT® 70554 MRI Brain, functional MRI, including test selection and administration of repetitive body part movement and/or visual stimulation, not requiring physician or psychologist administration
  - CPT® 70555 MRI Brain, functional MRI; requiring physician or psychologist administration of entire neurofunctional testing
  - If MRA Head (CPT® 70544) is medically necessary but Functional MRI (CPT® 70554 or CPT® 70555) was erroneously ordered, then CPT® 70544 may be substituted when appropriate
- MRI Brain (CPT® 70551 or CPT® 70553) and/or fMRI (CPT® 70554 or CPT® 70555) are medically necessary concurrently
  - See **Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)** in the Preface Imaging Guidelines if MRI Unlisted is requested for surgical planning

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Magnetic Resonance Spectroscopy (MRS) (HD-24.3)

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- MRS (CPT® 76390) involves analysis of the levels of certain chemicals in a pre-selected voxels (small regions) on an MRI scan done at the same time.
- When conventional imaging by magnetic resonance imaging (MRI) or computed tomography (CT) provides limited information regarding specific clinical questions, indications for MRS in adults and children include, but are not limited to, the following and are evaluated on a case-by-case basis:
  - Distinguish recurrent brain tumor from radiation necrosis as an alternative to PET (CPT® 78608)
  - Diagnosis of certain rare inborn errors of metabolism affecting the CNS (primarily pediatric individuals)
  - Evidence or suspicion of primary or secondary neoplasm (pre-treatment and post-treatment)
  - Grading of primary glial neoplasm, particularly high-grade versus low-grade glioma
  - Evidence or suspicion of brain infection, especially cerebral abscess (pre-treatment and post-treatment) and HIV-related infections
  - Seizures, especially temporal lobe epilepsy

Background and Supporting Information

- Evaluation of certain primary brain tumors where diagnostic accuracy has been established in peer-reviewed literature.
  - See **Primary Central Nervous System Tumors – General Considerations (ONC-2.1)**, **Low Grade Gliomas (ONC-2.2)**, and **High Grade Gliomas (ONC-2.3)** in the Oncology Imaging Guidelines

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CSF Flow Imaging (HD-24.4)

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- Pulse-gated MRI imaging or MRI CINE is generally performed as a part of an MRI Brain study. It is not coded separately for pre-operative evaluation of hydrocephalus, Chiari syndromes, Normal Pressure Hydrocephalus, Idiopathic Intracranial Hypertension (also known as pseudotumor cerebri), and spontaneous intracranial hypotension.
- There is no specific or unique procedure code for this study; it is done as a special sequence of a routine MRI Brain without contrast (CPT® 70551).
- If not previously performed as part of recent study, a second study for the purpose of evaluating CSF flow is medically necessary.

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CT or MRI Perfusion (HD-24.5)

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- Performed as part of a CT Head or MRI Brain examination in the evaluation of individuals with very new strokes or brain tumors.
- CT perfusion study, if performed in conjunction with a CT angiogram of the intracranial and/or cervical vessels, can be performed before, after, or concurrent with the CT angiogram.
  - CTA Head and/or Neck is medically necessary in conjunction with the CT Perfusion study (CPT® 0042T)
- CPT® 0042T - "cerebral perfusion analysis using CT" is medically necessary for the following:
  - Evaluation of acute stroke (<24 hours) to help identify individuals with stroke-like symptoms and to help identify those most likely to benefit from thrombolysis or thrombectomy
  - Follow-up for acute cerebral ischemia or infarction and/or reperfusion in the subacute or chronic phase of recovery
  - To assist in planning and evaluating the effectiveness of therapy for cervical or intracranial arterial occlusive disease (as an isolated test or in combination with a cerebrovascular reserve challenge) and/or chronic cerebral ischemia
  - Identifying cerebral hyperperfusion syndrome following revascularization
  - Evaluation of the vascular status of solid tumors where MRI is degraded due to susceptibility artifact from air-containing spaces, surgical clips, or dental work
  - Follow-up of tumor response to therapy
- MRI Perfusion is obtained with MRI Brain (CPT® 70551 OR CPT® 70552 OR CPT® 70553).
  - No additional CPT® codes are medically necessary to perform MRI perfusion.
- Medically necessary indications for perfusion magnetic resonance imaging (MRI) MRI Perfusion (CPT® 70551 OR CPT® 70552 OR CPT® 70553)[9] include the following:
  - Diagnosis and Characterization of Mass Lesions
    - Differential diagnosis (tumor versus tumor mimic)
    - Diagnosis of primary neoplasms (may include grading)
    - Surgical planning (biopsy or resection)
    - Targeting locations for biopsy
    - Guiding resection extent
  - Therapeutic follow-up
    - Radiation necrosis versus recurrent or residual tumor

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        - Chemonecrosis versus recurrent or residual tumor
        - Pseudoprogression and pseudoresponse
        - Monitor potential transformation of non-resectable low-grade neoplasms to higher grade
    - Assessment of Neurovascular Disease
        - Acute stroke (assessment of ischemic penumbra)
        - Assessment of the hemodynamic significance of cervical or intracranial vascular stenosis
        - Assessment of cervical or intracranial revascularization efficacy
        - Assessment of vasospasm
- Other indications are usually regarded as not medically necessary.

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Magnetic Resonance Neurography (MRN) (HD-24.6)

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- See **Magnetic Resonance Neurography (MRN) (PN-7.1)** in the Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines.

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Cone Beam Computed Tomography (CBCT) (HD-24.7)

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- CPT® Codes: CPT® 70486, CPT® 70487, CPT® 70488, CPT® 70480, CPT® 70482 (No separate 3-D rendering codes should be reported)
- An alternative to traditional CT imaging is in-office cone beam testing and possible decreased radiation dosage. The indications for office-based CT imaging are the same as for traditional scanners, and they should not be used for diagnosing or managing uncomplicated acute bacterial rhinosinusitis (ABRS).
- See **Temporomandibular Joint Disease (TMJ) (HD-30.1)** and **Dental/Periodontal/ Maxillofacial Imaging (HD-30.2)**

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Transcranial Doppler (CPT® 93886) (HD-24.8)

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- Transcranial Doppler (TCD) is a non-invasive ultrasonic technique that measures local blood flow velocity and direction in the proximal portions of intracranial and extracranial arteries

CPT Code	Description	Additional Notes
93886	Transcranial Doppler study of the intracranial arteries; complete study	
93888	Transcranial Doppler study of the intracranial arteries; limited study	
93892	Transcranial Doppler study of the intracranial arteries; emboli detection without intravenous microbubble injection	
93893	Transcranial Doppler study of the intracranial arteries; venous-arterial detection with intravenous microbubble injection	Report 93893 if the study is performed with intravenous microbubble injection. Transcranial Doppler studies described as “with contrast” are performed with intravenous microbubble injection. The bubbles serve to enhance ultrasound thus enabling better visualization of the intracranial arteries.

- Transcranial Doppler studies are ordered either as a single complete or limited study or as a combination of the complete or limited study with additional studies for further evaluation of the condition being investigated.
- Evaluation of Stroke/TIA usually includes CPT® 93886 (Vasoreactivity study) and either CPT® 93892 or CPT® 93893 (Emboli detection).

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  - Examples include:
    - Evaluation of right to left cardiac shunts: Detection of microemboli in individuals with stroke or TIA. (CPT® 93892 or CPT® 93893 added to CPT® 93886)
    - Evaluation of intracranial occlusive disease in individuals with documented stroke or TIA
    - Evaluation of hemodynamic effects of known severe extra-cranial occlusive disease
- TCD studies are medically necessary for the following:
  - Evaluation of severe stenosis or occlusion of the extracranial (≥60% diameter reduction) and major basal intracranial arteries (≥50% diameter reduction)
  - Detection and serial evaluation of cerebral vasospasm in subarachnoid hemorrhage
  - Evaluation of cerebral embolization including in COVID-19 and refractory encephalopathy
  - Assessing the extent of collateral circulation in individuals with known regions of severe stenosis or occlusion
  - To detect residual right to left shunting after repair/closure of an intracardiac or intrapulmonary shunt
  - Evaluation of AVM both pre- and post-surgical intervention.
  - Periprocedural monitoring to detect cerebral thrombosis, embolization, hypoperfusion, and hyperperfusion
  - Assessing the stroke risk in children aged two to sixteen with homozygous sickle cell disease
  - Annual screening for individuals with Sickle Cell Anemia (Hb-SS) and Sickle Beta Thalassemia (Sß) (CPT® 93886) up to the age of 16.
- TCD studies are not medically necessary for evaluation of:
  - Brain tumors
  - Familial and degenerative disease of the brain
  - Psychiatric disorders
  - Epilepsy
  - Migraine or other primary headache disorders
  - Infectious and inflammatory conditions

Background and Supporting Information

- Transcranial Doppler (TCD) ultrasound provides rapid, non-invasive, real-time measure of cerebrovascular function.
- TCD can be used to measure flow velocity in the proximal cerebral arteries to assess relative changes in flow, diagnose focal vascular stenosis, or to detect embolic signals within these arteries.

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- TCD can be used to measure blood flow responses to changes in blood pressure (cerebral autoregulation), changes in end-tidal CO2 (cerebral vasoreactivity), or cognitive and motor activation (neurovascular coupling or functional hyperemia).
- A technical limitation of TCD includes inadequate temporal bone acoustic windows due to a thickened skull which limits ultrasound penetration.
- Studies are ongoing regarding the use of TCD in the evaluation of dementia and psychiatric conditions such as depression.
- CPT® 93892 and CPT® 93893 represent add on services that require additional expertise, lab time, and equipment not included in the complete and limited codes. These additional codes may be appropriate during the same encounter if medical necessity is documented.
- CPT® 93892/CPT® 93893: Identification of right to left shunts (microembolic signals may be detected during TCD monitoring) and may indicate source of emboli in individuals with stroke or TIA. TCD bubble test is very sensitive and may be superior to transthoracic and transesophageal echocardiography in detection of right to left shunts.
- Transcranial Doppler (TCD) is considered not medically necessary for the following indications:
  - Assessing individuals with migraine;
  - Monitoring during cardiopulmonary bypass and other cerebrovascular and cardiovascular interventions, and surgical procedures (except during carotid endarterectomy, as noted above);
  - Evaluation of individuals with dilated vasculopathies such as fusiform aneurysms;
  - Assessing autoregulation, physiologic, and pharmacological responses of cerebral arteries; and/or
  - Evaluating children with various vasculopathies, such as moyamoya disease and neurofibromatosis.

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Evidence Discussion (HD-24)

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- Functional magnetic resonance imaging (fMRI) is useful for localizing eloquent cortex in relation to a focal brain lesion, for pre-surgical planning and therapeutic follow up.[7] Overall functional MRI imaging sensitivity is 83% with a specificity of 82%, for mapping language and motor functions. [22] Functional MRI is a useful tool for predicting post-operative outcomes in individuals with a single brain tumor.[23] Overall, fMRI studies are used in pre-operative decision making in 89% of tumor individuals and in 91% of epilepsy surgery individuals.[24,25] In 63% of epilepsy individuals undergoing surgical evaluation, fMRI imaging results helped to avoid further studies, including the Wada test.[25]
- For cases when conventional imaging by magnetic resonance imaging or computed tomography provides limited information regarding specific clinical questions, magnetic resonance spectroscopy (MRS) provides further characterization of brain tumors, radiation treatment changes, cerebral abscess, seizure disorders, and inherited metabolic disorders.[8,27,28] MRS has a 90% sensitivity and 86% specificity in distinguishing tumoral tissue from non-tumoral tissue.[27]
- Pulse-gated MRI imaging or MRI CINE is performed as part of an MRI Brain study and allows qualitative and quantitative analysis of oscillatory cerebrospinal fluid (CSF) movement in normal and abnormal conditions.[11,21] This imaging technique is useful for evaluation of hydrocephalus, Chiari syndromes, Normal Pressure Hydrocephalus, intracranial hypertension, and spontaneous intracranial hypotension.[10,11]
- MRI perfusion is useful for the diagnosis and characterization of mass lesions, surgical planning and therapeutic follow-up.[9,30] MR perfusion allows localization of tumor for higher yield on stereotactic biopsy and non-invasive differentiation between radiation necrosis from recurrent tumor when conventional MR findings are equivocal.[30]
- CT perfusion has multiple uses including in stroke diagnosis and treatment planning, characterization of neoplastic disease and response to treatment, and is alternative modality for those with contraindication to MRI-based perfusion imaging.[2,29]
- The American Heart Association/American Stroke Association guidelines for acute stroke management recommended CT Perfusion for selecting candidates for mechanical thrombectomy within 24 hours after last known well.[29]

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Epistaxis (HD-25)

Guideline

Epistaxis (HD-25.1)
References (HD-25)

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Epistaxis (HD-25.1)

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- After initial nasal endoscopy by ENT, if there are findings suspicious for a mass lesion, the following imaging is medically necessary:
  - CT Maxillofacial without or with contrast (CPT® 70486 or CPT® 70487) **AND/OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543)
- Individuals who have failed initial management with cauterization and packing and have persistent or recurrent epistaxis despite these primary interventions, should be referred to a clinician who can evaluate the individual for their candidacy for surgical ligation or endovascular embolization.
- Prior to embolization with surgical or endovascular technique, CT Maxillofacial without contrast (CPT® 70486) **OR** CT Maxillofacial with contrast (CPT® 70487) is medically necessary when requested by the clinician performing embolization or referring for embolization.
  - If endovascular embolization is planned, CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) is medically necessary ahead of the interventional radiologic procedure.

Background and Supporting Information

The American Academy of Otolaryngology Head and Neck Surgery recommended, in its most recent 2020 Clinical Practice Guidelines on Epistaxis, that the clinician should perform, or should refer to a clinician who can perform, nasal endoscopy to identify the site of bleeding and guide further management in individuals with recurrent nasal bleeding, despite prior treatment with packing or cautery, or with recurrent unilateral nasal bleeding. No recommendations for advanced imaging are outlined in this guideline without the exam findings (anterior rhinoscopy and/or nasal endoscopy) or the procedural needs of the individual indicating the need for such studies. If anterior rhinoscopy does not reveal the source of bleeding, it is recommended that the clinician perform nasal endoscopy or refer to a clinician who can perform nasal endoscopy, first.

Embolization procedures have shown an average nosebleed control rate of 87%, with minor transient complications in 20% (transient nasal ischemia, temporal-facial pain or numbness, headache, swelling, jaw claudication, trismus, and access site complications not requiring additional therapy) and major complications in up to 2.1% to 3.8% (skin/nasal necrosis, permanent facial nerve paralysis, monocular blindness, and stroke).

Detailed angiography, including internal and external carotid angiography, and precise embolization techniques are required. Despite use of meticulous techniques and knowledge of external carotid-internal carotid anastomoses, blindness and stroke are

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the most feared complications of endovascular embolization. These complications are rare but are more frequent than in individuals undergoing surgical arterial ligation. In one study, similar transient ischemic attacks are demonstrated across all groups but there is increased risk of stroke in the groups who underwent endovascular embolization alone (0.9%) or combined with surgical ligation (1.6%) as compared with surgical ligation alone (0.1%).

Evidence Discussion (HD-25)

- The American Academy of Otolaryngology - Head & Neck Surgery (AAO-HNS) recommendeds, in its most recent 2020 Clinical Practice Guidelines on Epistaxis, that the clinician should perform, or should refer to a clinician who can perform, nasal endoscopy to identify the site of bleeding and to guide further management in individuals with recurrent nasal bleeding despite prior treatment with packing or cautery, or with recurrent unilateral nasal bleeding. No recommendations for advanced imaging are outlined in this AAO-HNS Guideline without the exam findings (anterior rhinoscopy and/or nasal endoscopy) or the procedural needs of the individual, directing the need for such studies. If anterior rhinoscopy does not reveal the source of bleeding, it is recommended that the clinician perform nasal endoscopy or refer to a clinician who can perform nasal endoscopy.[3]
- Further characterization of any mass lesions suspected on initial nasal endoscopy may be evaluated with CT Maxillofacial, either with OR without contrast (CPT® 70487 or CPT® 70486), AND/OR MRI Orbit, Face, and/or Neck without and with contrast (CPT® 70543).[1,2]
- Because of the risks involved in embolization procedures (blindness, stroke, and others), CT or MRI imaging is supported prior to any planned intervention.[4]
- Embolization procedures have shown an average nosebleed control rate of 87%, with minor transient complications in 20% (transient nasal ischemia, temporal-facial pain or numbness, headache, swelling, jaw claudication, trismus, and access site complications not requiring additional therapy) and major complications in up to 2.1% to 3.8% (skin/nasal necrosis, permanent facial nerve paralysis, monocular blindness, and stroke).[4,5]

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Mastoid Disease or Ear Pain (HD-26)

Guideline

Mastoid Disease or Ear Pain (HD-26.1)
References (HD-26)

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Mastoid Disease or Ear Pain (HD-26.1)

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A pertinent clinical evaluation including a detailed history, physical examination (including otoscopic examination), must be performed on any individual with ear pain prior to considering advanced imaging. Common causes of ear pain include external and middle ear infections, dental problems, sinus infection, neck problems, tonsillitis, and pharyngitis.

Indications (Any one of the following)	Medically Necessary Imaging
• Persistent ear pain without obvious cause • Clinical suspicion for complicated or invasive infection such as mastoiditis • Clinical suspicion for complications from otitis media • Clinical suspicion of mass lesion causing ear pain • Significant trauma with concern for hematoma formation • Pre-operative planning • (For additional imaging requirements for cholesteatoma surgery, see bullet below) • (For skull base osteomyelitis, see **Skull Base Osteomyelitis (SBO) (HD-20.2)**)	• CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR** • CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR** • MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543)

- Advanced imaging is not medically necessary in the overwhelming majority of individuals with ear pain.
- The following imaging is medically necessary for **pre-operative evaluation** for cholesteatoma surgery when ordered by an otolaryngologist or in consultation with the otolaryngologist:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal Bone without and with contrast (CPT® 70482)

    AND/OR
  - MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR**

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  - MRI Internal Auditory Canal: (IAC) MRI can be reported as a limited study with one code from the set (CPT® 70540, CPT® 70542, or CPT® 70543), but should not be used in conjunction with MRI Brain codes (CPT® 70551, CPT® 70552, or CPT® 70553) if IAC views are performed as part of the brain
- The following imaging is medically necessary one time **post-operatively** to exclude residual or regrown cholesteatoma to avoid the need for a second-look surgery when ordered by an otolaryngologist or in consultation with the otolaryngologist:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **AND/OR**
  - MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), **OR**
  - MRI Internal Auditory Canal: (IAC) MRI can be reported as a limited study with one code from the set (CPT® 70540, CPT® 70542, or CPT® 70543), but should not be used in conjunction with MRI Brain codes (CPT® 70551, CPT® 70552, or CPT® 70553 if IAC views are performed as part of the brain
- Eustachian Tube Dilation: (endoscopic balloon dilatation of the Eustachian Tube, to treat persistent Eustachian tube dysfunction)
  - CT Orbit/Temporal Bone without contrast (CPT® 70480) is medically necessary for pre-operative evaluation of possible aberrant carotid.
- Concern for petrous apex lesions when requested by the otolaryngologist or in consultation with the otolaryngologist, the following are medically necessary:
  - CT Orbit/Temporal bone without contrast (CPT® 70480) **OR**
  - CT Orbit/Temporal bone without and with contrast (CPT® 70482) **AND/OR**
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - MRI Internal Auditory Canal: (IAC) MRI can be reported as a limited study with one code from the set (CPT® 70540, CPT® 70542, or CPT® 70543), but should not be used in conjunction with MRI Brain codes (CPT® 70551, CPT® 70552, or CPT® 70553 if IAC views are performed as part of the brain
- For concern related to non-resolving otalgia with chronic otorrhea:
  - See **Skull Base Osteomyelitis (SBO) (HD-20.2)**

Background and Supporting Information

- Common causes of ear pain include external and middle ear infections, dental problems, sinus infection, neck problems, and referred pain from the oral pharynx.
- Clinical suspicion for complications from otitis media such as coalescent mastoiditis, resulting in: subperiosteal abscess formation/Bezold's abscess, acute facial nerve paralysis, and intracranial abscess formation.
- Cholesteatomas are expansive cysts of the middle ear filled with cellular debris. They can be congenital or arise from recurrent middle ear infections or trauma to the tympanic membrane. Hearing loss is usually conductive, although if the lesion is

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large enough combined conductive and sensorineural hearing loss may be present. Otoscopic exam findings and symptoms may include a white mass in the middle ear cleft, painless drainage from the ear or chronic/recurrent ear infections.

- Petrous apex lesions/infections may include: cholesteatoma, cephalocele, mucocele, and cholesterol granuloma and can present with symptoms of pain, hearing loss, headache, vertigo, and Cranial nerve insults (including CN V VI, VII, IX, X, XI).

Evidence Discussion (HD-26)

- Contrast enhanced CT is commonly used for evaluation of head and neck infections due to its accessibility and short examination time. MRI provides better sensitivity of soft tissue infections in the setting of cholesteatoma, when there is concern for abscess formation or intracranial complications.[7]

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Hearing Loss and Tinnitus (HD-27)

Guideline

Hearing Loss (HD-27.1)
Tinnitus (HD-27.2)
References (HD-27)

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Hearing Loss (HD-27.1)

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- An initial evaluation including hearing tests, by bedside testing or by formal audiology, is necessary to determine whether an individual's hearing loss is conductive (external or middle ear structures) or sensorineural (inner ear structures, such as cochlea or auditory nerve) hearing loss. See **General Guidelines (HD-1.0)**
- CT Orbits/Temporal Bone without (CPT® 70480) **OR** MRI Brain without and with contrast (with IAC views) (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for the following:
  - Mixed conductive (MC)/Sensorineural (SN) hearing loss or any sensorineural hearing loss (MRI generally preferred for SN - See *Background and Supporting Information*)
  - Unilateral fluctuating or asymmetric hearing loss
  - Cholesteatoma (see **Mastoid Disease or Ear Pain (HD-26.1)**)
  - Congenital hearing loss
  - Surgical planning, including cochlear implants (both CT Temporal Bone and MRI Brain for surgical planning if requested by surgeon or any provider in consultation with the surgeon)
  - Hearing loss with vertigo (see **Dizziness/Vertigo (HD-23.1)**)
- CT Orbits/Temporal Bone without contrast (CPT® 70480) is medically necessary for the following:
  - Conductive hearing loss should have a CT Temporal Bone initially in the absence of an evident mass in the middle ear
- CT Orbits/Temporal Bone with contrast (CPT® 70481) is medically necessary for the following:
  - Glomus tumors or other vascular tumors of the middle ear, and/or surgical planning
  - Acquired sensorineural hearing loss if MRI unavailable or contraindicated
- Limited MRI of the internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) is medically necessary when requested by the provider in place of a complete MRI Brain. Note: Limited MRI codes should not be used in addition to MRI Brain codes; IAC views are performed as additional sequences as part of the brain study (see **General Guidelines – Anatomic Issues (HD-1.1)**)
- FIESTA protocol can be performed with MRI Brain without contrast (CPT® 70551) or limited MRI with attention to internal auditory canals.

Background and Supporting Information

- Sensorineural (SN) hearing loss – MRI is generally preferable to CT. CT Temporal bone is medically necessary in post-traumatic SN hearing loss, to evaluate for bony

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remodeling of the IAC due to vestibular schwannoma and labyrinthine ossification resulting from prior infection and for consideration of otospongiosis, a common cause of MC and SN hearing loss.

Evidence Discussion (HD-27.1)

- A complete history and otologic exam should be performed prior to advanced imaging for the workup of hearing loss. Formal audiometric testing is also necessary to determine whether the hearing loss is conductive, sensorineural, or mixed.[3,6]
- MRI brain is generally preferred for sensorineural and mixed hearing loss, particularly for unilateral hearing loss, congenital loss, or for surgical planning.[1,4,5]
- CT orbits/temporal bone is preferred for cases of conductive hearing loss, trauma, or suspected bony or middle ear disorders.[1,4,5]
- Both may be supported for surgical planning (cochlear implants, petrous apex disorders).[1,4,5]
- 3D-Fast imaging employing steady state acquisition (3D-FIESTA) demonstrates significantly higher spatial resolution with superior imaging contrast between cranial nerves and CSF with a shorter acquisition time than conventional MRI scan.[7]

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Tinnitus (HD-27.2)

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- A hearing evaluation is not required prior to imaging for tinnitus.
- The history in individuals with tinnitus should include a description of the tinnitus (episodic or constant, pulsatile or non-pulsatile, rhythmicity, pitch, quality of the sound), as well as inciting or alleviating factors. Continuous and pulsatile tinnitus are more concerning for an underlying and significant disorder. Audiometric assessment can be used as initial diagnostic testing particularly in individuals with tinnitus that is unilateral, persistent (>6 months) or associated with hearing difficulties (see **General Guidelines (HD-1.0)**)

Indications (Any one of the following)	Medically Necessary Imaging
• Clinical suspicion of mass lesion causing tinnitus • Asymmetric or unilateral non-pulsatile tinnitus (i.e., tinnitus that localizes to one ear) • Tinnitus associated with focal neurologic abnormalities, including asymmetric hearing loss • Persistent tinnitus after recent significant trauma. • Pulsatile tinnitus with or without concern for vascular lesion	• CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR** • CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR** • MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR** • MRI Brain without contrast with attention to internal auditory canals (CPT® 70551) **OR** • MRI Orbits/Face/Neck without contrast (CPT® 70540), with contrast CPT® 70542, or without and with contrast (CPT® 70543)
• Pulsatile tinnitus • Suspicion for vascular lesions	• MRA Head (CPT® 70544, CPT® 70545 OR CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548 or CPT® 70549) **OR** • CTA Neck (CPT® 70498)

- Imaging is not medically necessary for bilateral non-pulsatile tinnitus without other neurologic signs or symptoms
- Limited MRI of the internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) is medically necessary when requested by the provider in place of a complete MRI Brain. Note: Limited MRI codes should not be used in addition to MRI Brain

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codes; IAC views are performed as additional sequences as part of the brain study (see **General Guidelines – Anatomic Issues (HD-1.1)**)
- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures.

Background and Supporting Information

- Non-pulsatile tinnitus may be described as ringing, buzzing, or clicking sensations which is constant and non-synchronous.
- Pulsatile tinnitus is a repetitive sound coinciding with the individual's heartbeat. The symptom may be subjective or objective.

Evidence Discussion (HD-27.2)

- A targeted history and clinical examination should be performed as the initial evaluation of an individual with tinnitus and determination as to whether the tinnitus is bothersome or not should be made before any imaging is considered.[3,6]
- Both MRI and CT have utility in diagnosing the etiology of tinnitus, particularly for concerns of mass lesions, or for tinnitus in conjunction with hearing loss or trauma.[1,4,5]
- MRA or CTA of the head and neck are also useful in the workup of pulsatile tinnitus or for suspicion of a vascular lesion.[1,4,5]

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Neurosurgical Imaging (HD-28)

Guideline

Neurosurgical Imaging (HD-28.1)
Neuronavigation (HD-28.2)
Post-Operative Imaging (HD-28.3)
Evidence Discussion (HD-28)
References (HD-28)

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Neurosurgical Imaging (HD-28.1)

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- Typically, advanced imaging for monitoring disease for mass lesions occurs after biopsy (histologic) confirmation. This ensures appropriate determination related to phase of oncology imaging and alignment to appropriate diagnosis-specified guideline section.
  - However, repeat imaging by neurosurgeons or others of the management team for areas of the central nervous system (CNS) where permanent neurologic damage would be excessive with even a limited biopsy attempt is medically necessary.
  - Examples would include, but are not exclusive to: medically fragile individual, and tumors of the brainstem, eloquent areas of the brain, deep gray matter areas of the brain (ex. thalamus), and cavernous sinus.
- Repeat diagnostic head imaging is medically necessary when:
  - previous diagnostic head imaging is determined to be inadequate or additional imaging sequences/protocols are required by the neurosurgeon or the treatment team
  - prior imaging is greater than 6 months old

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Neuronavigation (HD-28.2)

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- Neurosurgical navigation is “image-based” meaning that the necessary pre-operative CT and MRI images are used for navigation in the operating room (image acquisition). Accurate registration (a process to match the pre-operative images to the individual position) of pre-operative images is necessary to guide surgery regardless of the navigation system that is used. Registration can be point-based or surface matched routines to allow the surgeon to view the overlapping data sets and the current situation to allow navigation.
- The process of registration for neuronavigation via the acquisition of pre-operative CT and MRI images does not require a radiologist interpretation.
  - Diagnostic imaging codes are not medically necessary for the purpose of registration for neuronavigation.
  - Can be referenced by proprietary brand systems such as Brainlab or Stealth imaging procedures
  - See **Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)** in the Preface Imaging Guidelines and **Unlisted Procedure Codes (ONC-1.5)** in Oncology in the Oncology Imaging Guidelines
- Advanced imaging for neuronavigation (image acquisition for registration for surgery) is medically necessary with one of each of the following as unlisted codes apply:
  - Unlisted MRI procedure code (CPT® 76498)
  - Unlisted CT procedure code (CPT® 76497)
  - Due to variances with techniques currently available for neuronavigation, the following are medically necessary:
    - CTA Head without and with contrast (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) (to avoid arterial and venous structures)
    - 3D (CPT® 76377 or CPT® 76376) (see **General Guidelines – Other Imaging Situations (HD-1.7)**)
  - Diagnostic imaging codes are only medically necessary if radiological supervision and interpretation of imaging is necessary with supporting documentation
    - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with contrast (CPT® 70552) **OR** MRI Brain without and with contrast (CPT® 70553) (contrast as requested) **AND/OR** CT Head without contrast (CPT® 70450) **OR** CT Head with contrast (CPT® 70460) **OR** CT Head without and with contrast (CPT® 70470) (contrast as requested)

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Post-Operative Imaging (HD-28.3)

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- Post-operative imaging including MRI Brain without contrast (CPT® 70551), or MRI Brain with contrast (CPT® 70552), or MRI Brain without and with contrast (CPT® 70553) (contrast as request) or CT Head without contrast (CPT® 70450), or CT Head with contrast (CPT® 70460), or CT Head without and with contrast (CPT® 70470) (contrast as request) is medically necessary per neurosurgeon's or in concert with management team's request that includes, but not exclusive to the following indications:
  - Within 24-72 hours following brain surgery including to document the need for repeat surgery or if adjuvant intervention is necessary, concern or rule out for complication(s), evaluation if incomplete resection vs. consideration for plan for gross resection
  - Signs or symptoms indicating concern of clinical deterioration
  - Development of new neurological signs or symptoms
  - Follow-up on blood products, edema, and/or concern of cerebrospinal fluid leak
  - Follow- up imaging per condition-based guideline
- See additional condition-based guidelines:
  - Pediatric Neurosurgeries
    - See **Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)** in the Pediatric Head Imaging Guidelines
    - See **Modality General Considerations (PEDONC-1.3)** and **Pediatric CNS Tumors (PEDONC-4)** in the Pediatric and Special Populations Oncology Guidelines
  - Epilepsy
    - See **Perioperative Evaluations for Drug-Resistant Epilepsy (HD-9.2)**
  - Movement Disorders
    - See **Movement Disorders (HD-15.1)**
  - Pituitary or Sella Surgery
    - See **Pituitary (HD-19.1)**
  - Acoustic Neuroma and Other Cerebellopontine Angle Tumors
    - See **Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)**
  - Central Nervous System Tumors
    - See **Primary Central Nervous System Tumors (ONC-2)** and **Brain Metastases (ONC-31.3)** in the Oncology Imaging Guidelines

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Evidence Discussion (HD-28)

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- Imaging modalities for neurosurgical planning include MRI Brain and CT Head, along with vascular imaging.
- MRI Brain is the preferred modality for the evaluation of intracranial neoplasms and other conditions affecting the brain parenchyma, meninges or cranium, due to its superior soft tissue resolution when compared to CT Head.[19]
- CT Head is the preferred modality for evaluation of bony structures in the pre-operative setting.[22]
- For localization of relevant vascular anatomy prior to surgery, MRI angiography (MRA) head and neck or CT angiography (CTA) head and neck, may be appropriate.[20,21]
- Pre-surgical navigational imaging, whether by CT or MRI, allows a spatially accurate anatomical individual model for use in the treatment-planning process.[1,18]
- The requirements for surgical planning images differ from the requirements for diagnostic images, especially regarding the spatial accuracy of the images in the stereotactic coordinates used for localization and targeting.[1,18]
- Navigation based on an immediate pre-operative scan optimizes the accuracy of data used for initial surgical planning.[1] Navigation systems reduce length of surgery, lower incidence of wound infection and shorten length of post-operative hospital stay.[23]
- For post-operative imaging, CT Head is also useful for follow-up of intracranial hemorrhage, edema, hydrocephalus, shunts, and general post-operative follow-up. CT Head has the benefit of providing rapid evaluation if a post-operative complication is suspected.[19] Post-operative MRI Brain provides superior soft tissue resolution in less urgent scenarios.[22]

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Sinus and Facial Imaging (HD-29)

Guideline

Sinus and Facial Imaging (HD-29.1)
Evidence Discussion (HD-29.1)
References (HD-29)

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Sinus and Facial Imaging (HD-29.1)

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- CT Maxillofacial without contrast (CPT® 70486) or limited CT Sinus without contrast (CPT® 76380) is medically necessary for ANY of the following:
  - Acute sinusitis without resolution of symptoms after a minimum of 4 weeks of treatment (treatment can include an appropriate course and duration of empiric oral antibiotic, topical intranasal steroid, and/or nasal saline rinses.)
  - Concern for potential or suspected complicated sinusitis, which is sinusitis with actual or threatened orbital or intracranial extension
  - Recurrent sinusitis (4 or more episodes of acute bacterial rhinosinusitis within the past 12 months without symptoms or signs between episodes)
    - In practice, recurrent acute exacerbations of chronic rhinosinusitis are seen as well as recurrent acute rhinosinusitis with disease free intervals between the acute episodes. CT Maxillofacial without contrast (CPT® 70486) may still be medically necessary under chronic sinusitis definitions.
  - Suspicion of chronic sinusitis (**Note:** A trial of antibiotic therapy is not required prior to imaging if individual meets criteria for chronic sinusitis)
    - Chronic sinusitis is suspected when **BOTH** of the following are met:
      - At least two of the following on history and physical exam:
        - Mucopurulent drainage
        - Nasal obstruction or congestion
        - Facial pain, pressure, and/or fullness (may involve the anterior face, periorbital region, or manifest with headache that is localized or diffuse)
        - Decreased sense of smell (see **Taste and Smell Disorders (HD-2.1)** if anosmia, hyposmia, or dysosmia is an isolated symptom)

        AND
      - The aforementioned findings on history and physical exam have been present for at least 12 weeks
  - Sinus surgery is being considered (including Balloon Sinus Ostial Dilation or Functional Endoscopic Sinus Surgery)
  - Follow-up on incidentally noted sinus pathology (i.e., mucosal thickening, partial opacification of a sinus, or other indeterminate finding in incompletely visualized sinuses) on other imaging studies not performed for the purpose of evaluating sinus pathology, such as MRI Brain for headache, when requested by ENT for clinical correlation.
- Studies requested for the sole purpose of navigation for sinus surgery should be coded CPT® 77011 (CT guidance for stereotactic localization).

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  - It is not appropriate to report both CPT® 70486 and CPT® 77011 for the same CT stereotactic localization imaging session (see **CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2)**) in the Preface Imaging Guidelines).
- For unexplained cough as the main symptom, and suspected Upper Airway Cough Syndrome (UACS) as the etiology, see **Cough (CH-3.1)** in the Chest Imaging Guidelines.
- For the immunocompromised individual with symptoms of sinusitis, the following are medically necessary:
  - CT Maxillofacial (contrast level as requested) when ordered by the surgeon or in consultation with the surgeon (i.e., ENT or ophthalmologist)
  - CT Maxillofacial with contrast (CPT® 70487) when there is suspicion for ANY of the following:
    - Orbital or facial cellulitis
    - Proptosis
    - Abnormal visual examination
    - Ophthalmoplegia
    - Fungal or vascular lesions visualized in nasal cavity
- CT Maxillofacial without contrast (CPT® 70486) **OR** CT Maxillofacial with contrast (CPT® 70487) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is medically necessary for the conditions below. However, CT Maxillofacial without contrast (CPT® 70486) may also be requested with MRI Orbits/Face/Neck without and with contrast (CPT® 70543) for surgical planning or osseous involvement.
  - Sinonasal obstruction, polyp, or suspected mass
  - Suspected orbital complication
  - Suspected invasive fungal sinusitis
  - Cystic fibrosis
  - Osteomyelitis and odontogenic infections, see **Skull Base Osteomyelitis (SBO) (HD-20.2)** and **Dental/Periodontal/Maxillofacial Imaging (HD-30.2)** for additional imaging modalities
- CT Maxillofacial without contrast (CPT® 70486) **AND** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) are medically necessary when ordered by the ENT/specialist prior to planned biopsy of an intranasal mass — the extent and origin of which could not be determined by complete rhinologic exam alone
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for suspected intracranial complication
- CT Orbits/Temporal bone without contrast (CPT® 70480) or CT Orbits/Temporal bone with contrast (CPT® 70481) is medically necessary performed alone or added to CT Maxillofacial for:
  - suspected orbital complications
- For skull base osteomyelitis (SBO), see **Skull Base Osteomyelitis (SBO) (HD-20.2)**

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- Repeat imaging is medically necessary for ANY of the following scenarios:
  - An ENT specialist or any provider in consultation with an ENT specialist requests the imaging **and** ONE or more of the following:
    - There has been a follow-up visit since the previous imaging and there is no improvement after an additional 3 weeks of conservative treatment after initial imaging was completed:
      - The following imaging is medically necessary: CT Maxillofacial without contrast (CPT® 70486) **OR** limited CT Sinus without contrast (CPT® 76380)
    - There is a new abnormality on exam such as obstructing mass
      - The following imaging is medically necessary: CT Maxillofacial without contrast (CPT® 70486) **OR** CT Maxillofacial with contrast (CPT® 70487)
    - If sinus surgery is planned (including but not limited to Balloon Sinus Ostial Dilation or Functional Endoscopic Sinus Surgery) **AND** the most recent diagnostic CT Maxillofacial without contrast (CPT® 70486) is greater than 6 months old **OR** there is a change in clinical status as described above (i.e. interval completion of provider-prescribed medical management after the last CT was performed), a repeat diagnostic CT Maxillofacial without contrast (CPT® 70486) is supported for surgical planning.
      - Repeat CT Maxillofacial solely for the use of navigation during the sinus surgery (i.e., the most recent diagnostic CT Maxillofacial performed within the prior six months was only inadequate due to lacking anatomic landmarks or insufficient thinness of cuts) should be requested with CPT® 77011, not the diagnostic CPT® 70486.
      - 3D Rendering (CPT® 76376 or CPT® 76377) should not be reported in conjunction with CPT® 77011 (or CPT® 70486 if used). The procedure inherently generates a 3D dataset.
- Complications of ABRS (acute bacterial rhinosinusitis) are suspected based on:
  - Any constellation of symptoms worrisome for intracranial extension of infection or meningitis (i.e., severe headache, photophobia, fever, neck stiffness)
  - Severe headache
  - Facial swelling
  - Cranial nerve palsies
  - Orbital signs (cellulitis, impaired extraocular motility, decrease in vision or proptosis)
- Complications of ABRS are best assessed using iodine contrast-enhanced CT Maxillofacial with contrast (CPT® 70487) **OR** gadolinium-based MR imaging (MRI Orbits/Face/Neck without and with contrast (CPT® 70543) to identify extra-sinus extension or involvement; thus, this imaging is medically necessary.

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- CT Maxillofacial without contrast (CPT® 70486) may also be requested with MRI Orbits/Face/Neck without and with contrast (CPT® 70543) for surgical planning or osseous involvement.
- Suspected complications are the only medically necessary indication for MR imaging of the paranasal sinuses in the setting of ABRS.

For additional medical necessity criteria for CT Maxillofacial, see **Cone Beam Computed Tomography (CBCT) (HD-24.7)** and **Dental/Peridontal/Maxillofacial Imaging (HD-30.2)**)

Evaluation of potential candidates for Eustachian Tube balloon dilation procedure is with a one-time CT of the temporal bone without contrast (CPT® 70480). See medical necessity discussion in **Mastoid Disease or Ear Pain (HD-26.1)**. CT Sinus/Maxillofacial irrespective of contrast level is not supported if the sole indication for medical necessity is to evaluate a potential candidate for Eustachian Tube balloon dilation procedure, without meeting other HD-29.1 medical necessity criteria.

Background and Supporting Information

- Rhinosinusitis is defined as inflammation of the nasal cavity and adjacent paranasal sinuses. Acute sinusitis refers to symptom duration <4 weeks, subacute 4 to 12 weeks, and chronic >12 weeks. Complicated sinusitis refers to symptoms suggesting spread of disease into adjacent structures, including orbital or intracranial complications.
- There is no evidence to support advanced imaging of acute (<4 weeks) and subacute (4 to 12 weeks) uncomplicated rhinosinusitis.
- There is no evidence to support routine follow-up advanced imaging after treatment with clinical improvement of sinusitis.

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Evidence Discussion (HD-29.1)

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- The American Academy of Otolaryngology – Head & Neck Surgery (AAO-HNS) recommended that clinicians should not obtain radiographic imaging for individuals with suspected uncomplicated Acute Rhinosinusitis (ARS), with imaging reserved for cases with clinically suspected complication. ARS refers to inflammation of the nasal cavity and paranasal sinuses lasting <4 weeks' duration.[1]
- Contrasted CT Maxillofacial is first line imaging for rhinosinusitis with suspected complications (orbital or intracranial). There is up to 91% accuracy with CT to detect orbital complications vs clinical exam alone. CT also is preferred for surgical planning. However, CT is often more useful for surgical planning and easier to perform. Non-contrasted CT sinus is not preferred but may be useful for surgical navigation. There is no relevant literature to support pre- and post-contrast CT imaging. MRI head or orbits/face/neck can be complementary with CT. MRI is more accurate than CT in the evaluation of soft tissues regarding intra-orbital and intracranial complications.[3,4]
- Chronic rhinosinusitis, acute recurrent bacterial sinusitis, non-invasive fungal sinusitis, and/or sinonasal polyposis are best evaluated initially with non-contrast CT Maxillofacial. CT is critical for surgical planning. Contrast is not necessary unless complications are suspected. MRI is not useful as the first-line study because of the lack of bony detail. In select cases, evaluation with MRI without and with IV contrast may be helpful to differentiate fluid secretions from inflamed mucosa and exclude an underlying obstructing mass.[3,4]
- Urgent CT Maxillofacial, either without or with IV contrast is first line imaging for any suspected invasive fungal sinusitis, as delay in diagnosis and surgical debridement could increase the already high risk of mortality. In cases of invasive fungal sinusitis, MRI without and with IV contrast of the head and/or orbits/face/neck is adjunctive to look for invasion into surrounding soft tissues as well as vascular complications.[3,4]
- CT and MRI are considered complimentary imaging modalities in the evaluation of a sinonasal mass—localizing and characterizing the lesions to determine their extent for treatment planning. If an MRI is planned, the CT may be performed without IV contrast since the main purpose of the CT is to evaluate osseous involvement.[3,4]

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Temporomandibular Joint Disease (TMJ) and Dental/ Periodontal/Maxillofacial Imaging (HD-30)

Guideline

Temporomandibular Joint Disease (TMJ) (HD-30.1)
Dental/Periodontal/Maxillofacial Imaging (HD-30.2)
References (HD-30)

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Temporomandibular Joint Disease (TMJ) (HD-30.1)

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- MRI TMJ (CPT® 70336) is the diagnostic study of choice and is medically necessary for those who fail a minimum of 6 weeks of non-surgical treatment **AND** who are actively being considered for TMJ surgery.
  - The exception to the conservative management and surgical consideration requirement above includes the need to evaluate a recent trauma, dislocation, severe malocclusion, dental infection, or abscess.
- CT Maxillofacial without contrast (CPT® 70486) or without and with contrast (CPT® 70488) when there is suspicion of bony involvement based on prior x-ray or MRI
- Ultrasound (CPT® 76536) is medically necessary to look for the presence of a joint effusion and to evaluate cartilage and disk displacement with open and closed mouth imaging and to guide injections
- For TMJ imaging in individuals with Juvenile Idiopathic Arthritis (see **Temporomandibular Joint (TMJ) Imaging in Children (PEDHD-25)** in the Pediatric Head Imaging Guidelines)
  - MRI TMJ (CPT® 70336) is medically necessary annually for detecting silent TMJ arthritis in children and young adults with juvenile idiopathic arthritis as requested by a rheumatologist and/or oral/maxillofacial surgeon (OMS) and/or any provider in consultation with a rheumatologist or OMS.
  - Repeat imaging with MRI TMJ (CPT® 70336) in individuals with JIA is medically necessary for any of the following:
    - Change in signs or symptoms suggesting progression of disease
    - To monitor the effects of treatment[11]
  - Bone Scintigraphy/Bone Scan 3 Phase Study (CPT® 78315) in individuals over 12 years of age is medically necessary in anticipation or consideration of surgery.
- Jaw Asymmetry - Unilateral condylar hyperplasia is manifested by slow growth in areas of the mandible causing facial asymmetry. It is usually a self-limiting condition seen predominantly in individuals 12 to 30 years of age.
  - CPT® 78315 Bone Scan 3 Phase Study is medically necessary in anticipation or consideration of surgery.

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Evidence Discussion (HD-30.1)

- MRI is preferred for evaluation of the temporomandibular joint (TMJ) due to its superior contrast resolution and its ability to acquire dynamic imaging for demonstration of the functionality of the joint.[3,10]
- MRI is the imaging modality of choice for the diagnosis of internal derangement with an accuracy of 95% in assessing the disc position and form and 93% accuracy in assessing the osseous changes.[3]
- MRI is reserved for individuals with persistent symptoms in whom conservative measures have been ineffective, or in those with suspected internal joint derangement.[2,8,9] Imaging the TMJ prematurely may lead to harms including unnecessary surgery.[7]
- CT is the alternative modality for evaluating bony anatomy of the TMJ, fractures, degenerative changes, erosions, infections, congenital anomalies, acute and chronic inflammatory conditions, pre-operative evaluation and follow- up after surgery.[3,5,10]
- For pre-operative planning of unilateral condylar hyperplasia, bone scintigraphy is useful to predict ongoing condylar growth.[16]
- The diagnosis of chronic rheumatoid arthritis of the TMJ is established with contrast-enhanced MRI.[11] It is the preferred imaging study for diagnosis, disease progression, treatment monitoring and annual surveillance of TMJ arthritis in juvenile idiopathic arthritis (JIA).[11]

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Dental/Periodontal/Maxillofacial Imaging (HD-30.2)

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- Initial imaging with plain x-rays/dental x-rays is required.
- Cone beam CT is required for surgical planning and is, therefore, medically necessary for the potential indications listed below.
- Potential indications include but are not limited to:
  - Osteomyelitis and odontogenic infections (x-ray **NOT** required)
  - Bisphosphonate-related osteonecrosis of the jaw (x-ray **NOT** required)
  - Impacted teeth
  - Supernumerary teeth
  - Dentoalveolar trauma
  - Root resorption
  - Foreign body
  - Odontogenic cysts, tumors, or other jaw pathology
  - Orthognathic surgery for dentofacial anomalies
  - Maxillofacial bone graft planning
  - Dental implants related to tooth loss from injury, trauma, or jaw pathology such as cysts, tumors, or cancer
  - Post-operative imaging, including dental implants
- Cone Beam CT: Report with CPT® Codes: CPT® 70486, CPT® 70487, CPT® 70488, CPT® 70480, CPT® 70482 (see **Cone Beam Computed Tomography (CBCT) (HD-24.7)**)
- 3-D rendering (CPT® 76376 or CPT® 76377) should **NOT** be reported separately
- Cone beam CT (CBCT) may also be called i-CAT scanner or mini-CAT scanner
- For cleft palate indications (see **Other Indications for Sinus Imaging (PEDHD-5.6)**)
- For salivary gland indications (see **Salivary Gland Disorders (Neck-11.1)**)

Background and Supporting Information

Dental x-rays include panoramic, periapical, and/or occlusal x-rays.

Evidence Discussion (HD-30.2)

- CT is the radiologic modality for evaluating the bony anatomy of the head, acute and chronic inflammatory conditions, paranasal sinuses, pre-operative evaluation and follow-up after surgery.[1,5]

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- Recommendations by the American Association of Endodontists and the American Academy of Oral and Maxillofacial Radiology supported the use of cone beam CT (CBCT) as a supplemental imaging technique when conventional radiography fails to answer the clinical question and for surgical planning.[1,4,12]

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Cranial Neuropathies (HD-31)

Guideline

Cranial Neuropathies (HD-31.1)
References (HD-31)

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Cranial Neuropathies (HD-31.1)

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Cranial Neuropathies Imaging Indications

- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for all individuals with new or worsening specific cranial nerve abnormalities.
- MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR** MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is also medically necessary for individuals with abnormalities in cranial nerves I, II, III, IV, V, VI, VII, IX, X, XI, or XII.
- CT Neck with contrast (CPT® 70491) is medically necessary for evaluation of abnormalities involving cranial nerves IX, X, XI, or XII.
- Imaging of the Brain and Orbit, Face and/or Neck is medically necessary concurrently when requested.
- For specific cranial neuropathies, see the corresponding guideline section listed below:
  - CN I: Olfactory nerve (see **Taste and Smell Disorders (HD-2.1)**)
  - CN II, III, IV, VI: Optic, Oculomotor, Trochlear and Abducens (see **Eye Disorders and Visual Loss (HD-32.1)**)
  - CN V: Trigeminal nerve (see **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10.1)**)
  - CN VII: Facial nerve (see **Facial Palsy (HD-6.1)**)
  - CN VIII: Vestibulocochlear nerve (see **Dizziness/Vertigo (HD-23.1)**, **Hearing Loss (HD-27.1)**, **Tinnitus (HD-27.2)**, **Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)**. For isolated nystagmus (see **Eye Disorders and Visual Loss (HD-32.1)**)
  - CN IX: Glossopharyngeal nerve (see **Glossopharyngeal Neuralgia/ Glossopharyngeal Neuropathy HD-10.2)**)
  - CN X: Vagal nerve, imaging as detailed above (see also **Recurrent Laryngeal Palsy/Vocal Cord Palsy (Neck-7.1)**)
  - CN XI: Spinal accessory nerve, imaging as indicated above
  - CN XII: Hypoglossal nerve, imaging as indicated above
- For cranial neuropathies, whether isolated or multiple, due to clinically suspected stroke and/or vascular dissection (see **General Guidelines - CT and MR Angiography (CTA/CTV and MRA/MRV) (HD-1.5)**, **Headache and Suspected Vascular Dissection (HD-11.1)** and **Stroke/TIA (HD-21.1)**

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Background and Supporting Information

If a detailed clinical evaluation is unable to localize the site of the lesion, imaging of the entire course of the relevant cranial nerve is required, as cranial neuropathy can result from pathology affecting the nerve fibers at any point along the course of the nerve, from the cranial nerve origin in the brainstem to the end organ supplied by the nerve, requiring multiple imaging modalities.

The spinal trigeminal tract and nucleus extend from the midpons caudally into the upper cervical cord at the C2-4 levels. For suspected lesions of the spinal trigeminal tract and nucleus, imaging the brain stem and cervical spinal cord is supported. See **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10.1)**.

Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
I	Olfactory (smell)	Anosmia, hyposmia, parosmia, phatosmia	2
II	Optic (vision)	Optic neuritis, disc edema, papilledema, afferent pupillary defect APD)	16, 17, 32
III	Oculomotor (eye and pupil movement)	Eye "down and out", +/- dilated pupil, ptosis, diplopia	32
IV	Trochlear (depresses the eye)	Inability to depress the eye, diplopia	32
V	Trigeminal (sensation, mastication, taste)	Pain, numbness, corneal reflex loss, jaw deviation, trigeminal neuralgia, loss of taste	10
VI	Abducens (lateral movement of the eye)	Eye turns medially, inability to abduct, lateral rectus palsy, diplopia	32

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Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
VII	Facial (movement facial muscles, taste at 2/3, salivation/ lacrimation)	Inability to close eyelid, smile, nasolabial fold flattening, hyperacusis, impaired taste, salivation, lacrimation	6
VIII	Auditory, Vestibular, Vestibulochochlear (hearing and balance)	Hearing loss, tinnitus, vertigo, nystagmus, abnormal gait/ balance, sway on Romberg	23, 27, 33
IX	Glossopharyngeal (swallow, sensation, pharynx, posterior 1/3 tongue, parotid salivary gland)	Depressed gag reflex and palate, dysphagia, uvula deviation, throat pain	10, 2
X	Vagus (swallow, speech, parasympathetic to heart, lungs, GI tract)	Vocal cord paralysis, recurrent laryngeal nerve palsy, spasmodic dysphonia	7.1, 1.1
XI	Spinal Accessory (motor function neck/shoulder)	Sternocleidomastoid (SCM) weakness when turning head opposite, shoulder elevation, winging scapula	1.1
XII	Hypoglossal (tongue movement)	Tongue deviation, atrophy, fasciculation	1.1

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Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
INO	Internuclear Opthalmoplegia (lesion of medial longitudinal fasciculus, CN III, CN VI)	Impaired adduction of ipsilateral eye with nystagmus of abducting eye	16, 21, 22
Horner Syndrome	Disruption of sympathetic innervation to eye and face	Ptosis, miosis (constricted pupil), facial anhidrosis (absence of sweating)	32.2, 11.3

Evidence Discussion (HD-31.1)

- Imaging of each body section along the entire course of the relevant cranial nerve may be medically necessary if detailed clinical evaluation is unable to localize the site of the lesion. Cranial neuropathy can result from pathology affecting the nerve fibers at any point along the course of the nerve, from the cranial nerve origin in the brainstem to the end organ supplied by the nerve, indicating need to image multiple body sections.[1,2] MRI Brain, Orbits, Face, Neck, or any combination may be necessary depending on the clinical need.[1,2]
- MRI is the standard modality for imaging the cranial nerves.[2,3]
- CT Neck is useful to exclude neck masses when evaluating either isolated or multiple lower cranial neuropathies.[2] CT may be complementary to MRI in characterizing skull base erosions, calcifications, and skull base foramina.[2]

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Eye Disorders and Visual Loss (HD-32)

Guideline

Eye Disorders and Visual Loss (HD-32.1)
Pupillary Abnormalities Including Horner's Syndrome (HD-32.2)
References (HD-32)

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Eye Disorders and Visual Loss (HD-32.1)

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- For specific conditions - See *Background and Supporting Information* that include table of abbreviations
- For eye pain, also refer to **Trigeminal Neuralgia/Trigeminal Neuropathy (HD-10.1)** and/or **Multiple Sclerosis and Related Conditions (HD-16)**
- Examination of ocular complaints and visual loss may include evaluation of pupillary responses, extraocular motility, visual acuity, visual field testing, intraocular pressures, external examination, slit lamp examination, and/or fundoscopic exam of retinae. An exam performed by a neuro-ophthalmologist, ophthalmologist, neurologist, or an optometrist meets this requirement.
- MRI Orbits/Face/Neck without contrast (CPT® 70540) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** CT Orbits/Temporal bone with contrast (CPT® 70481) **OR** CT Orbits/Temporal bone without contrast (CPT® 70480) **AND/OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for:
  - unexplained vision loss
  - optic atrophy (cranial nerve II)
  - optic neuropathy (cranial nerve II)
  - papilledema/optic disc swelling (cranial nerve II) (see **Cranial Neuropathies (HD-31.1)** and **Papilledema/Pseudotumor Cerebri (HD-17.1)**)
  - Afferent Pupillary Defect (APD) or Relative Afferent Pupillary Defect (RAPD)
  - chiasmal symptoms/signs (including bitemporal field deficit)
  - ophthalmoplegia, diplopia, and/or cranial nerve palsy (specifically CN III, IV, and VI, see **Cranial Neuropathies (HD-31.1)**)
  - nystagmus
- For optic disc edema/papilledema, CT Head without contrast (CPT® 70450) is medically necessary to assess for space-occupying processes such as intracranial hemorrhage, mass effect and hydrocephalus.
- For suspected optic neuritis, MRI is the preferred modality (see **Multiple Sclerosis (MS) (HD-16.1)** and **Neuromyelitis Optica Spectrum Disorders (HD-16.2)**)
- Visual field defects are associated with retrochiasmal pathology (see **Stroke/TIA (HD-21.1)** or **Primary Central Nervous System Tumors (ONC-2)** in the Oncology Imaging Guidelines or **Brain Metastasis (ONC- 31.3)** in the Oncology Imaging Guidelines)
- MRI Orbits/Face/Neck without contrast (CPT® 70540) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) or CT Orbits/Temporal bone with contrast (CPT® 70481) is medically necessary for:

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  - exophthalmos (including thyroid eye disease), enophthalmos or non-traumatic orbital asymmetry
  - suspected orbital cellulitis or atypical pre-septal cellulitis, uveitis, or scleritis
  - orbital mass or metastasis
  - orbital inflammatory syndrome (orbital pseudotumor) and dacryocystitis or dacryoadenitis
- CT Orbits/Temporal bone without contrast (CPT® 70480) and/or CT Head without contrast (CPT® 70450) is/are medically necessary for:
  - orbital trauma with visual defect
  - exophthalmos (including thyroid eye disease)
- CT Orbits/Temporal bone without contrast (CPT® 70480) or CT Orbits/Temporal bone with contrast (CPT® 70481) or CT Orbits/Temporal bone without and with contrast (CPT® 70482) or CT Maxillofacial without contrast (CPT® 70486) or CT Maxillofacial with contrast (CPT® 70487) or CT Maxillofacial without and with contrast (CPT® 70488) is medically necessary for:
  - pre-operative planning for procedures including dacryocystorhinostomy (DCR) to correct nasolacrimal duct obstruction (NLDO)
- When requested by the surgeon or in consultation with surgeon, contrast level as requested is medically necessary. This includes requests from ophthalmologists and oculoplastic surgeons. Contrast level preference may vary per institutional protocol.
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) is medically necessary for suspicion of intracranial aneurysm, including third nerve palsy with pupillary involvement (see **Cerebral Aneurysms (HD-12.1)**
- MRA Head (CPT®70544, CPT®70545, or CPT®70546) or CTA Head (CPT®70496) **AND/OR** MRA Neck (CPT®70547, CPT®70548, or CPT®70549) or CTA Neck (CPT® 70498) is medically necessary for evaluation of diplopia due to suspected stroke or TIA (see **Cerebral Aneurysms (HD-12.1)**)
- Amaurosis Fugax (see **Stroke/TIA (HD-21.1)**)
  - Individuals describe a transient darkening or loss of vision, typically monocular
- Central Retinal Artery Occlusion, Branch Retinal Artery Occlusion, and Ophthalmic Artery Occlusion (see **Stroke/TIA (HD-21.1)**)
  - Individuals describe a sudden monocular loss of vision or visual field. Etiology is usually embolic and is considered a stroke to the retina
- There is currently no data to support advanced imaging while on Tepezza® (teprotumumab). However, advanced imaging is medically necessary for any of the following concerns:
  - Neurologic symptoms or ophthalmologic symptoms
  - To reassess compressive optic neuropathy (symptoms/signs of compressive optic neuropathy include APD, decreased visual acuity, and/ or visual field defects)

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- For non-response to Tepezza® (teprotumumab) or relapses, worsening proptosis, diplopia, lid retraction, or optic neuropathy
- For surgical planning for orbital decompression, strabismus surgery, or lid surgery
  - When any of the above concerns are present, **ONE** of the following modalities is medically necessary:
    - MRI Orbits/Face/Neck without contrast (CPT® 70540)
    - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
    - CT Orbits/Temporal bone with contrast (CPT® 70481)
    - CT Orbits/Temporal bone without contrast (CPT® 70480)
    - CT head without contrast (CPT® 70450)

- Autoimmune Retinopathy
  - Suspicion for CAR (Cancer associated retinopathy) or MAR (melanoma associated retinopathy) syndromes (see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines)
- Oncologic conditions
  - Retinoblastoma (see **Retinoblastoma (PEDONC-12)** in the Pediatric and Special Populations Oncology Imaging Guidelines)
  - Uveal (choroidal) melanoma (see **Ocular Melanoma (ONC-5.9)** in the Oncology Imaging Guidelines)
  - Biopsy results are not required before initial staging
- Vasculitis including Temporal Arteritis (Giant Cell Arteritis) (see **Cerebral Vasculitis (HD-22.1)**)

Background and Supporting Information

- Imaging Non-Indications
  - Imaging is not necessary if visual loss or ocular symptom/sign is due to known intrinsic eye disease, such as refractive errors, amblyopia, pterygium, subconjunctival hemorrhage, conjunctivitis, cataracts, macular degeneration, central serous retinopathy, retinal vein occlusion, retinal detachment, etc. Monocular diplopia is not an indication for imaging. Physiologic anisocoria and surgically distorted pupils are not indications for imaging. Anisocoria may be physiologic or non-physiologic when the difference in pupil diameter between the two eyes is up to 2 mm.
  - Imaging is not typically necessary in cases of ptosis without concern for Horner's or 3rd nerve palsy
- Advanced imaging of the brain and orbit are not routinely paired.
  - Suspicion for disorders involving both regions is needed to image both regions.
  - Orbital imaging alone may be sufficient unless other signs or symptoms suggest brain involvement.

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- Thyroid function and iodine contrast: thyroid dysfunction can occur in susceptible individuals after iodine exposure.

List of Abbreviations and Meanings:

Abbreviation	Meaning
AC	anterior chamber
APD	afferent pupillary defect (see RAPD)
BCVA	best-corrected visual acuity
C3F8	gas bubble injected into vitreous cavity during retina surgery
cc	with correction (current new or old glasses or contact lenses)
CP	color plates
C/S	conjunctiva/sclera
CSME	clinically significant macular edema
CVF	confrontation visual field (testing of gross field of view)
D	disc, optic nerve head
DBH	dot blot hemorrhages
DCR	dacryocystorhinostomy
DFE	dilated fundus exam
E	esophoria at distance
E'	esophoria at near
EOM	extraocular movements
ERM	epiretinal membrane
ET	esotropia at distance
E(T)	iIntermittent esotropia at distance
ET'	esotropia at near

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Abbreviation	Meaning
E(T)'	intermittent esotropia at near
GVF	Goldmann visual field test
HT	hypertropia
HVF	Humphrey visual field test (automated perimetry)
I	iris
Ishihara	commonly used color plates
IOP	intraocular pressure
K	cornea
LF	levator function
LFH	lid fissure height
LLL	lids, lashes, lacrimal gland
M	macula
ME	macular edema
MH	macular hole
MP	membrane peel
MRD1	margin-reflex distance from upper lid margin to pupillary light reflex
MRx	manifest refraction
NI	no improvement
NLDO	nasolacrimal duct obstruction
NSC or NS	nuclear sclerotic cataract
OD	right eye
OS	left eye
ortho	eyes are aligned on the same target

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Abbreviation	Meaning
OCT	optical coherence tomography
P	periphery
PD	prism diopter
ph or PH	pinhole (crude assessment of best-corrected visual acuity)
PPV or PPVx	pars plana vitrectomy
PVD	posterior vitreous detachment
RAPD	relative afferent pupillary defect (see APD)
RD	retinal detachment
RT	retinal tear
SB	scleral buckle
sc	without correction
SF6	gas bubble injected into vitreous cavity during retina surgery
SLE	slit lamp examination
SO	silicone oil
SRF	subretinal fluid
Ta	applanation tonometry (intraocular pressure measurement)
Tp	tonopen tonometry (intraocular pressure measurement)
V	vessels
Va	visual acuity
VF	visual field testing (formal automated perimetry versus confrontation visual field testing)
X	exophoria at distance
X'	exophoria at near

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Abbreviation	Meaning
XT	exotropia
X(T)	intermittent exotropia at distance
XT'	exotropia at near
X(T)'	intermittent exotropia at near

Evidence Discussion (HD-32.1)

- When evaluating suspected or known issues involving the eye, orbit, and/or brain, consideration must be given to:
  - whether or not imaging is required,
  - which body area should be imaged, i.e., brain, orbits, or both, and
  - which modality, MRI or CT, would best provide the information needed while exposing an individual to the least risk.
- The body area imaged should be reasonably expected to be potentially involved in the suspected condition. The angles of and distance between each view differ between brain imaging and orbital imaging. There are circumstances in which imaging of both the brain and orbits may be useful, as in conditions that can affect both locations or for which evaluation by the different techniques provides useful information.
- Soft tissue detail such as neural tissue is well-visualized by MRI.
- Calcification, bone, and hemorrhages are well-visualized by CT.
- MRI carries no risk of radiation exposure but is sensitive to motion, takes longer, and may require sedation or anesthesia for a longer duration than would be required for CT. Certain populations may have psychological or physical difficulty undergoing MRI scans, including children, those with obesity, movement disorders, anxiety, or claustrophobia.
- CT carries risk of radiation exposure but is less sensitive to motion and has a shorter duration than MRI. Imaging more than one body area increases the exposure dose. Certain populations may carry higher risk of detrimental effects from exposure, including children.
- Radiation exposure of the ocular lens contributes to cataract formation. Radiation doses vary between CT scans due to differences in scanning technique, number of images taken per CT, body area scanned, CT machines used, and facility protocols. The cancer risk of radiation exposure in diagnostic CT is considered extremely low, and the benefit of an appropriately indicated CT examination far outweighs the potential risk.[25] Cataract formation is among the earliest radiation associated pathologies in the eye. The Beaver Dam Eye Study, a population-based study

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of common age-related eye diseases, found that nuclear sclerosis and posterior subcapsular opacity were significantly associated with CAT scans.[25]

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Pupillary Abnormalities Including Horner's Syndrome (HD-32.2)

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- Examination of ocular complaints may include evaluation of pupillary responses, extraocular motility, visual acuity, visual field testing, intraocular pressures, external examination, slit lamp examination and/or funduscopic exam of the retinae. An exam performed by a neuro-ophthalmologist, ophthalmologist, neurologist, or an optometrist meets this requirement.
- Anisocoria and Other Pupillary Disorders
  - Physiologic anisocoria (difference in pupil diameter between the two eyes of typically 2 mm or less) and surgically distorted pupils are not indications for advanced imaging.
  - Dilated pupil from suspected Third nerve palsy (see **Eye Disorders and Visual Loss (HD-32.1)**)
  - Horner's Syndrome (See below)
- Horner's Syndrome (anisocoria, ptosis, and ipsilateral anhidrosis) is caused by disruption of sympathetic innervation to the eye and face. Definitive diagnosis may be established by pharmacologic testing of the pupillary response with eye drops. Evaluation and imaging depends on determining whether the cause is a central lesion (brainstem or cervical spinal cord), pre-ganglionic lesion (spinal cord or sympathetic chain in the chest), or post-ganglionic lesion (neck or carotid artery).
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary for suspected intracranial or brainstem lesions
- MRI Cervical Spine without contrast (CPT® 72141) or MRI Cervical Spine without and with contrast (CPT® 72156) is medically necessary for suspected spinal cord abnormality
- MRI Brachial plexus for Horner syndrome with traction or trauma to the brachial plexus
  - Any **ONE** of the following are medically necessary:
    - MRI Upper Extremity other than joint without contrast (CPT® 73218)
    - MRI Upper Extremity other than joint without and with contrast (CPT® 73220)
    - MRI Chest without contrast (CPT® 71550)
    - MRI Chest without and with contrast (CPT® 71552)
    - MRI Neck without contrast (CPT® 70540)
    - MRI Neck without and with contrast (CPT® 70543)

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- CT Chest with contrast (CPT® 71260) or MRI Chest without and with contrast (CPT®71552) is medically necessary for suspected chest mass
- CT Neck with contrast (CPT® 70491) or MRI Face/Neck/Orbits without and with contrast (CPT®70543) is medically necessary for suspected neck mass
- CTA Neck without and with contrast (CPT® 70498) or MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) is medically necessary for suspected carotid injury or dissection
- MRI Orbits/Face/Neck without contrast (CPT® 70540), MRI Orbits/Face/Neck without and with contrast (CPT® 70543) or CT Orbits/Temporal bone with contrast (CPT® 70481) is medically necessary for suspected orbital lesion or mass

Evidence Discussion (HD-32.2)

- In the evaluation of Horner Syndrome, if a detailed clinical evaluation is unable to localize the site of the lesion, imaging of the entire course of the relevant oculosympathetic pathway is required, as symptoms may result from pathology affecting the nerve fibers at any point along the course of the pathway, requiring multiple imaging modalities.[35] MRI Brain, MRI Orbits/Face/Neck, MRI Cervical Spine and/or MRI Brachial plexus studies may be necessary, depending on the clinical presentation.[1,2,4,35]
- CT Neck is useful to exclude neck masses.[2,35] CT may be complementary to MRI in characterizing skull base erosions, calcifications, and skull base foramina.[35]
- For suspected lung masses associated with Horner syndrome, such as for evaluation of Pancoast tumors, chest imaging is recommended. A mass may be diagnosed on a CT Chest or an MRI Chest.[2,4,24] CT scans provide 60% sensitivity, 65% specificity, and 63% accuracy in defining the local extent of tumor, in contrast to MRI with a sensitivity of 88%, a specificity of 100%, and an accuracy of 94%.[24] MRI of the chest is a more accurate pre-operative examination in identifying the local extent of a Pancoast tumor.[24]
- For suspected carotid injury or dissection, vascular imaging with either CT Angiography (CTA) neck or MR Angiography (MRA) neck is medically necessary, depending on the individual's risk and benefit profile.[2,4,6]

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Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33)

Guideline

Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)
References (HD-33)

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Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)

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- Acoustic neuroma and vestibular schwannoma may be used interchangeably
- Initial diagnosis is usually made during evaluation for asymmetric hearing loss and/or vertigo (see **Dizziness, Vertigo and Syncope (HD-23)** and **Hearing Loss and Tinnitus (HD-27)**) for evaluation of those problems)
- MRI Brain without and with contrast (CPT® 70553), which should be done with attention to the internal auditory canals, is medically necessary for initial diagnosis.
- MRI Brain without contrast (CPT® 70551) is medically necessary if performed with FIESTA protocol
- MRI Orbits/Face/Neck without and with contrast (CPT® 70543) with audiologic or clinical features of retrocochlear hearing loss and a negative MRI Brain and in the rare individual in whom a detailed search is medically necessary for both a lesion of the cerebellopontine angle **and** lesions of the cerebral hemispheres
- Repeat MRI Brain (contrast as requested) is medically necessary 6 months after diagnosis, then annually for 5 years and thereafter per neurologist, neurosurgeon, or otolaryngologist, or any provider in consultation with a neurologist, neurosurgeon, or otolaryngologist.
- MRI Brain without and with contrast with attention to the internal auditory canals (CPT® 70553) is medically necessary after surgical resection and following stereotactic radiation therapy at 6 to 12 months to document the completeness of tumor removal and to serve as a baseline for further follow-up. Additional follow-up is done annually for 5 years and every 2 years thereafter.
- Limited MRI of the internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) is medically necessary when requested by the provider in place of a complete MRI Brain. Note: Limited MRI codes should not be used in addition to MRI Brain codes; IAC views are performed as additional sequences as part of the brain study. (See **General Guidelines – Anatomic Issues (HD-1.1)**)
- See **Primary Central Nervous System Tumors- General Considerations (ONC-2.1)** in the Oncology Imaging Guidelines for additional imaging requests for surgery

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Evidence Discussion (HD-33)

- MRI Brain is the preferred initial imaging modality for evaluation of persistent vertigo, vertigo associated with an abnormal neurologic exam, vertigo due to a suspected central cause, pulsatile or asymmetric tinnitus, and/or hearing loss.[8,9]
- CT Head is not recommended for the initial evaluation of suspected acoustic neuroma due to inferior soft tissue resolution when compared to MRI Brain. In addition, MRI Brain provides better visualization of the cerebellum, posterior fossa, and cranial nerves.[8]
- MRI Brain, in this clinical scenario, is performed using specialized internal auditory canal (IAC) protocols, which include thin-section sequences to evaluate for vascular loops and small vestibular schwannomas.[10]
- MRI Brain can diagnose lesions in the cerebellopontine angle including schwannoma, meningioma, and other posterior fossa tumors.[10]
- 3D-Fast imaging employing steady state acquisition (3D-FIESTA) demonstrates significantly higher spatial resolution with superior imaging contrast between cranial nerves and CSF with a shorter acquisition time than conventional MRI scan.[4]
- Follow-up imaging is recommended 6 months after diagnosis to evaluate for rapid growth, then annually for 5 years.[3,6,7] After 5 years, tumor growth that has remained stable is unlikely but may still occur, therefore, lifelong surveillance is advised with longer imaging intervals.[7]
- Follow-up imaging after surgical resection and/or stereotactic radiosurgery to assess residual tumor and treatment response is performed at 6-12 months with additional follow-up annually for 5 years and every 2 years thereafter.[5,6]

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Pineal/Colloid Cysts (HD-34)

Guideline

Pineal/Colloid Cysts (HD-34.1)
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Pineal/Colloid Cysts (HD-34.1)

HD.PT.0034.1.A
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Pineal Cysts

Pineal cysts are generally discovered incidentally and do not require surgical intervention.

- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary for initial evaluation of pineal cysts if not already completed.
- Repeat MRI Brain is not medically necessary for most individuals with pineal cysts, but MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary for the following:
  - New or worsening headache or focal neurologic deficits suggesting progression of cyst
  - Pre-operative planning

Colloid Cysts

- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary for the initial evaluation of colloid cysts if not already completed.
- Repeat MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is medically necessary for colloid cysts for the following:
  - In the presence of symptoms including syncope
  - Evaluation of CSF flow (CPT® 70551)
  - When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon

Evidence Discussion (HD-34)

- MRI Brain is the preferred modality for the evaluation of intracranial cysts, due to its superior soft tissue resolution when compared to CT Head.[1,4]
- Follow-up imaging of pineal cysts is supported for new or worsening headaches, focal neurologic deficits, and/or for surgical planning, otherwise, routine follow up is not supported.[2,3]
- In contrast to pineal cysts, colloid cysts may lead to sudden obstruction of cerebrospinal fluid flow at the foramen of Monro, resulting in neurologic symptoms, including syncope. Other than this scenario, follow-up imaging indications are similar to pineal cysts.[1,5]

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Arachnoid Cysts (HD-35)

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Arachnoid Cysts (HD-35.1)

HD.AR.0035.1.A
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Arachnoid cysts arise in the middle or posterior fossa, and the majority of lesions are discovered incidentally and do not require surgical intervention.

- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is medically necessary for initial evaluation of arachnoid cysts if not already completed.
- Repeat MRI Brain is not medically necessary for most individuals with arachnoid cysts, except in the following scenarios:
  - New or worsening headache or focal neurologic deficits suggesting progression of cyst
  - Pre-operative planning
  - When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon

Evidence Discussion (HD-35)

- MRI Brain is the preferred modality for evaluation and follow up of intracranial arachnoid cysts, due to its superior soft tissue resolution when compared to CT Head.[1,3]
- Most intracranial arachnoid cysts remain asymptomatic and follow up imaging is not routinely supported. Surgical intervention is reserved for those with symptoms.[1,2]

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Nuclear Medicine (HD-36)

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Nuclear Medicine (HD-36.1)
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Nuclear Medicine (HD-36.1)

HD.NM.0036.1.A
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Nuclear medicine studies used in the evaluation of some head/brain disorders, and other rare indications as well.

- Brain Scintigraphy with or without vascular flow (any one of CPT® 78600, CPT® 78601, CPT® 78605, or CPT® 78606)
- Brain Imaging Radiopharmaceutical Localization SPECT (CPT® 78803)
  - Immunocompromised individuals with mass lesion detected on CT or MRI for differentiation between lymphoma and infection
  - In distinguishing recurrent brain tumor from radiation necrosis
  - Can be performed with vasodilating agent acetazolamide (Diamox) to assess functional reserve capacity to predict critically reduced perfusion in individuals with chronic cerebrovascular disease (for example, in Moya-Moya disease) and identify individuals who might benefit from an extracranial-to-intracranial (EC-IC) bypass to augment Cerebral Blood Flow, and to assess pre-operatively the potential for ischemia following carotid artery sacrifice. See **Moyamoya Syndrome/Disease (HD-21.5)**
- Brain Imaging Vascular Flow (CPT® 78610)
  - Cerebral ischemia
  - Establish brain death
- CSF Leakage Detection (CPT® 78650)
  - Evaluation of CSF rhinorrhea, otorrhea, or refractory post-lumbar puncture headache
  - Suspected normal pressure hydrocephalus with gait disturbance and either dementia or urinary incontinence
- Radiopharmaceutical Dacryocystography (CPT® 78660)
  - Suspected obstruction of nasolacrimal duct due to excessive tearing
- Cisternogram (CPT® 78630), see **Low-Pressure Headache and CSF Leak (HD-11.15)** and **Facial Trauma (HD-13.2)**
- Cerebrospinal Ventriculography (CPT® 78635) and Nuclear Medicine Shunt Evaluation (CPT® 78645) and CSF Flow SPECT (CPT® 78803), see **Hydrocephalus Shunts (HD-11.14)**
- DAT-SPECT **OR** Fluorodopa F (F-DOPA) PET Brain see **Lewy Body Dementia (LBD) – SPECT Brain Scan and PET (HD-8.3)** and **Movement Disorders (HD-15.1))**
- Jaw Asymmetry - see **Temporomandibular Joint Disease (HD-30.1)**

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Evidence Discussion (HD-36)

- Nuclear medicine studies are adjunct, functional imaging modalities in the evaluation of a variety of neurologic conditions and are generally helpful when structural brain imaging modalities are unable to provide answers to complex clinical questions.
- The American Academy of Neurology practice guideline recommends brain SPECT to assist with prognostication in adults with traumatic brain injuries.[4]
- Single-photon emission CT (SPECT) is the most readily available nuclear medicine technique for assessment of cerebral hemodynamics.[2] Brain SPECT has also been validated for evaluation of cerebrovascular reactivity to acetazolamide (ACZ) challenge in individuals with various types of vaso-occlusive disease, including Moyamoya disease.[2,7]
- Brain SPECT has been used in the evaluation of stroke, TIA, monitoring of medical or surgical therapy, assessment of cerebral blood flow reserve, and estimation of prognosis. The sensitivity of brain SPECT for acute stroke localization is 85.5% with a specificity of 97.6%.[6]
- Brain SPECT used with specific perfusion agents is useful in distinguishing radiation effects from residual or recurrent tumor, and/or for distinguishing cerebral lymphoma from infection, a distinction not always possible with CT or MRI.[6]
- Dacryocystography is supported to localize the lacrimal drainage system and evaluate for suspected obstruction in the nasolacrimal duct.[9]
- Radionuclide CSF shunt studies can evaluate shunt patency, differentiate proximal versus distal obstruction, and localize the site of obstruction. The combination of CT and radionuclide is more sensitive than CT alone in diagnosing shunt malfunction.[8]
- CT head cisternography in the evaluation of CSF leak has a sensitivity range between 85%-92%. This modality is particularly useful when there are multiple potential CSF leak sites. Radionuclide cisternography is useful for confirming the presence of a CSF leak.[5]

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Sleep-Related Imaging (HD-37)

Guideline

General Guidelines Sleep-Related Imaging (HD-37.1)
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General Guidelines Sleep-Related Imaging (HD-37.1)

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- Hypersomnolence:
  - When there are focal neurologic signs or suspicion for an inflammatory neurologic process as the etiology. Recognition and treatment of a comorbid sleep disorders is paramount, and a complete neurologic history and examination should precede any request for advanced imaging.
    - MRI Brain without and with contrast (CPT® 70553) **OR**
    - MRI Brain without contrast (CPT® 70551)
- Central Sleep Apnea:
  - For unexplained central sleep apnea syndrome when a primary CNS etiology is suspected, i.e., unassociated with CHF, COPD, or other potential etiology. Specific etiologies should be stated for imaging requests, including but not limited to, suspected Chiari malformation, stroke, CNS demyelinating disease, posterior fossa lesion, anoxia, or infection.
    - MRI Brain without and with contrast (CPT® 70553) **OR**
    - MRI Brain without contrast (CPT® 70551)
- Obstructive Sleep Apnea (OSA):
  - Advanced imaging is not medically necessary in the evaluation and management of obstructive sleep apnea.
  - The gold standard for OSA diagnosis, evaluation and management is a sleep study.
  - See **Initial Sleep Diagnostic and Treatment Testing (SL-2.1)**
- Oral Appliance:
  - There is a lack of published case-controlled clinical studies in Sleep literature validating the use of advanced imaging with respect to oral appliance therapy (pre-treatment assessment).
  - Previous literature has demonstrated support for cephalometric studies (x-ray) in predicting treatment success.
  - Nasoendoscopy (sedated and non-sedated with provocative maneuvers such as Mueller maneuver) has been helpful as well in this regard.
  - Routine use of advanced imaging is not medically necessary at this time.
- Upper Airway Surgery:

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  - There is a lack of published case-controlled clinical studies in sleep literature validating the use of advanced imaging for upper airway surgery (pre-treatment assessment).
    - Examples of upper airway surgery include, but are not limited to, hypoglossal nerve stimulation, uvulopalatopharyngoplasty (UPPP), turbinate reduction, and nasal septoplasty
  - For congenital anomalies, see **Craniosynostosis Imaging (PEDHD-8.1)** and **Facial Anomalies (PEDHD-8.2)**
  - For dentofacial abnormalities, see **Dental/Periodontal/Maxillofacial Imaging (HD-30.2)**
  - Routine use of advanced imaging is not supported at this time.
- For suspected sleep-related seizure imaging (see **Epilepsy and Other Seizure Disorders (HD-9)**)

Evidence Discussion (HD-37.1)

- An individual's management is rarely impacted by structural brain imaging in the evaluation of unexplained hypersomnolence. Instead, a thorough evaluation can result in an accurate diagnosis while safeguarding individuals from unnecessary exposure to radiation and over-reliance on incidental imaging findings as potential contributors to the symptom(s).
- Literature supports use of drug-induced sleep endoscopy (DISE) to predict effectiveness of upper airway surgery, particularly in individuals seeking upper airway stimulation or maxillomandibular advancement.[16,17]
- Studies have demonstrated how upper airway MRI and CT can be used to determine sites of collapse in OSA patients.[14,15] Information from imaging could assist with determining which surgical interventions should be considered. Additionally, imaging may be helpful to determine why upper airway surgery is ineffective post-surgically.[14] However, there are no large clinical trials that have shown improved surgical outcomes utilizing data from pre-treatment advanced imaging for OSA.
- The appropriate step in care for individuals with disordered sleep is to evaluate their breathing with polysomnography. Advanced imaging can lead to gaps in care and ineffective treatment of disordered sleep patterns. Instead, emphasis should be placed on holistic evaluation, including sleep history and sleep testing. Radiography (x-rays), 3D Advanced Imaging, or dynamic nasopharyngoscopy are not supported by evidence to being superior over polysomnography at this time.

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Procedure Codes Associated with Musculoskeletal Imaging (MS)

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MRI/MRA	CPT®
MRI Upper Extremity, other than joint, without contrast	73218
MRI Upper Extremity, other than joint, with contrast	73219
MRI Upper Extremity, other than joint, without and with contrast	73220
MRI Upper Extremity, any joint, without contrast	73221
MRI Upper Extremity, any joint, with contrast	73222
MRI Upper Extremity, any joint, without and with contrast	73223
MR Angiography Upper Extremity without or with contrast	73225
MRI Lower Extremity, other than joint, without contrast	73718
MRI Lower Extremity, other than joint, with contrast	73719
MRI Lower Extremity, other than joint, without and with contrast	73720
MRI Lower Extremity, any joint, without contrast	73721
MRI Lower Extremity, any joint, with contrast	73722
MRI Lower Extremity, any joint, without and with contrast	73723
MR Angiography Lower Extremity without or with contrast	73725
MRI Pelvis without contrast	72195
MRI Pelvis with contrast	72196

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MRI/MRA	CPT®
MRI Pelvis without and with contrast	72197

CT/CTA	CPT®
CT Upper Extremity without contrast	73200
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CT Lower Extremity without contrast	73700
CT Lower Extremity with contrast	73701
CT Lower Extremity without and with contrast	73702
CT Angiography Lower Extremity without and with contrast	73706
CT Pelvis without contrast	72192
CT Pelvis with contrast	72193
CT Pelvis without and with contrast	72194
Bone Mineral Density CT, one or more sites, axial skeleton	77078

Ultrasound	CPT®
Ultrasound, complete joint (ie, joint space and peri-articular soft tissue structures) real-time with image documentation	76881
Ultrasound, limited, joint or other nonvascular extremity structure(s) (e.g., joint space, peri-articular tendon[s], muscle[s], nerve[s], other soft tissue structure[s], or soft tissue mass[es]), real-time with image documentation	76882

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Ultrasound	CPT®
Ultrasound, pelvic (nonobstetric), real time with image documentation	76857

Nuclear Medicine	CPT®
Bone Marrow Imaging, Limited	78102
Bone Marrow Imaging, Multiple	78103
Bone Marrow Imaging, Whole Body	78104
Bone or Joint Imaging Limited	78300
Bone or Joint Imaging Multiple	78305
Bone Scan Whole Body	78306
Bone Scan 3 Phase Study	78315
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, single area (e.g., head, neck, chest, pelvis), single day imaging	78800
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, 2 or more areas (eg, abdomen and pelvis, head and chest), 1 or more days imaging or single area imaging over 2 or more days	78801
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, single day imaging	78802
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), single area (e.g., head, neck, chest, pelvis), single day imaging	78803

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Nuclear Medicine	CPT®
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (e.g., head, neck, chest, pelvis), single day imaging	78830
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), minimum 2 areas (e.g., pelvis and knees, abdomen and pelvis), single day imaging, or single area imaging over 2 or more days	78831
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, minimum 2 areas (e.g., pelvis and knees, abdomen and pelvis), single day imaging, or single area imaging over 2 or more days	78832

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General Guidelines (MS-1.0)

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General Criteria

Information regarding the purpose, development, management and review/revision of these guidelines can be found in the **Preface to the Imaging Guidelines** (the Preface). The Preface provides guidance on the clinical documentation and information that should be submitted in order to establish the medical necessity of advanced imaging. The Preface also contains information on special considerations that may be pertinent to criteria-based reviews for advanced imaging (e.g., pediatric vs. adult, gender identity, health plan policies, national or local mandates). Providers should review and be familiar with all information in the Preface.

These musculoskeletal (MSK) imaging guidelines are intended for the further investigation of non-spinal, MSK conditions or concerns (e.g., shoulder, elbow, wrist, hand, arm, pelvis, hip, knee, ankle, leg, foot). Imaging guidelines for spinal conditions or concerns are located in **Spine Imaging Guidelines.**

Before any request for advanced diagnostic imaging of musculoskeletal concerns will be considered ALL of the following must be complete and clearly documented. The following criteria apply to all requests for advanced imaging, details, or exceptions, will be provided within each specific section throughout the guideline:

- There must be an in-person clinical evaluation, as well as a clinical re-evaluation following any required conservative care as outlined throughout the guideline.
- An in-person clinical evaluation that includes a complete relevant medical history, physical examination, appropriate laboratory studies and initial non-advanced imaging (e.g., plain radiographs) should have been completed after the current episode of symptoms began. The in-person clinical evaluation may be, at least, either the initial or the re-evaluation.
    - Prior to advanced imaging consideration, the results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the provider that is requesting the advanced imaging.
- When necessary, a failed trial of provider directed conservative treatment following the initial clinical evaluation and non-advanced imaging may be required:
    - The trial of conservative care lasting at least six weeks in duration
    - A clinical re-evaluation or documentation of other meaningful contact (e.g., email, telephone, video consult) with the provider's office following the initial evaluation

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- Details will be in the specific criteria sections throughout this publication regarding when conservative treatment is required.

The need for repeat or serial advanced imaging, whether CT or MRI, should be considered carefully and may not be medically necessary. This includes serial imaging for surveillance of healing or recovery from musculoskeletal disease. In the majority of musculoskeletal conditions, the medical evidence (i.e., documentation) does not support the need for repeat imaging.

Health Equity Considerations

Health equity is the highest level of health for all people; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which people are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include safe housing, transportation, and neighborhoods; racism, discrimination and violence; education, job opportunities and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Evidence Discussion (MS-1)

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Advanced imaging is typically not necessary for the initial evaluation of individuals with a musculoskeletal complaint. A diagnosis can often be made based on a detailed history, physical examination, appropriate laboratory tests and/or plain radiographs or initial ultrasound (US).[1-5] Many individual's complaints can improve within a few weeks to months with conservative care.

The American College of Radiology Appropriateness Criteria recommended that x-rays, or US in a few select scenarios, as the initial imaging modality for most musculoskeletal complaints.[1,3] These initial images can be helpful in determining if a course of conservative treatment is recommended. In some instances, the initial x-rays or US may help determine a diagnosis or that more advanced images or procedures are appropriate.[6] If conservative treatment is unsuccessful or more advanced study is needed the initial images can be helpful in determining the next course of action.[6,7] These initial images can be helpful in pre-operative planning when surgery is being considered.[1,3]

Advanced imaging can often demonstrate abnormalities that have no relevance to the patient's symptoms. Studies have shown that advanced imaging of asymptomatic middle age individuals commonly demonstrate musculoskeletal abnormalities that do not affect their quality of life.[8] Focusing on incidental findings can lead to overtreatment through unnecessary referral to medical specialists or unneeded medical procedures. Advanced imaging can have additional risks to the individual beyond those of initial plain images. There are risks associated with additional exposure to radiation. There can also be complications due to implantable devices or metallic foreign bodies in the individual. Risk mitigation needs to be weighed when advanced imaging is being considered, especially in relation to repeat or serial advanced imaging.[9]

In many instances for musculoskeletal complaints, current evidence has supported the recommendation of obtaining initial plain x-rays, or US when appropriate, and having the individual follow a course of provider directed conservative care prior to seeking advanced imaging studies.[1] The potential clinical benefit of starting conservative care and not needing advanced images or additional procedures may outweigh the potential harm of a brief delay of obtaining said imaging. It may also help to reduce the risk of unnecessary exposures to radiation/chemicals associated with the advanced imaging techniques.[7,9]

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Imaging Techniques (MS-2)

Guideline

Imaging Techniques (MS-2)

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Imaging Techniques (MS-2)

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Plain Radiograph

Plain radiographs (x-ray) are often the initial imaging for musculoskeletal conditions. The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the provider requesting advanced imaging, unless otherwise noted, to rule out those situations that do not often require advanced imaging, such as: osteoarthritis, acute/healing fracture, dislocation, osteomyelitis and bone tumor amenable to biopsy or radiation therapy (in known metastatic disease). Unless otherwise noted in the guidelines, x-rays should be performed before requesting advanced imaging.

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is often a preferred advanced imaging modality in MSK conditions because it can be superior in visualizing the soft tissues and physiological processes happening due to an injury or medical condition. As MRI uses magnetic fields, it does not expose the individual to ionizing radiation and may be a better choice when exposure is a concern. However, individuals with implanted devices may not be able to undergo MRI.

Currently there is inadequate scientific evidence to support the necessity of positional MRI (also referred to as dynamic, weight-bearing, standing or kinetic MRI) and delayed gadolinium enhanced MRI of cartilage (dGEMRIC). These imaging modalities should be considered not medically necessary.

Computed Tomography

Computed tomography (CT) is preferred for imaging cortical bone anatomy. It is useful for studying complex fractures, dislocations, and assessing delayed union or non-union of fractures when x-rays are equivocal. CT can also be the imaging choice in individuals who cannot undergo an MRI.

Positional CT (also referred to as weight-bearing or cone beam CT) may be useful in imaging of the foot and ankle. Requests for positional CT for the foot and ankle are subject to the same condition-specific criteria as contained throughout this guideline publication. Positional CT of anatomic areas other than the foot and ankle are considered not medically necessary.

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Ultrasound

Ultrasound (US) uses sound waves to produce images that can be used to evaluate a variety of musculoskeletal disorders. US is highly operator dependent and proper training and experience are required to perform consistent, high-quality evaluations.

Nuclear Medicine

A bone scan is a nuclear medicine imaging study in which an amount of radioactive material is injected, and images are obtained at different time intervals, depending on the condition. A bone scan is done to reveal problems with bone metabolism. Areas where bone cells are repairing themselves show the most activity. It can help diagnose a number of bone conditions, including cancer of the bone or metastasis, location of bone inflammation, fracture, and bone infection.

Nuclear Medicine WBC Scan is performed using radioactive material which is tagged to the white blood cells. When injected into the body, the material attaches to sites of inflammation/infection. Once distributed in these areas, the sites of suspected infection/ inflammation can be seen on nuclear imaging equipment. These can be imaged as a planar study, SPECT study, or SPECT/CT study.

Bone Marrow Imaging is used in combination with a WBC Scan to help differentiate between true infection and physiological marrow uptake. The bone marrow scan provides a map of the normal physiological white cell uptake that is then compared to the white blood cell scan. Any discordance in white cell uptake (e.g., more WBC uptake than marrow uptake) between the two studies indicates a focus of infection.

Positron Emission Tomography

Positron Emission Tomography (PET/CT) is a nuclear medicine/CT fusion study that uses a positron emitting radiotracer to create cross-sectional and volumetric images based on tissue metabolism. PET/CT allows for better anatomic localization of the areas of abnormal increased tissue activity. PET/CT can be helpful for imaging certain musculoskeletal conditions when MRI or CT is equivocal or cannot be performed.

- See: **Nuclear Medicine (MS-28)** and condition specific guidelines for specific indications.

Contrast Issues

Most musculoskeletal imaging (MRI or CT) is completed without contrast media; however, contrast may be useful and will be indicated throughout the condition-specific criteria in these guidelines. Some examples may be tumors, osteomyelitis, infection, arthrograms, detection of rheumatoid arthritis. For individuals with contrast contraindication, the corresponding advanced imaging without contrast may be approved as an alternative. However, in some cases the non-contrast study may not

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provide as adequate evaluation of the condition of concern and would therefore not be necessary.

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3D Rendering (MS-3)

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3D Rendering (MS-3.1)

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3-D image post-processing of conventional images is medically necessary for preoperative planning when conventional imaging is insufficient for the following:

- Complex fractures (comminuted or displaced)/dislocations of any joint
- Spine fractures, pelvic/acetabulum fractures, intra-articular fractures.
- Preoperative planning for other complex surgical cases.

The code assignment for 3-D rendering depends upon whether the 3-D post-processing is performed on the scanner workstation (CPT® 76376) or on an independent workstation (CPT® 76377)

- 2-D reconstruction (i.e., reformatting axial images into the coronal plane) is considered part of the tomography procedure, is not separately reportable, and does not meet the definition of 3-D rendering.
- It is not medically necessary to report 3-D rendering in conjunction with CTA and MRA because those procedure codes already include the post-processing.
- In addition to the term "3-D," the following terms may also be used to describe 3-D post-processing:
  - Maximum intensity projection (MIP)
  - Shaded surface rendering
  - Volume rendering
- Additionally - If multiple CPT codes are performed for the same indication on the same day, one 3D rendering code is required. If they are performed on separate days, 3D rendering codes are required for each study on each day.
- The 3-D rendering codes require concurrent supervision of image post-processing 3-D manipulation of volumetric data set and image rendering.

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Evidence Discussion (MS-3)

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In some complex clinical scenarios, initial and conventional advanced images alone may not sufficiently reveal bony displacement and other incongruities. Accurate radiographic characterization of complex fractures and injuries is important for appropriate treatment decisions. The American College of Radiology, in multiple of their Appropriateness Criteria (ACR) publications, supports the use of 3D image post-processing of conventional images of complex fractures and injuries when said images are insufficient to visualize bone displacement and/or incongruity, and further clarification will aid in treatment decisions. [1-3]

Evidence has shown that 3D modeling can improve the accuracy of fracture classification. In a cohort study comparing the use of 2D CT images alone or combined with 3D segmentation or 3D models, both trauma surgeons and residents were more accurate with their classification of proximal humerus fractures when they used digital 3D models.[4] In a systematic review, inter- and intra-observer agreement in proximal humeral fracture classification increased as image technology advanced from radiographs to 2D CT to 3D CT.[5] Fracture gaps of 100µm were identified at a consistent rate of 80-100% in CT images with displacements of up to 400µm visible in digital 3D models. However, rates were dependent on CT technology and post-processing effects of rendering, thus original CT images should still be used along with any 3D rendering for verification of fractures prior to surgery.[6]

In their Appropriateness Criteria for acute trauma to the hand and wrist, the ACR supports the use of 3D rendering as a visualization tool for more accurate preoperative planning in the management of some complex surgical procedures.[7] Additional evidence demonstrated the utility of 3D rendering in preoperative planning for various complex musculoskeletal conditions. In a retrospective study of the planning of reinforcement cages for total-hip replacement revision surgery 3D templating accurately predicted cage size in 29/27 surgeries, compared to only 15/27 using conventional 2D templating.[8] 3D preoperative planning for reverse total shoulder arthroplasty has been shown to result in minimal deviation of the procedure from the plan with high levels of agreement in screw angle, length and prosthetic component size.[9] In a comparison of 2D versus 3D preoperative planning in total hip arthroplasty, 3D templating was found to result in higher accuracy restoration of native joint mechanics.[10]

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Nuclear Medicine (MS-28)

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Nuclear Medicine (MS-28.1)

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As mentioned in General Guidelines (MS-1.0), results of plain x-rays after the current episode of symptoms started or changed must be available to the requesting provider, unless otherwise specified below, prior to advanced imaging. A trial of conservative care may also be required, as specified below.

NOTE: SPECT scan may be medically necessary for any of the same criteria for which a bone scan is medically necessary as listed in this section if:

- The request is for CPT® 78300 and CPT® 78803, then only CPT® 78803 is to be approved if medical necessity is established.
- The request is for CPT® 78305 or CPT® 78306 and CPT® 78803, then two CPT® codes may be approved if medical necessity is established.

Nuclear Medicine may be medically necessary in the evaluation of some musculoskeletal disorders, and other rare criteria as follows:

- Evaluation of suspected aseptic loosening of orthopedic prostheses when recent plain x-ray is nondiagnostic:
  - Bone scan (CPT® 78315) OR
  - Distribution Of Radiopharmaceutical Agent SPECT (CPT® 78803, or 78831) OR
  - Hybrid SPECT/CT (CPT® 78830)
  - Criteria for non-nuclear medicine imaging can be found in: **Joint Replacement Post-Operative Follow-Up (MS-16.1)**
- For detection of ischemic or infarcted regions in sickle cell disease:
  - Nuclear medicine bone marrow imaging (CPT® 78102, 78103, or 78104) OR
  - SPECT (CPT® 78803) OR
  - Hybrid SPECT/CT (CPT® 78830)
  - See also: **Modality General Considerations (PEDMS-1.3)**
- Evaluation of complex regional pain syndrome or reflex sympathetic dystrophy, after failure of six weeks provider-directed conservative treatment, including re-evaluation:
  - Triple phase bone scan (CPT® 78315)
  - Criteria for non-nuclear medicine imaging of CRPS of the foot can be found in: **General Pain (MS-32.1)**
- Evaluation of Paget's disease
  - Bone scan (CPT® codes: 78300, 78305, or 78306) OR
  - Distribution Of Radiopharmaceutical Agent SPECT (CPT® 78803) OR

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- Hybrid SPECT/CT (CPT® 78830)
- Criteria for non-nuclear medicine imaging can be found in: **Bone Lesion (MS-10.2)**

- Suspected fractures
  - If the following criteria listed below (as also found in **Occult, Stress or Insufficiency Fracture or Reaction (MS-5.2)**) (**excluding peri-prosthetic fractures**) are met:
    - For suspected occult/stress/insufficiency fractures NOT related to bisphosphonate use:
      - Initial plain x-rays obtained a minimum of 14 days after the onset of symptoms are non-diagnostic for fracture OR
      - Repeat plain x-rays remain non-diagnostic for fracture after a minimum of 10 days of provider-directed conservative treatment
      - For suspicion of shin splints, a 6-week trial of provider-directed conservative treatment, including clinical re-evaluation, is also required prior to requesting advanced imaging.
      - For stress reaction surveillance or "return to play" decisions, advanced imaging is not medically necessary regarding a stress reaction identified on an initial imaging study.
      - For stress fracture follow-up imaging for "return to play" evaluation at least 3 months after the initial imaging study, MRI without contrast of the area of interest.
    - Or for suspected occult/stress/insufficiency fractures of the **hip/femoral neck, tibia, pelvis/sacrum, tarsal, navicular, proximal fifth metatarsal, or scaphoid**, and **atypical femoral shaft** fractures related to bisphosphonate use when ALL of the following apply:
      - The initial evaluation, history, and physical exam fails to establish a definitive diagnosis AND
      - The initial plain x-ray findings are negative or equivocal
      - CT without contrast can be performed as an alternative to MRI for suspected occult/insufficiency fractures of the pelvis/hip and suspected atypical femoral shaft fractures related to bisphosphonate and suspected occult fractures of the scaphoid
  - **And MRI cannot be performed**, the following imaging is medically necessary:
    - Tc-99m bone scan whole-body (CPT® 78306) with SPECT of the area of interest (CPT® 78803) OR
    - Hybrid SPECT/CT (CPT® 78830) OR
    - Bone scan (CPT® 78315, 78305, or 78300)

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- **Or for peri-prosthetic fractures when MRI cannot be performed**, the following imaging is medically necessary:
  - Tc-99m bone scan whole-body (CPT® 78306) with SPECT of the area of interest (CPT® 78803) OR
  - Hybrid SPECT/CT (CPT® 78830) OR
  - Bone scan (CPT® 78315, 78305, or 78300)

- Evaluation of suspected bone infection if MRI or CT cannot be done and when infection is multifocal, or when the infection is associated with orthopedic hardware or chronic bone alterations from trauma or surgery:
  - **Note** - For suspected infection following joint replacement, joint aspiration is the initial evaluation after plain x-ray for a painful joint replacement. Joint aspiration should be negative or inconclusive prior to requesting advanced imaging.
    - Criteria for non-nuclear medicine imaging regarding specific joints post-operative from replacement surgery can be found in: **Joint Replacement Post-Operative Follow-Up (MS-16.1)**
  - FDG PET/CT (CPT® 78815 for multifocal infection, or CPT® 78811 for unifocal/ limited area of interest) if MRI or CT is equivocal or cannot be done
    - At this time, FDG is the only indicated radiotracer for use with PET/CT in the imaging of musculoskeletal conditions
  - Bone scan (CPT® 78315, 78300, 78305, or 78306) OR
  - Distribution of radiopharmaceutical agent SPECT (CPT® 78803 or 78831) OR
  - Hybrid SPECT/CT (CPT® 78830 or 78832)
  - A labeled leukocyte scan (radiopharmaceutical inflammatory imaging - one of CPT® codes: 78800, 78801, 78802, or 78803) in concert with Tc-99m sulfur colloid marrow imaging (one of CPT® codes: 78102, 78103, or 78104)

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Evidence Discussion (MS-28)

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Many musculoskeletal complaints can be diagnosed based on a detailed history, physical examination and initial x-rays. In more complex clinical situations, x-rays can be useful in determining whether advanced diagnostic imaging is needed and which specific advanced diagnostic imaging study is warranted. MRI or CT are typically the study of choice if x-rays are non-diagnostic or equivocal in evaluating musculoskeletal complaints. While nuclear medicine scans may demonstrate high sensitivity to certain diagnoses, the increased exposure to radiation must be considered in comparison to other advanced imaging options.[1,2]

The American College of Radiology (ACR) has recommended the use of MRI, when possible, as the next image after x-rays for investigation of suspected infection, stress fracture and post-surgical complications following joint replacement. MRI can be as sensitive as nuclear medicine scans but also considerably more specific, while creating less radiation exposure. However, there may be situations where MRI or CT are not available, contraindicated or image results would be unclear. [1, 3-7]

Implanted hardware, devices, or foreign objects may create image artifacts making diagnosis uncertain. In these situations, nuclear medicine scans may be the next best option in imaging. Practical planning to keep radiation exposure as low as possible must be taken into account.[2] Bone scans have been shown to be as sensitive to stress fracture as MRI, allowing earlier detection than may be shown on plain x-ray.[3] When an alternative imaging modality to other advanced imaging is necessary to evaluate suspected infection, bone scans have demonstrated high sensitivity to osteomyelitis. However, their specificity has been shown to be limited (45%), especially when there are other underlying conditions (e.g., arthritis), recent fracture, or recent surgery.[4,5] Bone scan may also have similar sensitivity and specificity as MRI when evaluating suspected complex regional pain syndrome, type 1 and may be useful to rule-out the diagnosis.[8]

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Arthritis and Joint Degeneration (MS-12)

Guideline

Osteoarthritis (MS-12.1)
Inflammatory Arthritis (MS-12.2)
Evidence Discussion (MS-12)
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Osteoarthritis (MS-12.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning of osteoarthritis. See specific details below regarding any requirements for conservative care prior to requesting advanced imaging.

Non-surgical and surgical treatment planning, other than joint replacement

- Osteoarthritis - CT without contrast for further investigation of osteoarthritis is medically necessary when ALL of the following apply:
  - Requested for treatment planning, AND
  - Congenital or significant atypical post-traumatic arthritic deformities are identified on plain x-ray, AND
  - The aforementioned deformities require further evaluation of their clinical significance, AND
  - The request is related to the shoulder, elbow, wrist, hip, knee, or ankle
- Reconstructive joint sparing/salvage surgery planning - MRI arthrogram or CT arthrogram for joint sparing/salvage reconstructive surgery planning is medically necessary for the following:
  - Suspected concomitant rotator cuff tear of the shoulder
  - Suspected concomitant labral tear of the shoulder or hip
  - Suspected concomitant internal derangement of the knee

Joint replacement planning, non-customized-to-patient (not for intra-operative navigation)

- **Elbow, wrist, hip, knee or ankle** - CT without contrast of the elbow (CPT® 73200), wrist (CPT® 73200), hip (CPT® 73700 or 72192), knee (CPT® 73700) or ankle (CPT® 73700) for non-customized-to-patient joint replacement planning (prior to surgery/not for intra-operative navigation) is medically necessary when ALL of the following apply:
  - Initial x-rays have been performed after the current episode of symptoms started or changed and results are available to the requesting provider AND
  - Congenital or significant atypical post-traumatic arthritic deformities are identified on plain x-ray AND
  - The aforementioned deformities require further evaluation of their clinical significance

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- **Shoulder** - CT without contrast (CPT® 73200) and/or MRI without contrast (CPT® 73221) of the shoulder for non-customized-to-patient joint replacement planning (prior to surgery/not for intra-operative navigation) is medically necessary when ALL of the following apply:
  - Initial x-rays have been performed after the current episode of symptoms started or changed and results are available to the requesting provider

Joint replacement planning and intraoperative navigation, customized-to-patient

- CT without contrast or MRI without contrast of the **shoulder, elbow, wrist, hip, knee, or ankle** for customized-to-patient joint replacement surgery planning and intraoperative navigation (e.g., MAKOplasty) is medically necessary when ALL of the following apply:
  - Initial x-rays have been performed after the current episode of symptoms started or changed and results are available to the requesting provider AND
  - The joint replacement surgery has been approved or does not require prior authorization
- The preoperative imaging listed above is considered not medically necessary if any of the following are deemed not medically necessary, not a covered benefit, or experimental, investigational, or unproven by the health plan:
  - Joint replacement surgery
  - Customized-to-patient implant
  - Computer assisted surgical navigation (e.g. MAKOplasty)
- See: Unlisted **Procedures/Therapy Treatment Planning (Preface-4.3)** in the Preface Imaging Guidelines

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Inflammatory Arthritis (MS-12.2)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning of inflammatory arthritis (e.g., Gout, Pseudogout, Pigmented Synovitis, Rheumatoid arthritis).

Gout/Pseudogout

CT without contrast, MRI without contrast, or MRI without and with contrast of the area of interest is medically necessary for investigation of suspected gout/pseudogout when ALL of the following are met:

- Initial plain x-ray to rule out other potential disease processes
- Infection or neoplasm is in the differential diagnosis for soft-tissue tophi

Pigmented Villonodual Synovitis (PVNS)

Results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider prior to requesting advanced imaging for PVNS

MRI without contrast of the affected joint, or CT with contrast if MRI contraindicated, is medically necessary for the investigation of PVNS following plain x-rays.

Rheumatoid Arthritis

Plain x-ray, physical exam, and appropriate laboratory studies are required prior to advanced imaging for rheumatoid arthritis

- Results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider
- Examples of appropriate laboratory studies may include: Lyme titers, rheumatoid factor (RF), anti-cyclic citrullinated peptide (anti-CCP), sedimentation rate (ESR), C-reactive protein (CRP), and antinuclear antibody (ANA), joint fluid analysis

US (CPT® 76881 or CPT® 76882), MRI without contrast OR MRI without and with contrast is medically necessary for investigation of rheumatoid arthritis in the most symptomatic joint, or of the dominant hand or wrist, in ALL of the following situations:

- When diagnosis is uncertain prior to initiation of drug therapy
- To study the effects of treatment with disease modifying anti-rheumatic drug (DMARD) therapy

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- To identify seronegative RA individuals that might benefit from early DMARD therapy
- To determine change in treatment, such as:
  - Switching from standard DMARD therapy to tumor necrosis factor (TNF) therapy.
  - Changing to a different TNF drug therapy, then one MRI (contrast as requested) of a single joint can be performed.
  - Addition of other treatments, including joint injections
- MRI or US is not medically necessary for routine follow-up of treatment

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Evidence Discussion (MS-12)

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Current ACR appropriateness criteria for chronic pain in the shoulder, knee, and hip all recommended plain x-ray as the initial image of choice when investigating suspected osteoarthritis (OA).[1-3] Advanced imaging is typically not necessary for the initial evaluation of OA, a diagnosis can be made based on history, physical exam and plain x-rays.[4] For most individuals diagnosed with OA, treatment does not rely on advanced imaging findings, and many can improve with conservative care.[5-8]

When individuals with OA poorly respond to conservative care or there is a concern for concomitant joint pathology, advanced imaging may be able to identify additional sources of symptoms.[9] MRI is generally considered the next appropriate imaging study for chronic joint pain when x-rays are inconclusive or further investigation is warranted. CT may be appropriate in specific situations to provide additional bony detail for treatment planning or when MRI is contraindicated.[1-3]

Plain x-rays are typically sufficient for preoperative planning for the majority of patients undergoing joint replacement surgery. However, for those with congenital or significant atypical post-traumatic arthritic deformities, CT scan can be of value for further evaluation/planning.[10] Advanced imaging will also be required prior to the replacement surgery if there will be the use of custom implants, patient specific instrumentation or computer assisted navigation.[11]

The American College of Radiology (ACR) Appropriateness Criteria for Chronic Extremity Joint Pain-Suspected Inflammatory Arthritis, Crystalline Arthritis, or Erosive Osteoarthritis (revised 2022) has recommended plain radiography as the initial imaging study for chronic extremity joint pain where crystalline arthritis is suspected.[12] X-ray sensitivity can be lower than other imaging modalities, however they may contain sufficient findings for the diagnosis of gout or calcium pyrophosphate deposition disease (CPPD). The benefits to using x-rays as an initial imaging study is to potentially avoid the need for further testing, treatment delays. Early onset gout/pseudogout may not be evident on any imaging, so reliance on a complete clinical history, evaluation and synovial fluid testing is important in any initial examination when gout/pseudogout is suspected.[13]

Advanced imaging is typically not required for the evaluation of patients with suspected crystalline arthropathy. The presence of monosodium urate crystals or calcium pyrophosphate crystals on synovial fluid microscopy can be definitive of a diagnosis of gout. However, additional imaging can be helpful in the evaluation of an individual's symptoms when differential diagnoses are suspected.[13]

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Significant advances in imaging have made the visualization of gout/pseudogout much better than in previous years, especially when using US and CT technology. The 2018 EULAR recommendations stated that US offers the best potential for diagnosis of gout and dual-energy CT is showing great promise in its utility when compared to other imaging options.[13] Research since those recommendations have added more support that US and dual-energy CT are modalities of choice. A small cross-sectional study demonstrated that US is more specific in detecting early stage gout and that both US and CT have similar accuracy for middle and late stage disease identification.[14] A systematic review in 2022 of 28 studies determined that the sensitivity and specificity for US to detect gout was 84% and 84% respectively for both early and late staged, and for dual-energy CT to be 89% and 91% for chronic gout (75% and 85% for earlier staged disease).[15] A 2019 meta-analysis found that dual-energy CT was only less sensitive for as early as 6 weeks since the start of symptoms.[16] In 2018, two systematic reviews analyzing the accuracy of and comparing dual energy CT other imaging modalities concluded it was better than x-ray or normal CT and equal to US in diagnosing gout.[17,18] There is limited evidence to lend any support of the utility of other advanced image options in the investigation of gout/pseudogout. The most recent ACR recommended that CT with contrast or MRI of any method are usually not appropriate when investigating gout/pseudogout.[12] The 2018 EULAR recommendations are similar, that while MRI has some ability to detect crystalline deposit there is limited research in the diagnostic utility of the images, especially when compared to other imaging options.[13] The utility of MRI may change as research and technology continue to advance.

The ACR Appropriateness Criteria for Chronic Extremity Joint Pain-Suspected Inflammatory Arthritis, Crystalline Arthritis, or Erosive Osteoarthritis (revised 2022) recommended plain radiography as the initial imaging study for chronic extremity joint pain where Rheumatoid arthritis (RA) is suspect.[12] Clinical and laboratory assessment of RA remains the cornerstone of diagnosis and determining a response to treatment.[19] Inconclusive or non-diagnostic imaging results can be further evaluated with advanced imaging. MRI allows assessment of all structures as well as bone edema and baseline bone edema on low and high field MRI in patients with early RA is predictive of future radiographic damage.[20] Joints and bones in the hand are often affected in RA and assessing changes in these joints can help in therapy monitoring.[21] MRI and US play important roles in detecting subclinical disease in patients with inflammatory arthritis. These modalities have higher sensitivity in detecting subclinical synovitis, tenosynovitis, osteitis, and early erosive disease compared with physical exam and xray, therefore useful in early diagnosis and evaluating response to treatment.[22]

Investigation of Pigmented Villonodular Synovitis (PVNS) may be further aided with advanced imaging. Radiographs are non-specific and may appear normal 20% of the time but joint effusion, soft-tissue swelling, extrinsic erosion of bone, absence of calcification, preservation of joint space, and/or normal bone mineralization may be seen

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in diffuse intraarticular PVNS. Localized form my appear normal on plain radiographs.[23] MRI may be necessary for diagnosis.[24] A retrospective review of 122 histologically confirmed PVNS cases demonstrated that MRI was helpful in obtaining a correct diagnosis 83% of the time.[25] MRI is also optimal for demonstrating the relationship of extraarticular lesions to the tendon sheath to suggest the diagnosis.[23]

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Capsular/Synovial and Meniscal Dysfunction (MS-29)

Guideline

Capsular/Synovial and Meniscal Dysfunction (MS-29.1)
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Capsular/Synovial and Meniscal Dysfunction (MS-29.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning for capsular and/or synovial dysfunction. See specific details below regarding any requirements for conservative care prior to requesting advanced imaging.

- Initial plain x-ray imaging of the suspected area must first be performed after the current episode of symptoms started or changed and results available to the requesting provider.
- Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started, **EXCEPT** as noted below

When the above criteria has been met, the following advanced imaging is medically necessary:

- **Shoulder**
  - Labral tear or dysfunction (e.g. SLAP, ALPSA, HAGL) – CT with contrast (CPT®73201), MRI without contrast (CPT®73221), or MRI with contrast (CPT®73222).
  - Adhesive capsulitis (frozen shoulder) – MRI without contrast (CPT®73221)
- **Hip**
  - Labral tear or dysfunction - CT with contrast (CPT® 73701), MRI without contrast (CPT® 73721), or MRI with contrast (CPT® 73722)
- **Knee**
  - Hemarthrosis, non-traumatic or spontaneous – MRI without contrast (CPT® 73721)
    - **trial of conservative care not required**
  - Hemarthrosis, traumatic – see specific section related to the tissue trauma (e.g., fracture, joint stability, ligament, osteochondral injury, etc)
  - Meniscus tear - MRI without contrast (CPT® 73721), or CT with contrast (CPT® 73701) if MRI cannot be performed, is medically necessary for further investigation of suspected meniscal tear when all of the following have been met:
    - Plain x-rays have been performed after the beginning of the current episode of symptoms started

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- Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started OR at least 2 of the following 4 criteria met:
  - positive McMurray's, Thessaly, or Apley's compression test
  - twisting or acute injury of the knee
  - locked knee: inability to fully extend knee on exam in comparison to the opposite knee
  - knee effusion
- For degenerative meniscal tear - MRI without contrast (CPT® 73721) is medically necessary for further investigation of suspected degenerative meniscus tear in an individual with osteoarthritis when all of the following have been met:
  - Plain x-rays have been performed after the beginning of the current episode of symptoms started
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started

◦ Synovial Plica or Medial Shelf – MRI without contrast (CPT® 73721)

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Evidence Discussion (MS-29)

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Advanced imaging of synovial joint symptoms is typically not recommended as an initial diagnostic study. Research has demonstrated that asymptomatic individuals may commonly have tears or other findings on advanced images of capsular and synovial tissues (e.g., labrum, capsule, synovial lining).[1,2] Abnormal findings could lead to a cascade of unnecessary treatment or even surgery as asymptomatic tissues typically do not require any treatment. The American College of Radiology (ACR) Appropriateness criteria recommended that plain x-ray is the initial imaging study for investigation of joint pain.[3-6] When initial x-rays are inconclusive or conservative care does not result in amelioration more advanced images may be necessary.

Both CT and MRI technology, when necessary, may be useful for further investigation of labral tear at either the shoulder or hip. MR arthrography is considered the standard for labral imaging due to its high sensitivity and improved soft tissue contrast. However, there may be times, such as with acute trauma, effusion and pain, that MRI without contrast is preferred.[3,5] MRI without contrast can show if capsular thickening is present in the investigation of suspected adhesive capsulitis at the shoulder. It has a relatively high sensitivity for detection of adhesive capsulitis. The introduction of contrast fluid within the rotator interval or along the axillary recess can create limitations in detecting findings of adhesive capsulitis, for this reason MRA is not recommended as next imaging when investigating adhesive capsulitis.[4]

When additional investigation of knee pain is appropriate following initial x-rays and possible conservative care, MRI without contrast is the recommended next image for suspicion of capsular or synovial pathology. MRI has been demonstrated to show the presence of medial plica.[6] Further evaluation of suspected hemarthrosis may involve the use of advanced imaging to help identify the cause of bleeding, especially in recurring cases.[7,8]

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Foreign and Loose Bodies (MS-6)

Guideline

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Foreign and Loose Bodies (MS-6.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning.

Foreign Body

US (CPT® 76881 or CPT® 76882), CT without contrast or MRI without contrast or MRI without and with contrast for further investigation of foreign bodies is medically necessary after plain x-rays have ruled out the presence of radiopaque foreign bodies as follows:

- US is the preferred imaging for radiolucent foreign bodies (e.g., wood, plastic)
- CT without contrast is recommended when plain x-rays are negative and a radiopaque foreign body is still suspected
- MRI without and with contrast is an alternative to US and CT for assessing the extent of infection associated with a suspected foreign body

Loose Body

When the initial x-rays have been taken and the results are available to the provider, advanced imaging of symptomatic loose bodies in a joint is medically necessary as follows:

- **Shoulder** - MRI without contrast (CPT®73221)
- **Elbow** - CT without contrast (CPT® 73200) , CT with contrast (CPT® 73201), MRI without contrast (CPT® 73221) or MRI with contrast (CPT® 73222)
- **Hip** - MRI without contrast (CPT® 73721)
- **Knee** - MRI without contrast (CPT® 73721) or CT Knee with contrast (arthrogram) (CPT® 73701) if MRI cannot be performed
- **Ankle** - MRI without contrast (CPT® 73721)

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Evidence Discussion (MS-6)

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Initial x-rays are recommended as a first imaging examination following acute trauma or when evaluating infection of the musculoskeletal system and there is suspicion of foreign objects or loose bodies.[1-3] X-rays are highly sensitive to embedded radiopaque materials, especially metals. X-rays provide an excellent overview of the anatomic area of interest allowing for a better overall assessment of symptoms.[4]

When a foreign body is not visualized on x-rays, ultrasound may be helpful to further evaluation.[1-3] Ultrasound has high sensitivity and specificity in detecting radiolucent objects like wood, and plastic. It has also been demonstrated as useful in detecting glass and metal.[4-6] Ultrasound is widely available, accessible, and does not involve ionizing radiation. Ultrasound can also help to evaluate complications of foreign body such as infections and vascular or tendon injuries. Ultrasound does have limitations, such as, visualization will be limited to more superficial tissue depths and variability of the technology available during the assessment may limit image quality.[5,6]

More advanced imaging may be necessary when suspected bodies are out of the range of ultrasound or are not clearly visualized on x-ray and ultrasound. CT has been shown as useful in detecting wood, aluminum, copper and some glass.[5] MRI is recommended when there is suspicion of infection or soft tissue and osseous change in relation to a foreign or loose body.[3] MRI is better than CT in the assessment of infection, however it was found to be less sensitive to actually visualizing a foreign body. There may also be image artifacts on an MRI if there are metallic foreign bodies.[4,5]

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Fractures (MS-5)

Guideline

Acute Fractures (MS-5.1)
Occult, Stress, Insufficiency Fracture or Reaction (MS-5.2)
Other Indications (e.g., Delayed Union, Preoperative) (MS-5.3)
Evidence Discussion (MS-5)
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Acute Fractures (MS-5.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning.

CT or MRI without contrast of the area of interest is medically necessary for the investigation of suspected acute complex fracture when ANY of the following apply:

- The initial plain x-ray findings demonstrate a complex (comminuted or displaced) fracture with or without dislocation
  - CT is preferred unless there is a known or suspected association of the fracture with neoplastic disease or other contraindication of CT imaging
  - If CT is contraindicated, then MRI without/with contrast is preferred
- Preoperative planning - Refer to **3D Rendering (MS-3.1)** for medical necessity criteria of 3D processing

For osteochondral fracture or osteochondral injury, see: **Osteochondral Lesions and Fractures (MS-13.1)**

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Occult, Stress, Insufficiency Fracture or Reaction (MS-5.2)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. See specific details below regarding any requirements for conservative care prior to requesting advanced imaging.

MRI without contrast of the **hip/femoral neck, tibia, pelvis/sacrum, tarsal, navicular, proximal fifth metatarsal, or scaphoid** is medically necessary for the investigation of suspected occult/stress/insufficiency fractures, and **atypical femoral shaft** fractures **related to bisphosphonate use** when ALL of the following apply:

- The initial evaluation, history, and physical exam fails to establish a definitive diagnosis AND
- The initial plain x-ray findings are negative or equivocal
- CT without contrast can be performed as an alternative to MRI for suspected occult/ insufficiency fractures of the pelvis/hip and suspected atypical femoral shaft fractures related to bisphosphonate and suspected occult fractures of the scaphoid

CT or MRI without contrast of the area is medically necessary for the investigation of all other suspected occult/stress/insufficiency fractures when ANY of the following apply, except as otherwise noted:

- Initial plain x-rays obtained a minimum of 14 days after the onset of symptoms are non-diagnostic for fracture OR
- Repeat plain x-rays remain non-diagnostic for fracture after a minimum of 10 days of provider-directed conservative treatment
- **For Tarsal/Navicular stress/occult fracture** - MRI Foot without contrast (CPT® 73718)
  - CT Foot without contrast (CPT® 73700) for follow-up of healing fractures
  - X-ray results are available to the requesting provider
- **Note** - When MRI cannot be performed to investigate suspected fractures see: **Nuclear Medicine (MS-28.1)**

MRI without contrast (CPT® 73718) is medically necessary for suspicion of shin splints when both of the following are met:

- Initial plain x-ray results are available to the requesting provider AND

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- Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started

For **stress fracture follow-up** imaging for "return to play" evaluation at least 3 months after the initial imaging study, MRI without contrast of the area of interest is medically necessary

For **stress reaction surveillance** or "return to play" decisions, advanced imaging is not medically necessary regarding a stress reaction identified on an initial imaging study.

Periprosthetic fractures related to joint replacement see: **Joint Replacement Post-Operative Follow-Up (MS-16.1)**

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Other Indications (e.g., Delayed Union, Preoperative) (MS-5.3)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning.

CT or MRI without contrast after recent (within 30 days) plain x-ray if ONE of the following is present:

- Concern for delayed union or non-union of fracture, osteotomy, or joint fusions
- Part of preoperative evaluation for a planned surgery of a complex fracture with or without dislocation

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Evidence Discussion (MS-5)

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It is widely accepted that the initial imaging for a patient with a suspected fracture should be plain radiographs. The American College of Radiology (ACR) has recommended plain x-ray as the initial image for a majority of both acute and chronic musculoskeletal pain situations, including suspected fractures.[1] When the initial x-rays are negative or unclear, yet fracture is still suspected the ACR has recommended CT as the next method of investigation as it can provide detailed information on bony structure and allow for further evaluation and treatment planning.[1] MRI may be an alternative image to CT in a situation where CT is contraindicated or there has been complex and/or extensive trauma and surrounding tissues also need to be evaluated.[2-4]

The American College of Radiology also recommended that x-rays should be the initial imaging for a suspected stress fracture. Initial x-rays may not identify the fracture and if there is continued suspicion repeat x-ray imaging in 10 - 14 days is supported.[5] Repeat radiographs may show osseous reaction confirming the presence of an occult or stress fracture. However, if repeat x-rays remain negative and there is still suspicion of an occult or stress fracture, MRI is recommended as it has been shown to be the most sensitive and specific imaging modality for workup of suspected stress injuries.[5,6] In addition, there exists a subset of occult/stress/insufficiency fractures that have an increased risk of fracture progression, delayed healing, non-union and avascular necrosis. For these high-risk injuries, advanced imaging is recommended if initial x-rays are negative or indeterminate as these injuries require early diagnosis and immediate treatment.[5,7,8]

The evaluation of patients with chronic stress syndromes (e.g., shin splints) includes a detailed history, physical examination and plain x-rays.[5] Many individuals can improve with conservative care. If the individual fails to respond to an adequate trial of conservative care, MRI is recommended as the next imaging option for further investigation.[9]

For the assessment of bony healing, serial x-ray imaging is usually sufficient. However, if there are still concerns for delayed union or non-union, CT scanning can provide detail as to the presence or absence of bridging callus. MRI can also assist in the evaluation of bone healing.

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Infection (MS-9)

Guideline

General Infection/Osteomyelitis (MS-9.1)
Charcot and Septic Joint (MS-9.2)
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General Infection/Osteomyelitis (MS-9.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning.

US (CPT® 76881 or CPT® 76882), MRI without contrast, MRI without and with contrast, CT without contrast, or CT with contrast of the affected area for further investigation of infection is medically necessary after plain x-ray(s) have been obtained in the following scenarios:

- Plain x-ray(s) do not demonstrate infection, AND
- Plain x-ray(s) do not suggest alternative diagnoses such as neuropathic arthropathy (see Charcot Joint (MS-9.2)) or fracture, AND
- Soft tissue or bone infection (osteomyelitis) is suspected

OR

- Plain x-ray(s) are positive for infection, AND
- The extent of infection into the soft tissues and any skip lesions require evaluation

For Individuals with **diabetic foot infections** after plain x-rays - as a compliment to plain x-ray - MRI without and with contrast (CPT® 73720), MRI without contrast (CPT® 73718), CT without contrast (CPT® 73700) or CT with contrast (CPT® 73701)

For individuals with suspected spinal infections please use the spine imaging guidelines, see: **Red Flag Indications (SP-1.2)**

For nuclear medicine studies appropriate in specific scenarios, see: **Nuclear Medicine (MS-28)**

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Charcot and Septic Joint (MS-9.2)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. See specific details below regarding any requirements for conservative care prior to requesting advanced imaging.

Charcot Joint

Following the initial x-ray and the failure of the required 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started, the following advanced imaging is medically necessary for the investigation of suspected Charcot joint:

- Charcot ankle - MRI without contrast (CPT® 73721)
- Charcot foot - MRI without contrast (CPT® 73718) or MRI without and with contrast (CPT® 73720)

Septic Joint

MRI without and with contrast, MRI without contrast, CT without contrast, CT with contrast, or Ultrasound (CPT® 76881 or CPT® 76882) of the affected joint for further investigation of joint sepsis is medically necessary when **standard or image-guided arthrocentesis is contraindicated, unsuccessful, or non-diagnostic**, and the clinical documentation satisfies ALL of the following criteria:

- History and physical examination findings [One of the following]:
  - Development of an acutely hot and swollen joint (< 2 weeks)
  - Decreased range of motion due to pain
  - Documented fever
- Laboratory tests [One of the following]:
  - Leukocytosis
  - Elevated ESR or C-reactive protein
  - Analysis of the joint fluid is non-diagnostic
- Plain x-ray of the joint - Results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider

MRI without and with contrast, MRI without contrast, CT without contrast, or CT with contrast of the affected joint to evaluate the extent of infection into the soft tissues and any skip lesions that would require evaluation is medically necessary after plain x-rays if

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the arthrocentesis is diagnostic and if there is a confirmed septic joint. Results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider.

For the criteria for further investigation of post-operative joint replacement septic infection please see joint specific criteria in **Joint Replacement Post-Operative Follow-Up (MS-16.1)**

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Evidence Discussion (MS-9)

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The American College of Radiology (ACR) has recommended that x-rays should be used for the initial evaluation of musculoskeletal infections, including osteomyelitis, septic arthritis, and soft tissue infection.[1] They also recommended x-rays as the initial image for the evaluation of feet in individuals with Diabetes Mellitus, which often present with symptoms consistent with infection.[2] Initial radiographs can provide an excellent overview of the anatomic area of interest and can help to exclude other causes of swelling or pain.[3] Radiographs can help with the interpretation of future imaging studies such as CT, MRI, US, and nuclear medicine scans.

The clinical presentation of a suspected infected area or joint has a wide differential diagnosis. Infection is primarily a clinical diagnosis based on physical examination and laboratory testing of blood and/or synovial fluid, which may require prompt arthrocentesis and culture.[3-5] In select situations when a full examination and initial imaging is unclear, advanced imaging may be necessary to better visual the extent of suspected infection and associated effusion or other tissue damage or injury.[3,4]

US, MRI, or CT can be appropriate as the next imaging study for suspected septic arthritis, soft tissue infection, or osteomyelitis following normal radiographs.[1,2] MRI is often the next image of choice for suspect musculoskeletal infection due to its superior contrast resolution of soft tissues which can be helpful in detecting the spread of infection.[3,4,6] They may also be helpful to evaluate the adjacent soft tissues for infection. MRI is also recommended as the next image for evaluation of suspected Charcot joint of the foot. Charcot joints often present with complex symptoms including infection and potential ulceration that may be best visualized with MRI.[2] US can be helpful in detecting effusion and for guiding aspiration for more superficial areas in question.[3,7] CT may also be appropriate in situations when MRI is contraindicated, such as in areas with implanted metal or suspected foreign metal, or MRI is unavailable.[3,4]

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Joint Instability and Dysfunction (MS-30)

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Joint Instability and Dysfunction (MS-30.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required, **EXCEPT** for situations specifically listed as not required in this section, prior to requesting advanced imaging for further investigation and/or treatment planning. See specific details throughout the section regarding any requirements for when conservative care prior to requesting advanced imaging is required.

- Initial x-rays must have been performed after the current episode of symptoms started or changed and the results are available to the requesting provider

Advanced imaging for further investigation of joint instability and/or articular dysfunction is medically necessary as follows:

- **Shoulder**:
  - Glenohumeral dislocation/subluxation and/or Bankart or Hill-Sachs lesion – MRI with contrast (CPT®73222), MRI without contrast (CPT®73221), CT with contrast (CPT®73201) or CT without contrast (CPT®73200)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required in individuals over age 40 with a first time dislocation
    - Conservative treatment is not required for individuals 40 years of age or younger with a first-time dislocation, and individuals with recurrent dislocations
  - Acromioclavicular dislocation/separation when ruling out possible rotator cuff tear – MRI without contrast (CPT® 73221)
  - Sternoclavicular (SC) dislocation/separation
    - **Initial x-rays are not required**
    - For evident or suspected SC dislocations - CT chest without contrast (CPT® 71250) or CT chest with contrast (CPT® 71260)
    - For differentiating physeal injury from SC dislocation in individuals 25 years of age or younger OR planning for operative repair - MRI chest without contrast (CPT® 71550) or MRI without and with contrast (CPT® 71552)
    - Proximal 1/3rd (one-third) clavicle fracture – CT chest with contrast (CPT® 71260), CT chest without contrast (CPT® 71250), MRI chest without contrast or MRI chest without and with contrast (CPT® 71552)
- **Wrist**:

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- Distal Radioulnar joint instability – CT of both wrists, pronated and supinated positions, without contrast (CPT® 73200)

- **Pelvis**:
  - Osteitis Pubis/ Symphysis Pubis Diastasis – MRI without contrast (CPT® 72195)
    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required
- **Hip**:
  - Femoroacetabular Impingement, for preoperative planning – MRI without contrast (CPT® 73721) or MRI with contrast (CPT® 73722) and CT without contrast (CPT® 73700) or CT Pelvis without contrast (CPT® 72192)
    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required
- **Knee**:
  - Tibiofemoral dislocation following a significant trauma to evaluate for ligament and vascular injury – MRI without contrast (CPT® 73721) and/either MRA lower extremity without and with contrast (CPT® 73725) or CTA lower extremity without and with contrast (CPT® 73706)
  - Patellar dislocation/subluxation following an acute injury with consideration of surgery and concern for osteochondral fragment – MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
  - Recurrent Patellar instability if consideration for surgery – MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required
- **Ankle**:
  - Impingement, anterior, anterior-lateral or posterior – MRI without contrast (CPT® 73721), MRI with contrast (CPT® 73722), or CT with contrast (CPT® 73701)
    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required
  - Instability, preoperative planning – MRI without contrast (CPT® 73721) or MRI with contrast (CPT® 73722)
    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required
- **Foot**:
  - Tarsal Coalition, preoperative planning – MRI Ankle without contrast (CPT® 73721) or CT Ankle without contrast (CPT® 73700)

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    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required is required
- Sinus Tarsi Syndrome, if diagnosis is unclear of for preoperative evaluation – MRI Ankle without contrast (CPT® 73721)
    - Failure of 6 weeks of provider-directed conservative treatment, including re-evaluation, occurring after the current episode of symptoms started is required

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Evidence Discussion (MS-30)

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The American College of Radiology (ACR) has recommended that x-rays be the initial image of choice for evaluation of acute and chronic joint instability symptoms (e.g., dislocation, subluxation) and articular dysfunction or abnormality (e.g., bony impingement, coalition).[1-6] X-rays, along with a full history and clinical examination, may be sufficient to determine a diagnosis and treatment plan. In many instances, having an individual follow a course of provider directed conservative care may be sufficient to reduce symptoms and aid in the return of normal function.[7,8]Early advanced imaging could expose individuals to unnecessary risks and additional healthcare costs.

Some acute injuries involving significant trauma may require advanced imaging prior to conservative care. Damage to surrounding anatomy can occur that would need prompt treatment. MRI is typically recommended as the next image for many acute joint injuries as it has excellent capability in discriminating soft tissues and surrounding anatomy. CT may also be appropriate when MRI is contraindicated, unavailable or better visualization of bony alignment is required, such as in surgical planning.[1,2,4,6,9]

When conservative care has not been effective at improving function, or an individual presents with re-occurring and chronic problems, both MRI and CT are recommended as next images to further investigate joint instability and dysfunction.[3,5,7,10] MRI is recommended as useful in assessing the integrity of tissues that support the joint (e.g., labrum, capsule, ligament) and damage that may have occurred over time to chondral surfaces of both the hip and shoulder.[7,10] MRI was shown to have good inter- and intra-rater reliability in assessing patellofemoral instability.[11]

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Ligament and Fascia Injury (MS-31)

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Ligament and Fascia Injury (MS-31.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. See specific details throughout the section regarding any requirements for when conservative care prior to requesting advanced imaging is required.

- Initial plain x-ray imaging of the suspected area must first be performed after the current episode of symptoms started or changed with results available to the requesting provider

Advanced imaging for further investigation of ligament and fascia injury is medically necessary as follows:

- **Elbow**
  - Ulnar Collateral Ligament tear, following acute or repetitive trauma (including overhead throwing athletes) – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221), MRI with contrast (CPT® 73222) or CT with contrast (CPT® 73201)
- **Wrist**
  - Intrinsic ligament tear and/or Triangular Fibrocartilage Complex injury – MRI with contrast (CPT® 73222) or CT with contrast (CPT® 73201).
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Hand**
  - Ulnar Collateral Ligament tear, if ruling out a Stener lesion or a complete ligament tear – US (CPT® 76881 or CPT® 76882), or MRI without contrast (CPT® 73218)
- **Knee**
  - Cruciate and/or collateral ligament tear – MRI without contrast (CPT® 73721)
    - A failed trial of conservative treatment is not required if any of the following signs are positive in comparison to the opposite knee:
      - Anterior Drawer
      - Lachman
      - Pivot Shift
      - Posterior Drawer or Posterior Sag

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        - Valgus Stress
        - Varus Stress
      - Otherwise, failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Patellofemoral and Patellotibial ligament tear – MRI without contrast (CPT® 73721)
      - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Ankle**
    - Lateral/Medial Ligament injury (e.g., sprained ankle), including avulsion fracture – MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
      - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Syndesmosis Injury (e.g., high ankle sprain) – MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
- **Foot**
    - Plantar Fascia
      - Fascia tear (partial or complete) – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73718)
        - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
      - Fasciitis preoperative planning – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73718)
        - Failure of provider-directed conservative treatment lasting at least 6 months or more, including clinical re-evaluation, occurring after the current episode of symptoms started is required
      - Plantar plate (e.g., Turf Toe injury) – MRI without contrast (CPT® 73718)
        - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required

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Evidence Discussion (MS-31)

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Initial imaging of MSK injuries where ligament or fascia is suspected to be involved is typically done with plain x-rays. The American College of Radiology has recommended x-ray as the initial image for evaluation of most acute injuries.[1] Even in suspected ligament or fascia injury, initial x-rays can be beneficial in determining if there is damage or malalignment to bone that needs to be considered during a thorough evaluation.

A trial of conservative care following negative x-rays may be beneficial for some suspected ligament and fascia injuries prior to immediately seeking advanced imaging. Individuals that show improvement over time with conservative care could avoid the additional potential exposure and expense of advanced imaging or unnecessary treatment or surgery. In their 2018 revised Appropriateness Criteria for Acute Hand and Wrist Trauma, the ACR recommended that symptomatic individuals with negative initial x-rays may use a trial of immobilization and repeat x-rays.[2] Current injury rehabilitation guidelines have recommended that individuals with suspected lateral ankle sprains follow a 4-6 week conservative care program with advanced imaging only being performed for those with persistent symptoms despite following the whole program.[3] Furthermore, guidelines also indicate that individuals with suspected plantar fascial pain usually do not require imaging studies unless they fail a comprehensive conservative care program.[4]

When medically necessary, advanced imaging for ligament and fascia injury has demonstrated high accuracy. More superficial ligaments such as those in the hand, wrist and elbow may be appropriately visualized through ultrasound alone. Ultrasound has been shown to be 81% sensitive and 91% specific in detecting Ulnar collateral Ligament injury at the elbow and 96% sensitive and 90% specific at the thumb.[5-7] Ultrasound demonstrated high sensitivity in evaluating heel/foot pain and may have utility in ruling out plantar plate injury.[8]

When further imaging beyond initial x-ray or ultrasound is necessary to investigate potential ligament or fascia injury, MRI is typically considered the standard next image option. ACR recommendations for imaging of the acute elbow, wrist, hand, knee, ankle and chronic foot all support MRI as the next imaging type for ligament or fascia investigation. Due to MRI's sensitivity to different tissues, it can be useful in evaluating not only ligament and fascia, but also any associated stress injury to bone or marrow, edema patterns, syndesmotic injury, meniscal involvement or other soft tissue injury.[2,9-12] Having accurate imaging of all aspects of any injury will aid in appropriate management and treatment planning .

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CT imaging may also be useful in some instances, such as if MRI is contraindicated for an individual or when evaluation of bone displacement is desired. When combined with contrast, CT can be helpful in more accurate visualization of malalignment of the small and compact joints of the wrist. CT arthrography has demonstrated sensitivity and specificity equal to, or higher than, MRI or arthroscopy in detection of scapholunate and lunotriquetral ligament injury.[2] CT imaging is supported in suspected "high-ankle" syndesmotic injury, while MRI is often still considered a better option, measurements from CT scans and displayed displacement have been demonstrated to identify syndesmotic injury seen on MRI.[12]

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Limb Length Discrepancy (MS-17)

Guideline

Limb Length Discrepancy (MS-17.1)
Evidence Discussion (MS-17)
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Limb Length Discrepancy (MS-17.1)

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A diagnostic advanced imaging CPT code (e.g., CPT® 73700, CPT® 73701, or CPT® 73702) is not medically necessary for the evaluation of limb length discrepancy.

Either plain radiograph or "CT scanogram," both reported with CPT® 77073, is appropriate to radiographically evaluate limb length discrepancy due to congenital anomalies, acquired deformities, growth plate (physeal injuries or surgery), or inborn errors of metabolism.

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Evidence Discussion (MS-17)

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The findings of recent systematic reviews support that x-ray is the most valid and reliable method for evaluation of limb length discrepancy. Advanced imaging modalities are not supported in current research as a method to evaluate leg length discrepancy. These technologies are more expensive, can have added risks such as sedation, and increased exposure to radiation. Furthermore, they can have added time it takes an individual to access the technology and may be contraindicated for some that have implantable devices. Full length, anterior-to-posterior x-ray images are recommended. Studies also reported excellent reliability using CT scanogram imaging, which may be an appropriate alternative to x-rays.[1,2]

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Muscle/Tendon Unit Injuries/Diseases (MS-11)

Guideline

Muscle and Tendon Injuries (MS-11.1)
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Evidence Discussion (MS-11)
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Muscle and Tendon Injuries (MS-11.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. See specific details throughout the section regarding any requirements for when conservative care is required prior to requesting advanced imaging.

- Initial plain x-ray of the suspected area must first be obtained after the current episode of symptoms started or changed and results are available to the provider

Muscle belly strains/tears - Advanced imaging is not medically necessary. Advanced imaging will not change the treatment plan for muscle belly strains/tears. There is lack of evidence to support surgical treatment for these injuries. These injuries are nearly always treated through conservative, non-operative, care.

Tendons - Advanced imaging for further investigation of tendon injury and/or pathology is medically necessary as follows:

- **Shoulder**
  - Impingement – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT®73221) or MRI with contrast (CPT®73222) or CT with contrast (CPT® 73201) if MRI is contraindicated
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Chronic tendon inflammation/irritation (e.g., tendinitis, bursitis) – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT®73221)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Bicep Tendon (Long Head), complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT®73221)
    - Clinical exam must be inconclusive, or advanced imaging for preoperative planning/surgery is being considered
  - Pectoralis Major/Minor complete tear – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT®73221) or MRI Chest without contrast (CPT®71550)
    - Clinical exam must be inconclusive or for preoperative planning
  - Rotator Cuff tendon tear

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        - Partial or complete tear from acute injury and surgery is being considered – MRI without contrast (CPT®73221), MRI with contrast (CPT®73222) or CT with contrast (CPT®73201)
        - Partial or complete tear – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT®73221) or MRI with contrast (CPT®73222) or CT with contrast (CPT®73201) if MRI is contraindicated
            - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Other tendons not otherwise mentioned above, partial or complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT®73221)
        - Specific tendon must be named for partial tendon tear
        - Complete tears must be in relation to preoperative planning of a specific named tendon
- **Elbow**
    - Chronic tendon inflammation/irritation (e.g., tendinitis, bursitis)
        - Bursitis – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221) or MRI without and with contrast (CPT® 73223)
            - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
        - Tendinitis – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73221)
            - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
        - Epicondylitis, medial or lateral – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73221)
            - Symptoms must persist for longer than 6 months despite trial of at least 6 weeks of conservative treatment. Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Bicep/Tricep insertion tendon, complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73221)
        - Clinical examination must be inconclusive or imaging is for preoperative planning

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- Other tendons not otherwise mentioned above, partial or complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73221)
  - Specific tendon must be named for partial tendon tear
  - Complete tears must be in relation to preoperative planning of a specific named tendon

- **Wrist and Hand**
  - Chronic tendon inflammation/irritation (e.g., tendinitis) – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221 for wrist or CPT® 73218 for hand) or MRI without and with contrast (CPT® 73223 for wrist or CPT® 73220 for hand)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Other tendons not otherwise mentioned above, partial or complete tear – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221 for wrist or CPT® 73218 for hand) or MRI without and with contrast (CPT® 73223 for wrist or CPT® 73220 for hand)
    - Specific tendon must be named for partial tendon tear
    - Complete tears must be in relation to preoperative planning of a specific named tendon
- **Pelvis**
  - Chronic tendon inflammation/irritation (e.g., tendinitis) – MRI without contrast (CPT® 72195) or MRI right and or left hip without contrast (CPT® 73721)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Tendon tear, partial or complete – MRI without contrast (CPT® 72195)
    - Specific tendon must be named
  - Athletic Pubalgia - MRI without contrast (CPT® 72195)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Piriformis Syndrome – This condition is imaged according to the criteria found in the Peripheral Nerve and Neuromuscular Guidelines see: **Focal Neurophathy (PN-2.1)**
- **Hip**
  - Chronic tendon inflammation/irritation (e.g., tendinitis, bursitis) – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)

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        - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Abductor tear/avulsion – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
    - Other tendons not otherwise mentioned above, partial or complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
        - Specific tendon must be named for partial tendon tear
        - Complete tears must be in relation to preoperative planning of a specific named tendon
    - Piriformis Syndrome – See above in **Pelvis**
- **Knee**
    - Chronic tendon inflammation/irritation (e.g., tendinitis) – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
        - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Quadriceps tendon (distal) complete tear, preoperative planning – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
    - Other tendons not otherwise mentioned above, partial or complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
        - Specific tendon must be named for partial tendon tear
        - Complete tears must be in relation to preoperative planning of a specific named tendon
- **Ankle**
    - Chronic tendon inflammation/irritation (e.g., tibial or peroneal tendon dysfunction, subluxation, Achilles tendinitis) – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
        - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - Achilles Tendon, complete tear, preoperative planning – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
    - Other tendons not otherwise mentioned above, partial or complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
        - Specific tendon must be named for partial tendon tear
        - Complete tears must be in relation to preoperative planning of a specific named tendon

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- **Foot**
  - Chronic tendon inflammation/irritation (e.g., tendinitis) – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73718)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Tendons, partial or complete tear – US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73718)
    - Specific tendon must be named for partial tendon tear
    - Complete tears must be in relation to preoperative planning of a specific named tendon

For **inflammatory muscle disease imaging**, please see either: **Muscle Diseases (PN-8.5)** in the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines or **Inflammatory Muscle Diseases (PEDMS-10.3)** in the Pediatric Musculoskeletal Imaging Guidelines.

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Compartment Syndrome (MS-11.2)

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Acute - This is a surgical emergency and advanced imaging not medically necessary as this diagnosis is made clinically by direct measurement of muscle compartment pressure.

Chronic/Exertional - Advanced imaging should only be considered when ruling out other potential causes of extremity pain following a plain x-ray and conservative treatment as indicated. Results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider.

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Evidence Discussion (MS-11)

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Initial evaluation of a patient with a suspected tendon injury should include an accurate history, careful examination and plain radiographs. The American College of Radiology has recommended x-ray as the initial image for evaluation of most acute and chronic injuries.[1] With negative x-rays, suspicion of tendon injury does not always require immediate advanced imaging. Best practice recommendations for MSK pain have stated that imaging be used selectively. Unnecessary imaging can drive up costs of care and may lead to over-medicalization of an individual's symptoms.[2,3] Many muscle and tendon injuries can significantly improve through conservative care without advanced imaging.

After x-rays have been done, and any conservative care recommended has not resulted in a meaningful improvement of symptoms, advanced imaging may be beneficial for further assessment and treatment planning of certain muscle and tendon injuries. Both US and MRI are supported by the American College of Radiology as a next appropriate image in the evaluation of acute and chronic shoulder, elbow, wrist, hip, knee, ankle and foot injuries when initial x-rays are negative or unclear and muscle or tendon involvement is suspected.[4-14] Evidence has shown that both US and MRI have been validated as excellent methods to evaluate the integrity of muscle and tendon, with MRI being reserved for instances of surgical planning, differential diagnosis or assessment of injury too deep for clear visualization by US.[15,16]

Advanced imaging will typically not change the treatment plan for muscle belly strains/ tears. These injuries are most often treated through conservative, non-operative, care. There is limited evidence to support the surgical repair of muscle belly strains/tears.[17]

Initial diagnosis of compartment syndrome is typically based on clinical presentation, perfusion pressure measurements and laboratory results. Acute compartment syndrome is considered a medical emergency for which a thorough clinical assessment and direct pressure measurements of the compartment can result in a diagnosis so the individual can quickly receive emergency care. Advanced imaging does not play a role in the diagnosis or management of this condition and may delay the time to surgical treatment.[18,19] Chronic exertional compartment syndrome can be determined through dynamic intra-compartmental pressure measurements. These are considered gold standard for diagnosis of chronic compartment pressure concerns. MRI has lacked validity as a non-invasive diagnostic tool for this condition. However, MRI may be useful to rule out other possible sources of pain if plain x-rays fail to find a source.[20,21]

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Chondral/Osteochondral Lesions (MS-13)

Guideline

Osteochondral Lesions and Fractures (MS-13.1)
Evidence Discussion (MS-13)
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Osteochondral Lesions and Fractures (MS-13.1)

MS.OC.0013.1
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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. The following must be met prior to requesting advanced imaging for further investigation and/or treatment planning of suspected osteochondral lesions or fractures (including osteochondritis dissecans):

- Initial plain x-ray imaging of the suspected area must first be performed after the current episode of symptoms started or changed
- If plain x-rays show a non-displaced osteochondral fragment, follow-up imaging should be with plain x-rays. Advanced imaging is not necessary

When the initial x-rays are negative and an osteochondral injury is still suspected or the x-rays and clinical examination suggest an unstable osteochondral injury the following advanced imaging is medically necessary:

- **Elbow** - MRI without contrast (CPT® 73221), MRI with contrast (CPT® 73222), CT without contrast (CPT® 73200) or CT with contrast (CPT® 73201)
- **Knee** MRI without contrast (CPT® 73721), MRI with contrast (CPT® 73722), or CT with contrast (CPT® 73701)
- **Ankle** - MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
- **Other Joints** - MRI without contrast, MRI with contrast (arthrogram), or CT with contrast (arthrogram)
- **Follow-up of healing (including post-operative fixation)** - MRI without contrast or CT without contrast when healing cannot be adequately assessed on follow-up plain x-rays.

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Evidence Discussion (MS-13)

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The American College of Radiography (ACR) has recommended that x-ray should be the first imaging test performed to evaluate joint pain suspected to be due to chondral/ osteochondral lesions.[1-3] Open physes have a much higher potential for healing with conservative treatment. So, radiographs may help to exclude other causes of pain and to determine skeletal maturity, which significantly affects prognosis and management of Osteochondritis Dissecans lesions (OCD). Radiographs may also be sufficient for classification of lower grade lesions avoiding unnecessary advanced imaging.[4] In clinical practice, serial radiographs may be sufficient for monitoring healing of juvenile OCD lesions.

In individuals with radiographs positive for Osteochondritis Dissecans (OCD) or subchondral insufficiency fracture, or with negative radiographs but osteochondral fracture is still suspected, MRI may be medically necessary to further evaluate cartilage for additional injuries and for grading of osteochondral fractures and OCD.[1-3] MRI has a high diagnostic value for assessing the stability of lesions, especially in higher grade lesions.[4,5] This makes MRI useful in determining the best method of treatment, be that conservative or surgical.[4,6] Repeat MRI is suggested only if radiographs are not diagnostic for healing and for worsening symptoms.[6] CT without contrast may be medically necessary to evaluate patients with OCD to confirm loose bodies or when MRI is not definitive.[1-3] MR arthrography or CT arthrography is an effective test for locating intra-articular osteochondral fragments, loose bodies and grading chondral and osteochondral lesions. A combination of x-rays and MRI or CT is recommended to best evaluate the stability of lesions.[7]

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Osteonecrosis/Avascular Necrosis (MS-4)

Guideline

Osteonecrosis/Avascular Necrosis (MS-4.1)
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Osteonecrosis/Avascular Necrosis (MS-4.1)

MS.AN.0004.1
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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. The following must be met prior to requesting advanced imaging for further investigation and/or treatment planning of suspected osteonecrosis (avascular necrosis):

- Initial plain x-ray imaging of the suspected area must first be performed after the current episode of symptoms started or changed

When the initial x-rays are negative or equivocal and the clinical symptoms warrant further investigation of osteonecrosis the following advanced imaging is medically necessary:

- MRI without contrast, MRI without and with contrast, or CT without contrast of the area of interest

When the initial x-rays have confirmed osteonecrosis and advanced imaging is needed for further treatment planning the following advanced imaging is medically necessary for the follow situations:

- Shoulder - Humeral head - MRI without contrast (CPT® 73221) or CT without contrast (CPT® 73200)
- Wrist – Lunate/Scaphoid - MRI without contrast (CPT® 73221) or CT without contrast (CPT® 73200)
- Hip – Femoral head - MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
- Knee – Distal Femur - MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
- Ankle – Talus - MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
- Foot – Tarsal Navicular/Metatarsal head - MRI without contrast (CPT® 73718) or CT without contrast (CPT® 73700)

Individuals with acute lymphoblastic leukemia and known or suspected osteonecrosis should be imaged according to guidelines in **Acute Lymphoblastic Leukemia (PEDONC-3.2)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Known or suspected osteonecrosis in long-term cancer survivors should be imaged according to guidelines in **Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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The American College of Radiology Appropriateness Criteria (ACR) for Osteonecrosis (revised 2022) has recommended radiography as the initial imaging study for clinically suspected osteonecrosis. Radiographs are less sensitive for detection of early osteonecrosis but they help to exclude other causes of extremity pain such as fracture, primary arthritis, or tumor. In late stage osteonecrosis, x-rays will also show findings of secondary osteoarthritis.[1,2]When x-rays are negative or equivocal, and osteonecrosis is still suspected, the ACR supports the use of advanced imaging to aid in further diagnosis and treatment planning.[1]

Both CT and MRI are recommended as next step imaging options after plain x-rays are obtained. MRI has been shown to be both highly sensitive and specific to the detection of osteonecrosis. A 2018 meta-analysis demonstrated that MRI was 93% sensitive and 91% specific in the early detection of osteonecrosis of the femoral head.[3] A recent Delphi model to better predict femoral head collapse identified subchondral fracture as a key predictor. CT scans can provide important insights into the structural integrity of subchondral bone allowing for better treatment planning for patients with osteonecrosis.[4] Preoperative CT has been shown to more clearly display the extent of articular collapse than radiographs.[5] Both MRI and CT have been shown to have similar accuracy in displaying femoral head lesions in more advanced stages of osteonecrosis (ARCO stage III or greater).[6]

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Osteoporosis (MS-14)

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Osteoporosis (MS-14.1)

MS.OP.0014.1
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Initial plain x-ray imaging is not required to request advanced imaging for osteoporosis screening.

Quantitative CT (CPT® 77078) can be approved for **screening** when DXA scanner is unavailable or known to be inaccurate for ANY of the following populations:

- Women 65 years of age or older, Men 70 years of age or older
- Women less than 65 years of age who have the following additional risk factors for osteoporosis:
  - Estrogen deficiency
  - A history of maternal hip fracture that occurred after age 50 years
  - Low body mass (less than 127 lb or 57.6 kg)
  - History of amenorrhea (greater than 1 year before age 42 years)
- Women less than 65 years or men less than 70 years of age who have the following additional risk factors:
  - Current use of cigarettes
  - Loss of height, thoracic kyphosis
- Individuals of any age with the following:
  - Bone mass osteopenia or fragility fractures on imaging studies such as x-rays, CT, or MRI
  - Develop 1 or more insufficiency fractures
- Individuals 50 years of age and older who develop a wrist, hip, spine, or proximal humerus fracture with minimal or no trauma, excluding pathologic fractures.
- Premenopausal females, or males age 20 to 50 years, with risk factors known to potentially alter bone mineral density such as:
  - Chronic renal failure
  - Rheumatoid or other inflammatory arthritis
  - Eating disorders, including anorexia nervosa and bulimia
  - Organ transplantation
  - Prolonged immobilization
  - Conditions associated with secondary osteoporosis, such as gastrointestinal malabsorption or malnutrition, sprue, osteomalacia, vitamin D deficiency, endometriosis, acromegaly, chronic alcoholism or established cirrhosis, and multiple myeloma

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  - Individuals who have had gastric bypass for obesity
  - Individuals with an endocrine disorder known to adversely affect bone mineral density (e.g., hyperparathyroidism, hyperthyroidism, or Cushing syndrome)
- Individuals receiving (or expected to receive) glucocorticoid therapy for >3 months
- Hypogonadal men older than 18 years of age and men with surgically or chemotherapeutically-induced castration
- Individuals beginning or receiving long-term therapy with medications known to adversely affect BMD (e.g., anti-convulsant drugs, androgen deprivation therapy, aromatase inhibitor therapy, or chronic heparin)

NOTE: Repeat screening quantitative computed tomography (QCT) can be approved no sooner than every two years.

Quantitative CT scan (CPT® 77078) can be approved for **monitoring/non-screening** when DXA scanner is unavailable or known to be inaccurate for ANY of the following circumstances:

- Follow-up in cases where QCT was the original study
- Multiple healed vertebral compression fractures
- Significant scoliosis
- Advanced arthritis of the spine due to increased cortical sclerosis often with large marginal osteophytes
- Obese individual over the weight limit of the dual-energy x-ray absorptiometry (DXA) exam table
- Individuals with BMI >35kg/m2
- Extremes in body height (i.e., very large and very small individuals)
- Individuals with extensive degenerative disease of the spine
- A clinical scenario that requires sensitivity to small changes in trabecular bone density (parathyroid hormone and glucocorticoid treatment monitoring).

NOTE: Repeat monitoring/non-screening QCT can be approved no earlier than one year following a change in treatment regimen, and only when the results will directly impact a treatment decision.

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The primary diagnostic test used to screen for osteoporosis is the central dual x-ray absorptiometry (DEXA) which accurately measures bone mineral density at the hip and lumbar spine. DEXA accuracy and reproducibility has led to the established standards for diagnosis of osteoporosis by the World Health Organization.[1] The USA Bone Health and Osteoporosis Foundation has recommended bone mineral density testing in all women age 65 and older and all men age 70 and older, and in postmenopausal women younger than 65 years and men aged 50-69 years based on their risk factor profile, including if they had a fracture as an adult.[2] The American College of Radiology Appropriateness Criteria supports DEXA as the primary diagnostic choice to screen women >65 years of age and men >70 years of age for osteoporosis, and for postmenopausal women <65 years of age with additional risk factors for fracture.[3] Repeat bone mineral density assessments one to two years after initiating medical therapy for osteoporosis and every two years thereafter, but recognizes that testing more frequently may be warranted in certain clinical situations, and may be needed less frequently in patients without major risk factors or significant bone density loss on initial BMD testing. DEXA is currently the preferred method for monitoring treatment response.[2]

Quantitative CT (QCT) is regarded as a secondary tool after DEXA for screening for osteoporosis. QCT is highly accurate in determining tissue density within a region of interest. Indications for QCT are the same as for DEXA, however DEXA is recommended as the first-line screening and follow-up test for bone density. If DXA is not available, QCT may be used as a secondary technique and for serial/follow-up purposes.[2,3,4] Selected conditions in which QCT is considered superior to DEXA include extremes in body height, BMI >35, clinical scenarios when an increased sensitivity to small changes in trabecular bone density is required, and in patients with advanced degenerative bony changes in the spine.[3] A potential harm of Quantitative CT is increased radiation exposure (1-10 mSv) as compared to <0.1 mSv for DEXA scan. Whereas the radiation dose for both lumbar spine and hip scanning in a DEXA scan is approximately equivalent to that of a chest x-ray.

There is insufficient evidence to support the use of x-ray as a screening tool in patients suspected of having low bone mineral density. Patients whose x-rays report osteopenia and/or fragility fractures should be referred for DEXA for further characterization of bone density. There is also insufficient evidence to support the current use of quantitative ultrasound as a screening tool in patients suspected of having low bone mineral density.[3]

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General Pain (MS-32)

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General Pain (MS-32.1)

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As outlined in General Guidelines (MS-1.0), initial images and physical exam are required prior to requesting advanced imaging for further investigation and/or treatment planning. See specific details throughout the section regarding any requirements for when conservative care prior to requesting advanced imaging is required. The criteria in this section applies only to general musculoskeletal pain conditions not already specified in any other section of this guideline.

- Initial plain x-ray imaging of the suspected area must first be performed after the current episode of symptoms started or changed and results are available to the requesting provider

Advanced imaging for further investigation of general musculoskeletal pain is medically necessary as follows:

- **Shoulder** - US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT®73221) or CT with contrast (CPT® 73201) if MRI contraindicated
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Elbow** - US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73221)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Wrist** - US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221), MRI with contrast (CPT® 73222), CT without contrast (CPT® 73200) or CT with contrast (CPT® 73201)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Hand** - US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73218) or MRI without and with contrast (CPT® 73220)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Pelvis** - MRI without contrast (CPT® 72195) or MRI right and/or left Hip without contrast (CPT® 73721)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required

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  - Imaging for Sacroiliac and Coccygeal pain is guided by the Spine Imaging Guidelines (Sacroiliac Joint Pain/Sacroiliitis (SP-10.1) and Coccydynia without Neurological Features (SP-5.2))
- **Hip** - US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Knee** - US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - For patellofemoral pain or tracking issues where surgery is being considered - MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Ankle** - US (CPT® 76881 or CPT® 76882) or MRI without contrast (CPT® 73721)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started
- **Foot** - MRI without contrast (CPT® 73718)
  - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - For investigation of sesamoid disorders:
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - MRI without contrast (CPT® 73718) or CT without contrast (CPT® 73700) of the foot
  - For investigation of Complex Regional Pain Syndrome (CRPS) (Type 1) at the foot:
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
    - MRI Foot without contrast (CPT® 73718)
    - Criteria regarding nuclear medicine imaging of CRPS is found in: **Nuclear Medicine (MS-28)**

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Evidence Discussion (MS-32)

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General, unspecific, musculoskeletal pain and trauma may not always present in a definitive manner. An individual may present with symptoms similar to multiple differential diagnoses. Imaging, as part of a comprehensive examination and assessment, can be useful in these situations to help rule-in or -out a certain diagnosis so as to develop a clinical course of action. The American College of Radiology (ACR) has recommended x-ray as the initial evaluation image for the vast majority of non-specific acute[1-7] and chronic[8-14] musculoskeletal pain. Initial x-rays may provide sufficient information, along with the clinical evaluation, to form a diagnosis. These initial images can be helpful in determining if a course of conservative treatment is recommended. In some instances, the initial x-rays or US may help determine a diagnosis or that more advanced images or procedures are appropriate.[15]

Advanced imaging can often demonstrate abnormalities that have no relevance to the patient's symptoms. Studies have shown that advanced imaging may demonstrate musculoskeletal abnormalities that do not affect their quality of life.[16] Focusing on incidental findings can lead to overtreatment through unnecessary referral to medical specialists or unneeded medical procedures. Advanced imaging can have additional risks to the individual beyond those of initial plain images. There are risks associated with additional exposure to radiation. There can also be complications due to implantable devices or metallic foreign bodies in the individual. Risk mitigation needs to be weighed when advanced imaging is being considered, especially in relation to repeat or serial advanced imaging.[17,18] Musculoskeletal complaints may respond to care prior to needing advanced imaging. Current ACR guidance has recommended obtaining initial plain x-rays and having the individual follow a course of provider directed conservative care prior to seeking advanced imaging for many nonspecific complaints.[8-14] The potential clinical benefit of starting conservative care and not needing advanced images or additional procedures may outweigh the potential harm of a brief delay in obtaining said imaging.

MRI is commonly recommended as the next image technology of choice.[1-14] The use of contrast media during imaging will depend on the clinical presentation and the particular tissues suspected to be injured. When x-rays are negative and there is suspicion of soft tissue involvement MRI can be helpful to visualize tissue damage and effusion. CT may also be helpful in situations where MRI is contraindicated, or x-rays were unclear and bony involvement continues to be suspect.[1-14]

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Post-Operative Follow-Up (MS-16)

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Joint Replacement Post-Operative Follow-Up (MS-16.1)

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As outlined in General Guidelines (MS-1.0), the following must be met prior to requesting advanced imaging for further investigation and/or treatment planning of joint replacement post-op symptoms (e.g., aseptic loosening, infection, fracture, pseudotumor, implant performance):

- Plain x-ray imaging of the suspected area must first be performed after the current post-operative episode of symptoms started with results available to the requesting provider
- See specific details throughout the section regarding any requirements for when conservative care prior to requesting advanced imaging is required
- For suspected periprosthetic infection - Joint aspiration culture is the initial evaluation after plain x-ray for a painful joint replacement when periprosthetic infection is suspected
- Criteria regarding any **nuclear medicine imaging** of suspected post-operative joint replacement complications (e.g., aseptic loosening, peri-prosthetic fracture, suspected infection) is found in: **Nuclear Medicine (MS-28)**

Advanced imaging for further investigation of joint replacement post-operative follow-up is medically necessary as follows:

- **Shoulder**:
  - Suspected aseptic loosening or fracture – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221), OR CT without contrast (CPT® 73200)
  - Suspected infection with negative or inconclusive aspiration culture – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73321), MRI without and with contrast (CPT® 73223) OR CT with contrast (CPT® 73201)
  - Possible rotator cuff tear – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221) OR CT with contrast (CPT® 73201)
  - Possible nerve injury – US (CPT® 76881 or CPT® 76882) OR MRI without contrast (CPT® 73221)
- **Elbow/Wrist**:
  - Suspected aseptic loosening or fracture – CT without contrast (CPT® 73200)
  - Suspected infection with negative or inconclusive aspiration culture – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221), MRI without and with contrast (CPT® 73223) OR CT with contrast (CPT® 73201)

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- **Hip**:
  - Suspected aseptic loosening or fracture – CT without contrast (CPT® 73700)
  - Suspected infection with negative or inconclusive aspiration culture – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73721), MRI without and with contrast (CPT® 73723), CT with contrast (CPT® 73701), OR CT without contrast (CPT® 73700)
  - Suspected particle disease (aggressive granulomatosis) after infection has been excluded – MRI without contrast (CPT® 73721), OR CT without contrast (CPT® 73700)
  - Suspected periprosthetic fracture after non-diagnostic x-ray – CT without contrast (CPT® 73700)
  - Suspected component malposition or heterotopic bone – CT without contrast (CPT® 73700)
  - Suspected metal-on-metal implant with high-risk of performance issues from cup-neck impingement and adverse local tissue reaction (ALTR, with Co and Cr ion levels greater than 10 ppb) – US (CPT® 76881 or CPT® 76882) OR MRI without contrast (CPT® 73721)
  - Suspected Aseptic lymphocytic-dominated vasculitis lesion (ALVAL) pseudotumor surrounding metal-on-metal prosthesis – US (CPT® 76881 or CPT® 76882) OR MRI without contrast (CPT® 73721)
  - Suspected tendinitis/bursitis, abductor injury or other soft tissue abnormality – US (CPT® 76881 or CPT® 76882) OR MRI without contrast (CPT® 73721)
    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
  - Possible nerve injury – MRI without contrast (CPT® 73721)
- **Knee**:
  - Suspected aseptic loosening or fracture – MRI without contrast (CPT® 73721) OR CT without contrast (CPT® 73700)
  - Suspected infection with negative or inconclusive aspiration culture – US (CPT® 76881 or 76882), MRI without contrast (CPT® 73721), MRI without and with contrast (CPT® 73723) OR CT with contrast (CPT® 73701)
  - Suspected periprosthetic fracture – MRI without contrast (CPT® 73721), OR CT without contrast (CPT® 73700)
  - Suspected osteolysis or component instability, rotation, or wear – MRI without contrast (CPT® 73721), OR CT without contrast (CPT® 73700)
  - Suspected periprosthetic soft tissue abnormality unrelated to infection (e.g. tendinopathy, arthrofibrosis, patellar clunk, impingement of nerves or soft tissue) - US (CPT® 76881 or CPT® 76882), OR MRI without contrast (CPT® 73721)

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    - Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started is required
- **Ankle**:
    - Suspected aseptic loosening or fracture – CT without contrast (CPT® 73700)
    - Suspected infection with negative or inconclusive aspiration culture – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73721), MRI without and with contrast (CPT® 73723) OR CT with contrast (CPT® 73701)

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General Musculoskeletal Post-Operative Follow-Up (MS-16.2)

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As outlined in General Guidelines (MS-1.0), the following must be met prior to requesting advanced imaging for further investigation and/or treatment planning of general musculoskeletal post-operative (post-op) symptoms (e.g., fracture fixation, tendon/ligament repair, labral repair):

- Plain x-ray imaging of the suspected area must first be obtained after the current post-operative episode of symptoms started and results are available to the provider
- Failure of 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current post-op episode of symptoms started

When all of the above criteria have been met, further investigation in symptomatic individuals is medically necessary for post-operative follow-up as follows:

- **Shoulder**:
  - Post rotator cuff repair – US (CPT® 76881 or CPT® 76882), MRI without contrast (CPT® 73221), MRI with contrast (CPT® 73222) OR CT with contrast if MRI contraindicated (CPT® 73201)
  - Post impingement or labral repair - MRI without contrast (CPT® 73221), MRI with contrast (CPT® 73222) OR CT with contrast if MRI contraindicated (CPT® 73201)
- **Elbow**:
  - Post complex fracture surgery – CT without contrast (CPT® 73200)
  - Post soft-tissue surgery – MRI without contrast (CPT® 73221)
- **Wrist**:
  - Post complex fracture surgery – CT without contrast (CPT® 73200)
  - Post instability and/or ligament surgery (e.g., TFCC, DRUJ instability) surgery – MRI with contrast (CPT® 73222)
- **Hand**:
  - Post soft-tissue or complex fracture surgery – MRI without contrast (CPT® 73218) OR CT without contrast (CPT® 73200)
- **Pelvis**:
  - Post complex pelvic ring/acetabular fracture surgery – CT without contrast (CPT® 72192)
- **Hip**:

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  - Fracture surgery or post-op for avascular necrosis – MRI without contrast (CPT® 73721) or CT without contrast (CPT® 73700)
  - Post Femoroacetabular Impingement or labral repairs – MRI with contrast (CPT® 73722)
- **Knee**:
  - Post fracture or dislocation surgery – CT without contrast (CPT® 73700)
  - Post ligament reconstruction or meniscus tear surgery – MRI without contrast (CPT® 73721) OR MRI with contrast (CPT® 73722)
- **Ankle**:
  - Post complex fracture surgery – CT without contrast (CPT® 73700)
  - Post ligament or tendon injury surgery – US (CPT® 76881 or CPT® 76882) OR MRI without contrast (CPT® 73721)
- **Foot**:
  - Post complex fracture surgery – CT without contrast (CPT® 73700)
  - Post ligament, tendon or plantar plate surgery – US (CPT® 76881 or CPT® 76882) OR MRI without contrast (CPT® 73718)

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Evidence Discussion (MS-16)

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The American College of Radiology (ACR) has recommended plain x-rays as the initial study for routine follow up of asymptomatic patients and symptomatic individuals who have undergone surgery for musculoskeletal injury.[1-4] Plain x-rays can identify fractures and show signs of hardware loosening, wear, osteolysis or infection. Blood tests and joint aspiration with synovial fluid laboratory analysis may be sufficient to diagnosis post-surgical infection. Joint aspiration with synovial fluid analysis remains the most useful test for confirming the presence or absence of infection and identifying the causative organism.[2-7] When plain x-rays, and laboratory tests as applicable, are negative or inconclusive and there is a suspicion of post-surgical complications, advanced imaging can help to identify what may be the cause of the individual's symptoms.

When there is suspicion of a post-surgical soft tissue abnormality (e.g., tendinitis, tendinopathy, bursitis, arthrofibrosis), a course of conservative care prior to advanced imaging can allow many individuals to improve[8,9], thus eliminating the need for advanced imaging. This may avoid unnecessary exposure to radiation from imaging and additional costs of imaging. If there is failure to improve, advanced imaging would be appropriate. Suspicion of more significant complications following joint surgery may benefit from advanced imaging without a trial of conservative care.

The ACR further recommended MRI, CT, and US as advanced imaging options when further assessing post-surgical complications. Their recommendations vary based on the clinical scenario.[2-4] Complications with implanted hardware (e.g., loosening, component rotation, hardware fracture) may greatly impair an individual's function and may lead to additional negative sequelae if not addressed in a timely manner. MRI has been identified as accurate diagnostic tools in evaluating aseptic loosening following knee replacement.[10] Suspected torn rotator cuff tendons in individuals that have undergone total shoulder arthroplasty may also be difficult to diagnose without advanced imaging due to the various techniques used to release the subscapularis tendon during surgery.[3] MRI has been demonstrated to have good intra- and inter-rater reliability in determining the structural integrity of tendons following surgery.[11,12] Individuals with metal-on-metal hip replacements are at risk for adverse local tissue reactions including metallosis, pseudotumor and generalized synovitis that could result in tissue damage. Prompt evaluation, including advanced imaging, would be appropriate to best mitigate these risks.[13]

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Tissue Growths, Masses and Cysts (MS-10)

Guideline

Soft Tissue Mass and Morton's Neuroma (MS-10.1)
Bone Lesion (MS-10.2)
Cysts (MS-10.3)
Evidence Discussion (MS-10)
References (MS-10)

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Soft Tissue Mass and Morton's Neuroma (MS-10.1)

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As outlined in General Guidelines (MS-1.0), the following must be met prior to requesting advanced imaging for further investigation and/or treatment planning of soft tissue masses and Morton's neuroma:

- Initial plain x-ray imaging of the suspected area must first be performed after the current episode of symptoms started. Results of these plain x-rays need to be available to the requesting provider
- Additional documentation of clinical features if specifically stated below
- See specific details throughout the section regarding any requirements for when conservative care prior to requesting advanced imaging is required

Soft Tissue Mass

Plain x-ray is indicated as the initial imaging study, with the exception of individuals with cancer predisposition syndrome (If there is a known or suspected soft tissue mass in an individual with a cancer predisposition syndrome, see: Screening Imaging in **Cancer Predisposition Syndromes (PEDONC-2)** in the Pediatric and Special Populations Oncology Imaging Guidelines). History and physical exam of any palpable soft tissue mass should include documentation of any one or more of the following clinical features:

- Increase in volume/size
- More than 5cm in diameter
- Painful
- Deep or subfascial location

After the initial history, exam and plain imaging have been performed and further investigation is needed the following advanced imaging is medically necessary for evaluation of soft tissue masses:

- MRI without and with contrast, or MRI without contrast, or US of the area of interest (CPT® 76881 or 76882) is appropriate when ANY of the following are met after plain x-ray:
  - Soft tissue mass(es) which are increasing in volume/size, more than 5 cm in diameter, painful or deep or in a subfascial location
  - Surgical planning

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- CT with contrast or CT without and with contrast is appropriate when MRI is contraindicated or after a metal limiting MRI evaluation

Advanced imaging is not medically necessary for subcutaneous lipoma with no surgery planned

Tarsal Tunnel Syndrome soft tissue mass entrapment (Baxter's Neuropathy)

After the x-rays have been performed and results are available to the provider, and there has been failure of at least 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started, the following advanced imaging is medically necessary for preoperative planning if mass is suspected as etiology of entrapment:

- US Foot (CPT® 76881 or CPT® 76882) or MRI Foot without contrast (CPT® 73718)

Morton's Neuroma

After the x-rays have been performed and results are available to the provider, and there has been failure of at least 6 weeks of provider-directed conservative treatment, including clinical re-evaluation, occurring after the current episode of symptoms started, the following advanced imaging is medically necessary for further investigation of Morton's Neuroma for preoperative planning:

- US Foot (CPT® 76881 or CPT® 76882), MRI Foot without contrast (CPT® 73718) or MRI Foot without and with contrast (CPT® 73720)

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Bone Lesion (MS-10.2)

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As outlined in General Guidelines (MS-1.0), the following must be met prior to requesting advanced imaging for further investigation and/or treatment planning of suspected abnormal bone growth or lesion:

- Initial plain x-ray imaging of the entire bone containing the lesion must first be performed after the current episode of symptoms started. Results need to be available to the requesting provider.

After the x-rays have been performed and further investigation is needed advanced imaging is medically necessary for the following scenarios:

- Diagnosis uncertain based on plain x-ray appearance - MRI without contrast, MRI without and with contrast, or CT without contrast
- Imaging requested for preoperative planning – MRI without contrast, MRI without and with contrast, or CT without contrast
- Plain x-ray reveals an osteochondroma with clinical concern of malignant transformation - MRI without contrast or MRI with and without contrast
- For Paget's Disease - MRI (contrast as requested) can be considered if the diagnosis (based on plain x-rays and laboratory studies) is in doubt or malignant degeneration is suspected
    - Criteria regarding **nuclear medicine imaging** is found in: **Nuclear Medicine (MS-28)**

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Cysts (MS-10.3)

As outlined in General Guidelines (MS-1.0), the following must be met prior to requesting advanced imaging for treatment planning of soft tissue masses and cysts:

- Initial plain x-rays must be performed after the current episode of symptoms started or changed. Results need to be available to the requesting provider.

After the initial imaging has been performed and further investigation is needed for surgical planning to address ganglion or other musculoskeletal related cysts, (e.g., Popliteal) the following advanced imaging is medically necessary:

- **Ganglion Cyst** - US (CPT® 76881 or CPT® 76882), MRI without contrast or MRI without and with contrast
- **Popliteal (Baker's) Cyst** - US (CPT® 76881 or CPT® 76882), MRI Knee without contrast (CPT® 73721)

Advanced imaging is not medically necessary for:

- Ganglia that can be diagnosed by history and physical examination
- Sebaceous cyst, as these cysts are benign and additional imaging typically will not change the treatment plan

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Evidence Discussion (MS-10)

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The American College of Radiology (ACR) has recommended the use of x-rays as the initial image when evaluating suspected bone tumor. They recommended that a complete x-ray of the entire lesion-containing bone be evaluated prior to considering any advanced imaging.[1] Many benign bone tumors have a characteristic appearance on plain x-ray. Along with a relevant history and physical examination, x-rays may be sufficient for diagnosis and would avoid additional cost and risks associated with advanced images. If plain x-rays are equivocal, they may still direct further imaging with the correct modality. Furthermore, plain x-ray may provide complementary information to advanced imaging, allowing for a better overall interpretation. Clearly benign findings on exam (lipoma, ganglion, sebaceous cyst) do not need additional imaging prior to treatment unless the imaging was necessary for surgical management or for a possible malignancy.

When diagnostic uncertainty remains and concerns for malignant degeneration still exist, or imaging is requested for surgical planning, advanced imaging would be medically necessary. MRI is a superior modality for evaluation of many bone tumors. The ACR recommended MRI as the primary modality for advanced imaging of primary bone tumors. MRI can be useful in narrowing down differential diagnoses and in determining the need for biopsy.[1] CT may be appropriate when an individual presents with contraindications to MRI.

Current ACR recommendations have supported plain x-ray as an initial imaging option when evaluating individuals with suspicion of superficial soft tissue masses.[2] X-ray can be helpful in visualizing soft tissue masses and aid in further management planning. It is often the modality of choice for initial imaging. US is being recommended as an excellent triage tool more frequently for soft tissue masses that are more superficial, further supporting the recommendation for its use by the ACR.[3,4]US has been found to have high accuracy in defining superficial (superior to investing fascia) soft tissues masses with a sensitivity of 93% and specificity of 98%.[5] A recent study found that US was more predictive of the histology of lipomatous tumors than the use of predictive clinical indicators.[6]

Common clinical features of soft tissue sarcoma can be helpful in the differentiation of soft tissue masses and decisions for additional biopsy. In many cases, a thorough exam, history and initial imaging may be sufficient to determine a diagnosis and management plan.[7] When clinical findings and initial imaging is insufficient to diagnose suspicious soft tissue masses MRI without and with IV contrast is primarily recommended as the next advanced image of choice.[2-4] MRI capabilities present many advantages to other imaging techniques for the use of advanced diagnostics of soft tissue masses. Improved

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contrast, multiplanar views, visualization of involved vascular structures and better demarcation between tissue types/levels (with the aid of contrast agents) are among the most prominent advantages to MRI.[2,3] When MRI is contraindicated, CT imaging without and with IV contrast can provide similarly diagnostic data as MRI and is considered an important adjunct option to MRI.[2]

ACR imaging recommendations supports both plain xray and ultrasound as initial imaging options when evaluating individuals with suspicion of soft tissue cysts.[2] While x-ray findings may not fully demonstrate a small ganglionic mass they can still be beneficial in visualizing the surrounding anatomy and in determining the next steps for care. In a recent systematic review of 941 ganglion cyst cases MRI and US were the most commonly reported imaging study. It was recommended that US can be a beneficial option for triage. However, it was found that MRI was much better at showing the underlying pathoanatomy of masses and their surrounding area than US when evaluating intraneural ganglion cysts.[8] The utility of MRI for preoperative visibility of dorsal wrist ganglion and any associated pathoanatomy has also been demonstrated. MRI would be more useful for preoperative planning.[9]

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General (Neck-1)

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Abbreviations for Neck Imaging Guidelines

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Abbreviations for Neck Imaging Guidelines	
ACSS	anterior cervical spine surgery
ALS	amyotrophic lateral sclerosis
CT	computed tomography
ENT	Ear, Nose, Throat
FNA	fine needle aspiration
FEES	fiberoptic endoscopic evaluation of swallowing
FFL	flexible fiberoptic laryngoscopy
FOE	fiberoptic exam for laryngoscopy
GERD	gastroesophageal reflux disease
GI	gastrointestinal
HIV	human immunodeficiency virus
HPV	human papillomavirus
MEN-2	multiple endocrine neoplasia
MRI	magnetic resonance imaging
TNE	transnasal esophagoscopy
VFSS	videofluoroscopic swallow study

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General Guidelines (Neck-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms including a detailed history, physical examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.
- Advanced imaging of the neck covers the following areas:
  - Skull base (thus, a separate CPT® code for head imaging in order to visualize the skull base is not medically necessary)
  - Nasopharynx
  - Upper oral cavity to the head of the clavicle
  - Parotid glands and the supraclavicular region
- Ultrasound of neck soft tissues including thyroid, parathyroid, parotid and other salivary glands, lymph nodes, cysts, etc. is coded as CPT® 76536. This can be helpful in more ill-defined masses or fullness and differentiating adenopathy from mass or cyst, to define further advanced imaging.
- CT Neck
  - CT Neck is usually obtained with contrast only (CPT® 70491).
    - With the exception of 4D CT Neck without and with contrast (CPT® 70492) for parathyroid adenoma localization, little significant information is added by performing a CT Neck without and with contrast (CPT® 70492), and there is the risk of added radiation exposure, especially to the thyroid.
    - CT Neck without contrast (CPT® 70490) can be difficult to interpret due to difficulty identifying the blood vessels.
      - Exceptions include:
        - Contrast is generally not medically necessary when evaluating known or suspected tracheal anomalies with CT.
        - Additionally, non-contrast CT is medically necessary for the evaluation of salivary duct stones in the appropriate clinical circumstance where intravenous contrast may obscure high attenuation stones. Dual-phase CT imaging (without and with IV contrast) is not medically necessary in this situation.
    - Contrast enhanced CT is helpful in the assessment of cervical adenopathy and preoperative planning, including in the setting of thyroid carcinomas.

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  - Contrast may cause intense and prolonged enhancement of the thyroid gland which interferes with radioactive iodine nuclear medicine studies.
  - Use of IV contrast is an important adjunct, however, because it helps to delineate the anatomic relationship between the primary tumor and metastatic disease. Iodine is generally cleared within four to eight weeks in most individuals, so concern about iodine burden from IV contrast causing a clinically significant delay in subsequent whole-body scans (WBSs) or radioactive iodine (RAI) treatment after the imaging followed by surgery is generally unfounded. The benefit gained from improved anatomic imaging generally outweighs any potential risk of a several week delay in RAI imaging or therapy. Where there is concern, a urinary iodine to creatinine ratio can be measured.

- MRI Neck
  - MRI Neck is used less frequently than CT Neck.
  - MRI Neck without and with contrast (CPT® 70543) is medically necessary if CT suggests the need for further imaging or if ultrasound or CT suggests any of the following:
    - Neurogenic tumor (schwannoma, neurofibroma, glomus tumor, etc.)
    - Vascular malformations
    - Deep neck masses
    - Angiofibromas
    - Cystic neck mass
    - Concern for malignancy (see **Squamous Cell Carcinomas of the Head and Neck (ONC-3)**, **Salivary Gland Cancers (ONC-4)**, or **Thyroid Cancer (ONC-6)** as appropriate)
  - MRI Neck without and with contrast (CPT® 70543) is medically necessary if the head and neck surgeon or neurosurgeon, or the provider in consultation with the head and neck surgeon or neurosurgeon, has reasonable clinical concern:
    - for a skull base or nasopharyngeal neoplasm, or potential perineural invasion/ cranial nerve involvement.[2]
    - that extensive dental amalgam may obscure the anatomy on CT in individuals with oral cavity neoplasm.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence;

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education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Dysphagia and Upper Digestive Tract Disorders (Neck-3)

Guideline

Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)
References (Neck-3)

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Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)

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- **General considerations**
  - A detailed history of the dysphagia symptoms is important to distinguish neurogenic, pharyngeal and esophageal disorders.
  - Dysphagia (difficulty swallowing) can be the result of a wide range of benign and malignant processes that affects the body's ability to move food or liquid from the mouth to the pharynx and into the esophagus.
  - A short duration (weeks to months) of rapidly progressive esophageal dysphagia with associated weight loss is highly suggestive of esophageal cancer.
  - Advanced imaging for individuals presenting with isolated globus rarely impacts clinical management. In a study of 148 neck CTs and 104 barium esophagrams done for the evaluation of globus sensation, there were no malignancies detected.
- **Gastroesophageal reflux disease (GERD)**
  - Advanced imaging is not medically necessary for the evaluation of GERD, the diagnosis of which is usually made on the basis of clinical history, in conjunction with endoscopy, pH monitoring, Upper GI Barium Studies, and occasionally manometry. Exceptions would include the following:
    - Non-cardiac chest pain suspected of being GERD should be evaluated first to exclude cardiac and other etiologies. See **Non-Cardiac Chest Pain-Imaging (CH-4.1)** in the Chest Imaging Guidelines.
    - Gastric emptying study (CPT® 78264) for individuals with refractory GERD symptoms, and gastroparesis is being considered.
- **Suspected foreign body impaction and ingested foreign bodies**
  - Plain x-rays are medically necessary as initial imaging.
  - If imaging is inconclusive, and/or there is suspicion of a radiolucent foreign body (such as fish or chicken bones, wood, plastic, thin metal objects, aluminum can pop-ups, etc.):
    - CT Neck and/or Chest with or without contrast (CPT® 70491 or CPT® 70490 or CPT® 71260 or CPT® 71250)
    - If requested, 3-D reconstruction (CPT® 76377 or CPT® 76376) is medically necessary in this setting.
  - The use of oral contrast is discouraged (to avoid the aspiration of contrast material) for acute dysphagia or foreign body impaction, as the contrast may not pass, may be aspirated, and can interfere with subsequent endoscopic intervention.
- **Oropharyngeal dysphagia**

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- Oropharyngeal dysphagia (difficulty in transferring food from the mouth to the pharynx)
  - Suspected neurologic causes: See appropriate sections in Head Imaging Guidelines.
  - Initial evaluation is with A) a swallow study AND B) direct visualization (with laryngoscopy and/or upper endoscopy).
    - A) Video fluoroscopic swallowing study (VFSS) – (Dynamic radiographic evaluation of swallowing during speech pathologist-guided oral intake of various consistencies). Also referred to as a modified barium swallow study (MBSS).
    - B) Flexible fiberoptic laryngoscopy (FFL), FiberOptic Exam for laryngoscopy (FOE), TransNasal Esophagoscopy (TNE) and/or FEES (Fiberoptic Endoscopic Evaluation of Swallowing). FEES is a dynamic evaluation of swallowing via direct visualization using transnasal laryngoscopy during speech pathologist-guided oral intake of various consistencies.
    - CT Neck with contrast (CPT® 70491) is medically necessary for any documented anatomic abnormalities suggested by direct visualization—i.e., on exam with flexible laryngoscopy or rigid video stroboscopy or FEES.
      - Completion of a radiographic swallow study, though potentially helpful, is NOT medically necessary prior to the requested advanced imaging in such a case.

- **Esophageal dysphagia**
  - Esophageal dysphagia (difficulty in transferring food down the esophagus in the retrosternal region, e.g., food sticking in the neck or chest)
    - Initial evaluation is with barium esophagram or upper gastrointestinal endoscopy.
    - If requested, esophageal manometry is medically necessary
    - Advanced imaging is medically necessary for the evaluation of structural abnormalities demonstrated on either esophagram or direct visualization (i.e., laryngoscopy/upper GI endoscopy), such as an external compression, tumor, stricture, diverticulum, etc.
      - Contrasted CT Neck (CPT® 70491), CT Chest (CPT® 71260), **and/or** CT Abdomen (CPT® 74160) depending on the location of the abnormality identified.
- **Suspected perforation, abscess, or fistula**
  - CT Neck, Chest, **and/or** Abdomen, contrast is medically necessary (preferably with contrast- CPT® 70491, CPT® 71260, CPT® 74160), depending on location.
- **Hiatal hernia**
  - See **Hiatal Hernia (AB-12.3)** in the Abdomen Imaging Guidelines.
- **Globus sensation**

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- Globus sensation is a feeling of a lump or foreign body in the throat. In general, laryngoscopy, endoscopy, and physical examination will rule out malignant causes and advanced imaging is not medically necessary for evaluation. It is considered a mild form of dysphagia.
  - Direct visualization with laryngoscopy and/or upper endoscopy should be performed prior to advanced imaging.
    - Unremarkable laryngoscopy and/or upper endoscopy does not preclude advanced imaging if **red flag** symptoms are also present:
      - weight loss
      - odynophagia/throat pain
      - referred otalgia
      - hoarseness
      - hemoptysis, **AND/OR**
      - other unilateral presentation of concerning symptoms.
    - CT Neck with contrast (CPT® 70491) for ANY of the following:
      - Negative or equivocal findings on laryngoscopy and/or upper endoscopy with any **red flag** present
      - Known history of upper aerodigestive or esophageal malignancy
      - Known history of lymphoma
      - History of previous neck, esophageal, or gastric surgery–see below, and see **Background and Supporting Information** for post-operative oropharyngeal dysphagia associated with Anterior Cervical Spine Surgery (ACSS).
      - Palpable abnormality on physical examination such as neck mass

- **Post-operative dysphagia**
  - Dysphagia following surgery on the oropharynx, soft tissues of the neck, cervical spine, esophagus, or stomach:
    - In the immediate post-operative period, within 3 months of the surgery, the concern is for fluid collections, anastomotic leaks, perforations, and abscess. Prior laryngoscopy/upper endoscopy and barium esophagram are not medically necessary initially.
      - CT Neck with contrast (CPT® 70491) **AND/OR** CT Chest with contrast (CPT® 71260)
    - In the delayed post-operative period—three months or greater from surgery, the primary modalities for evaluation are history, physical, endoscopy (laryngoscopy or EGD) and/or barium esophagram/videofluoroscopic swallow study (VFSS) to direct any additional advanced imaging. See also **Background and Supporting Information**.
      - If the results of endoscopy and/or barium esophagram or VFSS are abnormal or inconclusive, the following is medically necessary:

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    - CT Neck with contrast (CPT® 70491) **AND/OR** CT Chest with contrast (CPT® 71260)
- **Suspected vascular ring** (See **Dysphagia (PEDNECK-5)**)
    - Advanced imaging can be performed if a vascular ring is suspected by, or in consultation with, the treating specialty, i.e., cardiothoracic surgery, cardiology, otolaryngology, and/or pulmonology. More commonly, this congenital pathology would be suspected in a much younger population; however, dysphagia lusoria is a relatively rare condition involving a vascular ring (usually an aberrant right subclavian artery). As children, these individuals are asymptomatic but develop worsening dysphagia later in adulthood, presumably secondary to increasing calcification and blood pressure.
        - CTA Chest (CPT® 71275) **OR** MRA Chest (CPT® 71555) are the preferred imaging studies in the evaluation of a suspected vascular ring.
        - CT Chest with contrast (CPT® 71260) **OR** MRI Chest without contrast (CPT® 71550) **OR** MRI Chest without and with contrast (CPT® 71552) can be performed as alternative exams in the evaluation of suspected vascular ring.

Background and Supporting Information

- Postoperative oropharyngeal dysphagia is one of the most common complications following anterior cervical spine surgery (ACSS), and is considered by some to be an inevitable result of this surgery, rather than a surgical complication.
- Severe dysphagia after ACSS should prompt immediate evaluation to exclude any potentially reversible surgical complication such as bone graft dislodgement, hematoma or retropharyngeal abscess.
- In general, history, exam, plain films, laryngoscopy, and videofluoroscopic swallow studies are considered the primary modalities for evaluation. The videofluoroscopic swallow study, in fact, is the gold standard in evaluation, and is very sensitive in individuals post-ACSS, and should be considered the initial evaluation in individuals who are status post ACSS with globus sensation, or mild dysphagia.

Evidence Discussion

Dysphagia

- Imaging studies are complementary to endoscopy (and in certain cases, also manometry) in the evaluation of dysphagia. The optimal imaging study depends on the nature and location of the dysphagia, as well as clinical setting. Fluoroscopy, however, is usually the first line choice.[4]
- Modified barium swallow study (videofluoroscopic procedure performed in conjunction with a speech therapist) is generally first line for oropharyngeal dysphagia. CT is generally not medically necessary because it does not assess motility, nor the oropharyngeal and esophageal mucosa as well. CT may be helpful for subsequent

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evaluation if initial studies are not revealing, or if there are suspicious findings on direct visualization.[10]

- Barium esophagram is medically necessary for initial imaging of retrosternal dysphagia. CT is generally not medically necessary because it does not assess motility, nor esophageal mucosa as well. CT may be helpful if subsequent evaluation if initial studies are not revealing, or if there are suspicious findings on direct visualization and/or barium esophagram.[10]
- Fluoroscopy still remains the imaging of choice for initial evaluation of early (as well as late) post-operative dysphagia, within the oropharyngeal or retrosternal regions. Utilizing a water-soluble contrast first, followed by barium if necessary, suspected leaks or fistulas may be investigated. Esophagrams are highly specific for leaks, but not as sensitive as CT. If high clinical suspicion remains following negative esophagram, esophagography and CT combined have a sensitivity and negative predictive value of 100%, but a specificity of 27% and positive predictive value of 56%.[10]
- For oropharyngeal and retrosternal dysphagia, contrasted CT of the neck and/or chest is medically necessary when there is concern of early post-operative complications, such as leak, fluid collection, abscess, or hematoma.[10]

Dysphagia and Upper Digestive Tract Disorders Health Equity Considerations

- Dysphagia is more common in individuals who are female, White/Caucasian, non-Hispanic, of older age, low income, of public insurance, and unemployed.
- Individuals who are female and White/Caucasian are less likely to pursue diagnostic testing for upper digestive tract disorders.
- GERD is more common in females, but the rate of complications is higher in males.
- Hispanic and Latino Americans have the highest prevalence of GERD among all minority populations.
- Reflux is more common in individuals who are female, White/Caucasian, non-Hispanic, and of public insurance.

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Neck Mass/Swelling/ Adenopathy (Neck-5)

Guideline

Neck Mass/Swelling/Adenopathy (Neck-5.1)
References (Neck-5)

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Neck Mass/Swelling/Adenopathy (Neck-5.1)

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- Cervical lymphadenitis is common and follows most viral or bacterial infections of the ears, nose and throat. Painful acute lymphadenopathy should be treated with a trial of conservative therapy for 2-weeks, including antibiotics if appropriate. If there is improvement with conservative treatment, advanced imaging is not medically necessary. If the adenopathy persists, it can be imaged as per below**.**
- Ultrasound (CPT® 76536) is medically necessary for **ANY** of the following:
  - Cervical adenopathy/lymphadenitis or an inflammatory, infective, or reactive mass that has failed a 2-week trial of treatment (including antibiotics if appropriate) or observation
  - Anterior neck masses
  - Posterior neck masses
  - Any ill-defined mass, fullness or asymmetry
- CT Neck with contrast (CPT® 70491) is medically necessary if:
  - Neck mass with any ONE of the following:
    - Size ≥1.5cm
    - Mass present ≥2 weeks or of uncertain duration
    - Non-tender neck masses
    - Firm texture or fixation of the mass
    - Suspected peritonsillar, retropharyngeal or other cervical space abscess
    - Ulceration of skin overlying the neck mass
    - Ear pain ipsilateral to the neck mass
    - Associated onset of hoarseness persistent for greater than 3-weeks
    - Associated onset of throat pain, tonsil asymmetry, oral or oropharyngeal ulceration, weight loss, or hemoptysis
    - History of malignancy that would be primary or metastatic to the neck
    - Prior ultrasound results, if performed, are suspicious or indeterminate for malignancy
  - Isolated tonsil asymmetry with concerning features such as suspicious appearance, firmness, palatal immobility, rapid unilateral enlargement, history of malignancy, or immune compromise.
  - Carcinoma found in a lymph node or other neck mass
  - Suspected or known sarcoidosis
  - Preoperative evaluation of any neck mass

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  - Foreign body migration with neck swelling
- MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary if:
  - CT suggests the need for further imaging
  - Ultrasound or CT suggests neurogenic tumor (schwannoma, neurofibroma, glomus tumor, etc.), vascular malformations, cystic neck mass, deep neck masses, or angiofibroma.
- MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is also medically necessary without prior CT Neck or ultrasound requirement, if the head and neck specialist, or the provider in consultation with the head and neck specialist, has reasonable clinical concern for:
  - skull base or nasopharyngeal neoplasm, **OR**
  - potential perineural invasion/cranial nerve involvement, **AND/OR**
  - extensive dental amalgam which may obscure the anatomy on CT in individuals with oral cavity neoplasm.
- Congenital cervical sinus, fistula, or cyst for preoperative planning:
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) **OR**
  - CT Neck with contrast (CPT® 70491)
    - For fourth branchial cleft cyst/sinus/fistula, barium swallow is medically necessary in addition to the above conventional imaging.

Background and Supporting Information

- Inflammatory neck adenopathy is often associated with upper respiratory infection, pharyngitis, dental infection, HIV and toxoplasmosis. Occasionally it is associated with sarcoidosis and tuberculosis.
- Malignancy is a greater possibility in adults that are heavy drinkers and smokers, but HPV associated disease is on the rise and there can be a high suspicion for malignancy even without these traditional risk factors.
- ENT evaluation can be helpful in determining the need for advanced imaging.
- Although CT and MRI can have characteristic appearances for certain entities, biopsy and histological diagnosis are the only way to obtain a definitive diagnosis. The preferred initial method of biopsy is Ultrasound guided core needle biopsy of the mass.
- The most common causes of neoplastic cervical adenopathy are metastasis from head and neck tumors or lymphoma.
- Tonsil asymmetry is a common exam finding in both adults and children and is often benign. In cases of associated suspicious characteristics, neck imaging is medically necessary, even if there is not an associated neck mass.

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- MRI has great specificity for determining the boundaries and prevalence of developmental neck cysts. It may thus be considered optimal to use only MRI, which leads to a correct diagnosis in more than 90% of cases.
- For the evaluation of neck masses by CT or MRI, the diagnostic accuracy is 96%, the sensitivity is 96.30%, and the specificity is 95.65%.

Evidence Discussion

- CT has several benefits that support its utilization as a primary imaging modality, including its availability and cost. CT imaging is generally easily tolerated by individuals because of short scanning time (<5 minutes) and large scanner bore. While CT utilizes ionizing radiation, the average dose of 3 mSv (equivalent to approximately 150 chest x-rays) is considered acceptable in the adult population.[17]
- MRI is preferred for tumors of the nasopharynx or when there is a cranial nerve concern on physical examination because of its sensitivity to abnormalities of the skull base and in the detection of perineural spread. MRI also offers improved tissue contrast and can help detect subclinical tumors not evident with nasal endoscopy.[17]
- Ultrasound is also used to characterize neck masses, to guide tissue sampling, and to search for additional masses. It is both noninvasive and inexpensive. Ultrasound is, however, best suited for evaluation of superficial tissue, in situations where there will be a delay in obtaining CT or MRI, if the use of contrast medium is contraindicated, or as an adjunct to expedite FNA biopsy.[11]

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Recurrent Laryngeal Nerve Palsy (Neck-7)

Guideline

Recurrent Laryngeal Nerve Palsy (Neck-7.1)
References (Neck-7)

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Recurrent Laryngeal Nerve Palsy (Neck-7.1)

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- The following are medically necessary with new diagnosis of idiopathic unilateral vocal fold paralysis/immobility or weakness, as identified on videostroboscopy or laryngoscopy by an Otolaryngologist—Head and Neck surgeon, or a clinician in consultation with such a specialist:
  - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)

    AND
  - MRI Orbit/Face/Neck with and without contrast (CPT® 70543) **OR** CT Neck with contrast (CPT® 70491)

    AND
  - CT Chest with contrast (CPT® 71260) is medically necessary for left vocal fold paralysis and may be medically necessary for right vocal fold paralysis. If requested, see **Background and Supporting Information**.

Background and Supporting Information

- The right and left recurrent laryngeal nerves supply the motor innervation of the right and left vocal folds/cords, respectively. They are branches off of the Vagus Nerve, CN X. The entire pathway from origin to endpoint of this nerve must be visualized in cases of presumed idiopathic vocal fold paralysis, newly identified by laryngoscopy, to search for a possible cause. From the origin of the vagus at the medulla oblongata to the looping down into the superior mediastinum and back to the neck of its branching nerve (the recurrent laryngeal nerve)—advanced imaging is medically necessary to screen for a cause for otherwise idiopathic vocal fold paralysis/paresis. The greater the degree of motion impairment, the more likely it is to find a cause on imaging.
- The superior mediastinum is most noteworthy for containing the take-off point of the three great branches of the aortic arch: the brachiocephalic trunk (also known as the innominate artery), the left common carotid artery, and the left subclavian artery.
- The extent of the CT Neck with contrast, to be inclusive of the entirety of the course of the recurrent laryngeal nerve in question, would have to extend to the "thoracic inlet" portion of the superior mediastinum on the right, and the "aortic triangle" portion of the superior mediastinum on the left.
- Contrasted CT Chest is medically necessary with left vocal cord palsy due to the lower course of the recurrent laryngeal nerve branch on the left side of the body. It curves inferior to the aortic arch and ascends in the groove between the trachea and

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the esophagus. However, the course of the recurrent laryngeal nerve on the right side may be as low as the level at which the brachiocephalic artery meets the subclavian artery, and this area of the thoracic inlet may or may not be contained within the anatomic extent of a CT neck at all institutions.

Evidence Discussion

Evaluation of Idiopathic Unilateral Vocal Fold Paralysis/Immobility

- Idiopathic unilateral vocal fold immobility is a diagnosis of exclusion. When it is noted that a vocal cord is immobile on laryngoscopy, the etiology must be sought. If it is not apparent on history and laryngoscopy, this work-up involves imaging the entire pathway of the motor nerve supply to the involved side of the larynx—from the brain to the superior mediastinum.[1]
- Repeat imaging for a pre-existing diagnosis of idiopathic unilateral vocal cord paralysis may be considered on a case by case basis. Recommendations include possibly repeating advanced imaging as above within 5 years after initial diagnosis and workup, or performing regular (annual) clinical evaluations with imaging reserved for the development of new symptoms or exam findings.[5]

Recurrent Laryngeal Nerve Palsy Health Equity Considerations

- Individuals with a higher socioeconomic status typically receive a greater number of interventions for recurrent laryngeal nerve palsy, as well as a longer duration of care.

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Thyroid and Parathyroid (Neck-8)

Guideline

Thyroid Nodule (Neck-8.1)
Hyperthyroidism and Hypothyroidism (Neck-8.2)
Parathyroid Imaging (Neck-8.3)
Parathyroid Incidentaloma (Neck-8.4)
References (Neck-8)

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Thyroid Nodule (Neck-8.1)

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- **Serum thyrotropin (TSH)** should be measured in the initial evaluation of thyroid nodule/mass/asymmetry/goiter but is not medically necessary for follow-up imaging.
- **Ultrasound (US) Neck (CPT® 76536)** is medically necessary prior to advanced imaging for a palpable thyroid nodule/mass/asymmetry/goiter, including such examples as:
  - Nodules incidentally found on CT, MRI, or PET (focal activity)
  - Palpable solitary nodule
  - Palpable multinodular goiter
  - Suspicion of a nodule in an individual with a difficult neck examination
  - Prior history of neck radiation
  - Family history of medullary carcinoma, multiple endocrine neoplasia (MEN-2), or papillary thyroid carcinoma
  - Unexplained cervical lymphadenopathy
  - Preoperative thyroidectomy
- See **Thyroid Cancer-Surveillance/Follow-Up (ONC-6.4)** for thyroid nodules that are biopsy proven thyroid cancer but are being monitored on active surveillance.
- A thyroid nodule detected for the first time during pregnancy should be managed in the same way as in non-pregnant individuals, except for avoiding the use of radioactive agents for diagnostic and therapeutic purposes.
- **Nuclear scan (CPT®78013 or CPT® 78014)** is medically necessary if the serum TSH is subnormal and ANY of the following:
  - Single or multiple thyroid nodules
  - Suspicion of ectopic thyroid tissue
  - Presence of thyroid nodule in the setting of Grave's disease
- **Nuclear medicine thyroid scan (CPT® 78013 or CPT® 78014)** is medically necessary for ANY of the following (TSH is not medically necessary prior to imaging in the below settings):
  - Evaluate eligibility for radioiodine therapy
  - Select nodules to biopsy in multinodular goiter even if TSH not low
  - Substernal goiter with compressive symptoms (e.g., dyspnea, stridor, cough, dysphonia, dysphagia)
  - Non-diagnostic or indeterminate FNA of thyroid nodule (e.g., follicular lesion of undetermined significance), to see if hot (functioning) nodule that may be benign vs cold nodule

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- **CT Neck with contrast (CPT® 70491), or CT Neck without contrast (CPT® 70490), or MRI Orbit/Face/Neck without contrast (CPT® 70540), or MRI Orbit/Face/Neck without and with contrast (CPT® 70543):**

  Imaging should be done only after initial thyroid ultrasound has been performed.

  MRI and CT are not medically necessary for routine thyroid nodule evaluation and should only be considered after US for:
  - evaluation of extent of known substernal goiter.
  - suspected airway compression (i.e., subjective sense of dyspnea or choking sensation in the clinical history with known multinodular goiter).
  - presence of pathologic lymph nodes in cervical regions not visualized on ultrasound.
  - clinically suspected advanced thyroid disease, including invasive primary tumor.
  - any preoperative planning for thyroid disease.
- **CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260)** is also medically necessary for:
  - preoperative planning for individuals with substernal extension of the thyroid, pulmonary symptoms, or abnormalities on chest x-ray, and should be ordered by a surgeon or in consultation with a surgeon.
- **Thyroglossal duct cysts (TGDC)** are the most common type of congenital neck cyst of the midline neck, and may be first diagnosed in adulthood, though more commonly in early childhood.
  - A physical exam feature includes the rise and fall of the midline mass with protrusion and retraction of the tongue, due to its embryonal connection to the foramen cecum.
  - There is a small risk (about 1%) of incidental malignant degeneration within the TGDC, particularly within adults, and therefore, it is uniformly managed surgically. The Sistrunk procedure, which involves resection of the TGDC and its complete tract within the surrounding midline tissues—to include the middle third of the hyoid bone, is considered the gold standard in surgical management with a less than 5% risk of recurrence.
  - Advanced imaging, per surgeon's request—or a provider in consultation with the head and neck surgeon, to include Neck Ultrasound (CPT® 76536) **AND/OR** CT Neck with contrast (CPT® 70491) or MRI Neck with and without contrast (CPT® 70543), is medically necessary pre-operatively, or for the evaluation of a suspected recurrence.

Background and Supporting Information

- The American College of Radiology (ACR) Thyroid Imaging, Reporting, and Data System (TI-RADS), consisting of five levels, is utilized for recommendations in

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determining US follow-up vs FNA of thyroid nodule(s). TI-RADS levels are determined based on the ultrasound appearance of the nodule.

- The American Thyroid Association (ATA) guidelines from 2015 also use imaging characteristics and size for thyroid nodule risk stratification; however, size cutoffs are slightly more generous when compared to ACR-TIRADs. Sonographic imaging and/or biopsy requests in accordance with ATA criteria are appropriate.
- Fine-Needle Aspiration (FNA) biopsy is medically necessary for suspicious and/or large thyroid nodules prior to CT or MRI imaging.
- Ultrasound is not used to screen: 1) the general population, 2) individuals with normal thyroid on palpation with a low risk of thyroid cancer, 3) individuals with hyperthyroidism, 4) individuals with hypothyroidism or 5) individuals with thyroiditis. Conversely, US is medically necessary in individuals who have no symptoms but are high-risk as a result of: history of head and neck irradiation, total body irradiation for bone marrow transplant, exposure to fallout from radiation during childhood or adolescence, as well as family history of thyroid cancer syndromes such as MEN-2, medullary or papillary thyroid cancer, Cowden's disease, familial adenomatous polyposis, Carney complex, and Werner syndrome/progeria.
- There is insufficient evidence supporting the use of PET to distinguish indeterminate thyroid nodules that are benign from those that are malignant.
- 18FDG-PET imaging is not medically necessary for the evaluation of thyroid nodules with indeterminate cytology. Routine preoperative 18FDG-PET scanning is not medically necessary.
- Elastography provides information about nodule stiffness that is complementary to gray scale ultrasound findings in nodules with indeterminate cytology or ultrasound findings. It should not be used as a substitute for gray scale ultrasound.
- Use of ultrasound contrast medium is not medically necessary for the diagnostic evaluation of thyroid nodules and its current use is restricted to definition of size and limits of necrotic zones after minimally invasive nodule ablation techniques.
- For prior history of neck radiation and family history of medullary carcinoma, MEN-2, or papillary thyroid carcinoma, US Neck can be ordered as a baseline test. With an indeterminate or abnormal initial US, follow-up studies should be directed by the relevant Neck or Oncology guideline criteria.

Evidence Discussion

- Ultrasound (US) plays a key role in determining which nodules display characteristics suspicious for malignancy and warrant biopsy. Of palpable thyroid nodules, the American College of Radiology (ACR) states, "US provides high-resolution imaging to show that the palpable abnormality is within the thyroid and is the best study to characterize the nodule for the risk of malignancy".[5] US is also the best study to demonstrate goiter size and evaluate thyroid morphology.[5] US has a sensitivity of 88%, a specificity of 86%, and a diagnostic accuracy of 92%.[14]

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- Nuclear imaging with a radionuclide uptake and scan also plays an important role in thyroid imaging. In the setting of a suppressed thyroid stimulating hormone (TSH) level, nuclear imaging helps to determine whether the individual has thyrotoxicosis.[5]
- A thyroid nuclear scan, in conjunction with thyroid ultrasound, is also useful in the setting of a multinodular goiter and a normal TSH to identify hypofunctioning or isofunctioning nodules which should be targeted for biopsy.[5]
- Neck CT is useful in determining the extent of a substernal goiter, evaluating whether a goiter is causing tracheal compression or deviation, assessing the extent of invasive thyroid cancer, or for preoperative imaging. It does not have a role in distinguishing benign from malignant thyroid nodules.[5]
- Neck CT is preferred to neck MRI, as there is less respiratory motion artifact.[5]
- FDG-PET/CT does not have a role in the initial imaging of a thyroid nodule.[5]

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Hyperthyroidism and Hypothyroidism (Neck-8.2)

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- Hyperthyroidism suspected
  - Thyroid Uptake Study (CPT® 78012 or CPT® 78014) if ONE of the following:
    - TSH below normal range and elevated free T4 and/or free T3, OR
    - Subclinical hyperthyroidism with TSH <0.1 mU/L and normal free T4 and free T3, OR
    - Subclinical hyperthyroidism with TSH below the lower limit of normal but ≥0.1 and normal free T4 and free T3 in the setting of any of the following:
      - Age ≥65
      - Symptoms of hyperthyroidism
      - Presence of cardiac disease or osteoporosis
  - Ultrasound (US) Neck (CPT® 76536) if any of the following:
    - Palpable nodule on examination
    - Nuclear scanning is suggestive of thyroid nodular disease
    - Diagnostic uncertainty regarding the etiology of hyperthyroidism based on clinical presentation and initial biochemical evaluation
    - To evaluate thyroid dimensions for planning RAI treatment
    - Nuclear scanning is contraindicated (i.e., pregnancy, breastfeeding etc)
- Hyperthyroidism on therapy---For individuals with thyroid hormone levels (TSH, free T4 and free T3) within the normal, hypothyroid, or hyperthyroid range while receiving treatment with an anti-thyroid medication (methimazole or propylthiouracil/PTU):
  - Nuclear Scan (CPT® 78013 or CPT® 78014) if ONE of the following:
    - To determine the cause of hyperthyroidism if there was no diagnostic scan prior to the start of medical therapy
    - To characterize the uptake in a thyroid nodule(s) to properly triage the nodule for FNA if there was no diagnostic scan prior to the start of medical therapy
  - Thyroid Uptake Study (CPT® 78012 or CPT® 78014) if:
    - plan is for radioactive iodine therapy as definitive hyperthyroidism treatment.
- Hypothyroidism: There is no role for thyroid imaging in the workup of hypothyroidism in adults. Imaging for thyroid morphology does not help differentiate among causes of hypothyroidism, and all causes of hypothyroidism will have decreased radioiodine uptake.

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Evidence Discussion

- The etiology of thyrotoxicosis is not always apparent in clinical presentation and therefore the use of diagnostic imaging studies is medically necessary to determine the etiology of hyperthyroidism.
- Thyroid nuclear scans play a central role in thyroid disease evaluation. They provide a planar image of the thyroid gland using a gamma camera to assess potential variability in the concentration of the radioisotope within thyroid tissue and can be combined with measurements of uptake of specific tracers and provide very useful information regarding thyroid pathology and function.[5]
- Imaging with a radioiodine uptake and scan provides a valuable benefit as it can help confirm the cause for thyrotoxicosis and helps to decide the most appropriate treatment, which may vary depending on cause of hyperthyroidism.[5]
- Imaging with a thyroid nuclear scan also has a role in planning therapy with radioactive iodine.
- Radioiodine uptake and scans can distinguish between high-uptake causes of thyrotoxicosis, such as Graves disease, toxic adenoma, and toxic multinodular goiter, and low-uptake causes, such as subacute thyroiditis.[5]
- The scan component is helpful in differentiating between the high-uptake causes that show a focal uptake pattern, such as toxic adenoma or toxic multinodular goiter, and the high uptake causes that show a diffuse uptake pattern such as Graves disease.[5]
- If radioiodine therapy is planned, the uptake component of the scan can then help determine the dose.[5]
- A neck ultrasound is the best imaging study to evaluate thyroid morphology and can be a helpful adjunct study to a radioiodine uptake. Although a Doppler ultrasound may help to distinguish the cause of hyperthyroidism, a radionuclide uptake study is still preferred because it directly measures thyroid activity rather than inferring it based on blood flow.[5]
- When a radioactive uptake scan shows nodules from toxic multinodular goiter or toxic adenoma, a neck ultrasound can confirm presence of nodules and also evaluate for suspicious features of malignancy.[5]
- Though radioactivity exposure exists with the use of radioactive scans, the studies are an invaluable resource when helping to establish the cause of hyperthyroidism and guide treatment; guidelines as to when it is appropriate to order should be followed to avoid unnecessary radiation exposure.[5]
- There is no role for imaging in the workup of hypothyroidism in adults. Imaging for thyroid morphology does not help differentiate among causes of hypothyroidism, and all causes of hypothyroidism will have decreased radioiodine uptake.[5]

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Thyroid Health Equity Considerations

- Racial and ethnic minorities with thyroid disease have worse health outcomes for both benign and malignant disease.
- Socioeconomic status, insurance status, and differential access to healthcare all contribute to disparities in health outcomes.

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Parathyroid Imaging (Neck-8.3)

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- Classic primary hyperparathyroidism
  - Parathyroid Planar Imaging (CPT® 78070), Parathyroid Planar Imaging with SPECT (CPT® 78071), or Parathyroid Planar Imaging with SPECT/CT (preferred study) (CPT® 78072) AND/OR Ultrasound (CPT® 76536) AND/OR 4D CT Neck without and with contrast (CPT® 70492) are medically necessary if BOTH of the following conditions are met:
    - PTH and Calcium levels are elevated (See **Background and Supporting Information**)
    - Intention of the study is preoperative localization
      - All parathyroid nuclear scan codes (CPT® 78070, CPT® 78071, CPT® 78072) include thyroid subtraction when performed and no additional thyroid nuclear scan CPT codes are required unless otherwise indicated in **Thyroid Nodule (Neck-8.1)** or **Hyperthyroidism and Hypothyroidism (Neck 8.2)**.
      - Reporting or billing CPT® 78800 for the purpose of intraoperative parathyroid localization using a gamma probe is not medically necessary if performed along with a parathyroid nuclear scan (CPT® 78070, CPT® 78071, CPT® 78072).
      - Ultrasound (CPT® 76536) can be ordered independently to evaluate the thyroid per criteria in **Thyroid Nodule (Neck-8.1)** or **Hyperthyroidism and Hypothyroidism (Neck 8.2)**.
      - 3D Imaging (CPT® 76376 or CPT® 76377) is medically necessary with a 4D CT Neck.
  - MRI Neck without and with contrast (CPT® 70543) for cases of re-operation, difficult localization or ionizing radiation contraindication as ordered by an Endocrinologist, Parathyroid surgeon, or Radiologist or any provider in consultation with one of these specialists.
  - CT Chest with contrast (CPT® 71260) in rare circumstances in the evaluation of ectopic mediastinal parathyroid adenomas as ordered by an Endocrinologist, Parathyroid surgeon, or Radiologist or any provider in consultation with one of these specialists.
  - Choline PET/CT (CPT® 78815 or CPT® 78816) is considered experimental and investigational for preoperative localization in cases of primary hyperparathyroidism.
  - Repeat imaging is medically necessary both in individuals with prior non-localizing imaging who have not yet undergone parathyroid exploration OR in cases of hyperparathyroidism that recurs or persists after parathyroid surgery if reimaging is

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being ordered by a surgeon or any provider after consultation with a surgeon with expertise in parathyroidectomy.

- Primary hyperparathyroidism variants
  - Primary hyperparathyroidism with non-elevated serum calcium (serum calcium level normal and PTH elevated)
    - Confirmatory study is elevated ionized calcium, elevated albumin corrected calcium or elevated historic calcium levels.
  - Hypercalcemia with inappropriately non-suppressed PTH (calcium level elevated and PTH normal)
    - PTH level ≥25 pg/mL is consistent with primary hyperparathyroidism.
    - See **Background and Supporting Information** for more information.
  - Intention of parathyroid imaging should be for pre-operative localization.
  - Use the same guidance on imaging modalities as described in "classic" primary hyperparathyroidism.

Primary Hyperparathyroidism variants:

	Calcium	PTH	Confirms/strongly suggests primary hyperparathyroidism
Classic primary hyperparathyroidism	High	High	Yes
Primary hyperparathyroidism with non-elevated serum calcium	Normal	High	Elevated ionized albumin corrected or historic calcium levels*
Hypercalcemia with inappropriately non-suppressed PTH	High	Normal	PTH ≥25 pg/ml

- Normocalcemic hyperparathyroidism
  - Serum calcium levels (including ionized calcium levels) are always normal and PTH levels are elevated.
  - Secondary causes of PTH elevation are excluded. See **Background and Supporting Information** for differential diagnosis of secondary hyperparathyroidism.
  - Calcium, PTH and clinical status should be monitored annually.
    - In the event of laboratory progression to hypercalcemia, refer to "classic" primary hyperparathyroidism for imaging guidance.

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    - In the event of clinical progression (decline in bone mineral density or new fracture/renal stone/nephrocalcinosis), imaging for the intent of preoperative localization is as requested by or after consultation with a specialist or any provider in consultation with a specialist.
- Secondary renal hyperparathyroidism
  - Serum calcium levels are low or normal (but may also be elevated in more advanced disease) and PTH levels are very elevated.
  - Imaging for the intent of preoperative localization as requested by or after consultation with a specialist if all of the following are met:
    - Individual has stage 3a-stage 5 chronic kidney disease (GFR<60).
    - PTH level is >9x upper limit of normal reference range for the lab testing facility (~585 pg/mL) despite standard medical or pharmacologic therapy (calcimimetics, calcitriol and/or vitamin D analogs).
- Tertiary hyperparathyroidism
  - Serum calcium and PTH levels are elevated as a result of long standing secondary hyperparathyroidism in individuals on renal replacement therapy or after renal transplant.
  - Imaging for the intent of preoperative localization as requested by or in consultation with a specialist.

Hyperparathyroidism subtypes:

	Calcium	PTH	Clinical Hallmarks
Normocalcemic Hyperparathyroidism	Normal	High	Calcium never elevated
Secondary Renal Hyperparathyroidism	Low/Normal/High	Very High	Stage 3a-5 CKD, PTH >9x ULN
Tertiary Hyperparathyroidism	High	High	ESRD/renal transplant

Background and Supporting Information

- Hypercalcemia in individuals with primary hyperparathyroidism may be determined by elevated serum calcium, elevated serum ionized calcium, elevated serum calcium level corrected for albumin, or historic calcium elevation. A comparison of serial measurements of calcium is helpful in determining the presence of true hypercalcemia as calcium levels may be variable over time.
- Parathyroidectomy candidacy should be determined by the provider; however, national guidelines recognize the following criteria for surgery:

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  - All individuals <50 years of age, regardless of whether objective features are present or absent
  - All symptomatic individuals, including those with kidney stones, hypercalcemic crises, pathologic fractures or other associated symptoms
  - Individuals with findings concerning for parathyroid cancer (very high calcium >13)
  - All asymptomatic individuals with the following:
    - Serum calcium >1.0 mg/dl (0.25 mmol/l) above the normal range
    - BMD by DEXA: T-score ≤2.5 at the lumbar spine, total hip femoral neck or distal 1/3 radius (The forearm- i.e., distal 1/3 radius is preferentially impacted by primary hyperparathyroidism as this area is rich in cortical bone.)
    - Vertebral fracture by x-ray, CT, MRI and vertebral fracture assessment
    - Estimated glomerular filtration rate of less than 60 ml/min
    - Urinary calcium excretion >400 mg in 24 hours
    - Nephrolithiasis or nephrocalcinosis by x-ray, ultrasound or CT
  - Asymptomatic individuals who cannot participate in appropriate medical surveillance
  - Asymptomatic individuals desiring definitive surgical management
- For cases of “normocalcemic hyperparathyroidism” in which primary hyperparathyroidism is not confirmed, additional investigation for secondary causes of hyperparathyroidism (renal insufficiency, hypercalciuria as a primary renal abnormality, vitamin D deficiency and gastrointestinal malabsorption problems such as short gut syndrome, celiac disease, Crohn's disease or a prior Roux-en-Y bypass surgery) is medically necessary.
- For cases of hypercalcemia in which primary hyperparathyroidism is not confirmed, additional consideration for other causes of hypercalcemia (malignancy including PTH-RP mediated and myeloma, granulomatous disease, FHH, medications including thiazide diuretics, excessive calcium/vitamin D supplementation and the history of or present lithium use) is medically necessary.

Evidence Discussion

- The purpose of parathyroid imaging is to aid in localizing hyperfunctioning parathyroid gland(s) for the purpose of curative surgery. Imaging has no role in the diagnosis of hyperparathyroidism.[1]
- There may be a need for more than one modality in the localization of a parathyroid adenoma as studies have shown no one modality is superior over others.[1]
- The sensitivity and positive predictive value of imaging modalities will vary in different situations, such as whether the individual has a single parathyroid adenoma or if multiple parathyroid glands are involved, if the imaging is for an initial surgical intervention versus a re-operation, and whether the individual has primary, secondary, or tertiary hyperparathyroidism.[1]

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Parathyroid Incidentaloma (Neck-8.4)

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- A mass incidentally found on neck imaging that may represent an enlarged parathyroid gland should prompt laboratory testing including calcium and PTH levels.
  - If laboratory abnormalities suggest hyperparathyroidism, i.e., "functioning parathyroid incidentaloma", see **Hyperparathyroidism (NECK- 8.3)** for imaging recommendations.
  - If there are no laboratory abnormalities and diagnoses other than parathyroid incidentaloma are suspected, see **Neck Mass/Swelling/Adenopathy (NECK- 5.1)** for imaging recommendations.
  - Parathyroid nuclear scans are commonly requested for an evaluation of a parathyroid incidentaloma; however, the sensitivity of these scans are low in individuals with normal calcium/PTH and no clinical symptoms of primary hyperparathyroidism. Reliance on either a positive scan or negative scan to decide if surgery is medically necessary is not supported by current literature.
- If a parathyroid incidentaloma is suspected on imaging prior to planned thyroid surgery or other head/neck surgery, the following studies are medically necessary if ordered by the surgical team or any provider in consultation with the surgical team:
  - Parathyroid Planar Imaging (CPT® 78070), Parathyroid Planar Imaging with SPECT (CPT® 78071), or Parathyroid Planar Imaging with SPECT/CT (CPT® 78072) AND/OR Ultrasound (CPT® 76536) AND/OR 4D CT Neck without and with contrast (CPT® 70492)
- Ultrasound (US) Neck (CPT® 76536) is medically necessary annually if the mass was not removed surgically.

Background and Supporting Information

- "Parathyroid incidentalomas" include parathyroid adenomas found unexpectedly at the time of surgery or seen on ultrasound.
- Normal sized parathyroid glands (~6mm) are not usually identified by most imaging modalities, so enlargement warrants laboratory evaluation to rule out pathologic causes such as primary hyperparathyroidism or rarely parathyroid carcinoma.
- Sonographic imaging features of a parathyroid incidentaloma (ovoid, hypoechoic, well circumscribed and adjacent to but separate from the thyroid either posteriorly or inferiorly) may have overlap with perithyroidal lymph nodes and exophytic thyroid nodules in a multinodular goiter.
- The literature does report cases of pathologically confirmed parathyroid adenomas/ hyperplasia in individuals with normal serum calcium and PTH levels, so these enlarged parathyroid glands may represent an early stage of hyperparathyroidism.

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It is unclear what percentage of non-functioning PTIs become hyper-secreting over time, but many of these masses are surgically managed.
- Normally sized and normally functioning parathyroid glands do not take up sestamibi or tetrofosmin. The likelihood of a positive parathyroid nuclear scan is low in the setting of normal calcium and PTH levels.
- Parathyroid fine needle aspiration biopsy has been used historically; however, its diagnostic use is limited due to the potential for hemorrhage and fibrosis which make eventual surgical dissection and pathologic interpretation more difficult.

Evidence Discussion
- With the advent of high resolution CT scans, ultrasounds, and other imaging modalities, along with their widespread use as diagnostic modalities, parathyroid lesions are increasingly being incidentally found on these imaging studies.[1]
- The distinct features of parathyroid lesions are readily seen on ultrasound imaging.[1]
- If lab work does not indicate hyperfunctioning of the parathyroid gland, then these lesions can be monitored annually with ultrasound, as parathyroid nuclear scans have low sensitivity for detection in individuals with normal parathyroid function.[1]
- Parathyroid nuclear scans, 4D CT of the Neck, and Neck ultrasound studies can be used in evaluation of parathyroid incidentalomas found prior to a pending neck surgery to aid in preoperative evaluation, as determined by the surgical team.[1]

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Parathyroid

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Imaging of the Larynx, Trachea, and Bronchus (Neck-9)

Guideline

Imaging of the Larynx, Trachea, and Bronchus (Neck-9.1)
References (Neck-9)

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Imaging of the Larynx, Trachea, and Bronchus (Neck-9.1)

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- Initial evaluation for suspected laryngotracheal pathology:
  - Direct visualization of the upper airway (via laryngoscopy, with or without bronchoscopy), and can also include
  - Plain x-rays of the neck with or without chest x-ray
- To further evaluate definite abnormalities found on either of the above, including laryngotracheal, tracheal, or bronchial anomalies, foreign bodies or persistent segmental or lobar lung collapse:
  - CT Neck with contrast (CPT® 70491) **OR** CT Neck without contrast (CPT® 70490) **AND/OR**
  - CT Chest with contrast (CPT® 71260) **OR** CT Chest without contrast (CPT® 71250), depending on the anatomic level of the lesion.
    - MRI Orbit/Face/Neck without contrast (CPT® 70540) OR MRI Neck without and with contrast (CPT® 70543) is medically necessary as a supplement to CT Neck or CT Chest if additional assessment is needed.
    - See **Squamous Cell Carcinomas of the Head and Neck—Suspected/ Diagnosis (ONC-3.1)** for suspected laryngotracheal tumor.
- For suspected subglottic stenosis (SGS) after evaluation by a specialist or in consultation with a specialist who has directly visualized the upper airway:
  - CT Neck with contrast (CPT® 70491) **OR** CT Neck without contrast (CPT® 70490) is medically necessary.
- For obstructive physiology in the setting of tracheomalacia:
  - Expiratory HRCT (CPT® 71250) is medically necessary.

Background and Supporting Information

- Bronchoscopy can further evaluate the distal endobronchial tree.
- Suspected laryngotracheal disease can be identified by inspiratory or biphasic stridor and a characteristic flow-volume loop of PFTs.
- The visualization of tracheal or bronchial "inspissation" or thickening of secretions without an abnormality, is not a risk for malignancy.
- CT with multiplanar reformatting has proven comparable to rigid bronchoscopy with a 100% sensitivity and specificity of detecting SGS and for measuring length and grade of stenosis.

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Evidence Discussion

- Radiographs by means of neck and chest radiographs are sensitive for only those radioopaque foreign bodies but have value in providing information regarding other chest pathology such as presence of consolidation, atelectasis and bronchiectasis.[6]
- CT is more sensitive than x ray in identifying airway foreign bodies due to its higher resolution.[7]
- CT scan can rapidly and accurately identify laryngo-tracheo-bronchial abnormalities prior to bronchoscopy such as pneumonia, airway stenosis, atelectasis, effusion and consolidation.[7-9]
- CT provides a sensitivity of more than 96% and a specificity of more than 97% in comparison to bronchoscopy for detecting tracheomalacia. CT provides the advantage of simultaneously evaluating mediastinal, vascular and lung pathologies.[10]

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Neck Pain/Neck Trauma (Neck-10)

Guideline

Neck Pain (Cervical) (Neck-10.1)
Torticollis and Dystonia (Neck-10.2)
Eagle Syndrome (Neck-10.3)
Neck Trauma (Neck-10.4)
References (Neck-10)

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Neck Pain (Cervical) (Neck-10.1)

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- Neck pain is usually related to a specific process including pharyngitis, radiculopathy, adenopathy, mass, carotid dissection and torticollis, and therefore found elsewhere in these guidelines.
- For the evaluation of neck pain or other symptoms which may involve the cervical spine, including myelopathy and cervical radiculopathy, see Spine Imaging Guidelines .

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Torticollis and Dystonia (Neck-10.2)

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Older Child (beyond infancy) or Adult

- To identify fracture or malalignment in cases of trauma:
  - Initial evaluation of trauma (without a high-risk mechanism of injury–see **SP-3.2 Neck (Cervical Spine) Trauma**) is by plain radiographs of the cervical spine. If inconclusive:
    - CT Neck with contrast (CPT® 70491) **AND/OR**
    - CT Cervical Spine without contrast (CPT® 72125)
- In the clinical setting of cervical spine trauma with an associated neurologic deficit:
  - MRI Cervical Spine without contrast (CPT® 72141) is medically necessary.
- To evaluate for soft tissue or neurological cause in cases with no trauma history:
  - CT Neck with contrast (CPT® 70491), **AND/OR**
  - MRI Cervical Spine without contrast (CPT® 72141), **OR**
  - CT Cervical Spine without contrast (CPT® 72125)
    - Positive→ Further advanced imaging is not medically necessary if CT Neck or CT/MRI Cervical Spine has identified local cause.
    - Negative→ MRI Brain without and with contrast (CPT® 70553) to exclude CNS cause.

Evidence Discussion

- Plain radiography continues to be the primary imaging modality for the initial diagnosis of neck concerns in cases of trauma without high-risk mechanism of injury.[9,10]
- CT offers superior depiction of cortical bone and is more sensitive than radiographs in assessing facet degenerative disease, osteophyte formation, and other osseous structures.[9,10]
- Individuals with abnormal neurological findings benefit from MRI to help identify spinal cord abnormalities and soft tissue causes in the workup of dystonia and/or torticollis.[9,10]
- Risks associated with imaging include, but are not limited to, radiation exposure and contrast complications.[9,10]

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Eagle Syndrome (Neck-10.3)

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Also known as "calcified stylohyoid ligament", "elongation of styloid process", or "stylocarotid artery syndrome".

- **"Classic Eagle Syndrome"**
  - Typically seen in individuals after pharyngeal trauma or tonsillectomy
  - Characterized by ipsilateral dull, persistent pharyngeal pain, centered in the ipsilateral tonsillar fossa, that can be referred to the ear, and exacerbated by rotation of the head
  - Other symptoms may include dysphagia, sensation of foreign body in the throat, tinnitus, or cervicofacial pain.
  - If Eagle Syndrome is suspected on exam and/or lateral neck x-ray:
    - CT Maxillofacial with contrast (CPT® 70487) **OR** CT Maxillofacial without contrast (CPT® 70486) **AND/OR**
    - CT Neck with contrast (CPT® 70491) **OR** CT Neck without contrast (CPT® 70490)
- **"Stylocarotid Artery Syndrome"** (i.e., the anterior circulation equivalent of "bow hunter syndrome"—rotational vertebral artery occlusion syndrome)
  - It is characterized by the compression of the internal or external carotid artery (with their peri-vascular sympathetic fibers) by a laterally or medially deviated styloid process.
  - It is related to a pain along the distribution of the artery, which is provoked and exacerbated by rotation and compression of the neck.
  - It is not correlated with tonsillectomy.
  - If Stylocarotid Artery Syndrome is suspected on exam and/or lateral neck x-ray:
    - CT Maxillofacial with contrast (CPT® 70487) **AND/OR**
    - CT Neck with contrast (CPT® 70491)
    - Dynamic/Positional CTA (CPT® 70498) is also medically necessary to assess for concerns of vascular compression (see also **General Guidelines - CT and MR Angiography (CTA and MRA) (HD-1.5)** and **General Guidelines - Other Imaging Situations (HD-1.7)**
    - In cases of impingement of the internal carotid artery, there may be referred supraorbital pain and parietal headache. In cases of external carotid artery irritation, the pain radiates to the infraorbital region.
- CT scanning (and in particular, 3-D CT scanning) represents an extremely valuable imaging tool in individuals with Eagle syndrome. 3-D rendering (CPT® 76376 or CPT®

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76377) is medically necessary as an add on to CT Neck (CPT® 70491 **or** CPT® 70490), if requested, for accurate evaluation of the styloid process in relation to its anatomic relationship with the other head and neck structures, in surgical planning. See **3D Rendering (Preface-4.1)**.

Background and Supporting Information

- Torticollis or cervical dystonia is an abnormal twisting of the neck resulting in head rotation. Its causes are many and may be congenital or acquired and caused by trauma, infection/inflammation, neoplasm and/or idiopathic. It occurs more frequently in children and on the right side (75%).
- Eagle syndrome is characterized by recurrent pain in the oropharynx and face due to an elongated styloid process or calcified stylohyoid ligament. The styloid process is a slender outgrowth at the base of the temporal bone, immediately posterior to the mastoid apex.

Evidence Discussion

- If Eagle syndrome is suspected on exam and/or lateral neck x-ray, CT Maxillofacial or CT Neck are medically necessary.[4,7]
- CT is the preferred modality for evaluation of bony structures and detection of abnormalities associated with calcifications.[13]
- Indications for cervicocerebral computed tomography angiography (CTA) or magnetic resonance angiography (MRA) of the head and neck vessels or include the diagnosis, characterization and/or surveillance of a variety of vascular conditions, including vascular compression of the internal or external carotid artery by an elongated styloid process (Eagle Syndrome).[6]
- Risks of CTA include exposure to ionizing radiation; thus, magnetic resonance angiography (MRA) is available as an alternative to reduce radiation exposure. In addition, MRA is an alternative for individuals with iodinated contrast allergies or other contraindications to iodinated contrast.[6,14]
- MRA, as an alternative modality, is noninvasive, and does not require iodinated contrast. Limitations include artifacts due to motion, slow or turbulent flow, susceptibility effects, and claustrophobia. Additionally, MRA may not be a feasible option for those with contraindications to MRI such as incompatible pacemakers, cochlear implants, neurostimulators or other devices. In these scenarios, CTA may be the appropriate alternative.[6,14,15]

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Neck Trauma (Neck-10.4)

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- CT Neck with contrast (CPT® 70491) OR CT Neck without contrast (CPT® 70490) is medically necessary in individuals with neck trauma who have an altered state of consciousness, distracting injuries or intoxication, midline tenderness to palpation, or neurological deficits.
- Penetrating neck trauma
  - CTA Neck (CPT® 70498) is medically necessary for evaluating vascular injury and/or aerodigestive injuries.
- Blunt neck trauma
  - For any evidence of blunt neck trauma, including but not limited to: neurologic deficit, infarction on CT Head, cervical seatbelt sign, and fractures of the skull, face, or spine, a CTA Neck (CPT® 70498) is medically necessary.
  - If CTA is negative but high clinical suspicion remains for esophageal injury, MRA Neck with contrast (CPT® 70548) OR MRA Neck without contrast (CPT® 70547) OR MRA Neck without and with contrast (CPT® 70549) is medically necessary.
- Laryngeal neck trauma
  - For any clinical suspicion of injury, CT Neck without contrast (CPT® 70490) is medically necessary.

Evidence Discussion

- CTA has high sensitivity and specificity for stable individuals with penetrating neck trauma.[18]
- CTA is the primary imaging modality to assess aerodigestive injuries from penetrating neck trauma. However, a high number of esophageal injuries are missed on CTA, so a negative CTA with suspicion for esophageal injury warrants additional testing.[16,19]
- MRA Neck can be useful to detect dissections and intramural hematoma from blunt neck trauma.[20]
- CT Neck without contrast is useful to assess laryngeal neck trauma, as it can assess the cartilaginous and bony components, as well as determine any fractures that may need stabilization.[21]

Background and Supporting Information

- For penetrating neck trauma, if diagnostic uncertainty remains after CTA Neck, follow-up tests may include ultrasound, esophagram, angiography, and triple endoscopy.

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Salivary Gland Disorders (Neck-11)

Guideline

Salivary Gland Disorders (Neck-11.1)
References (Neck-11)

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Salivary Gland Disorders (Neck-11.1)

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- **Xerostomia (Dry Mouth)**
  - Salivary Gland Nuclear Imaging (one of CPT® 78230, CPT® 78231, or CPT® 78232) is medically necessary for any one of the following:
    - Dry mouth and either:
      - Sjögren's syndrome
      - Sialadenitis
      - History of head or neck radiation therapy
      - History of cerebral palsy
      - Parotid mass to allow preoperative diagnosis of Warthin's tumor
- **Salivary Gland Stones, Sialadenitis or Stenosis**:
  - Sialography (contrast dye injection) under fluoroscopy is medically necessary to rule out a salivary duct stone or stricture, using
    - Post-sialography CT (CT Maxillofacial without contrast [CPT® 70486] for Stensen's duct of the parotid gland, which would be most common; **or** CT Neck without contrast [CPT® 70490] for the level of the Wharton's Duct); **or** post-sialography MRI (MRI Orbit/Face/Neck without contrast [CPT® 70540]) **OR**
  - CT Maxillofacial area with contrast (CPT® 70487) **OR**
  - CT Neck with contrast (CPT® 70491)
    - CT performed only without IV contrast (CPT® 70490) may be helpful in a small minority of cases, such as cases of follow-up for known salivary stones, or for post-sialography imaging, as described above **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543)
- **Parotid or Other Salivary Gland Mass**
  - The following are appropriate:
    - Ultrasound (CPT® 76536) is medically necessary as initial or additional imaging and does not need to be completed prior to the performance of advanced imaging.
    - MRI Orbit/Face/Neck without and with contrast preferred (CPT® 70543) **or** MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR**
    - CT Neck with contrast (CPT® 70491) preferred **or** CT Maxillofacial area with contrast (CPT® 70487)
- **Repeat Imaging (CT or MRI, as above)**
  - There is currently no standard timeframe for repeat advanced imaging to follow known benign pathology of the salivary gland that has been resected—partially

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or completely, or only observed. This holds true even if the salivary lesion has the potential for recurrence or malignant transformation (i.e., pleomorphic adenoma).
  - Repeat advanced imaging, as requested by the surgeon or those in consultation with the surgical team, is medically necessary if relevant history and exam demonstrate signs:
    - concerning for complications of surgery, or
    - recurrence or progression of neoplasm/lesion

Evidence Discussion

- Current history and comprehensive head & neck exam are required prior to any advanced imaging for suspected salivary gland pathology.[1]
- CT should be performed with IV contrast to distinguish vessels from lymph nodes and to confirm if a mass is hypervascular. *Dual-phase CT imaging (without and with IV contrast) is not medically necessary*.[2]
  - CT performed only without IV contrast may be helpful in a small minority of cases including cases of follow-up for known salivary stones or post-sialography studies.
- A recent study in the American Journal of Neuroradiology comparing contrast enhanced and non-contrast enhanced CT in the evaluation of sialolithiasis demonstrated excellent sensitivity and specificity with no false-positive results using contrast-enhanced CT alone (without the addition of non-contrasted images for comparison).[3] The sensitivity of contrast-enhanced CT is 96-98%, and the specificity is 88-100%.[7] Benefits of initial only contrast-enhanced CT include better evaluation of the ductal system, improved soft tissue contrast in assessing salivary masses and decreased radiation dose (compared to dual phase CT imaging (without and with IV contrast)).[3]
- The preferred modality to evaluate suspected parotid lesions is MRI of the face and/or neck with and without IV contrast. It can provide comprehensive information about the full extent of the mass (i.e., deep lobe involvement) and other local invasion (such as perineural tumor spread and possible extension into the temporal bone).[4]

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Sore Throat, Odynophagia, and Hoarseness (Neck-12)

Guideline

Definitions (Neck-12.0)
Sore Throat/Throat Pain/Odynophagia (Neck-12.1)
Hoarseness (Neck-12.2)
References (Neck-12)

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Definitions (Neck-12.0)

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- Hoarseness – Altered voice quality reported by the individual
- Dysphagia – Disordered or impaired swallowing i.e., food impactions, globus sensation, choking/aspiration, regurgitation (see **Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)**)
- Odynophagia – Painful swallowing

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Sore Throat/Throat Pain/Odynophagia (Neck-12.1)

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- See **Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)** for dysphagia as the primary symptom.
- Sore Throat/Throat Pain/Odynophagia
  - Uncomplicated viral or streptococcal pharyngitis with sore throat
    - Imaging studies are not medically necessary. See **Neck Mass/Swelling/ Adenopathy (Neck-5.1)** for suspected complications of pharyngitis/tonsillitis, such as a cervical space abscess.
  - Postoperative throat pain or odynophagia after head and neck procedure with suspected complication of procedure:
    - CT Neck with contrast (CPT® 70491)
  - Sore throat/throat pain/odynophagia that is persistent or progressive for two or more weeks, in spite of any treatment measures or observation:
    - Initial evaluation is laryngoscopy
      - If the initial laryngoscopy is abnormal, or if it is negative, and if there is a continued concern for an occult lesion within the pharynx due to any red flag symptoms (weight loss, referred otalgia, hoarseness, hemoptysis, and/or unilateral presentation of symptoms):
        - CT Neck with contrast (CPT® 70491) **OR**
        - MRI Orbit/Face/Neck without and with contrast (CPT® 70543)
      - Alarm symptoms of persistent unilateral throat pain or odynophagia with ipsilateral referred otalgia is especially suspicious for a submucosal tumor of the head and neck (versus more distal esophageal pathology).
      - If subjective dysphagia AND odynophagia are both present and the initial laryngoscopy and neck exam are normal (i.e., no cervical space abscess or post-surgical complication is suspected), and no red flag symptoms are present, then barium esophagram (or GI upper endoscopy) is medically necessary prior to the advanced imaging studies of the neck listed above.

Evidence Discussion

- Both CT and MRI can be used to delineate masses and provide cross sectional visualization of lesions that are deep to the mucosal surface. CT has the advantage to being readily available, requiring less time and less expense when compared to MRI.[7,8]

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Hoarseness (Neck-12.2)

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- Laryngoscopy is the primary diagnostic modality for evaluating individuals with hoarseness. Imaging studies, including CT and MRI, are not medically necessary in most individuals with hoarseness because most hoarseness is self-limited or caused by pathology that can be identified by laryngoscopy alone.
- The need for advanced imaging is based upon abnormal findings upon laryngoscopy, such as:
  - Immobile or partially mobile vocal cord [see **Recurrent Laryngeal Nerve Palsy (NECK-7.1)**]
  - Any growth, asymmetry, ulceration, or other suspected neoplasm of the glottis or supraglottis [see **Neck Mass/Swelling/Adenopathy (Neck 5.1)**; see also **ONC-3.0-3.4**].

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General Guidelines (ONC-1)

Guideline

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Abbreviations for Oncology Imaging Guidelines

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Abbreviations for Oncology Imaging Guidelines	
ACTH	adrenocorticotropic hormone
AFP	alpha-fetoprotein
ALKP	alkaline phosphatase
AP	anteroposterior
betaHCG	beta human chorionic gonadotropin
CA 125	cancer antigen 125 test
CA 19-9	cancer antigen 19-9
CA 15-3	cancer antigen 15-3
CA 27-29	cancer antigen 27-29
CBC	complete blood count
CEA	carcinoembryonic antigen
CNS	central nervous system
CR	complete response
CTA	computed tomography angiography
DCIS	ductal carcinoma in situ
DLBCL	diffuse large B cell lymphomas
DRE	digital rectal exam
EGD	esophagogastroduodenoscopy
ENT	ear, nose, throat
EOT	end of therapy
ERCP	endoscopic retrograde cholangiopancreatography
ESR	erythrocyte sedimentation rate

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Abbreviations for Oncology Imaging Guidelines	
EUA	exam under anesthesia
EUS	endoscopic ultrasound
FDG	fluorodeoxyglucose
FNA	fine needle aspiration
FUO	fever of unknown origin
GE	gastroesophageal
GI	gastrointestinal
GU	genitourinary
GTR	gross total resection
HG	high-grade
HIV	human immunodeficiency disease
HRPC	hormone refractory prostate cancer
hypermet	hypermetabolic
IFRT	involved field radiation therapy
inv	invasive
LAR	low anterior resection
LCIS	lobular carcinoma in situ
LDH	lactate dehydrogenase
LFT	liver function tests
LND	lymph node dissection
MALT	mucosa associated lymphoid tissue
maint	maintenance
MEN	multiple endocrine neoplasia
MG	myasthenia gravis
MGUS	monoclonal gammopathy of unknown significance
MIBG	I-123 metaiodobenzylguanidine scintigraphy
MRA	magnetic resonance angiography

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Abbreviations for Oncology Imaging Guidelines	
MRCP	magnetic resonance cholangiopancreatography
MRI	magnetic resonance imaging
MUGA	'multiple gated acquisition' cardiac nuclear scan
MWA	microwave ablation
NaF	sodium fluoride
NET	neuroendocrine tumor
NCCN®	National Comprehensive Cancer Network
NHL	non-Hodgkin's lymphoma
NPC	nasopharyngeal carcinoma
NSABP	National Surgical Adjuvant Breast and Bowel Project
NSAIDS	nonsteroidal anti-inflammatory drugs
NSCLC	non-small cell lung cancer
NSGCT	non-seminomatous germ cell tumor
PA	posteroanterior
PCI	prophylactic cranial irradiation
PET	positron emission tomography
COG	Children's Oncology Group
PSA	prostate specific antigen
RFA	radiofrequency ablation
RPLND	retroperitoneal lymph node dissection
SqCCa	squamous cell carcinoma
SCLC	small cell lung cancer
SIADH	syndrome of inappropriate secretion of antidiuretic hormone
TCC	transitional cell carcinoma
TLH	total laparoscopic hysterectomy
TNM	tumor node metastasis staging system
TSH	thyroid-stimulating hormone

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Abbreviations for Oncology Imaging Guidelines	
TURBT	trans-urethral resection of bladder tumor
VIPoma	vasoactive intestinal polypeptide
WLE	wide local incision
WB-MRI	whole body MRI
WM	Waldenstrom's macroglobulinemia
WBXRT	whole brain radiation therapy

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General Guidelines (ONC-1.0)

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- A relevant clinical evaluation or meaningful contact (telephone call, electronic mail or messaging) should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled off therapy surveillance evaluation or cancer screening. The clinical evaluation may include a relevant history and physical examination, including biopsy, appropriate laboratory studies, and results of non-advanced or advanced imaging modalities.
- Unless otherwise stated in the disease-specific guideline, a histological confirmation of malignancy (or recurrence) and the stage of disease is required to perform a medical necessity review of the requested imaging.
- Generally, the studies listed in the disease-specific sections reflect the studies supported by current literature and research for that condition. If a study is not listed, then it is not supported.
- Routine imaging of brain, spine, neck, chest, abdomen, pelvis, bones, or other body areas is not indicated except where explicitly stated in a diagnosis-specific guideline section, or if one of the following applies:
  - Known prior disease involving the requested body area
  - New or worsening symptoms or physical exam findings involving the requested body area (including non-specific findings such as ascites or pleural effusion)
  - New finding on basic imaging study such as plain x-ray or ultrasound
  - New finding on adjacent body area CT/MRI study (i.e., pleural effusion observed on CT Abdomen)
- Unless otherwise stated in the disease-specific guideline, advanced imaging of asymptomatic individuals is not routinely supported without signs or symptoms of systemic involvement of cancer.
- Repeat imaging studies are not generally necessary unless there is evidence of disease progression, recurrence of disease, and/or the repeat imaging will affect an individual's clinical management.
- Conventional imaging performed prior to diagnosis should not be repeated unless there is a delay of at least 6 weeks since previous imaging and treatment initiation or there are new or significantly worsening clinical signs or symptoms

Phases of Oncology Imaging and General Phase-Related Considerations

The following provides general clarity regarding the categories used to describe the phases of imaging throughout these guidelines.

- Screening

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  - Imaging requested for individuals at increased risk for a particular cancer in the absence of known clinical signs or symptoms
- Suspected diagnosis
  - Imaging requested to evaluate a suspicion of cancer, prior to histological confirmation
- Initial work-up and staging
  - Imaging requested after biopsy confirmation and prior to starting specific treatment
- Treatment response or interim restaging
  - Imaging performed during active treatment with chemotherapy, targeted therapy, immunotherapy, radiation therapy, or endocrine therapy. Can be curative or non-curative.
- Restaging of locally treated lesions
  - Imaging performed to evaluate response to primary or metastatic lesion(s) following lesion directed therapy (e.g. cryotherapy, ablation, radioembolization, chemoembolization, radiation therapy).
- Restaging/suspected recurrence
  - Imaging requested when there is suspicion for progression or recurrence of known cancer based on clinical signs or symptoms, laboratory tests, or basic imaging studies.
- Surveillance
  - Imaging performed in individuals who are asymptomatic or have chronic stable symptoms, have no clinical suspicion of change in disease status, and are not receiving active anti-tumor treatment or are receiving maintenance treatment.

The table below provides descriptions of treatment phases, their definitions, and general timeframes.

Treatment Phase	Phase Definition	Imaging Timeframe
After definitive local therapy of primary tumor	Treatment intended to be curative typically using a combination of surgery, radiation therapy, and chemotherapy	Follow surveillance guidelines
During/after neoadjuvant chemotherapy	Treatment given prior to definitive intended curative therapy (usually surgery and/or radiation therapy based), to shrink the cancer to make the definitive curative therapy more effective	Follow initial work-up/staging or restaging/recurrence guidelines

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Treatment Phase	Phase Definition	Imaging Timeframe
During adjuvant chemotherapy	Adjuvant is given after the primary cancer treatment (primary treatment is usually surgery or radiation) with the goal of destroying any remaining cancer cells that cannot be visualized but may still be present, to reduce the risk of recurrence and improve cure/ survival rates beyond initial curative treatments (e.g., colorectal, breast).	Follow surveillance guidelines
After ablative therapy	Minimally invasive medical procedure that destroys abnormal tissue (cancer cells) using heat, cold or other methods, often used as an alternative to surgery or in conjunction with other treatments. Ablative therapy can be considered curative or palliative depending on numerous factors such as other sites of disease, cancer subtype, and ablative technique used.	See disease-specific guidelines

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Treatment Phase	Phase Definition	Imaging Timeframe
Chemotherapy or immunotherapy for active disease	• Measurable disease: Cancerous lesions that can be accurately measured in size to allow for assessment of tumor response. Solid lesions need to be 1 cm of greater while lymph nodes should be 1.5 cm or greater (short axis) • Non-measurable active disease: Lesions that cannot be accurately measured based on size (see above) in addition to lesions such as bone, pleural effusions, ascites, and carcinomatosis. • Active disease: Cancer thought to be "active" whether it is measurable or non-measurable. For lesions that are not clear (i.e., due to small size, chronically stable, not proven to be malignant), provider discretion is to be used to include modality.	Every 2 cycles (generally every 6 to 8 weeks)
During endocrine/ hormonal therapy alone for active disease	E.g., tamoxifen, letrozole, anastrozole, exemestane	Every 3 months (12 weeks)
Active disease on maintenance therapy	Maintenance therapy is given to help keep cancer from coming back after it has shrunk or disappeared following initial therapy and may be given for a finite or indefinite timeframe.	See disease-specific guidelines
Active metastatic disease being monitored off therapy		Every 3 months (12 weeks)

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Treatment Phase	Phase Definition	Imaging Timeframe
Metastatic disease in complete response on or off therapy	Defined as all measurable and non-measurable disease has disappeared (to include normalized tumor markers if applicable) and rarely translates into cure but remission can be sustained for years	• Once complete remission (CR) has been achieved, frequency of imaging should be no sooner than every 3 months and continue for up to 5 years from the time of CR • Follow surveillance guidelines

- Advanced imaging is not indicated for evaluation of in situ or non-invasive cancers or cancer surveillance after complete surgical removal of primary disease unless otherwise stated in the cancer-specific guidelines.
- Advanced imaging is not indicated for monitoring disease in individuals who choose to not receive standard oncologic therapy but may be receiving alternative therapies or palliative care and/or hospice. All advanced imaging indicated for initial staging of the specific cancer type can be approved once when the individual is considering initiation of a standard therapeutic approach (surgery, chemotherapy, or radiation therapy).
- Brain imaging is performed for signs or symptoms of brain disease
  - MRI Brain without and with contrast (CPT® 70553) is the recommended study for evaluation of suspected or known brain metastases. If a non-contrast CT head shows suspicious lesion, MRI Brain may be obtained to further characterize the lesion.
  - CT without and with contrast (CPT® 70470) can be approved when MRI is contraindicated or not available, or if there is skull bone involvement.
  - Certain malignancies including, but not limited to, melanoma and lung cancer have indications for brain imaging for asymptomatic individuals.
  - If stage IV disease is demonstrated elsewhere or if systemic disease progression is noted, refer to disease specific guidelines.
  - Initiation of angiogenesis therapy is not an indication for advanced imaging of the brain in asymptomatic individuals (Avastin/Bevacizumab; <3% risk of bleeding and <1% risk of serious bleeding).
- Bone Scan:

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  - Primarily used for evaluation of bone metastases in individuals with solid malignancies.
  - Indications for bone scan in individuals with history of malignancy include – bone pain, rising tumor markers, elevated alkaline phosphatase or in individuals with primary bone tumor.
  - For evaluation of suspected or known bony metastases, CPT® 78306 (Nuclear bone scan whole body), may be approved.
  - Radiopharmaceutical Localization scan SPECT (CPT® 78803 or CPT® 78831) or SPECT/CT (CPT® 78830 or CPT® 78832) may be approved as an add-on test for further evaluation of a specific area of interest.
  - CPT® codes 78300 (Nuclear bone scan limited), 78305 (Nuclear bone scan multiple areas) or 78315 do not have any indications in oncology nuclear medicine imaging.
- Bone scan supplemented by plain x-rays are the initial imaging modalities for suspected malignant bone pain. For specific imaging indications, see also:
  - **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)**
  - **Bone (Non-Vertebral) Metastases (ONC-31.5)**
  - **Spinal/Vertebral Metastases (ONC-31.6)**
  - **Carcinoma of Unknown Primary Site (ONC-31.7)**
- Advanced imaging used for radiation therapy treatment planning should not be authorized using any of the diagnostic imaging codes for CT, MRI, or PET.
  - Advanced imaging performed in support of radiation therapy treatment planning should be reported with CPT® 76498 for Unlisted MRI or CPT® 76497 for Unlisted CT scan.
- Delay PET/CT for at least 12 weeks after completion of radiation treatment, unless required sooner for imminent surgical resection.
- PET/CT may be considered prior to biopsy in order to determine a more favorable site for biopsy when a prior biopsy was nondiagnostic or a relatively inaccessible site is contemplated which would require invasive surgical intervention for biopsy attempt.
- PET/CT may be indicated if:
  - conventional imaging (CT, MRI or bone scan) reveals findings that are inconclusive or negative, with continued suspicion for recurrence
- Unless specified in diagnosis-specific guideline section PET/CT Imaging is NOT medically necessary for:
  - infection, inflammation, trauma, post-operative healing, granulomatous disease, rheumatological conditions
  - concomitant use with separate diagnostic CT studies
  - conclusive evidence of distant or diffuse metastatic disease on recent conventional imaging studies
  - metastatic disease in the central nervous system (CNS)

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  - lesions less than 8 mm in size
  - follow up after localized therapy (i.e. radiofrequency ablation, embolization, stereotactic radiation, etc.)
  - rare malignancies, due to lack of available evidence regarding the diagnostic accuracy of PET in rare cancers
  - surveillance
    - Serial monitoring of individuals who are not currently receiving anti-tumor treatment or are receiving maintenance treatment
    - Serial monitoring of FDG avidity until resolution.
    - PET/CT avidity in a residual mass at the end of planned therapy is not an indication for PET/CT imaging during surveillance.
    - Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging
- Unless otherwise specified for a specific cancer type, once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET should not be used for continued disease monitoring or surveillance.
- PET/MRI is generally not supported for a vast majority of oncologic conditions due to lack of standardization in imaging technique and interpretation. However, it may be approved select circumstances when the following criteria are met:
  - The individual meets condition-specific guidelines for PET/MRI OR
  - The individual meets ALL of the following:
    - The individual meets guideline criteria for PET/CT, AND
    - PET/CT is not available at the treating institution, AND
    - The provider requests PET/MRI in lieu of PET/CT
  - When the above criteria are met, PET/MRI may be reported using the code combination of PET Whole-Body (CPT® 78813) and MRI Unlisted (CPT® 76498). All other methods of reporting PET/MRI are inappropriate.
    - When clinically appropriate, diagnostic MRI codes may be indicated at the same time as the PET/MRI code combination.
- The specific radiotracer planned to be used with PET/CT imaging is required to perform a medical necessity review. Indications for PET/CT imaging using non-FDG radiotracers are listed in diagnosis-specific guidelines.
  - Supported radiotracers:
    - ^{18}F-FDG
    - ^{68}Gallium DOTATATE (NETSPOT®) for low-grade neuroendocrine tumors and medullary thyroid cancer
    - ^{64}Cu-DOTATATE (DETECTNET®) for low-grade neuroendocrine tumors
    - ^{68}Ga-DOTA-TOC for low-grade neuroendocrine tumors

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        - ^{11}C Choline for prostate cancer
        - ^{18}F-Fluciclovine (AXUMIN®) for prostate cancer
        - ^{68}Ga PSMA-11 for prostate cancer
        - ^{18}F Piflufolastat (Pylarify®) for prostate cancer
        - ^{68}Ga Gozetotide (Illuccix® and Locametz®) for prostate cancer
        - ^{18}F Flotufolastat (Posluma®) for prostate cancer
        - ^{18}F Fluoroestradiol (Cerianna®) for breast cancer
        - ^{18}F Na Fluoride PET bone scan for breast cancer and prostate cancer
    - Unsupported radiotracers:
        - PET/CT imaging using isotopes other than those specified above
- Octreotide scan:
    - Specific for low and intermediate grade neuroendocrine tumors which express specific cell surface somatostatin receptors. See cancer specific guidelines for recommended use.
    - One of the following codes may be approved when Octreotide scan is requested:
    - CPT® 78802 (Radiopharmaceutical localization of tumor whole-body single day study)
    - CPT® 78804 (Radiopharmaceutical localization of tumor whole-body two or more days)
    - In addition to one of the above CPT codes, CPT® 78803 (Radiopharmaceutical localization of tumor SPECT), SPECT CPT® 78831, or hybrid SPECT/CT (CPT® 78830 or 78832) may be approved as an add-on test for further evaluation of a specific area of interest.

Clinical Trials

- Similar to investigational and experimental studies, clinical trial imaging requests will be considered to determine whether they meet these evidence-based guidelines.
- Imaging studies which are inconsistent with established clinical standards, or are requested for data collection and not used in direct clinical management are not supported.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Key Principles (ONC-1.1)

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AGE APPROPRIATE GUIDELINES	
Age of Individual	**Appropriate Imaging Guidelines**
≥18 years old at initial diagnosis	• General Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section
<18 years old at initial diagnosis	• Pediatric and Special Populations Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section
15 to 39 years old at initial diagnosis (defined as Adolescent and Young Adult (AYA) oncology individuals)	• When unique guidelines for a specific cancer type exist only in either General Oncology or Pediatric and Special Populations Oncology, AYA individuals should be imaged according to the guideline section for their specific cancer type, regardless of the individual's age • When unique guidelines for a specific cancer type exist in both General Oncology and Pediatric and Special Populations Oncology, AYA individuals should be imaged according to the age rule in the previous bullet

- Conventional Imaging (mostly CT, sometimes MRI or bone scan) of the affected area(s) drives much of initial and re-staging and surveillance. PET is not indicated for surveillance imaging unless specifically stated in the diagnosis-specific guideline sections.
- Brain imaging is performed for signs or symptoms of brain disease.
  - MRI Brain without and with contrast (CPT® 70553) is the recommended study for evaluation of suspected or known brain metastases.
  - MRI Brain without and with contrast (CPT® 70553) may be obtained if a non-contrast CT Head shows suspicious lesion.
  - CT Head without and with contrast (CPT® 70470) can be approved when MRI is contraindicated or not available, or if there is skull bone involvement.
  - Initiation of angiogenesis therapy is not an indication for advanced imaging of the brain in asymptomatic individuals (Avastin/Bevacizumab; <3% risk of bleeding and <1% risk of serious bleeding).

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- Individuals receiving cardiotoxic chemotherapy (such as doxorubicin, trastuzumab, pertuzumab, mitoxantrone, etc.) may undergo cardiac evaluation – at baseline and for monitoring while on active therapy.
  - Echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308) rather than MUGA scan for determination of LVEF and/or wall motion.
    - MUGA Scan may be performed instead of ECHO in individuals who have a low LV ejection fraction of <50% on a prior ECHO or MUGA, pre-existing left ventricular wall motion abnormalities from ischemic or non-ischemic cardiomyopathies, congestive heart failure or when ECHO is technically limited and prevents accurate assessment of LV function.
    - A prior MUGA is not a reason to approve another MUGA (it is not necessary to compare LVEF by the same modality).
  - The timeframe for monitoring the ejection fraction should be determined by the provider, but no more often than baseline and at every 6 weeks.
  - May repeat every 4 weeks if cardiotoxic chemotherapeutic drug is withheld for significant left ventricular cardiac dysfunction.
  - See: **Oncologic Indications for Cancer Therapeutics-Related Cardiac Dysfunction (CTRCD) (CD-12.1)** in the Cardiology Imaging Guidelines
- CTA or MRA of a specific anatomic region is indicated when requested for surgical planning when there is suspected vascular proximity to proposed resection margin.
- Adults (≥18 years) with a diagnosis of Li-Fraumeni Syndrome (LFS) may be screened for malignancy with a Whole-Body MRI (CPT® 76498) on an annual basis. Annual Brain MRI (CPT® 70553) may be performed as part of Whole-Body MRI or as a separate exam. Due to lack of standardization of technique, interpretation, and availability of Whole-Body MRI, individuals with LFS are encouraged to participate in clinical trials.

Use of Contrast

- CT imaging should be performed with contrast for known or suspected body regions, unless contraindicated.
  - Shellfish allergy is not a contraindication to contrast. Individuals with known shellfish allergy do not have contrast reaction any more often than other atopic individuals or individuals with other food allergies.
  - For iodinated contrast dye allergy, either CT scans without contrast or MRI scans without and with contrast are indicated.
  - If CT scanning is considered strongly indicated in an individual with known contrast allergy, CT with contrast may be considered to be safely performed following prednisone premedication over a 24-hour period prior to the study.
- For individuals with renal insufficiency which precludes contrast use, CT without contrast appropriate disease-specific areas should be offered. Further imaging (such as MRI) may be indicated if non-contrast CT results are inconclusive.

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- Severe renal insufficiency, i.e. an eGFR less than 30, is a contraindication for an MRI using a gadolinium-based contrast agent (GBCA) as well. In individuals with eGFR greater than 40, GBCA administration can be safely performed. GBCA administered to individuals with acute kidney injury or severe chronic kidney disease can result in a syndrome of nephrogenic systemic fibrosis (NSF), but GBCAs are not considered nephrotoxic at dosages approved for MRI.
- Gadolinium deposition has been found in individuals with normal renal function following the use of gadolinium based contrast agents (GBCAs).
  - The U.S. Food and Drug Administration (FDA) is investigating the risk of brain deposits following repeated use of GBCAs.
  - The FDA has noted that, “It is unknown whether these gadolinium deposits are harmful or can lead to adverse health effects.” and have recommended:
    - To reduce the potential for gadolinium accumulation, health care professionals should consider limiting GBCA use to clinical circumstances in which the additional information provided by the contrast is necessary.
    - Health care professionals are also urged to reassess the necessity of repetitive GBCA MRIs in established treatment protocols.

Radiation Exposure

- The use of MRI in place of CT scans to reduce risk of secondary malignancy from radiation exposure during CT is not supported by the peer-reviewed literature. Unless otherwise specified in the Guidelines, MRI in place of CT scans for this purpose alone is not indicated. In some instances (i.e., testicular cancer surveillance), MRI may be considered inferior to CT scans.

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Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)

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- This section does not apply to PET imaging. PET imaging considerations can be found in **PET Imaging in Oncology (ONC-1.4)**
- Bone Scan:
  - Primarily used for evaluation of bone metastases in individuals with solid malignancies.
  - Indications for bone scan in individuals with history of malignancy include – bone pain, rising tumor markers, elevated alkaline phosphatase or in individuals with primary bone tumor.
  - For evaluation of suspected or known bony metastases, CPT® 78306 (Nuclear bone scan whole-body), may be approved.
    - Radiopharmaceutical Localization scan SPECT (CPT® 78803 or CPT® 78831) or SPECT/CT (CPT® 78830 or CPT® 78832) may be approved for further evaluation of unclear findings on a whole body scan or clinically suspected lesions with negative whole body bone scan or documented bone metastasis.
  - CPT® codes 78300 (Nuclear bone scan limited), 78305 (Nuclear bone scan multiple areas) or 78315 do not have any indications in oncology nuclear medicine imaging.
- Octreotide scan:
  - Specific for low and intermediate grade neuroendocrine tumors which express specific cell surface somatostatin receptors. See cancer specific guidelines for recommended use.
  - One of the following codes may be approved when Octreotide scan is requested:
    - CPT® 78802 (Radiopharmaceutical localization of tumor whole-body single day study)
    - CPT® 78804 (Radiopharmaceutical localization of tumor whole-body two or more days)
  - In addition to one of the above CPT codes, CPT® 78803 (Radiopharmaceutical localization of tumor SPECT), SPECT CPT® 78831, or hybrid SPECT/CT (CPT® 78830 or 78832) may be approved as an add-on test for further evaluation of a specific area of interest.
- Bone marrow imaging:
  - This study is rarely performed for evaluation of the entire bone marrow in conditions like myeloproliferative disorders, sickle cell bone infarct or ischemia, avascular necrosis or myeloma.

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  - The correct CPT code for this study is CPT® 78104 (Diagnostic Nuclear Medicine Procedures on the Hematopoietic, Reticuloendothelial and Lymphatic System).
- Brain imaging SPECT with Technetium-99m or thallium-201 (CPT® 78803 or CPT® 78830):
  - Immunocompromised individuals with mass lesion detected on CT or MRI for differentiation between lymphoma and infection
  - In distinguishing recurrent brain tumor from radiation necrosis
- Radiopharmaceutical localization of tumor or distribution of radiopharmaceutical agent(s):
  - CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78804, CPT® 78803, CPT® 78831 (SPECT), or CPT® 78830 or CPT® 78832 (SPECT/CT)
  - for evaluation of fever of unknown origin and osteomyelitis
  - for suspected infections such as infected central lines, grafts or shunts
- Gallium Isotope Scan:
  - Radiopharmaceutical Localization of tumor (CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, or CPT® 78804), SPECT CPT® 78831, or hybrid SPECT/CT CPT® 78830 or 78832
  - This may be rarely used in place of PET/CT scan when PET/CT scan not available and PET/CT is indicated by guidelines for lymphoma, sarcoma, melanoma or myeloma.

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PET Imaging in Oncology (ONC-1.4)

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- CPT codes:
  - PET imaging in oncology should use PET/CT fusion (CPT® 78815 or CPT® 78816) Unbundling PET/CT imaging into separate PET and diagnostic CT codes is otherwise not supported.
  - The decision whether to use skull base to mid-femur (“eyes to thighs”) procedure code for PET (CPT® 78812 or CPT® 78815) or whole-body PET (CPT® 78813 or CPT® 78816) is addressed in the diagnosis-specific guideline sections.
  - ”Limited area” protocol is done infrequently, but may be considered, and is reported with PET (CPT® 78811) or for PET/CT (CPT® 78814).
- Radiotracers:
  - Unless specified otherwise, the term “PET” refers to ^{18}F-FDG-PET and PET/CT fusion studies.
  - Indications for PET/CT imaging using non-FDG radiotracers are listed in diagnosis-specific guidelines. The indications may be as follows:
- Supported radiotracers:
  - ^{18}F-FDG
  - ^{68}Gallium DOTATATE (NETSPOT®) for low-grade neuroendocrine tumors and medullary thyroid cancer
  - ^{64}Cu-DOTATATE (DETECTNET®) for low-grade neuroendocrine tumors
  - ^{68}Ga-DOTA-TOC for low-grade neuroendocrine tumors
  - ^{11}C Choline for prostate cancer
  - ^{18}F-Fluciclovine (AXUMIN®) for prostate cancer
  - ^{68}Ga PSMA-11 for prostate cancer
  - ^{18}F Piflufolastat (Pylarify®) for prostate cancer
  - ^{68}Ga Gozetotide (Illuccix® and Locametz®) for prostate cancer
  - ^{18}F Flotufolastat (Posluma®) for prostate cancer
  - ^{18}F Fluoroestradiol (Cerianna®) for breast cancer
  - ^{18}F Na Fluoride PET bone scan for breast cancer and prostate cancer
- Unsupported radiotracers:
  - PET/CT imaging using isotopes other than those specified above

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CPT/ HCPCS Code	Code Description	Brand or common name	Guideline Section and Cancer Type
A9552	^{18}F Fluoro deoxyglucose	FDG	Various guideline sections where PET is indicated
A9580	^{18}F Sodium fluoride	N/A	ONC-11: Breast Cancer, ONC-19: Prostate Cancer
A9587	^{68}Ga-68 Dotatate	NETSPOT®	ONC-15: Low-grade neuroendocrine tumors, ONC-6: Medullary thyroid cancer
A9515	^{11}C Choline	N/A	ONC-19, Prostate Cancer
A9588	^{18}F-Fluciclovine	AXUMIN®	ONC-19, Prostate Cancer
A9593 A9594	^{68}Ga PSMA-11	N/A	ONC-19, Prostate Cancer
A9595	^{18}F Piflufolastat	Pylarify®	ONC-19, Prostate Cancer
A9596	^{68}Ga Gozetotide	Illuccix®	ONC-19, Prostate Cancer
A9800	^{68}Ga Gozetotide	Locametz®	ONC-19, Prostate Cancer
A9608	^{18}F Flotufolastat	Posluma®	ONC-19, Prostate Cancer
A9591	^{18}F Fluoroestradiol	Cerianna®	ONC-11, Breast Cancer
A9592	^{64}Cu Copper dotatate	Detectnet®	ONC-15, Low-grade neuroendocrine tumors
C9067	^{68}Ga Gallium-DOTA-TOC	N/A	ONC-15, Low-grade neuroendocrine tumors

- Unless specified in diagnosis-specific guideline section PET/CT Imaging is NOT medically necessary for:
  - infection, inflammation, trauma, post-operative healing, granulomatous disease, rheumatological conditions

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- concomitant use with separate diagnostic CT studies
- conclusive evidence of distant or diffuse metastatic disease on recent conventional imaging studies
- metastatic disease in the central nervous system (CNS)
- lesions less than 8 mm in size
- follow up after localized therapy (i.e. radiofrequency ablation, embolization, stereotactic radiation, etc.)
- rare malignancies, due to lack of available evidence regarding the diagnostic accuracy of PET in rare cancers
- surveillance
  - Serial monitoring of individuals who are not currently receiving anti-tumor treatment or are receiving maintenance treatment
  - Serial monitoring of FDG avidity until resolution.
  - PET/CT avidity in a residual mass at the end of planned therapy is not an indication for PET/CT imaging during surveillance.
  - Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging
- Unless otherwise specified for a specific cancer type, once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET should not be used for continued disease monitoring or surveillance.

- PET/CT may be indicated if:
  - Conventional imaging (CT, MRI or bone scan) reveals findings that are inconclusive or negative, with continued suspicion for recurrence
  - The individual is undergoing salvage treatment for a recurrent solid tumor with residual measurable disease on conventional imaging and confirmed repeat negative PET imaging will allow the individual to transition from active treatment to surveillance.
  - PET/CT may be considered prior to biopsy in order to determine a more favorable site for biopsy when a prior biopsy was nondiagnostic or a relatively inaccessible site is contemplated which would require invasive surgical intervention for biopsy attempt.
- PET/CT for rare malignancies is not covered by these guidelines due to lack of available evidence regarding diagnostic accuracy of PET/CT in the majority of rare cancers. Conventional imaging studies should be used for initial staging and treatment response for these diagnoses. PET/CT can be approved if all of the following apply:
  - Conventional imaging (CT, MRI or bone scan) reveals equivocal or suspicious findings.

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  - No other specific metabolic imaging (MIBG, octreotide, technetium, etc.) is appropriate for the disease type.
  - The submitted clinical information describes a specific decision regarding the individual's care that will be made based on the PET/CT results.
- Delay PET/CT for at least 12 weeks after completion of radiation treatment, unless required sooner for imminent surgical resection.
- PET mammography (PEM, generally reported with CPT® 78811) is considered experimental and investigational at this time.
- PET/MRI is generally not supported for a vast majority of oncologic conditions due to lack of standardization in imaging technique and interpretation. However, it may be approved in select circumstances when the following criteria are met:
  - The individual meets condition-specific guidelines for PET/MRI OR
  - The individual meets ALL of the following:
    - The individual meets guideline criteria for PET/CT, AND
    - PET/CT is not available at the treating institution, AND
    - The provider requests PET/MRI in lieu of PET/CT
  - When the above criteria are met, PET/MRI may be reported using the code combination of PET Whole-Body (CPT® 78813) and MRI Unlisted (CPT® 76498). All other methods of reporting PET/MRI are inappropriate.
    - When clinically appropriate, diagnostic MRI codes may be indicated at the same time as the PET/MRI code combination.

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Unlisted Procedure Codes in Oncology (ONC-1.5)

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- There is often no unique procedure code for a service performed solely for treatment planning purposes. AMA instructions in the CPT state that if no specific code exists for a particular service, the service is reported with an unlisted code.
- Advanced imaging being used for radiation therapy treatment planning should not be authorized using any of the diagnostic imaging codes for CT, MRI or PET. Advanced imaging performed in support of radiation therapy treatment planning should be reported with:
  - **CPT® 76498 for Unlisted MRI** – when MRI will be used for treatment planning of radiation therapy to be delivered ONLY to the brain, prostate and cervix. The use of this code for radiation treatment planning of any other cancers/body parts not listed above may be reviewed on a case-by-case basis.
  - **CPT® 76497 for Unlisted CT** – may NOT be used for radiation treatment planning. CT imaging performed in support of radiation therapy treatment planning is bundled in with the concurrent radiation treatment authorization codes and a separate authorization for treatment planning is not required.
  - Imaging associated with image-directed biopsy should be reported with the corresponding interventional codes. See also: CT-, MR-, or Ultrasound-Guided Procedures (Preface-4.2) in the Preface Imaging Guidelines.
  - For advanced imaging used solely for the purpose of Surgical planning, see: **Unlisted Procedures/Therapy treatment planning (Preface-4.3)** in the Preface Imaging Guidelines

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Predisposition Syndromes (ONC-1.6)

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- For predisposition syndrome screening in adult individuals, see: **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)** in the Pediatric and Special Populations Oncology Imaging Guidelines

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Primary Central Nervous System Tumors (ONC-2)

Guideline

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Primary Central Nervous System Tumors – General Considerations (ONC-2.1)

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- This guideline section applies to primary CNS tumors only. For imaging guidelines in metastatic brain cancer, see the appropriate diagnosis-specific section or **Brain Metastases (ONC-31.3)** for imaging guidelines.
- Primary brain tumors presenting only with uncomplicated headache are very uncommon. Most primary brain tumors present with specific CNS symptoms.
- Histologic confirmation is critical. Therapeutic decisions should not be made on radiographic findings alone, except for ANY of the following:
  - Medically fragile individuals for whom attempted biopsy carries excess medical risk, as stated in writing by both the attending physician and surgeon.
  - Brain stem tumors or other sites where the imaging findings are pathognomonic and the risk of permanent neurological damage is excessive with even a limited biopsy attempt.
- For evaluation of known or suspected spinal cord compromise, see: **Spinal/Vertebral Metastases (ONC-31.6)**
- For suspected brain tumors in neurofibromatosis, see: **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)** in the Pediatric and Special Populations Oncology Imaging Guidelines
- Rare tumors occurring more commonly in the pediatric population should be imaged according to the imaging guidelines in: **Pediatric Central Nervous System Tumors (PEDONC-4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

Indication	Medically Necessary Imaging Study
Characterization and follow up of all brain tumors	• MRI Brain without and with contrast (CPT® 70553) • CT Head without and with contrast (CPT® 70470) when MRI is contraindicated or not available, or there is skull bone involvement • CT Head (contrast as requested) for preoperative planning when requested by the operating surgeon
Preoperative planning or to clarify inconclusive findings on MRI or CT	• MRA Head (CPT® 70544) or CTA Head (CPT® 70496)

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Indication	Medically Necessary Imaging Study
Within 24 to 72 hours following brain tumor surgery	• MRI Brain without and with contrast (CPT® 70553)
Clinical deterioration or development of new neurological features	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) for signs/symptoms of spinal involvement or if spinal involvement is suspected

MR Spectroscopy in Brain Tumors (MRS, CPT® 76390)

- MRS is only medically necessary for use in brain tumors of specified histologies where diagnostic accuracy has been established in peer-reviewed literature.
  - See diagnosis-specific guidelines for MRS indications
- MRS is considered not medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section.

PET Brain Imaging (CPT® 78608 and CPT® 78609)

- PET Brain Metabolic Imaging (CPT® 78608) is only medically necessary for use in brain tumors of specified histologies where diagnostic accuracy has been established in peer-reviewed literature.
- PET Brain metabolic imaging (CPT® 78608) is considered not medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section.
- PET Brain perfusion imaging (CPT® 78609) is not medically necessary in the evaluation or management of primary CNS tumors.
- Body PET studies (CPT® 78811, CPT® 78812, and CPT® 78813) and fusion PET/CT studies (CPT® 78814, CPT® 78815, or CPT® 78816) are not medically necessary in the evaluation or management of primary CNS tumors.
- See: **Other Imaging Studies (HD-24)** in the Head Imaging Guidelines for details on other advanced neuro-imaging studies.

Evidence Discussion

Primary central nervous system tumors account for 1.4% of all new cancer diagnoses in the United States and 2.7% of deaths due to cancer. Primary central nervous system tumors develop within any region of the brain. Utilizing the World Health Organization (WHO) classification of tumors, Low grade tumors (WHO I, II) are the most common primary brain tumors. (71.7% of all tumors). High grade tumors (WHO II/IV) account for

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28.3% of all tumors. Meningioma is the most common low grade tumor accounting for 39.7% of all tumors. Glioblastoma is the most common malignant glioma accounting for 15.4% of all tumors. The most recent classification of these tumors is based on histology and on molecular diagnostics.

The primary imaging modality for the evaluation of primary brain tumors is an MRI Brain and Spine (with and without contrast). The standard MRI protocol minimally includes T1 and T2, fluid-attenuated inversion recovery (FLAIR), gradient-echo/susceptibility, diffusion-weighted imaging, and post contrast T1-weighted images to characterize the tumor. MRI provides much better characterization of intracranial parenchymal tumors in comparison to CT. MRI is more sensitive in detecting lesions in the posterior fossa and in evaluation of leptomeningeal spread of tumor. CT imaging (with and/or without contrast) is valuable in the emergent scenario to assist in initial description of the disease. CT imaging of brain and spine should be used in individuals with a contraindication for use of MRI (those with metallic implants or those who experience claustrophobia). If there is bone involvement, CT imaging may be included with MRI for disease assessment.

Advanced imaging modalities may be included to complement standard imaging to further characterize tumors and assist in treatment decisions. MRI perfusion measures blood flow in the tumor and can be useful in differentiating viable tumor versus radiation necrosis, in determining tumor grade and in determining optimal site for biopsy. MR Spectroscopy involves analysis of the levels of certain chemicals in pre-selected voxels (small regions) on an MRI scan done at the same time. MR spectroscopy may be useful in defining grade of tumor or differentiate viable tumor from radiation necrosis. The use of MR spectroscopy is limited to specific histologies based on peer-reviewed literature. A major limitation of both modalities is the added imaging time. Brain FDG-PET imaging may also be considered to differentiate viable tumor versus radiation necrosis, to determine optimal biopsy site and to determine tumor grade. The use of Brain FDG-PET imaging is limited to specific histologies based on peer-reviewed literature.[1,7,8,9,10,11,12]

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Low Grade Gliomas (ONC-2.2)

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- These tumors are defined as having a World Health Organization (WHO) histologic grade of I or II (out of IV), can occur anywhere in the CNS, and includes the following tumors:
  - Pilocytic Astrocytoma
  - Fibrillary (or Diffuse) Astrocytoma
  - Optic Pathway Gliomas
  - Pilomyxoid Astrocytoma
  - Oligodendroglioma
  - Oligoastrocytoma
  - Oligodendrocytoma
  - Subependymal Giant Cell Astrocytoma (SEGA)
  - Ganglioglioma
  - Gangliocytoma
  - Dysembryoplastic infantile astrocytoma (DIA)
  - Dysembryoplastic infantile ganglioglioma (DIG)
  - Dysembryoplastic neuroepithelial tumor (DNT)
  - Tectal plate gliomas
  - Cervicomedullary gliomas
  - Pleomorphic xanthoastrocytoma (PXA)
  - Any other glial tumor with a WHO grade of I or II

Indication	Medically Necessary Imaging Study
Initial Staging	• MRI Brain without and with contrast (CPT® 70553) if not already done • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) ◦ MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) if being performed immediately following a contrast-enhanced MRI Brain

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Indication	Medically Necessary Imaging Study
After initial resection or other treatment (radiation therapy, etc.)	• MRI Brain without and with contrast (CPT® 70553)
For individuals undergoing chemotherapy treatment	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles • Individuals with spinal cord involvement at diagnosis can have MRI without and with contrast of the involved spinal region on the same schedule as MRI brain
ONE of the following: • Determine need for biopsy when transformation to high-grade glioma is suspected based on clinical symptoms or recent MRI findings • Evaluate a brain lesion of indeterminate nature when the study will be used to determine whether biopsy/resection can be safely postponed	ANY of the following: • PET Brain Metabolic Imaging (CPT® 78608) • MRI Perfusion imaging (CPT® 70553)
ONE of the following: • Distinguish low-grade from high-grade gliomas • Evaluate a brain lesion of indeterminate nature when the study will be used to determine whether biopsy/resection can be safely postponed • Distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy	ANY of the following: • MR Spectroscopy (CPT® 76390) • MRI Perfusion imaging (CPT® 70553)
Suspected intracranial or intraspinal recurrence	• All imaging supported for initial staging

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Indication	Medically Necessary Imaging Study
Surveillance, grade I	• MRI Brain without and with contrast (CPT® 70553) every 3 months for up to 5 years • Individuals with spinal cord involvement at diagnosis: MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) on the same schedule as MRI Brain
Surveillance, grade II (not poor performance status as defined below)	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 5 years, then every 6 months thereafter • Individuals with spinal cord involvement at diagnosis: MRI Spine without and with contrast (Cervical- CPT® 72156, Thoracic- CPT® 72157, Lumbar- CPT® 72158) on the same schedule as MRI Brain
Surveillance, grade II and poor performance status (Karnofsky Performance Score <60)	• MRI Brain without and with contrast (CPT® 70553) every 2 months for 3 years, then every 3 months thereafter • Individuals with spinal cord involvement at diagnosis: MRI Spine without and with contrast (Cervical- CPT® 72156, Thoracic- CPT® 72157, Lumbar- CPT® 72158) on the same schedule as MRI Brain

Evidence Discussion

The primary brain tumors classified as low grade gliomas are listed in the guideline. Initial staging in low grade glioma includes both MRI Brain as well as MRI Whole Spine. Whole spine MRI imaging is indicated for initial staging as the finding of spinal metastases, leptomeningeal disease will impact prognosis, and treatment approaches. MRI studies should be completed both without and with gadolinium contrast. However, MRI Spine with contrast only can be approved if being performed immediately following a contrast-enhanced MRI Brain for patient-centricity to limit time in the MRI machine if requested since the non-contrast component is less essential for the evaluation of spine. MRI imaging is completed after initial resection or radiation to establish a new

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baseline for disease monitoring. If intracranial or intraspinal recurrence is suspected or documented, MRI imaging that was completed for initial staging is repeated.

In individuals undergoing active therapy, MRI imaging may be repeated after every 2 cycles of therapy for disease assessment. If there is spine involvement, MRI Spine of the involved spinal region can be included on this same schedule.

Advanced imaging modalities such as MRI perfusion imaging, MR Spectroscopy and/or PET Brain Metabolic Imaging used in conjunction with standard MRI imaging can be performed to characterize non-invasively changes in the tumor not noted on standard MRI or as problem solving tools with inconclusive findings on MRI imaging. Results of these advanced imaging modalities may be the basis to pursue additional treatment and/or surgical intervention; to define transition of the tumor to a higher grade or to distinguish between radiation-induced radiation necrosis and progressive disease within 18 months of completing radiation therapy.

Surveillance imaging is conducted at a frequency and interval based on published standards noted in the NCCN guidelines. Individuals with poor performance status require more frequent imaging than others because of their higher risk for complications or recurrence.[1] More frequent imaging may be done as clinically indicated by the treating physician, in the event of clinical changes such as development of seizures or neurologic deterioration that are suspicious for disease progression.[8,1,9,7,10,11,12]

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High Grade Gliomas (ONC-2.3)

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- These tumors are defined as having a WHO histologic grade of III or IV (out of IV can occur anywhere in the CNS (though the majority occur in the brain), and include the following tumors:
  - Anaplastic astrocytoma
  - Glioblastoma multiforme
  - Diffuse intrinsic pontine glioma (DIPG, or “brainstem glioma”)
  - Gliomatosis cerebri
  - Gliosarcoma
  - Anaplastic oligodendroglioma
  - Anaplastic ganglioglioma
  - Anaplastic mixed glioma
  - Anaplastic mixed ganglioneuronal tumors
  - Any other glial tumor with a WHO grade of III or IV

Indication	Medically Necessary Imaging Study
Initial Staging	• MRI Brain without and with contrast (CPT® 70553) if not already done • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) ◦ MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) if being performed immediately following a contrast-enhanced MRI Brain
Immediately following partial or complete resection	• MRI Brain without and with contrast (CPT® 70553)
Immediately following radiation therapy (XRT)	• MRI Brain without and with contrast (CPT® 70553) once within 2 to 8 weeks following completion of treatment, and then go to surveillance imaging

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Indication	Medically Necessary Imaging Study
For individuals undergoing chemotherapy treatment	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles • Individuals with spinal cord involvement at diagnosis: MRI without and with contrast of the involved spinal region on the same schedule as MRI Brain
ONE of the following: • Distinguish low-grade from high-grade gliomas • Evaluate a brain lesion of indeterminate nature when the study will be used to determine whether biopsy/resection can be safely postponed • Distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy	ANY of the following: • MR Spectroscopy (CPT® 76390) • MRI Perfusion imaging (CPT® 70553)
ONE of the following: • Distinguish radiation-induced tumor necrosis from progressive disease • Evaluate inconclusive MRI findings when the study will be used to determine need for biopsy or change in therapy, including a change from active therapy to surveillance • Evaluate a brain lesion of indeterminate nature when the study will be used to determine whether biopsy/resection can be safely postponed	ANY of the following: • MRI Perfusion imaging (CPT® 70553) • PET Brain metabolic imaging (CPT® 78608) ◦ PET Brain is not medically necessary in gliomas occurring in the brain stem due to poor uptake and lack of impact on individual outcomes
Suspected intracranial or intraspinal recurrence	• All imaging supported for initial staging

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Indication	Medically Necessary Imaging Study
Surveillance	• MRI Brain without and with contrast (CPT® 70553) every 2 months for 3 years and every 3 months thereafter • Individuals with spinal cord involvement at diagnosis: MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) on the same schedule as MRI Brain

Evidence Discussion

The primary brain tumors classified as high grade gliomas are listed in the guideline. Initial staging in high grade glioma includes both MRI Brain as well as MRI Whole spine. Whole spine MRI imaging is indicated for initial staging as the finding of spinal metastases and leptomeningeal disease will impact prognosis and treatment approaches. MRI studies should be completed both without and with gadolinium contrast. However, MRI Spine with contrast only can be approved if being performed immediately following a contrast-enhanced MRI Brain for patient-centricity to limit time in the MRI machine if requested, since the non-contrast component is less essential for the evaluation of spine. MRI imaging is indicated after initial resection or radiation to establish a new baseline for disease monitoring. If intracranial or intraspinal recurrence is suspected or documented, MRI imaging that was completed for initial staging is repeated.

In individuals undergoing active therapy, MRI imaging may be repeated after every 2 cycles of therapy for disease assessment. If there is spine involvement, MRI spine of the involved spinal region can be included on this same schedule.

Advanced imaging modalities such as MRI perfusion imaging, MR Spectroscopy and/or PET Brain Metabolic Imaging used in conjunction with standard MRI imaging can be performed as problem solving tools to characterize non-invasively changes in the tumor noted on standard MRI. Results of these advanced imaging modalities may be the basis to pursue additional treatment and/or surgical intervention; to distinguish low-grade from high-grade gliomas; to distinguish between radiation-induced radiation necrosis and progressive disease within 18 months of completing radiation therapy.

Surveillance imaging is conducted at a frequency and interval based on published standards noted in the NCCN guidelines. More frequent imaging may be done as clinically indicated by the treating physician, in the event of a clinical change such as development of seizures or neurologic deterioration that is suspicious for disease progression.[8,1,9,7,10,11,12]

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Medulloblastoma and Supratentorial Primitive Neuroectodermal Tumors (sPNET) (ONC-2.4)

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- Medulloblastoma and sPNET imaging indications in adult individuals are identical to those for pediatric individuals. See: **Medulloblastoma (MDB), Supratentorial Primitive Neuroectodermal Tumors (sPNET), and Pineoblastoma (PEDONC-4.4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Ependymoma (ONC-2.5)

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- Ependymoma imaging indications in adult individuals are identical to those for pediatric individuals. See: **Ependymoma (PEDONC-4.8)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Central Nervous System Germ Cell Tumors (ONC-2.6)

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- Central nervous system germ cell tumor imaging indications in adult individuals are identical to those for pediatric individuals. See: **CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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CNS Lymphoma (Also Known as Microglioma) (ONC-2.7)

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Indication	Medically Necessary Imaging Study
Initial Staging	ALL of the following: • MRI Brain without and with contrast (CPT® 70553) • MRI Cervical Spine without and with contrast (CPT® 72156) • MRI Thoracic Spine without and with contrast (CPT® 72157) • MRI Lumbar Spine without and with contrast (CPT® 72158)
Extra-neural evaluation to confirm CNS primary *Individuals with CNS Lymphoma that is metastatic should be imaged according to: • **Non-Hodgkin Lymphomas (ONC-27)** for individuals age ≥18 years • **Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)** in the Pediatric Oncology Imaging Guidelines for individuals age ≤17 years	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • PET/CT (CPT® 78815) for evaluation of inconclusive findings on CT imaging
Treatment Response	• MRI without and with contrast of all positive disease sites every 2 cycles
Suspected intracranial or intraspinal recurrence	• All imaging supported for initial staging

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Indication	Medically Necessary Imaging Study
Surveillance	• MRI without and with contrast of all positive disease sites every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter

Evidence Discussion

Primary central nervous system lymphoma is an aggressive non-Hodgkin lymphoma that can occur in any location within the intracranial neuraxis (brain, spine, cranial nerves, and leptomeninges). This malignancy can occur in immunocompromised individuals or immunocompetent individuals and represents approximately 4% of all intracranial malignancies. Individuals may present with focal neurological deficits or nonspecific neurological findings depending on the specific location of tumor involvement.

For initial staging, MRI Brain without and with contrast and whole spine imaging without and with gadolinium contrast are indicated. CNS lymphoma has potential to spread throughout the intracranial neuraxis. For confirmation as a primary central nervous system lymphoma, extra neural evaluation is indicated and follows the ONC-27 Non-Hodgkin Lymphoma Guideline. This evaluation includes CT Chest with contrast and CT Abdomen and Pelvis with contrast. FDG PET/CT can be approved if extra-neural CT imaging is inconclusive. Evaluation of treatment response can be assessed after every 2 cycles of treatment with MRI without and with contrast of all positive disease sites. All imaging obtained for initial staging is repeated for suspected disease recurrence to evaluate for metastatic disease. Surveillance imaging includes MRI Brain without and with of all positive disease sites on a schedule outlined in the guideline.[1,12,16]

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Meningiomas (Intracranial and Intraspinal) (ONC-2.8)

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Indication	Medically Necessary Imaging Study
Initial Staging of Intracranial Meningioma	ANY or ALL of the following: • MRI Brain without and with contrast (CPT® 70553) • CT Head (contrast as requested)
Initial staging of Intraspinal Meningioma	ONE of the following: • MRI without and with contrast of appropriate spinal region (Cervical CPT® 72156, Thoracic CPT® 72157, and Lumbar CPT® 72158) OR • CT without and with contrast of the appropriate spinal region (Cervical CPT® 72127, Thoracic CPT® 72130, and Lumbar CPT® 72133)
Inconclusive MRI Brain or CT Head and further imaging is needed to confirm diagnosis	• Dotatate PET/CT Brain (CPT® 78814)
Treatment Response	• MRI without and with contrast of all positive disease sites every 2 cycles
Suspected recurrence of intracranial or intraspinal disease	• All imaging supported for initial staging
Suspected recurrence with inconclusive findings on MRI	Any ONE of the following studies: • Octreotide SPECT Brain (CPT® 78803) • Octreotide SPECT/CT Brain (CPT® 78830) • Dotatate PET/CT Brain (CPT® 78814)

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Indication	Medically Necessary Imaging Study
Surveillance for Grade I (low-grade) and Grade II (atypical) intracranial meningioma (completely resected, partially resected, and unresected)	• MRI Brain without and with contrast (CPT® 70553) every 3 months in year 1, then every 6 months for 5 years ◦ Imaging every 1-3 years thereafter as clinically indicated
Surveillance for Grade I (low-grade) and Grade II (atypical) intraspinal meningioma (completely resected, partially resected, and unresected)	ONE of the following at 3, 6, and 12 months, and then every 6 months for 5 years: • MRI without and with contrast (CPT® 72156 [Cervical spine], CPT® 72157 [Thoracic spine], CPT® 72158 [Lumbar spine]) of the involved spinal level OR • CT without and with contrast (CPT® 72127 [Cervical spine], CPT® 72130 [Thoracic spine], CPT® 72133 [Lumbar spine]) of the involved spinal level • Imaging every 1-3 years thereafter as clinically indicated
Surveillance for Grade III (malignant or anaplastic) Meningioma	• Intracranial Meningioma: MRI Brain without and with contrast (CPT® 70553) every 2 months for 3 years, and then every 3 months thereafter • Intraspinal Meningioma: MRI or CT without and with contrast of the involved spinal region every 2 months for 3 years and then every 3 months thereafter

Evidence Discussion

Meningiomas are the most frequent primary central nervous system tumors, accounting for approximately 34% of all primary brain and spine tumors. Meningiomas are extra-axial, dural-based tumors that are derived from the dura and occur throughout the neuroaxis. Meningiomas are a heterogeneous group of tumors that have been classified in three histologic grades, WHO Grades I (benign), II and III (aggressive). Meningiomas can involve bone resulting in bone overgrowth or infiltration into bony structures. Meningiomas are associated with genetic syndromes and molecular alterations.

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Approaches for classification are evolving to incorporate histopathologic, genetic and molecular characteristics.

The standard imaging modality is MRI without and with contrast. CT imaging is supported, as there is potential for bone involvement. Meningioma overexpresses somatostatin receptors. PET imaging using various radiolabeled somatostatin receptor ligands (SSAs) such as 68Ga-DOTA-Tyr3-octreotide (DOTATOC), 68Ga-DOTA-d-Phe1-Tyr3-octreotide (DOTATATE), or 68Ga-DOTA-l-Nal3-octreotide (DOTANOC) have been used for the diagnostic evaluation of meningioma. PET imaging with these ligands is supported for restaging to clarify inconclusive findings on MRI imaging. ^{111}In-octreotide scintigraphy (octreotide) imaging has a similar imaging indication as ^{68}Ga-DOTATATE PET/CT. In surveillance, the schedule for follow-up MRI/CT imaging is based on tumor grade and extent of residual disease.[1,17,18,19]

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Spinal Cord Tumors (Benign and Malignant) (ONC-2.9)

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- See: **Low Grade Gliomas (ONC-2.2)** and **High Grade Gliomas (ONC-2.3)** for imaging guidelines of low-grade and high-grade gliomas of the spinal cord
- See: **Malignant Tumors of the Spinal Cord (PEDONC-4.9)** in the Pediatric and Special Populations Oncology Imaging Guidelines for other malignant spinal cord tumors
- See: **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines for spinal tumors in individuals with Neurofibromatosis 1 or 2
- See: **Spinal/Vertebral Metastases (ONC-31.6)** for known secondary malignancy involving the spine/spinal canal/spinal cord

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Choroid Plexus Tumors (ONC-2.10)

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- Choroid Plexus Tumor imaging indications in adult individuals are identical to those for pediatric individuals. See: **Choroid Plexus Tumors (PEDONC-4.13)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Squamous Cell Carcinomas of the Head and Neck (ONC-3)

Guideline

Squamous Cell Carcinomas of the Head and Neck – General Considerations (ONC-3.0)
Squamous Cell Carcinomas of the Head and Neck – Suspected/Diagnosis (ONC-3.1)
Squamous Cell Carcinomas of the Head and Neck – Initial Work-up/Staging (ONC-3.2)
Squamous Cell Carcinomas of the Head and Neck – Restaging/Recurrence (ONC-3.3)
Squamous Cell Carcinomas of the Head and Neck – Surveillance/Follow-up (ONC-3.4)
References (ONC-3)

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Squamous Cell Carcinomas of the Head and Neck – General Considerations (ONC-3.0)

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- Individuals with esthesioneuroblastoma should be imaged according to this guideline section.
- Stage III/IV disease encompasses any primary tumor larger than 4 cm or documented lymph node positive disease.

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Squamous Cell Carcinomas of the Head and Neck – Suspected/Diagnosis (ONC-3.1)

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- See: **Neck Masses - Imaging (NECK-5.1)** in the Neck Imaging Guidelines for evaluation of suspected malignancy in the neck.
- PET is medically necessary prior to biopsy in order to determine a more favorable site for biopsy when:
  - a prior biopsy was nondiagnostic or
  - a relatively inaccessible site is contemplated which would require invasive surgical intervention for biopsy attempt

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Squamous Cell Carcinomas of the Head and Neck – Initial Work-up/Staging (ONC-3.2)

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Indication	Medically Necessary Imaging Study
All Stages of Disease	• CT Neck with contrast (CPT® 70491) **or** MRI Orbits/Face/Neck (OFN) without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)
For sentinel lymph node evaluation when nodes are not clinically positive	• Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
Nasal cavity and paranasal sinuses (bony erosion or skull base and intracranial involvement)	ONE of the following: • CT Maxillofacial with contrast (CPT® 70487) • CT Neck with contrast (CPT® 70491) • MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
Nasopharyngeal (NPC) Cancer	• PET/CT (CPT® 78815) • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is the preferred study ◦ CT Neck (CPT® 70491) **and/or** CT Maxillofacial (CPT® 70487) with contrast if contraindication to MRI • CT Chest with contrast (CPT® 71260)

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Indication	Medically Necessary Imaging Study
For ANY of the following: • Known stage III or IV disease • To determine role for upfront surgery vs chemoradiation in T3-T4 size tumor • Prior to start of primary chemoradiotherapy and have not undergone definitive surgical resection • Inconclusive findings on conventional imaging (CT, MRI) • In order to direct laryngoscopy/exam under anesthesia for biopsy • Pulmonary nodule(s) ≥8 mm in size • Cervical lymph node biopsy positive for squamous cell carcinoma and no primary site identified on CT or MRI Neck and Chest • Inconclusive findings suggestive of disease outside the head and neck area	• PET/CT (CPT® 78815)
Signs or symptoms of abdominal metastatic disease including, but not limited to, elevated liver function tests	• CT Abdomen with contrast (CPT® 74160)
Any head and neck cancer with neurological findings or suspicion of skull base invasion	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

Accurate initial staging guides prognosis and management options. CT Neck with contrast or MRI Neck with and without contrast is required for correct tumor, nodal, and metastases (TNM) staging. A contrasted CT of the chest is also supported if requested.

- Classification of tumor staging involves determination of mass size and extent of invasion, if present, of surrounding structures.
- Size and location (including laterality and nodal basin) of pathologic lymph nodes is also required for accurate nodal staging, which will further direct treatment planning to include the extent of potential neck dissection and/or field of radiation.

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Lymphoscintigraphy is supported, with SPECT if requested, for sentinel node evaluation when nodes are not clinically obviously positive.

- Assessment for potential distant metastases ("M") is based on clinical signs/symptoms and the presence of advanced locoregional primary disease. Discovery of distant metastasis, or a second primary, shifts management to more systemic options. A heavy smoking history also may be a separate indication for advanced imaging of the chest. Up to 7% to 14% of individuals may have a separate lung primary at the time of initial staging of head and neck SCCa. The use of IV contrast improves the detection of mediastinal and hilar adenopathy, and generally, CT Chest with contrast is preferred. Given the rarity of abdominal or pelvic metastatic disease, abdominopelvic imaging is only supported for signs and symptoms of metastatic disease.
  - Nasopharyngeal carcinoma (NPC) has a relatively high rate of distant metastases compared with other head and neck cancers, being found in 5% to 11% of individuals at the time of initial diagnosis. The most common sites of metastasis are bone (20%), lung (13%), and liver (9%).
- FDG-PET/CT Skull Base to Mid-Thigh detects and localizes primary tumor site, and can be helpful in squamous cell carcinoma (SCCa) of the head and neck with unknown primary. It is also equivalent to and possibly superior to contrast-enhanced CT Neck for accurate diagnosis of regional nodal disease. It is helpful in confirming distant metastases as well. The National Comprehensive Cancer Network (NCCN) recommended FDG-PET/CT for initial staging of any NPC, as well as for individuals with locoregionally advanced SCCa (ie, T3-T4 primary or >/= N1 nodal staging).
  - PET/CT alone, however, is not sufficient for initial staging. It does not provide the necessary anatomic detail of the primary tumor's extent for accurate "T" staging, which is required for best selection of local disease management options. Contrast-enhanced CT Neck or MRI Neck are necessary adjuncts.
  - If imaging fails to reveal an obvious primary, PET/CT should be completed before exam under anesthesia, biopsies, and tonsillectomy, to help identify potential primary sites before any intervention occurs.[1,6,7,8,9,10,11,12]

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Squamous Cell Carcinomas of the Head and Neck – Restaging/Recurrence (ONC-3.3)

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Indication	Medically Necessary Imaging Study
Following complete resection and/or radical neck dissection	See: **Surveillance/Follow-up (ONC-3.4)**
Following primary chemoradiotherapy or radiation therapy in individuals who have not undergone surgical resection of primary tumor or neck dissection	ONE of the following: • CT Neck with contrast (CPT® 70491); **or** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543); **or** • PET/CT (CPT® 78815) no sooner than 12 weeks (3 months) post completion of radiation therapy ◦ If post-treatment PET/CT scan is negative, further surveillance imaging is not routinely indicated.
Post-treatment PET is positive for residual disease	ONE of the following: • CT Neck with contrast (CPT® 70491); **or** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
Post-treatment PET is equivocal for residual disease	PET/CT (CPT® 78815) at 3-6 months from prior post-treatment PET/CT
Induction chemotherapy response	• CT Neck with contrast (CPT® 70491) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • PET not indicated to assess response to induction chemotherapy
Concern for progression seen on imaging performed at completion of induction chemotherapy	EITHER or ALL of the following: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • PET/CT (CPT® 78815)

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Indication	Medically Necessary Imaging Study
Measurable or metastatic disease undergoing active treatment	Every 2 cycles (6-8 weeks): • CT Neck with contrast (CPT® 70491) OR • MRI Orbits/ Face/Neck without and with contrast (CPT® 70543) AND • CT with contrast of involved body sites
Suspected local recurrence	• CT Neck with contrast (CPT® 70491) or MRI Orbits/ Face/Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260)
Biopsy proven local recurrence	ONE of the following: • PET/CT (CPT® 78815) **or** • CT Neck with contrast (CPT® 70491) or MRI Orbits/ Face/Neck without and with contrast (CPT® 70543) and CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)
Inconclusive conventional imaging (CT or MRI)	• PET/CT (CPT® 78815)
Any of the following: • Known prior thoracic disease • New or worsening pulmonary symptoms • New or worsening chest x-ray findings	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)

Evidence Discussion

Follow-up imaging is required for the evaluation of treatment response. In alignment with the NCCN, a PET/CT is supported following primary chemoradiotherapy in individuals who have not undergone surgical resection of the primary tumor or neck dissection. For individuals receiving induction chemotherapy prior to definitive therapy, a CT or MRI of the primary tumor site to assess response is recommended by NCCN after 2-3 cycles

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of induction, but a repeat PET/CT is not routinely recommended by NCCN unless there are unclear findings on this CT or MRI. For individuals with metastatic disease on active treatment, cross sectional imaging of involved body areas is supported every 2 cycles. PET/CT is supported for only as a problem-solving tool for inconclusive conventional imaging, as the incidence of false positive findings is high in the setting of ongoing inflammation with known disease.

For individuals treated with primary chemoradiotherapy, a negative PET at the 3-6 month timeframe predicts improved survival at 2 years, with a negative predictive value of 95% to 97%. PET/CT performed earlier than this timeframe is associated with higher false-positive findings and should be avoided. CT or MRI neck may be performed **in lieu of** PET/CT, but PET/CT has excellent sensitivity and specificity in this setting, so these studies are generally not supported **in addition to** a PET/CT as they add additional radiation without a clear impact on management.

For suspected local recurrence, CT or MRI of the primary site (neck/face) is supported, as well as CT of the chest as lung and mediastinal nodes are the most common site of metastatic disease at recurrence, often without pulmonary symptoms. PET/CT has a relatively high false-positive rate due to ongoing inflammatory changes, and thus these guidelines do not support PET/CT for suspected recurrence until recurrence is proven by biopsy.[1,6,7,8,9,10,11,12]

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Squamous Cell Carcinomas of the Head and Neck – Surveillance/Follow-up (ONC-3.4)

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Indications	Medically Necessary Imaging Study
Individuals treated with surgical resection of primary site and/or neck dissection (with or without postoperative radiation therapy)	Once within 6 months of completing all treatment: • CT Neck with contrast (CPT® 70491) **or** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT with contrast of any other involved body area
If above post-treatment CT or MRI imaging shows residual abnormalities	ONE of the following, once within 6 months of prior imaging: • CT Neck with contrast (CPT® 70491) OR • MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
Individuals treated with definitive radiation therapy or combined chemoradiation, and post-treatment imaging is negative	Further surveillance imaging is not routinely medically necessary
After initial post-treatment study, for ANY of the following: • Nasopharyngeal primary site • Difficult physical exam due to the effects of treatment (e.g., scarring, etc.) or unable to visualize deep-seated primary site	Annually for 3 years: • CT Neck with contrast (CPT® 70491) **or** MRI Orbits/Face/Neck without and with contrast (CPT® 70543)

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Indications	Medically Necessary Imaging Study
• CT Chest is not medically necessary for surveillance. Individuals with smoking history may undergo annual low dose CT cancer screening if criteria are met (See: **Lung Cancer Screening (CH-33)** in the Chest Imaging Guidelines)	

Evidence Discussion

Timely detection and accurate assessment of the extent of recurrent disease will direct salvage therapy and improve prognosis. A thorough head and neck clinical examination will typically guide any additional imaging that may be necessary, after post-treatment baseline imaging. There is no controlled prospective data showing a survival benefit for long term surveillance imaging. 3-year disease free survival in individuals undergoing surveillance imaging vs those undergoing clinical surveillance only is not significantly different (41% vs 46%, P=0.91). Given the excellent NPV of PET/CT 3-6 months post therapy, and the fact that median time to recurrence is 6 months, eviCore guidelines support cross sectional imaging once within 6 months of completion of therapy, following the initial post treatment PET/CT. For individuals whose primary tumor site cannot be evaluated with physical exam and for patients with nasopharyngeal primary tumors, CT Neck or MRI Face/Orbit/Neck are supported annually for 3 years, as 80% to 90% of recurrences occur within 3 years.

The role of annual CT Chest screening for surveillance of lung metastasis is controversial in head and neck cancer, following primary definitive treatment (surgery, XRT, or systemic therapy/XRT). Further study is needed to determine the extent of the positive effect and/or cost-effectiveness of this approach. Individuals with a heavy smoking history may be at increased risk, and may meet criteria for low-dose CT lung cancer screening as defined in CH-33 in the Chest Imaging Guidelines.[1,6,7,8,9,10,11,12]

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Salivary Gland Cancers (ONC-4)

Guideline

Salivary Gland Cancers – General Considerations (ONC-4.0)
Salivary Gland Cancers – Suspected/Diagnosis (ONC-4.1)
Salivary Gland Cancers – Initial Work-up/Staging (ONC-4.2)
Salivary Gland Cancers – Restaging/Recurrence (ONC-4.3)
Salivary Gland Cancers – Surveillance/Follow-up (ONC-4.4)
References (ONC-4)

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Salivary Gland Cancers – General Considerations (ONC-4.0)

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- Salivary gland tumors may originate within the parotid, submandibular, sublingual or minor salivary glands in the mouth.
- Histological subtypes include:
  - mucoepidermoid
  - acinic
  - adenocarcinoma
  - adenoid cystic carcinoma
  - malignant myoepithelial tumors
  - squamous cell carcinoma
  - lymphoma and metastatic squamous carcinoma can occur in the parotid gland
- Over 80% of parotid gland tumors are benign. A bilateral parotid tumor is most likely Warthin's tumor.

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Salivary Gland Cancers – Suspected/ Diagnosis (ONC-4.1)

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- See: **Salivary Gland Disorders (NECK-11)** and **Neck Masses – Imaging (NECK-5.1)** in the Neck Imaging Guidelines for evaluation of salivary gland masses, salivary gland stones, and neck masses.

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Salivary Gland Cancers – Initial Work-up/ Staging (ONC-4.2)

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Indication	Medically Necessary Imaging Study
Biopsy-proven malignancy	ONE of the following: • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT Neck with contrast (CPT® 70491) • CT Neck without contrast (CPT® 70490)
Skull base invasion	• MRI Brain without and with contrast (CPT® 70553)
• Adenoid cystic carcinoma • Lymphadenopathy in the neck • Pulmonary signs or symptoms • Abnormal chest x-ray	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

There are over 40 histologies of salivary gland malignancies, with different patterns of presentation and invasiveness. The choice of MRI vs CT depends on location of tumor, specific symptoms, and individual characteristics. CT may be useful to assess stones and sialadenitis, which may mimic tumor, and is superior for assessing cortical bone erosion. MRI is superior in the assessment of extent of soft tissue disease and perineural invasion. Contrast is recommended in all studies to better outline primary site and to better assess nodal involvement.

In individuals who present with metastatic disease outside the neck, 90% are lung/ chest node metastases. Metastatic disease to lung is most common with adenoid cystic carcinoma and chest imaging is supported in all individuals with this histology. Contrast should be used to allow for assessment of nodal disease in the chest. In other histologies, metastatic disease is less common, and thus chest imaging is only supported in individuals with neck adenopathy, abnormal chest x-ray, or pulmonary signs and symptoms.

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Perineural and skull base invasion may occur with salivary gland cancers, particularly with adenoid cystic carcinoma, where perineural spread is seen in 50% to 60% of individuals. When skull base invasion is clinically suspected an MRI Brain with and without contrast is supported by these guidelines in the interest of patient safety. MRI with and without gadolinium and with fat-saturated, T1 weighted MRI sequences is the most sensitive technique to evaluate for invasion of skull base and perineural invasion.

The role of PET/CT remains controversial in salivary gland cancers. Several studies show no statistically significant different in outcomes with imaging with PET/CT vs conventional imaging. The rate of change in treatment plan based on imaging with PET/CT is widely variable across studies, ranging from 15% to 47%. PET/CT is not adequate to distinguish benign from malignant parotid tumors. Benign tumors such as Warthin's tumor can have FDG uptake, and low-grade malignant tumors may not take up FDG. Healthy salivary glands may also exhibit FDG uptake and obscure tumors. While there is emerging evidence in the use of FDG-PET/CT and PET/MRI to assess for distant disease and perineural spread, it is not considered routine at this time and is not routinely recommended by the NCCN. PET/CT thus remains reserved as a problem-solving tool when conventional imaging is inconclusive.[1-13]

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Salivary Gland Cancers – Restaging/ Recurrence (ONC-4.3)

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Indication	Medically Necessary Imaging Study
After complete surgical resection	One-time imaging to establish baseline within 3-4 months of surgical resection is medically necessary using ONE of the following: • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT Neck with contrast (CPT® 70491) • CT Neck without contrast (CPT® 70490)
Individuals with unresected disease receiving systemic therapy (chemotherapy)	Every 2 cycles: • CT Neck with contrast (CPT® 70491) OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT with contrast or MRI without and with for any other sites of disease
Recurrence or progression suspected based on new or worsening signs or symptoms	ONE of the following: • CT Neck with contrast (CPT® 70491) • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) In addition, for all individuals: • CT Chest with contrast (CPT® 71260)
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)
All other individuals	• No routine advanced imaging

Evidence Discussion

NCCN supported one-time imaging with CT or MRI to establish a new baseline for future comparison.[1] CT or MRI based on initial tumor and individual characteristics

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is supported every 2 cycles of systemic chemotherapy. If recurrence of progression is clinically suspected at any time, CT neck with contrast or MRI without and with contrast is supported based on prior tumor characteristics and symptoms, per NCCN recommendations and American College of Radiology ACR Appropriateness Criteria®. The incidence of metastatic disease to the chest is higher at recurrence than at initial presentation, with 63% of individuals with metastatic recurrence presenting with metastatic disease to the chest, so CT Chest is supported for suspected recurrence. Contrast should be used to allow better assessment of nodal disease, in addition to parenchymal lesions. PET/CT remains a tool for the clarification of inconclusive findings on conventional imaging. Any CNS symptoms warrant MRI with further guidance in guideline ONC-31.3.[1-13]

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Salivary Gland Cancers – Surveillance/ Follow-up (ONC-4.4)

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Indication	Medically Necessary Imaging Study
Total surgical resection	• No routine advanced imaging
Unresectable or partially resected disease, including those treated with radiation therapy	• Either CT Neck with contrast (CPT® 70491) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) once within 6 months of completion of treatment
Adenoid cystic carcinoma	ANY of the following, annually for up to 10 years: • CT Neck with contrast (CPT® 70491) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)

Evidence Discussion

The mainstay of surveillance for head and neck cancers including salivary gland carcinoma are frequent history and physical examination. For most histologies, no survival benefit has been documented with imaging surveillance over clinical surveillance. The NCCN notes most recurrences are picked up by patient report of symptoms. For all histologies other than adenoid cystic carcinoma, guidelines support imaging of the primary tumor site once within 6 months from completion of therapy to establish post-treatment baseline, with further imaging guided by signs and symptoms of recurrence. Adenoid cystic carcinoma has the highest incidence of metastatic disease, with over 60 percent of individuals presenting with metastatic disease at recurrence having a history of this histology. They also have the longest risk of recurrence, with a median time to recurrence of 3 years with some recurrences occurring as late as 10 years from diagnosis. For individuals with a history of adenoid cystic carcinoma, CT Neck with contrast or MRI Orbit/Face/Neck as well as CT Chest with or without contrast are supported annually for up to 10 years.[1-13]

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Melanomas and Other Skin Cancers (ONC-5)

Guideline

Melanoma – General Considerations (ONC-5.0)
Melanoma – Suspected/Diagnosis (ONC-5.1)
Melanoma – Initial Work-up/Staging (ONC-5.2)
Melanoma – Restaging/Recurrence (ONC-5.3)
Melanoma – Surveillance/Follow-up (ONC-5.4)
Non-Melanoma Skin Cancers – General Considerations (ONC-5.5)
Non-Melanoma Skin Cancers – Initial Work-up/Staging (ONC-5.6)
Non-Melanoma Skin Cancers – Restaging/Recurrence (ONC-5.7)
Non-Melanoma Skin Cancers – Surveillance/Follow-up (ONC-5.8)
Ocular Melanoma (ONC-5.9)
References (ONC-5)

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Melanoma – General Considerations (ONC-5.0)

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- Melanomas can metastasize in an unpredictable fashion.

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Melanoma – Suspected/Diagnosis (ONC-5.1)

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Indication	Medically Necessary Imaging Study
All	• Imaging is not medically necessary until histologic diagnosis is confirmed

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Melanoma – Initial Work-up/Staging (ONC-5.2)

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Indication	Medically Necessary Imaging Study
Stage 0 or IA (in situ or disease <1 mm)	• Routine advanced imaging is not medically necessary
• Stage IB (<0.8 mm with ulceration or 0.8-1 mm without or with ulceration) • Stage II (lesions >1 mm thick, but node negative)	• CT with contrast or MRI without and with contrast of specific areas, only if signs or symptoms indicate need for further evaluation
For sentinel lymph node evaluation in stages IB and II	• Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
Any of the following: • Stage III (sentinel node positive, palpable regional nodes) • Stage IV (metastatic)	• MRI Brain without and with contrast (CPT® 70553) AND one of the following: • PET/CT (CPT® 78815 or CPT® 78816) OR • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)
• Head or neck primary site • Palpable lymphadenopathy in the neck • Mucosal melanoma of the head or neck region	In addition to above initial staging imaging, if PET/CT not performed: • CT Neck with contrast (CPT® 70491)
• Primary site of melanoma is unknown **and** CT Chest, Abdomen, and Pelvis are negative	• PET/CT (CPT® 78815 or CPT® 78816)

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Evidence Discussion

Formal diagnosis and clinical staging of melanoma needs to take place before any imaging is completed as 84% of individuals present with localized disease, 9% with regional disease, and 4% with distant metastatic disease. Stage 0 (in situ) or 1A does not require routine advanced imaging as the 5 year survival rate is >98% with very little risk for recurrence or metastases. For Stage IB or II disease, sentinel lymph node mapping is indicated and depending on results, survival rates range from 50% to 90% that also incorporates tumor thickness, ulceration and mitotic rate. The yield of imaging in screening individuals with clinical Stage 0-II disease for asymptomatic distant metastatic disease is very low due to low sensitivity and false positive findings. Therefore, the NCCN does not recommend imaging unless needed for surgical planning or to evaluate specific signs or symptoms of disease. Stage III or IV disease can be staged using PET/CT or CT Chest/Abdomen/Pelvis, with PET/CT often preferred due to its superiority over CT in detecting distant metastases. Baseline MRI brain is indicated with or without symptoms due to high risk of CNS involvement estimated to be 15.8% at 5 years for Stage III and up to 60% overall in individuals with advanced stage disease. If primary site of melanoma is unknown and CT Chest/Abdomen/Pelvis negative, PET/CT can be performed due to its higher sensitivity and ability to image the extremities.[1,7,17-19]

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Melanoma – Restaging/Recurrence (ONC-5.3)

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- All recurrences should be confirmed histologically, except when excessive morbidity from a biopsy may occur, such as a biopsy requiring craniotomy.

Indication	Medically Necessary Imaging Study
Individuals receiving chemotherapy, with measurable disease, every 2 cycles (commonly every 6 to 8 weeks)	• CT Chest with contrast (CPT® 71260); **and** CT Abdomen and Pelvis with contrast (CPT® 74177)
All in situ recurrences	• Restaging imaging is not medically necessary after adequate aggressive local therapy (See Surveillance below)
Documented or clinically suspected (see top of page regarding biopsy morbidity) recurrence at: • True scar recurrence (if re-excision suggests more than in situ disease) • In-transit disease • Regional lymph nodes • Metastatic site	• CT Chest with contrast (CPT® 71260); **and** CT Abdomen and Pelvis with contrast (CPT® 74177) In addition, for all individuals: • MRI Brain without and with contrast (CPT® 70553) In addition, for head or neck primary: • CT Neck with contrast (CPT® 70491)
ANY of the following: • Inconclusive findings on conventional imaging • Isolated metastatic site found on conventional imaging	• PET/CT (CPT® 78815 or CPT® 78816)

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Indication	Medically Necessary Imaging Study
Brain imaging is indicated for: • New discovery of metastatic disease or progression of metastatic disease • Signs or symptoms of CNS disease • If considering Interleukin (IL-2) therapy	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

Individuals receiving chemotherapy with measurable disease can undergo CT Chest/Abdomen/Pelvis every 2 cycles. In situ recurrences do not require restaging imaging due to its high cure rate while recurrence at the primary site, in-transit disease, regional nodes and metastatic site can undergo CT Chest/Abdomen/Pelvis and MRI Brain due to the higher risk of CNS involvement. PET/CT is reserved for inconclusive findings or isolated metastatic site found on CT imaging to guide decisions on local versus systemic therapy. MRI Brain is also indicated for newly diagnosed or progressive metastatic disease, signs or symptoms of CNS involvement or if IL-2 therapy is being considered.[1]

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Melanoma – Surveillance/Follow-up (ONC-5.4)

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Indication	Medically Necessary Imaging Study
Stage 0, IA, IB and IIA Melanomas	• No routine advanced imaging
Stage IIB, IIC, IIIA and IIIB Melanomas	• CT Chest (CPT® 71260) **and** CT Abdomen and Pelvis (CPT® 74177) with contrast every 6 months for 2 years, then annually for 3 years • For melanoma arising from extremities, advanced imaging of the primary site is not routinely medically necessary for surveillance in asymptomatic individuals.
Stage IIIC and IV Melanomas	• CT Chest (CPT® 71260) **and** CT Abdomen and Pelvis (CPT® 74177) with contrast every 3 months for 2 years, then every 6 months for 3 years • MRI Brain without and with contrast (CPT® 70553) annually for 3 years • For melanoma arising from extremities, advanced imaging of the primary site is not routinely medically necessary for surveillance in asymptomatic individuals.
Mucosal Melanoma of the head or neck region	In addition to above stage-based surveillance imaging, ONCE within 6 months of completing all treatment: • CT Neck with contrast (CPT® 70491) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT with contrast of any other involved body area
Liver metastases treated with focal therapy	• See: **Liver Metastases (ONC-31.2)**

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Evidence Discussion

The majority of recurrences, especially in those with early stage disease, are detected clinically by either the patient or during physical exam thus supporting no surveillance imaging in stage 0, 1A, 1B and IIA disease. Furthermore, additional studies have reported low yield, significant false positivity (often associated with increased anxiety and medical costs), and risks of cumulative radiation exposure. Individuals with more advanced disease are more likely to recur, and recur more quickly, with the risk of recurrence reaching low levels after only 2.7 years, thereby supporting stage IIB/IIC as well as stage IIIA/IIIB undergoing CT Chest/Abdomen/Pelvis every 6 months for 2 years then annually for 3 years. Due to the even high risk of early distant recurrence, Stage IIIC/IV should undergo CT Chest/Abdomen/Pelvis every 3 months for 2 years then every 6 months for 3 years as well as MRI brain annually for 3 years. The utility of PET/CT scan in sentinel lymph positive stage III melanoma was minimal with only 2 out of 38 patients (108 total scans) being true positive with 9 scans showing false positive results thus supporting CT rather than PET/CT imaging in this setting. For melanoma arising from extremities, advanced imaging of the primary site is not indicated in asymptomatic individuals. Mucosal melanoma of the head or neck region can also undergo on a one-time basis CT Neck or MRI Orbits/Face/Neck or CT of any other involved body area within 6 months of completing treatment.

The NCCN did not specify the type of imaging required during the workup, response assessment, or surveillance other than listing "cross-sectional with or without brain imaging" thus allowing the provider to determine if CT or PET/CT may be most appropriate. Among the many factors playing a role in this decision are cost, convenience, false positives/false negatives, dye, and radiation exposure.[1,21,22]

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Non-Melanoma Skin Cancers – General Considerations (ONC-5.5)

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- Advanced imaging is generally not medically necessary for basal cell and squamous cell skin cancers.
- PET/CT scan is not medically necessary for evaluation of non-melanoma skin cancers unless specified within the guidelines below (e.g., Merkel cell carcinoma).
- Merkel cell carcinoma is an unusual skin cancer with neuroendocrine-like histologic features, which has a high propensity (25% to 33%) for regional lymph node spread and occasionally, metastatic spread to lungs.
- Merkel cell carcinoma may present as a primary cancer or as a skin metastasis from a non-cutaneous primary neuroendocrine carcinoma (i.e., small cell lung cancer), therefore conventional imaging is indicated initially to confirm the absence of metastasis prior to considering PET scan.

Evidence Discussion

Advanced imaging to include PET/CT is generally not indicated for basal cell (BCC) or squamous cell (SCC) skin cancers. The incidence of metastatic BCC was found to be <1% at 14 years of follow-up while metastatic SCC is noted to be rare. Merkel cell carcinoma, due to its high propensity (25% to 33%) for regional lymph node spread as well as distant metastases (12% to 20%), conventional imaging is indicated initially prior to considering PET scan.[3,15,16]

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Non-Melanoma Skin Cancers – Initial Work-up/Staging (ONC-5.6)

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Indication	Medically Necessary Imaging Study
Body area with unexplained signs or symptoms	• CT with contrast of that body area
Perineural invasion or local regional extension (i.e. bone; deep soft tissue) involvement	ONE of the following of the primary site: • MRI without contrast **or** without and with contrast • CT (contrast as requested)
Skin lesion may be a dermal metastasis from distant primary	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • PET/CT (CPT® 78815 or CPT® 78816) if conventional imaging (CT or MRI) is unable to identify a primary site
Squamous cell carcinoma head or neck skin with regional lymphadenopathy	• CT Neck (CPT® 70491) and CT Chest (CPT® 71260) with contrast
Merkel Cell carcinoma	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of other involved body area(s) • PET/CT (CPT® 78815 or CPT® 78816) if inconclusive conventional imaging • Lymph system imaging (lymphoscintigraphy, CPT® 78195) for sentinel lymph node evaluation ◦ SPECT/CT (CPT® 78830) if requested
Signs or symptoms of CNS involvement	• MRI Brain with and without contrast (CPT® 70553)

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Evidence Discussion

A body area with unexplained signs or symptoms can undergo imaging with CT with contrast. For SCC, MRI or CT of the primary site with MRI favored if there is perineural or deep soft tissue involvement while CT is preferred for bone disease. If the skin lesion is felt to be a dermal metastasis from a distant primary, CT Chest/Abdomen/Pelvis with contrast is the initial recommended imaging with PET/CT indicated if conventional imaging is unable to identify a primary site and especially if the primary tumor may involve an extremity.[3-5]

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Non-Melanoma Skin Cancers – Restaging/Recurrence (ONC-5.7)

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- All recurrences should be confirmed histologically, except when excessive morbidity from a biopsy may occur, such as a biopsy requiring craniotomy.

Indication	Medically Necessary Imaging Study
Recurrence where planned therapy is more extensive than simple wide local excision	• CT with contrast of the primary and recurrent site(s)
Suspected or biopsy-proven recurrence of Merkel cell carcinoma	• CT Chest (CPT® 71260) **and** CT Abdomen and Pelvis (CPT® 74177) with contrast • CT with contrast of other symptomatic body area(s)
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815 or 78816)
Signs or symptoms of CNS involvement	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

For recurrences where planned therapy is more extensive than simple wide local excision, CT with contrast of primary site and recurrent site(s) is indicated. Merkel cell carcinoma recurrence can be evaluated with CT Chest/Abdomen/Pelvis plus any other symptomatic areas. PET/CT can be done for inconclusive findings on conventional imaging. MRI Brain indicated for signs or symptoms of CNS involvement.[3-5]

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Non-Melanoma Skin Cancers – Surveillance/Follow-up (ONC-5.8)

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Indication	Medically Necessary Imaging Study
Merkel cell cancer with any of the following high-risk factors: • Stage II-IV • Individuals assigned male at birth • Immunosuppression • Merkel Cell polyomavirus negative status • Age ≥70 • Non-sentinel lymph node metastases	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast every 6 months for 5 years • Add CT Neck with contrast (CPT® 70491) if known prior neck disease or scalp/facial/neck disease
All others	• Routine advanced imaging for surveillance is not medically necessary • Imaging only for signs and symptoms of recurrent disease

Evidence Discussion

In node positive Merkel cell carcinoma, recurrence rates have been found to be up to 33% at 5 years. In addition, individuals assigned male at birth are twice as likely to develop Merkel Cell carcinoma. Also, the likelihood of developing Merkel Cell increases with age. One study showed that 73.6% of Merkel Cell cases are diagnosed at age greater than 70.[24] Additional risk factors for recurrence of Merkel Cell are polyomavirus negative status, non-sentinel lymph node metastases, and immunosuppression. Routine imaging is not indicated for SCC or BCC in the absence of signs and symptoms of recurrence.[3-5]

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Ocular Melanoma (ONC-5.9)

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General Considerations

- Approximately 95% of ocular melanomas arise from the uvea (iris, ciliary body and choroid) and 5% arise from the conjunctiva or orbit.
- Biopsy is usually not necessary for initial diagnosis of uveal melanoma but may be useful in cases when diagnosis is uncertain (e.g., amelanotic tumors, retinal detachment) or for prognostic analysis and risk stratification.
- Treatment is directed to the affected eye with systemic therapy reserved only for known metastatic disease.
- The most common site of metastatic disease is the liver.
- Surveillance of the affected eye is with clinical examination only; advanced imaging is supported for surveillance of systemic metastatic disease based on individual risk factors. See risk categories below for surveillance recommendations.

Ocular Melanoma Risk Categories

Low Risk	Medium Risk	High-Risk
T1	T2 and T3	T4
Class IA	Class IB	Class 2
Spindle cell histology	Mixed Spindle and Epitheloid cells	Epitheloid cell histology
No extraoccular extension	No extraoccular extension	Extraoccular extension present
No ciliary body involvement	No ciliary body involvement	Ciliary body involvement present
Chromosome mutations: • Disomy 3 • EIF1AX mutation • Gain of chromosome 6p	Chromosome mutations: • SF3B1 mutation	Chromosome mutations: • BAP1 mutation • PRAME mutation • Monosomy 3 • Gain of chromosome 8q

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Indication	Medically Necessary Imaging Study
Initial staging of suspected or biopsy-proven uveal melanoma	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177), OR MRI Abdomen without and with contrast (CPT® 74183) with CT Pelvis with contrast (CPT® 72913), OR MRI Abdomen without and with contrast (CPT® 74183) with MRI Pelvis without and with contrast (CPT® 72197) • MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
Neurological signs/ symptoms	• MRI Brain without and with contrast (CPT® 70553)
Restaging/Suspected Recurrence	ANY or ALL of the following: • MRI Brain without and with contrast (CPT® 70553) • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260) AND one of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Abdomen without and with contrast (CPT® 74183) with CT Pelvis with contrast (CPT® 72913) OR ◦ Ultrasound Abdomen substituted for MRI Abdomen if requested • MRI Abdomen without and with contrast (CPT® 74183) with MRI Pelvis without and with contrast (CPT® 72197)
Surveillance for Low Risk disease	Annually for 10 years: • CT Chest with contrast (CPT® 71260) • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Surveillance for Medium Risk disease	Every 6 months up to year 10: • CT Chest with contrast (CPT® 71260) • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)

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Indication	Medically Necessary Imaging Study
Surveillance for High-risk disease	Every 3 months for 5 years, then every 6 months up to year 10: • CT Chest with contrast (CPT® 71260) • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)

Evidence Discussion

Approximately 95% of ocular melanoma occur in the uvea and 5% from the conjunctiva or orbit. Biopsy may not be mandatory for diagnosis but should be performed if diagnosis is uncertain or for prognostic analysis and risk stratification. Less than 3% of cases present with metastatic disease with 5 year risk of metastasis ranging from 3% to 5% in Stage I to 44% or higher in Stage III. The most common site of metastatic disease is the liver (80%) but may also spread to the lungs, bone, skin/soft tissue and lymph nodes. Surveillance of the affected eye is with clinical examination only with advanced imaging supported based on individual risk factors. Initial staging includes MRI orbits/face/neck to determine extraocular extension that impacts treatment planning (radiation therapy versus enucleation). While the risk of baseline metastases may be low, the NCCN favors baseline staging before treatment to include CT Chest/Abdomen/Pelvis in addition to aforementioned MRI. MRI Brain is indicated with any neurologic signs or symptoms. Restaging/recurrence with same imaging as initial staging as well as including MRI Brain. Surveillance: Local recurrence is rare (<10%) and the development of metastatic disease is much more common (up to 70% up to 20 years after initial diagnosis) hence the following recommendations for surveillance that does not include local imaging unless clinically indicated. For low risk disease, CT Chest with CT or MRI of the abdomen annually for 10 years. For medium risk disease, same as low risk but every 6 months for a total of 10 years. For high-risk disease, every 3 months for 5 years, then annually for a total of 10 years. The NCCN recognized the optimal surveillance strategy is an issue of debate due to overall low yield of testing and the risk of cumulative radiation exposure.[2, 25]

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Thyroid Cancer (ONC-6)

Guideline

Thyroid Cancer – General Considerations (ONC-6.0)
Thyroid Cancer – Suspected/Diagnosis (ONC-6.1)
Thyroid Cancer – Initial Work-up/Staging (ONC-6.2)
Thyroid Cancer – Restaging/Recurrence (ONC-6.3)
Thyroid Cancer – Surveillance/Follow-up (ONC-6.4)
References (ONC-6)

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Thyroid Cancer – General Considerations (ONC-6.0)

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- Individuals of all ages with thyroid cancer are imaged according to this guideline.
- Whole-Body Thyroid Nuclear scan (also known as whole-body radioiodine scan) is coded with CPT® 78018. If CPT® 78018 is obtained and found to be positive, CPT® 78020 is medically necessary as an add-on test to evaluate the degree of iodine uptake.
- Single photon emission computed tomography (SPECT) imaging – Radiopharmaceutical Localization of Tumor SPECT (CPT® 78803 or CPT® 78831) or SPECT/CT Hybrid study (CPT® 78830 or CPT® 78832) may complement planar and pinhole imaging and can be approved as an add-on wherever radioiodine (RAI) scans are indicated.
- Whole-Body Thyroid Nuclear scan (also known as whole-body RAI scan) is the imaging modality of choice for differentiated thyroid cancers, as these are usually not well visualized on FDG-PET/CT scans. Individuals who have RAI-diagnostic scan negative and PET-positive disease will generally not respond to RAI treatment, whereas individuals who have PET-negative and RAI-diagnostic scan negative disease may still be candidates for empiric RAI treatment.
- Radioiodine (RAI) refractory disease is defined as: (i) the malignant/metastatic tissue does not ever concentrate RAI (no uptake outside the thyroid bed at the first therapeutic WBS), (ii)the tumor tissue loses the ability to concentrate RAI after previous evidence of RAI-avid disease (in the absence of stable iodine contamination), (iii) RAI is concentrated in some lesions but not in others, and (iv) metastatic disease progresses despite significant concentration of RAI.

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Thyroid Cancer – Suspected/Diagnosis (ONC-6.1)

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- See: **Thyroid Nodule (NECK-8.1)** in the Neck Imaging Guidelines for suspected thyroid malignancies.

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Thyroid Cancer – Initial Work-up/Staging (ONC-6.2)

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Follicular, Papillary, and Hürthle Cell (Oncocytic) Carcinomas	Medically Necessary Imaging Study
ONE of the following: • Locally advanced disease or fixation suggested by clinical exam and/or ultrasound • Substernal or bulky disease • Disease precluding full ultrasound examination • Vocal cord paresis	ONE of the following: • MRI Neck without contrast (CPT® 70540) • MRI Neck without and with contrast (CPT® 70543) • CT Neck with contrast (CPT® 70491) if contrast study is necessary for complete pre-operative assessment and use of IV contrast will not delay post-operative use of RAI therapy.
• Extent of thyroid remnant cannot be accurately ascertained from the surgical report, neck ultrasound, or cross-sectional imaging • Unresectable residual disease • To assess for iodine-avid metastatic disease • When the results may alter the decision to treat • Prior to administration of RAI therapy	• Whole-Body Thyroid Nuclear scan (CPT® 78018) ◦ CPT® 78020 as an add-on test to evaluate the degree of iodine uptake AND/OR • SPECT (CPT® 78803, or CPT® 78831), OR SPECT/CT Hybrid study (CPT® 78830, or CPT® 78832)

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Follicular, Papillary, and Hürthle Cell (Oncocytic) Carcinomas	Medically Necessary Imaging Study
Skeletal pain	• Bone scan (CPT® 78306) See also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes • Whole-Body Thyroid Nuclear scan (CPT® 78018) ◦ CPT® 78020 as an add-on test to evaluate the degree of iodine uptake AND/OR • SPECT (CPT® 78803 or CPT® 78831), OR SPECT/CT Hybrid study (CPT® 78830, or CPT® 78832)
Suspicious findings on chest x-ray, US, or substernal extension of mass	• CT Chest without contrast (CPT® 71250)
All other individuals	• Routine preoperative advanced imaging is not medically necessary

Medullary Thyroid Carcinomas	Medically Necessary Imaging Study
ANY of the following: • Elevated CEA levels • Calcitonin level >300pg/mL • Positive lymph nodes	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen with contrast (CPT® 74160) **or** CT Abdomen without and with contrast (CPT® 74170) • MRI Abdomen without and with contrast (CPT® 74183) • Bone scan (CPT® 78306) see also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes

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Medullary Thyroid Carcinomas	Medically Necessary Imaging Study
Skeletal pain	• Bone scan (CPT® 78306) see also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes
Inconclusive findings on conventional imaging	• ^{68}Gallium-labeled PET/CT (CPT® 78815)

Anaplastic/Poorly Differentiated Thyroid Carcinomas	Medically Necessary Imaging Study
All	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) or MRI Orbit, Face, Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • FDG PET/CT (CPT® 78815) • CT Head with contrast (CPT® 70460)
Skeletal pain	• Bone scan see also: **Nuclear Medicine (NM) Imaging (ONC-1.3)** in Oncology

Evidence Discussion

For follicular, papillary and Hurthle cell carcinomas, focused imaging of the neck using contrast enhanced CT or MRI is recommended to assess extent of local disease and guide pre-surgical planning. CT Chest with contrast may be indicated based on these results to include substernal extension of the thyroid mass. There are no established guidelines regarding the minimum gap between contrast enhanced CT with iodinated contrast agents and iodine-131/123 for whole body scintigraphy (WBS) in the treatment of residual disease and distant metastases, with majority recommendation of a gap of 4 weeks and 2 months. In the post-thyroidectomy setting, WBS is recommended to assess for either extent of thyroid remnant, when results may alter the decision to treat and prior to administration of radioactive iodine (RAI) therapy. In the presence of skeletal pain, whole body bone scan or WBS to assess for osseous metastases. Medullary thyroid carcinoma is frequently aggressive with 48% of patients having localized disease, 35% with tumors extending beyond the thyroid into surrounding tissues or regional nodes and 13% with distant metastases typically to the lung, liver

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or bones. Due to these concerns, more extensive staging is indicated that includes contrast enhanced CT of neck, chest and abdomen as well as bone scan if there are elevated CEA levels, calcitonin level >300 pg/nL or positive lymph nodes. Skeletal pain can be imaged with bone scan. Gallium-68 labelled dotatate PET/CT or if not available Indium-111-pentetreotide (Octreoscan) is useful due to high expression of somatostatin receptors in MTC and is indicated if conventional imaging is inconclusive due to its high sensitivity compared to other imaging modalities especially if calcitonin levels are >500. Anaplastic thyroid carcinoma is the most aggressive variant of thyroid cancer with distant metastases in over 50% of cases at presentation most commonly involving the lung, bone and brain with 5 year survival <10%. Complete staging with CT Neck, Chest, Abdomen and Pelvis is supported in conjunction with FDG PET/CT as the diagnostic contrasted CTs may better elucidate vascular and airway involvement. MRI of the brain is also indicated.[4,6,9,10,13]

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Thyroid Cancer – Restaging/Recurrence (ONC-6.3)

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Follicular, Papillary, and Hürthle Cell (Oncocytic) Carcinomas	Medically Necessary Imaging Study
Post-thyroidectomy, if gross residual disease	ONE of the following: • MRI Neck without contrast (CPT® 70540) • MRI Neck without and with contrast (CPT® 70543) • CT Neck with contrast (CPT® 70491)
Within 2 weeks (ideally 7 to 10 days) following the administration of Radioactive Iodine therapy	• Whole-body thyroid nuclear scan (CPT® 78018) • The following may be approved as an add-on test: ◦ CPT® 78020 to evaluate the degree of iodine uptake ◦ SPECT (CPT® 78803, or CPT® 78831), or SPECT/CT Hybrid study (CPT® 78830, or CPT® 78832)
ANY of the following: • Recurrence documented by biopsy • Increasing thyroglobulin level without Thyrogen® stimulation • Thyroglobulin level >2 ng/mL or higher than previous after Thyrogen® stimulation • Anti-thyroglobulin antibody present • Evidence of residual thyroid tissue on ultrasound or physical exam after thyroidectomy or ablation	ALL of the following: • CT Neck with contrast (CPT® 70491) or MRI Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260) • CT with contrast of any symptomatic body area • Whole-body Thyroid Nuclear Scan (CPT® 78018) ◦ As an add-on test: ▪ CPT® 78020 to evaluate the degree of iodine uptake ▪ SPECT (CPT® 78803 or CPT® 78831), or SPECT/CT Hybrid study (CPT® 78830, or CPT® 78832)

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Follicular, Papillary, and Hürthle Cell (Oncocytic) Carcinomas	Medically Necessary Imaging Study
ANY of the following: • Rising thyroglobulin level with negative CT scans AND radioiodine scan • Inconclusive findings on conventional imaging (CT scans and radioiodine scan) • Known radioiodine-refractory disease and CT scans are negative or inconclusive	• FDG PET/CT (CPT® 78815)
Measurable metastatic disease on systemic therapy (no more often than every 2 cycles)	• CT Chest with contrast (CPT® 71260) • CT with contrast of affected or symptomatic body area

Medullary Thyroid Carcinoma	Medically Necessary Imaging Study
ANY of the following: • Elevated CEA levels • Calcitonin level ≥150 pg/mL • Signs or symptoms of recurrence	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) or MRI Neck without and with contrast (CPT® 70543) • CT Chest with contrast (CPT® 71260) • CT Abdomen either with (CPT® 74160) or without and with contrast (CPT® 74170) • Bone scan (CPT® 78306) See also: **Nuclear Medicine (NM) Imaging (ONC-1.3)** in Oncology
Inconclusive conventional imaging with calcitonin ≥150 pg per mL	• ^{68}Gallium-labeled DOTATATE PET/CT (CPT® 78815)
Calcitonin level >500	ONE of the following: • MRI Abdomen without and with contrast (CPT® 74183) • Whole-body MRI (CPT® 76498)

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Anaplastic/Poorly Differentiated Thyroid Carcinoma	Medically Necessary Imaging Study
Measurable metastatic disease on systemic treatment	Any of the following every 2 cycles (usually every 6-8 weeks): • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT of any other involved/symptomatic sites
Signs or symptoms of recurrence	ONE of the following combinations, not both: • CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177) **OR** • FDG PET/CT (CPT® 78815) In addition to one of the above studies: • MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

For Follicular, Papillary and Hurthle Cell carcinomas, CT or MRI neck can be performed if gross residual disease is found in the neck post-thyroidectomy. RAI therapy is administered after thyroidectomy for several reasons to include remnant ablation, treat presumed foci of neoplastic cells and/or treat persistent or recurrent disease. Within 2 weeks of treatment, WBS is indicated to stage the disease and document the I-131 avidity of any structural lesion. Follow-up is usually a combination of exam, laboratories (thyroglobulin and anti-thyroglobulin antibody levels) and ultrasound. If there is concern for recurrence, CT of the neck/chest as well as any symptomatic body area along with WBS should be performed to complete restaging. In the setting of rising thyroglobulin level with negative conventional imaging, inconclusive conventional imaging or known RAI-refractory disease with negative/inconclusive CT scans, FDG PET/CT can be performed due to its high sensitivity (94%) and specificity (80% to 84%) compared to conventional imaging. FDG uptake is associated with a worse prognosis and refractoriness to RAI therapy. Initial imaging for MTC recurrence based on elevated CEA levels, calcitonin level >150 pg/mL or signs/symptoms of recurrence should

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undergo CT or MRI Neck, CT Chest and Abdomen, and bone scan. If this imaging is inconclusive and calcitonin is >150, Gallium-68 labeled dotatate PET/CT or if not available Indium-111-pentetreotide (Octreoscan) is indicated as outlined in the initial work-up/staging section. In individuals with very high calcitonin levels, whole-body MRI or MRI liver are useful for imaging evaluation to evaluate for metastases.[1] Initial imaging for ATC recurrence includes either CT Neck, Chest, Abdomen and Pelvis or FDG PET/CT as well as MRI Brain due to its aggressive and widespread behavior. For individuals on systemic therapy, CT of neck/chest/abdomen/pelvis with any additional involved/symptomatic sites can be done every 2 cycles.[11]

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Thyroid Cancer – Surveillance/Follow-up (ONC-6.4)

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Follicular, Papillary, and Hürthle Cell (Oncocytic) Carcinomas	Medically Necessary Imaging Study
Individuals being monitored on active surveillance	• Neck ultrasound (CPT® 76536) every 6 months for 2 years, and then annually thereafter
All other individuals post-treatment	• Neck ultrasound (CPT® 76536) once at 6-12 months post-treatment, and then annually thereafter
For individuals with ANY of the following: • Node positive disease • RAI-avid metastases	• Whole-body Thyroid Nuclear Scan annually (CPT® 78018) ◦ CPT® 78020 as an add-on test to evaluate the degree of iodine uptake AND/OR • SPECT (CPT® 78803, or CPT® 78831), OR SPECT/CT Hybrid study (CPT® 78830, or CPT® 78832)

Medullary Carcinomas	Medically Necessary Imaging Study
All individuals	• CEA and calcitonin are required for monitoring medullary carcinomas • Routine surveillance imaging is not medically necessary

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Anaplastic/Poorly Differentiated Thyroid Carcinomas	Medically Necessary Imaging Study
All individuals	Every 3 months for 2 years: • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

For follicular, papillary and Hurthle cell carcinomas, all individuals are monitored with ultrasound either every 6 months for 2 years then annually (active surveillance) or once at 6-12 months then annually (post-treatment). For node positive disease or RAI-avid metastases, WBS annually. MTC is monitored with CEA and calcitonin levels with no routine imaging indicated. ATC requires close monitoring with CT neck/chest/abdomen/pelvis and MRI Brain every 3 months for 2 years as the vast majority of relapses occur within this timeframe.[1,9]

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Small Cell Lung Cancer (ONC-7)

Guideline

Small Cell Lung Cancer – General Considerations (ONC-7.0)
Small Cell Lung Cancer – Suspected/Diagnosis (ONC-7.1)
Small Cell Lung Cancer – Initial Work-up/Staging (ONC-7.2)
Small Cell Lung Cancer – Restaging/Recurrence (ONC-7.3)
Small Cell Lung Cancer – Surveillance/Follow-up ONC-7.4
References (ONC-7)

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Small Cell Lung Cancer – General Considerations (ONC-7.0)

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- Combined histologies of small and non-small cell are considered small cell lung cancer. Use this guideline for imaging recommendations for small and large cell high-grade (poorly differentiated) neuroendocrine tumors of the lung.
- Imaging is presently guided by traditional staging of limited or extensive disease.
  - Extensive stage is either metastatic disease or an extent which cannot be encompassed by a single radiotherapy portal.
  - Limited staging is confined to one side of the chest.
- Individuals treated curatively for SCLC are at increased risk for developing a second lung cancer. If new lung nodule is seen on imaging without any evidence of other systemic disease, follow **Lung Metastases (ONC-31.1)** for work-up of nodule.
- For carcinoid (low-grade neuroendocrine tumors) of the lung, see: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)**.

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Small Cell Lung Cancer – Suspected/ Diagnosis (ONC-7.1)

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Indication	Medically Necessary Imaging Study
• Abnormal chest x-ray or clinical suspicion remains high despite a normal chest x-ray in symptomatic individual	• CT Chest without contrast (CPT® 71250) **or** • CT Chest with contrast (CPT® 71260)
• Pulmonary nodule <8 mm in size noted on CT Chest	• See: **Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)** in the Chest Imaging Guidelines
• Pulmonary nodule 8 mm (0.8 cm) to 30 mm (3 cm) seen on CT Chest or MRI Chest	• PET/CT (CPT® 78815) • PET is Positive: Qualifies as initial staging PET/CT
• Pulmonary mass 31 mm (3.1 cm) or greater seen on CT or MRI	• PET/CT (CPT® 78815) prior to biopsy if ONE or MORE of the following applies: ◦ Definitive treatment with resection or radiation will be utilized instead of biopsy if PET confirms limited disease ◦ Multiple possible biopsy options are present within the chest and PET findings will be used to determine the most favorable biopsy site • Biopsy is indicated prior to PET imaging for all other indications in pulmonary masses ≥31 mm (3.1 cm) in size
• Mediastinal/Hilar Mass	See: **Lymphadenopathy (CH-2)** in the Chest Imaging Guidelines
• Paraneoplastic syndrome suspected	See: **Paraneoplastic Syndromes (ONC-30.3)**

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Evidence Discussion

For individuals with suspected lung cancer and an abnormal chest x-ray or a high suspicion for lung cancer with symptoms of lung cancer, a CT Chest is indicated, with or without contrast. If a PET/CT is performed in the workup of a pulmonary nodule and is positive, it qualifies as the initial staging PET. The radiotracer supported for PET/CT for lung cancer is 18-FDG.[6,9] While some small cell lung cancer (SCLC) has a neuroendocrine component, the sensitivity, specificity and predictive value of dotatate PET/CT are uncertain at this time and dotatate PET is not supported.[6,9] Lesions 31mm or greater are considered masses rather than nodules, and should be biopsied rather than re-imaged with PET/CT.[5]

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Small Cell Lung Cancer – Initial Work-up/ Staging (ONC-7.2)

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Indication	Medically Necessary Imaging Study
Initial staging	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Brain without and with contrast (CPT® 70553) • Bone scan (CPT® 78306), if PET/CT not being done or not available (See also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)
• To confirm the extent of disease when initial CT Chest/Abdomen/Pelvis and MRI Brain indicate limited stage disease (confined to one side of the chest) • To evaluate inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)
• Inconclusive bone findings on PET/CT or bone scan	• MRI without and with contrast of the involved body site

Evidence Discussion

SCLC has widespread distant metastatic potential, with 2/3 of individuals having metastatic disease at diagnosis and 10% to 15% including central nervous system disease.[6,9] Diagnostic contrasted CTs of chest, abdomen and pelvis as well as MRI Brain with and without contrast are supported.[1] If PET has been completed in the pulmonary nodule workup, a repeat PET/CT is generally not supported. If a PET/CT was done prior to diagnosis and conventional imaging clearly shows extensive stage disease, a PET/CT does not change management. However, if a PET/CT was not done prior to diagnosis, a PET/CT is supported to confirm limited stage disease prior to treatment, as FDG PET/CT changes stage versus conventional imaging in up to 25% of

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individuals.[6,9] Bony metastatic disease is not unusual in SCLC; thus, evaluation for bony metastatic disease is supported .[1,6]

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Small Cell Lung Cancer – Restaging/ Recurrence (ONC-7.3)

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Indication	Medically Necessary Imaging Study
Treatment response in individuals treated with sequential chemotherapy-radiation or chemotherapy alone* *For individuals receiving systemic therapy and concurrent radiation therapy, response assessment is recommended only after completion of initial therapy (see box below).	After every 2 cycles, ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Brain without and with contrast (CPT® 70553) for measurable brain metastases being treated with systemic therapy • Bone scan (CPT® 78306) (See also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) • PET is not indicated for evaluation of treatment response in SCLC but can be considered on a case-by-case basis.
Following completion of planned treatment: • Chemoradiation • Adjuvant chemotherapy • Radiation	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Brain without and with contrast (CPT® 70553) for measurable brain metastases being treated with systemic therapy • Bone scan (CPT® 78306) (See also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3**) for additional bone scan codes)

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Indication	Medically Necessary Imaging Study
Restaging (suspected recurrence)	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Brain without and with contrast (CPT® 70553) • Bone scan (CPT® 78306) (See: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) • PET is not indicated for evaluation of recurrent SCLC but can be considered on a case-by-case basis.
For response assessment following primary treatment	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

Conventional imaging with contrasted CT Chest, Abdomen and Pelvis is supported every 2 cycles of chemotherapy and at the end of chemoradiation. MRI Brain with and without contrast is supported when there is measurable CNS disease being treated with systemic therapy. If prophylactic cranial irradiation (PCI) is planned, an MRI brain is supported at end of initial treatment as some individuals will harbor asymptomatic brain metastases and will require different management, and PCI would expose the individual to radiation doses and neurotoxicity without benefit.[1,7] CT Chest, Abdomen and Pelvis as well as MRI Brain and bone scan are supported if recurrence is suspected. Further literature is emerging to determine the role of FDG PET-CT for treatment response and suspected recurrence; these guidelines do not routinely support PET/CT for treatment response or suspected recurrence of SCLC, but provide flexibility on a case by case basis, particularly for individuals with bony metastatic disease.[1,8,9]

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Small Cell Lung Cancer – Surveillance/ Follow-up ONC-7.4

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Indication	Medically Necessary Imaging Study
Limited stage SCLC	Every 3 months for two years, every 6 months during year three, and then annually: • CT Chest without (CPT® 71250) or CT Chest with (CPT® 71260) contrast • CT Abdomen and Pelvis with contrast (CPT® 74177)
Extensive stage SCLC	Every 2 months for one year, every 3 months for two years, every 6 months for two years, and then annually: • CT Chest without (CPT® 71250) or CT Chest with (CPT® 71260) contrast • CT Abdomen and Pelvis with contrast (CPT® 74177)
Screening for brain metastases, regardless of PCI status	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 1 year and then every 6 months thereafter
Surveillance of known and/or treated brain metastases	• See: **Brain Metastases (ONC-31.3)**
New lung nodule(s)	• See: **Lung Metastases (ONC-31.1)**

Evidence Discussion

Surveillance with CT contrasted chest, abdomen and pelvis is supported. MRI chest is less sensitive than CT chest and usually not supported as a substitution for lung cancer. Follow up is supported more frequently in the first two years post treatment, as that is when recurrence is most common.[1,6,9] The surveillance timeframe is determined by the initial extent of disease. For those with limited stage disease, these guidelines support the above CT imaging every 3 months for 2 years, every 6 months during year 3, and then annually. For those with extensive stage disease, imaging with above CTs is supported every 2 months for 1 year, every 3 months for two years, every 6 months for 2 years, then annually. This is in alignment with the follow up visit timeframe recommended by NCCN. Up to 30% of individuals develop metastatic disease to the

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brain. Screening for brain metastases is supported to allow early treatment of brain metastases prior to potentially impairing neurologic symptoms. MRI is preferred over CT for its increased sensitivity and specificity, at an interval of every 3 months for 1 year then every 6 months indefinitely.[1,7,9] PET/CT is not supported for surveillance due to excessive radiation exposure, false positive incidental findings, and financial toxicity.

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Non-Small Cell Lung Cancer (ONC-8)

Guideline

Non-Small Cell Lung Cancer – General Considerations (ONC-8.0)
Non-Small Cell Lung Cancer – Asymptomatic Screening (ONC-8.1)
Non-Small Cell Lung Cancer – Suspected/Diagnosis (ONC-8.2)
Non-Small Cell Lung Cancer – Initial Work-up/Staging (ONC-8.3)
Non-Small Cell Lung Cancer – Restaging/Recurrence (ONC-8.4)
Non-Small Cell Lung Cancer – Surveillance/Follow-up (ONC-8.5)
References (ONC-8)

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Non-Small Cell Lung Cancer – General Considerations (ONC-8.0)

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- Non-small cell lung cancer includes adenocarcinoma, squamous cell carcinoma, adenosquamous and large cell tumors.
- See: **Bronchopulmonary or Thymic Carcinoid – Initial Staging (ONC-15.6)** for evaluation of low-grade neuroendocrine tumors (carcinoid) of the lung.
- See: **Small Cell Lung Cancer (ONC-7)** for evaluation of high-grade small cell and large cell neuroendocrine tumors of the lung.
- PET/CT may be considered to confirm solitary focus of extra-pulmonary metastatic disease (i.e., brain or adrenal) if the individual is being considered for an aggressive treatment for oligometastatic disease.

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Non-Small Cell Lung Cancer – Asymptomatic Screening (ONC-8.1)

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- See: **Lung Cancer Screening (CH-33)** in the Chest Imaging Guidelines for criteria for Low-dose CT Chest for lung cancer screening.

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Non-Small Cell Lung Cancer – Suspected/Diagnosis (ONC-8.2)

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Indication	Medically Necessary Imaging Study
Abnormal chest x-ray or clinical suspicion remains high despite a normal chest x-ray in symptomatic individual	• CT Chest without contrast (CPT® 71250) **or** • CT Chest with contrast (CPT® 71260)
Pulmonary nodule <8 mm in size noted on CT Chest	• See: **Incidental Pulmonary Nodules Detected on CT Images (CH-16.2)** in the Chest Imaging Guidelines
Pulmonary nodule 8 mm (0.8 cm) to 30 mm (3 cm) seen on CT Chest or MRI Chest	• PET/CT (CPT® 78815) • If PET is Positive: Qualifies as initial staging PET/CT
Pulmonary mass 31 mm (3.1 cm) or greater seen on CT or MRI	• PET/CT (CPT® 78815) prior to biopsy if ONE or MORE of the following applies: ◦ Definitive treatment with resection or radiation will be utilized instead of biopsy if PET confirms limited disease ◦ Multiple possible biopsy options are present within the chest and PET findings will be used to determine the most favorable biopsy site • Biopsy is indicated prior to PET imaging for all other indications in pulmonary masses ≥31 mm (3.1 cm) in size
Mediastinal/Hilar Lymphadenopathy	See: **Mediastinal Lymphadenopathy (CH-2.3)** in the Chest Imaging Guidelines
Mediastinal/Hilar Mass	See: **Mediastinal Mass (CH-20)** in the Chest Imaging Guidelines
Paraneoplastic syndrome suspected	See: **Paraneoplastic Syndromes (ONC-30.3)**

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Evidence Discussion

For individuals with suspected lung cancer and an abnormal chest x-ray or a high suspicion for lung cancer with symptoms of lung cancer, a CT Chest is indicated, with or without contrast. If a PET/CT is performed in the workup of a pulmonary nodule and is positive, it qualifies as the initial staging PET. The radiotracer supported for PET/CT for lung cancer is 18-FDG.[3,14] Lesions 31mm or greater are considered masses rather than nodules.*3* There is no clear evidence for PET/CT over biopsy in this case. Generally, masses should be biopsied rather than re-imaged. PET/CT is supported if definitive treatment with resection or radiation will be utilized instead of biopsy (if PET confirms limited disease), or if multiple biopsy sites are present within the chest and PET findings will be used to determine the most favorable biopsy site. This maximizes safety when making decisions regarding invasive procedures.

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Non-Small Cell Lung Cancer – Initial Work-up/Staging (ONC-8.3)

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Indication	Medically Necessary Imaging Study
All individuals	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen with contrast (CPT® 74160) ◦ CT Abdomen can be omitted if CT Chest report clearly documents upper abdomen through level of adrenals • PET/CT (CPT® 78815) (if not already completed prior to histological diagnosis) • Bone scan (CPT® 78306, if PET/CT not being done) See also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes
Stage IB-IV disease	• MRI Brain without and with contrast (CPT® 70553) ◦ If MRI Brain is contraindicated or not available, CT Head with contrast (CPT® 70460) or CT Head without and with contrast (CPT® 70470)
Superior sulcus (Pancoast) tumor suspected	ANY or ALL of the following: • MRI Chest without and with contrast (CPT® 71552) • MRI Cervical Spine without and with contrast (CPT® 72156) • MRI Thoracic Spine without and with contrast (CPT® 72157)

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Evidence Discussion

CT Chest and upper abdomen to the level of the adrenals is supported for initial staging, as liver and adrenal metastatic disease are common in NSCLC. Pelvic disease is rare and imaging of the pelvis without pelvic symptoms is not recommended by NCCN.[1] FDG PET-CT is supported by NCCN in all individuals.[1]

Over 10% of individuals with stage III or IV disease present with metastatic disease to the brain, and 4% to 5% of individuals with stage II disease. MRI Brain with and without contrast has a higher detection rate for metastatic disease to the brain than CT, and is indicated in all individuals with stage II-IV disease.[1] For individuals with stage IB disease, MRI Brain with and without contrast is optional in this setting but can be considered to prevent under-staging and under-treatment, since a small number of individuals with apparent stage I disease and no CNS symptoms will have occult brain lesions and will require additional therapies.[1,14]

For individuals with superior sulcus (Pancoast) tumor, MRI Chest and MRI Cervical and Thoracic Spine with and without contrast have higher specificity for chest wall invasion, neurologic involvement, and fibrosis than CT alone, and are supported in addition to the imaging stated above.[1,13]

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Non-Small Cell Lung Cancer – Restaging/Recurrence (ONC-8.4)

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Indication	Medically Necessary Imaging Study
Stage I or II individuals who undergo definitive local treatment with surgery, radiation, or radiosurgery	• Restaging imaging is not medically necessary. See: **Surveillance/Follow-up (ONC-8.5)**
Measurable disease, undergoing active treatment	ANY or ALL of the following every 2 cycles: • CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) • CT Abdomen with contrast (CPT® 74160) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for known pelvic disease or pelvic symptoms • MRI Brain without and with contrast (CPT® 70553) for measurable brain metastases being treated with systemic therapy
ANY of the following: • After neoadjuvant treatment for evaluation of surgical resectability • Prior to starting adjuvant therapy • Inadequately resected disease	• CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250)
Suspected recurrence	ANY or ALL of the following: • CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) • CT Abdomen with contrast (CPT® 74160) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for known pelvic disease or pelvic symptoms

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Indication	Medically Necessary Imaging Study
Newly identified lung nodule(s)	• See: **Lung Metastases (ONC-31.1)** for new nodule evaluation
ANY of the following: • Suspected/biopsy proven recurrence localized to the chest cavity • Inconclusive findings conventional imaging • To differentiate tumor from radiation scar/fibrosis • Stage IV with oligometastatic disease on conventional imaging and individual is a candidate for aggressive surgical resection or other localized treatment of metastases with a curative intent	• PET/CT (CPT® 78815)
ANY of the following: • Following a demonstrated adequate response to neoadjuvant therapy if intracranial disease will preclude surgery • Documented recurrence/progression • New or worsening neurological signs or symptoms	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

In alignment with NCCN, CT Chest and Abdomen with contrast are supported every two cycles, with pelvic imaging only for a history of pelvic disease or new pelvic symptoms. These are also supported at any time for clinically suspected recurrence. MRI Brain with and without contrast is supported every 2 cycles for individuals with known brain metastases being treated with systemic therapy, or at any time for individuals with new neurologic symptoms or documented systemic progression.[1] An additional CT chest is supported if requested after neoadjuvant therapy to evaluate for resectability, in the interest of safe resection. CT is also supported post-operatively to assess baseline prior to starting adjuvant therapy, in alignment with NCCN.[1]

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Non-Small Cell Lung Cancer – Surveillance/Follow-up (ONC-8.5)

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Indication	Medically Necessary Imaging Study
Stage I-II (treatment including surgery and/ or chemotherapy, but no radiation)	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) every 6 months for 3 years and then annually
Stage I-II treated with radiation therapy Stage III-IV (metastatic sites treated with definitive intent)	• CT Chest with contrast (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) every 3-6 months for 3 years, every 6 months for 2 years and then annually
New lung nodule	• See: **Lung Metastases (ONC-31.1)**

Evidence Discussion

CT Chest with or without contrast is recommended by national guidelines every 6 months for 3 years and then annually for individuals with stage I or II disease. To prevent under-treatment, individuals treated with radiation who have residual abnormalities on imaging may undergo more frequent imaging every 3-6 months for the first year then every 6 months for 2 years, then annually thereafter. Individuals with stage II disease or definitively treated metastatic disease are at higher risk for relapse particularly in the first 2 years, and NCCN recommended CT Chest every 3-6 months for 3 years, every 6 months for 2 years, then annually. Asymptomatic abdominal and pelvic imaging exposes to radiation with low-yield for metastatic disease detection and is not supported. FDG-PET is not supported for surveillance due to excessive false positive rates, radiation exposure, and increased risk of unnecessary procedures for incidental false positive findings. MRI Brain for asymptomatic surveillance is low yield in asymptomatic NSCLC surveillance and is not routinely recommended.[1,2]

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Esophageal and GE Junction Cancer (ONC-9)

Guideline

Esophageal and GE Junction Cancer – General Considerations (ONC-9.0)
Esophageal and GE Junction Cancer – Suspected/Diagnosis (ONC-9.1)
Esophageal and GE Junction Cancer – Initial Work-up/Staging (ONC-9.2)
Esophageal and GE Junction Cancer – Restaging/Recurrence (ONC-9.3)
Esophageal and GE Junction Cancer – Surveillance/Follow-up (ONC-9.4)
References (ONC-9)

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Esophageal and GE Junction Cancer – General Considerations (ONC-9.0)

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- Imaging for esophageal cancer is determined by cell type and in which third of the esophagus it occurs.
- Cancers of the upper and middle third are usually squamous cell and are highly associated with tobacco and alcohol abuse.
- Cancers of the gastroesophageal (GE) junction are treated as lower third cancers. Lower third cancers are usually adenocarcinomas; 62% of these arise in the setting of Barrett's esophagus, a condition associated with high body mass index (BMI).

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Esophageal and GE Junction Cancer – Suspected/Diagnosis (ONC-9.1)

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- See: **Dysphagia and Upper Digestive Tract Disorders (NECK-3.1)** in the Neck Imaging Guidelines for evaluation of suspected esophageal malignancy.

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Esophageal and GE Junction Cancer – Initial Work-up/Staging (ONC-9.2)

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Indication	Medically Necessary Imaging Study
Biopsy proven	• CT Chest (CPT® 71260) and CT Abdomen (CPT® 74160) with contrast ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) instead of CT Abdomen if there are pelvic signs or symptoms
In addition to the above, for any of the following: • Upper 1/3 of esophagus • Neck mass	• CT Neck with contrast (CPT® 70491)
If no evidence of metastatic disease on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

- Upon initial diagnosis of cancer, CT Chest/Abdomen is recommended with the addition of pelvis if there are signs/symptoms of disease. NCCN stated that "CT can be used to determine the location of the primary tumor and its proximity to other structures".[1]
- Cancers diagnosed in the upper 1/3 of the esophagus should also obtain CT of the neck due to concern for nodal spread.
- If CT imaging does not show evidence of metastatic disease, PET/CT is indicated to assess for occult metastases to help finalize treatment options (curative versus palliative). PET/CT is not sensitive for locoregional nodal assessment as often these nodes are obscured by metabolic activity in the primary tumor but is more sensitive than CT for detecting distant metastases.
- NCCN also stated that PET/CT has "limited ability to ability to differentiate between cT1, cT2, and cT3 tumors. Therefore, CT should be performed as part of initial workup (as well as pelvic CT scan with contrast if clinically indicated) while FDG-PET/CT should be reserved for individuals with no evidence of M1 disease".[1]

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Esophageal and GE Junction Cancer – Restaging/Recurrence (ONC-9.3)

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Indication	Medically Necessary Imaging Study
After primary chemoradiation therapy prior to surgery	Any ONE of the following, not both: • PET/CT (CPT® 78815) no sooner than 5 weeks post completion of radiation therapy OR • CT Chest (CPT® 71260) and CT Abdomen (CPT® 74160) with contrast ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) instead of CT Abdomen if there are pelvic signs or symptoms or known pelvic disease
Post-surgical resection	• See: **Surveillance/Follow-up (ONC-9.4)**
Monitoring response to chemotherapy for stage IV/ metastatic disease	Every 2 cycles of treatment (~every 6-8 weeks): • CT Abdomen with contrast (CPT® 74160) • CT Chest with contrast (CPT® 71260)
• If conventional imaging is inconclusive **or** • Salvage surgical candidate with recurrence and no metastatic disease documented by conventional imaging	• PET/CT (CPT® 78815)
For ANY of the following: • Signs or symptoms of recurrence • Biopsy proven on follow-up endoscopy • Recurrence suggested by other imaging (i.e. chest x-ray or barium swallow)	• CT Chest (CPT® 71260) and CT Abdomen (CPT® 74160) with contrast

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Indication	Medically Necessary Imaging Study
If previously involved or new signs or symptoms	• CT Pelvis with contrast (CPT® 72193) and/or CT Neck with contrast (CPT® 70491)

Evidence Discussion

- Primary treatment typically involves chemoradiotherapy alone, surgery alone or both.
- After primary chemoradiation has been completed and prior to surgery, one of the following is recommended: CT Chest/Abdomen or PET/CT, with the latter ideally no sooner than 5 weeks after completion of radiation to minimize risk of false positives.
- Post-surgical resection is handled as disease surveillance.
- For Stage IV disease on chemotherapy, CT Chest/Abdomen indicated every 2 cycles.
- If conventional imaging is inconclusive or the member is a candidate for salvage surgery upon recurrence with no evidence of metastatic disease, PET/CT is indicated.
- CT Chest/Abdomen for signs/symptoms of recurrence, biopsy proven recurrence on follow-up endoscopy and recurrence suggested by other imaging.
- CT imaging of any appropriate area (e.g. neck, pelvis) if new signs/symptoms or known previous involvement.[1]

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Esophageal and GE Junction Cancer – Surveillance/Follow-up (ONC-9.4)

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Indication	Medically Necessary Imaging Study
Stage 0-IA (Tis, T1a) disease	• No routine advanced imaging
Stage IB (T1b)-III disease	• CT Chest (CPT® 71260) and CT Abdomen (CPT® 74160) with contrast every 6 months for 2 years and then annually for 3 more years
Stage IV disease	• See: **Phases of Oncology Imaging and General Phase-Related Considerations (ONC-1.2)**

Evidence Discussion

- Stage 0-IA (Tis, T1a): No routine advanced imaging indicated. Fully treated Tis and T1aN0 disease have prognoses that approximate a non-cancer cohort.[1]
- Stage IB (T1b): CT Chest/Abdomen with contrast annually for 3 years. T1b does not perform as well as fully treated Tis and T1aN0 disease, thus supporting current recommendations.[1]
- Stage II-III: CT Chest/Abdomen every 6 months for 2 years then annually for 3 years.[1]
- Stage IV: CT Chest/Abdomen (additional sites as clinically indicated) every 3 months while on maintenance therapy or every 3 months up to 5 years if being monitored off therapy.[1]

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Other Thoracic Tumors (ONC-10)

Guideline

Malignant Pleural Mesothelioma – Suspected/Diagnosis (ONC-10.1)
Malignant Pleural Mesothelioma – Initial Work-up/Staging (ONC-10.2)
Malignant Pleural Mesothelioma – Restaging (ONC-10.3)
Malignant Pleural Mesothelioma – Surveillance (ONC-10.4)
Thymoma and Thymic Carcinoma – Suspected/Diagnosis (ONC-10.5)
Thymoma and Thymic Carcinoma – Initial Work-up/Staging (ONC-10.6)
Thymoma and Thymic Carcinoma – Restaging (ONC-10.7)
Thymoma and Thymic Carcinoma – Surveillance (ONC-10.8)
References (ONC-10)

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Malignant Pleural Mesothelioma – Suspected/Diagnosis (ONC-10.1)

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- See: **Asbestos Exposure (CH-9.1)** in the Chest Imaging Guidelines for evaluation of suspected mesothelioma.

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Malignant Pleural Mesothelioma – Initial Work-up/Staging (ONC-10.2)

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Indication	Medically Necessary Imaging Study
Cytologically or pathologically proven	• CT Chest (CPT® 71260) **and** CT Abdomen (CPT® 74160) with contrast ◦ CT Abdomen and Pelvis with contrast (CPT® 74177)instead of CT Abdomen if there are pelvic signs or symptoms
For EITHER of the following: • No evidence of distant metastatic disease on CT imaging • Inconclusive conventional imaging	• PET/CT (CPT® 78815)
Suspicion of possible chest wall, spinal, diaphragmatic, or vascular involvement based on CT imaging	• MRI Chest without and with contrast (CPT® 71552)

Evidence Discussion

Contrasted CT Chest and CT Abdomen are supported for initial staging once mesothelioma is proven with lung fluid cytology or tissue biopsy. Contrast allows better evaluation of nodal disease in addition to parenchymal disease and offers improved characterization of direct extrapulmonary tumor invasion. MRI is inferior for parenchymal lung imaging thus CT is essential. Anatomic soft tissue detail, differentiation from progressive benign fibrosis, and brachiocephalic vascular involvement may be better demonstrated on MRI without and with contrast, so this study is supported for suspected chest wall, spine, diaphragm, or vascular involvement. The most common site of metastatic disease outside the chest is the liver, so CT Abdomen is an important part of

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the initial workup. However, pelvic disease is rare and pelvic CT exposes to additional radiation and is low yield in individuals without pelvic signs and symptoms. Pelvis may be added to contrasted CT in individuals with signs and symptoms of pelvic involvement, including direct abdominoperitoneal invasion. National Cancer Database review of over 40,000 individuals treated between 2004 and 2020 reveals that 50% of individuals are metastatic upon presentation.[17] Signs and symptoms of metastatic disease in body areas not addressed in ONC-10 may be imaged according to their respective sections in ONC-31, for which separate evidence summaries are provided.

PET/CT is not first line imaging for mesothelioma as it is inadequate for differentiating benign vs malignant changes in exposure-related progressive massive pulmonary fibrosis, but of which take up FDG in unpredictable fashions. Understaging of the primary site is common with PET/CT alone. Conventional imaging is essential. However, PET/CT is supported to confirm the absence of metastatic disease on conventional imaging (negative or inconclusive) prior to resection, as up to 29% of individuals initially identified as operable may be reclassified as inoperable due to identification of distant metastatic disease on PET/CT during pre-operative evaluation.[1]

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Malignant Pleural Mesothelioma – Restaging (ONC-10.3)

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Indication	Medically Necessary Imaging Study
Signs or symptoms of recurrence	• CT Chest (CPT® 71260) **and** CT Abdomen (CPT® 74160) with contrast ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) instead of CT Abdomen if there are pelvic signs or symptoms
Treatment with chemotherapy	Every 2 cycles: • CT Chest (CPT® 71260) **and** CT Abdomen (CPT® 74160) with contrast ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) instead of CT Abdomen if there are pelvic signs or symptoms
Following induction chemotherapy prior to surgical resection	• CT Chest (CPT® 71260) **and** CT Abdomen (CPT® 74160) with contrast ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) instead of CT Abdomen if there are pelvic signs or symptoms • PET/CT (CPT® 78815) if no evidence of distant metastatic disease
Inconclusive CT Chest	• MRI Chest without and with contrast (CPT® 71552)

Evidence Discussion

For individuals on chemotherapy, contrasted CT of chest and abdomen are supported every 2 cycles. Pelvic imaging may be added for signs and symptoms of peritoneal/ pelvic disease or known pelvic/peritoneal disease as noted in the initial staging section. The same logic applies to any individuals with signs and symptoms of recurrence. For individuals receiving induction chemotherapy, CT Chest and Abdomen (with pelvis if previously reviewed indications are present) are supported at end of induction. Response is categorized by validated mRECIST criteria on CT. If there is no metastatic

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disease, this may again be confirmed with PET/CT prior to attempted resection, ensuring the patient is not subjected to futile invasive surgery when RT or further systemic therapy may be more appropriate. If CT Chest is inconclusive, MRI Chest without and with contrast is supported. MRI Chest may differentiate between treatment-related changes (fibrosis) and persistent mesothelioma, which is not well-differentiated on PET/CT as both may have unpredictable FDG uptake.[1,10-13,17]

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Malignant Pleural Mesothelioma – Surveillance (ONC-10.4)

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Indication	Medically Necessary Imaging Study
All	• CT Chest with contrast (CPT® 71260) and previously involved regions every 3 months for 2 years, then annually thereafter

Evidence Discussion

There is no clear consensus for surveillance imaging for malignant pleural mesothelioma, and NCCN offered no guidance on this topic. The European Society of Medical Oncology (ESMO) advised CT for surveillance without a specific timeframe.[13] Given the known value of CT in assessing primary mesothelioma and abdomino/peritoneal metastatic disease, these guidelines support CT for surveillance. The timeframe is based on National Cancer Database survival statistics for malignant pleural mesothelioma.[2] In review of 40,000+ individuals treated between 2004 and 2020, individuals undergoing surgery had a median survival time for 19.8 months, compared with 7.9 months in those who had not undergone surgery. The 2-year survival for those who underwent surgery was 44%, with 18% 2-year survival in unresectable disease. 5-year survival is 5% in unresected disease, and 16% in those who underwent surgery. These guidelines support CT Chest and Abdomen with contrast every 3 months for the first 2 years, then annually. As noted in the restaging section, CT imaging is always supported for new signs and symptoms.[1]

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Thymoma and Thymic Carcinoma – Suspected/Diagnosis (ONC-10.5)

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- The proceeding guidelines are intended to be used for imaging of malignant mediastinal masses.
- See: **Mediastinal Mass (CH-20.1)** in the Chest Imaging Guidelines for evaluation of mediastinal mass without confirmation of malignancy.
- See: **Bronchopulmonary or Thymic Carcinoid – Initial Staging (ONC-15.6)** for imaging guidelines for thymic carcinoid.

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Thymoma and Thymic Carcinoma – Initial Work-up/Staging (ONC-10.6)

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Indication	Medically Necessary Imaging Study
Encapsulated or invasive limited disease	• CT Chest with contrast (CPT® 71260)
Extensive mediastinal involvement on CT Chest	• CT Abdomen with contrast (CPT® 74160) • CT Neck with contrast (CPT® 70491)
Inconclusive finding on CT	ONE of the following: • PET/CT (CPT® 78815) • MRI Chest without and with contrast (CPT® 71552)
Preoperative planning	• MRI Chest without and with contrast (CPT® 71552)
Thymic Carcinomas	• Image according to **Non-Small Cell Lung Cancer - Initial Work-up/Staging (ONC-8.3)**

Evidence Discussion

Thymomas and thymic carcinomas originate in the thymus and are epithelial tumors. Thymomas are rare tumors (though most common primary tumor of anterior mediastinum) that typically spread locally with 5-year survival rates of 90% while thymic carcinomas are very rare, more invasive and often present with metastases with 5-year survival rates of 55%.[3] Initial imaging for thymoma includes CT Chest with contrast that usually shows a well-defined rounded or oval mass without adenopathy. If there is extensive mediastinal involvement, CT Neck/Abdomen with contrast can be performed. If CT imaging is inconclusive, PET/CT or MRI Chest with and without contrast may be indicated, with MRI preferred in thymic carcinoma.[14,15] For preoperative planning, MRI Chest is also indicated.

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Thymoma and Thymic Carcinoma – Restaging (ONC-10.7)

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Indication	Medically Necessary Imaging Study
Adjuvant therapy following surgical resection	• Follow surveillance imaging
Following induction chemotherapy prior to surgical resection, if no evidence of metastatic disease	ONE of the following: • PET/CT (CPT® 78815) • CT Chest with contrast (CPT® 71260) • MRI Chest without and with contrast (CPT® 71552)
For suspected recurrence	• CT Chest with contrast (CPT® 71260)
Recurrence with extensive mediastinal involvement on CT Chest	• CT Abdomen with contrast (CPT® 74160) • CT Neck with contrast (CPT® 70491)
Inconclusive finding on CT	ONE of the following: • PET/CT (CPT® 78815) • MRI Chest without and with contrast (CPT® 71552)
Metastatic disease on chemotherapy	• CT Neck (CPT® 70491), CT Chest (CPT® 71260), and CT Abdomen (CPT® 74160) with contrast, every 2 cycles of therapy
Thymic carcinomas	• See: **Non-Small Cell Lung Cancer Restaging/ Recurrence (ONC-8.4)**

Evidence Discussion

If induction chemotherapy is given, PET/CT can be obtained prior to surgical resection as studies have shown a correlation of radiographic response to pathologic response to help guide resectability.[16] Chest imaging with CT or MRI are also accepted imaging choices for this purpose. For recurrence, CT Chest with CT Neck/Abdomen as clinically

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indicated. PET/CT or MRI Chest indicated if CT Chest is inconclusive. For individuals on chemotherapy for metastatic disease, CT Neck/Chest/Abdomen with contrast can be given every 2 cycles.[3]

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Thymoma and Thymic Carcinoma – Surveillance (ONC-10.8)

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Indication	Medically Necessary Imaging Study
Thymoma, R0 resection	• CT Chest with contrast (CPT® 71260) and previously involved regions every 6 months for 2 years, then annually until year 10
Thymoma, R1 or R2 resection	• CT Chest with contrast (CPT® 71260) and previously involved regions every 6 months for 2 years, then annually until year 10
Thymic carcinoma, R0 resection	• CT Chest with contrast (CPT® 71260) every 6 months for 2 years and then annually until year 5
Thymic carcinoma, R1 or R2 resection	• CT Chest with contrast (CPT® 71260) and previously involved regions every 3 months for 2 years, then annually until year 5

Evidence Discussion

Thymoma surveillance is stratified into resection categories by NCCN. Thymoma surveillance should be with CT Chest with contrast and any previously involved areas for 10 years due to the risk of late recurrence.[3] Thymic carcinoma, like thymoma, management depends on the extent of resection. NCCN stated "patients who have an R0 resection have a 5-year survival of approximately 60%".[3] Thymic carcinoma surveillance recommendations from NCCN are based on consensus as there is a paucity of published studies specifically addressing surveillance imaging of thymic carcinoma. The recommendation includes imaging for 5 years.[3]

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Breast Cancer (ONC-11)

Guideline

Breast Cancer – General Considerations (ONC-11.0)
Breast Cancer – Suspected/Diagnosis (ONC-11.1)
Breast Cancer – Initial Work-up/Staging (ONC-11.2)
Breast Cancer – Restaging/Recurrence (ONC-11.3)
Breast Cancer – Surveillance/Follow-up (ONC-11.4)
References (ONC-11)

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Breast Cancer – General Considerations (ONC-11.0)

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- This guideline applies to invasive carcinoma and ductal carcinoma in situ histologies of breast cancer.
- Advanced imaging to evaluate for distant metastases is not indicated for asymptomatic individuals with invasive carcinoma or ductal carcinoma in situ (DCIS).
- High-risk lesions, such as LCIS, ADH, and ALH are discussed in **Breast MRI Indications (BR-5.1)**.
- Bone scan has a high concordance rate with PET for detecting bone metastases.
- Scintimammography and Breast Specific Gamma Imaging (BSGI) are considered experimental, investigational, or unproven.

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Breast Cancer – Suspected/Diagnosis (ONC-11.1)

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- See: **Breast MRI Indications (BR-5.1)** in the Breast Imaging Guidelines for evaluation of suspected breast cancer.

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Breast Cancer – Initial Work-up/Staging (ONC-11.2)

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Indication	Medically Necessary Imaging Study
Newly diagnosed breast cancer or ductal carcinoma in situ	• Diagnostic bilateral mammogram and/or Ultrasound Breast (CPT® 76641 or CPT® 76642) are imaging modalities of choice • MRI Breast bilateral without and with contrast (CPT® 77049)
ANY of the following: • Atypical ductal hyperplasia (ADH) • Atypical lobular hyperplasia (ALH) • Lobular carcinoma in situ (LCIS)	See: **Breast MRI Indications (BR-5.1)**
ANY of the following: • Ductal carcinoma in situ • Stage I-III	• For sentinel lymph node evaluation: Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
Stages I, II, and operable stage III (N1 disease)	• Routine systemic imaging is not medically necessary for initial staging of non-metastatic breast cancer in the absence of signs or symptoms

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Indication	Medically Necessary Imaging Study
ANY of the following: • Clinically suspected or biopsy-proven distant metastases/Stage IV disease (not locally advanced disease with three or less positive axillary nodes) • Signs or symptoms of systemic disease • Elevated liver function tests or tumor markers • Inflammatory breast cancer (stage T4d) • 4 or more axillary lymph nodes positive for cancer involvement (i.e., N2 disease)	ANY or ALL of the following: • CT Chest (CPT® 71260) **and** CT Abdomen and Pelvis (CPT® 74177) with contrast • Bone scan (CPT® 78306) (if PET/CT is not being performed) See: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes
ANY of the following: • Stage III N2 disease • T4 disease • Inflammatory breast cancer (stage T4d) • Inconclusive CT and/or bone scan	In addition to the above: • FDG PET/CT (CPT® 78815)
Bone pain	• Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)**) for additional bone scan codes • PET/CT (CPT® 78815) with Sodium Fluoride radiotracer if CT, MRI, Bone scan and FDG PET/CT scan are inconclusive for bone metastases • See: **Bone (Non-Vertebral) Metastases (ONC-31.5)** • See: **Spinal/Vertebral Metastases (ONC-31.6)**

Evidence Discussion

Evaluation of disease in the breast/axilla with breast MRI:

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National Comprehensive Cancer Network (NCCN) guidelines stated that breast MRI "may be used for staging evaluation to define extent of cancer or presence of multifocal or multicentric cancer in the ipsilateral breast, or to screen the contralateral breast at the time of initial diagnosis".[1] Disagreement continues to exist as to whether the benefit of breast MRI in initial staging outweighs the drawbacks. Detractors cite evidence of increased time-to-surgery, high false-positive rates among the reasons to avoid universal usage of breast MRI in initial staging.[12,32] The American Society of Breast Surgeons Consensus (ASBrS) statement recommended against the use of routine MRI in the preoperative workup of individuals with breast cancer.[11] MRI may be helpful in elucidating whether disease exists in the intervening breast tissue thereby aiding in tumor size estimation and the decision to opt for mastectomy vs. lumpectomy. NCCN cautioned that "surgical decisions should not be based solely on MRI findings".[1] Breast MRI has been shown to find occult primary cancers in roughly two thirds of patients who present with positive axillary lymph nodes, allowing for definitive surgical management.[31] NCCN stated that breast MRI "may be helpful for evaluation before and after preoperative systemic therapy to define extent of disease, response to treatment, and potential for breast-conservation therapy".[1] Breast MRI has also been shown to more accurately predict tumor size in individuals with invasive lobular carcinoma than conventional imaging.[36] While data are limited on the use of breast MRI in the setting of Paget's disease of the breast, one study found that for individuals with a histologic diagnosis of Paget's disease and a negative mammogram, MRI was able to detect occult cancer in 4/8 (50%) individuals.[34] NCCN guidelines noted that there is considerable controversy in the use of MRI based on breast density.[1] They noted that MRI advocates argue that it has a high sensitivity to find occult disease in "dense breasts where mammographically occult disease is more likely to elude preoperative detection"; however, MRI detractors note the high percentage of false- positive findings, resulting in further workup, overestimation of extent of disease and increased frequency of mastectomy. Given the recommendation by NCCN that breast MRI may be useful for specific circumstances in initial staging, these guidelines allow for the approval of breast MRI in initial staging to empower individuals and providers to discuss the risks and benefits of this modality and move forward with the most advantageous choice for the individual's specific circumstances.

Lymphatic mapping with lymphoscintigraphy and/or SPECT/CT:

Sentinel node biopsy is important in the staging of individuals with breast cancer. However, lymphoscintigraphy has limited utility in this setting.[21] In individuals with recurrent disease who have had previous axillary surgery, lymphoscintigraphy with SPECT/CT may be helpful in delineating alternate drainage pathways.[20] In individuals who have had a positive axillary node prior to neoadjuvant chemotherapy, some authors have also found the technique to be helpful in identifying the previously positive clipped node, which may not be subsequently identified as a sentinel node.[23]

Systemic staging with CT Chest, Abdomen, Pelvis, and bone scan vs. PET/CT:

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The NCCN guidelines stated to "consider additional imaging studies only in the presence of signs and symptoms of metastatic disease and for patients who are clinically high risk".[1] This is in keeping with the ACR Appropriateness Criteria which stated that systemic staging is "usually not appropriate" for all newly diagnosed clinical Stage I-IIA (early stage) breast cancer, and clinical Stage IIB-III (late stage) with ER +/HER2- breast cancer .[37] Over a third of individuals with inflammatory breast cancer will have distant metastatic disease at presentation and NCCN recommended staging studies in these individuals.[1,33]

NCCN guidelines recommended CT scan for the work up for distant metastatic disease.[1] NCCN also stated that the "routine use of FDG PET/CT is not recommended in the staging of clinical stage I, II, or operable III (T3,N1) breast cancer.[1] However, NCCN indicated that PET/CT may be performed at the same time as diagnostic CT and can be helpful as a problem-solving tool when conventional imaging is suspicious or inconclusive. It is also noted to be "helpful in identifying unsuspected regional nodal disease and/or distant metastases when used in addition to standard staging studies".[1] Bone imaging is not recommended if FDG PET/CT is already going to be performed.[1] Given the above, these guidelines allow for the selected use of concomitant PET/CT for initial staging in specific clinical circumstances.

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Breast Cancer – Restaging/Recurrence (ONC-11.3)

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- For imaging related to breast reconstruction, see: **Breast Reconstruction (BR-3.1)** in the Breast Imaging Guidelines

Indication	Medically Necessary Imaging Study
ANY of the following: • End of planned neoadjuvant chemotherapy to determine resectability • Biopsy proven local recurrence • Suspicion of recurrence with inconclusive mammogram and/or ultrasound (BIRADS 0) • Mammogram and ultrasound conflicts with physical exam	• MRI Breast Bilateral without and with contrast (CPT® 77049)
After neoadjuvant chemotherapy, if sentinel lymph node evaluation is planned	• Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
ANY of the following: • Assessing for residual disease after surgery • Assessing response to neoadjuvant chemotherapy • After lumpectomy or mastectomy, prior to adjuvant therapy	• Neither PET nor CT are medically necessary for systemic restaging after neoadjuvant chemotherapy or after surgery

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Indication	Medically Necessary Imaging Study
• Treatment response in individuals with metastatic disease and measurable disease on imaging ◦ For individuals receiving chemotherapy, imaging is indicated after every 2 cycles ◦ For individuals receiving hormonal or endocrine therapy, imaging is indicated every 3 months	• CT Chest with contrast (CPT® 71260); and CT Abdomen and Pelvis with contrast (CPT® 74177) • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) In addition to the above options, for individuals receiving systemic treatment for brain metastases: • MRI Brain without and with contrast (CPT® 70553)
ANY of the following: • Elevated LFTs • Elevated tumor markers • Signs or symptoms of recurrence • Biopsy proven recurrence	Any or all of the following: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • Bone scan (CPT® 78306) (See also: **Nuclear Medicine (NM) Imaging in Oncology) (ONC-1.3)** for additional bone scan codes) For individuals with prior history of brain metastases or current signs or symptoms of brain metastases: • MRI Brain without and with contrast (CPT® 70553)
• Inconclusive CT, MRI, and/or bone scan • Treatment response assessment for bone-only metastases (excluding brain metastases) and a prior bone scan has not been performed for serial comparison	• ^{18}F-FDG PET/CT (CPT® 78815)

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Indication	Medically Necessary Imaging Study
Individuals with known or suspected bone metastases AND inconclusive bone findings on ALL of the following: • CT, MRI, bone scan, and FDG PET/CT scan	• ^{18}F Sodium Fluoride PET/CT (CPT® 78815)
To determine the ER-status of suspected/known metastatic recurrence noted on CT/bone scan **and** any one of the following: • Biopsy of metastatic site is non-diagnostic/inconclusive • Biopsy of metastatic site is risky & cannot be performed (metastatic sites in the brain, spine or near vascular structures)	• ^{18}F-FES (fluoroestradiol) PET/CT scan (CPT® 78815 or CPT® 78816)

Evidence Discussion

Evaluation of disease in the breast/axilla with breast MRI:

Breast MRI has been shown to predict extent of pathological tumor response in the breast and lymph nodes after neoadjuvant systemic therapy better than conventional imaging, although may over- or under-estimate residual tumor size.[35]

While some authors have found the use of MRI to be helpful in terms of estimating size of ipsilateral breast tumor recurrences and finding multifocal or multicentric disease[27], others have found that the addition of MRI in this context did not significantly change management and increased time to definitive therapy.[26] However, as previously noted, MRI may be a useful adjunct in situations where conventional imaging is inconclusive.

Systemic staging with CT Chest, Abdomen, Pelvis, and bone scan vs. PET/CT:

As individuals with symptoms for distant metastatic disease would have had systemic staging prior to neoadjuvant chemotherapy and/or surgery, there is no indication to repeat these until treatment is completed. However, for individuals with metastatic disease, NCCN guidelines recommended CT Chest, Abdomen, and Pelvis with contrast every 2-4 cycles of chemotherapy or every 2-6 months of endocrine therapy and bone scan is recommended every 4-6 cycles of chemotherapy or every 2-6 months of endocrine therapy. Restaging using these modalities is also advised if there is concern for progression of disease. They noted that PET/CT is not routinely indicated for restaging "because of the absence of a reproducible, validated, and widely accepted set of standards for disease activity assessment". PET is instead preferred to be reserved

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as a problem-solving tool when conventional imaging is inconclusive. Additionally, NCCN noted sodium fluoride PET/CT may be helpful to detect bone metastases where other imaging is inconclusive.[1]

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Breast Cancer – Surveillance/Follow-up (ONC-11.4)

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Indication	Medically Necessary Imaging Study
Measurable metastatic disease on maintenance therapy or being monitored off therapy	ANY or ALL of the following, every 3 months for up to 5 years after completion of active treatment: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)
• Asymptomatic non-metastatic disease • Individuals receiving post-operative adjuvant therapy	• No advanced imaging
Individuals with a personal history of breast cancer (not treated with bilateral mastectomy)	• Annual MRI Breast Bilateral without and with contrast (CPT® 77049)

Evidence Discussion

Evaluation of disease in the breast/axilla with breast MRI:

NCCN guidelines suggested that "the utility of MRI in follow-up screening of most patients with prior breast cancer is undefined", but recommend annual MRI in individuals with a personal history of breast cancer who were either younger than age 50 or who have dense breasts.[1] ACR also addressed the use of MRI breast in individuals with dense breast tissue. They did not differentiate between heterogeneously and extremely dense breasts in the recommendation. ACR considered dense breasts to be heterogeneously dense (Category C) and extremely dense (Category D).[29] As individuals with breast cancer may have residual breast tissue in the ipsilateral or contralateral breast for which certain genetic mutations may increase the risk of subsequent cancers, breast MRI would also be indicated in such individuals.[1] Individuals with a clinical lifetime risk estimated to be ≥20% lifetime risk prior to their diagnosis of breast cancer and/or who had a history of ADH or lobular neoplasia would

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have been candidates for breast cancer screening with breast MRI regardless (see BR 5.1) and therefore, would equally be eligible for this screening modality after breast cancer treatment, as long as they had not had bilateral mastectomies. Individuals who have had bilateral mastectomies have little residual tissue, and therefore, surveillance with breast imaging would be of little value. A recent meta-analysis found that the rate of occult cancer in individuals with mastectomy and the rate at which MRI detected cancer in individuals after mastectomy was well below the current BIRADS benchmark for women with genetic predispositions to cancer.[25]

Systemic surveillance with CT Chest, Abdomen, Pelvis, and bone scan vs. PET/CT:

The NCCN guidelines stated "In the absence of clinical signs and symptoms suggestive of recurrent disease, there is no indication for laboratory or imaging studies for metastases screening".[1] This is in keeping with ASCO's Choosing Wisely guideline which similarly recommended against surveillance testing with biomarkers or imaging for asymptomatic breast cancer in individuals who have been treated with curative intent.[28] Several studies have shown no benefit from routine imaging which can result in unnecessary radiation exposure and biopsies, and lead to misdiagnosis and treatment related complications.[24] A recent study also found that more intensive screening for metastasis did not result in improved survival.[22]

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Sarcomas – Bone, Soft Tissue, and GIST (ONC-12)

Guideline

Bone and Soft Tissue Sarcomas – General Considerations (ONC-12.1)
Soft Tissue Sarcomas – Initial Work-up/Staging (ONC-12.2)
Soft Tissue Sarcomas – Restaging/Recurrence (ONC-12.3)
Soft Tissue Sarcomas Surveillance/Follow-up (ONC-12.4)
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Bone and Soft Tissue Sarcomas – General Considerations (ONC-12.1)

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- Sarcomas are tumors of mesenchymal origin, classified as high-, intermediate-, and low-grade (G) tumors (sometimes described as “spindle cell” cancers). They can arise in any bony, cartilaginous, smooth muscle, skeletal muscle, or cardiac muscle tissue.
- Malignant nerve sheath tumor cell types should be imaged as high-grade sarcoma.
- Sarcomas occur in both adult and pediatric individuals, but some are more common in one age group than the other. Unless specified below, individuals age ≥18 years old should be imaged according to this guideline section.
- Exceptions include:
  - Rhabdomyosarcoma in individuals of all ages should be imaged according to guidelines in **Rhabdomyosarcoma (RMS) (PEDONC-8.2)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
  - Osteogenic sarcoma (Osteosarcoma) in individuals of all ages should be imaged according to guidelines in **Osteogenic Sarcoma (OS) (PEDONC-9.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
  - Ewing sarcoma and Primitive Neuroectodermal Tumor in individuals of all ages should be imaged according to guidelines in **Ewing Sarcoma and Primitive Neuroectodermal Tumors (ESFT) (PEDONC-9.4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
  - Kaposi’s sarcoma in individuals of all ages should be imaged according to guidelines in **Kaposi’s Sarcoma (ONC-31.10)**.
  - See: **Uterine Cancer (ONC-22)** for imaging recommendations for uterine sarcoma.
  - Desmoplastic small round cell tumor in individuals of all ages should be imaged according to guidelines in **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

Evidence Discussion

The choice of imaging modality is driven by the primary tumor site. Cross sectional imaging of the primary site with MRI with and without contrast or CT with contrast is recommended for best illustration of anatomic detail and vascular and nodal involvement and provides flexibility for clinician discretion for choice of modality for all sites for maximum tumor definition with consideration of minimizing radiation exposure.[1,2] CT is superior for evaluation for metastatic disease of the lung, which is supported for all individuals with a newly diagnosed malignant sarcoma. Imaging both with and without

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contrast for evaluation of metastatic disease to the lung is not supported as this exposes individuals to higher radiation doses without significant clinical benefit.

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Soft Tissue Sarcomas – Initial Work-up/ Staging (ONC-12.2)

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Indication	Medically Necessary Imaging Study
All individuals	• CT Chest with (CPT® 71260) or CT Chest without (CPT® 71250) contrast • MRI without and with contrast of involved area ◦ CT of involved area if contraindication to MRI
ANY of the following: • Angiosarcoma • Alveolar soft part sarcoma • Clear cell sarcoma • Epithelioid sarcoma • Hemangiopericytoma • Leiomyosarcoma • Liposarcoma • Retroperitoneal or intra-abdominal primary site (including pelvic primary site) • Other histologies documented to have propensity for lymphatic spread and deep-seated tumors	In addition to the above, one of the following combinations: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • MRI Abdomen (CPT® 74183) with and without contrast and CT Pelvis (CPT® 72193) with contrast • CT Abdomen (CPT® 74160) with contrast and MRI Pelvis (CPT® 72197) without and with contrast
ANY of the following: • Myxoid round cell liposarcoma • Angiosarcoma • Alveolar soft part sarcoma • Cardiac sarcoma • Neurologic signs or symptoms raising suspicion of CNS metastases	In addition to the above: • MRI Brain without and with contrast (CPT® 70553) • MRI Cervical/Thoracic/Lumbar Spine without and with contrast (CPT® 72156, CPT® 72157, and CPT® 72158)

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Indication	Medically Necessary Imaging Study
Leiomyosarcoma	In addition to the above: • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) Imaging in Oncology [ONC-1.3]** for additional bone scan codes)
ANY of the following: • Grade of tumor in doubt following biopsy • Conventional imaging suggests solitary metastasis amenable to surgical resection • Inconclusive conventional imaging • Prior to planned neoadjuvant therapy	• PET/CT (CPT® 78815 or CPT® 78816)
Desmoid Tumors	ONE of the following: • CT without contrast or with contrast of the affected body part • MRI without contrast or without and with contrast of the affected body part • Imaging of lung, lymph node, and metastatic site for these tumors is not medically necessary
Dermatofibrosarcoma Protuberans (DFSP)	ONE of the following: • CT without contrast or with contrast of the affected body part • MRI without contrast or without and with contrast of the affected body part • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast for: ◦ Pulmonary symptoms ◦ Abnormal chest x-ray ◦ Sarcomatous differentiation

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Evidence Discussion

- PET/CT is supported where the grade of tumor is in doubt following biopsy or to confirm oligometastatic disease amenable to local treatment, to support treatment decision making. NCCN also supported the use of PET/CT for initial staging prior to planned neoadjuvant therapy. It is also a useful tool to follow inconclusive conventional imaging.[1]
- Different subtypes of soft tissue sarcomas have different patterns of spread; thus a histologic diagnosis is essential to determine imaging strategy.
- Abdominal and pelvic imaging is not supported for extremity, trunk or head and neck primary sites, unless documented histologies with propensity for lymphatic spread.[14] Due to the propensity of myxoid and round cell liposarcomas for leptomeningeal spread, initial evaluation of the spine with MRI is supported. MRI of the brain is supported for those with CNS signs and symptoms, and for all individuals with angiosaroma, alveolar soft part sarcomas, and cardiac sarcoma.[1]
- For desmoid tumors, disease biology and patterns of recurrence do not support metastatic disease workup, CT Chest or body areas outside of the primary site subject individuals to additional radiation and incidental finding risk.[7]
- For dermatofibrosarcoma protuberans (DFSP), CT Chest is supported for sarcomatous differentiation as noted for other sarcoma histologies above, or for pulmonary symptoms. In the absence of these features, CT exposes to risk with no statistically significant clinical benefit.[10,15]

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Soft Tissue Sarcomas – Restaging/ Recurrence (ONC-12.3)

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Indication	Medically Necessary Imaging Study
ANY of the following: • After preoperative radiotherapy • After surgical resection • After adjuvant radiotherapy	• MRI without and with contrast or CT with contrast of affected body area • Chest or lymph node imaging is not medically necessary if no abnormality on previous imaging
Baseline end of therapy evaluation	• Same studies as initial work-up
ANY of the following: • Differentiate tumor from radiation or surgical fibrosis • Determine response to neoadjuvant therapy • Confirm oligometastatic disease prior to curative intent surgical resection	• PET/CT (CPT® 78815 or CPT® 78816) ◦ If treated with radiation therapy, PET/CT no sooner than 12 weeks (3 months) post completion of radiation therapy
EITHER of the following: • Chemotherapy response for individuals with measurable disease • Individuals on neoadjuvant treatment without known lung involvement	• CT with contrast or MRI without and with contrast of affected body area every 2 cycles

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Indication	Medically Necessary Imaging Study
Unresectable or metastatic disease on chemotherapy	Every 2 cycles: • CT with contrast or MRI without and with contrast of affected body area AND • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)
Recurrence suspected	• Repeat all imaging for initial workup of specific histology and/or primary site and other symptomatic areas
Preoperative planning prior to resection	ANY or ALL of the following: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area
Dermatofibrosarcoma Protuberans (DFSP)	• CT without contrast or with contrast of the affected body part or MRI without contrast or without and with contrast of the affected body part • CT Chest with contrast (CPT® 71260) or CT Chest without (CPT® 71250) contrast for: ◦ Known prior thoracic disease ◦ New or worsening pulmonary symptoms ◦ New or worsening chest x-ray ◦ Sarcomatous differentiation

Evidence Discussion

- Local therapy is a cornerstone of treatment for sarcomas. Cross-sectional imaging of primary site, with modality used at diagnosis specific to tumor site, is supported after pre-operative radiotherapy, and after resection of adjuvant radiotherapy to determine response, as well as for pre-operative planning prior to resection and every two cycles of treatment during active therapy. NCCN supported imaging for a baseline at the end of therapy. In the absence of known lung involvement or pulmonary symptoms, restaging of lung on active treatment does not provide benefit in most histologies and exposes individuals to additional radiation and risk of incidental findings.[1]
- NCCN indicated that PET/CT may be useful for therapy response. Individuals with baseline tumor SUVmax ≥6 and <40% decrease in FDG avidity after neoadjuvant

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therapy are at high risk for disease recurrence. Pretreatment tumor SUVmax and change in SUV max after neoadjuvant therapy has been shown to identify individuals at high risk of tumor recurrence and may be used to identify individuals most likely to benefit from additional chemotherapy.[13] PET/CT is supported to assess neoadjuvant therapy response, to differentiate scarring from disease, or to confirm oligometastatic disease prior to resection.[1,13]

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Soft Tissue Sarcomas Surveillance/ Follow-up (ONC-12.4)

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Indication	Medically Necessary Imaging Study
For any of the following: • Retroperitoneal/intra-abdominal primary site (including pelvic primary site) • Angiosarcoma • Epithelioid sarcoma	ANY or ALL of the following every 3 months for 3 years, then every 6 months for 2 more years, then annually: • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast or MRI without and with contrast of any other involved body areas
Myxoid/round cell liposarcoma	ANY or ALL of the following every 3 months for 2 years, then every 6 months for 2 more years, then annually: • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast or MRI without and with contrast of any other involved body areas • MRI Cervical/Thoracic/Lumbar Spine without and with contrast (CPT® 72156, CPT® 72157, and CPT® 72158)
Low-grade/Stage I extremity or trunk, primary site	ANY or ALL of the following every 6 months for 2 years, then annually thereafter: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT with contrast, MRI without contrast, or MRI without and with contrast of primary site

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Indication	Medically Necessary Imaging Study
ANY of the following: • Extremity/trunk primary site - grade II/stage II or higher • Head/neck primary site	ANY or ALL of the following every 3 months for 2 years, then every 6 months for 2 more years, then annually: • CT with contrast, MRI without contrast, or MRI without and with contrast of primary site • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • CT with contrast or MRI without and with contrast of any other involved body areas
ANY of the following: • Angiosarcoma • Alveolar soft part sarcoma • Cardiac sarcoma	In addition to the above studies: • MRI Brain without and with contrast (CPT® 70553) annually For surveillance of individuals with known brain metastases, see: **Brain Metastases (ONC-31.3)**
Desmoid tumors	ONE of the following every 3 months for 3 years, then every 6-12 months: • CT without contrast or with contrast of the affected body part • MRI without contrast or without and with contrast of the affected body part
Dermatofibrosarcoma Protuberans	• No routine imaging unless clinical signs/symptoms of recurrence

Evidence Discussion

Time frames, modality, and body site for surveillance by histology are based on tumor recurrence patterns specific to primary site and tumor biology. PET/CT is not supported for asymptomatic surveillance, as this can lead to unnecessary radiation exposure and invasive procedures or excess treatment.[1,7,10]

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Gastrointestinal Stromal Tumor (GIST) (ONC-12.5)

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General Considerations

- GISTs are mesenchymal neoplasms of the gastrointestinal (GI) tract, mostly found in the stomach and upper small bowel, commonly metastasizing to the liver and abdominal cavity and primarily treated with surgery.
- Recurrence risk of GIST is estimated by prognostic model based on location, size of primary tumor, and mitotic rate per high power field (HPF).
- High-risk category includes any tumor >5 cm with >5 mitoses/50 HPF and any tumors >10 cm in size regardless of mitotic rate.

Indication	Medically Necessary Imaging Study
Suspected/Diagnosis	• CT Abdomen and Pelvis with contrast (CPT® 74177)
Initial Work-up/Staging	• CT Chest (CPT® 71260) with contrast or CT Chest without contrast (71250) and • CT Abdomen and Pelvis (CPT® 74177) with contrast, if not already performed
In addition to the above imaging for initial staging of either of the following: • Liver lesions noted on initial CT • Preoperative assessment of liver	• MRI Abdomen without and with contrast (CPT® 74183)
Monitoring response to treatment (every 8 to 12 weeks) in any of the following: • Unresectable primary disease • Metastatic disease • Resectable disease with significant morbidity	EITHER of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
Additional treatment response, in addition to the above imaging, for known or suspected chest involvement	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)
Evaluation of inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)
Surveillance for any of the following: • Incompletely resected • Metastatic disease • High-risk disease	Every 3 months for 5 years, then annually: • CT Abdomen and Pelvis with contrast (CPT® 74177)
Surveillance for completely resected, low-risk disease	Every 6 months for 5 years, then annually: • CT Abdomen and Pelvis with contrast (CPT® 74177)
Known or suspected recurrence	ANY or ALL of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) for known or suspected liver involvement • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for known or suspected chest involvement

Evidence Discussion

Suspected/Diagnosis and Initial Staging

CT Abdomen and Pelvis are recommended by NCCN for suspected GIST.[3]

NCCN stated that CT is the "imaging modality of choice to characterize an abdominal mass, as well as to evaluate its extent and the presence or absence of metastasis at the initial staging workup".[3] While CT Chest is supported to rule out metastatic disease in biopsy-proven GIST, advanced imaging prior to confirmation of diagnosis may expose individuals to unnecessary radiation and increased irrelevant incidental findings. MRI is an adjunct to CT to provide anatomical definition of liver metastases.[3] NCCN noted that PET/CT is not a substitute for diagnostic CT, which has superior sensitivity for this tumor

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site, but PET/CT is supported for inconclusive CT findings as it can "help differentiate tumor from necrotic or inactive scar tissue, malignant from benign tissue, and recurrent tumor from nondescript benign changes".[3].

Restaging

Abdominal/pelvic imaging every 8-12 weeks to assess response to TKI is appropriate given the typical response timeframe for this treatment.[17] NCCN stated "since there is a 95% correlation between the information from regular contrast-enhanced CT and PET/CT, CT with IV contrast is the preferred routine imaging modality for patients with GIST on TKI therapy".[3] More frequent imaging may lead to premature or incorrect treatment decisions. Chest imaging in the absence of prior chest findings or signs and symptoms of chest involvement is low-yield and not recommended by NCCN, it poses potential for risk of increased incidental findings and increased radiation exposure.[3]

Surveillance

Surveillance guidance from NCCN stated that imaging should be performed every three to six months for five years. More frequent surveillance imaging is recommended by NCCN for high-risk tumors. Less frequent imaging is acceptable for low-risk tumors.[3]

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Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)

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- Osteogenic sarcoma (Osteosarcoma) in individuals of all ages should be imaged according to guidelines in **Osteogenic Sarcoma (OS) (PEDONC-9.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Ewing sarcoma and Primitive Neuroectodermal Tumor in individuals of all ages should be imaged according to guidelines in **Ewing Sarcoma and Primitive Neuroectodermal Tumors (ESFT) (PEDONC-9.4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

Indication	Medically Necessary Imaging Study
Chondrosarcoma • Low-grade intra-compartmental • High-grade (grade II or grade III) • Clear cell • Extra-compartmental	ANY or ALL of the following: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast
Dedifferentiated chondrosarcoma	See: **Osteogenic Sarcoma (OS) (PEDONC-9.3)** for imaging
Mesenchymal chondrosarcoma	See: **Ewing's Sarcoma Family of Tumors (PEDONC-9.4)** for imaging

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Indication	Medically Necessary Imaging Study
Chordoma	ANY or ALL of the following: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar (CPT® 72158) Spine without and with contrast • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)
Chordoma with inconclusive findings on conventional imaging	PET/CT (CPT® 78815 or CPT® 78816)

Evidence Discussion

The choice of imaging modality is driven by the primary tumor site. Cross-sectional imaging of the primary site with MRI with and without contrast or CT with contrast is recommended for best illustration of anatomic detail and vascular and nodal involvement. Our guideline provides flexibility for provider discretion for choice of modality for all sites. Providers should choose modality expected to offer maximum primary tumor definition with consideration of minimizing radiation exposure.[2] CT is more sensitive and specific than MRI for evaluation for metastatic disease of the lung and is supported for all individuals with a newly diagnosed malignant sarcoma. Imaging both with and without contrast for evaluation of metastatic disease to the lung is not supported as this exposes individuals to higher radiation doses without significant clinical benefit. CT either with or without contrast should be selected at discretion of the provider.

Chordomas have a propensity for the distant disease at presentation including spine, so unlike other bone sarcomas, imaging of the abdomen and pelvis with contrasted CT as well as bone scan and MRI of the spine are supported. FDG PET/CT is supported only for inconclusive conventional imaging. PET is not generally supported for initial staging of other histologies discussed in this section due to low sensitivity and specificity.[2]

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Bone Sarcomas – Restaging/Recurrence (ONC-12.7)

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Indication	Medically Necessary Imaging Study
Chondrosarcoma • Low-grade intra-compartmental • High-grade (grade II or grade III) • Clear cell • Extra-compartmental	ANY or ALL of the following, after completion of radiotherapy or every 2 cycles of chemotherapy: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast
Suspected recurrence of atypical cartilaginous tumor (low-grade intra-compartmental appendicular tumor)	• MRI without contrast or without and with contrast of involved area or CT (contrast as requested) of involved area • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) Imaging in Oncology [ONC-1.3]** for additional bone scan codes)
Dedifferentiated chondrosarcoma	See: **Osteogenic Sarcoma (OS) (PEDONC-9.3)** for imaging recommendations
Mesenchymal chondrosarcoma	See: **Ewing's Sarcoma Family of Tumors (PEDONC-9.4)** for imaging recommendations
Chordoma	ANY or ALL of the following, after completion of radiotherapy or every 2 cycles of chemotherapy: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) Imaging in Oncology [ONC-1.3]** for additional bone scan codes)

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Indication	Medically Necessary Imaging Study
Chordoma with inconclusive findings on conventional imaging	PET/CT (CPT® 78815 or CPT® 78816)

Evidence Discussion

CT Chest is supported for restaging for all bone sarcomas, as well as MRI of primary site. CT of other body areas is driven by clinical symptoms and patterns of spread at primary site and not routine across all cell types .[2] PET/CT for restaging is not routinely supported but may be used for inconclusive conventional imaging.[2]

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Bone Sarcomas – Surveillance/Follow-up (ONC-12.8)

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Indication	Medically Necessary Imaging Study
• Intra-compartmental Chondrosarcoma	ANY or ALL of the following every 6-12 months for 2 years, then annually for 10 years: • Plain x-ray of primary site ◦ MRI without and with contrast for new findings on plain x-ray or new/worsening clinical symptoms. • Chest x-ray ◦ CT Chest with (CPT® 71260) or without (CPT® 71250) contrast for new findings on chest x-ray, or new/worsening signs/symptoms
• Grade I, II, or III Chondrosarcoma • Clear Cell Chondrosarcoma • Extra-compartmental Chondrosarcoma • Low-grade extra-compartmental appendicular tumors	ANY or ALL of the following every 6 months for 5 years, then annually for 10 years: • Plain x-ray of primary site ◦ MRI without and with contrast for new findings on plain x-ray or new/worsening clinical symptoms. • CT Chest with (CPT® 71260) **or** CT Chest without (CPT® 71250) contrast
Dedifferentiated chondrosarcoma	See: **Osteogenic Sarcoma (OS) (PEDONC-9.3)** for imaging recommendations
Mesenchymal chondrosarcoma	See: **Ewing's Sarcoma Family of Tumors (PEDONC-9.4)** for imaging recommendations

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Indication	Medically Necessary Imaging Study
Chordoma	• Plain x-ray of primary site every 6 months for 5 years and then annually until year 10 ◦ MRI without and with contrast for new findings on plain x-ray or new/worsening clinical symptoms. • Chest x-ray every 6 months for 5 years and then annually until year 10 ◦ CT Chest with (CPT® 71260) or without (CPT® 71250) contrast annually or for evaluation of any new findings on chest x-ray or new/worsening signs/symptoms

Evidence Discussion

Data do not show an overall survival benefit with advanced imaging surveillance of sarcomas. Less than 20% of local recurrences are detected based on advanced imaging surveillance in asymptomatic individuals. Sensitivity and specificity of chest imaging are higher in grade two or higher disease.[18]

For grades I-III disease, plain x-ray is supported as primary tool for primary site surveillance, with cross sectional advanced imaging for signs and symptoms of progression or changes on x-ray. Chest imaging is supported with either plain imaging or CT. Recurrence beyond 10 years is rare; asymptomatic surveillance imaging beyond 10 years is low yield and is not generally supported.[2]

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Benign Bone Tumors – General Considerations (ONC-12.9)

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- Variety of diagnoses, including osteoid osteochondroma, chondroblastoma, desmoplastic fibroma, Paget's disease, osteoid osteoma, and others.
- Plain x-ray appearance is diagnostic for many benign bone tumors and advanced imaging is generally not medically necessary except for preoperative planning.
- MRI without and with contrast is the primary modality for advanced imaging of bone tumors and is medically necessary to help narrow differential diagnoses and determine whether biopsy is indicated.
- Some benign bone tumor types carry a risk of malignant degeneration over time, but routine advanced imaging surveillance has not been shown to improve outcomes for these individuals.
- MRI without and with contrast can be approved to evaluate new findings on plain x-ray new/worsening clinical symptoms not explained by a recent plain x-ray.
- There are no data to support the use of PET/CT in the evaluation of benign bone tumors, and PET requests are not medically necessary without biopsy confirmation of a malignancy.
- Other benign bone tumors should be imaged according to guidelines in **Bone Lesion (MS-10.2)** in the General Musculoskeletal Imaging Guidelines or **Mass Involving Bone (including Lytic and Blastic Metastatic Disease) (PEDMS-3.4)** in the Pediatric Musculoskeletal Imaging Guidelines.

Evidence Discussion

Many benign bone tumors have characteristic appearance on plain x-ray, particularly in conjunction with history, individual age, and size and growth characteristics. Lesions without aggressive appearing characteristics on x-ray generally do not require further evaluation. Advanced imaging modalities are supported when x-ray is indeterminate for malignancy to determine management strategy. Thus, the advanced imaging guidelines in this section pertain to enchondromas, which often appear indeterminate on plain x-ray, and giant cell tumor of bone (GCTB), which have potential for malignant degeneration and metastasis.[2,19]

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Benign Bone Tumors – Initial Work-up/ Staging (ONC-12.10)

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Indication	Medically Necessary Imaging Study
Giant Cell Tumor of Bone (GCTB)	ANY or ALL of the following: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • Bone scan (CPT® 78306) (see also: **Nuclear Medicine (NM) in Oncology [ONC-1.3]** for additional bone scan codes)
Enchondroma	• MRI without contrast or without and with contrast of primary site

Evidence Discussion

- Giant Cell Tumor of Bone
  - MRI can help distinguish malignant transformation, while complex bony anatomy maybe better visualized on CT. To establish management strategy, our guidelines support using both modalities for involved areas in alignment with NCCN and ACR.[2,16]
  - CT Chest and whole-body bone scan are supported at time of initial staging given the malignant and metastatic potential of GCTB. CT Abdomen and Pelvis are not supported without symptoms in these areas as this would not be a typical pattern of metastasis in the setting of malignant degeneration of GCTB. CT of abdomen and pelvis increases radiation exposure with low yield.[2]
- Enchondroma
  - MRI can help distinguish suspected enchondroma on plain film from other more malignant entities and is supported for initial staging to confirm characteristic appearance.[19]

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Benign Bone Tumors – Restaging/ Recurrence (ONC-12.11)

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Indication	Medically Necessary Imaging Study
Giant Cell Tumor of Bone (GCTB)	ANY or ALL of the following, after completion of radiotherapy or every 2 cycles of chemotherapy: • MRI without contrast or without and with contrast of involved area • CT (contrast as requested) of involved area • Bone scan (CPT® 78306 (see also: **Nuclear Medicine (NM) Imaging in Oncology [ONC-1.3]** for additional bone scan codes)
Enchondroma	Plain films of primary site

Evidence Discussion

- GCTB
  - For individuals requiring chemotherapy, repeat of all imaging done at initial staging may be done every two cycles to assess treatment response or need to change therapy. For individuals treated with radiotherapy, repeat imaging may be done at completion of radiotherapy to verify treatment response and establish baseline for surveillance.[2,16]
- Enchondroma
  - Once initial staging with advanced imaging has been completed, plain films should be adequate to ensure stability or for suspected recurrence, restaging after local therapy, or surveillance. Further advanced imaging is generally low yield unless there are indeterminate findings on the plain films.[19]

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Benign Bone Tumors – Surveillance/ Follow-up (ONC-12.12)

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Indication	Medically Necessary Imaging Study
Giant Cell Tumor of Bone (GCTB)	ANY or ALL of the following every 6-12 months for 4 years, then annually thereafter: • Plain x-ray of primary site ◦ MRI without and with contrast is indicated for new findings on plain x-ray or new/worsening clinical symptoms. • Chest x-ray ◦ CT Chest with (CPT® 71260) or without (CPT® 71250) contrast for new findings on chest x-ray, or new/worsening signs/ symptoms.
Enchondroma	Plain films of primary site

Evidence Discussion

- GCTB
  - The role of advanced imaging in asymptomatic surveillance is not well established for GCTB. Though late recurrences can occur, there is not strong data to support advanced imaging over plain film in asymptomatic individuals. These guidelines allow advanced imaging if there are indeterminate findings on plain film, both for primary site and chest. Time frames for plain films are in alignment with NCCN. [2,16]
- Enchondroma
  - Once initial staging with advanced imaging has been completed, plain films should be adequate to ensure stability or further imaging for recurrence, restaging after local therapy, or surveillance . Further advanced imaging is generally low yield unless there are indeterminate findings on the plain films.[19]

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Pancreatic Cancer (ONC-13)

Guideline

Pancreatic Cancer – General Considerations (ONC-13.0)
Pancreatic Cancer – Screening Studies for Pancreatic Cancer (ONC-13.1)
Pancreatic Cancer – Suspected/Diagnosis (ONC-13.2)
Pancreatic Cancer – Initial Work-up/Staging (ONC-13.3)
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References (ONC-13)

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Pancreatic Cancer – General Considerations (ONC-13.0)

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- This guideline refers only to adenocarcinoma of the exocrine pancreas, which accounts for over 90% of pancreatic malignancies. This guideline may also be used for cancer of the Ampulla of Vater.
- Neuroendocrine and carcinoid tumors of the pancreas are not included in this guideline, see: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)**.

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Pancreatic Cancer – Screening Studies for Pancreatic Cancer (ONC-13.1)

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- Detailed history of any known inherited syndrome in the individual and detailed family history in first- and second-degree relatives, including the age and lineage, is essential to guide screening recommendations. See table below for age- and risk-specific screening recommendations.
- New onset of diabetes in individuals older than 50 has been recognized as a potential indicator of the development of pancreatic cancer. Approximately 1% of individuals in this category are diagnosed with cancer within 3 years. A prediction model has been established which identifies those individuals at greatest risk for pancreatic malignancy. The scoring system, known as ENDPAC (Enriching New-Onset Diabetes for Pancreatic Cancer) is based on 3 discriminatory factors, including change in blood glucose, change in weight, and age of onset at the time of the new diagnosis of diabetes. A score of >3 imparts an elevated risk of pancreatic cancer (3.6%), and these individuals should be screened. Screening is not medically necessary at this time for scores of 0-2.

Indications	Medically Necessary Imaging Study
Individuals who meet **BOTH** of the following criteria: • One or more first- or second-degree relative affected with pancreatic cancer **AND** • Known mutation carrier of ONE of the following genes: ◦ Lynch Syndrome (MLH1, MSH2, or MSH6 gene mutations) ◦ BRCA1 ◦ PALB2 mutation ◦ EPCAM ◦ TP53	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) starting at age 50 or 10 years earlier than the youngest family member affected with exocrine pancreatic cancer, repeat annually

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Indications	Medically Necessary Imaging Study
Individuals with family history of pancreatic cancer but no known genetic mutation: • Individuals with 2 relatives with pancreatic cancer where one is a first-degree relative • Individuals with 3 or more relatives with pancreatic cancer	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) starting at age 45 or 10 years earlier than the youngest family member affected with exocrine pancreatic cancer, repeat annually
Pancreatic Cancer Kindred (individuals who have at least one first-degree relative with pancreatic cancer who in turn also has a first-degree relative with pancreatic cancer) and NO known genetic germline mutations	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) starting at age 50 or 10 years earlier than the youngest family member affected with exocrine pancreatic cancer, repeat annually
Hereditary Pancreatitis (PRSS1, CPA1, and CTRC gene mutations)	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) beginning at age 40 or 20 years after the first pancreatitis attack, repeat annually.
Peutz-Jeghers Syndrome (LKB1/STK11 gene mutation)	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) starting at age 30 or 10 years earlier than the youngest family member affected with exocrine pancreatic cancer, repeat annually ◦ Family history is not required to begin imaging at age 30

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Indications	Medically Necessary Imaging Study
CDKN2A mutation (also known as p16, p16INK4a, and MTS1, FAMM-Familial Atypical Multiple Melanoma and Mole Syndrome)	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) beginning at age 40 or 10 years earlier than the youngest family member affected with exocrine pancreatic cancer, repeat annually. ◦ Family history is not required to begin imaging at age 40
• BRCA2 • ATM (Ataxia-Telangiectasia)	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) starting at age 50 or 10 years earlier than the youngest family member affected with exocrine pancreatic cancer, repeat annually. ◦ Family history is not required to begin imaging at age 50
Screening MRI reveals cystic lesion of the pancreas	• Repeat MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) in 6 months
Screening MRI reveals indeterminate solid lesion	• CT Abdomen with contrast – pancreatic protocol (CPT® 74160) • Repeat MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) in 3 months after the CT scan
Screening MRI reveals pancreatic stricture and/or dilation ≥6 mm without a mass	• CT Abdomen with contrast – pancreatic protocol (CPT® 74160) • Repeat MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) in 3 months after the CT scan

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Indications	Medically Necessary Imaging Study
New onset diabetes in adults with ENDPAC score of ≥3	• CT Abdomen without and with contrast (CPT® 74170) or MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) at baseline; if negative, repeat once after 6 months

Background and Supporting Information

ENDPAC scoring:

Blood Glucose (BG) Categories		Change BG Category Score (NOD-1y)(A)
BG Range (mg/dl)	Score	Score Range
BG Category at -1 years		1-4
<100	1	
100-109	2	
110-125	3	
BG Category at Glycemically-Defined New-Onset Diabetes		
126-160	4	
>160	5	
Change Weight Categories		Change Weight Score (B)
Change Weight (kg)	Score	Score Range
≤-6.0	+6	-6 to +6
-5.9 to -4.0	+4	
-3.9 to -2.0	+2	
-1.9 to +1.9	0	
+2.0 to +3.9	-2	
+4.0 to +5.9	-4	
≥+6.0	-6	
Age (years) at glycemically-defined new-onset diabetes		Age Score (C)

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Blood Glucose (BG) Categories		Change BG Category Score (NOD-1y)(A)
Age Range	Score	Score Range
≤59	-1	-1 to +1
60 to 69	0	
≥70	+1	
Total Score		A + B + C

Evidence Discussion

International Cancer of the Pancreas Screening Consortium (CAPS) recommended screening for those with an estimated lifetime risk of pancreatic cancer >5% to facilitate early detection, as the survival of individuals diagnosed with advanced disease at presentation is extremely poor. Individuals may be high-risk due to family history of 2-3 relatives with pancreatic adenocarcinoma with first degree relative affected, or those with known deleterious genetic mutations in conjunction with one first- or second-degree relative with pancreatic adenocarcinoma.[3,14,17] Asymptomatic screening has not been shown to improve outcomes in those without an established lifetime risk of >5%, including those with only more distant relatives affected, a single relative affected and no known high-risk mutation, or those with mutations whose risk is unknown.[3,14]

The lifetime risk and age at presentation varies with each genetic mutation. Updated CAPS recommendations generally support imaging at age 50 or ten years younger than the age of the youngest relative with pancreatic adenocarcinoma. These guidelines align with CAPS and NCCN with multiple exceptions to this to allow screening at younger ages for individuals with mutations known to be higher risk or present at younger ages.[17,13] US Preventative Services Task Force (USPSTF) did not generally recommend pancreatic cancer screening, though they did not evaluate individuals with high-risk criteria in this data. The USPSTF noted that screening has moderate risk due to unnecessary imaging and management of incidental findings.[4] These findings further support that screening for those that do not meet the clearly defined risk factors listed in the guideline is not indicated.

MRI without and with contrast shows up to 93% sensitivity for pancreatic lesions and better illustrates pancreatic ducts, which may indirectly identify pancreatic malignancy, and is the preferred modality for screening for most individuals in these guidelines. This is superior to both CT and endoscopic ultrasound, which may not detect smaller lesions.[15] CT pancreatic protocol is supported in these guidelines if indeterminate findings on MRI in the interest of early detection.

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A unique group is individuals with new onset diabetes and ENDPAC score >3. These individuals are at an elevated risk of developing pancreatic cancer within 6 months even in the absence of family history or known deleterious mutations. MRI or CT is supported in this population at diagnosis of diabetes and again in 6 months.[16,12]

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Pancreatic Cancer – Suspected/ Diagnosis (ONC-13.2)

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Indication	Medically Necessary Imaging Study
For any suspected symptoms only (e.g., epigastric pain, weight loss, pain radiating to back, etc.)	• Ultrasound (CPT® 76700 or CPT® 76705) • Also see: **Epigastric Pain and Dyspepsia (AB-2.5)**
Symptoms suspicious for pancreatic cancer AND any one of the following: • Abnormal labs (e.g., elevated CA 19-9, ALKP, bilirubin, or GGTP) • Abnormal physical exam findings (e.g., abdominal mass) • Abnormal or non-diagnostic ultrasound/ERCP	Any ONE of the following: • CT Pancreatic Protocol (CT Abdomen with contrast with dual phase imaging, CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377)
Preoperative studies for potentially resectable tumors without confirmed histologic diagnosis	• See: **Pancreatic Cancer – Initial Work-up/ Staging (ONC-13.3)**

Evidence Discussion

For individuals with symptoms suspicious for pancreatic cancer with abnormal labs and physical findings or abnormal or non-diagnostic ultrasound or ERCP, CT pancreatic protocol (abdomen without and with contrast) or MRI abdomen without and with contrast are supported. CT pancreatic protocol has a sensitivity and specific or 90 and 87 percent respectively.[20,19] Sensitivity and specificity of MRI and 89 and 90% respectively.[20,19] CT has the advantage of being less costly, more accessible, and faster, but exposes to more radiation. MRI may be more sensitive for visualization of pancreatic ducts and has less radiation exposure, but can be less accessible, more costly, takes longer, and can pose difficulties with claustrophobia and has more contraindications.[19] Given their similar sensitivity and specificity, these guidelines allow flexibility for individuals and providers to weigh risks and benefits for each individual for suspected disease.

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Pancreatic Cancer – Initial Work-up/ Staging (ONC-13.3)

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Indication	Medically Necessary Imaging Study
All individuals	• CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with (CPT® 74177) or CT Abdomen and Pelvis without and with contrast (CPT® 74178)
For any of the following: • Preoperative planning • CT insufficient to determine resectability • Evaluation of indeterminate liver lesions	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377)
No evidence of metastatic disease on CT or MRI AND any of the following: • Equivocal or indeterminate conventional imaging findings • Markedly elevated CA 19-9 • Large primary tumor(s) • Enlarged regional lymph nodes • Planned neoadjuvant therapy	• PET/CT (CPT® 78815)

Evidence Discussion

For biopsy proven pancreatic adenocarcinoma, evaluation for metastatic disease with contrasted CTs of the chest, abdomen and pelvis are supported, given that approximately 80% of individuals have metastatic or locally advanced disease at presentation.[20] Resectability has a dramatic impact on prognosis, and MRI abdomen with and without contrast is supported in addition to CT if CT insufficient to determine resectability, for preoperative planning, or to evaluate indeterminate liver lesions.[1] Given the high incidence of metastatic disease at presentation, if high-risk features are noted but no metastatic disease is visible on conventional imaging, a PET/CT is

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supported to assess for occult extra-pancreatic metastatic disease, in alignment with the NCCN. This is not a substitute for a diagnostic-quality contrasted CT or MRI, which is superior for detecting pancreatic disease.[1,19] PET/MRI has a weak expert consensus without sufficient data, and as such these guidelines do not routinely support this modality.[1,21,22,24]

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Pancreatic Cancer – Restaging/ Recurrence (ONC-13.4)

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Indication	Medically Necessary Imaging Study
For ANY of the following: • After neoadjuvant therapy • Post-operative baseline • Suspected recurrence	• CT Chest with contrast (CPT® 71260) • Abdominal imaging with one of the following, using same modality as used at initial evaluation: ◦ CT Abdomen and Pelvis with (CPT® 74177) ◦ CT Abdomen and Pelvis without and with contrast (CPT® 74178) ◦ MRI Abdomen without and with contrast (CPT® 74183) ◦ MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377) • CT with contrast of other involved or symptomatic areas
Unresectable disease or metastatic disease on chemotherapy	Every 2 cycles of treatment (commonly every 6 to 8 weeks): • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with (CPT® 74177) or CT Abdomen and Pelvis without and with contrast (CPT® 74178) • CT with contrast of other involved or symptomatic areas
Unexplained elevated liver enzymes or inconclusive recent CT abnormality	• MRI Abdomen without and with contrast (CPT® 74183) or MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377)
If complete surgical resection was initial therapy	• See: **Pancreatic Cancer – Surveillance/Follow-up for surveillance imaging (ONC-13.5)**
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

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Evidence Discussion

For individuals with unresectable or metastatic disease on chemotherapy, imaging of the chest, abdomen and pelvis with contrast (or without and with contrast for the abdomen and pelvis) are supported every 2 cycles of treatment, given the high rate of progression and metastases, to allow for prompt consideration of changes in treatment. CT of any involved or symptomatic area is also supported. The same imaging is supported after neoadjuvant chemoradiation to assess resectability and rule out metastatic disease. Resection is the mainstay of curative therapy in pancreatic cancer, but only 20% of individuals achieve this status. Individuals with unresectable or metastatic disease are unlikely to benefit from surgery, thus pre-operative evaluation for metastasis is essential to minimize unnecessary surgical risk.[20,1] As noted in the initial staging section, both CT and MRI have advantages and disadvantages. MRI is more specific for liver disease and offers better soft tissue differentiation for inconclusive findings.[1,24] The same imaging guidance applies to suspected recurrence of disease. Restaging studies are also supported as a post-operative baseline, to ensure the absence of residual disease and make decisions regarding adjuvant treatment, and to ensure accurate comparison for surveillance imaging.

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Pancreatic Cancer – Surveillance/Follow-up (ONC-13.5)

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Indication	Medically Necessary Imaging Study
All individuals (other than resected ampullary cancer)	Every 3 months for 2 years, then annually: • CT Chest with contrast (CPT® 71260) And ANY ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Resected ampullary cancer	Every 3 months for 2 years, then every 6 months for up to 5 years: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Measurable metastatic disease on maintenance therapy or being monitored off therapy	Every 3 months for up to 5 years after completion of definitive treatment, then annually: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

Evidence Discussion

Data on the role of surveillance in pancreatic cancer are limited, due to the poor prognosis and small numbers of individuals with long-term follow up. The NCCN recommendations for surveillance are based on consensus rather than data, and SEER-Medicare data shows no significant survival benefit for individuals who received regular surveillance scans/no improved outcome with earlier detection of recurrence.[1,23] The role of ongoing imaging in individuals with metastatic disease on maintenance or observation is also unclear. There is no clear data on length of time for maintenance

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therapy in those with metastatic disease, and treatment 'holidays' are often interjected in therapy. Outside of situations where there is a clear impact on management decisions, ASCO stated that imaging should be supplanted by clinical evaluation.[24] However, the NCCN offered several second and third-line treatment options. These guidelines provide some flexibility for patient-centricity and provider preference in this setting, allowing for CT Chest, Abdomen and Pelvis every 3 months for up to 5 years and then annually after completion of definitive treatment.[1] Specifically for individuals with resected ampullary cancer, NCCN recommended surveillance after adjuvant therapy every three to six months for two years, then every six to 12 months for up to five years. Imaging studies as well as history, physical exam, and tumor marker studies are recommended components of surveillance for these individuals.[25]

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Upper GI Cancers (ONC-14)

Guideline

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Hepatocellular Carcinoma (HCC) – General Considerations (ONC-14.1)

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- A biopsy is not always required for the diagnosis of Hepatocellular carcinoma (HCC). A dedicated triple-phase CT or MRI may be obtained. MRI with contrast is the test of choice for the evaluation of liver masses. It offers soft tissue contrast resolution superior to CT as well as the possibility of using two different contrast agents, one of which if more blood flow based and the other which also is blood flow based and demonstrates hepatobiliary function (Eovist). Classical imaging findings include:
  - arterial phase hyper-enhancement
  - venous phase washout appearance
  - capsule appearance
  - threshold growth
- For individuals who are high-risk for developing HCC (cirrhosis, chronic Hepatitis B or current or prior HCC), if the liver lesion is >1 cm with 2 classic enhancements on triple-phase CT or MRI, the diagnosis is confirmatory and biopsy is not needed.
- For lesions less than 1 cm or with less than 2 classical enhancements or for any liver lesions in individuals who are not high-risk, a biopsy is needed for histological confirmation. PET/CT scan is considered not medically necessary for the diagnosis or staging of HCC.

Evidence Discussion

HCC does not necessarily require a biopsy for diagnosis as a triple phase CT or MRI can support the diagnosis in the absence of biopsy as these lesions are characterized by arterial hypervascularity and "wash out" on portal venous phases unlike the surrounding liver. For individuals who are high-risk for developing HCC (cirrhosis, chronic Hepatitis B or current or prior HCC), if the liver lesion is >1 cm with 2 classic enhancements on triple-phase CT or MRI, the diagnosis is confirmatory and biopsy is not needed. For lesions less than 1 cm or with less than 2 classical enhancements or for any liver lesions in individuals who are not high-risk, a biopsy may be needed for histological confirmation. Serum biomarkers such as AFP are not sensitive or specific enough to establish a diagnosis. PET/CT scan is not medically necessary for the diagnosis or staging of HCC due to limited sensitivity.[2,15]

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Hepatocellular Carcinoma (HCC) – Suspected/Diagnosis (ONC-14.2)

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- See: **Chronic Liver Disease, Cirrhosis and Screening for HCC (AB-26.1)** in the Abdomen Imaging Guidelines.
- See: **Liver Lesion Characterization (AB-29.1)** in the Abdomen Imaging Guidelines.

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Hepatocellular Carcinoma (HCC) – Initial Work-up/Staging (ONC-14.3)

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Indication	Medically Necessary Imaging Study
All individuals	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) And ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen without and with contrast (CPT® 74170) • CT Abdomen and Pelvis with contrast (CPT® 74177) or without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • Bone scan (CPT® 78306) (for skeletal symptoms)
Inconclusive conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

Initial imaging of hepatocellular carcinoma seeks to establish the tumor burden in the liver. CT or MRI imaging is preferred over the use of ultrasound. Additionally, common sites of metastases in individuals with hepatocellular carcinoma include the lungs, adrenal glands, peritoneum, and bone. Thus, initial work-up for all newly diagnosed individuals requires chest imaging. Bone scan completes this initial work-up by allowing dedicated imaging to identify potential bone metastases if suspected. PET/CT is a useful tool for evaluating inconclusive findings on the initial imaging studies due to its limited specificity but high sensitivity.[2,8]

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Hepatocellular Carcinoma (HCC) – Restaging/Recurrence (ONC-14.4)

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Indication	Medically Necessary Imaging Study
ONE of the following: • After initial therapy • For suspected recurrence or new liver lesions • Individuals receiving systemic therapy (every 2 cycles)	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) And ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen without and with contrast (CPT® 74170) • CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Inconclusive conventional imaging	• PET/CT (CPT® 78815)
Hepatocellular Carcinoma treated with local therapy (ablation, embolization)	See: **Liver Metastases (ONC-31.2)** for imaging studies prior to and post-procedure
Hepatocellular Carcinoma awaiting liver transplant	See: **Liver Transplant, Pre-Transplant (AB-42.1)** in the Abdomen Imaging Guidelines

Evidence Discussion

To assess response after treatment, dedicated imaging of the chest, abdomen, and pelvis are indicated. NCCN stated, "CT of the chest and CT or MRI of the abdomen and pelvis are the preferred modalities as they reliably assess intranodular arterial vascularity, a key feature of residual or recurrent tumor".[2] Thus, these studies are also recommended for suspected recurrence or new liver lesions. PET/CT remains a useful tool for evaluation of inconclusive conventional studies due to its limited sensitivity but high specificity.[2] For individuals undergoing liver embolization, CTA Abdomen can be obtained immediately prior to this procedure. In addition, either MRI or CT of the

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Abdomen with and without contrast can be obtained immediately prior and 1 month post-ablation.[2,8] ONC-31.2 provides a broader description of additional appropriate studies prior to and after embolization for liver metastases.

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Hepatocellular Carcinoma (HCC) – Surveillance/Follow-up (ONC-14.5)

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Indication	Medically Necessary Imaging Study
Hepatocellular Carcinoma: • Treated with surgical resection • Treated with embolization • Being monitored off therapy	Every 3 months for 2 years, then every 6 months until year 5: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) And ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen without and with contrast (CPT® 74170) • CT Abdomen and Pelvis with contrast (CPT® 74177) or without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast
Hepatocellular Carcinoma treated with liver transplant	• See: **Liver Transplant, Post-transplant Imaging (AB-42.3)** in the Abdomen Imaging Guidelines

Evidence Discussion

Individuals treated with surgical resection, embolization or being monitored off therapy, recommendation is to obtain CT Chest with or without contrast every 3 months for 2 years then every 6 months until year 5 as well as one of the following: CT Abdomen with or with/without contrast, CT Abdomen/Pelvis with or with/without contrast or MRI Abdomen/Pelvis with and without contrast. Multiphasic cross-sectional imaging with CT or MRI is preferred due to its reliability in assessing arterial vascularity, which is associated with increased risk of recurrence following treatment.[2,9,10] HCC treated with transplant is addressed in AB-42.3, Liver Transplant and Post-Transplant Imaging.

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Gallbladder and Biliary Tumors – Initial Work-up/Staging (ONC-14.6)

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Indication	Medically Necessary Imaging Study
All individuals	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) And ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen without and with contrast (CPT® 74170) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • MRCP with 3D rendering (CPT® 74183 and CPT® 76376 or CPT® 76377)
Inconclusive liver findings on CT (if MRI not already performed)	MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Inconclusive findings on MRI Abdomen	• PET/CT (CPT® 78815)

Evidence Discussion

High-quality contrast-enhanced cross-sectional imaging (CT and/or MRI) of the chest, abdomen, and pelvis is recommended to evaluate tumor penetration through the wall of the gallbladder and the presence of nodal and distant metastases, and to detect the extent of direct tumor invasion of other organs/biliary system or major vascular invasion. PET/CT is a useful tool for inconclusive findings on CT/MRI, with downsides including false-positives related to inflammation of the gallbladder.[3,11,12]

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Gallbladder and Biliary Tumors – Restaging/Recurrence (ONC-14.7)

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Indication	Medically Necessary Imaging Study
ANY of the following: • After initial therapy • For suspected recurrence or new liver lesions • Individuals receiving systemic chemotherapy (every 2 cycles)	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) And ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Inconclusive liver findings on CT (if MRI not already performed for restaging)	• MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Inconclusive findings on MRI Abdomen	• PET/CT (CPT® 78815)

Evidence Discussion

CT Chest with or without contrast plus one of the following: CT Abdomen with or with/without contrast, CT Abdomen/Pelvis with contrast and MRI Abdomen/Pelvis with and without contrast. High-quality contrast-enhanced cross-sectional imaging (CT and/or MRI) of the chest, abdomen, and pelvis is recommended to evaluate tumor penetration through the wall of the gallbladder and the presence of nodal and distant metastases, and to detect the extent of direct tumor invasion of other organs/biliary system or major vascular invasion. PET/CT for inconclusive findings on CT/MRI keeping in mind false positives related to an inflamed gallbladder are problematic.[3,11,12]

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Gallbladder and Biliary Tumors – Surveillance/Follow-up (ONC-14.8)

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Indication	Medically Necessary Imaging Study
All individuals	Every 3 months for 2 years, and then every 6 months up to year 5: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) And ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen without and with contrast (CPT® 74170) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Biliary carcinoma treated with liver transplant	See: **Liver Transplant, Post-transplant Imaging (AB-42.3)** in the Abdomen Imaging Guidelines

Evidence Discussion

NCCN stated "there are no data to support a specific surveillance schedule following resection of gallbladder cancer".[3] However, NCCN did suggest consideration of imaging studies every three to six months for two years, and then every six to twelve months for up to five years. Recurrence typically occurs within two years.[3] Thus, this guideline allows for all individuals to receive imaging based upon that schedule. Biliary cancer treated with liver transplant would follow AB-42.3, Liver Transplant, Post-transplant Imaging.

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Gastric Cancer – Initial Work-up/Staging (ONC-14.9)

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Indication	Medically Necessary Imaging Study
All individuals	• CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Gastric cancer ≥T2 or higher with no metastatic disease by conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

All individuals should get CT Chest/Abdomen/Pelvis with contrast. For T2 or higher stage disease with no metastatic disease by CT, PET/CT recommended to complete staging. FDG-PET alone is not an adequate diagnostic procedure in the detection and preoperative staging of gastric cancer, but can be helpful when used in conjunction with CT.[1,13,14]

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Gastric Cancer – Restaging/Recurrence (ONC-14.10)

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Indication	Medically Necessary Imaging Study
• After initial therapy for presumed surgically resectable disease • Post curative chemoradiation being treated without surgery • For suspected recurrence	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)
Monitoring response to chemotherapy (every 2 cycles, ~every 6-8 weeks) for: • Unresected primary disease • Metastatic disease	• CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Chest with contrast (CPT® 71260)
New liver lesion(s) and primary site controlled	• CT Abdomen without and with contrast (CPT® 74170) or MRI Abdomen without and with contrast (CPT® 74183)
EITHER of the following: • Inconclusive findings on conventional imaging • Restaging of individuals with unresectable disease who cannot undergo CT with contrast due to renal insufficiency or contrast allergy	• PET/CT (CPT® 78815)

Evidence Discussion

CT Chest/Abdomen/Pelvis should be obtained after initial therapy for presumed resectable disease, post curative chemoradiation (no surgery) and for suspected recurrence. Monitoring chemotherapy response should include CT Abdomen/Pelvis and include CT Chest for known disease, new/worsening pulmonary symptoms or abnormal chest x-ray. PET/CT can be considered with inconclusive findings on conventional imaging. PET/CT is also the preferred option for imaging individuals who cannot undergo CT evaluations due to contrast allergies or renal insufficiency.[1]

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Gastric Cancer – Surveillance/Follow-up (ONC-14.11)

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Indication	Medically Necessary Imaging Study
Stage I (treated with resection alone)	• No routine imaging unless clinical signs/ symptoms of recurrence
ANY of the following: • Stage I treated with systemic therapy • Stages II-III • Stage IV - Metastatic disease with no measurable disease post definitive treatment	Every 6 months for 2 years, and then annually for 3 more years: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) and CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ In individuals who cannot undergo CT with contrast due to renal insufficiency or contrast allergy, MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) are the medically necessary substitute.
Measurable metastatic disease on maintenance therapy or being monitored off therapy	Every 3 months for up to 5 years after completion of active treatment: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)

Evidence Discussion

Stage I treated with resection alone does not require routine imaging in the absence of signs/symptoms of recurrence. Stage I treated with systemic therapy, Stages II-III and Stage IV s/p definitive treatment of all measurable disease or being observed off therapy should undergo CT Chest/Abdomen/Pelvis every 6 months for 2 years then annually up to 5 years. Measurable metastatic disease on maintenance therapy or being monitored off therapy should undergo CT Chest/Abdomen/Pelvis every 3 months for up to 5 years after completion of active treatment.[1]

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Neuroendocrine Cancers and Adrenal Tumors (ONC-15)

Guideline

General Considerations (ONC-15.1)
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Bronchopulmonary or Thymic Carcinoid – Initial Staging (ONC-15.6)
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General Considerations (ONC-15.1)

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This guideline includes low-grade or well-differentiated carcinoid and endocrine tumors of the lung, thymus, pancreas, gastrointestinal tract or unknown primary site; including insulinoma, glucagonoma, VIPoma, gastrinoma, somatostatinoma and others as well as catecholamine-secreting tumors of the adrenal gland such as pheochromocytoma, paraganglioma, adrenocortical carcinoma, and others.

- For poorly-differentiated or high-grade small cell or large cell neuroendocrine tumors arising outside the lung or from an unknown primary site, see: **Extrathoracic Small Cell and Large Cell Neuroendocrine Tumors (ONC-31.8)**.
- For poorly-differentiated or high-grade neuroendocrine tumors of the lung, see: **Small Cell Lung Cancer (ONC-7)**.
- Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma occurring in adults should be imaged according to **Neuroblastoma (PEDONC-6)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Many are associated with Multiple Endocrine Neoplasia (MEN) familial syndromes.
  – See: **Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)** in the Pediatric and Special Populations Oncology Imaging Guidelines for screening recommendations.
- Somatostatin receptor (SSR) based imaging is more sensitive and specific for evaluation of well-differentiated neuroendocrine tumors and may be performed using ^{111}In DTPA Octreotide scintigraphy or PET/CT scan with SSR radiotracers (such as ^{68}Ga–DOTATATE, ^{68}Ga-DOTATOC, or ^{64}Cu-DOTATATE). This study is not part of evaluation of poorly-differentiated or high-grade neuroendocrine tumors, which are imaged according to: **Extrathoracic Small Cell and Large Cell Neuroendocrine Tumors (ONC-31.8)**.

Evidence Discussion

This guideline includes low-grade or well-differentiated (Grade 1, 2, or 3; Ki-67 >20% and <55%) carcinoid and endocrine tumors of the lung, thymus, pancreas, gastrointestinal tract or unknown primary site; including insulinoma, glucagonoma, VIPoma, gastrinoma, somatostatinoma and others as well as catecholamine-secreting tumors of the adrenal gland such as pheochromocytoma, paraganglioma, adrenocortical carcinoma, and others. These tumors are particularly sensitive and specific to somatostatin receptor (SSR) based imaging (nearly 80% express SSR on the cell surface) while poorly differentiated or high-grade tumors typically are not with imaging recommendations being addressed in separate guidelines.[1,9]

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Gastrointestinal/Pancreatic Neuroendocrine Cancers – Suspected/ Diagnosis (ONC-15.2)

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Indication	Medically Necessary Imaging Study
• Systemic symptoms strongly suggestive of functioning neuroendocrine tumor • Suspicious findings on other imaging studies • Unexplained elevation in ANY of the following: ◦ Chromogranin A ◦ 5HIAA ◦ Insulin ◦ VIP ◦ Glucagon ◦ Gastrin ◦ Substance P ◦ Serotonin ◦ Somatostatin	ANY of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) or without and with contrast (CPT® 74178) **OR** MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT with contrast or MRI without and with contrast of any other symptomatic body areas

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Indication	Medically Necessary Imaging Study
• Continued suspicion with negative/ inconclusive CT or MRI	ONE of the following: • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE • If PET/CT is not available, octreotide scan ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, or 78804 **AND** ◦ Any one of the follow SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832

Evidence Discussion

Neuroendocrine tumors (NETs) arise from cells of the endocrine system that can be found throughout the body. They can occur sporadically or arise in the context of an inherited genetic syndrome. Presentation is usually attributable to hormonal hypersecretion (functional tumors) that can include flushing/diarrhea/wheezing (Carcinoid syndrome), hypertension and hypoglycemia versus being found incidentally on various imaging studies. If these symptoms/signs are suspicious for a NET, appropriate serologic/urinary workup may include chromogranin A, 5HIAA, insulin, VIP, glucagon, gastrin, substance P, serotonin and somatostatin. If these markers are elevated, CT chest with or without contrast as well as CT or MRI of abdomen/pelvis/ any other symptomatic body area is indicated. CT is best for detection of primary small bowel lesions and lymphadenopathy while MRI is preferred for pancreatic NETs and detecting hepatic metastases. If these imaging studies are negative/inconclusive, SSR based imaging with either Octreotide scan or Dotatate/Dotatoc (Gallium-68, Copper-64) may be indicated (SSR PET/CT). All three of these functional imaging modalities are FDA approved and can be performed in individuals on somatostatin analog therapy and are considered to be superior to Octreotide scan. FDG-based PET/CT has limited use as the majority of NETs are metabolically inactive and fail to take up the tracer well.[1,9,10,11]

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Gastrointestinal/Pancreatic Neuroendocrine Cancers – Initial Work-up/Staging (ONC-15.3)

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- For undifferentiated neuroendocrine tumors, see: **Extrathoracic Small Cell and Large Cell Neuroendocrine Tumors ONC-31.8**

Indication	Medically Necessary Imaging Study
ANY of the following: • Jejunal • Ileal • Colon • Gastric type 3 • Neuroendocrine tumors of unknown primary • Grade 3 well-differentiated tumors	If not already done, ANY or ALL of the following: • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ 68Ga-DOTATATE ◦ 68Ga-DOTATOC ◦ 64Cu-DOTATATE • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • And ONE of the following: ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ CT Abdomen and Pelvis without and with contrast (CPT® 74178) ◦ MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast

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Indication	Medically Necessary Imaging Study
All other GI or pancreatic neuroendocrine (carcinoid) tumors	If not already done: • CT Abdomen and Pelvis with contrast (CPT® 74177) or without and with contrast (CPT® 74178) OR MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250)
Inconclusive CT or MRI scans	ONE of the following: • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE • If PET is not available, octreotide scan (ANY one of the following): ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, or 78804 AND ◦ Any one of the following SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832
ANY of the following: • Markers fail to normalize after complete resection AND CT/MRI and somatostatin-receptor based study are negative • Biopsy-proven neuroendocrine tumor of unknown primary site AND CT/MRI and somatostatin-receptor based study are negative	• FDG-PET/CT (CPT® 78815)

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Evidence Discussion

For initial evaluation of all neuroendocrine tumors, the National Comprehensive Cancer Network (NCCN) recommended that cross-sectional imaging be performed of the primary site of disease utilizing either CT or MRI. Imaging with chest CT is also an NCCN recommended consideration. For certain disease types including neuroendocrine tumors of the jejunum, ileum, and colon, as well as gastric type 3, neuroendocrine tumors of unknown primary, and grade 3 well-differentiated tumors, the NCCN recommends that a PET/CT or PET/MRI utilizing SSR radiotracers be performed in addition to diagnostic CT or MRI imaging. Brain imaging is not generally recommended for well-differentiated NET without signs and symptoms of neurologic involvement. The NCCN states octreotide scans are "much less sensitive for defining SSTR-positive disease than PET". Thus, these guidelines reserve octreotide scans as a problem-solving tool if PET is inconclusive or not available.[1,10,11]

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Gastrointestinal/Pancreatic Neuroendocrine Cancers – Restaging/ Recurrence (ONC-15.4)

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Indication	Medically Necessary Imaging Study
All after surgical resection	• See: **Gastrointestinal/Pancreatic Neuroendocrine Cancers – Surveillance (ONC-15.5)**
Unresectable/metastatic disease on treatment with somatostatin analogues	• CT or MRI of involved body area no more frequently than every 3 months
Unresectable/metastatic disease on treatment with chemotherapy	• CT or MRI of involved body area every 2 cycles (6 to 8 weeks)
Suspected recurrence	• PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE AND • CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) And ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast

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Indication	Medically Necessary Imaging Study
Continued suspicion for recurrence with negative or inconclusive CT or MRI, and PET not available	• Octreotide scan: ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, or 78804 **AND** ◦ Any one of the following SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832
To assess candidacy for peptide receptor radionuclide therapy (PRRT) with Lutetium ^{177}Lu-dotatate	• PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE

Evidence Discussion

NCCN recommended that for any suspected recurrence of disease, PET/CT or PET/MRI be performed in addition to systemic diagnostic imaging. For individuals with unresectable/metastatic disease, NCCN stated that "anatomic imaging should generally be performed every 12 weeks-12 months based on clinical or pathologic signs of aggressiveness". Thus, CT imaging of involved body area is recommended every 3 months if on somatostatin therapy versus every 2 cycles (6-8 weeks) if on chemotherapy. To assess for candidacy for peptide receptor radionuclide therapy (PRRT) with Lutetium Lu-177 dotatate (Lutathera), any of the 3 SSR PET/CT or PET/MRI options are supported. The NCCN stated octreotide scans are "much less sensitive for defining SSTR-positive disease than PET". Thus, these guidelines reserve octreotide scans as a problem-solving tool if PET is inconclusive or not available.[1]

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Gastrointestinal/Pancreatic Neuroendocrine Cancers – Surveillance (ONC-15.5)

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Indication	Medically Necessary Imaging Study
ANY of the following: • Appendix carcinoid <1 cm, completely resected • Rectal carcinoid <1 cm, completely resected • Gastric carcinoid treated with complete endoscopic resection	• Advanced imaging is not medically necessary for surveillance
Rectal carcinoid 1-2 cm, completely resected	• MRI Pelvis without and with contrast (CPT® 72197) at 6 and 12 months post resection. No further routine surveillance.
All other GI neuroendocrine tumors (stomach, large and small intestine)	• CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast once at 3 to 12 months post-operatively and every 12-24 months up to year 10 ◦
Unresected GI neuroendocrine tumors being monitored with observation alone	• CT Abdomen with contrast (CPT® 74160) once at 3 to 12 months from initial diagnosis, then every 3 months for 1 year, then every 6 months in year 2, and then annually up to year 10
Pancreatic neuroendocrine tumors	• CT Abdomen with contrast (CPT® 74160) once at 3 to 12 months post-operatively then annually up to year 10

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Indication	Medically Necessary Imaging Study
Unresected pancreatic neuroendocrine tumors being monitored with observation alone	• CT Abdomen with contrast (CPT® 74160) once at 3 to 12 months from initial diagnosis then annually up to year 10
Measurable metastatic disease on maintenance treatment or off therapy	• CT of involved body area no more frequently than every 3 months

Evidence Discussion

In the absence of signs/symptoms of recurrence, advanced imaging is not routinely indicated for completely resected appendiceal carcinoid <1 cm, completely resected rectal carcinoid <1 cm, and gastric carcinoid treated with complete endoscopic resection due to their excellent prognosis and low risk of recurrence.[6,13] For completely resected rectal carcinoid 1-2 cm, MRI pelvis with and without contrast at 6 and 12 months post-resection and if clear, no further imaging. Due to the indolent nature of NETs, long term follow-up is recommended. For all other GI NETs, CT or MRI abdomen/pelvis once 3-12 months post-operatively and then every 12-24 months for 10 years is supported by NCCN. Unresected GI NETs on observation should undergo CT abdomen with contrast once at 3-12 months from initial diagnosis then annually up to year 10. Resected pancreatic NETs should undergo CT abdomen with contrast once at 3-12 months post-operatively then annually up to year 10. Unresected pancreatic NETs should undergo CT Abdomen with contrast once at 3-12 months from initial diagnosis then annually up to year 10. For individuals with measurable metastatic disease on maintenance or off therapy, CT of involved body area no more frequently than every 3 months. After 10 years, surveillance should be as clinically indicated.[1]

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Bronchopulmonary or Thymic Carcinoid – Initial Staging (ONC-15.6)

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Indication	Medically Necessary Imaging Study
Initial diagnosis	If not already done: • CT Chest with contrast (CPT® 71260) • CT Abdomen with contrast (CPT® 74160) or without and with contrast (CPT® 74170) or MRI Abdomen (CPT® 74183) without and with contrast
Inconclusive CT or MRI scans	ONE of the following: • Octreotide scan (ANY ONE of the following): ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, or 78804 **AND** ◦ Any one of the following SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832 • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE

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Indication	Medically Necessary Imaging Study
ANY of the following: • Markers fail to normalize after complete resection AND CT/MRI and somatostatin-receptor based study are negative • Biopsy-proven neuroendocrine tumor of unknown primary site AND CT/MRI and somatostatin-receptor based study are negative	• FDG-PET/CT (CPT® 78815) OR PET/MRI (CPT® 78813 and CPT® 76498)

Evidence Discussion

More than 80% of lung carcinoids are diagnosed at Stage I or II with the most common sites of metastases being liver, bone, and lung.[14] Most thymic carcinoids are diagnosed at Stage III or IV with the most common sites of metastases being pleura, pericardium, bone, lung, and liver. Recommended initial imaging includes CT Chest with contrast and CT or MRI of the abdomen. Imaging of the brain, pelvis or osseous structures is based on signs/symptoms of disease. If the CT/MRI is inconclusive, Octreotide scan or SSR-based PET can be completed. In the setting where markers fail to normalize after surgery AND CT/MRI and SSR based study are negative OR there is biopsy proven NET of unknown origin AND CT/MRI and SSR based study are negative, FDG-based PET is indicated for the concern of a higher-grade NET being present.[10,11]

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Bronchopulmonary or Thymic Carcinoid – Restaging/Recurrence (ONC-15.7)

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Indication	Medically Necessary Imaging Study
All after surgical resection	• See: **Bronchopulmonary or Thymic Carcinoid - Surveillance (ONC-15.8)**
Unresectable/metastatic disease on treatment with somatostatin analogues	• CT or MRI of involved body area no more frequently than every 3 months
Unresectable/metastatic disease on treatment with chemotherapy	• CT or MRI of involved body area every 2 cycles (6 to 8 weeks)
Progression of symptoms or elevation of tumor markers	• CT Chest without (CPT® 71250) or CT Chest with contrast (CPT® 71260) And ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast

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Indication	Medically Necessary Imaging Study
Continued suspicion for recurrence with negative or inconclusive CT or MRI	ONE of the following: • Octreotide scan ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, or 78804 **AND** ◦ Any one of the following SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832 • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498)with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE

Evidence Discussion

For individuals with unresectable/metastatic disease, CT imaging of involved body area is recommended every 3 months if on somatostatin therapy versus every 2 cycles (6-8 weeks) if on chemotherapy. CT Chest and CT/MRI Abdomen/Pelvis is indicated for progression of symptoms or elevation of tumor markers while Octreotide scan or SSR PET if conventional imaging is negative/inconclusive.[10,11]

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Bronchopulmonary or Thymic Carcinoid – Surveillance (ONC-15.8)

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Indication	Medically Necessary Imaging Study
Carcinoid tumors of lung or thymus	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) and CT Abdomen with contrast (CPT® 74160) once at 3 to 12 months post-resection and then annually up to year 10
Unresected primary tumors being monitored with observation alone	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) and CT Abdomen with contrast (CPT® 74160) once at 3 to 12 months from initial diagnosis then annually up to year 10
Asymptomatic lung or thymus distant metastasis on observation alone	Every 3 months for 1 year, and then every 6 months: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Measurable metastatic disease on maintenance treatment or off therapy	• CT Chest with contrast (CPT® 71260) • CT of involved body area no more frequently than every 3 months

Evidence Discussion

For carcinoid tumors of the lung or thymus, prognosis varies. In typical lung carcinoid, 5-year and 10-year overall survival (OS) is approximately 81% to 96% and 74% to 91% respectively for both node negative and node positive disease. With atypical histology, 5-year OS is 82% to 90% in node negative disease and 58-71% in node positive disease. Thymic carcinoid 5-year OS is <50%. Due to lack of long-term follow-up imaging studies, it is recommended to minimize risk of radiation exposure using CT or MRI.[14] In individuals with resected disease, CT Chest 3-12 months post resection and then annually for up to 3 years then every 2 years up to year 10. Unresected primary tumors on observation follow a similar schedule with CT Chest 3-12 months from initial diagnosis and then annually for up to 3 years then every 2 years up to year 10. Measurable metastatic disease on maintenance therapy or observation undergoes CT of

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involved body area no more frequently than every 3 months. After 10 years, surveillance should be as clinically indicated.[1]

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Adrenal Tumors – Suspected/Diagnosis (ONC-15.9)

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- See: **Adrenal Cortical Lesions (AB-16.1)** in the Abdomen Imaging Guidelines for evaluation of indeterminate adrenal masses.
- Adrenal tumors that involve the adrenal medulla or neural crest tissue outside the adrenal gland include pheochromocytoma, paraganglioma, and paraganglioneuroma.
  - These tumors are imaged according to sections **ONC-15.10 through ONC-15.12**.
  - Malignant adrenal tumors that involve the adrenal cortex are addressed in **Adrenocortical Carcinoma (ONC-15.13).**
- Adrenocortical carcinoma is imaged according to **Adrenocortical Carcinoma (ONC-15.13)**.
- If concern for genetic predisposition syndrome such as MEN, neurofibromatosis, or Von Hippel-Lindau disease, see screening recommendations in **Screening Imaging and Cancer Predisposition Syndromes (PEDONC-2)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Adrenal Tumors – Initial Work-up/Staging (ONC-15.10)

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- This guideline can be applied to **any primary site** (including beyond adrenal gland) for pheochromocytoma, paraganglioma, or paraganglioneuroma.

Indication	Medically Necessary Imaging Study
For ANY of the following: • Pheochromocytoma • Paraganglioma • Paraganglioneuroma	If not already done: • CT Chest without (CPT® 71250) or CT Chest with contrast (CPT® 71260) And ONE of the following (if not already done): • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • CT with contrast or MRI without and with contrast of primary site (i.e., neck) and any other symptomatic body areas
Hypercortisolemia with ANY of the following: • Tumor greater than 4 cm • Inhomogeneous • Irregular margins • Local invasion • Other malignant imaging characteristics	ANY or ALL of the following: • FDG PET/CT (CPT® 78815) • CT Abdomen with contrast (CPT® 74160) or CT Abdomen without and with contrast (CPT® 74170) or MRI Abdomen without and with contrast (CPT® 74183)

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Indication	Medically Necessary Imaging Study
Continued suspicion with negative/ inconclusive CT or MRI	ONE of the following: • Octreotide or MIBG scan: ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, 78804 **AND** ◦ Any one of the following SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832 • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE
All above studies done and negative/ inconclusive	• FDG PET/CT scan (CPT® 78815)

Evidence Discussion

Radiologic evaluation of adrenal tumors should follow biochemical conformation of the diagnosis of pheochromocytoma and paraganglioglioma. Ninety-five percent of this group of tumors occur in the abdomen and pelvis. Paragangliomas account for 15% of this group of tumors. The initial work-up/staging of this group of tumors includes CT Chest (with contrast or without contrast) as approximately 10% of paragangliomas are found in the chest and pheochromocytoma has a potential to metastasize to chest. CT imaging with or without and with contrast or MRI imaging without and with contrast of the abdomen, pelvis and any other symptomatic body areas are indicated for initial staging. Hypercortisolemia can be caused by benign and malignant tumors. NCCN stated, "Malignancy should be suspected if the tumor is larger than 4 centimeters or is inhomogeneous with irregular margins and/or has local invasion and other malignant imaging characteristics" and went on to recommend imaging with PET/CT, CT Chest, and CT or MRI abdominal imaging.[1]

Functional imaging is a valuable problem-solving tool when there is continued suspicion for this group of tumors with inconclusive or negative findings on CT/MRI. ^{123}I/ ^{131}I-MIBG sensitivity is higher for detecting pheochromocytoma than for detecting paraganglioma, at 88% and 67%, respectively. Another approach to functional imaging utilizes ^{68}Ga-DOTA-somatostatin analogs including DOTATOC, DOTANOC, and DOTATATE (SSR-PET/CT. In meta-analyses, the sensitivity of 68Ga-DOTA-somatostatin

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analogs (93%) is superior to ^{18}F-FDG (74%), and ^{123}I/ ^{131}I-MIBG (38%). Imaging with ^{111}In-pentetreotide (octreotide) (24%) is less sensitive that imaging with ^{68}Ga-DOTA-somatostatin analogs. Of note, functional imaging using ^{18}F-FDG PET/CT can be useful for evaluation in the scenario that CT/MRI and other functional imaging studies are negative/inconclusive.[1,15-18]

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Adrenal Tumors – Restaging/Recurrence (ONC-15.11)

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- This guideline can be applied to **any primary site** (including beyond adrenal gland) for pheochromocytoma, paraganglioma, or paraganglioneuroma.

Indication	Medically Necessary Imaging Study
If surgery is primary therapy	See: **ONC-15.12** for surveillance recommendations
Recurrence, progression of symptoms, or elevation of tumor markers	• CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) • CT with contrast of primary site (i.e., neck) and involved areas And ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Continued suspicion for recurrence with negative or inconclusive CT or MRI	ONE of the following: • Octreotide or MIBG scan (ANY ONE of the following): ◦ Any one of the following planar imaging codes - CPT® 78801, 78802, or 78804 **AND** ◦ Any one of the following SPECT/SPECT-CT codes - CPT® 78803, 78830, 78831, 78832 • PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}Cu-DOTATATE

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Indication	Medically Necessary Imaging Study
To assess candidacy for peptide receptor radionuclide therapy (PRRT) with Lutetium ^{177}Lu-dotatate	• PET/CT (CPT® 78815) or PET/MRI (CPT® 78813 and CPT® 76498) with any ONE of the following SSR radiotracers: ◦ ^{68}Ga-DOTATATE ◦ ^{68}Ga-DOTATOC ◦ ^{64}CU-DOTATATE
For ANY of the following: • All above studies done and negative/ inconclusive • Hypercortisolemia with inconclusive findings on CT • Pheochromocytoma bone-dominant disease • Paraganglioma bone-dominant disease	• FDG-PET/CT scan (CPT® 78815)

Evidence Discussion

In individuals who underwent surgery for resection of localized disease, CT Abdomen with contrast is supported within the first year post resection. Follow-up imaging is based on surveillance recommendations noted in Adrenal Tumors – Surveillance (ONC 15.12). In individuals who develop recurrence, progression of symptoms or elevation of tumor markers, the extent of disease must be determined with CT/MRI imaging. CT chest (with contrast or without contrast), CT imaging with or without and with contrast or MRI imaging without and with contrast of the abdomen, pelvis and any any other symptomatic body areas are indicated. Functional imaging with ^{111}In-pentetreotide (octreotide scan) or ^{68}Ga-DOTA-somatostatin analog PET is a valuable problem solving tool when there is continued suspicion of recurrence with inconclusive or negative findings on CT/MRI. Further evaluation using ^{18}F-FDG PET/CT can be useful for evaluation in the scenario that CT/MRI and other functional imaging studies are negative/inconclusive. ^{18}F-FDG PET/CT is also supported by NCCN for the evaluation of hypercortisolemia with inconclusive findings on CT, pheochromocytoma bone-dominant disease, and paraganglioma bone-dominant disease.[1,15-18]

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Adrenal Tumors – Surveillance (ONC-15.12)

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- This guideline can be applied to **any primary site** (including beyond adrenal gland) for pheochromocytoma, paraganglioma, paraganglioneuroma, or hypercortisolemia.

Indication	Medically Necessary Imaging Study
All individuals	Once within 3-12 months post resection and then annually for 10 years: • CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and 72197) • CT with contrast of primary site (i.e., neck) and other involved body areas
Measurable metastatic disease being observed off therapy or on maintenance treatment	• CT of involved body area no more frequently than every 3 months for up to 5 years after completion of definitive therapy and annually thereafter

Evidence Discussion

In individuals who had resectable disease, CT imaging of the chest and either CT or MR imaging of the abdomen, pelvis, and any other previously involved body areas are indicated once within 3-12 months post resection then annually for 10 years. In individuals with measurable metastatic disease being observed off therapy or on maintenance therapy are at greater risk for recurrence. In addition to the imaging schedule noted above, imaging of the involved body area is indicated with greater frequency (3 months) for up to 5 years followed by annual imaging.[1,15-18]

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Adrenocortical Carcinoma (ONC-15.13)

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Indication	Medically Necessary Imaging Study
Initial Staging	• FDG PET/CT (CPT® 78815) • CT Chest without (CPT® 71250) or CT Chest with contrast (CPT® 71260) And ONE of the following (if not already done): • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Suspected recurrence	• CT Chest without (CPT® 71250) or CT Chest with contrast (CPT® 71260) And ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen and Pelvis without and with contrast (CPT® 74178) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
• Inconclusive findings on conventional imaging	• FDG PET/CT (CPT® 78815)
Surveillance after complete response to definitive treatment	Annually for 5 years: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen (CPT® 74183) without and with contrast • CT of other involved body areas with contrast

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Indication	Medically Necessary Imaging Study
Measurable metastatic disease on maintenance therapy or being monitored off therapy	Every 3 months for up to 5 years after completion of definitive therapy: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • CT with contrast of other involved body areas

Evidence Discussion

Adrenocortical carcinoma (ACC) is a rare, aggressive tumor arising from the adrenal cortex. Most cases of ACC are sporadic; however, ACC has been described as a component of hereditary cancer syndromes that include Li-Fraumeni syndrome, Beckwith-Weidemann syndrome and multiple endocrine neoplasia type 1 (MEN1). Recommended imaging guidelines for screening of individuals with these syndromes are summarized in Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2). ACCs that secrete excess adrenal hormones are classified as functional tumors and occur in 15-30% of cases in adults. Those patients with functional tumors present with Cushing's syndrome and/or virilization. The majority of adults with non-functioning ACCs present symptomatically with abdominal pain or flank pain; however, these tumors may present asymptomatically as a large palpable intra-abdominal mass or incidentally as a small adrenal mass. Approximately 30% of ACCs present with metastatic disease in lymph nodes, lung, live and bone. ACCs may also present with invasion of adjacent structures and with venous extension.

CT and MRI cross-sectional imaging are the standard imaging modalities used for the evaluation of ACC. Due to the measurable risk for widely metastatic disease at initial presentation, cross-section imaging of the chest and abdomen/pelvis is indicated. CT Chest (with contrast or without contrast) is indicated to evaluate for lung metastases. Cross-sectional imaging of the abdomen/pelvis using CT Abdomen/Pelvis (with or with and without contrast) or MRI Abdomen as well as MRI Pelvis (with and without contrast) are indicated for characterization of the primary tumor and evaluation for metastatic disease. If recurrence is suspected, the same imaging studies should be completed as was done for initial staging. Additional imaging may be needed to assess other suspicious sites of disease based on clinical signs/symptoms.

NCCN endorsed the use of FDG PET/CT in addition to diagnostic CT or MRI for initial imaging of suspected adrenocortical carcinoma. FDG PET/CT scan is a valuable

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problem-solving tool; thus, FDG PET/CT is supported to characterize inconclusive findings CT/MRI.

CT imaging is the standard approach in surveillance. In individuals who had complete response to definitive therapy, the recommendation is annual imaging for 5 years after completion of treatment as the rate of recurrence is <25% and overall survival rate approaches 74% to 95% at 5 years. In individuals with metastatic disease on maintenance therapy or being monitored off treatment, the frequency of CT imaging is shortened and repeated at 3 month intervals for up to 5 years.[1,19-22]

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Colorectal and Small Bowel Cancer (ONC-16)

Guideline

Colorectal Cancer – General Considerations (ONC-16.0)
Colorectal Cancer – Suspected/Diagnosis (ONC-16.1)
Colorectal Cancer – Initial Work-up/Staging (ONC-16.2)
Colorectal Cancer – Restaging/Recurrence (ONC-16.3)
Colorectal Cancer – Surveillance/Follow-up (ONC-16.4)
Small Bowel Cancer – Initial Work-up/Staging (ONC-16.5)
Small Bowel Cancer – Restaging/Recurrence (ONC-16.6)
Small Bowel Cancer – Surveillance/Follow-up (ONC-16.7)
References (ONC-16)

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Colorectal Cancer – General Considerations (ONC-16.0)

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- Neuroendocrine tumors of the bowel are covered in: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)**.
- Appendiceal adenocarcinoma (including pseudomyxoma peritonei) follows imaging guidelines for colorectal cancer.
- For squamous cell carcinoma of the rectum, see: **Anal Carcinoma (ONC-24)**.

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Colorectal Cancer – Suspected/ Diagnosis (ONC-16.1)

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- See: **GI Bleeding (AB-22)** or **CT Colonography (CTC) (AB-25.1)** in the Abdomen Imaging Guidelines for evaluation of suspected colorectal malignancies.
- See: **Abnormal Findings on Endoscopy/Colonoscopy (AB-13.3)** in the Abdomen Imaging Guidelines for evaluation of abnormal findings on endoscopy/colonoscopy.

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Colorectal Cancer – Initial Work-up/ Staging (ONC-16.2)

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Indication	Medically Necessary Imaging Study
Carcinoma within a polyp that is completely removed	• No advanced imaging
• Biopsy-proven invasive adenocarcinoma	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)
• Further evaluation of an inconclusive liver lesion seen on CT • Potentially resectable liver metastases	• MRI Abdomen without and with contrast (CPT® 74183)
Rectal adenocarcinoma	• MRI Pelvis without and with contrast (CPT® 72197) or MRI Pelvis without contrast (CPT® 72195) (can be obtained in addition to CT scans for initial staging)
Rectal adenocarcinoma with ANY one of the following: • Rectal MRI is contraindicated • Rectal MRI is inconclusive • Superficial lesions	• Endorectal ultrasound (CPT® 76872)
ONE of the following: • Isolated metastatic lesion(s) on other imaging and individual is a candidate for aggressive surgical resection or other localized treatment to metastasis for curative intent • Inconclusive conventional imaging	• PET/CT (CPT® 78815)

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Evidence Discussion

- Carcinoma within a polyp that is completely resected does not require advanced imaging.[1]
- Invasive cancer at any stage requires advanced imaging with CT of Chest/Abdomen/Pelvis with contrast needed to adequately visualize lung, nodal, and especially liver lesions with MRI of abdomen appropriate in the event of unclear liver lesions. The chest CT can identify lung metastases, which occur in approximately 4% - 9% of individuals with colon and rectal cancer.[11]
- Rectal cancer requires additional dedicated imaging with MRI Pelvis that is superior to CT imaging to locally stage this form of colon cancer with endorectal/endoscopic ultrasound (EUS), providing additional staging information when MRI is contraindicated/inconclusive/superficial. MRI is considered superior to EUS due to the latter's limitations in regard to high/bulky tumors, tumor deposits, or vascular invasion.[12]
- PET/CT is reserved for inconclusive CT/MRI imaging and to confirm isolated metastases that are amenable to definitive localized treatment with curative intent.[1]

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Colorectal Cancer – Restaging/ Recurrence (ONC-16.3)

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Indication	Medically Necessary Imaging Study
• Complete resection • Individuals receiving post-operative adjuvant chemotherapy	• See: **Surveillance/Follow-up (ONC-16.4)**
Recurrence suspected (i.e., signs or symptoms, elevated CEA)	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast
After completion of planned neoadjuvant therapy	Prior to surgical resection in individuals with non-metastatic rectal cancer: • CT Chest with contrast (CPT® 71260) and Any ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) and MRI Pelvis without and with contrast (CPT® 72197)
Unresected primary disease or metastatic disease on chemotherapy	Every 2 cycles of chemotherapy treatment and at the completion of chemoradiotherapy: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of other involved or symptomatic areas
• Further evaluation of an inconclusive liver lesion seen on CT • Potentially resectable liver metastases	• MRI Abdomen without and with contrast (CPT® 74183)

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Indication	Medically Necessary Imaging Study
ONE of the following: • Postoperative elevated or rising CEA or LFTs with negative recent conventional imaging • Isolated metastatic lesion(s) on other imaging and individual is a candidate for aggressive surgical resection or other localized treatment to metastasis for curative intent • Differentiate local tumor recurrence from postoperative and/or post-radiation scarring	• PET/CT (CPT® 78815)
New or worsening pelvic pain and recent CT imaging negative or inconclusive	• MRI Pelvis without and with contrast (CPT® 72197)

Evidence Discussion

- If recurrence is suspected, CT Chest/Abdomen/Pelvis with contrast is the first line of imaging.[1]
- NCCN stated that ctDNA is not recommended for surveillance.[1]
- Upon completion of neoadjuvant therapy, to ensure the cancer has not progressed prior to definitive surgery, repeat imaging of the chest, abdomen and pelvis is medically necessary.[1]
- With measurable disease or unresected primary disease on chemotherapy, CT imaging is medically necessary every 2 cycles of treatment to assess response and appropriateness to continue the same treatment or change to new therapy.[1]
- MRI Abdomen is medically necessary for inconclusive liver lesion or to better define resectability.[1]
- PET/CT may be used in specific situations to better determine cancer recurrence if CT/MRI is inconclusive. These results may allow aggressive interventions (e.g., surgery, radiation, liver directed therapy) to take place with goal of cure, explain elevated or rising CEA level or LFTs with negative conventional imaging and can also be useful to differentiate surgical/radiation scarring from cancer. A systemic review and meta-analysis of 11 studies using PET/CT with elevated CEA and negative CT Chest/Abdomen/Pelvis showed a sensitivity of 94% and specificity of 77% in detection of tumor recurrence.[1,6]

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Colorectal Cancer – Surveillance/Follow-up (ONC-16.4)

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Indication	Medically Necessary Imaging Study
Colon and rectal adenocarcinoma: • Stage I	• No routine advanced imaging
Colon and rectal adenocarcinoma: • Stage II-III	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) after completion of surgery and then annually for 5 years
Colon and rectal adenocarcinoma: • Stage IV or distant metastatic disease (post definitive treatment of all measurable disease or being observed off therapy)	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) every 6 months for 2 years and then annually for 3 years
Measurable metastatic disease on maintenance therapy	Every 3 months for up to 5 years after completion of active treatment: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Rectal cancer treated with transanal excision alone	Any one of the following every 6 months for 5 years: • Endorectal ultrasound (CPT® 76872) • MRI Pelvis without and with contrast (CPT® 72197)
Stage II-III rectal cancer treated with chemoradiation alone (no surgical treatment)	In addition to the above stage-specific surveillance: • MRI Pelvis (CPT® 72197) without and with contrast every 6 months for 3 years

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Indication	Medically Necessary Imaging Study
Pseudomyxoma peritonei	ONE of each of the following, every 3 months for first year, then every 6 months for 4 more years: • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast

Evidence Discussion

- Up to 95% of recurrences occur in the first 5 years after surgery with 80% occurring in the first 3 years, many of which can still be cured supporting the need for CT imaging.
- Given the >90% cure rate with Stage I cancer, routine imaging is not usually indicated.[1]
- Given the recurrence rate between 20%-30% in Stage II-III disease, CT Chest/Abdomen/Pelvis is indicated after completion of surgery (new baseline) and annually for 5 years. More frequent imaging has a lower level of support based on the following trials: FACS, COLOFOL, CEAwatch, and PRODIGE 13.[15]
- For Stage IV cancer status post definitive treatment or being observed off therapy, CT imaging is recommended every 6 months for 2 years then annually for 3 years.[15]
- For Stage IV cancer that is measurable and on maintenance therapy, CT imaging is recommended every 3 months for up to 5 years after completion of active treatment.
- For rectal cancer treated with transanal excision alone, due to higher risk of local recurrence, endorectal ultrasound (EUS) should be performed every 6 months for 5 years with pelvic MRI reserved for abnormal findings on EUS or EUS can't be performed as well as new signs/symptoms concerning for local recurrence.[2]
- Treatment with chemoradiation alone (no surgery) is becoming more common in the setting of rectal cancer in the presence of a complete response to therapy with 5-year survival rates exceeding 80% in several trials. These individuals require additional follow-up studies to include MRI Pelvis every 6 months for 3 years as well as digital rectal exam (DRE)/endoscopy every 3-4 months for 2 years and every 6 months for 3 more years to assess for local recurrence even without signs/symptoms of recurrence.[2]
- Pseudomyxoma peritonei is a condition associated with appendiceal cancer and requires close imaging follow-up due to high risk of recurrence, thus imaging with CT/MRI is indicated every 3 months for year 1 and every 6 months for 4 more years.[1]

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Small Bowel Cancer – Initial Work-up/ Staging (ONC-16.5)

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This section provides imaging guidelines for small bowel adenocarcinoma arising from the duodenum, jejunum, and ileum.

Indication	Medically Necessary Imaging Study
Carcinoma within a polyp that is completely removed	• No advanced imaging
Invasive adenocarcinoma	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast ◦ MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) if CT is inconclusive or cannot be performed

Evidence Discussion

- Cancerous polyps that are completely removed do not require imaging.[1]
- Invasive cancer at any stage requires advanced imaging with CT of Chest/Abdomen/ Pelvis with contrast needed to adequately visualize lung, nodal, and especially liver lesions with MRI of abdomen/pelvis appropriate in the event the CT is inconclusive or cannot be performed.[1]

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Small Bowel Cancer – Restaging/ Recurrence (ONC-16.6)

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Indication	Medically Necessary Imaging Study
Complete resection	• See Surveillance below
Recurrence suspected	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast
Unresected primary disease or metastatic disease on chemotherapy	Every 2 cycles of chemotherapy: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Further evaluation of an inconclusive liver lesion seen on CT	• MRI Abdomen without and with contrast (CPT® 74183)
ONE of the following: • Postoperative elevated or rising CEA or LFTs with negative recent conventional imaging • Isolated metastatic lesion(s) on other imaging and individual is a candidate for aggressive surgical resection or other localized treatment to metastasis for curative intent	• PET/CT (CPT® 78815)

Evidence Discussion

- If recurrence is suspected, CT Chest/Abdomen/Pelvis with contrast is the first line of imaging.
- With measurable disease or unresected primary disease on chemotherapy, CT imaging is medically necessary every 2 cycles of treatment to assess response and appropriateness to continue the same treatment or change to new therapy.
- MRI Abdomen is medically necessary for inconclusive liver lesion on CT

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- PET/CT may be used in specific situations to better determine cancer recurrence if CT/MRI is inconclusive. These results may allow aggressive interventions (surgery, radiation, liver directed therapy) to take place with goal of cure and also explain elevated or rising CEA level or LFTs with negative conventional imaging.[3]

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Small Bowel Cancer – Surveillance/ Follow-up (ONC-16.7)

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Indication	Medically Necessary Imaging Study
Stage I-III	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast after completion of surgery, and then annually for 5 years
Stage IV - Metastatic disease (post definitive treatment of all measurable disease, or being observed off therapy)	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast every 6 months for 2 years and then annually for 3 years
Measurable metastatic disease on maintenance therapy	Every 3 months for up to 5 years after completion of active treatment: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)

Evidence Discussion

- Stage I, II and III cancers undergo CT Chest/Abdomen/Pelvis at completion of surgery then annually for 5 years, similar to Colorectal guidelines due to lack of data regarding optimal approach.[3,15]
- For Stage IV cancer s/p definitive treatment or being observed off therapy, CT imaging is recommended every 6 months for 2 years then annually for 3 years.
- For Stage IV cancer that is measurable and on maintenance therapy, CT imaging is recommended every 3 months for up to 5 years after completion of active treatment.

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Renal Cell Cancer (RCC) (ONC-17)

Guideline

Renal Cell Cancer (RCC) – General Considerations (ONC-17.0)
Renal Cell Cancer (RCC) – Suspected/Diagnosis (ONC-17.1)
Renal Cell Cancer (RCC) – Initial Work-up/Staging (ONC-17.2)
Renal Cell Cancer (RCC) – Restaging/Recurrence (ONC-17.3)
Renal Cell Cancer (RCC) – Surveillance (ONC-17.4)
References (ONC-17)

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Renal Cell Cancer (RCC) – General Considerations (ONC-17.0)

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- PET is considered not medically necessary for initial diagnosis, staging or restaging of renal cell cancer.
- A minority of adult individuals with renal cell cancer (RCC) will have translocations in TFE3 or TFEB, which have a different natural history than “adult type” RCC. Individuals of any age with TFE3 or TFEB translocated RCC should be imaged according to guidelines in **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Individuals of any age with a Wilms tumor should be imaged according to guidelines in section **Unilateral Wilms Tumor (UWT) (PEDONC-7.2)** or **Bilateral Wilms Tumor (BWT) (PEDONC-7.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Oncocytoma in individuals of all ages should be imaged according to these guidelines.

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Renal Cell Cancer (RCC) – Suspected/ Diagnosis (ONC-17.1)

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Indication	Medically Necessary Imaging Study
• Solitary renal mass suspicious for renal cell cancer	• See: **Indeterminate Renal Lesion (AB-35.1)** in the Abdomen Imaging Guidelines for evaluation of suspected renal malignancies • CT Chest with contrast with (CPT® 71260) or without contrast (CPT® 71250)

Evidence Discussion

In individuals with RCC or suspicion of RCC, complete staging is typically completed with chest radiography (CXR) or chest CT. CT Chest is preferred.[1,17]

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Renal Cell Cancer (RCC) – Initial Work-up/Staging (ONC-17.2)

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Indication	Medically Necessary Imaging study
All individuals	If not done previously: • CT Chest with (CPT® 71260) or without (CPT® 71250) contrast • CT Abdomen and Pelvis, contrast as requested
ANY of the following: • Extension of tumor into the vena cava by other imaging • Inconclusive findings on CT	• MRI Abdomen without and with contrast (CPT® 74183)
Bone pain	• Bone scan (CPT® 78306) (See: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)
EITHER of the following: • Signs/symptoms suspicious for brain metastases • Newly diagnosed stage IV/ metastatic RCC	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

American Urological Association (AUA) and the National Comprehensive Cancer Network (NCCN) guidelines stated that pre- and post-contrast enhanced axial imaging, either CT or MRI, is the ideal imaging technique for the diagnosis and staging of clinically localized renal masses. Contrast-enhanced abdominal imaging best characterizes the mass, provides information regarding both the affected and unaffected renal unit, can assess extrarenal tumor spread (venous invasion or regional lymph nodes), and evaluates the adrenal glands and other abdominal organs for visceral metastasis.[1,20,21]

Masses initially diagnosed by ultrasound should be confirmed with pre- and post-contrast enhanced imaging.[1]

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In individuals with RCC or suspicion of RCC, complete staging is typically completed with chest radiography (CXR) or chest CT. Chest CT is preferred.[1] Bone scans should be reserved primarily for individuals with bone pain as the prevalence of osseous metastasis for localized renal cell cancer has been shown to be low in individuals without symptoms suggestive of osseous metastasis. Brain imaging is reserved for individuals with neurologic symptoms, as most individuals with metastasis to the central nervous system are symptomatic, or individuals with newly diagnosed metastatic disease.[1]

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Renal Cell Cancer (RCC) – Restaging/ Recurrence (ONC-17.3)

ON.RC.0017.3.A
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Indication	Medically Necessary Imaging Study
Unresectable disease or metastatic disease on systemic therapy	Every 2 cycles of treatment (commonly every 6 to 8 weeks): • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis, contrast as requested (CPT® 74177, CPT® 74178, or CPT® 74176) • CT with contrast of other involved or symptomatic areas
Recurrence suspected	• CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis, contrast as requested (CPT® 74177, CPT® 74178, or CPT® 74176)
EITHER of the following: • Biopsy-proven recurrent/ metastatic disease • Signs or symptoms concerning for brain metastases	• MRI Brain without and with contrast (CPT® 70553)

Evidence Discussion

Individuals presenting with findings suggesting a new renal primary or local recurrence of renal malignancy should undergo metastatic evaluation including chest and abdominal imaging.[1,20,21] The most common sites of distant metastasis include lung, bone, retroperitoneal and mediastinal nodes, liver, brain, or multiple sites.[29,30]

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Renal Cell Cancer (RCC) – Surveillance (ONC-17.4)

ON.RC.0017.4.A
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Indication	Medically Necessary Imaging Study
RCC on active surveillance of renal mass <1 cm	ALL of the following, once within 6 months of surveillance initiation and annually thereafter: • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150) or MRI Abdomen without and with contrast (CPT® 74183) • Chest x-ray ◦ CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) is indicated instead of chest x-ray for any of the following: ▪ Known prior thoracic disease ▪ New or worsening pulmonary symptoms ▪ New or worsening chest x-ray findings
RCC on active surveillance of renal mass ≥1 cm	ALL of the following, every 3 months for year 1, every 6 months for years 2 and 3 and annually thereafter: • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150) OR MRI Abdomen without and with contrast (CPT® 74183) • Chest x-ray ◦ CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) is indicated instead of chest x-ray for any of the following: ▪ Known prior thoracic disease ▪ New or worsening pulmonary symptoms ▪ New or worsening chest x-ray findings

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Indication	Medically Necessary Imaging Study
Follow up after post-ablation therapy of RCC	EITHER of the following, at 1 to 3 months, 6 months, and 12 months post-ablation and then annually thereafter: • CT Abdomen, contrast as requested (CPT® 74170), CPT® 74160, or CPT® 74150 or • MRI Abdomen without and with contrast (CPT® 74183) AND Annually for 5 years: • Chest x-ray or CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250)
Follow up after SBRT	ONE of each of the following, every 3 months for 1 year, every 6 months in year 2, every 9 months in years 3-4, and once in year 5 (to complete 5 years surveillance): • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150) or MRI Abdomen without and with contrast (CPT® 74183)
Stage I RCC, after partial or radical nephrectomy	ONE of each of the following, 3 to 12 months post-resection: • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150) or MRI Abdomen without and with contrast (CPT® 74183) ONE of each of the following, annually for 5 years: • CT Chest with (CPT® 71260) or CT Chest without (CPT® 71250) contrast • MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150)

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Indication	Medically Necessary Imaging Study
Stage II RCC, post-nephrectomy	ONE of each of the following, 3 to 6 months post-resection: • CT Chest with (CPT® 71260) or CT Chest without (CPT® 71250) contrast • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150), or CT Abdomen and Pelvis, contrast as requested (CPT® 74177, CPT® 74176, or CPT® 74178), or MRI Abdomen without and with contrast (CPT® 74183) +/- MRI Pelvis without and with contrast (CPT® 72197) ONE of each of the following, every 6 months for 2 years, then annually until year 5: • CT Chest with (CPT® 71260) or CT Chest without (CPT® 71250) contrast • MRI Abdomen without and with contrast (CPT® 74183) +/- MRI Pelvis without and with contrast (CPT® 72197), or CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150), or CT Abdomen and Pelvis, contrast as requested (CPT® 74177, CPT® 74176, CPT® 74178)
Stage III RCC, post-nephrectomy	ONE of each of the following, 3 to 6 months post-resection: • CT Chest with (CPT® 71260) or CT Chest without (CPT® 71250) contrast • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150), or CT Abdomen and Pelvis, contrast as requested (CPT® 74177, CPT® 74176, or CPT® 74178), or MRI Abdomen without and with contrast (CPT® 74183) +/- MRI Pelvis without and with contrast (CPT® 72197) ONE of each of the following, every 3 months for 3 years, then annually to year 5: • CT Chest with (CPT® 71260) or CT Chest without (CPT® 71250) contrast • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150), or CT Abdomen and Pelvis, contrast as request (CPT® 74177, CPT® 74176, CPT® 74178), or MRI Abdomen without and with contrast (CPT® 74183)

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Indication	Medically Necessary Imaging Study
Stage IV/metastatic disease on maintenance therapy or being observed off therapy	ONE of each of the following, every 3 months for up to 5 years after completion of active treatment: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen, contrast as requested (CPT® 74170, CPT® 74160, or CPT® 74150), or CT Abdomen and Pelvis contrast as requested (CPT® 74177, CPT® 74176, or CPT® 74178), or MRI Abdomen without and with contrast (CPT® 74183) +/- MRI Pelvis without and with contrast (CPT® 72197)

Evidence Discussion

Active surveillance Stage T1a:

Active surveillance entails serial abdominal imaging in order to study the growth rate of the tumor and it is recommended that abdominal imaging (CT or MRI with contrast) within 6 months from initiation of active surveillance; subsequent imaging (with CT, MRI, or ultrasound [US]) may be performed annually thereafter. CT and MRI have both been found to accurately predict pathologic tumor size in a retrospective analysis.[23] Therefore, best clinical judgment should be used in choosing the imaging modality. A chest x-ray or chest CT at baseline and annually as clinically indicated to assess for pulmonary metastases. Repeat chest imaging can be considered if intervention is being contemplated.[1]

Follow up after ablative therapy for Stage T1a:

An abdominal imaging, either CT or MRI with and without IV contrast (unless otherwise contraindicated), at 1 through 6 months to assess treatment response, followed by annual abdominal CT or MRI (preferred) for 5 years or longer as clinically indicated. If the individual cannot receive IV contrast, MRI is preferred.[1]

Follow up after partial or radical nephrectomy for Stages 1-2:

Stage I RCC, it is recommended that abdominal CT or MRI (preferred) within 3 to 12 months following renal surgery, then annually for up to 5 years or longer as clinically indicated. For individuals with stage II RCC, it is recommended to increase in abdominal imaging frequency, with baseline abdominal CT or MRI (preferred) every 6 months for 2 years, then annually for up to 5 years or longer, as clinically indicated.[1]

It is also recommended that yearly chest x-ray or CT for at least 5 years and as clinically indicated thereafter.[1]

Follow up for individuals with Stage 3 RCC:

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It is recommended to obtain a baseline abdominal CT or MRI within 3 to 6 months following surgery, followed by CT, MRI (preferred), or US every 3 to 6 months for at least 3 years, and annually thereafter for up to 5 years.[1]

A baseline chest CT within 3 to 6 months following surgery is also recommended along with continued imaging (CT preferred) every 3 to 6 months for at least 3 years, and annually thereafter for up to 5 years.[1]

CT is the preferred modality for those with a high risk of recurrence.

Follow up for an individual with relapse or unresectable disease or Stage 4 RCC:

It is recommended to obtain chest, abdominal and pelvic imaging at baseline and then as clinically indicated based on clinical status, and therapeutic schedule.[1]

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Transitional Cell Cancer (ONC-18)

Guideline

Transitional Cell Cancer – General Considerations (ONC-18.0)
Transitional Cell Cancer – Suspected/Diagnosis (ONC-18.1)
Transitional Cell Cancer – Initial Work-up/Staging (ONC-18.2)
Transitional Cell Cancer – Restaging/Recurrence (ONC-18.3)
Transitional Cell Cancer – Surveillance/Follow-up (ONC-18.4)
References (ONC-18)

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Transitional Cell Cancer – General Considerations (ONC-18.0)

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- Transitional cell cancers can include: tumors of the bladder, ureters, prostate, urethra, or renal pelvis. For primary cancer of the kidney, see: **Renal Cell Cancer (RCC) (ONC-17)**.
- Most common histology of bladder cancer is transitional cell (TCC) or urothelial carcinoma (UCC). Rare histologies include adenocarcinoma, squamous cell (imaged according to **Transitional Cell Cancer (ONC-18)**), or small cell (imaged according to **Extrathoracic Small Cell and Large Cell Neuroendocrine Tumors (ONC-31.8)**).
- Urachal cancer is rare type of bladder cancer; the most common histology is adenocarcinoma. These are imaged according to muscle invasive bladder cancer.
- PET not routinely indicated in transitional cell cancer with exception noted below in **Transitional Cell Cancer – Initial Work-up/Staging (ONC-18.2)**.

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Transitional Cell Cancer – Suspected/ Diagnosis (ONC-18.1)

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- See: **Hematuria and Hydronephrosis (AB-39)** in the Abdomen Imaging Guidelines for evaluation of suspected transitional cell malignancies.

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Transitional Cell Cancer – Initial Work-up/Staging (ONC-18.2)

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Indication	Medically necessary Imaging Study
All individuals	ONE of the following: • CT Abdomen and Pelvis without and with contrast (CPT® 74178) ◦ MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast if contraindication to CT contrast • CT Abdomen and Pelvis without contrast (CPT® 74176) with retrograde pyelogram or renal ultrasound (CPT® 76770 or CPT® 76775) in individuals who cannot receive either CT or MRI contrast
Sessile or high-grade tumors	In addition to the imaging above, if not already performed: • MRI Pelvis without and with contrast (CPT® 72197)
ANY of the following: • Muscle invasive bladder carcinoma • Urethral carcinoma • Urothelial carcinoma of the prostate	• CT Chest without (CPT® 71250) or with (CPT® 71260) contrast
Individuals without metastatic disease, when requested by operating surgeon for operative planning	• CT with contrast or MRI without and with contrast of all operative sites

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Indication	Medically necessary Imaging Study
Evaluation of suspected bone metastasis: • Individuals with symptoms of bone metastasis • Laboratory indicators of bone metastasis • Individuals with muscle invasive disease • Suspected sites of extra-osseous metastatic disease	Any ONE of the following: • Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology [ONC-1.3]** for additional bone scan codes) • MRI without and with contrast of areas of suspected involvement • FDG PET/CT (CPT® 78815)
To evaluate inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)
Suspected brain metastases	See: **Brain Metastases (ONC-31.3)**

Evidence Discussion

Initial staging of non-muscle invasive bladder cancer (NMIBC)

A clinician should perform upper tract imaging as a component of the initial evaluation of an individual with bladder cancer. NCCN guidelines recommend CT urography CTU (CT of the abdomen and pelvis without and with IV contrast with excretory imaging), which is an imaging study that is tailored to improve visualization of both the upper and lower urinary tracts. CT scan can require a significant dose of ionizing radiation but the speed of image acquisition reduces the potential for motion artifact.

MR urography (MRU) may be appropriate, especially in an individual with poor renal function or iodinated contrast allergy but with GFR >30 and no acute renal failure. MRU can be degraded due to motion artifact, but there is no ionizing radiation with this imaging modality.

CTU and MRU allow for comprehensive evaluation of the genitourinary tract, as well as assessment of retroperitoneal and pelvic lymph nodes.

Renal ultrasound (US) or CT without contrast may be utilized in conjunction with retrograde ureteropyelography in individuals who cannot receive either iodinated or gadolinium-based contrast material. Ultrasound requires no ionizing radiation, but is not sufficient for evaluation alone, and must be combined with either retrograde ureteropyelography or ureteroscopy to completely evaluate the upper urinary tract.[1]

NCCN guidelines stated that chest imaging may not be necessary in the initial staging of non-invasive disease, as the risk of chest metastasis in individuals with TA or T1 NMBIC is low.[1,8]

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FDG PET/CT is not recommended to delineate the anatomy of the upper urinary tract as it does not provide detailed imaging of the bladder. Thus, it is reserved as a problem-solving tool when other imaging studies are inconclusive.[1]

Initial staging of non-metastatic muscle-invasive bladder cancer (MIBC)

Prior to muscle-invasive bladder cancer management, clinicians should perform a complete staging evaluation, including imaging of the chest and cross-sectional imaging of the abdomen and pelvis with intravenous contrast if not contraindicated. Laboratory evaluation should include a comprehensive metabolic panel (complete blood count, liver function tests, alkaline phosphatase, and renal function).

All individuals with MIBC require imaging of the thorax. Chest radiography is an effective screening exam. Any abnormality should be followed up with a CT exam.

For sessile or high-grade tumors, it is recommended that MRI Pelvis be performed in addition to CTU for local staging.[1]

NCCN indicated that bone imaging is medically necessary if there are clinical indications of bone metastases or in individuals with muscle-invasive disease.[1]

FDG PET/CT is not recommended to delineate the anatomy of the upper urinary tract as it does not provide detailed imaging of the bladder. Thus, it is reserved as a problem-solving tool when other imaging studies are inconclusive.

Initial staging of Urothelial Carcinoma of the prostate/primary carcinoma of the urethra

Initial work up is similar to non-metastatic muscle invasive bladder cancer and should include Chest CT, CTU or MRU.

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Transitional Cell Cancer – Restaging/ Recurrence (ONC-18.3)

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Indication	Medically Necessary Imaging Study
After definitive surgery	• CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen and Pelvis without and with contrast (CPT® 74178) for post-operative baseline
Recurrence suspicion	• CT Abdomen and Pelvis with contrast (CPT® 74177) or with and without contrast (CPT® 74178) • CT Chest with contrast (CPT® 71260) for ANY of the following: ◦ Known prior thoracic disease ◦ New or worsening pulmonary symptoms ◦ New or worsening chest x-ray findings ◦ Known prior or suspected muscle invasive disease
After neoadjuvant therapy and before resection	• CT Chest with contrast (CPT® 71260) and CT Urogram (CPT® 74178)
Monitoring therapy for metastatic disease	Every 2 cycles of therapy: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Chest with contrast (CPT® 71260) for ANY of the following: ◦ Known prior thoracic disease ◦ New or worsening pulmonary symptoms ◦ New or worsening chest x-ray findings
To evaluate inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

Individuals presenting with findings suggesting a new primary or local recurrence of malignancy should undergo metastatic evaluation including abdominal and pelvic

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imaging. The most common sites for metastasis include lymph nodes, bone, lung, liver, and peritoneum.[10] The recommended imaging for restaging of suspected recurrence mirrors that which is performed in initial staging.[1,6]

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Transitional Cell Cancer – Surveillance/ Follow-up (ONC-18.4)

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Indication	Medically Necessary Imaging Study
ANY of the following: • Papillary urothelial neoplasm of low malignant potential • Low risk lesions ◦ Solitary Ta lesions ≤3cm • Intermediate risk lesions ◦ Low-grade >3cm ◦ Low-grade multifocal ◦ T1 lesions ◦ High-grade solitary Ta ≤3cm	One-time baseline imaging at the end of therapy: • CT Urogram (CPT® 74178) or MR Urogram (CPT® 74183 and CPT® 72197) • Further advanced imaging is not routinely medically necessary for surveillance
ANY of the following high-risk non-muscle invasive transitional cell carcinoma of the bladder or upper tracts: • Multifocal high-grade lesions • High-grade lesions >3 cm • Superficial and minimally invasive (Tis and T1) • BCG unresponsive • Lymphovascular invasion • Prostatic urethral invasion	• CT Urogram (CPT® 74178) or MR Urogram (CPT® 74183 and CPT® 72197) (imaging as performed at initial staging) every 1-2 years for 10 years
Non-muscle-invasive transitional carcinoma of the bladder treated with cystectomy	• CT Urogram (CPT® 74178) or MR Urogram (CPT® 74183 and CPT® 72197) (imaging as performed at initial staging) at 3 and 12 months post-cystectomy, and then annually for years 2-5

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Indication	Medically Necessary Imaging Study
Muscle invasive lower and upper genitourinary tumors treated with cystectomy, nephrectomy, or chemoradiation	Every 3-6 months for 2 years, then annually for 3 more years: • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250), and CT Abdomen and Pelvis with contrast (CPT® 74177) or without and with contrast (CPT® 74178) OR • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250), and MR Urogram (CPT® 74183 and CPT® 72197)
Measurable metastatic disease on maintenance therapy or being monitored off therapy	Every 3 months for up to 5 years after completion of active treatment: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177), CT Urogram (CPT® 74178), or MR Urogram (CPT® 74183 and CPT® 72197) (imaging as performed at initial staging)
Urethral cancers (high-risk T1 or greater) and urothelial carcinoma of the prostate	Every 3-6 months for 2 years, then annually: • CT Abdomen and Pelvis with contrast (CPT® 74177) or without and with contrast (CPT® 74178) OR • MR Urogram (CPT® 74183 and CPT® 72197) AND • Chest x-ray ◦ CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for any of the following: ▪ Signs/symptoms of pulmonary disease ▪ Abnormal chest x-ray ▪ Prior involvement of the chest

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Evidence Discussion

Surveillance of non-muscle invasive bladder cancer[1,7]

In an asymptomatic individual with a history of low-risk NMIBC a clinician should not perform routine surveillance upper tract imaging, after the initial baseline.

For an intermediate or high-risk individual, a clinician should consider performing surveillance upper tract imaging at 1-2 year intervals. Initially obtain imaging at 12 months, then every 1-2 years up to 10 years. CT urography (CTU), MRU, or retrograde ureteropyelography with non-contrast CT or US or ureteroscopy with a non-contrast CT or US.

Routine chest imaging is not appropriate for individuals with NMBIC unless an abnormality is identified with chest radiography.

Surveillance of non-metastatic muscle-invasive bladder cancer[1,7]

Clinicians should obtain chest imaging and cross-sectional imaging of the abdomen and pelvis with CT or MRI at 6-12 month intervals for 2-3 years and then may continue annually.

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Prostate Cancer (ONC-19)

Guideline

Prostate Cancer – General Considerations (ONC-19.0)
Suspected Prostate Cancer (ONC-19.1)
Prostate Cancer – Initial Work-up/Staging (ONC-19.2)
Prostate Cancer – Restaging/Recurrence (ONC-19.3)
Prostate Cancer – Follow-up On Active Surveillance (ONC-19.4)
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Prostate Cancer – General Considerations (ONC-19.0)

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- Prostate cancer screening begins at age 45 for individuals at average risk of prostate cancer. However, individuals at high-risk may begin screening at age 40. High-risk features include:
  - African ancestry
  - germline mutations (BRCA1 or 2, HOXB13, ATM, CHEK2, or mismatch repair genes (MLH1, MSH2, MSH6, PMS2) that increase the risk of prostate cancer
  - family history of first or second-degree relative with prostate, male breast, colorectal, pancreatic, endometrial or female breast cancer at age <45 years
- Treatment of benign prostatic hyperplasia with 5-alfa reductase inhibitors (such as finasteride and dutasteride) can falsely reduce the measured PSA levels by 50%. Thus, the reported PSA level should be doubled when prostate cancer is suspected in individuals on these medications.
- Individuals with high-risk adverse clinical and pathological factors may benefit from a more aggressive diagnostic and therapeutic approach at the time of relapse after initial treatment. These factors include pre-treatment Gleason score of ≥8, pre-treatment clinical stage of cT3b or higher, positive surgical margins, post-treatment PSA doubling time of <3 months, and an interval to biochemical failure of <3 years after initial treatment.
- PET/CT scans using ^{18}F-FDG radiotracer are not medically necessary for evaluation of prostate cancer.
- ^{11}C Choline, ^{18}F-Fluciclovine (AXUMIN®), and PSMA-specific radiopharmaceuticals have recently gained FDA approval for evaluation of prostate cancer. Optimal detection rates for these radiotracers vary greatly with PSA levels. False positive rate is high and histological confirmation of positive sites is recommended.
- PSMA-specific PET radiopharmaceuticals that are currently FDA-approved and indicated in prostate cancer are: ^{68}Ga PSMA-11 (UCSF & UCLA), ^{18}F Piflufolastat (Pylarify®), ^{18}F Flotufolastat (Posluma®), and ^{68}Ga Gozetotide (Illuccix® and Locametz®).
- While early detection of low-volume recurrence after treatment of prostate cancer using PET/CT scans may influence therapeutic decisions, there is lack of evidence that this approach has any meaningful impact on overall survival.
- As high-intensity focused ultrasound prostate ablation is considered investigational and experimental at this time, and advanced imaging for treatment planning and/or

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surveillance of high intensity focused ultrasound prostate ablation is not medically necessary.
- MR Spectroscopy (CPT® 76390) is considered not medically necessary in the evaluation of prostate cancer at this time.
- As laser prostate ablation is considered investigational and experimental at this time, advanced imaging for treatment planning and/or surveillance of laser prostate ablation is not medically necessary.
- Monitoring an elevated prostate-specific antigen level (PSA) with serial MRI is not medically necessary for suspected prostate cancer.
- Requests for imaging based on PSA must provide a recent PSA.

ISUP Prostate Cancer Grade Groups

Grade Group	Gleason Score	Gleason Pattern
1	≤6	≤3+3
2	7	3+4
3	7	4+3
4	8	4+4, 3+5, 5+3
5	9 or 10	4+5, 5+4, or 5+5

NCCN Initial Risk Stratification
- Very Low Risk
  - ALL of the following features are present:
    - Tumor not clinically palpable, but present on one or both lobes on biopsy (cT1a, cT1b, or cT1c)
    - PSA (ng/mL) <10
    - Gleason Grade Group = 1
    - <3 prostate biopsy cores positive, ≤50% cancer in each core
    - PSA Density <0.15 ng/mL/g
- Low Risk
  - ALL of the following features are present but does not qualify for very low risk:
    - Clinical T Stage = cT1-cT2a (palpable tumor limited to ≤1/2 of one side)
    - PSA (ng/mL) <10
    - Gleason Grade Group = 1
- Intermediate Risk

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- No "high-risk" or "very high-risk" features and one or more of the following intermediate risk factors (IRFs):
  - cT2b-cT2c
  - Grade group 2 or 3
  - PSA 10-20 ng/mL
- Favorable Intermediate Risk
  - ALL of the following features must be present:
    - Only 1 IRF
    - Gleason Grade Group = 1 or 2
    - <50% biopsy cores positive (e.g., <6 of 12 cores)
- Unfavorable Intermediate Risk
  - One or more of the following must be present:
    - 2 or 3 IRFs
    - Gleason grade group = 3
    - ≥50% biopsy cores positive (e.g., ≥6 of 12 cores)

- High-Risk
  - ONE or more of the following high-risk features is present:
    - Clinical T Stage = cT3-cT4
    - PSA (ng/mL) >20
    - Gleason Grade Group = 4 or 5
- Very High-Risk
  - At least TWO of the following features is present:
    - Clinical T stage = cT3-cT4 (extension into the seminal vesicles or invasion into adjacent structures)
    - Primary Gleason Pattern = 5
    - Gleason Grade Group = 4 or 5
    - PSA (ng/mL) >40

3D Rendering of MRI for MRI / Ultrasound Fusion Biopsy:

- When specific target lesion(s) is (are) detected on multi-parametric MRI (mpMRI) prostate and classified as PIRADS 4 or 5, 3D Rendering (CPT® 76377) to generate prostate segmentation data image set for target identification on MRI/Transrectal ultrasound (TRUS) fusion biopsy is medically necessary as:
  - a subsequent separate standalone request; or
  - a retrospective request for medical necessity
- For MRI/TRUS fusion biopsy of a PIRADS 1-3 lesion, approval of 3D rendering at independent workstation (CPT® 76376 or CPT® 76377) can be considered on a case-by-case basis.

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- If there is no target lesion identified on MRI, then 3D rendering and MRI/TRUS fusion biopsy is generally not medically necessary.
- The 3D rendering for the TRUS component of the fusion is a part of the UroNav Fusion Equipment Software and an additional 3D code CPT® 76376 or CPT® 76377 is not medically necessary.

Evidence Discussion

Screening can begin as early as age 40 for high-risk individuals (Black/African-American identity, certain germline mutations, or concerning family history) and 45 for individuals with average risk. Those with a first-degree relative diagnosed at age less than 60 years have a more than 2-fold increase in likelihood of prostate cancer diagnosis. Individuals with African ancestry have a 60% higher incidence of prostate cancer. Individuals with high-risk adverse clinical and pathologic factors may benefit from a more aggressive diagnostic and therapeutic approach at the time of relapse after initial treatment.[2]

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Suspected Prostate Cancer (ONC-19.1)

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- MR guidance for needle placement (CPT® 77021) is not medically necessary for prostate biopsy.

Indication	Medically Necessary Imaging Study
ANY of the following: • Age 40-75 years with PSA >3 ng/ml or very suspicious DRE and ONE of the following high-risk features: ◦ African ancestry ◦ Germline mutations that increase the risk of prostate cancer ◦ Family history of first- or second-degree relative with prostate, male breast, pancreatic, or ovarian cancer ◦ Family history of first- or second-degree relative diagnosed at age ≤45 years with female breast cancer ◦ Family history of first- or second-degree relative diagnosed at age ≤50 years with colorectal or endometrial cancer • Age 45-75 years and ONE of the following: ◦ PSA >3 ng/ml ◦ Very suspicious DRE • Age >75 years and ONE of the following: ◦ PSA ≥4 ng/ml ◦ Very suspicious DRE • At least one negative/non-diagnostic TRUS biopsy and ANY of the following: ◦ Rising PSA ◦ Abnormal DRE ◦ Need for confirmatory MR/US fusion biopsy	ANY of the following: • Transrectal ultrasound (CPT® 76872) • TRUS-guided biopsy (CPT® 76942) • MRI/US fusion biopsy (CPT® 76942) • If results of MRI will be used to determine the need for, and subsequently perform, MR fusion biopsy: ◦ MRI Pelvis without and with contrast (CPT® 72197) OR ◦ MRI Pelvis without contrast (CPT® 72195)

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Indication	Medically Necessary Imaging Study
• PIRADS 4 or 5 lesion identified on recent diagnostic MRI Pelvis (CPT® 72195 or CPT® 72197) and planning for biopsy to be done by MRI/TRUS fusion technique	• 3D Rendering (CPT® 76376 or CPT® 76377)
ANY of the following: • Multifocal (2 or more lesions) high-grade prostatic intraepithelial neoplasia (PIN) • Atypia on biopsy	• Extended pattern re-biopsy within 6 months by TRUS-guided biopsy (CPT® 76942)
• Solitary PIN (1 lesion)	ONE of the following, to be repeated in one year: • MRI Pelvis without contrast (CPT® 72195) • MRI Pelvis without and with contrast (CPT® 72197) • MRI/US fusion biopsy (CPT® 76942 or 76872) ◦ These codes are medically necessary in addition to MR Pelvis codes above, if requested
In office, transperineal, MRI guided biopsy without US fusion (i.e., ProMaxo)	Diagnostic MRI code (CPT® 77021) is not medically necessary in conjunction with procedure codes (see: **CT-, MR-, or US-Guided Procedures (Preface-4.2)**)

Evidence Discussion

Based on the high-risk factors outlined above along with age, digital rectal exam (DRE) findings and PSA level, further imaging work-up may be medically necessary to include transrectal ultrasound with or without biopsy, MRI of the pelvis without and/or with contrast as well as MRI/US fusion biopsy is indicated. These interventions will help dictate the appropriate therapy for each individual diagnosed with prostate cancer.[1]

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Prostate Cancer – Initial Work-up/ Staging (ONC-19.2)

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Indication	Medically Necessary Imaging Study
Localized prostate cancer with any of the following risk groups (see: **ONC-19.0** for definition of risk groups): • Very low risk • Low risk • Favorable intermediate risk	Advanced imaging is not routinely medically necessary for initial staging If not already performed prior to biopsy, MRI Pelvis without and with contrast (CPT® 72197) is appropriate for any of the following: • Prior to planned treatment (surgery and/or radiation therapy) • To establish candidacy for active surveillance
Localized prostate cancer with any of the following risk groups (see: **ONC-19.0** for definition of risk groups): • Unfavorable intermediate risk • High-risk • Very high-risk	Any ONE of the following combinations, not all (may be obtained in addition to mpMRI prostate): • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) • CT Chest with contrast (CPT® 71260), CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197) if not previously performed, and Bone scan (CPT® 78306) • PSMA PET/CT scan (CPT® 78815 or CPT® 78816) using any one of the following radiotracers: ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) ◦ ^{18}F Flotufolastat (Posluma®)

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Indication	Medically Necessary Imaging Study
To confirm oligo- or low volume metastatic prostate cancer suspected on conventional imaging in individuals with biopsy-proven prostate cancer	• PSMA PET/CT (CPT® 78815 or CPT® 78816) or PSMA PET/MRI (CPT® 78813 and CPT® 76498) with any of the following radiotracers: ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{18}F Flotufolastat (Posluma®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®)
Known diffuse metastatic prostate cancer (including prior to prostate biopsy)	Any ONE of the following combinations, not all: • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) • CT Chest with contrast (CPT® 71260), CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197) if not previously performed, and Bone scan (CPT® 78306)
Inconclusive bone scan	Any ONE of the following: • CT with contrast of involved body site • MRI without and with contrast of involved body site • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) using any one of the following radiotracers: ◦ ^{18}F Fluciclovine ◦ ^{11}C Choline ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{18}F Flotufolastat (Posluma®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) ◦ ^{18}F sodium fluoride

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Evidence Discussion

Risk stratification uses a minimum of stage, Gleason grade, and PSA to assign individuals to risk groups that in turn help select the imaging options and predict the probability of biochemical recurrence after definitive local therapy. The current prostate cancer grading system was adopted from the ISUP 2014 consensus conference with the goal to limit overtreatment. The grading system is divided into 6 risk groups.

The goal of imaging is to detect and characterize disease in order to guide disease management. Very low risk, low risk, and favorable intermediate risk does not routinely require advanced imaging. NCCN stated that "conventional bone scans are rarely positive in asymptomatic individuals with PSA <10 ng/mL".[1] Very low risk, low risk, and favorable intermediate risk groups have very low risk of positive bone scan or CT scan.[1]

Unfavorable intermediate risk, high-risk, and very high-risk do require imaging that can be a combination of CT/MRI Pelvis or PSMA PET using specific radiotracers. In individuals with known or suspected metastatic disease, CT Chest, Abdomen, and Pelvis along with bone scan are indicated. MRI Pelvis is an acceptable choice in lieu of CT Pelvis due to MRI's ability to provide detailed soft tissue characterization and contrast. NCCN supported bone imaging for those at high risk for skeletal metastases[1]

For inconclusive bone findings, NCCN stated that additional imaging using CT, MRI, PSMA-PET/CT or PSMA-PET/MRI, or PET/CT or PET/MRI with F-18 sodium fluoride, C-11 choline, or F-18 fluciclovine can be considered. For conventional imaging suggesting oligo- or low-volume metastatic disease, PET/CT using specific radiotracers can be performed to confirm the individual is a candidate for localized treatment. F18-FDG PET/CT scans are considered not medically necessary for evaluation of prostate cancer because, per NCCN, there is a lack of data surrounding the use of this radiotracer in individuals with prostate cancer. Other radiotracers (C11 choline, F18-Fluciclovine, PSMA-specific) are FDA approved and have influenced treatment planning but the impact on long-term survival remains to be studied. However, data does indicate that PSMA-PET has a high sensitivity for metastatic disease and small-volume disease compared to conventional imaging and therefore it has become a useful tool for the evaluation of individuals with prostate cancer.[1]

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Prostate Cancer – Restaging/Recurrence (ONC-19.3)

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Indication	Medically Necessary Imaging Study
For ANY of the following: • Obvious progression by DRE with plans for prostatectomy or radiation therapy • Repeat TRUS biopsy for rising PSA shows progression to a higher Gleason's score with plans for prostatectomy or radiation therapy • Inconclusive findings on CT scan	• MRI Pelvis without and with contrast (CPT® 72197)
Non-metastatic prostate cancer previously treated with ablation, hormonal therapy, or chemotherapy **and** any one of the following: • Clinical suspicion of relapse/recurrence • Palpable anastomotic recurrence • PSA rises above post-treatment baseline to >0.2 ng/mL on two consecutive measurements	Any ONE of the following combinations: • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) • CT Chest with contrast (CPT® 71260), CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197), and Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)

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Indication	Medically Necessary Imaging Study
Non-metastatic prostate cancer previously treated with prostatectomy and **all** of the following are met: • Persistent detectable PSA after prostatectomy, or • Undetectable PSA that subsequently becomes detectable with two consecutive increases in PSA (to any amount), or • Any increase in PSA to 0.1 ng/mL or higher AND: • Individual is a candidate for salvage local therapy	Any ONE of the following: • PSMA PET/CT scan (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) with any of the following radiotracers: ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{18}F Flotufolastat (Posluma®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) • CT Chest with contrast (CPT® 71260), CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197), and Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)

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Indication	Medically Necessary Imaging Study
Non-metastatic prostate cancer previously treated with radiation therapy and **both** of the following are met: • Two consecutive increases in PSA above nadir AND • Individual is a candidate for salvage local therapy	Any ONE of the following: • PSMA PET/CT scan (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) with any of the following radiotracers: ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{18}F Flotufolastat (Posluma®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) • CT Chest with contrast (CPT® 71260), CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197), and Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)
Non-metastatic prostate cancer previously treated with prostatectomy or radiation therapy, and **all** of the following are met: • PSA rises on two consecutive measurements above post-treatment baseline **and** • PSA ≥1 ng/mL **and** • Recent CT scan and bone scan are negative for metastatic disease **and** • Individual is a candidate for salvage local therapy	• PET/CT scan (CPT® 78815 or CPT® 78816) using any **ONE** of the following radiotracers: ◦ ^{18}F-Fluciclovine ◦ ^{11}C Choline

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Indication	Medically Necessary Imaging Study
Suspected progression of known metastatic disease based on: • New or worsening signs/symptoms • Rising PSA levels	• CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) • CT with contrast of any involved or symptomatic body part
Metastatic prostate cancer receiving treatment with chemotherapy or Pluvicto®	One of the following combinations, every 2 cycles (6 to 8 weeks), while on chemotherapy: • CT Abdomen and Pelvis with contrast (CPT® 74177) and CT scan with contrast of any involved body part OR • CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197) AND, every 3-6 months: • Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)

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Indication	Medically Necessary Imaging Study
Metastatic prostate cancer receiving anti-androgen therapy	One of the following combinations, every 3 months, while on anti-androgen therapy: • CT Abdomen and Pelvis with contrast (CPT® 74177) and CT scan of any involved body part OR • CT Abdomen with contrast (CPT® 74160), MRI Pelvis without and with contrast (CPT® 72197) AND, every 3-6 months: • Bone scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes)
Previously treated metastatic prostate cancer progressed on conventional imaging and being considered for ^{177}Lu-PSMA-617 (Pluvicto®) treatment[31, 32]	• PSMA PET/CT scan (CPT® 78815 or CPT® 78816) with one of the following agents: ◦ ^{68}Ga PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) ◦ ^{18}F Flotufolastat (Posluma®)
Prior to start of Xofigo® (Radium-223) therapy	• ONE time CT Chest, Abdomen, and Pelvis with contrast (CPT® 71260 and CPT® 74177)

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Indication	Medically Necessary Imaging Study
Inconclusive bone scan	Any ONE of the following: • CT with contrast of involved body site • MRI without and with contrast of involved body site • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) using any one of the following radiotracers: ◦ ^{18}F Fluciclovine ◦ ^{11}C Choline ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{18}F Flotufolastat (Posluma®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) ◦ ^{18}F sodium fluoride
Conventional imaging studies (CT and bone scan) suggests oligo- or low volume metastatic disease that needs further confirmation	• PET/CT scan (CPT® 78815 or CPT® 78816) using any one of the following radiotracers: ◦ ^{18}F Fluciclovine ◦ ^{11}C Choline ◦ ^{68}Ga-PSMA-11 ◦ ^{18}F Piflufolastat (Pylarify®) ◦ ^{68}Ga Gozetotide (Illuccix® and Locametz®) ◦ ^{18}F Flotufolastat (Posluma®)

Evidence Discussion

The use of PSMA PET radiotracers has been shown to impact treatment planning but more research is needed on the impact to long-term outcomes. However, the benefit to individuals of initiating treatment earlier for recurrent disease detected on PSMA PET outweighs the disadvantages of possible false-positives. For non-metastatic prostate cancer previously treated with prostatectomy, NCCN stated that PET can be used without prerequisite conventional imaging for suspected biochemical recurrence. This recommendation is due to "increased sensitivity and specificity of PSMA-PET tracers for detecting micrometastatic disease compared to conventional imaging". Conventional imaging is also noted as useful for workup of recurrence or progression. For individuals

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treated with radiation therapy, NCCN stated that suspected recurrence can also be imaged using these modalities. In the setting of distant metastatic disease, CT imaging along with bone scan is the mainstay of imaging with PET/CT reserved for inconclusive conventional imaging or oligo-/low volume disease that needs confirmation. NCCN stated "CT provides a high level of anatomic detail, and may detect gross extracapsular disease, nodal metastatic disease, and/or visceral metastatic disease." PSMA imaging at baseline is medically necessary for individuals being considered for 177Lu-PSMA-617 (Pluvicto®) treatment.[1]

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Prostate Cancer – Follow-up On Active Surveillance (ONC-19.4)

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Active surveillance is being increasingly utilized in prostate cancer, and this therapeutic option involves regimented monitoring of an individual with known diagnosis of low-risk prostate cancer for disease progression, without specific anticancer treatment. While being treated with active surveillance, an individual is generally considered a potential candidate for curative intent treatment approaches in the event that disease progression occurs.

It is important to distinguish active surveillance from watchful waiting (or observation), which is generally employed in individuals with limited life expectancy. Watchful waiting involves cessation of routine monitoring and treatment is initiated only if symptoms develop.

Current active surveillance guidelines suggest the following protocol:

- PSA every 6 months
- Digital Rectal Exam (DRE) every 12 months
- Repeat prostate biopsy every 12 months
- Repeat mpMRI (CPT® 72195 or CPT® 72197) no more often than every 12 months

Indication	Medically Necessary Imaging Study
Routine monitoring on active surveillance protocol	• MRI Pelvis without (CPT® 72195) or without and with contrast (CPT® 72197) at initiation of active surveillance, and every 12 months thereafter
Planning for re-biopsy to be done by MRI/US fusion technique	• 3D Rendering (CPT® 76376 or CPT® 76377)

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Indication	Medically Necessary Imaging Study
For ANY of the following: • Progression is suspected based on DRE changes or rising PSA and a recent TRUS biopsy was negative • Repeat TRUS biopsy shows progression to a higher Gleason score	• MRI Pelvis without (CPT® 72195) or MRI Pelvis without and with contrast (CPT® 72197)
Individuals on active surveillance who are noted to have progression and have plans to initiate treatment	• Imaging studies for initial staging as per **ONC-19.2**

Evidence Discussion

For certain individuals who fall into a low-risk category, close monitoring in the absence of treatment can be pursued with the intent to offer curative therapy in the event progression occurs. Current guidelines include PSA every 6 months, DRE every 12 months, repeat prostate biopsy every 12 months and repeat mpMRI no more often than every 12 months. NCCN stated that a metastatic staging evaluation is not medically necessary for those on active surveillance. With progression and the decision to pursue treatment, imaging is performed according to the same principles as stated in initial staging.[1]

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Surveillance/Follow-up For Treated Prostate Cancer (ONC-19.5)

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Indication	Medically Necessary Imaging Study
ANY of the following: • Asymptomatic or stable chronic symptoms • Stable DRE findings • Stable PSA levels	• Advanced imaging is not routinely medically necessary for surveillance

Evidence Discussion

For individuals who are asymptomatic or have chronic stable findings to include DRE and PSA, advanced imaging is not routinely indicated. This form of monitoring is also referred to as observation. NCCN stated that the advantages of observation avoidance of "possible side effects of unnecessary confirmatory testing and definitive therapy".[1]

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Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)

Guideline

Testicular, Ovarian and Extragonadal Germ Cell Tumors – General Considerations (ONC-20.0)
Testicular, Ovarian and Extragonadal Germ Cell Tumors – Initial Work-Up/Staging (ONC-20.1)
Testicular, Ovarian and Extragonadal Germ Cell Tumors – Restaging/Recurrence (ONC-20.2)
Testicular, Ovarian and Extragonadal Germ Cell Tumors – Surveillance (ONC-20.3)
References (ONC-20)

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Testicular, Ovarian and Extragonadal Germ Cell Tumors – General Considerations (ONC-20.0)

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- This section applies to primary germ cell tumors occurring outside the central nervous system for individual's aged >15 years at the time of initial diagnosis. Individuals aged ≤15 years at diagnosis should be imaged according to pediatric guidelines in: **Pediatric Germ Cell Tumors (PEDONC-10)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- These guidelines are for germ cell tumors of the testicle, ovary and extragonadal sites as well as malignant sex cord stromal tumors (granulosa cell and Sertoli-Leydig cell tumors).
- Requests for imaging must state the histologic type of the cancer being evaluated.
- Classified as pure seminomas (dysgerminomas, 40%) or non-seminomatous germ cell tumors (NSGCT, 60%):
  - Pure seminomas are defined as pure seminoma histology with a normal serum concentration of alpha fetoprotein (AFP). Seminomas with elevated AFP are by definition mixed.
  - Required for TNM staging are the tumor marker levels indicated by "S" (TNMS)
  - Mixed tumors are treated as NSGCTs, as they tend to be more aggressive.
  - The NSGCT histologies include:
    - yolk-sac tumors
    - immature (malignant) teratomas
    - choriocarcinomas (<1%)
    - embryonal cell carcinomas (15% - 20%)
    - endodermal sinus tumors (ovarian)
    - combinations of all of the above (mixed)
- MRI in place of CT scans to reduce risk of secondary malignancy is not supported by the peer-reviewed literature. CT scans are medically necessary for surveillance and are the preferred modality of imaging to assess for recurrence.
- PET/CT is considered not medically necessary for evaluation of non-seminomatous germ cell tumors.
- Active surveillance in testicular cancer refers to treatment with surgery (orchiectomy) alone without any additional post-operative treatment such as chemotherapy or radiotherapy.

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Testicular, Ovarian and Extragonadal Germ Cell Tumors – Initial Work-Up/ Staging (ONC-20.1)

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Indication	Medically Necessary Imaging Study
Suspicious testicular mass on ultrasound	EITHER of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) or • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
After orchiectomy or oophorectomy: • Orchiectomy/oophorectomy is both diagnostic and therapeutic, and imaging was not performed prior to surgery	EITHER of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) or • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Prior to initiation of chemotherapy to confirm staging, if prior imaging was performed ≥4 weeks ago	Repeat abdominal imaging with ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) or • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
For ANY of the following: • Non-seminoma histology • Ovarian germ cell tumor • Abdominal lymphadenopathy noted on CT scan • Abnormal chest x-ray or signs/ symptoms suggestive of chest involvement	• CT Chest with contrast (CPT® 71260)

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Indication	Medically Necessary Imaging Study
Extragonadal Germ Cell Tumor	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)
Pelvic mass suspicious for ovarian etiology	See: **Ovarian Cancer - Suspected/Diagnosis (ONC-21.2)**

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Testicular, Ovarian and Extragonadal Germ Cell Tumors – Restaging/ Recurrence (ONC-20.2)

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Indication	Medically Necessary Imaging Study
Treatment response for individuals with metastatic disease, every 2 cycles	• CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) AND • CT with contrast of previously involved body areas
Seminoma with residual mass >3 cm after completion of chemotherapy	• PET/CT (CPT® 78815)
Nonseminoma or pure seminoma with partial response or with residual mass and ALL of the following: • Abnormal AFP or beta-hCG levels • Need to evaluate for occult brain metastases	• MRI Brain without and with contrast (CPT® 70553)
Stage IIA pure seminoma or stage IIA nonseminoma with borderline retroperitoneal lymph nodes on preoperative imaging, 4-8 weeks after preoperative imaging	ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) or • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
End of therapy evaluation for NSGCT post chemotherapy or post retroperitoneal lymph node dissection (RPLND)	• CT Abdomen and Pelvis with contrast (CPT® 74177)

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Indication	Medically Necessary Imaging Study
Recurrence suspected, including increased tumor markers	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • Ultrasound (CPT® 76856 or CPT® 76857) of the remaining gonad if applicable
Unexplained pulmonary symptoms despite a negative chest x-ray, or new findings on chest x-ray	• CT Chest with contrast (CPT® 71260)
All others	• See: **Surveillance (ONC-20.3)**

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Testicular, Ovarian and Extragonadal Germ Cell Tumors – Surveillance (ONC-20.3)

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Indication	Medically Necessary Imaging Study
Stage I seminoma treated with orchiectomy alone (no radiotherapy or chemotherapy, also called active surveillance)	ONE of the following, every 4 months in year 1, then every 6 months for years 2 and 3, and then annually until year 5: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage I seminoma treated with radiotherapy and/or chemotherapy	ONE of the following, annually for 3 years: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
Stage IIA and non-bulky stage IIB seminomas treated with radiotherapy or chemotherapy	ONE of the following, once at 3 months, then once at 9-12 months after completion of therapy, then annually for 2 additional years: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage II seminoma treated with primary RPLND and NOT treated with adjuvant chemotherapy	ONE of the following, every 4 months in year 1, every 6 months in year 2, and annually in year 3: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage II seminoma treated with primary RPLND AND adjuvant chemotherapy	ONE of the following, at 6 months, 12 months, and 24 months: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
Bulky stage IIB, IIC, and III seminomas treated with chemotherapy	For individuals with ≤3 cm residual mass, ONE of the following every 4 months for 1 year, every 6 months for 1 year, and then annually for 2 additional years: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) For individuals with >3 cm residual mass and negative PET scan, ONE of the following at 6 and 12 months after completion of therapy, then annually until year 5: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) For individuals with thoracic disease: • CT Chest with contrast (CPT® 71260) every 2 months for 1 year, then every 3 months for 1 year, then annually until year 5 after completion of therapy

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Indication	Medically Necessary Imaging Study
Stage IA non-seminomatous germ cell tumors treated with orchiectomy alone (without risk factors)	ONE of the following, every 4 months for one year, every 6 months in the second year, and then annually for years 3, 4, and 5: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage IB non-seminomatous germ cell tumors treated with orchiectomy alone (with risk factors – lymphovascular invasion or invasion into spermatic cord/scrotum)	ONE of the following, every 4 months for years 1 and 2, then every 6 months for year 3, and then once in year 4: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage IA/IB non-seminomatous germ cell tumors treated with chemotherapy and/or primary RPLND	ONE of the following, annually for 2 years: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
Stage II-III non-seminomatous germ cell tumors with complete response to chemotherapy +/- post-chemotherapy RPLND	ONE of the following, every 4 months in year 1, every 6 months in year 2, and then annually in year 3: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) For individuals with known thoracic disease or thoracic symptoms, every 4 months in year 1, every 6 months in year 2, then annually until year 4 after completion of therapy: • CT Chest with contrast (CPT® 71260)
Stage IIA, IIB, or IIC non-seminomatous germ cell tumors treated with post-primary RPLND AND adjuvant chemotherapy	ONE of the following, once at 3-4 months after completion of RPLND: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage IIA or IIB non-seminomatous germ cell tumors treated with post-primary RPLND WITHOUT adjuvant chemotherapy	ONE of the following, once at 4 months after completion of therapy and once in year 2: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) • MRI Abdomen without and with contrast (CPT® 74183) • MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
All stages of ovarian dysgerminoma germ cell tumors	• CT Abdomen and Pelvis with contrast (CPT® 74177) every 4 months for 1 year, every 6 months for 1 year and then annually for 3 years after completion of therapy
All ovarian non-dysgerminoma germ cell tumors • Embryonal tumor • Endodermal sinus tumor • Immature teratoma • Non-gestational choriocarcinoma • Yolk sac • Mixed germ cell tumor	• CT Abdomen and Pelvis with contrast (CPT® 74177) every 4 months for 1 year, every 6 months for 1 year and then annually for 3 years after completion of therapy • CT Chest with contrast (CPT® 71260) every 4 months for 1 year and every 6 months for 1 year after completion of therapy
• Sex cord stromal tumors (male and female) • Mature teratoma	• No routine advanced imaging indicated
Extragonadal germ cell tumors	• CT of the involved region every 3 months for one year and every 6 months for one year.

Evidence Discussion - ONC-20

Initial Evaluation

- American Urological Association guideline recommended scrotal ultrasound with Doppler should be obtained in individuals with a unilateral or bilateral scrotal mass suspicious for neoplasm and that MRI should not be used in the initial evaluation and diagnosis of a testicular lesion suspected of being a neoplasm.[12]
- Ultrasound requires no ionizing radiation and is readily available. Overall, ultrasound is relatively quick and non-invasive modality to evaluate a testicular lesion. There are relatively few disadvantages of ultrasound for testicular lesions and primarily relate to sonographer experience.
- As advanced imaging modalities, CT and MRI offer excellent 3-D resolution. A CT scan can require a significant dose of ionizing radiation, but the speed of image acquisition reduces the potential for motion artifact. MRI yields better soft contrast resolution than CT and does not expose individuals to ionizing radiation, but due to longer image time is motion artifact-prone and may require sedation.

Seminoma:

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- NCCN recommended CT of the abdomen and pelvis with contrast or MRI of the abdomen and pelvis with and without contrast, and a chest x-ray. Chest CT with contrast is recommended if there is a positive finding on abdomen CT or abnormal chest x-ray.[1]

Nonseminoma:

- NCCN recommended CT of the abdomen, pelvis and chest with contrast or MRI of the abdomen and pelvis with and without contrast in addition to a chest CT with contrast.[1]

Surveillance

Pure seminoma:[1,13-16]

- Chest radiography is sufficient when compared with CT for follow-up of stage 1 pure seminoma and is recommended by NCCN. While CT is more sensitive than radiography for detecting recurrent disease in the chest, this added sensitivity is offset by lower specificity and a higher false positive detection rate for abnormalities that are not metastatic.
- Scrotal US does not have a role in the restaging of men with testicular cancer that has been established by orchiectomy, unless there is concern for contralateral tumor or equivocal clinical exam.
- CT Abdomen and Pelvis is the reference standard imaging test used for assessing the retroperitoneum. It is rapid, reproducible, and provides excellent imaging of the para-aortic and paracaval regions but does expose individuals to significant ionizing radiation.
- MRI of the abdomen and pelvis
  - MRI has comparable accuracy with CT for the detection of metastatic retroperitoneal lymph nodes and has the benefit of no ionizing radiation. The disadvantage of MRI is longer imaging time which can lead to motion artifact.
- CT of the abdomen and pelvis is the standard imaging test used for assessing the retroperitoneum for nodal metastasis but does expose the individual to high levels of ionizing radiation.
- NCCN guidelines recommended chest radiography for the surveillance of stage I nonseminomatous testicular cancer but chest CT with contrast is preferred in the presence of thoracic symptoms.

Nonseminoma:[1]

- CT of the abdomen and pelvis is the standard imaging test used for assessing the retroperitoneum for nodal metastasis but does expose the individual to high levels of ionizing radiation.

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- NCCN guidelines recommended chest radiography for the surveillance of stage 1 nonseminomatous testicular cancer but chest CT with contrast is preferred in the presence of thoracic symptoms.
- MRI of the abdomen and pelvis shows comparable accuracy with CT in the detection of metastatic retroperitoneal lymph nodes and does not have high levels of ionizing radiation but is subject to motion artifact due to longer imaging time.
- Scrotal US does not have a role in the restaging of men with testicular cancer that has been established by orchiectomy, unless there is concern for contralateral tumor or equivocal clinical exam.

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Ovarian Cancer (ONC-21)

Guideline

Ovarian Cancer – General Considerations (ONC-21.0)
Screening for Ovarian Cancer (ONC-21.1)
Ovarian Cancer – Suspected/Diagnosis (ONC-21.2)
Ovarian Cancer – Initial Work-Up/Staging (ONC-21.3)
Ovarian Cancer – Restaging/Recurrence (ONC-21.4)
Ovarian Cancer – Surveillance (ONC-21.5)
References (ONC-21)

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Ovarian Cancer – General Considerations (ONC-21.0)

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- Ovarian cancers include: epithelial ovarian cancers, ovarian cancers of low malignant potential and mixed Müllerian tumors, primary peritoneal and fallopian tube cancers.
  - There are five main types of epithelial ovarian cancers:
    - High-grade serous carcinoma (HGSC) (70%)
    - Endometrioid carcinoma (EC) (10%)
    - Clear cell carcinoma (CCC) (10%)
    - Mucinous carcinoma (MC) (3%)
    - Low-grade serous carcinoma (LGSC) (<5%)
- Borderline tumors (formerly referred to as tumors of low malignant potential) usually have some feature of carcinoma when they recur.
- Fallopian tube and primary peritoneal are usually serous carcinoma.
- Germ cell tumors and sex cord stromal tumors (granulosa cell tumors), are imaged according to **Testicular, Ovarian and Extragonadal Germ Cell Cancer (ONC-20)**.

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Screening for Ovarian Cancer (ONC-21.1)

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Indication	Medically Necessary Study
High-Risk Factors: • Family history of BRCA 1 or BRCA 2 mutations • Family history of ovarian cancer • Hereditary ovarian cancer syndrome that includes ovarian, breast, and/or endometrial and gastrointestinal cancers [Lynch II syndrome] in multiple members of two to four generations • Low parity • Decreased fertility • Delayed childbearing	• The use of advanced imaging for ovarian cancer screening is considered not medically necessary. • Genetic counseling is recommended for women with an increased-risk family history.
Known BRCA 1 or BRCA 2 mutation	• Transvaginal ultrasound (CPT® 76830), combined with CA-125 for ovarian cancer screening is medically necessary annually starting at age 30, until risk-reducing salpingo-oophorectomy is performed.

Evidence Discussion

According to ACR Appropriateness Criteria for ovarian cancer screening, any imaging in an average-risk individual is "usually not appropriate."[5]

There is much debate about the role of imaging in screening for ovarian cancer. This cancer has a low prevalence but is the leading cause of mortality in women in the United States. Average lifetime risk is 1.3%. There are factors that increase the risk such as family history, BRCA 1 or BRCA 2 mutations, nulliparity, lack of hormonal contraceptive use, and lack of breastfeeding. Genetic predisposition is associated with the highest increase in risk.

In individuals who are considered high-risk (personal or family history, genetic predisposition or elevated CA-125) ultrasound and color Doppler may be appropriate annual screening along with CA-125. Lu, et al. demonstrated a PPV 40% with specificity

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of 99.9% using ROCA.[11] Studies showed that the risk for developing ovarian cancer increases with age beginning around age 30 years.[12]

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Ovarian Cancer – Suspected/Diagnosis (ONC-21.2)

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- See: **Complex Adnexal Masses (PV-5.3)** for imaging guidelines for evaluation of suspected ovarian malignancies.
- To differentiate the origin of pelvic masses that are not clearly of ovarian origin, see: **Suspected Adnexal Mass (PV-5.1)**.

Indication	Medically Necessary Imaging
• Pelvic signs or symptoms • Palpable pelvic mass	• Transvaginal (TV) ultrasound imaging (CPT® 76830) and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857)
• Ultrasound shows complex and/or solid adnexal mass suspicious for ovarian malignancy or • Any suspicious signs/symptoms for ovarian malignancy (ascites, abdominal symptoms such as distension or tenderness, elevated CA-125, elevated LFTs, obstructive uropathy*)	ANY of the following: • MRI Pelvis without and with contrast (CPT® 72197) ◦ MRI Pelvis without contrast (CPT® 72195) if contrast is contraindicated • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) are medically necessary in lieu of CT for individuals with renal insufficiency or contrast allergy. ◦ CT Abdomen and Pelvis without and with contrast (CT Urogram – CPT® 74178) only for symptoms of obstructive uropathy*

Evidence Discussion

If there is a question about the pelvic mass evaluation by ultrasound, transvaginal or pelvic ultrasound using O-RADS or IOTA is medically necessary to clarify risk before surgery. Accurate diagnosis of adnexal pathology is imperative in order to limit invasive interventions for benign lesions and improve preoperative triage to a gynecologic oncologist for high-risk lesions.[16] In order to standardize reporting of adnexal lesions, the American College of Radiology (ACR) has created the Ovarian-Adnexal Reporting and Data-System (O-RADS). A meta-analysis of 26 studies demonstrated that O-RADS

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has high sensitivity for detection of malignancy (95%).[16] A classification of O-RADS US Category 2 has an extremely low risk of malignancy (<1%), while a Category 5 has a high risk of malignancy (≥50%). For an indeterminate lesion on ultrasound or features concerning for malignancy, adjunct imaging with MRI is supported to aid in preoperative triage. For individuals who have renal insufficiency and therefore are unable to undergo contrasted CT imaging of the abdomen and pelvis, the accepted substitute imaging is MRI with contrast.[1] This imaging will provide the necessary level of soft tissue detail that would not be achievable with a non-contrasted CT.

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Ovarian Cancer – Initial Work-Up/Staging (ONC-21.3)

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Indication	Medically Necessary Imaging Study
Initial staging of all newly diagnosed ovarian cancer, if imaging has not yet been performed	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT for individuals with renal insufficiency or contrast allergy • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for: ◦ Abnormal signs/symptoms of pulmonary disease ◦ Abnormal chest x-ray ◦ Concern for metastatic disease (i.e., elevated tumor markers, abdominal symptoms)
ANY of the following: • Primary peritoneal disease with biopsy-proven malignancy consistent with ovarian carcinoma • Elevated tumor markers with negative or inconclusive CT imaging	ONE of the following: • MRI Abdomen without and with contrast (CPT® 74183) and/or MRI Pelvis without and with contrast (CPT® 72197) OR • PET/CT (CPT® 78815)
If both CT and MRI are inconclusive	• PET/CT (CPT® 78815)
If CT, MRI, and PET/CT are inconclusive	• PET/MRI (CPT® 78813 and 76498)

Evidence Discussion

If advanced imaging was not already performed, it is necessary to image the abdomen and pelvis with CT. NCCN stated that advanced imaging may improve assessment of metastases, staging, and preoperative planning.[1] For individuals who have renal

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insufficiency and therefore are unable to undergo contrasted CT imaging of the abdomen and pelvis, the preferred substitute imaging is MRI with contrast. This imaging will provide the necessary level of soft tissue detail that would not be achievable with a non-contrasted CT. For those with primary peritoneal disease or inconclusive or conflicting CT imaging, NCCN stated "MRI has been shown to provide equivalent accuracy for staging and comparable accuracy for predicting peritoneal tumor volume, and can be useful if CT results are inconclusive".[1] Chest imaging via CT either with or without contrast is necessary for suspected pulmonary disease or if there is concern for metastatic disease. NCCN stated this imaging is helpful to detect pleural or pulmonary metastases, as well as pleural effusion, which may help with treatment planning.[1] PET is a helpful tool in evaluation of indeterminate lesions on other imaging as it is shown to have high accuracy for detection of metastases.[1]

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Ovarian Cancer – Restaging/Recurrence (ONC-21.4)

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Indication	Medically Necessary Imaging Study
Completely resected or definitively treated with chemotherapy and normal(ized) tumor markers	• No advanced imaging
ANY of the following: • Unresected disease • Unknown preoperative markers • Difficult or abnormal examination • Elevated LFTs • Elevated tumor markers (CA-125, inhibin) • Signs or symptoms of recurrence	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT for individuals with renal insufficiency or contrast allergy. • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for ANY of the following: ◦ Known prior thoracic disease ◦ New or worsening pulmonary symptoms ◦ New or worsening chest x-ray findings ◦ Rising tumor markers (CA-125, inhibin)
Monitoring response to treatment (every 2 cycles, or ~every 6 to 8 weeks)	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT for individuals with renal insufficiency or contrast allergy. • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for ANY of the following: ◦ Known prior thoracic disease ◦ New or worsening pulmonary symptoms ◦ New or worsening chest x-ray findings

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Indication	Medically Necessary Imaging Study
• CT negative or inconclusive and CA-125 continues to rise or elevated LFTs • Conventional imaging failed to demonstrate tumor or if persistent radiographic mass with rising tumor markers	ONE of the following: • MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) OR • PET/CT (CPT® 78815)
If both CT and MRI are inconclusive	• PET/CT (CPT® 78815)
If CT, MRI, and PET/CT are inconclusive	• PET/MRI (CPT® 78813 and 76498)

Evidence Discussion

If disease was completely resected and normal CA-125 or other markers normal, then no imaging is needed. If there any question or signs or symptoms of recurrence then the appropriate imaging is similar to that of initial staging. To monitor response to treatment, CT is appropriate every two cycles.

NCCN stated that advanced imaging may improve assessment of metastases and assist in staging.[1] For individuals who have renal insufficiency and therefore are unable to undergo contrasted CT imaging of the abdomen and pelvis, the preferred substitute imaging is MRI with contrast. This imaging will provide the necessary level of soft tissue detail that would not be achievable with a non-contrasted CT. NCCN stated "MRI has been shown to provide equivalent accuracy for staging and comparable accuracy for predicting peritoneal tumor volume, and can be useful if CT results are inconclusive".[1] Thus, MRI is a tool presented for use in the event that CT results are inconclusive or conflict with the clinical picture. Chest imaging via CT either with or without contrast is necessary for suspected pulmonary disease or if there is concern for metastatic disease. NCCN stated this imaging is helpful to detect pleural or pulmonary metastases, as well as pleural effusion, which may help with treatment planning.[1] PET/CT is a helpful tool in evaluation of indeterminate lesions on other imaging as it is shown to have high accuracy for detection of metastases.[1]

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Ovarian Cancer – Surveillance (ONC-21.5)

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Indication	Medically Necessary Imaging Study
Stages I-III	• Advanced imaging is not routinely indicated for surveillance
Measurable metastatic disease on maintenance therapy or being monitored off therapy	Every 3 months for up to 5 years after completion of active treatment: • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT for individuals with renal insufficiency. • CT with contrast of previously involved body areas • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for known history of chest involvement

Evidence Discussion

Stages I to III there is no need for advanced imaging if there are no signs or symptoms. If on maintenance or if there is measurable disease present, CT of the areas involved every three months for up to 5 years. For individuals who have renal insufficiency and therefore are unable to undergo contrasted CT imaging of the abdomen and pelvis, the preferred substitute imaging is MRI with contrast. This imaging will provide the necessary level of soft tissue detail that would not be achievable with a non-contrasted CT.[1]

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Uterine Cancer (ONC-22)

Guideline

Uterine Cancer – General Considerations (ONC-22.0)
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Uterine Cancer – Restaging/Recurrence (ONC-22.3)
Uterine Cancer – Surveillance (ONC-22.4)
Gestational Trophoblastic Neoplasia (GTN) (ONC-22.5)
References (ONC-22)

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Uterine Cancer – General Considerations (ONC-22.0)

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- Gestational trophoblastic neoplasia (GTN) – see: **Molar Pregnancy and Gestational Trophoblastic Neoplasia (GTN) (PV-16.1)** in the Pelvic Imaging Guidelines.
- Most common cell type is adenocarcinoma. Uterine sarcomas are also imaged according to this guideline.
- Staging of uterine cancer is primarily surgical. Advanced imaging is not routinely medically necessary pre-operatively for laparoscopic/minimally invasive surgery unless initial staging criteria are met. Pelvic and para-aortic lymphadenectomy can still be performed.

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Uterine Cancer – Suspected/Diagnosis (ONC-22.1)

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- See: **Abnormal Uterine Bleeding (PV-2.1)** in the Pelvic Imaging Guidelines for evaluation of suspected uterine malignancies.

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Uterine Cancer – Initial Work-Up/Staging (ONC-22.2)

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Indication	Medically Necessary Imaging Study
ANY of the following: • Extra-uterine disease suspected • To assess local extent of tumor prior to fertility-sparing surgery for well-differentiated Stage IA (grade 1) uterine cancer • Poor surgical candidate (due to medical comorbidities) considering medical therapy	• MRI Pelvis without and with contrast (CPT® 72197) • Transvaginal ultrasound (CPT® 76830) if MRI is contraindicated • Chest x-ray ◦ CT Chest with contrast (CPT® 71260) for: ▪ New or worsening pulmonary symptoms ▪ New or worsening chest x-ray findings In addition, for surgical staging: • Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
ANY of the following: • Abdominal symptoms or abnormal examination findings • Elevated LFTs • Other imaging studies suggest liver involvement	ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160)

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Indication	Medically Necessary Imaging Study
ANY of the following high-risk histologies: • Papillary serous • Clear cell • High-grade/poorly differentiated endometrioid carcinoma • Uterine sarcomas: ◦ Carcinosarcoma ◦ Soft tissue sarcoma of the uterus ◦ Leiomyosarcoma ◦ Rhabdomyosarcoma ◦ Undifferentiated sarcoma ◦ Endometrial stromal sarcoma	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
Tumors detected incidentally or incompletely staged surgically ***and*** ANY of the following high-risk features: • Myoinvasion >50% • Cervical stromal involvement • Lymphovascular invasion • Tumor >2 cm	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast
Inconclusive findings on conventional imaging	• PET/CT scan (CPT® 78815)

Evidence Discussion

- The National Comprehensive Cancer Network guidelines (NCCN) for non-fertility sparing treatment recommended considering chest imaging with chest x-ray.[1] If an abnormality is seen on chest x-ray, or if there are symptoms of chest involvement, then chest CT may be performed. Both NCCN and the American College of Radiology noted that MRI of the pelvis is the preferred imaging modality when pretreatment assessment of local tumor extent is indicated due to suspected extra-uterine disease or prior to fertility sparing treatment.[1,6]
- Transvaginal ultrasound can be done if MRI is contraindicated or unavailable.[1]
- If distant metastatic disease is suspected based on abnormal physical examination findings, or for high-grade endometrioid carcinoma, serous, clear cell or

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carcinosarcoma, cross-sectional imaging with CT Chest and CT Abdomen and Pelvis can be considered.[1]

- For incidental findings of endometrial cancer after hysterectomy or incompletely surgically staged with uterine risk factors (myoinvasion of over 50%, cervical stromal involvement or tumor larger than 2 cm), consideration should be given to CT Chest/ Abdomen and Pelvis to evaluate for distant metastatic disease per NCCN guidelines.[1]
- FDG-PET/CT in select individuals if metastases is suspected and other cross-sectional imaging is inconclusive.[1]
- Although endometrial cancer (EC) is surgically staged, preoperative imaging can help tailor surgery and medical treatment in cases as outlined by NCCN and ACR.[1,6] MRI or transvaginal ultrasound is valuable to assess local tumor extent. CT and/or PET-CT is valuable to assess lymph node metastases and distant spread. Preoperative imaging may identify deep myometrial invasion, cervical stromal involvement and metastatic disease. Although these imaging methods may provide information about likely tumor stage, the reported accuracies for preoperative staging of endometrial cancer by conventional imaging have not yet been good enough to replace surgical staging.[8]
- MRI Pelvis has long been established as a valuable imaging method in the preoperative staging of endometrial cancer because it allows the most accurate evaluation of the extent of the pelvis tumor. A meta-analysis showed that the efficacy of contrast-enhanced MRI is significantly better than that of non-contrast MRI and US, and tended toward better results than CT, in evaluating the depth of myometrial invasion in individuals with EC.[6]
- Transvaginal ultrasound may be used if MRI is contraindicated. A study found this imaging modality to have a 79.5% sensitivity and an 89.6% specificity for detecting deep myometrial invasion. However, MRI showed greater accuracy than ultrasound and ultrasound is limited in the setting of concomitant benign disease.[6]
- CT Chest, Abdomen and Pelvis may be used for detection of lymph node metastases, if distant metastatic disease is suspected as per NCCN and ACR listed above.[1,6]
- FDG-PET/CT may be indicated if distant metastatic disease is suspected and CT scans are inconclusive. A meta-analysis reported that the overall pooled sensitivity, specificity, and accuracy of using FDG-PET/CT for detection of lymph node metastasis in EC was 72.0%, 94.0%, and 88.0%, respectively.[6]

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Uterine Cancer – Restaging/Recurrence (ONC-22.3)

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Indication	Medically Necessary Imaging Study
• Unresected disease • Medically inoperable disease • Incomplete surgical staging • Difficult or abnormal examination • Elevated LFTs or rising tumor markers • Signs or symptoms of recurrence	• CT Chest (CPT® 71260) **and** • CT Abdomen and Pelvis with contrast (CPT® 74177)
Monitoring response to chemotherapy (every 2 cycles, ~every 6-8 weeks) for: • Unresected primary disease • Metastatic disease	• CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Chest with contrast (CPT® 71260) for: ◦ Known prior thoracic disease ◦ New or worsening pulmonary symptoms ◦ New or worsening chest x-ray findings
Any of the following: • After fertility sparing treatment • Inconclusive CT scan findings	• MRI Pelvis without and with contrast (CPT® 72197)
• Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

- Recurrence rates for low- or intermediate-risk individuals with endometrial cancer are infrequent. A recent review of post-treatment surveillance and diagnosis of recurrence in people assigned female at birth, or individuals with gynecologic cancers sponsored by the Society of Gynecologic Oncology recommends that radiologic evaluation be used only to investigate suspicion of recurrent disease because of symptoms or physical exam and not for routine surveillance after treatment.[5]
- CT Chest, Abdomen, and Pelvis is useful for suspected recurrence of disease based on abnormal physical examination findings and/or new pelvis, abdominal or pulmonary symptoms.[1] A study reported that 45 asymptomatic individuals had

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routine CT scans, and recurrence was diagnosed by CT in only 2 (4.4%), whereas 37 symptomatic individuals had CT scans for suspicion of recurrence, and it was confirmed by CT in 17 (46%).[6]

- MRI of the pelvis may be done after fertility sparing treatment for persistent endometrial carcinoma.[1] In individuals with persistent endometrial carcinoma after 6 months of failed hormonal therapy, pelvic MRI to exclude myoinvasion and nodal/ovarian metastasis is recommended before continuing fertility-sparing therapy.[1]
- FDG-PET/CT may give further clinically applicable information in cases where conventional imaging is inconclusive. As per the prior meta-analysis, the overall pooled sensitivity, specificity, and accuracy of using FDG-PET/CT for detection of lymph node metastasis in EC was 72.0%, 94.0%, and 88.0%, respectively.[6]

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Uterine Cancer – Surveillance (ONC-22.4)

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Indication	Medically Necessary Imaging Study
Stage I-III of uterine carcinoma	Advanced imaging is not routinely medically necessary
Measurable metastatic disease on maintenance therapy or being monitored off therapy	Every 3 months for up to 5 years after completion of definitive treatment: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved body areas
All stages of uterine sarcoma: • Soft tissue sarcoma of the uterus • Leiomyosarcoma • Adenosarcoma • Carcinosarcoma • Rhabdomyosarcoma • Undifferentiated sarcoma • Endometrial stromal sarcoma	CT Chest (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) every 3 months for 2 years, every 6 months for 3 years, and then every 1-2 years until year 10

Evidence Discussion

- Advanced imaging is not routinely indicated for surveillance for non-metastatic, asymptomatic disease in endometrial cancer. A review of post-treatment surveillance and diagnosis of recurrence in individuals with gynecologic cancers sponsored by the Society of Gynecologic Oncology recommended that radiologic evaluation be used only to investigate suspicion of recurrent disease because of symptoms or physical exam and not for routine surveillance after treatment.[5]
- Measurable metastatic disease can be followed with CT Abdomen and Pelvis and CT of previously involved body areas every 3 months for 5 years after treatment.
- All stages of uterine sarcoma, CT Chest, Abdomen and Pelvis every 3 months for 2 years, every 6 months for 3 years and then every 1-2 years until year 10.[1]

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Gestational Trophoblastic Neoplasia (GTN) (ONC-22.5)

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- The most common form of gestational trophoblastic disease (GTD) is hydatidiform mole (HM), a benign form, also known as molar pregnancy.
  - See: **Molar Pregnancy and GTN (PV-16.1)**
- Gestational trophoblastic neoplastic disorders including a malignant form of GTD, and can present as invasive mole, choriocarcinoma, placental site trophoblastic tumor (PSTT), or epithelioid trophoblastic tumor (ETT). GTN cells are malignant and can metastasize to other organs such as lungs, brain, bone and vagina. These tumors have a high likelihood of cure and treatment with methotrexate usually allows for fertility preservation.
- Surveillance is generally with serial monitoring of hCG levels, and advanced imaging is reserved for high-risk histologies where hCG levels may not be a reliable marker.

Indication	Medically Necessary Imaging Study
Initial staging	• CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
EITHER of the following: • Pulmonary metastases noted on CT scan • Signs/symptoms of CNS involvement	• MRI Brain without and with contrast (CPT® 70553)
EITHER of the following: • Monitoring response to systemic therapy (every 2 cycles, i.e., 6-8 weeks) • Suspected progression	• CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)

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Indication	Medically Necessary Imaging Study
Surveillance for any of the following high risk histologies: • Placental site trophoblastic tumor (PSTT) • Epithelioid trophoblastic tumor (ETT)	Annually for 2 years: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)

Evidence Discussion

- The most common form of GTD is hydatidiform mole (HM), also known as molar pregnancy. HMs are considered a benign, premalignant disease.[7]
- Initial determination of suspected HM is often made based on ultrasound findings in combination with clinical symptoms and hCG levels.[7]
- Gestational trophoblastic neoplastic disorders include a malignant form of GTD, and can present as invasive mole, choriocarcinoma, placental site trophoblastic tumor (PSTT), or epithelioid trophoblastic tumor (ETT). GTN cells are malignant and can metastasize to other organs such as lungs, brain, bone and vagina.
- Initial staging of GTN includes a CT of chest and CT of abdomen and pelvis.
- If pulmonary metastases are noted on CT Chest or for signs or symptoms of central nervous system (CNS) involvement, MRI Brain is medically necesssary to evaluate for metastatic disease. Rates of CNS metastases are low with post-molar GTN, but approximately 20% of individuals with choriocarcinoma have CNS involvement.[7]
- For monitoring response to treatment or for suspected progression CT Chest, Abdomen and Pelvis is performed.
- Post-treatment surveillance in general is done with monitoring of hCG levels in individuals with post-molar GTN or choriocarcinoma, where hCG is a reliable tumor marker.[7]
- Surveillance imaging for placental site or epithelioid trophoblastic tumor can be done with CT Chest, Abdomen, and Pelvis annually for 2 years. Post-treatment imaging is medically necessary for follow-up after treatment of PSTT and ETT, where hCG is a less reliable tumor marker.[7]

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Cervical Cancer (ONC-23)

Guideline

Cervical Cancer – General Considerations (ONC-23.0)
Cervical Cancer – Suspected/Diagnosis (ONC-23.1)
Cervical Cancer – Initial Work-Up/Staging (ONC-23.2)
Cervical Cancer – Restaging/Recurrence (ONC-23.3)
Cervical Cancer – Surveillance (ONC-23.4)
References (ONC-23)

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Cervical Cancer – General Considerations (ONC-23.0)

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- Primary histology for cervical cancer is squamous cell. Other, less common histologies are adenosquamous and adenocarcinoma. If biopsy is consistent with one of these less common histologies, it is necessary to clarify that tumor is not of primary uterine origin.
- If the primary histology is uterine in origin, follow imaging recommendations for uterine cancer, see: **Uterine Cancer (ONC-22)**.

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Cervical Cancer – Suspected/Diagnosis (ONC-23.1)

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Indication	Medically Necessary Imaging Study
All	• Advanced imaging is not medically necessary prior to biopsy

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Cervical Cancer – Initial Work-Up/ Staging (ONC-23.2)

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Indication	Medically Necessary Imaging Study
All individuals with one of the following: • Stage I-IVA • Stage IVB amenable to local treatment	• MRI Pelvis without and with contrast (CPT® 72197)
Stage IB1-IB3, non-fertility sparing treatment	In addition to MRI Pelvis above, any ONE of the following combinations: • PET/CT (CPT® 78815) or • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) or • PET/MRI (CPT® 78813 and CPT® 76498)
ANY of the following: • Stage 1B1-IB3, fertility-sparing treatment • Stage II-IVA • Any size cervical cancer incidentally found in a hysterectomy specimen	In addition to MRI Pelvis above, any ONE of the following combinations: • PET/CT (CPT® 78815) or • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) or • CT Chest without contrast (CPT® 71250) and CT Abdomen and Pelvis with contrast (CPT® 74177)

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Indication	Medically Necessary Imaging Study
Small cell neuroendocrine carcinoma of the cervix (NECC)	MRI Brain without and with contrast (CPT® 70553) and any ONE of the following combinations: • PET/CT (CPT® 78815) or • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) or • CT Chest without contrast (CPT® 71250) and CT Abdomen and Pelvis with contrast (CPT® 74177)
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

- MRI Pelvis is recommended by NCCN in virtually all individuals, with exception of stage IVB not amenable to local treatment. MRI Pelvis imaging is used to assess local disease extent, to help with surgical treatment planning, and to help with any radiation treatment planning.[1]
- Systemic imaging is indicated to assess for lymphadenopathy and distant metastases. The goal of systemic imaging is to evaluate for metastatic disease. CT imaging of the chest, abdomen, and pelvis are useful; PET/CT is the recommended study of choice in higher stages of disease. NCCN stated of non-fertility-sparing approaches, "depending on their stage and disease bulk, individuals with stage IB or IIA tumors can be treated with surgery, RT, or concurrent chemoradiation. A combined PET/CT scan can be performed to rule out extra pelvic disease before deciding how to treat these individuals. Neck/chest/abdomen/pelvis/groin FDG-PET/CT is preferred to evaluate metastatic disease. A chest/abdomen/pelvis CT or FDG-PET/MRI can also be performed."[1]
- Meta-analyses have shown CT with intravenous (IV) contrast to have 43%-55% sensitivity and 71% specificity for parametrial invasion, and 41% sensitivity and 92% specificity for bladder invasion In comparison, MRI demonstrated 71% specificity (95% confidence interval [CI], 62%-79%) and 91% sensitivity (95% CI, 88%-93%) for parametrial invasion, and 84% sensitivity (95% CI, 57%-95%) and 95% specificity (95% CI, 87%-98%) for bladder invasion.
- Small cell neuroendocrine carcinoma of the cervix (NECC) is considered an aggressive form of cervical cancer. The NCCN-recommended studies are adapted

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from their own guidelines on neuroendocrine tumors and non-small cell lung cancers.[1] Brain imaging is medically necessary in this cell type to evaluate for metastatic disease to the brain.

- PET/CT remains a useful tool for evaluation of inconclusive findings on conventional imaging.

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Cervical Cancer – Restaging/Recurrence (ONC-23.3)

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Indication	Medically Necessary Imaging Study
Stage I treated with definitive surgery	• See: **Cervical Cancer – Surveillance (ONC-23.4)**
EITHER of the following: • Stage IB3 • Stage I treated with post-operative adjuvant radiation or chemoradiation due to positive nodes, positive parametria, positive margins, or local cervical factors	• PET/CT (CPT® 78815) 3-6 months post-completion of therapy
Stage II-IV	ONE of the following within 3-6 months of completion of therapy: • PET/CT (CPT® 78815) or • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast)
Small cell NECC, treated with primary chemoradiation	ONE of the following after completion of therapy: • PET/CT (CPT® 78815) and MRI Brain without and with contrast (CPT® 70553) or • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast and MRI Brain without and with contrast (CPT® 70553)
Inconclusive findings on CT scan or PET/CT	MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
Unresectable disease or metastatic disease on systemic treatment	Every 2 cycles of treatment (commonly every 6 to 8 weeks): • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of other involved or symptomatic areas
Inconclusive conventional imaging	• PET/CT (CPT® 78815)
Suspected or biopsy proven recurrence	EITHER of the following, not both: • PET/CT (CPT® 78815) OR • CT Chest (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)

Evidence Discussion

- The main purpose of imaging after primary surgery is to determine whether distant metastases are present.[1] Thus, repeating systemic imaging for certain individuals in whom surgery was not definitive treatment is recommended.
- For unresectable or metastatic disease on systemic treatment, CT Chest/Abdomen/Pelvis every 2 cycles is appropriate.
- PET/CT remains a useful tool for evaluation of inconclusive conventional imaging. MRI Pelvis, given its excellent resolution of soft-tissue imaging, is a recommended tool for evaluation of an inconclusive CT or PET/CT.[1]

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Cervical Cancer – Surveillance (ONC-23.4)

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Indication	Medically Necessary Imaging Study
Stage I disease treated with fertility sparing approach	• MRI Pelvis without and with contrast (CPT® 72197) at 6 months after surgery and then annually for 2-3 years
Small cell NECC	Once, 3-6 months after completion of therapy: • PET/CT (CPT® 78815) Then, every 3 months for 1 year, then every 6 months: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast and MRI Brain without and with contrast (CPT® 70553)
All other individuals	• No routine advanced imaging in asymptomatic individuals.

Evidence Discussion

With stage I post fertility-sparing treatment, MRI Pelvis is supported by NCCN for surveillance imaging at 6 months post-operatively and then annually for 2-3 years. For small-cell NECC, NCCN supported PET/CT at completion of therapy and supported ongoing surveillance, but a timeframe is not provided. There are little data in the medical literature specific to this topic. It is therefore reasonable to follow the NCCN-recommended surveillance schedule for extrathoracic small-cell neuroendocrine tumors for small-cell NECC. For small-cell NECC, NCCN supported PET/CT at completion of therapy and supported ongoing surveillance, but a timeframe is not provided. There are little data in the medical literature specific to this topic. It is therefore reasonable to follow the NCCN-recommended surveillance schedule for extrathoracic small cell neuroendocrine tumors for small cell NECC.[1]

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Anal Cancer & Cancers of the External Genitalia (ONC-24)

Guideline

Anal Carcinoma – General Considerations (ONC-24.0)
Anal Carcinoma – Suspected/Diagnosis (ONC-24.1)
Anal Carcinoma – Initial Work-up/Staging (ONC-24.2)
Anal Carcinoma – Restaging/Recurrence (ONC-24.3)
Anal Carcinoma – Surveillance (ONC-24.4)
Cancers of External Genitalia – General Considerations (ONC-24.5)
Cancers of External Genitalia – Initial Work-Up/Staging (ONC-24.6)
Cancers of External Genitalia – Restaging/Recurrence (ONC-24.7)
Cancers of External Genitalia – Surveillance (ONC-24.8)
References (ONC-24)

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Anal Carcinoma – General Considerations (ONC-24.0)

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- Most are squamous cell carcinomas, although some transitional and cloacogenic carcinomas are seen.
- Adenocarcinoma of the anal canal is managed as rectal cancer according to **Colorectal and Small Bowel Cancer (ONC-16)**.
- Squamous cell carcinoma of the perianal region (up to 5 cm radius from the anal verge) are imaged according to anal carcinoma guidelines.
- Bowen's disease and Paget's disease of the perianal and perigenital skin are considered non-invasive/in-situ conditions and do not routinely require advanced imaging. See: **Non-Melanoma Skin Cancers – Initial Work-up/Staging (ONC-5.6)**.

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Anal Carcinoma – Suspected/Diagnosis (ONC-24.1)

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Indication	Medically Necessary Imaging Study
All	• No advanced imaging prior to biopsy

Evidence Discussion

Advanced imaging prior to biopsy is not medically necessary as most tumors are staged clinically by direct examination and microscopic confirmation (biopsy).[1]

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Anal Carcinoma – Initial Work-up/ Staging (ONC-24.2)

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Indication	Medically Necessary Imaging Study
All individuals	• CT Chest with contrast (CPT® 71260) and Any ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Abdomen with contrast (CPT® 74160) and MRI Pelvis without and with contrast (CPT® 72197)
• Stage II-III Squamous Cell Carcinoma of the Anal Canal and no evidence of metastatic disease by conventional imaging • Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

All individuals undergo CT Chest with contrast and either CT Abdomen and Pelvis with contrast or CT Abdomen with contrast and MRI Pelvis without/with contrast. CT allows information on whether there is other organ involvement or possible disease spread. PET/CT is supported in stage II-III disease with no evidence of distant metastatic disease by conventional imaging or if conventional imaging is inconclusive. PET/CT is useful in assessing pelvic nodes and has been shown to change the nodal status/TNM stage in up to 41% of individuals. PET/CT does not replace a diagnostic CT in initial staging.[1,11]

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Anal Carcinoma – Restaging/Recurrence (ONC-24.3)

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Indication	Medically Necessary Imaging Study
Stage I treated with complete surgical resection	• See: **Anal Carcinoma – Surveillance (ONC-24.4)** for surveillance guidelines
Stages I, II and III – post chemoradiation evaluation	Any ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Metastatic (stage IV) disease	• CT Abdomen and Pelvis with contrast (CPT® 74177) every 2 cycles (generally 6 to 8 weeks) on treatment • CT Chest with contrast (CPT® 71260) if chest x-ray is abnormal or if symptoms of chest involvement
• Difficult or abnormal examination • Elevated LFTs • Signs or symptoms of recurrence • Biopsy proven recurrence	Preferably using same imaging studies as used in initial imaging: • CT Chest (CPT® 71260) with contrast and Any ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

Due to low risk of recurrence, stage I treated with complete surgical resection would follow surveillance guidelines that do not recommend any routine imaging. For stages I, II, and III treated with chemoradiation, CT Abdomen and Pelvis, or MRI Abdomen and Pelvis should be obtained upon completion of therapy. Stage IV disease on treatment should undergo CT Abdomen and Pelvis every 2 cycles with imaging of the chest if chest x-ray is abnormal or symptoms develop. If recurrence is suspected, CT Chest

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with either CT Abdomen and Pelvis or MRI Abdomen and Pelvis should be performed. NCCN stated that imaging for recurrence is preferred to mirror the same imaging studies as used in initial imaging. PET/CT is indicated for inconclusive findings on conventional imaging.[1]

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Anal Carcinoma – Surveillance (ONC-24.4)

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Indication	Medically Necessary Imaging Study
Stage I	• Advanced imaging is not routinely indicated for surveillance
• Stage II • Stage III • Local recurrence treated definitively	• CT Chest (CPT® 71260) with contrast or CT Chest without contrast (CPT® 71250) annually for 3 years • And ANY one of the following annually for three years: ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Stage IV – measurable metastatic disease on maintenance treatment or being observed off treatment	Every 3 months for up to 5 years after completion of all treatment: • CT Chest (CPT® 71260) with contrast • CT Abdomen and Pelvis with contrast (CPT® 74177)

Evidence Discussion

For individuals with Stage II-III disease or had definitive treatment of a local recurrence, CT Chest with or without contrast plus either CT Abdomen and Pelvis with contrast or MRI Abdomen and Pelvis without/with contrast is medically necessary annually for 3 years. NCCN stated that FDG PET/CT is not indicated for surveillance of local recurrence treated definitively.[1]

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Cancers of External Genitalia – General Considerations (ONC-24.5)

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- These imaging guidelines are applicable for squamous cell carcinomas arising from the vulva, vagina, penis, urethra, and scrotum.

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Cancers of External Genitalia – Initial Work-Up/Staging (ONC-24.6)

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Indication	Medically Necessary Imaging Study
Clinical node negative **vulvar** cancer with ANY of the following: • Lesion >2 cm • Any size with stromal invasion >1 mm	• For planned sentinel lymph node evaluation: Lymph system imaging (lymphoscintigraphy, CPT® 78195) ◦ SPECT/CT (CPT® 78830) if requested
For stage II or higher **vulvar** cancer or suspected metastatic vulvar cancer based on one of the following: • Abnormal physical exam findings • Bulky vulvar tumor (≥4 cm or close to critical structures) • Vaginal, urethral, or anal involvement • Delay in presentation or treatment • Pelvic, abdominal, or pulmonary symptoms	ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) OR • CT Abdomen with contrast (CPT® 74160) and MRI Pelvis without and with contrast (CPT® 72197) • CT Chest with contrast (CPT® 71260) is medically necessary only for: ◦ Signs/symptoms suggestive of chest involvement ◦ Abnormal findings on chest x-ray

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Indication	Medically Necessary Imaging Study
Vaginal carcinoma	ONE of the following combinations: • PET/CT (CPT® 78815) and MRI Pelvis without and with contrast (CPT® 72197) or • CT Abdomen and Pelvis with contrast (CPT® 74177) and CT Chest with contrast (CPT® 71260) or • CT Abdomen with contrast (CPT® 74160) and MRI Pelvis without and with contrast (CPT® 72197) and CT Chest with contrast (71260)
Penile carcinoma with ANY of the following: • Non-palpable inguinal lymph nodes and intermediate or high-risk lesion (T1b or any T2 or greater) • Palpable inguinal lymph nodes • Difficult pelvic node assessment on physical exam	Any ONE of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • CT Abdomen with contrast (CPT® 74160) and MRI Pelvis without and with contrast (CPT® 72197) • CT Chest with contrast (CPT® 71260)
• Inconclusive findings on conventional imaging • Pelvic nodes enlarged but biopsy not technically feasible	• PET/CT (CPT® 78815)

Evidence Discussion

Lymphoscintigraphy is appropriate for the workup of vulvar cancer since the disease spreads through skin layer and into the lymph system. FIGO surgical staging is used after superficial removal of the lesion. If it is a large (>2 cm) or stromal invasion (>1 mm) then spread is evaluated with CT. CT imaging plays an important role in stage II or higher disease by helping to delineate the extent of tumor and assisting in treatment planning.[3] CT of the chest is valuable if there are signs or symptoms of chest involvement. In the evaluation of vaginal carcinoma, NCCN stated that PET/CT is the preferred imaging for evaluation of metastatic disease (conventional CT of the

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chest, abdomen, and pelvis remains a recommended option). MRI Pelvis is medically necessary in addition to PET/CT or conventional CT for assessment of local disease extent.[14] For the initial workup of penile cancer, cross-sectional imaging with CT or MRI of the abdomen and pelvis are necessary for pelvic node evaluation in circumstances where there is greater chance of lymph node involvement, palpable lymph nodes, or when it is difficult to physically assess potential lymph node involvement.[2] Evaluation of inguinal lymph nodes "has been identified as the single most important prognostic indicator in determining long-term survival in individuals with invasive penile squamous cell carcinoma".[2] MRI can also provide information on depth of tumor invasion.[2] PET/CT remains a problem-solving tool for inconclusive conventional imaging. PET/CT has higher sensitivity for individuals with node-positive penile cancer than those without, thus it is used as a tool of lymph node assessment when biopsy is not technically feasible.[2]

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Cancers of External Genitalia – Restaging/Recurrence (ONC-24.7)

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Indication	Medically Necessary Imaging Study
• Difficult or abnormal examination • Elevated LFTs • Signs or symptoms of recurrence • Biopsy proven recurrence	• CT Chest with contrast (CPT® 71260) And ANY one of the following: • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast
EITHER of the following: • individuals receiving systemic treatment for measurable disease • At the end of definitive therapy	Any ONE of the following, every 2 cycles (generally 6 to 8 weeks) during treatment at at the end of planned chemotherapy treatment: • CT Abdomen and Pelvis with contrast (CPT® 74177) or • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast or • CT Abdomen with contrast (CPT® 74160) and MRI Pelvis (CPT® 72197) AND • CT Chest with contrast (CPT® 71260)
Vaginal primary tumor treated with upfront radiation therapy	• PET/CT (CPT® 78815) at least 12 weeks after completion of radiation therapy ◦ MRI Pelvis without and with contrast (CPT® 72197) is medically necessary if PET/CT not available (can be performed sooner than 12 weeks after completion of therapy) ◦ MRI Pelvis without and with contrast (CPT® 72197) is medically necessary for clarification of PET/CT findings
Vulvar carcinoma following definitive primary therapy with chemoradiation or surgery	3-6 months after primary therapy: • MRI Pelvis without and with contrast (CPT® 72197)

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Indication	Medically Necessary Imaging Study
Inconclusive findings on conventional imaging	• PET/CT (CPT® 78815)

Evidence Discussion

Signs or symptoms of recurrence should be imaged with CT or MRI to evaluate for metastatic disease. Specifically for vaginal cancer, PET/CT is preferred to re-image three to four months after the completion of radiation therapy to determine response to treatment while avoiding false-positive from recent radiation. If PET/CT is inconclusive, NCCN stated that MRI is the preferred imaging for clarification.[14] For vulvar cancer, reassessment with MR imaging is necessary to assess response to treatment and re-evaluate lymph node involvement.[3] PET/CT can be used as a tool for evaluation of inconclusive conventional imaging.

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Cancers of External Genitalia – Surveillance (ONC-24.8)

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Indication	Medically Necessary Imaging Study
• All stages of vulvar and vaginal cancers	• See: **ONC-24.7** for post-treatment imaging 3-6 months after completion of therapy. No additional routine advanced imaging for asymptomatic surveillance.
• Penile Cancer: stage I-IIIA	• CT Abdomen and Pelvis with contrast (CPT® 74177) every 6 months for 2 years, and then every 12 months for 2 years
• Penile cancer: stages IIIB-IV	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) every 3 months for one year, then every 6 months for 3 more years

Evidence Discussion

If no symptoms or findings on recent physical examination then advanced imaging is not medically necessary for all stages of vulvar and vaginal cancer.[3,14] For stage I-IIIA penile cancer, the NCCN supported CT Abdomen and Pelvis for surveillance, with chest imaging via radiograph only on the same schedule as CTs. For stages IIIB-IV penile cancer, chest CT is supported for surveillance with more frequent CT Abdomen and Pelvis than in early stage disease.[2]

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Multiple Myeloma and Plasmacytomas (ONC-25)

Guideline

Multiple Myeloma and Plasmacytomas – General Considerations (ONC-25.0)
Multiple Myeloma and Plasmacytomas –Suspected/Diagnosis (ONC-25.1)
Multiple Myeloma and Plasmacytomas – Initial Work-Up/Staging (ONC-25.2)
Multiple Myeloma and Plasmacytomas – Restaging/Recurrence (ONC-25.3)
Multiple Myeloma and Plasmacytomas – Surveillance (ONC-25.4)
References (ONC-25)

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Multiple Myeloma and Plasmacytomas – General Considerations (ONC-25.0)

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- Multiple myeloma (MM) is a neoplastic disorder characterized by the proliferation of a single clone of plasma cells derived from B cells which grows in the bone marrow and adjacent bone, producing skeletal destruction.
- Multiple myeloma group of disorders can be classified as below, which influence imaging modality of choice.

Condition	Monoclonal protein	Bone marrow plasma cells	CRAB criteria**
Solitary Plasmacytoma (biopsy proven tumor containing plasma cells)	<3 gm/dL	Absent	Absent
Monoclonal Gammopathy of Unknown Significance (MGUS)	<3 gm/dL	<10%	Absent
Smoldering Myeloma (SMM) (stage I MM or asymptomatic MM)	≥3 gm/dL	10% - 60%	Absent
Multiple Myeloma (MM)	≥3 gm/dL	≥10%	Present

**CRAB criteria = hypercalcemia, renal insufficiency, anemia, lytic bony lesions

- Diagnosis and monitoring of response to therapy is primarily with laboratory studies that include urine and serum monoclonal protein levels, serum-free light chain levels, LDH and beta-2 microglobulin. Routine advanced imaging to monitor response to treatment is not medically necessary.
- Rarely, (<5%), an individual may have nonsecretory myeloma, which does not produce measurable M-protein. These individuals require imaging as primary method to monitor disease.
- Other conditions that may present with monoclonal gammopathy include:
  - POEMS syndrome: Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein and Skin Changes – may also have sclerotic bone lesions and Castleman's disease. See: **Multiple Myeloma and Plasmacytomas – Initial Work-up/Staging (ONC-25.2)** for imaging recommendations.

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- Waldenström's Macroglobulinemia: IgM monoclonal protein along with bone marrow infiltration of small lymphocytes. See: **Waldenström Macroglobulinemia or Lymphoplasmacytic Lymphoma (ONC-27.10)** for imaging recommendations.
- Systemic Light chain Amyloidosis: light chain monoclonal protein in serum or urine with clonal plasma cells in bone marrow, systemic involvement of the kidneys, liver, heart, gastrointestinal tract or peripheral nerves due to amyloid deposition. See: **Multiple Myeloma and Plasmacytomas – Initial Work-up/Staging (ONC-25.2)** and **Cardiac Amyloidosis (CD-3.8)**for imaging recommendations for systemic light chain amyloidosis.

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Multiple Myeloma and Plasmacytomas – Suspected/Diagnosis (ONC-25.1)

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See: **Initial Work-up/Staging (ONC-25.2)**

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Multiple Myeloma and Plasmacytomas – Initial Work-Up/Staging (ONC-25.2)

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Indication	Medically Necessary Imaging Study
Either of the following: • Confirmed myeloma • Myeloma suspected based on any of the following: ◦ Abnormal skeletal survey or other imaging ◦ Abnormal myeloma labs ◦ Signs/symptoms of multiple myeloma	EITHER of the following: • Whole-body low-dose skeletal CT (CPT® 76497) or • Whole-body FDG PET/CT (CPT® 78816)
Inconclusive findings on whole-body low-dose skeletal CT	• PET/CT (CPT® 78815 or CPT® 78816)
To discern smoldering myeloma from active myeloma and whole-body CT or PET are negative or inconclusive	ONE of the following: • MRI Bone Marrow Blood Supply (CPT® 77084) • Whole-body MRI (CPT® 76498) • CT contrast as requested of a specific area to determine radiotherapy or surgical candidacy, or for suspected extra-osseous plasmacytoma
Initial work-up of suspected or known POEMS syndrome	• CT Chest with contrast (CPT® 71260) and • CT Abdomen and Pelvis with contrast (CPT® 74177) • Whole-body FDG PET/CT (CPT® 78816) for sclerotic bone lesions on other imaging

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Indication	Medically Necessary Imaging Study
Initial work-up of suspected or known systemic light chain amyloidosis	• Whole-body low-dose skeletal CT (CPT® 76497) OR Whole-body FDG PET/CT (CPT® 78816) • Additional imaging, base on suspected organ involvement: ◦ CT Chest with contrast (CPT® 71260) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) For additional imaging related to cardiac involvement, see: **Cardiac Amyloidosis (CD-3.8)**

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Multiple Myeloma and Plasmacytomas – Restaging/Recurrence (ONC-25.3)

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Indication	Medically Necessary Imaging Study
Extra-osseous plasmacytoma response to initial therapy	Repeat imaging with ONE of the following, whichever modality was used at initial diagnosis: • Whole-body low-dose skeletal CT scan (CPT® 76497) • Whole-body FDG PET/CT (CPT® 78816) • Whole-body MRI (CPT® 76498) • CT of any previously involved area, contrast as requested • MRI of any previously involved area, contrast as requested
Known spine involvement with new neurological signs/symptoms or worsening pain	• MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), Lumbar spine (CPT® 72158) without and with contrast
Treatment response assessment • After completion of primary therapy • Non-secretory multiple myeloma • To determine therapy response with inconclusive labs	Repeat imaging with ONE of the modalities below, whichever was used at initial diagnosis: • Whole-body low-dose skeletal CT scan (CPT® 76497) • Whole-body FDG PET/CT (CPT® 78816)
CAR-T cell therapy	Once before treatment and once 30-60 days after completion of treatment: • PET/CT (CPT® 78815 or CPT® 78816)
ANY of the following: • Suspected progression or relapse/recurrence • Suspected progression of MGUS or SMM to a more malignant form	EITHER of the following: • Whole-body low-dose skeletal CT (CPT® 76497) • Whole-body FDG PET/CT (CPT® 78816)

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Indication	Medically Necessary Imaging Study
ANY of the following: • Negative PET will allow change in management from active treatment to maintenance or surveillance. • Inconclusive findings on conventional imaging • Whole-body low-dose skeletal CT (CPT® 76497) is unfeasible and recurrence or progression is suspected	• PET/CT (CPT® 78815 or CPT® 78816)
To discern smoldering myeloma from active myeloma and whole-body CT or PET are negative or inconclusive	ONE of the following: • MRI Bone Marrow Blood Supply (CPT® 77084) • Whole-body MRI (CPT® 76498) • MRI without contrast, or MRI without and with contrast for any previously involved bony area or symptomatic area (including whole spine for spinal symptoms)
Stem cell transplant recipients	ONE of the following, once before transplant and once within 30-100 days after transplant: Imaging should use same modality as initial diagnosis. • Whole-body low-dose skeletal CT scan (CPT® 76497) • MRI Bone Marrow Blood Supply (CPT® 77084) • Whole-body MRI (CPT® 76498) • PET/CT (CPT® 78815 or CPT® 78816)

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Multiple Myeloma and Plasmacytomas – Surveillance (ONC-25.4)

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Indication	Medically Necessary Imaging Study
• Smoldering myeloma • Multiple myeloma	ANY ONE of the following (using same imaging modality used at diagnosis) annually: • Whole-body low-dose skeletal CT (CPT® 76497) • MRI Bone Marrow Blood Supply (CPT® 77084) • Whole-body MRI (CPT® 76498) • Whole-body FDG PET/CT (CPT® 78816)
Solitary plasmacytomas	Any ONE of the following (using same imaging modality used at diagnosis) annually for 5 years: • Whole-body low-dose skeletal CT (CPT® 76497) • MRI Bone Marrow Blood Supply (CPT® 77084) • Whole-body MRI (CPT® 76498) • Whole-body FDG PET/CT (CPT® 78816)

Evidence Discussion

- Bone disease is the most frequent feature of multiple myeloma (MM), occurring in approximately two-thirds of individuals at diagnosis and in nearly all individuals during their disease.[13] Imaging is a key part of the evaluation of all individuals with suspected MM and plays a very important role in the management of MM.[26] It is necessary for detection of lytic bone lesions, which represent a marker of disease-related end-organ damage and are traditionally used to diagnose MM and to establish the need for immediate therapy.[3]
- The detection of bone and bone marrow lesions is crucial in the investigation of multiple myeloma and often dictates the decision to start treatment. Cross-sectional imaging (i.e., CT, PET/CT, MRI) is preferred because these modalities are more sensitive than plain radiographs for the detection of most skeletal lesions in MM.
- Whole-body low-dose CT (WBLDCT) can be used as a baseline assessment of bone involvement. CT is quick, convenient, relatively sensitive, and cost-effective in this scenario. WBLDCT was introduced to detect osteolytic lesions in the whole skeleton, with high accuracy, no need for contrast agents, and twofold to threefold lower radiation dose exposure compared with standard CT.[17,18] Low-dose whole-

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body CT has increased sensitivity compared with conventional skeletal survey in the detection of bone disease, which can reveal information leading to changes in therapy and disease management that could prevent or delay the onset of clinically significant morbidity and mortality as a result of skeletal-related events.[16]

- 18F-FDG PET/CT imaging could identify sites of extra-medullary disease (EMD), which represent an unfavorable prognostic feature, and it helps to accurately differentiate between solitary plasmacytoma (SP) and MM, as well as to predict the risk of early progression from smoldering MM (SMM) to active disease. This is more sensitive than CT for the detection of extra medullary disease. The combination of functional imaging with positron emission tomography (PET) plus morphological assessment with CT makes this technique the most effective in identifying potential sites of EMD.[19] The use of FGD PET/CT can also confirm a suspected diagnosis of solitary plasmacytoma and can distinguish between smoldering and active multiple myeloma. Thus, NCCN stated that FDG PET/CT is the preferred imaging for initial diagnostic workup.[1]
- Whole-body MRI can distinguish between smoldering and active multiple myeloma, acting as a problem-solving tool when PET/CT or whole-body CT is inconclusive.[1]
- MRI is the elective imaging technique to assess the degree of BM PC infiltration, even before bone destruction is present, owing to its ability to visualize large volumes of BM.[7] MRI is highly sensitive for the detection of bone and bone marrow focal lesions and predictive of progression. Unlike CT and PET/CT, MRI can detect focal bone lesions that are not yet lytic (i.e., without advanced cortical bone destruction). Up to half of individuals without other evidence of end-organ damage with normal plain films may demonstrate tumor-related lesions on MRI.[21,22]
- Whole-body diffusion weighted MRI (DW-MRI), also known as MRI Bone Marrow Blood Supply, is a non-contrast study that covers from the vertex to the heels. Diffusion-weighted magnetic resonance imaging (DWI or DW-MRI) is the use of specific MRI sequences as well as software that generates images from the resulting data that uses the diffusion of water molecules to generate contrast in MR images. This produces images where the contrast between tissues is based on differences in the motion of water at a cellular level. As cellularity in marrow increases secondary either to disease or increased hematopoietic tissue, the amount of free water increases. The capability of WB DW-MRI to demonstrate both focal and diffuse marrow infiltration throughout the whole skeleton makes this extremely useful as a subjective tool for monitoring disease status and assessment of response.[23]

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Leukemias, Myelodysplasia and Myeloproliferative Neoplasms (ONC-26)

Guideline

Leukemias, Myelodysplasia and Myeloproliferative Neoplasms – General Considerations (ONC-26.1)
Acute Leukemias (ONC-26.2)
Chronic Myeloid Leukemias, Myelodysplastic Syndrome and Myeloproliferative Disorders (ONC-26.3)
Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Lymphoma (SLL) (ONC-26.4)
References (ONC-26)

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Leukemias, Myelodysplasia and Myeloproliferative Neoplasms – General Considerations (ONC-26.1)

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- Routine advanced imaging is not medically necessary for the evaluation and management of hairy cell leukemia in the absence of specific localizing clinical symptoms.[7,8]

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Acute Leukemias (ONC-26.2)

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- Imaging indications for acute lymphoblastic leukemia in adult individuals are identical to those for pediatric individuals. See: **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Imaging indications for acute myeloid leukemia in adult individuals are identical to those for pediatric individuals. See: **Acute Myeloid Leukemia (AML) (PEDONC-3.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Chronic Myeloid Leukemias, Myelodysplastic Syndrome and Myeloproliferative Disorders (ONC-26.3)

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- Routine advanced imaging is not medically necessary in the evaluation and management of chronic myeloid leukemias, myelodysplastic syndromes or myeloproliferative disorders in the absence of specific localizing clinical symptoms or clearance for hematopoietic stem cell transplantation.
- See: **Hematopoietic Stem Cell Transplantation (ONC-29)** for imaging guidelines related to transplant.
- For work-up of elevated blood counts, see: **Paraneoplastic Syndromes – General Considerations (ONC-30.3)**.

Evidence Discussion

It is not routinely recommended to utilize advanced imaging for chronic myeloid leukemia, myelodysplastic syndromes, and myeloproliferative disorders. In the interest of safety such that infectious and iatrogenic complications are assessed in a timely manner, these guidelines provide flexibility for approval of advanced imaging for specific localizing symptoms, and a separate guideline section for imaging related to stem cell transplantation.[6,12,13]

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Chronic Lymphocytic Leukemia (CLL)/ Small Lymphocytic Lymphoma (SLL) (ONC-26.4)

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- PET imaging is not medically necessary in the evaluation of CLL/SLL with the exception of suspected Richter's transformation (see suspected transformation, below).
- CLL/SLL is monitored with serial laboratory studies. Routine advanced imaging is not medically necessary for monitoring treatment response or surveillance, except when initial studies reveal bulky disease involvement.
- Bulky disease is defined as lymph node mass >10 cm or spleen >6 cm below costal margin.

Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	• Routine advanced imaging is not medically necessary for initial evaluation of asymptomatic individuals
For ANY of the following: • Bulky lymph node mass (>10 cm) • Splenomegaly >6 cm below costal margin • Presence of B symptoms • Progressive anemia and thrombocytopenia • Prior to planned systemic therapy	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Treatment Response	• For individuals with bulky nodal disease at diagnosis, CT with contrast of previously involved area(s) every 2 cycles of therapy • Routine imaging is not medically necessary for individuals without bulky nodal disease at diagnosis

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Indication	Medically Necessary Imaging Study
End of Therapy Evaluation	• For individuals with bulky nodal disease at diagnosis, CT with contrast of previously involved area(s)
Suspected Progression	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)
Suspected transformation (Richter's) from a low-grade lymphoma to a more aggressive type based on one or more of the following: • New B symptoms • Rapidly growing lymph nodes • Extranodal disease develops • Significant recent rise in LDH above normal range	• PET/CT (CPT® 78815 or CPT® 78816)
Surveillance	For individuals with bulky nodal disease at diagnosis, every 6 months for two years, then annually: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) Routine imaging is not medically necessary for individuals without bulky nodal disease at diagnosis

Evidence Discussion

Suspected/Diagnosis (whenever applicable)

- Diagnosis is by flow cytometry and biopsy. No imaging is supported.[1]

Initial staging

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- Routine advanced imaging for CLL/SLL is not supported. However, in the interest of individual safety, to recognize mass effect as assess risk of tumor lysis syndrome prior to treatment, CT imaging of the chest, abdomen and pelvis are supported. CT Neck may be added if neck symptoms, per general oncology guidelines.[1,12,13]

Restaging

- Routine imaging is not supported unless there is bulky nodal disease, as noted in 'initial staging' section. For bulky nodal disease, treatment response imaging with CT is supported every 2 cycles or for signs and symptoms of disease progression.
- There are no data-supported benefits to routine monitoring with PET/CT, and PET/CT is significantly more radiation than CT alone. PET/CT is supported only for signs and symptoms of Richter's transformation to high grade lymphoma, where the diagnosis can be made without invasive procedure using this modality.[1]

Surveillance

- There is no benefit to advanced imaging for surveillance of individuals without bulky nodal disease at diagnosis, and there is a risk of increased radiation exposure and invasive pursuit of incidental findings. In individuals with bulky nodal disease at diagnosis, flexibility is provided for surveillance imaging every 6 months for 2 years to assess for mass effect or progression.[131]

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Non-Hodgkin Lymphomas (ONC-27)

Guideline

Non-Hodgkin Lymphomas – General Considerations (ONC-27.1)
Diffuse Large B Cell Lymphoma (DLBCL) (ONC-27.2)
Follicular Lymphoma (ONC-27.3)
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Mantle Cell Lymphoma (ONC-27.5)
Burkitt's Lymphomas (ONC-27.6)
Lymphoblastic Lymphomas (ONC-27.7)
T Cell Lymphomas (ONC-27.8)
Post-Transplant Lymphoproliferative Disorders (ONC-27.9)
Waldenström Macroglobulinemia or Lymphoplasmacytic Lymphoma (ONC-27.10)
References (ONC-27)

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Non-Hodgkin Lymphomas – General Considerations (ONC-27.1)

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- Lymphoma is often suspected when individuals have any of the following:
  - Bulky lymphadenopathy (lymph node mass >10 cm in size), hepatomegaly or splenomegaly
  - The presence of systemic symptoms (fever, drenching night sweats or unintended weight loss of >10%, called “B symptoms”)
- Individuals with AIDS-related lymphoma should be imaged according to the primary lymphoma histology.
- See: **Castleman’s Disease (unicentric and multicentric) (ONC-31.11)** for guidelines covering Castleman’s disease.
- See: **Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Lymphoma (SLL) (ONC-26.4)** for guidelines covering Chronic Lymphocytic Leukemia (CLL)/ Small Lymphocytic Lymphoma (SLL).

Indication	Medically Necessary Imaging Study
• Biopsy proven lymphoma, or • Suspected lymphoma and any of the following: ◦ Bulky lymphadenopathy (LN mass >10 cm) ◦ Hepatomegaly ◦ Splenomegaly ◦ B symptom: Unexplained fever, drenching night sweats, unintended weight loss >10% total body weight	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast ◦ MRI without and with contrast for individuals who cannot tolerate CT contrast due to allergy or impaired renal function
Signs or symptoms of disease involving the neck	• CT Neck with contrast (CPT® 70491)
Signs or symptoms suggesting CNS involvement with lymphoma.	• MRI Brain without and with contrast (CPT® 70553) • See: **CNS Lymphoma (also known as Microglioma) (ONC-2.7)**

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Indication	Medically Necessary Imaging Study
Known or suspected bone involvement with lymphoma	• MRI without and with contrast of symptomatic or previously involved bony areas ◦ Bone scan is inferior to MRI for evaluation of known or suspected bone involvement with lymphoma
Determine a more favorable site for biopsy when a relatively inaccessible site is contemplated	• PET/CT (CPT® 78815 or CPT® 78816) ◦ PET/CT is not medically necessary for all other indications prior to histological confirmation of lymphoma
CAR-T cell therapy	Once before treatment and once 30-60 days after completion of treatment: • PET/CT (CPT® 78815 and CPT® 78816)

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Diffuse Large B Cell Lymphoma (DLBCL) (ONC-27.2)

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- Grey zone lymphomas, primary mediastinal B cell lymphomas, Grade 3 (high) follicular lymphoma, double-hit or triple-hit lymphomas, and primary cutaneous diffuse large B cell lymphoma should also be imaged according to these guidelines.

Indication	Medically Necessary Imaging Study
Initial Staging/ Diagnosis	ONE of the following: • PET/CT (CPT® 78815 or CPT® 78816) OR • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177)
Treatment response for all stage I, and stage II without extensive mesenteric disease	ANY of the following: • CT with contrast of previously involved area(s) may be approved every 2 cycles (6-8 weeks) of therapy • PET/CT (CPT® 78815 or CPT® 78816) after 3-4 cycles of chemotherapy (in lieu of CT or for inconclusive CT)
Treatment response for stage II WITH extensive mesenteric disease, and stages III-IV	ANY of the following: • CT with contrast of previously involved area(s) may be approved every 2 cycles (6-8 weeks) of therapy • PET/CT (CPT® 78815 or CPT® 78816) after 2-4 cycles of chemotherapy (in lieu of CT or for inconclusive CT)
End of Chemotherapy and/or Radiation Therapy Evaluation	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) at the end of chemotherapy and again at the end of radiation • CT with contrast of previously involved area(s)

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Indication	Medically Necessary Imaging Study
Suspected Recurrence	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) • PET/CT can be considered in rare circumstances (e.g., bone involvement).
Biopsy-proven recurrence	• PET/CT (CPT® 78815 or CPT® 78816)
CAR-T cell therapy	Once before treatment and once 30-60 days after completion of treatment: • PET/CT (CPT® 78815 or CPT® 78816)
Surveillance for ANY of the following: • All stages of DLBCL • Relapsed lymphoma • Primary mediastinal large B cell lymphoma • Primary cutaneous diffuse large B cell lymphoma	• Every 6 months for 2 years after completion of treatment: ◦ CT Chest with contrast (CPT® 71260) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ CT with contrast of previously involved area(s)

Evidence Discussion

Initial Staging

- These guidelines for initial staging align with NCCN and support either PET/CT or diagnostic CTs. Greater than 97% of DLBCL are FDG-avid, though it is not known to be more accurate than CT for DLBCL at initial staging. However, baseline PET may be useful for comparison, as end of treatment remission assessment with PET/CT is more accurate than CT alone in DLBCL.20

Restaging

- CT is supported by NCCN every 2 cycles for restaging. PET/CT is supported by NCCN for interim restaging after 3-4 cycles of therapy and at the end of

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chemotherapy and/or radiation for lower stage disease. For stage II with extensive mesenteric disease, stage III, and stage IV, NCCN supported PET/CT more frequently - after 2-4 cycles of therapy and at the end of therapy. PET/CT remains standard for remission assessment at end of therapy, where its accuracy is greater that CT alone for DLBCL .[1,14] Documentation of residual tissue at end of therapy is useful for monitoring for relapse, and as such diagnostic, contrast enhanced CT is supported if requested in addition to PET at end of therapy .[1,14]

- FDG avidity is prognostic for relapsed/refractory DLBCL and may have a role in patient selection for CAR-T therapy and to assess response, so is supported as a baseline before CAR-T and once 30-60 days after completion, to assess response and identify individuals who may be candidates for further salvage therapy .[1,14]

Surveillance

- The false-positive rate with PET scans for surveillance in various studies is 16%-20%, potentially leading to unnecessary investigations, radiation exposure, biopsies, expense, and anxiety.[3,13] Several small studies have failed to note an improvement in relapse detection with CT over clinical observation in DLBCL, however, there is no definitive standard for surveillance imaging with CT.[5,6] The majority of relapses occur in the first 2 years, and the NCCN supported CT imaging of involved areas and chest, abdomen, and pelvis every 6 months for the first two year. In the interest of individual- and provider-centricity, these guidelines align with the NCCN with respect to surveillance in DLBCL.[1]

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Follicular Lymphoma (ONC-27.3)

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- This section applies to follicular lymphomas with WHO grade of 1 (low) or 2 (intermediate) and primary cutaneous follicle center lymphoma. Grade 3 (high) follicular lymphomas should be imaged according to guidelines found in: **Diffuse Large B Cell Lymphoma (DLBCL) (ONC-27.2)**.

Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	EITHER of the following:: • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • PET/CT (CPT® 78815)
For ANY of the following, if PET/CT has not yet been performed: • If radiation therapy is being considered for stage I or II disease • CT results lead to a plan for systemic therapy	• PET/CT (CPT® 78815 or CPT® 78816)
Treatment Response	• CT with contrast of previously involved area(s) every 2 cycles of therapy
End of Therapy Evaluation	ONE of the following may be approved: • PET/CT (CPT® 78815 or CPT® 78816) OR • CT Chest with contrast (CPT® 71260) and • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)

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Indication	Medically Necessary Imaging Study
Suspected Recurrence	ANY or ALL of the following may be approved: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)
Suspected transformation (Richter's) from a low grade lymphoma to a more aggressive type based on one or more of the following: • New B symptoms • Rapidly growing lymph nodes • Extranodal disease develops • Significant recent rise in LDH above normal range	• PET/CT (CPT® 78815 or CPT® 78816)
Surveillance for ANY of the following: • After completion of active treatment • On maintenance treatment • Observation without any treatment	For all stages, every 6 months for two years, then annually: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)
Surveillance of pediatric-type follicular lymphoma in adults	Routine advanced imaging is not medically necessary after complete response

Evidence Discussion

Initial Staging

Diagnostic quality CT with contrast or PET/CT may be used for initial staging. Clinical stage is modified in only 15-20% of individuals with use of PET/CT and results in a change in treatment in only 8% of individuals.1 A baseline PET/CT is useful for comparison for treatment response and to determine if further treatment intensification is necessary and is recommended if systemic therapy is planned. PET/CT is particularly important in the setting of localized disease with a plan for radiation therapy only to rule out any other systemic disease and to serve as a baseline for treatment response.[1,3,4,20]

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Restaging

PET/CT is of unclear utility for interim restaging, as interim PET/CT response shows no association with overall survival, thus conventional CT is supported.[21] PET/CT does identify individuals at risk of progression at end of induction therapy, where initial studies showed that 69% of individuals who were classified as not having complete remission on CT were re-classified as complete metabolic remission when staged with PET/CT at end of induction.[20] This increased sensitivity and specificity more accurately identifies individuals at risk of poor progression-free survival who may be candidates for consolidative therapy and prevents over and under-treatment. These results only apply to end of induction PET/CT.[1,4,20]

While PET/CT alone is not sufficient to diagnose transformation of follicular lymphoma to diffuse large B cell lymphoma, when clinical signs and symptoms and lab values suggest transformation, PET/CT can be useful to detect transformation. SUV >10 predicts aggressive lymphoma with 80% certainty and PPV increases at higher SUVs.[1,18] FDG avidity is also standard of care to select biopsy site in suspected transformation.[1,18]

Surveillance

There are little data on the role of surveillance imaging in indolent lymphomas including follicular lymphoma. The majority of relapses occur within the first 2 years post completion of therapy, and these guidelines align with the NCCN support of CT no more than every 6 months in the first two years and no more than annually following. Given that indolent lymphoma is considered a chronic condition, there is no endpoint for this imaging if requested .[1] The exception is pediatric-type follicular lymphoma, for which there is no survival benefit with detection of recurrence via surveillance imaging vs clinical detection; surveillance imaging is not supported in this population.[1,13] PET/CT surveillance is generally not supported, due to a false positive rate as high as 20%, with no documented survival benefit, and increased radiation, invasive procedures, anxiety and cost.[13]

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Marginal Zone Lymphomas (ONC-27.4)

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- MALT lymphomas in any location and primary cutaneous marginal zone lymphoma should also be imaged according to these guidelines.
- Splenic Marginal Zone Lymphoma is diagnosed with splenomegaly, peripheral blood flow cytometry and bone marrow biopsy. Splenectomy is diagnostic and therapeutic. PET scan is not routinely medically necessary prior to splenectomy.

Indication	Medically Necessary Imaging Study
Initial Staging/ Diagnosis	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
EITHER of the following: • If radiation therapy is being considered for stage I or II disease • If systemic therapy is planned	• PET/CT (CPT® 78815 or CPT® 78816)
Treatment Response	• CT with contrast of previously involved area(s) every 2 cycles of therapy
End of Therapy Evaluation	ONE of the following: • CT with contrast of previously involved area(s) • PET/CT (CPT® 78815 or CPT® 78816)
Suspected Recurrence	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)

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Indication	Medically Necessary Imaging Study
Suspected recurrence and ANY of the following: • Symptoms of end organ dysfunction • Clinically significant or progressive cytopenias • Bulky disease (single mass of ≥7 cm or ≥3 or more nodal sites 3 cm in diameter) • Steady or rapid progression	In addition to the CT imaging above: • PET/CT (CPT® 78815 or CPT® 78816)
Surveillance of all stages of nodal marginal zone lymphoma for any of the following: • After completion of active treatment • On maintenance treatment • Observation without any treatment	Every 6 months for two years, then annually: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)
Surveillance of all stages of extranodal marginal zone lymphoma	Advanced imaging is not routinely medically necessary for surveillance of asymptomatic individuals

Evidence Discussion

Initial Staging

Diagnostic, contrasted CT of chest, abdomen and pelvis is supported for all individuals to assess extent of disease. PET avidity of extranodal marginal zone lymphoma is unreliable, as only 50%-75% of these tumors are FDG avid.[4] A baseline PET/CT is

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useful for comparison for treatment response and to determine if further treatment intensification is necessary, and so is recommended if systemic therapy is planned. PET/CT is particularly important in the setting of localized disease with a plan for RT only, to rule out any other systemic disease and to serve as a baseline for treatment response.[1,3,4]

Restaging

There is no clear role for PET in interim restaging of marginal zone lymphoma; CT is supported every 2 cycles in alignment with NCCN. End of therapy PET/CT is supported to identify individuals without a complete metabolic response who are candidates for extended therapy, to prevent over- or under- treatment.[1,4]

Surveillance

There are little data on the role of surveillance imaging in indolent lymphomas including marginal zone lymphoma. Extranodal marginal zone lymphoma typically remains localized, and asymptomatic surveillance with advanced imaging is not supported.[19] The majority of relapses occur within the first 2 years post completion of therapy. These guidelines align with NCCN support of CT no more than every 6 months in the first two years and no more than annually following. Given that indolent lymphoma is considered a chronic condition, there is no endpoint for this imaging if requested.[1] PET/CT surveillance is not universally supported, due to a false positive rate as high as 20%, with no documented survival benefit, and increased radiation, invasive procedures, anxiety, and cost.[13] However, PET/CT is a supported imaging modality for specific circumstances of recurrence; NCCN supported PET/CT in recurrence when there are indications for treatment.[1]

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Mantle Cell Lymphoma (ONC-27.5)

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Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	ONE of the following: • CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • PET/CT (CPT® 78815 or CPT® 78816)
Treatment Response	• CT with contrast of previously involved area(s) every 2 cycles of therapy • PET/CT is not medically necessary for monitoring treatment response but can be considered in rare circumstances when CT did not show disease (e.g., bone).
End of Therapy Evaluation	ONE of the following: • CT with contrast of previously involved area(s) • PET/CT (CPT® 78815 or CPT® 78816)
Suspected Recurrence	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) • PET/CT can be considered in rare circumstances (e.g., bone involvement).
Surveillance for all stages	Every 6 months for 2 years, and then annually: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)

Evidence Discussion

Initial Staging

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Diagnostic CT with contrast or PET/CT is supported. PET/CT is the preferred modality particularly when systemic therapy is planned, in order to prevent under treatment.[1,20]

Restaging

Interim restaging with PET/CT has not been shown to change outcomes, thus CT alone is supported every two cycles unless the sites of disease are only visible on PET/CT. However, PET/CT is supported at end of planned therapy as a lack of complete metabolic response may require maintenance treatment .[1]

Surveillance

Late relapses, as far as 15 years out, can occur with mantle cell lymphoma. The benefit of detection of with imaging vs clinical detection remains unclear, and some studies have shown no significant advantage in survival for relapses after first remission detected by surveillance imaging.[28] However, this is still an active point for discussion among treating providers, and the NCCN supported surveillance imaging with CT scan.[1,28] Given that most providers consider NCCN the standard of care, this guideline aligns with the more conservative NCCN recommended timeframe, to acknowledge this data while maintaining an individual- and provider-centric approach. Surveillance scanning with PET/CT has a positive predictive value of only 24% in this entity and is not supported.[1,28]

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Burkitt's Lymphomas (ONC-27.6)

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Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Treatment Response	• CT with contrast of previously involved area(s) every 2 cycles of therapy • PET/CT is not medically necessary for monitoring treatment response but can be considered in rare circumstances when CT did not show disease (e.g., bone).
End of Therapy Evaluation	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) at the end of chemotherapy and again at the end of radiation • CT with contrast of previously involved area(s)
Suspected Recurrence	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) • PET/CT can be considered in rare circumstances (e.g., bone involvement).
Surveillance	Every 6 months for 2 years after completion of treatment: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)

Evidence Discussion

Initial Staging/Diagnosis

In alignment with NCCN, both diagnostic quality CT scan with contrast and PET/CT are supported in Burkitt's Lymphoma. Diagnostic, contrasted CT is helpful for clarifying

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node bulk (individual nodes vs conglomerates), anatomic relationships of bulky disease, abdominal and Waldeyer's ring involvement, as well as abdominal visceral involvement, all of which are relevant to treatment decisions and emergency management in Burkitt's lymphoma. Baseline metabolic activity is useful for comparison at end of therapy, where FDG avidity defines treatment response.[1,3]

Treatment Response

CT alone (rather than PET/CT) should be used to assess response between cycles, as complete response is not defined until completion of upfront therapy regimen. At the end of all planned upfront therapy a PET/CT is supported, even in addition to diagnostic contrasted CTs. Metabolic response at this time point determines whether therapy can be considered complete or whether local therapy or intensification of treatment will be necessary.[1,3,4]

Surveillance

The role of surveillance in Burkitt's Lymphoma is somewhat controversial. Clinically evident symptoms of recurrence develop quickly in this aggressive entity, and recurrence is as likely to be diagnosed based on symptoms as by surveillance imaging in some studies (Lynch 2014). PET/CT surveillance of Burkitt's Lymphoma is widely discouraged as it increases false positive findings, radiation exposure, and does not improve outcomes .[4,13] NCCN is considered the standard for care in the U.S. and NCCN supported CT with contrast every 6 months for 2 years, with which these guidelines align for an individual- and provider-centric approach. Relapse after 2 years is rare, imaging after this point for asymptomatic surveillance has not been shown to improve outcomes.[1,13]

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Lymphoblastic Lymphomas (ONC-27.7)

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- Individuals with lymphoblastic lymphoma (even those with bulky nodal disease) are treated using the leukemia treatment plan appropriate to the cell type (B or T cell). Imaging indications in adult individuals are identical to those for pediatric individuals. See: **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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T Cell Lymphomas (ONC-27.8)

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- Includes Peripheral T-Cell Lymphomas, Mycosis Fungoides/Sézary Syndrome, Anaplastic Large Cell Lymphoma (ALCL) including breast implant-associated ALCL, Angioimmunoblastic lymphoma, and Primary Cutaneous CD30+T Cell Lymphoproliferative Disorders

Indication	Medically Necessary Imaging Study
Initial Staging/ Diagnosis	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
T-cell lymphoma of nasal cavity, oropharynx, or nasopharynx	In addition to the imaging above: • MRI Orbits, Face, and Neck without and with contrast (CPT® 70543)
Breast implant-associated ALCL	In addition to the above initial staging studies: • Ultrasound Breast (CPT® 76641 or CPT® 76642) ◦ MRI Breast (CPT® 77049) for evaluation of inconclusive ultrasound findings
Treatment Response	Any ONE of the following after 3-4 cycles: • PET/CT (CPT® 78815 or 78816) or • CT Chest with contrast (CPT® 71260), and • CT Abdomen and Pelvis with contrast (CPT® 74177) and • CT with contrast of previously involved area(s)

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Indication	Medically Necessary Imaging Study
End of Therapy Evaluation	Any ONE of the following at the end of chemotherapy and again at the end of radiation therapy: • PET/CT (CPT® 78815 or CPT® 78816) or • CT Chest with contrast (CPT® 71260), and • CT Abdomen and Pelvis with contrast (CPT® 74177), and • CT with contrast of previously involved area(s)
Suspected Recurrence	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) • PET/CT can be considered in rare circumstances (e.g., bone involvement).
Surveillance, all stages	Every 6 months for 2 years, then annually for 5 years: • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and CT of previously involved areas

Evidence Discussion

Initial Staging

FDG PET/CT fusion (PET with attenuation CT) is valuable for initial staging as individuals with T-cell lymphoma often have extranodal disease, which may be missed on body area specific diagnostic CTs. Noncontiguous nodes, Waldeyer ring, and GI/liver involvement is also common in T-cell lymphomas and may be difficult to distinguish on PET/CT fusion studies alone, thus our guidelines support the use of diagnostic quality, contrasted CTs when requested to ensure correct staging and treatment stratification and prevent under- or over-treatment, in addition to PET/CT fusion imaging.[1,2,29]

Treatment Response

Modality for restaging should be determined by which studies best illustrated disease at initial staging. PET/CT fusion imaging or body area specific diagnostic quality, contrasted CTs are generally adequate for comparison to initial staging to assess response[2] after 3-4 cycles and again at end of chemotherapy and at end of radiation. Imaging prior to 3-4 cycles may result in over- or under-treatment, and thus is not supported.[1,2]

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Surveillance

There is no evidence illustrating an overall survival advantage in detection of relapse from imaging vs clinical detection, but data suggests better progression-free survival after second line treatment in individuals undergoing imaging surveillance.[13] Considering these perspectives and to align with NCCN, CT of viscera and all previously involved areas is supported every 6 months for 2 years, then annually for 5 years.[2] PET/CT surveillance is not supported as no survival improvement is noted with PET/CT surveillance, and it subjects individuals to increased radiation, increased costs, and increased risk of invasive procedures for incidental findings, as the false positive rate in this setting is as high as 20%.[13,20] CT is generally supported for suspected recurrence as well, with PET reserved for biopsy-proven recurrence, by the same rationale.

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Post-Transplant Lymphoproliferative Disorders (ONC-27.9)

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- Post-transplant lymphoproliferative disorder (PTLD) or viral-associated lymphoproliferative disorder can rarely occur following solid organ or hematopoietic stem cell transplantation, or in primary immunodeficiency. When reduction of immunosuppression is unsuccessful, these are often treated with chemoimmunotherapy similar to high-grade NHL.
- This section applies to Monomorphic (B-cell type) PTLD and Polymorphic PTLD.
- For Hodgkin-lymphoma subtype of PTLD, see: **Hodgkin Lymphomas (ONC-28)** for imaging recommendations.

Indication	Medically Necessary Imaging Study
Initial Staging/ Diagnosis	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Treatment Response	ANY or ALL of the following after 4 weeks of reducing immunosuppression or every 2 cycles (6-8 weeks) of chemo/ immunotherapy: • CT Chest with contrast (CPT® 71260), and • CT Abdomen and Pelvis with contrast (CPT® 74177), and • CT with contrast of previously involved area(s)
End of Therapy Evaluation	ANY one of the following at the end of treatment: • PET/CT (CPT® 78815 or CPT® 78816) or • CT Chest with contrast (CPT® 71260), and • CT Abdomen and Pelvis with contrast (CPT® 74177), and • CT with contrast of previously involved area(s)
Suspected recurrence	• CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)

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Indication	Medically Necessary Imaging Study
Surveillance	No advanced imaging for surveillance

Evidence Discussion

Initial staging

FDG PET/CT fusion (PET with attenuation CT) is valuable for initial staging as individuals with PTLD often have extranodal and/or multi-site disease, which may be missed on body area specific diagnostic CTs. Individuals with single sites of disease may be managed with local treatment alone, thus thorough assessment for systemic disease is essential to prevent under-treatment. Node bulk has prognostic value and is used for treatment stratification, and bulky masses vs noncontiguous nodes may be difficult to distinguish on PET/CT fusion studies alone. These guidelines thus support the use of diagnostic quality, contrasted CTs when requested in addition to PET/CT fusion imaging.[1,29]

Restaging

Restaging with CT is supported every 2 cycles of chemotherapy as is standard for most disease processes .[1] Median time to failure of reduction of immunosuppression as first line therapy, however, is only 45 days, so for individual safety these guidelines support earlier restaging in this scenario as soon as 4 weeks after reduction of immunosuppression.[22] Changing therapy based on interim PET/CT alone is not supported and thus our guidelines support CT alone for interim restaging, with biopsy for concerning findings).[1,3,4] These guidelines do support PET/CT at end of planned treatment to ensure a complete metabolic response. [1,3,4] Concurrent diagnostic CTs may be done in lieu of PET/CT if requested, but diagnostic CTs in addition to PET/CT fusion studies do not offer additional information in the setting of a complete metabolic response .[3,4]

Surveillance

Advanced imaging surveillance is not supported for PTLD.[1,13] Surveillance imaging has not been shown to improve outcomes for PTLD and it subjects individuals to increased radiation, increased costs, and increased risk of invasive procedures for incidental findings.[13] Surveillance is predominantly via EBV PCR.[1]

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Waldenström Macroglobulinemia or Lymphoplasmacytic Lymphoma (ONC-27.10)

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Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • PET/CT (CPT® 78815 or CPT® 78816)
Treatment Response	• CT with contrast of previously involved area(s) every 2 cycles of therapy
End of Therapy Evaluation	• CT with contrast of previously involved area(s)
Suspected Recurrence	ANY or ALL of the following: • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)
Surveillance	No advanced imaging for surveillance

Evidence Discussion

Initial Staging

NCCN supported contrasted CT of chest, abdomen, and pelvis.[14,17] PET/CT fusion is recommended equally with diagnostic CT by the NCCN.[14]

Restaging

NCCN supported contrasted, diagnostic quality CTs every 2 cycles of chemotherapy and at end of treatment to determine response and prevent under-treatment. Diagnostic, contrasted CTs of chest, abdomen, pelvis and previously involved areas are supported for suspected recurrence.[14,17] PET/CT is not consistently correlated with monoclonal

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protein response, which is the primary means of monitoring this entity and PET/CT is not recommended for restaging of this entity.[14,15,16]

Surveillance

Surveillance of lymphoplasmacytic lymphoma is based on laboratory monitoring of blood counts and chemistries, serum proteins, and immunoglobulins. There is no established role for imaging surveillance in this entity.[14,16]

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Hodgkin Lymphoma (ONC-28)

Guideline

Hodgkin Lymphoma – General Considerations (ONC-28.1)
Classical Hodgkin Lymphoma (ONC-28.2)
Nodular Lymphocyte – Predominant Hodgkin Lymphoma (ONC-28.3)
References (ONC-28)

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Hodgkin Lymphoma – General Considerations (ONC-28.1)

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- Lymphoma is often suspected when individuals have any of the following:
  - Bulky lymphadenopathy (lymph node mass >10 cm in size), hepatomegaly or splenomegaly
  - The presence of systemic symptoms (fever, drenching night sweats or unintended weight loss of >10%, called “B symptoms”)
- Individuals with AIDS-related lymphoma should be imaged according to the primary lymphoma histology.
- The **Deauville Criteria** are internationally accepted criteria, which utilize a 5-point scoring system for the FDG avidity of a Hodgkin's lymphoma or Non-Hodgkin's lymphoma tumor mass as seen on FDG PET.
  - Score 1: No uptake above the background
  - Score 2: Uptake ≤mediastinum
  - Score 3: Uptake >mediastinum but ≤liver
  - Score 4: Uptake moderately increased compared to the liver at any site
  - Score 5: Uptake markedly increased compared to the liver at any site
  - Score X: New areas of uptake unlikely to be related to lymphoma

Indication	Medically Necessary Imaging Study
• Biopsy proven lymphoma, or • Suspected lymphoma and any one of the following: ◦ Bulky lymphadenopathy (LN mass >10 cm) ◦ Hepatomegaly ◦ Splenomegaly ◦ B symptom: Unexplained fever, drenching night sweats, unintended weight loss >10% total body weight	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast ◦ MRI without and with contrast for individuals who cannot tolerate CT contrast due to allergy or impaired renal function
Signs or symptoms of disease involving the neck	• CT Neck with contrast (CPT® 70491)

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Indication	Medically Necessary Imaging Study
Signs or symptoms suggesting CNS involvement with lymphoma	• MRI Brain without and with contrast (CPT® 70553) • See: **CNS Lymphoma (also known as Microglioma) (ONC-2.7)**
Known or suspected bone involvement with lymphoma	• MRI without and with contrast of symptomatic or previously involved bony areas ◦ Bone scan is inferior to MRI for evaluation of known or suspected bone involvement with lymphoma
Determine a more favorable site for biopsy when a relatively inaccessible site is contemplated	• PET/CT (CPT® 78815 or CPT® 78816) ◦ PET/CT not medically necessary for all other indications prior to histological confirmation of lymphoma
CAR-T cell therapy	Once before treatment and once 30-60 days after completion of treatment: • PET/CT (CPT® 78815 or CPT® 78816)

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Classical Hodgkin Lymphoma (ONC-28.2)

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- This section applies to nodular sclerosis, mixed cellularity, lymphocyte-depleted and lymphocyte-rich subtypes of Hodgkin lymphoma.
- Pregnancy-specific considerations: US Abdomen (CPT® 76700) or MRI Abdomen without contrast (CPT® 74181) and MRI Pelvis without contrast (CPT® 72195) are medically necessary in lieu of PET/CT or CT imaging.

Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Prior to initiation of therapy if one month or more has passed since previous PET/CT was performed	• PET/CT (CPT® 78815 or CPT® 78816)
Treatment Response	• PET/CT (CPT® 78815 or CPT® 78816) as frequently as every 2 cycles
End of Chemotherapy and/or Radiation Therapy Evaluation	• PET/CT (CPT® 78815 or CPT® 78816) at the end of chemotherapy and again at the end of radiation (at least 12 weeks after completion of radiation therapy)
Suspected or biopsy proven recurrence	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) • PET/CT (CPT® 78815 or CPT® 78816) if suspected recurrence ≥3 months after completion of therapy

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Indication	Medically Necessary Imaging Study
Surveillance	ANY or ALL of the following, every 6 months for 2 years after completion of therapy: • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) In addition to the above studies: • A single follow-up PET/CT at three months if end of therapy PET/CT shows Deauville 4 or 5 FDG avidity

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Nodular Lymphocyte – Predominant Hodgkin Lymphoma (ONC-28.3)

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Indication	Medically Necessary Imaging Study
Initial Staging/Diagnosis	ANY or ALL of the following: • PET/CT (CPT® 78815 or CPT® 78816) • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177)
Treatment Response	• CT with contrast of previously involved areas as frequently as every 2 cycles
End of Chemotherapy and/or Radiation Therapy Evaluation	• PET/CT (CPT® 78815 or CPT® 78816) at the end of chemotherapy and again at the end of radiation (at least 12 weeks after completion of radiation therapy)
Suspected Recurrence	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s)
Biopsy proven recurrence	• PET/CT (CPT® 78815 or CPT® 78816)

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Indication	Medically Necessary Imaging Study
Suspected transformation (Richter's) from a low-grade lymphoma to a more aggressive type based on one or more of the following: • New B symptoms • Rapidly growing lymph nodes • Extranodal disease develops • Significant recent rise in LDH above normal range	• PET/CT (CPT® 78815 or CPT® 78816)
Surveillance	ANY or ALL of the following every 6 months for 2 years after completion of therapy: • CT Neck with contrast (CPT® 70491) • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) • CT with contrast of previously involved area(s) In addition to the above studies: • A single follow-up PET/CT at three months if end of therapy PET/CT shows Deauville 4 or 5 FDG avidity

Evidence Discussion

Initial staging and Restaging

PET imaging is useful for staging, prognosis, and treatment stratification in all subtypes of Hodgkin Lymphoma. Staging with PET/CT rather than CT often confirms a higher stage of disease. While overall survival outcome is not clearly improved by staging/ restaging with PET/CT, stratification of treatment based on PET/CT benefits individuals by preventing over- and under-treatment. PET is thus supported for initial staging, re-staging every 2 cycles, and at the end of therapy. False-positive rates are elevated in the weeks following radiation, reaching up to 20%. Decisions based on scans done in close proximity to radiation may result in over-treatment. Therefore, PET/CT should not be performed until 12 weeks after completion of radiation.[1,2,11]

Diagnostic CT with contrast is supported concurrently with PET/CT for initial staging as it may better differentiate nodal conglomerates from individual nodes in close

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proximity, and node bulk is prognostic and also used for treatment stratification. However, performing diagnostic CTs concurrently with PET/CT at restaging does not provide a benefit, as FDG avidity is highly indicative of response in Hodgkin lymphoma, where CT alone may over- or under- estimate response. FDG avidity guides response assessment and informs subsequent treatment decisions. This includes intensifying therapy if PET avidity persists after 2-4 cycles, or omitting consolidative radiotherapy in cases of good response on PET/CT after 4 cycles for low-stage disease. A complete metabolic response (Deauville score of 3 or less) should be confirmed to determine the end of treatment. If the end-of-therapy PET/CT shows a Deauville score of 4-5, repeating the PET/CT 3 months later is appropriate to confirm the metabolic status of residual masses and to prevent under-treatment.[1,2,11]

While PET/MRI shows high concordance with PET/CT at a decreased radiation dose, it is inferior for assessing disease in the lungs, more time-consuming, and more costly. Furthermore, it has not been established as a standard for treatment stratification in adult Hodgkin Lymphoma and is therefore not recommended over PET/CT.[6]

Surveillance

Surveillance imaging with PET/CT is not supported, as the false-positive rate with PET scans in this context is greater than 20%, leading to unnecessary investigations, radiation exposure, biopsies, expense, and individual anxiety. In addition, no statistically significant difference in survival has been noted with CT surveillance imaging in Hodgkin Lymphoma, despite a statistically significant increase in radiation exposure and cost. However, given that many existing protocols still require surveillance imaging, the NCCN continued to support CT surveillance every 6 months in the first two years post therapy if requested. Given that NCCN is viewed as the standard of care in most U.S. oncology treatment centers, this guideline aligns with this current NCCN recommendation for an individual- and provider-centric approach.[1,2,4]

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Hematopoietic Stem Cell Transplantation (ONC-29)

Guideline

General Considerations for Stem Cell Transplant (ONC-29.1)
Reference (ONC-29)

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General Considerations for Stem Cell Transplant (ONC-29.1)

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Transplant Types:

Allogeneic ("allo"): The donor and recipient are different people, and there are multiple types depending on the source of the stem cells and degree of match between donor and recipient. This is most commonly used in diseases originating in the hematopoietic system, such as leukemias and lymphomas, and bone marrow failure syndromes or metabolic disorders. Common types are:

- Matched sibling donor (MSD or MRD): Donor and recipient are full siblings and HLA-matched
- Matched unrelated donor (MUD): Donor and recipient are HLA matched but not related to each other
- Cord blood: Donor stem cells come from frozen umbilical cord blood not related to the recipient, sometimes from multiple different donors at once
- Haploidentical transplant (haplo): Donor is a half-HLA match to the recipient, usually a parent

Autologous ("auto"): The donor and recipient are the same person. The process involves delivery of high dose chemotherapy that is ablative to the bone marrow, followed by an infusion of one's own harvested stem cells.

Allogeneic HSCT results in a much greater degree of immunosuppression than autologous HSCT because of the need to allow the new immune system to chimerize with the recipient's body. Immune reconstitution commonly takes more than a year for individuals who receive allogeneic HSCT, and individuals remain at high- risk for invasive infections until that has occurred.

Pre-Transplant Imaging in HSCT:

- Pre-transplant imaging in HSCT generally takes place within 30 days prior to transplant and involves a reassessment of the individual's disease status as well as infectious disease clearance.

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Indication	Medically Necessary Imaging Study
Immediate pre-transplant period	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) for ANY of the following: ◦ Any individual prior to undergoing allogeneic transplant ◦ New findings on chest x-ray ◦ New/worsening signs/symptoms of chest involvement • CT Sinus (CPT® 70486) for any clinical signs or symptoms • CT of other body areas is not indicated without clinical signs or symptoms of involvement
Assess cardiac function	• Echocardiogram (CPT® 93306, CPT® 93307 or CPT® 93308) ◦ MUGA scan (CPT® 78472) in specific circumstances, see: **Oncologic Indications for Cancer Therapeutics-Related Cardiac Dysfunction (CTRCD) (CD-12.1)** in the Cardiac Imaging Guidelines
Assess pulmonary function	• Pulmonary function tests
Assess primary disease status	• See disease-specific guidelines for end of therapy response assessment

Post-Transplant Imaging in HSCT:

- There are many common complications from HSCT, including infection, acute and chronic graft versus host disease (GVHD), hepatic sinusoidal obstruction syndrome, restrictive lung disease, among others.
- Disease response generally takes place at ~Day +30 (autos and some allos) or ~Day +100 (allos) post-transplant.

Indication	Medically Necessary Imaging Study
Assess known or suspected HSCT complications	• Site-specific imaging

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Indication	Medically Necessary Imaging Study
Suspected hepatic GVHD (elevated liver enzymes)	• Abdominal US (CPT® 76700 or CPT® 76705)
Suspected Bronchiolitis Obliterans Syndrome (BOS)	• CT Chest without contrast (CPT® 71250)
Assess primary disease status post-transplant	• See disease-specific guidelines for end of therapy evaluation and surveillance
Individuals receiving tandem auto transplants (2-4 autos back-to-back, spaced 6 to 8 weeks apart)	• Guideline recommended imaging repeated after each transplant

Evidence Discussion

Pre-Transplant imaging in Hematopoietic Stem Cell Transplant (HSCT)

This refers to imaging in the immediate pre-transplant period, approximately 30 days prior to anticipated HSCT. There is not a clear consensus for pre-transplant infectious screening with imaging, but a CT Chest and CT Sinus are supported for any clinical signs and symptoms of respiratory or sinus infection. NCCN does not support CT imaging for infection screening in asymptomatic individuals.[1] There are no clear data to support pre-transplant sinus imaging in adults; extrapolation from pediatric data shows no change in pre-transplantation management nor prediction of post-transplant sinusitis based on pre-transplant imaging of asymptomatic individuals.[2,3] Screening for infection of abdomen and pelvis with advanced imaging is not supported as it has not been shown to change management or outcomes yet increases cost and radiation exposure.[1,4] Echocardiogram is supported prior to transplant conditioning for all patients, to assure the safest possible dosing for cardiotoxic agents. MUGA scan is supported to supplement echocardiogram in individuals with a previous low ejection fraction (LVEF <50%).[5]

Post-Transplant imaging in HSCT

Timing of post-transplant disease restaging varies by disease process. Generally, repeat imaging follows the disease-specific guidelines for end of therapy evaluation and surveillance. For individuals receiving tandem auto transplants, disease-specific imaging can be repeated after each transplant. Imaging for post-transplant complications maximizes safety and allows for early intervention.

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Medical Conditions with Cancer in the Differential Diagnosis (ONC-30)

Guideline

Fever of Unknown Origin (FUO) (ONC-30.1)
Unexplained Weight Loss (ONC-30.2)
Paraneoplastic Syndromes (ONC-30.3)
References (ONC-30)

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Fever of Unknown Origin (FUO) (ONC-30.1)

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- FUO is defined as a persistent fever ≥101°F and ≥3 weeks with unidentified cause.
- While fever is a classic "B" symptom of advanced lymphoma, a cancer-related fever presenting in isolation without any other signs or symptoms of neoplastic disease is rare.

Indication	Medically Necessary Imaging Study
If physical examination, Chest x-ray, and laboratory studies are non-diagnostic	• Echocardiogram (CPT® 93306) • Abdominal ultrasound (CPT® 76700) • MRI Brain without and with contrast (CPT® 70553)
Above studies (including PE/ENT exam, pelvic exam, and DRE with laboratory studies) have failed to demonstrate site of infection	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • Radiopharmaceutical localization of tumor or distribution of radiopharmaceutical agent(s): CPT® 78800, CPT® 78801, or CPT® 78802, CPT® 78804, CPT® 78803 or CPT® 78831 (SPECT), or CPT® 78830, or CPT® 78832 (SPECT/CT)
"B" symptoms	• See: **Non-Hodgkin Lymphomas (ONC-27)**
Any CNS sign/symptom accompanied by fever	• MRI Brain without and with contrast (CPT® 70553)
All individuals	• PET is not indicated in the work-up of individuals with FUO

Evidence Discussion

The widely accepted definition of "Fever of Unknown Origin" is persistent fever of at least 101 degrees F for at least 3 weeks with unidentified cause. Most published recommendations are based on expert consensus rather than data. Malignancy accounts for 20%-30% of FUO in adults.[14,15] The remainder are infectious, inflammatory, and immune mediated. Physical exam, chest x-ray, and laboratory findings

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including workup for specific infections should guide workup, and all routine age-based cancer screening should be complete. If these are negative, further workup may be indicated. Echocardiogram, abdominal ultrasound, and MRI Brain without and with contrast are supported, with sensitivity rates of 80%-86%, as abdominal and pelvic abscess, endocarditis, and viral and bacterial CNS processes remain more common causes of fever than malignancy.[14,15,16,17] These modalities limit radiation exposure and are recommended as first line imaging in most algorithms.[14,15,16,17]

If the above workup has not demonstrated a source of infection, CT with contrast of the chest, abdomen and pelvis is supported as second line imaging, with sensitivity of up to 90% and specificity up to 70% for determining the cause of fever.[14,15] Technitium-based scans are insensitive but highly specific (93%-94%), with the advantage of lower radiation exposure than CT, and are supported to localize infectious or inflammatory foci.[14,18,21] MRI Brain without and with contrast is supported for any CNS symptoms accompanied by fever, as supported by several FUO algorithms and as outlined in HD-14.1 CNS and Head Infection. B symptoms with concern for lymphoma also warrant CTs, with further details outlined in EviCore ONC-27.1. The utility of PET/CT in workup of FUO is emerging, but specificity is variable, ranging from 52%-85%.[16,17,19-22] At this time it is not routinely supported in the workup of FUO.

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Unexplained Weight Loss (ONC-30.2)

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- Unintentional weight loss is defined as loss of ≥10 lbs. or ≥5% of body weight over 6 months or less, without an identifiable reason.
- Initial workup for all individuals may include appropriate detailed history, physical exam, baseline laboratory studies (e.g., CBC, CMP, HgbA1c, ESR/CRP, infectious workup, stool hemoccult, endocrine evaluation to rule out thyroid, pituitary, or gonadal dysfunction, etc.), chest x-ray, age-appropriate cancer screening, and neurological evaluation to rule out depression/dementia.
- Additional workup is directed to evaluate specific signs, symptoms, red flags, or abnormalities detected on initial workup. See condition-specific imaging guidelines for additional details.
- PET is not medically necessary in the work-up of individuals with unexplained weight loss.

Indication	Medically Necessary Imaging Study
CNS symptoms or abnormal pituitary hormones	• MRI Brain or Sella Turcica without and with contrast (CPT® 70553)
Abnormal thyroid function	• Thyroid ultrasound (CPT® 76536)
Abnormal liver function	• Abdominal ultrasound (CPT® 76700)
Abnormal kidney function	• Ultrasound Kidney and Bladder (CPT® 76770 or CPT® 76775)
Suspected cardiac dysfunction	• Echocardiogram (CPT® 93306)
Non-smokers	• Chest x-ray ◦ CT Chest with contrast (CPT® 71260) to evaluate abnormalities on chest x-ray
Current or former smokers	• CT Chest with contrast (CPT® 71260)
Dysphagia or early satiety	See: **Dysphagia and Esophageal Disorders (NECK-3)**
GI bleeding	See: **GI Bleeding (AB-22)**

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Indication	Medically Necessary Imaging Study
Abdominal pain without red flag signs	See: **Abdominal Pain (AB-2)**
Suspected pancreatic cancer in individuals aged ≥60 years with weight loss and at least one of the following[13]: • Diarrhea • Back pain • Abdominal pain • Nausea/vomiting • Constipation • New onset diabetes • Abnormal labs (CA 19-9, LFTs) • Non-diagnostic or negative abdominal ultrasound	Any ONE of the following: • CT Abdomen with contrast (CPT® 74160) • CT Abdomen and Pelvis with contrast (CPT® 74177) • MRI Abdomen without and with contrast (CPT® 74183) See also: **Epigastric Pain and Dyspepsia (AB-2.5)**
If all of the above do not identify cause of weight loss	Any of the following, if not previously performed: • CT Chest (CPT® 71260) • CT Abdomen and Pelvis (CPT® 74177) with contrast

Evidence Discussion

Gradual weight loss is a common occurrence in the elderly. Unintentional weight loss that is associated with in increased risk of morbidity and mortality is generally defined as a weight loss of 5% body weight over a period of 6-12 months.[23,26] Among individuals with unintentional weight loss, a minority are diagnosed with malignancy.[24,25] There is no unified published consensus or guideline to guide the workup for weight loss, but most publications recommend that the primary workup should be symptom-focused and include laboratory studies and age-appropriate cancer screening. Workup for particular symptoms or lab findings should be guided by condition-specific guidelines. Based on symptoms and lab findings, thyroid ultrasound, abdominal/renal ultrasounds, and/or echocardiogram are supported by these guidelines. All individuals with CNS symptoms or abnormal pituitary hormones warrant an MRI. A chest x-ray is reasonable in all individuals.[23,26,27]

A negative baseline evaluation is reassuring; with at least one prospective study showing that no individuals with a negative baseline clinical and laboratory evaluation were found to have malignancy on subsequent studies.[23,27] However, other prospective studies do illustrate that underlying malignancy may be detected on advanced imaging,

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with the highest predictive value for lung, pancreatic, lymphomas, prostate and colorectal cancers.[24,25] Based on this, these guidelines support contrasted CT Chest as part of initial workup for all smokers with clinically significant weight loss. These guidelines also support CT or MRI Abdomen (or CT Abdomen and Pelvis) as part of initial workup for individuals age 60+ with clinically significant weight loss and additional signs and symptoms significantly associated with pancreatic cancer.[13] For all other individuals, if the individuals have clinically significant unintentional weight loss as defined in paragraph 1, and the initial baseline evaluations above are negative, CT with contrast of the chest, abdomen, and pelvis are appropriate and supported by these guidelines.[23,24,26]

No published algorithm routinely supports PET/CT in the evaluation of unexplained weight loss, and there are no prospective studies illustrating the sensitivity or specificity of PET in this scenario. There may be individuals who meet evidence-based criteria for PET/CT based on their specific signs, symptoms and findings, particularly in the lymphomas (refer to guidelines ONC-27, ONC-28, and ONC-1.4).

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Paraneoplastic Syndromes (ONC-30.3)

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- Paraneoplastic syndromes are metabolic and neuromuscular disturbances. These syndromes are not directly related to a tumor or to metastatic disease. There may be a lead time between initial finding of a possible paraneoplastic syndrome and appearance of the cancer with imaging. Limited studies suggest annual imaging for 2 years after diagnosis of possible paraneoplastic syndrome may detect cancer, however benefit after 2 years is not well documented.
- The following are the most common symptoms of paraneoplastic syndromes known to arise from various malignancies:
  - Hypertrophic Pulmonary Osteoarthropathy: Often presents as a constellation of rheumatoid-like polyarthritis, periostitis of long bones, and clubbing of fingers and toes
  - Amyloidosis
  - Hypercalcemia
  - Hypophosphatemia
  - Cushing's Syndrome
  - Somatostatinoma syndrome (vomiting, abdominal pain, diarrhea, cholelithiasis)
  - Syndrome of inappropriate antidiuretic hormone secretion (SIADH)
  - Polymyositis/dermatomyositis
  - Opsoclonus
  - Paraneoplastic sensory neuropathy
  - Subacute cerebellar degeneration
  - Eaton-Lambert syndrome (a myasthenia-like syndrome)
  - Second event of unprovoked thrombosis
  - Disseminated Intravascular Coagulation
  - Migratory thrombophlebitis
  - Polycythemia
  - Chronic leukocytosis and/or thrombocytosis
  - Elevated tumor markers
  - Cryptogenic stroke (see also: **HD-21.3)**
- See: **Muscle Disorders (PN-6)** in the Peripheral Nerve Disorders Imaging Guidelines.
- See: **Multiple Myeloma and Plasmacytomas (ONC-25)** for evaluation of possible multiple myeloma.

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Indication	Medically Necessary Imaging Study
Initial evaluation	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast
ANY of the following: • Abnormality on conventional imaging difficult to biopsy • Inconclusive conventional imaging • Documented paraneoplastic antibody and conventional imaging fails to demonstrate primary site	• PET/CT (CPT® 78815 or CPT® 78816)
Subsequent evaluation for known paraneoplastic syndrome	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast every 6 months for 2 years after initial imaging for Lambert-Eaton Myasthenia syndrome • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast every 6 months for 4 years for all other paraneoplastic syndromes
Systemic mastocytosis	ANY ONE of the following: • CT Abdomen and Pelvis (CPT® 74177) with contrast • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • PET/CT scan is not medically necessary for evaluation of mastocytosis
First episode of unprovoked DVT/VTE	• Imaging to evaluate for malignancy is not medically necessary
Second unprovoked DVT/PE	• Imaging may be considered in the setting of a negative work-up for inherited thrombophilia and antiphospholipid syndrome
Thyroid US is recommended for elevated CEA, and upper/lower endoscopy is recommended for elevated CEA or CA 19-9.	

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Evidence Discussion

Cross-sectional imaging with contrasted CT of the chest, abdomen and pelvis is generally considered first-line to look for visceral malignancy in most paraneoplastic syndromes. While sensitivity varies widely (30%-82% across studies), specificity is reasonable (71%-100%).[33] PET/CT is supported in individuals with documented paraneoplastic antibodies, inconclusive conventional imaging, or to assess for alternate biopsy sites when an abnormality is found on conventional imaging that is inaccessible for biopsy. The sensitivity and specificity of PET/CT is approximately 80%, when used to evaluate individuals who had negative or unclear conventional imaging.[29] PET/CT is not supported as first line imaging as PET may miss smaller tumors, and has a false negative rate of approximately 20% in this setting .[33] While there is a lack of prospective data on monitoring paraneoplastic syndromes, it is known that these phenomena may precede detectable malignancy. In the interest of individual safety, these guidelines support repeat CT imaging every 6 months for 4 years; for Lambert-Eaton Syndrome, 2 years is sufficient as 96% of associated SCLC is detected in the first year, with later reports generally from an era of lesser quality CTs.[30,31,34]

Venous thromboembolism in the absence of a hypercoagulable risk factor may suggest occult malignancy. Blood testing, exam and non-advanced imaging have been shown to be helpful in most cancers that present with a first unprovoked DVT, but other advanced imaging is not cost-effective without other symptoms suggesting malignancy in this setting. In the setting of a second unprovoked DVT, cross-sectional imaging with contrasted CT may be considered and is supported by these guidelines.[31,35,36]

Systemic mastocyctosis may also develop extramedullary involvement and end-organ dysfunction, particularly involving liver and spleen. CT or MRI of abdomen and pelvis are supported in alignment with NCCN. There is no NCCN recommendation for PET/CT in systemic mastocytosis.

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Metastatic Cancer, Carcinoma of Unknown Primary Site, and Other Types of Cancer (ONC-31)

Guideline

General Guidelines (ONC-31.0)
Lung Metastases (ONC-31.1)
Liver Metastases (ONC-31.2)
Brain Metastases (ONC-31.3)
Adrenal Gland Metastases (ONC-31.4)
Bone (Non-Vertebral) Metastases (ONC-31.5)
Spinal/Vertebral Metastases (ONC-31.6)
Carcinoma of Unknown Primary Site (ONC-31.7)
Extrathoracic Small Cell and Large Cell Neuroendocrine Tumors (ONC-31.8)
Primary Peritoneal Mesothelioma (ONC-31.9)
Kaposi's Sarcoma (ONC-31.10)
Castleman's Disease (Unicentric and Multicentric) (ONC-31.11)
References (ONC-31)

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General Guidelines (ONC-31.0)

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- Guideline sections Lung Metastases (ONC-31.1) through Spinal Vertebral Metastases (ONC-31.6) are designed to supplement the disease-specific guidelines when the disease-specific guidelines do not provide guidance on the clinical scenario in question.
- These sections can also be used if the cancer type is rare and does not have its own diagnosis-specific imaging guidelines.
- These sections may also be used for lesions suspicious for cancer that do not have guidance in other disease-specific areas of the guidelines.

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Lung Metastases (ONC-31.1)

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Indication	Medically Necessary Imaging Study
New or worsening signs or symptoms suggestive of metastatic lung involvement or new or worsening chest x-ray abnormality	• CT Chest with contrast (CPT® 71260) • CT Chest without contrast (CPT® 71250) if there is a contraindication to CT contrast or only parenchymal lesions are being evaluated
Chest wall or brachial plexus involvement	• MRI Chest without and with contrast (CPT® 71552)
ONE of the following **and** no diagnosis-specific guideline regarding PET imaging: • Lung nodule(s) ≥8 mm • Confirm solitary metastasis amenable to resection on conventional imaging	• PET/CT (CPT® 78815) • When primary cancer known, PET request should be reviewed by primary cancer guideline
Previous or current malignancy and pulmonary nodule(s) that would reasonably metastasize to the lungs	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) at 3, 6, 12, and 24 months from the first study

Evidence Discussion

All individuals with a history of cancer who have new signs or symptoms suggestive of metastatic disease to the lung, or who have new or worsening findings on chest x-ray, warrant CT Chest. Contrast is preferable in most scenarios to include evaluation of soft tissue and nodes, but a non-contrast study may be approved if there is a contraindication to contrast or if only parenchymal lung lesions are being evaluated. CT with contrast (or without if for parenchymal lesion only) is supported to follow up new lung nodules in individuals with a history of malignancy.[55] There is, however, no clear consensus on a timeline for this follow up across all malignancy types. Where guidance is not provided in the disease-specific guidelines, these guidelines suggest a follow up timeline of CT at 3, 6, 12, and 24 months from discovery of nodule, extrapolating from Fleischner and Lung-RADS data that a nodule stable >24 months is exceedingly unlikely to be malignant.[54,55] MRI with and without contrast is supported for suspected malignant infiltration of the brachial plexus or chest wall infiltration, for better soft tissue delineation.[52,53]

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PET/CT is generally addressed in the guidelines for each specific cancer. If no specific guidance is provided, PET/CT is supported for lung nodules greater than or equal to 8mm.[54] In the interest of safety to prevent futile invasive procedures on individuals with occult metastatic disease, PET/CT is also supported by these guidelines to confirm solitary metastasis on conventional imaging that may be amenable to curative-intent resection. PET/CT surveillance is not generally supported due to high radiation exposure, financial toxicity, and excess radiation exposure.

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Liver Metastases (ONC-31.2)

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- Yitrium-90 Radioembolization (Y90-RE) is also known either as Selective Internal Radiation Therapy (SIRT) or trans-arterial radioembolization (TARE). Y90-RE is medically necessary for inoperable hepatocellular carcinoma and metastatic disease to the liver. Yitrium-90 resin or glass microsphere is injected into the hepatic vessel which supplies the tumor bed. This delivers high radiation dose to the tumor selectively.
- Yitrium-90 Radioembolization consists of three parts:

1. The pre-treatment planning angiogram with Technetium 99m macroaggregated albumin (Tc-MAA). The Tc-MAA acts as surrogate for biodistribution of application of Y-90. Planar or SPECT/CT are performed for calculation of lung shunt fraction and identification of extra hepatic uptake. The assessment of hepatopulmonary shunt is important in the determination eventual radiation dose. Presence of extra-hepatic uptake may preclude treatment or require coil embolization.
2. Yitrium-90 Radioembolization treatment typically done 7-10 days after mapping.
3. Post-treatment imaging may be done to confirm tumor localization.

- Ablation of liver metastases or primary HCC may be performed utilizing chemical, chemotherapeutic, radiofrequency, or radioactive isotope. Regardless of the modality of ablation, PET is not medically necessary for assessing response to this mode of therapy.

Indication	Medically Necessary Imaging Study
New or worsening signs or symptoms suggestive of metastatic liver involvement or new elevation in LFTs	• CT Abdomen with (CPT® 74160) or without and with (CPT® 74170) contrast
ANY of the following: • Considering limited resection • Inconclusive CT findings	• MRI Abdomen without and with contrast (CPT® 74183)

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Indication	Medically Necessary Imaging Study
ONE of the following **and** no diagnosis-specific guideline regarding PET imaging: • Confirm solitary metastasis amenable to resection on conventional imaging • LFT's and/or tumor markers continue to rise and CT and MRI are negative	• PET/CT (CPT® 78815) ◦ When primary cancer known, PET request should be reviewed by primary cancer guideline
Monitoring of liver metastases that have been surgically resected	• Review according to primary cancer guideline
Evaluation of hepatic artery chemotherapy infusion or TACE (transarterial chemoembolization)	• CTA Abdomen (CPT® 74175) immediately prior to embolization ONE of the following studies immediately prior to and one month post-embolization, if not previously done: • CT Abdomen without and with contrast (CPT® 74170) • MRI Abdomen without and with contrast (CPT® 74183)

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Indication	Medically Necessary Imaging Study
Evaluation for hepatic artery radioembolization with Y-90 radioactive spheres (TheraSphere or SIR Spheres) for liver metastases or primary liver tumors	To assess hepatic vascular anatomy before the procedure, any ONE of the following: • 3D Rendering (CPT® 76377) if conventional hepatic angiogram is being performed • CTA Abdomen (CPT® 74175) ONE of the following studies PRE-treatment based upon provider preference: • Radiopharmaceutical Localization Limited Area (CPT® 78800 or CPT® 78801) • SPECT or SPECT/CT (CPT® 78803, 78831, 78830, or 78832) • CPT® 78835 may be approved as an add-on code with SPECT/CT codes only (CPT® 78803, 78831, 78830, or 78832) for calculation of lung shunt fraction if planar imaging (CPT® 78800 or CPT® 78801) not performed. Liver-lung shunt calculation is included in planar scans and does not require additional Lung Perfusion Scan ONE of the following studies POST-treatment based upon provider preference: • Radiopharmaceutical Localization Limited Area (CPT® 78800 or CPT® 78801) • SPECT or SPECT/CT (CPT® 78803, 78831, 78830, or 78832)
Monitoring of ablated liver metastases or primary tumors	ONE of the following, immediately prior to ablation, 1 month post-ablation, then every 3 months for 2 years, and then every 6 months until year 5: • CT Abdomen without and with contrast (CPT® 74170) • MRI Abdomen without and with contrast (CPT® 74183)

Evidence Discussion

For individuals with known malignancy with new symptoms suggestive of metastatic liver involvement or increase in LFTs, these guidelines support CT of the abdomen

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with contrast or without and with contrast as first line imaging. This helps differentiate vascular enhancement patterns, number of lesions, and associated abdominal findings. If the CT remains indeterminate, MRI without and with contrast is supported as MRI enables better characterization of the internal features of the lesion.[56,57] For safety, MRI is also supported if limited resection is being considered.

PET/CT is less specific than conventional imaging for liver lesions, and is not first line to clarify indeterminate liver findings on CT.[56] However, if LFTs or tumor markers continue to rise and CT and MRI are negative, PET/CT may be used as a problem-solving tool to look for occult metastatic disease.[56] In the interest of safety, if a curative-intent resection of a liver lesion is planned, PET/CT may be used to confirm the liver metastasis is solitary to prevent subjecting the individual to a futile resection.

Imaging is medically necessary to evaluate for hepatic artery chemotherapy infusion or transarterial chemoembolization (TACE). Either CT or or MRI with and without contrast may be used for this purpose, per provider preference based on individual tumor characteristics, per the logic noted in paragraph 1. These guidelines for imaging for radioembolization align with international working group TheraSphere Global Dosimetry Steering Committee (DSC) recommendations.[59] Vascular mapping prior to radioembolization with CTA abdomen (with 3D rendering if requested), as well as a single nuclear medicine liver planar study or SPECT/SPECT-CT study, based on provider preference and individual tumor characteristics.[59] Liver-lung shunt calculations can generally be calculated from pre-treatment scans and an addition lung perfusion scan is not generally supported.[59] The nuclear imaging used pre-treatment is supported once post-treatment, and cross-sectional imaging with CT or MRI to evaluate response is supported 1 month post treatment.[59]

Monitoring of ablated liver tumors, metastatic or primary, is with cross sectional imaging with CT or MRI Abdomen, with and without contrast, per provider preference based on individual and tumor characteristics. In alignment with NCCN hepatocellular carcinoma surveillance recommendations, this guideline supports this imaging 1 month post ablation, every 3 months for 2 years, then every 6 months until year 5.[58] PET is not routinely supported for follow up for ablated liver lesions, regardless of ablation modality.[58,60] The sensitivity of PET is only 65% in this setting, compared with 96% for MRI, and the positive and negative predictive values are also significantly superior for MRI vs PET.[60]

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Brain Metastases (ONC-31.3)

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Indication	Medically Necessary Imaging Study
Individual with cancer and signs or symptoms of CNS disease or known brain metastasis with new signs or symptoms.	• MRI Brain without and with contrast (CPT® 70553)
To determine candidacy for SRS, and a diagnostic thin-slice MRI Brain has not been performed in the preceding 30 days	• MRI Brain without and with contrast (CPT® 70553)
Stereotactic radiosurgery planning	• Unlisted MRI for treatment planning purposes (CPT® 76498)
Monitoring of brain metastases treated with surgery or radiation therapy	Post-treatment, then every 3 months for 1 year and every 4 months thereafter: • MRI Brain without and with contrast (CPT® 70553) ***Individuals treated with stereotactic radiosurgery alone may have MRI Brain without and with contrast (CPT® 70553) immediately after stereotactic radiosurgery, then every 2 months for the first year, and then every 6 months thereafter
Brain metastases treated with radiation therapy, with recent MRI Brain indeterminate in distinguishing radiation necrosis vs. tumor progression	• MRI Perfusion imaging (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Brain metastases treated with radiation therapy, with recent MRI Brain and MR Perfusion studies both unable to distinguish radiation necrosis vs. tumor progression	• PET Metabolic Brain (CPT® 78608)
Any of the following: • Solitary brain metastasis suspected in individual with prior diagnosis of cancer and no diagnosis-specific guideline regarding PET imaging • Brain metastases and no known primary tumor	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • Mammography for individuals assigned female at birth • PET/CT (CPT® 78815 or CPT® 78816) for ANY of the following: ◦ Inconclusive conventional imaging ◦ Confirm either stable systemic disease or absence of other metastatic disease ◦ When primary cancer known, PET request should be reviewed by primary cancer guideline
Primary brain tumors	See: **Primary Central Nervous System Tumors (ONC-2)**
MR Spectroscopy (CPT® 76390) is not medically necessary for evaluation of metastatic brain cancer	

Evidence Discussion

Brain metastases are the most common malignant intracranial tumors with an incidence 10-fold higher than primary central brain tumors. Common presenting signs and symptoms of brain metastases include headache, nausea, vomiting, focal neurologic deficits, and mental status changes. The most common cancer associated with brain metastases is lung cancer approaching 50% of the cases. Melanoma is associated with the highest incidence of brain metastases.

These guidelines support MRI Brain without and with contrast as the standard imaging modality for evaluation of an individual with suspected or known brain metastases. The post-treatment monitoring after surgery or radiation is based on NCCN guidelines. Specifically for stereotactic radiosurgery planning, a diagnostic MRI is not supported and the MRI request is based on an unlisted procedure code. Advanced imaging with MR perfusion imaging is a problem-solving tool, complementing a standard MRI Brain,

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to distinguish between radiation necrosis and tumor progression. PET Metabolic Brain imaging is a useful tool to distinguish between radiation necrosis and tumor progression with recent indeterminate MRI Brain and MR Perfusion study.

In individuals who present with brain metastases and no known primary tumor, an evaluation to define a primary cancer is supported. Imaging studies that are supported include CT Chest and CT Abdomen and Pelvis with contrast. Mammography is supported for this staging in individuals assigned female at birth. PET/CT is indicated for inconclusive standard imaging, for evaluation of other metastatic disease or for staging if supported by primary cancer guideline. Biopsy or resection of a suspicious lesion is needed to establish a definitive diagnosis.

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Adrenal Gland Metastases (ONC-31.4)

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Indication	Medically Necessary Imaging Study
Differentiate benign adrenal adenoma from metastatic disease	• See: **Adrenal Cortical Lesions (AB-16.1)** in the Abdomen Imaging Guidelines
Known cancer and no known systemic metastases: • New adrenal mass • Enlarging adrenal mass • Inconclusive findings on recent CT	If not done previously, ANY of the following: • CT Abdomen without contrast (CPT® 74150) • CT Abdomen without and with contrast (CPT® 74170, adrenal protocol) • MRI Abdomen without contrast (CPT® 74181) • MRI Abdomen without and with contrast (CPT® 74183) • CT-directed needle biopsy (CPT® 77012)
One of the following and no diagnosis-specific guideline regarding PET imaging: • Biopsy is not feasible or is non-diagnostic • Isolated metastasis on conventional imaging and individual is a candidate for aggressive surgical management	• PET/CT (CPT® 78815) When primary cancer known, PET request should be reviewed by primary cancer guideline
Known extra-adrenal malignancy and undiagnosed adrenal mass being monitored off treatment	See: Phases of **Oncology Imaging and General Phase-Related Considerations (ONC-1.2)**

Evidence Discussion

In individuals with known extra-adrenal malignancy and no known systemic metastatic disease who have been found to have a new, enlarging or inconclusive adrenal mass on other imaging, a non-contrast CT adrenal protocol or CT Abdomen with and without

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contrast adrenal protocol or an MRI Abdomen without or without and with contrast are all supported by ACR appropriateness criteria.[62] Non-contrast images allow for initial attenuation measurements, but contrast-enhanced images with imaging for washout characteristics can help differentiate adenomas from metastatic disease.[7,62] Non-contrast chemical shift MRI can help detect intracytoplasmic fat, providing insight into benign vs malignant characteristics, but post-contrast imaging adds further specificity for adenoma.[7,62] CT-directed needle biopsy may also be appropriate and is supported by the guidelines if requested.[7,62]

The utility of PET/CT varies with histology and type of radiotracer. Generally, the use of PET/CT for a given malignancy is addressed in the disease-specific guidelines. For lesions with no known history of malignancy, there is no primary evidence supporting the use of FDG PET-CT for initial evaluation.[62] In one study of 1,049 incidental adrenal masses in individuals with no known history of cancer, zero were malignant.[27] Mild SUV uptake can also be seen in benign adenomas, bringing the sensitivity of PET/CT to only 85% .[28,29,62] False-positive interpretations potentially result in unnecessary invasive procedures. When other adrenal-specific cross sectional imaging is suspicious for malignancy by size and other criteria, biopsy is preferred. If a biopsy is not feasible or non-diagnostic, PET/CT may show increased SUV uptake in malignant lesions and guide further decision making, and is supported by the ACR in this context.[7,62]

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Bone (Non-Vertebral) Metastases (ONC-31.5)

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- This guideline may be used for individuals with or without a history of malignancy, depending on the indications as listed below.

Indication	Medically Necessary Imaging Study
Individuals ≥40 years of age with no history of malignancy who have a symptomatic bone lesion seen on x-ray and malignancy is suspected	EITHER of the following: • CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and Bone scan (CPT® 78306) OR • FDG PET/CT (CPT® 78815 or CPT® 78816)
Individuals less than 40 years of age with a bone lesion on x-ray	See: **Lesion of Bone (MS-10.2)**
Suspected bone metastasis due to ANY of the following in an individual with a current or prior malignancy: • Bone pain • Rising tumor markers • Elevated alkaline phosphatase	• Bone scan (CPT® 78306) supplemented by plain x-rays (see: **Nuclear Medicine (NM) Imaging in Oncology [ONC-1.3]** for additional bone scan codes)
ANY of the following: • Bone scan is not feasible or readily available • Bone scan is equivocal or indeterminate • Continued suspicion despite negative/inconclusive bone scan or other imaging modalities • Soft tissue component suspected on other imaging modalities • Differentiate neoplastic disease from Paget's disease of the bone	EITHER of the following: • MRI without and with contrast of the involved body site • CT without or with contrast of the involved body site

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Indication	Medically Necessary Imaging Study
Bone metastases suspected and **both** bone scan and either CT or MRI are inconclusive	• ^{18}F-FDG-PET/CT (CPT® 78815 or CPT® 78816)
EITHER of the following: • No prior cancer history with suspected bone metastatic disease or pathologic fracture on plain x-ray • Adult individuals with a biopsy-proven carcinoma of unknown origin in a bone lesion	• See: **Carcinoma of Unknown Primary Site (ONC-31.7)**

Evidence Discussion

Bone scan supplemented by plain x-ray is the generally the first-line modality for individuals with current or history of malignancy who have new bone pain, rising tumor markers or elevated alkaline phosphatase. Bone scan is 79%-86% sensitive and 81%-88% specific for metastatic lesions.[41,44] Bone scan allows rapid whole-skeletal evaluation, to ensure additional bony disease is not missed by focusing on a single site of cross-sectional imaging. MRI is a useful problem-solving tool if there is continued suspicion for bony metastatic disease with a negative bone scan.[41,43] MRI is supported when a soft tissue component is suspect to avoid understaging and undertreatment.[43] MRI is also the most specific study to supplement plain x-ray to differentiate Paget disease of bone from neoplastic disease.[45]

MRI is more accurate than CT or bone scan for the evaluation of malignant vertebral compression fractures and additionally can assess for cord compression, edema or leptomeningeal disease.[44] Individuals with known stage IV cancer with new back pain or any sings of neurologic compromise may be immediately evaluated by MRI of the whole spine without or without and with contrast.[44] MRI is also indicated for suspected leptomeningeal disease.[44] CT has the lower accuracy than MRI or bone scan in this setting and is only supported when MRI is contraindicated.[44] New leptomeningeal disease should prompt an MRI of the brain for complete neuroaxis imaging.

Where imaging is suspicious for bony metastatic disease and a workup for multiple myeloma is negative, CT Chest, Abdomen, and Pelvis with contrast are supported to look for a primary malignancy.[33]

The sensitivity and specificity of FDG PET-CT for bony metastatic disease varies with the malignancy. For example, for breast cancer, PET may be more sensitive (96% vs 76% for bone scan) but may be less specific (92% vs 95% for bone scan). However,

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for some cancers sensitivity of PET is as low as 56%.[42,44] Given this variability, PET/CT is supported as a problem-solving tool when both bone scan and MRI or CT are inconclusive. NCCN does support the use of FDG PET/CT specifically in individuals who 40 years of age or older and who have a symptomatic bone lesion with an abnormal x-ray.[63] These guidelines allow the flexibility of using FDG PET/CT in this specific scenario if it is desired in lieu of diagnostic CT and bone scan. NaF PET is considered investigational due to varying sensitivity and specificity.[46,47]

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Spinal/Vertebral Metastases (ONC-31.6)

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- Individuals with stage IV cancer with new onset back pain can forgo a bone scan (and plain films) in lieu of an MRI with and without contrast of the spine.

Indication	Medically Necessary Imaging Study
Known cancer history and spinal cord compression suspected based on signs/ symptoms of neurological compromise, including, but not limited to: • Unexpected, sudden loss of bowel or bladder control • Sudden loss of ability to ambulate • Complete loss of pinprick sensation corresponding to a specific vertebral level • Loss of pain at a site that had previously been refractory to pain management	MRI Cervical (CPT® 72156), MRI Thoracic (CPT® 72157), and MRI Lumbar Spine (CPT® 72158) without and with contrast OR without contrast • CT Cervical (CPT® 72126), CT Thoracic (CPT® 72129), and CT Lumbar (CPT® 72132) Spine if MRI is contraindicated
Individual with a known history of cancer and ANY of the following: • Metastatic or stage IV cancer with new or worsening back pain • Continued suspicion despite negative/ inconclusive bone scan or other imaging modalities • Back pain and suspicion of spinal malignancy based on any one of the following: ◦ Night pain ◦ Age >70 years ◦ Uncontrolled or unintentional weight loss ◦ Pain unrelieved by change in position ◦ Severe or worsening spinal pain	ONE of the following: • MRI of the relevant spinal level without contrast • MRI of the relevant spinal level without and with contrast • CT of the relevant spinal level without contrast • CT Myelogram of the relevant spinal level

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Indication	Medically Necessary Imaging Study
Monitoring untreated vertebral metastases	One of the following, every 3 months for 1 year: • MRI without and with contrast of the involved spinal level • CT without or with contrast of the involved spinal level **Imaging beyond 1 year is based on any new clinical signs/symptoms
Monitoring metastases within the spine treated with surgery and/or radiation therapy	One of the following, once within 3 months post-treatment, and then every 3 months for 1 year: • MRI without and with contrast of the involved spinal level • CT without or with contrast of the involved spinal level **Imaging beyond 1 year is based on any new clinical signs/symptoms

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Indication	Medically Necessary Imaging Study
Leptomeningeal involvement with cancer	Suspected: • MRI Brain without and with contrast (CPT® 70553) and MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar spine (CPT® 72158) without and with contrast • CT Cervical (CPT® 72127), Thoracic (CPT® 72130), and Lumbar Spine (CPT® 72133) without and with contrast can be approved if MRI is contraindicated or not readily available On active treatment: • MRI Brain without and with contrast (CPT® 70553) and MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar spine (CPT® 72158) without and with contrast every 2 cycles • CT without or with contrast of the involved spinal level if MRI is contraindicated Once treatment completed: • Routine advanced imaging not medically necessary for surveillance in asymptomatic individuals

Evidence Discussion

The incidence of malignant cord compression varies with cancer type but is rarely the first sign of systemic cancer. Back pain is the most common presenting symptoms and is reported in 80%-95% of individuals. Pain is often refractory to traditional pain medications. Sensory and motor deficits occur in 35%-75% of individuals, and acute bowel/bladder dysfunction are other red flags for cord compression. Up to 35% of individuals have multiple levels of compression, which may be non-contiguous, and as such where symptoms as above suggest cord compression in an individual with a history of malignancy, imaging of the whole spine is warranted. MRI with and without contrast has a sensitivity and specificity of 93% and 97%, respectively. ACR Appropriateness Criteria® stated CT Myelogram 'may be appropriate' in this setting, may be faster to obtain, and may be necessary to plan surgical intervention; thus, it is also supported by these guidelines for suspected malignant cord compression.[24,25,26]

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Some individuals will present with more localized symptoms suggestive of localized nerve root involvement but not consistent with the above symptoms of cord compression. Unilateral symptoms suggest a lower motor neuron lesion. Other symptoms suggestive of nerve root involvement are night pain, refractory pain, and pain unrelieved by a change in position. Elderly individuals with a cancer history are also at higher risk for nerve root involvement. Unintentional weight loss without other localizing symptoms may also suggest nerve root involvement. In individuals with a history of malignancy with any of the above, MRI without and with contrast of the involved spinal level of symptoms is supported. CT is less sensitive than MRI in this setting and is supported only when MRI is contraindicated.[24,25,26]

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Carcinoma of Unknown Primary Site (ONC-31.7)

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General Considerations

- Defined as carcinoma found in a lymph node or in an organ known not to be the primary for that cell type (e.g., adenocarcinoma arising in the brain or in a neck lymph node).
- This guideline also applies to a pathologic fracture that is clearly due to metastatic neoplastic disease in an individual without a previous cancer history.
- Detailed history and physical examination including pelvic and rectal exams and laboratory tests to be performed before advanced imaging.
- Individuals presenting with a thoracic squamous cell carcinoma described as metastatic appearing on chest imaging, or in lymph nodes above the clavicle, should undergo a detailed head and neck examination by a clinician skilled in laryngeal and pharyngeal examinations, especially in smokers.
- Individuals with suspected unknown primary based on only suspicious lytic bone lesions should be considered for serum protein electrophoresis (SPEP); urine protein electrophoresis (UPEP) and serum free light chains prior to consideration of extensive imaging.

Indication	Medically Necessary Imaging Study
Carcinoma found in a lymph node or in an organ known not to be primary	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Neck with contrast (CPT® 70491) if cervical or supraclavicular involvement • CT with contrast or MRI without and with contrast of any other symptomatic site • For individuals assigned female at birth: ◦ Diagnostic (not screening) mammogram and full pelvic exam ◦ MRI Breasts Bilateral (CPT® 77049) if pathology consistent with breast primary and mammogram is inconclusive

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Indication	Medically Necessary Imaging Study
Sebaceous carcinoma of the skin (can be associated with underlying primary malignancy)	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) • CT Neck with contrast (CPT® 70491) if cervical or supraclavicular involvement • CT with contrast or MRI without and with contrast of any other symptomatic site
Axillary adenocarcinoma	• Diagnostic (not screening) mammogram and full pelvic exam • MRI Breasts Bilateral (CPT® 77049) if pathology consistent with breast primary and mammogram is inconclusive • If the above are non-diagnostic for primary site: ◦ CT Neck (CPT® 70491), CT Chest (CPT® 71260), and CT Abdomen (CPT® 74160) with contrast ◦ CT with contrast or MRI without and with contrast of any other symptomatic site
EITHER of the following: • Carcinoma found within a bone lesion • Unexplained pathologic fracture	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) • Bone Scan (CPT® 78306) (see: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) • CT with contrast or MRI without and with contrast of any symptomatic site
ANY of the following: • Above studies have failed to demonstrate site of primary • CT scans reveal isolated metastatic disease for which definitive curative therapy is planned	• PET/CT (CPT® 78815 or CPT® 78816)
Post-treatment surveillance	• No advanced imaging for routine surveillance of asymptomatic individuals after treatment completion

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Evidence Discussion

Carcinoma of unknown primary site (CUP) is defined as carcinoma found in a lymph node or in an organ known not to be the primary for that cell type (e.g., adenocarcinoma arising in the brain or in a neck lymph node). This guideline also applies to a pathologic fracture that is clearly due to metastatic neoplastic disease in an individual without a previous cancer history. Individuals with suspected unknown primary based on only suspicious lytic bone lesions should be considered for serum protein electrophoresis (SPEP); urine protein electrophoresis (UPEP) and serum free light chains prior to consideration of extensive imaging.

CUP generally occurs in older adults, the majority 6-75 years, and accounts for 2-9% of all tumors. Median survival is poor, at 3-10 months. Twenty percent of individuals fall into a more favorable risk group with median survival >1 year, and imaging may help identify this group.[1,30] The primary step in workup of CUP, before advanced imaging, is a detailed history and physical examination including pelvic and rectal exams and laboratory tests, including basic CBC/Chemistries/LFTs but also tumor markers, immunohistochemistry, and PSA (for individuals assigned male at birth over 40). Endoscopy should also be considered if pathology suggests a GI primary. CT of the chest, abdomen, and pelvis with contrast is supported for all individuals where primary site is not suggested on physical and lab evaluation, in alignment with NCCN and the European Society of Medical Oncology (ESMO).[1,30] CT Neck may be included if cervical or supraclavicular involvement, as well as CT or MRI for other symptomatic sites or abnormal sites on physical, with the choice of modality driven by body site of interest. If the site of carcinoma is a bone lesion that is not consistent with multiple myeloma, a bone scan should be added to the workup. Morphology on bone scan can help determine the primary site, where lytic lesions are most suggestive of myeloma, renal cell, GI and melanoma, and blastic lesions most commonly occur with prostate cancer and GI carcinoid. Other morphologic features such as location and expansile nature can also help guide workup and treatment toward a particular primary site.[33] PET/CT has not been shown to be superior to bone scan for this purpose, and in fact may be less sensitive than bone scan for lesions <1cm.[1,33]

It is essential that individuals assigned female at birth have a diagnostic (not screening) mammogram and full pelvic exam. If pathology is consistent with breast cancer from axillary node or other metastatic site, but mammogram is inconclusive, a bilateral breast MRI with and without contrast is supported, as MRI may identify the breast as the primary site in approximately half of the individuals presenting with axillary adenocarcinoma metastases.[1,32] If a primary site is still not found, CT Neck, Chest, Abdomen, and Pelvis are supported.

These guidelines align with NCCN and support PET/CT as a problem-solving tool to look for a primary site of disease when the studies described above still do not reveal a primary site.[1] PET is of intermediate specificity in this setting and large randomized trials

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are lacking.[1] A meta-analysis on the use of PET/CT in individuals with CUP found that primary tumors were detected in 37% of 433 individuals across 11 studies, with pooled sensitivity and specificity of 84%.[1,31] In addition, if CT scans reveal oligometastatic disease and definitive curative therapy is planned, the absence of other sites of disease may be confirmed with PET/CT to prevent over- or under-treatment.[1,30]

Subsequent imaging and surveillance should follow the guideline for each primary site, once a likely primary has been established. NCCN stated follow-up should be with history and physical with subsequent diagnostic testing based on symptoms. In 20%-50% of individuals, the primary site remains unidentified even after postmortem examination, thus continued imaging is low-yield and may contribute to the significant distress associated with the uncertainty of this condition.[1,30] There is no data-driven algorithm for imaging surveillance when the primary site of disease remains undiscovered.[1]

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Extrathoracic Small Cell and Large Cell Neuroendocrine Tumors (ONC-31.8)

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- All poorly-differentiated or high-grade, small cell and large cell neuroendocrine tumors arising outside the lungs or of unknown primary origin are imaged according to these guidelines.
- For intrathoracic poorly differentiated neuroendocrine cancer, see: **Small Cell Lung Cancer (ONC-7)**

Indication	Medically Necessary Imaging Study
Initial staging	ONE of the following combinations: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • CT Chest with contrast (CPT® 71260) and MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)
Inconclusive findings on conventional imaging studies	• PET/CT (CPT® 78815)
ANY of the following: • Poorly differentiated neuroendocrine cancers of the head or neck • Signs or symptoms of CNS involvement	• MRI Brain without and with contrast (CPT® 70553)
Restaging during treatment	ONE of the following combinations, every 2 cycles: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) and CT of any known sites of disease with contrast • CT Chest with contrast (CPT® 71260) and MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) and CT with contrast of any known sites of disease

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Indication	Medically Necessary Imaging Study
Suspected Recurrence	ANY or ALL of the following are medically necessary: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast or CT Chest with contrast (CPT® 71260) and MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) • MRI Brain without and with contrast (CPT® 70553) • Bone scan (CPT® 78306) (See: **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)** for additional bone scan codes) • PET imaging is generally **not** medically necessary but can be considered for rare circumstances.
Surveillance	ONE of the following combinations every 3 months for 1 year, then every 6 months: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) • CT Chest with contrast (CPT® 71260) and MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197)

Evidence Discussion

Poorly differentiated/high-grade neuroendocrine tumors (NET) may occur anywhere in the body and exhibit more aggressive behavior than other neuroendocrine tumors. When these tumors occur in the lung, they are managed like small cell lung cancer and the small-cell lung cancer guidelines apply. This section refers to extrathoracic, poorly-differentiated or high-grade NETs.

Initial Staging

Initial staging with contrasted CT of the chest, abdomen and pelvis is supported, with sensitivity and specificity ranging from 82%-100% .[9,51] Metastatic disease, particularly to the liver, is common with this entity within this setting .[48,49] MRI is equally supported by NCCN for imaging of the abdomen and pelvis. Given the undifferentiated nature of these tumors, dotatate PET/CT is not routinely supported as they do not consistently have somatostatin receptors, with some studies showing this modality missing 50% of tumors.[49] Sensitivity and specificity of FDG PET/CT is superior to somatostatin-receptor based imaging for undifferentiated tumors, but is still widely variable and is not supported for first line imaging but may be used as a problem-solving tool when conventional imaging is inconclusive.[9,50] Poorly differentiated NETs do have a

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propensity for CNS involvement, and MRI brain with and without contrast is supported for initial staging with head and neck primary site or for any signs and symptoms suggestive of CNS involvement .[9,49] Suspected bony metastatic disease may be evaluated using guideline **ONC-31.5**.

Restaging and suspected recurrence

In alignment with NCCN, conventional imaging may be repeated every 2 cycles of treatment. In the case of suspected recurrence, CT Chest, Abdomen, and Pelvis with contrast are supported as well and MRI brain and Bone scan, in alignment with NCCN .[9] FDG PET/CT is not routinely supported in this setting for the reasons cited in the section on initial staging but may be utilized in rare circumstances in the interest of safety.

Surveillance

Guidelines support conventional imaging studies every 3 months for the first year, then every 6 months for 4 additional years, then annually indefinitely due to the long term-risk of recurrence in this entity.[9,48]

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Primary Peritoneal Mesothelioma (ONC-31.9)

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Indication	Medically Necessary Imaging Study
Initial staging	• CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • PET/CT (CPT® 78815) if there is no evidence of metastatic disease or conventional imaging is inconclusive
Recurrence/ Restaging	• If there is known prior disease, CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • PET for inconclusive finding on conventional imaging
Surveillance	• CT Abdomen and Pelvis with contrast (CPT® 74177) every 3-6 months for 5 years, then annually until year 10

Evidence Discussion

Contrasted CT of chest, abdomen and pelvis is essential to assess the degree of dissemination, verify that peritoneal disease is not metastatic from another primary site, evaluate lymphadenopathy, and to identify metastatic disease. For primary peritoneal mesothelioma, most individuals present with advanced locoregional disease.[35] Spread into pleural space and local lymph nodes are the primary sites of metastatic disease, with more distant/diffuse metastatic disease much less common.[35,36] Sensitivity of CT is superior to MRI for this entity. As with other malignancies, if CT shows a liver lesion indeterminate for metastatic disease, MRI may be used for further assessment per guideline section ONC-31.2. The sensitivity of PET-CT for malignant peritoneal mesothelioma ranges from 58%-100%, so it is not a primary imaging tool for staging. However, PET/CT may detect small peritoneal implants that are missed on CT and alter management.[34] These guidelines support PET/CT when no metastatic disease is detected on conventional imaging to ensure individuals are not under-staged.

Contrasted CT of the abdomen and pelvis are supported for restaging, as sensitivity of CT is superior to MRI for this disease process. Given that progression to chest disease is rare, CT Chest is supported for restaging only if there is known disease in the chest or if new chest symptoms develop. Given the widely variable sensitivity of PET/CT for peritoneal mesothelioma, it is supported only for inconclusive findings on conventional imaging.[35]

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NCCN guidelines and outcome data supported contrasted CT of the Chest/Abdomen/ Pelvis every 3 months for 2 years then annually until year 10.[34,35] Frequent imaging is supported only within the first two years as 68% of recurrences occur within the first 2 years, then annual imaging moving forward.[35]

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Kaposi's Sarcoma (ONC-31.10)

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Indication	Medically Necessary Imaging Study
Kaposi's Sarcoma	• Advanced imaging is not medically necessary since disease is generally localized to skin. • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast at initial diagnosis. If initial scans are negative then future imaging would be based on signs or symptoms.

Evidence Discussion

Most Kaposi Sarcoma (KS) is most often confined to skin; however, it can sometimes be found in viscera, particularly in HIV-associated disease. To prevent under-staging and to assess the need for systemic therapy, CT of the Chest, Abdomen, and Pelvis with contrast are supported at initial diagnosis.[61]

Routine advanced imaging is not supported if there is no visceral disease at diagnosis, but restaging CTs may be approved for individuals with visceral disease on systemic therapy per the timeframes offered in ONC-1.2. Contrasted CTs may also be approved to evaluate areas with specific signs and symptoms of new involvement.[61]

There are no data or expert consensus that support routine surveillance imaging for Kaposi's Sarcoma.[61]

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Castleman's Disease (Unicentric and Multicentric) (ONC-31.11)

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Indication	Medically Necessary Imaging Study
Initial staging	• Either CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast **or** PET/CT (CPT® 78815) • CT Neck with contrast (CPT® 70491) if cervical or supraclavicular involvement • If CT scans were utilized initially and suggested unicentric disease, and surgical resection is being considered, PET/CT (CPT® 78815) can be approved to confirm unicentric disease • If unicentric disease is surgically removed, proceed to surveillance section
Restaging: • Multicentric disease or surgically unresected unicentric disease on chemotherapy	ONE of the following every 2 cycles: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • PET/CT (CPT® 78815)
ANY of the following: • Suspected recurrence • Recurrent B symptoms • Rising LDH/IL-6/ VEGF levels	ONE of the following: • CT Chest (CPT® 71260) and CT Abdomen and Pelvis (CPT® 74177) with contrast • PET/CT (CPT® 78815)
Surveillance	• CT with contrast of involved areas no more than every 6 months up to 5 years

Evidence Discussion

Initial Staging

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PET/CT fusion is supported for initial staging of Castleman's Disease.[37] PET/CT not only assesses for multicentric disease, but the SUV can be used to determine Castleman's disease vs frank lymphoma.[39] Diagnostic, contrasted CTs of the chest, abdomen, pelvis, as well as neck (if suspected neck disease) may be substituted for PET/CT, but diagnostic CT is not generally supported concurrently with PET.[37] However, if diagnostic CTs were utilized initially and surgical resection is being considered, these guidelines allow a PET/CT to be done subsequently in the interest of safety, to confirm unicentric disease and prevent under-staging.

Restaging/Recurrence

Unicentric disease that is surgically resected is considered to be in surveillance and imaging follows surveillance guidelines. For multicentric disease (MCD) or unresected unicentric disease on chemotherapy, disease may be monitored every 2 cycles with contrasted CT of the chest, abdomen and pelvis or PET/CT fusion studies, in alignment with NCCN. The same imaging is supported for suspected recurrence or labs concerning for development of POEMS-associated MCD or HHV-8 MCD, as these entities are rapidly aggressive[38,39] Concurrent contrasted diagnostic CTs with PET/CT fusion studies do not generally change management and as such are not supported, nor are concurrent scans suggested by NCCN or international consensus recommendations.[37-40]

Surveillance

There are no clear consensus guidelines for imaging surveillance of Castleman's Disease. In the interest of safety, given a multitude of curative treatment options for recurrent disease, these guidelines support surveillance imaging with CT with contrast of involved body areas no more than every 6 months up to 5 years. PET/CT is not supported for surveillance in alignment with ASCO Choosing Wisely campaign.

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Abbreviations for Pelvis Imaging Guidelines

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Abbreviations for Pelvis Imaging Guidelines	
CA-125	cancer antigen 125 test
CT	computed tomography
FSH	follicle-stimulating hormone
GTN	gestational trophoblastic neoplasia
HCG	human chorionic gonadotropin
IC/BPS	interstitial cystitis/bladder pain syndrome
IUD	intrauterine device
KUB	kidneys, ureters, bladder (frontal supine abdomen radiograph)
LH	luteinizing hormone
MRA	magnetic resonance angiography
MRI	magnetic resonance imaging
MSv	millisievert
PA	posteroanterior projection
PID	pelvic inflammatory disease
TA	transabdominal
TSH	thyroid-stimulating hormone

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Abbreviations for Pelvis Imaging Guidelines	
TV	transvaginal
UCPPS	Urologic Chronic Pelvic Pain Syndrome
WBC	white blood cell count

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General Guidelines (PV-1.0)

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- A current clinical evaluation since the onset or change in symptoms is required before advanced imaging can be considered. The clinical evaluation should include a relevant history and physical examination including a pelvic and/or urological exam, appropriate laboratory studies, and non-advanced imaging modalities such as plain x-ray or Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or Transvaginal ultrasound (CPT® 76830) and/or Transperineal ultrasound (CPT® 76872).
  - Other meaningful contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms for follow up visit by an established individual can substitute for a face-to-face clinical evaluation.
- The use of gynecology CPT codes for pregnant females is not supported. Therefore, transvaginal ultrasound (CPT® 76830) and pelvic ultrasound (CPT® 76856 or CPT® 76857) are not medically necessary for those with a positive pregnancy test or known pregnancy. If a pregnancy test is positive, then obstetrical CPT codes are medically necessary.
- Abdominal imaging begins at the diaphragm and extends to the umbilicus or iliac crest.
- Pelvic imaging begins at the iliac crest and extends to the pubis.
- The uterus, tubes and ovaries arise out of the pelvis and are considered pelvic organs. If the uterus or adnexa rise out of the pelvic cavity, above the level of the iliac crest or umbilicus on physical exam or on previous imaging, abdominal imaging may be added for the completion of imaging.
- MRI (MRI Pelvis without contrast CPT® 72195) for Defecography is considered investigational/experimental by UHC.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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General Guidelines – Overview (PV-1.1)

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- When medically necessary, pregnant females should be evaluated with ultrasound or MRI without contrast to avoid radiation exposure. In carefully selected clinical circumstances, evaluation with CT may be medically necessary with careful attention to technique and radiation protection as deemed clinically appropriate.

Ultrasound

- Transvaginal ultrasound is the standard modality for imaging; no alternative modality has demonstrated sufficient superiority to justify routine use, and Transvaginal (TV) ultrasound (CPT® 76830) is the optimal study to evaluate adult female pelvic organ pathology.
- Pelvic ultrasound (complete CPT® 76856, or limited CPT® 76857) is medically necessary if it is a complementary study to the TV ultrasound. It may substitute for TV in pediatric individuals or non-sexually active females assigned at birth.
- Transperineal ultrasound (CPT® 76872) is medically necessary for cases of suspected urethral abnormalities, urinary incontinence, pelvic prolapse, or vaginal cysts.
- CPT® 76942 is used to report ultrasound imaging guidance for needle placement during biopsy, aspiration, and other percutaneous procedures.

Soft Tissue Ultrasound

- Pelvic wall, buttocks, and penis - CPT® 76857

Scrotal Ultrasound

- See
  - **Impotence/Erectile Dysfunction (PV-17.1)**
  - **Penis-Soft Tissue Mass (PV-18.1)**
- Ultrasound scrotum and contents - CPT 76870

3D Rendering with Ultrasound

- 3D Rendering (CPT® 76376 or CPT® 76377)
  - CPT® 76377 (3D rendering requiring image post-processing on an independent work station) or CPT®

    76376 (3D rendering not requiring image post-processing on an independent workstation) in the following clinical scenarios is medically necessary:

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  - Uterine intra-cavitary lesion when initial ultrasound is equivocal (See **Abnormal Uterine Bleeding (AUB) (PV-3.1)** and **Leiomyoma/Uterine Fibroids (PV-12.1)**
  - Hydrosalpinges or peritoneal cysts when initial ultrasound is equivocal (See **Complex Adnexal Masses (PV-5.3)**)
  - Lost IUD (inability to feel or see IUD string) with initial ultrasound (See **Intrauterine Device (PV-10.1)**)
  - Uterine anomaly is suspected on ultrasound (See **Uterine Anomalies (PV-14.1)**)
  - Infertility if ultrasound is indeterminate or there is clinical suspicion for intra-cavitary lesion (such as polyp or fibroid), hydrosalpinx, uterine synechia, adenomyosis or uterine anomalies (See **Initial Infertility Evaluation, Female (PV-9.1)**)
- 3D-4D (CPT® 76376 or CPT® 76377) rendering is medically necessary in certain situations of abnormal pregnancy implantation like suspected C-section scar pregnancies or suspected cornual (interstitial) ectopic pregnancy, or to locate an IUD.
- 3D-4D (CPT® 76376 or CPT® 76377) rendering is medically necessary for surgical planning with diagnosis of complex CHD in the fetus or for surgical planning of other complex fetal malformations.

Other Ultrasound

- CPT® 93975 Duplex scan (complete) of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study.
- CPT® 93976 Duplex scan (limited) of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study.
- CPT® 93975 and CPT® 93976 should not be reported together during the same session.

CT

- CT is not medically necessary for evaluating pelvic anatomy because it is limited due to soft tissue contrast resolution.

MRI

- Is medically necessary when used as a more targeted study or for individuals allergic to iodinated contrast.
  - MRI Pelvis without contrast (CPT® 72195)
  - MRI Pelvis without and with contrast (CPT® 72197)
  - MRI Pelvis with contrast only (CPT® 72196) is rarely performed

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Evidence Discussion (PV-1.1)

- Ultrasonography and magnetic resonance imaging (MRI) are the imaging techniques of choice for the pregnant individual, they should be used prudently and only when use is expected to answer a relevant clinical question[11].
- CT is not generally warranted for evaluating pelvic anatomy because it is limited due to soft tissue contrast resolution. Computed tomography (CT) scans are generally not recommended during pregnancy unless the benefits clearly outweigh the potential risks[12]. Computed tomography (CT) scan if necessary in addition to ultrasonography or MRI if more readily available for the diagnosis in question, should not be withheld from a pregnant individual. The risk of adverse effects from ionizing radiation should always be weighed against the risk of not performing the procedure and the benefit derived from the procedure[12].
- Ultrasound is the recommended modality for imaging the female pelvic organs; no alternative modality has demonstrated sufficient superiority to justify routine use[3], and transvaginal ultrasound is the optimal study to evaluate adult female pelvic pathology. Transabdominal pelvic ultrasound is a useful complementary study to transvaginal ultrasound and may substitute for transvaginal ultrasound in pediatric individuals or non-sexually active females. The American Institute of Ultrasound in Medicine (AIUM.org) launched an initiative in 2012 "Ultrasound First," which advocates the use of ultrasound examinations before other imaging modalities when the evidence shows that ultrasound imaging is at least equally, if not more, effective for the target anatomic area[13]. This applies particularly to obstetric and gynecologic patients for whom a skillfully performed and well-interpreted ultrasound image usually obviates the need to proceed to additional more costly and complex cross-sectional imaging techniques[3].
- Transperineal ultrasound can be useful for cases of suspected urethral abnormalities, urinary incontinence, pelvic prolapse, or vaginal cysts[14,15]. A study by Yang, et al confirmed transvaginal or transperineal ultrasound to be a non-invasive and cost-effective modality for diagnosis of urethral and periurethral masses[14]. Vaginal and urethral imaging is limited on transvaginal ultrasound due to the position of the endovaginal probe rendering the vagina out of the field, on computed tomography (CT) due to poor soft tissue discrimination of the vaginal walls and on magnetic resonance imaging (MRI). MRI of the vagina should be done with thin slice thickness and proper choice of the degree of angulation and used MR sequence, otherwise there is limited evaluation of the vagina[15]. Transperineal ultrasound is also a dynamic real-time examination, and can detect subtle abnormalities that are not seen in static imaging[15].
- Scrotal ultrasound is supported for evaluation of scrotal pain or suspected mass[16,17]. The American Urological Association recommends scrotal ultrasound for initial evaluation of unilateral or bilateral scrotal mass suspicious for neoplasm[17].
- Three-dimensional (3D) rendering with ultrasound can be considered when ultrasound shows suspected uterine anomaly, uterine intra-cavitary lesion,

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hydrosalpinges or peritoneal cysts[2,4,5,6]. A study by Laskshmy et al found 3D ultrasound to be a highly sensitive and specific tool for accurately diagnosing congenital uterine anomalies[2]. 3D rendering has shown a high degree of concordance with MRI and laparoscopy for congenital uterine anomalies, and is non-invasive, readily available and relatively cost-effective[4]. Three-dimensional ultrasound is a noninvasive method for evaluation of adnexal pathology[18].

- Doppler scan can be of benefit in addition to ultrasound for further evaluation of suspected uterine or ovarian abnormalities. Doppler flow mapping is useful in diagnosing submucosal fibroids and endometrial polyps[19]. ACOG (American College of Obstetrics and Gynecology)recommended that color Doppler ultrasonography is useful to evaluate the vascular characteristics of adnexal masses[1]. MRI pelvis is useful in cases such as inconclusive ultrasound for adenomyosis, "MRI is a second-line examination in the diagnosis of internal adenomyosis, mainly after a non-conclusive US evaluation. In addition, MRI can differentiate between the subtypes of adenomyosis." MRI pelvis is also useful for further evaluation of indeterminate adnexal masses. A study by Dirrichs, et al found MRI to improve sensitivity and specificity of diagnosis of indeterminate adnexal masses detected at TVUS, and use of MRI changed therapeutic management in 34% of cases[21]. MRI can aid in the diagnosis of deep pelvic endometriosis[22]. MRI pelvis is useful for further evaluation of unexplained pelvis pain when ultrasound evaluation is inconclusive[23]. Pelvic MRI is useful for evaluation of fibroids prior to uterine-sparing interventional techniques. "Although a high-quality ultrasonography (US) examination may be sufficient for evaluation in patients with straightforward cases of fibroids (for instance to estimate the size of a dominant fibroid), imaging evaluation is most reliably performed with magnetic resonance (MR) imaging to determine the characteristics, number, size, and location of fibroids and to assess for other pathologic conditions such as adenomyosis."[24]
- There is currently insufficient data to generate appropriateness criteria for the use of 3D and 4D rendering in conjunction with Obstetrical ultrasound imaging. Per ACOG, proof of a clinical advantage of 3-dimensional ultrasonography in prenatal diagnosis, in general, is still lacking.

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Abnormal Uterine Bleeding (PV-2)

Guideline

Abnormal Uterine Bleeding (AUB) (PV-2.1)
Retained Products of Conception (PV-2.2)
References (PV-2)

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Abnormal Uterine Bleeding (AUB) (PV-2.1)

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- Pregnancy test should be done initially if premenopausal
- If pregnancy test is negative or post menopausal initial evaluation includes ANY or ALL of the following:
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or Transvaginal ultrasound (CPT® 76830), D&C and/or endometrial biopsy
- Advanced imaging is not medically necessary for Endometrial Intraepithelial Neoplasia or Atypical Endometrial Hyperplasia (EIN-AEH) as imaging has not been found to improve outcomes for this condition.
- In females assigned at birth with postmenopausal bleeding
  - Those with thickened endometrium on ultrasound, those whose ultrasound failed to identify a thin, distinct endometrial strip and/or those with continued vaginal bleeding should all undergo endometrial biopsy to rule out endometrial carcinoma
- If biopsy confirms a malignancy, then see the appropriate oncology guideline.
- If ultrasound is equivocal for intracavitary lesion the following is medically necessary:
  - Duplex (Doppler) scan (CPT® 93975 complete; CPT® 93976 limited) as an add-on to TV ultrasound (CPT® 76830).
  - 3-D Rendering (CPT® 76377 or CPT® 76376) as an add-on.
- CT is not generally warranted for evaluating AUB since uterine anatomy is limited due to soft tissue contrast resolution.
  - An abnormal endometrium found incidentally on CT should be referred for TV ultrasound for further evaluation.
- MRI is not indicated for evaluation of abnormal uterine bleeding, please see specific Pelvis Imaging sections for MRI indications for ultrasound findings such as adnexal mass or uterine fibroids. See **Adnexal Mass/Ovarian Cysts (PV-5)** and **Leiomyomata (PV-12.1)**.

Evidence Discussion (PV-2.1)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for evaluation of abnormal uterine bleeding[2,12]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow and distinguish fluid and cysts from soft tissue[12]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure. 3-D Rendering has

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been shown to a useful adjunct for analysis of suspected lesions the endometrial cavity.

- MRI is not supported as an initial imaging modality for the diagnosis of abnormal uterine bleeding[12]. While MRI is accepted as an adjunct modality to ultrasound in cases where ultrasound may not fully characterize a soft tissue abnormality, imaging should be directed by the type of suspected soft tissue abnormality (i.e. adenomyosis, endometriosis, fibroids, and adnexal mass) and is addressed in additional sections of these guidelines. CT is of limited use in the evaluation of abnormal uterine bleeding given its suboptimal evaluation of the soft tissue of female pelvic organs[12].
- In premenopausal females assigned at birth presenting with abnormal uterine bleeding a pregnancy test should be performed. For those with a positive pregnancy test, imaging with appropriate obstetric ultrasound should be performed.
- Vaginal bleeding is the presenting symptom in 90% of postmenopausal females assigned at birth with endometrial cancer[10]. An endometrial strip of 4mm or less on ultrasound has been found to have a greater than 99% negative predictive value for endometrial cancer[10]. However, this cutoff may be inadequate in Black women, as it missed five-fold more cases than in White women[13]. Endometrial tissue sampling remains the gold standard for diagnosis of endometrial carcinoma. As such, those with thickened endometrium on ultrasound, those who ultrasound failed to identify a thin, distinct endometrial strip and those with continued vaginal bleeding should all undergo endometrial sampling to rule out endometrial carcinoma[10].
- The incidence of concurrent endometrial cancer with the diagnosis of Endometrial Intraepithelial Neoplasia or Atypical Endometrial Hyperplasia (EIN-AEH) is approximately 30% to 50%[1]. This makes evaluation for concurrent carcinoma imperative in the diagnosis of EIN-AEH for those considering a fertility-sparing treatment. The most accurate method for diagnosis is hysteroscopic-guided uterine sampling[1] which has the added benefit of direct visualization of any intrauterine pathology such as endometrial polyps.

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Retained Products of Conception (PV-2.2)

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- For abnormal uterine bleeding and/or pelvic pain with concern for retained products of conception (RPOC):
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or Transvaginal ultrasound (CPT® 76830) is medically necessary one time, repeat US is medically necessary for continued symptoms
  - Color Doppler ultrasonography (CPT® 93975 or CPT® 93976) may be added to ultrasound to aid in diagnosis of RPOC
  - CT Pelvis with and without contrast (CPT® 72194) OR MRI Pelvis with and without contrast (CPT® 72197) is medically necessary if US with Color Doppler is equivocal AND further imaging is needed for surgical planning

Evidence Discussion (PV-2.2)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for evaluation of suspected retained products of conception (RPOC)[15,16]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow within the endometrial complex, which improves the specificity and negative predictive value of detecting RPOC[15]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure.
- For most cases ultrasound is sufficient for detection of RPOC. For cases where ultrasound is inconclusive additional imaging with MRI or CT may provide additional information to aid in surgical planning[16].

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Amenorrhea (PV-3)

Guideline

Secondary Amenorrhea (PV-3.1)
Primary Amenorrhea (PV-3.2)
References (PV-3)

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Secondary Amenorrhea (PV-3.1)

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- Pregnancy test should be done initially
- If a pregnancy test is positive:
  - Refer to the member's individual coverage policy regarding obstetrical imaging indications and appropriate obstetrical imaging procedural codes. Billing of gynecology codes during pregnancy is not supported.
- If a pregnancy test is negative, further evaluation includes any of the following:
  - FSH, TSH, estradiol, and/or prolactin levels are medically necessary depending on clinical suspicion.
  - Serum free and total testosterone and/or DHEAS levels are indicated if there is evidence of hyperandrogenism
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) for suspected uterine or ovarian pathology are medically necessary
- The results of test(s) above determine the next steps, which include:
  - For suspected adrenal tumor, See **Adrenal Cortical Lesions (AB-16)** in the Abdomen Imaging Guidelines.
  - For suspected pituitary tumor or hypothalamic, See **Pituitary, Sella, Hypothalamus(HD-19)** in the Head Imaging Guidelines
  - For suspected Asherman's Syndrome the following are medically necessary:
    - Hysterosalpingogram (CPT® 74740), sonohysterosalpingography (CPT® 76831), and/or hysteroscopy if ultrasound is indeterminate for Asherman's syndrome
    - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) if hysterosalpingogram (CPT® 74740), sonohysterosalpingography (CPT® 76831), or hysteroscopy is indeterminate for Asherman's Syndrome.

Background and Supporting Information

- Asherman's syndrome: an acquired condition which refers to having scar tissue in the uterus

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Primary Amenorrhea (PV-3.2)

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- Prior to imaging a history, physical examination and Tanner stage should be evaluated.
- Initial evaluation may include pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) as medically necessary if ANY of the following:
  - Normal pubertal development and negative pregnancy test
  - Pelvic exam is indeterminate or unable to be performed
  - Delayed puberty with follicle-stimulating hormone (FSH) or luteinizing hormone (LH) that is elevated for the individual's age and Tanner stage
- If ultrasound defines a uterine or vaginal anomaly see **Uterine Anomalies (PV-14.1)**
- For suspected pituitary or hypothalamic tumor, See **Pituitary, Sella, Hypothalamus (HD-19)** in the Head Imaging Guidelines

Background and Supporting Information

- Evaluation of an individual without a uterus (determined by imaging or examination) may include karyotype and/or testosterone levels.
- TV ultrasound (CPT® 76830) is medically necessary in pediatric individuals who are sexually active or use a tampon and consent to the study.

Evidence Discussion (PV-3)

- The initial work up of amenorrhea should include a physical exam, pregnancy test and hormonal work up. For those with a positive pregnancy test, imaging with obstetric ultrasound should be performed. Hormonal testing can help to further direct imaging[7,2,5,6].
- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for evaluation of amenorrhea[7,2,5,6]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow and distinguish fluid and cysts from soft tissue[7]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure.
- MRI is supported as an adjunct to inconclusive ultrasound imaging, especially if the ultrasound is suggestive of a congenital uterine or vaginal anomaly[2]. CT is of limited use in the evaluation of amenorrhea given its suboptimal evaluation of the soft tissue of female pelvic organs.
- For suspected Asherman's syndrome, the gold standard for diagnosis remains hysteroscopy which has the added benefit of allowing for simultaneous treatment of adhesive disease[8,9]. However, hysteroscopy carries with it risks of anesthesia and

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uterine perforation. Hysterosaplingogram (HSG) allows for simultaneous evaluation of tubal patency[8]. Sonohsyterography (SHG) has a high negative predictive value (98%), but only a modest positive predictive value (43%)[8]. MRI may be a useful adjunct to HSG, SHG and hysteroscopy, especially in cases where there is complete obstruction of the endometrial cavity limiting the diagnostic ability of these tests[8,9].

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Adenomyosis (PV-4)

Guideline

Adenomyosis (PV-4.1)
References (PV-4)

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Adenomyosis (PV-4.1)

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- TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857) is the medically necessary diagnostic procedure of choice for the initial evaluation of suspected adenomyosis. Duplex Doppler (CPT® 93975 or CPT® 93976) can be added if requested.
- MRI Pelvis without contrast (CPT® 72195) or MRI Pelvis without and with contrast (CPT® 72197) is considered a medically necessary second-line imaging option after transvaginal ultrasound if:
  - Diagnosis is inconclusive for adenomyosis after an ultrasound and further delineation would affect management
  - MRI needed to guide the treatment of adenomyosis in an individual with an enlarged uterus, and coexisting leiomyoma/fibroid following indeterminate ultrasound
  - Uterine artery embolization is being considered
    - If MRI is equivocal, MRA Pelvis (CPT® 72198) or CTA Pelvis (CPT® 72191) if requested by or in consultation with the interventional radiologist planning the uterine artery embolization

Background and Supporting Information

Adenomyosis is when endometrial tissue, which normally lines the uterus, moves into the outer muscular walls of the uterus. Adenomyosis is a histologic diagnosis and is suspected by history and physical examination. Ultrasound findings of adenomyosis include heterogeneous myometrium, myometrial cysts, asymmetric myometrial thickness, and subendometrial echogenic linear striations.

Evidence Discussion (PV-4.1)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for evaluation of adenomyosis[10,1,2,3]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow and distinguish fluid and cysts from soft tissue[10]. In the presence of features mimicking leiomyomas, Doppler US displaying vessels perpendicular to the endometrial interface, is suggestive of adenomyosis[2]. Transvaginal ultrasound has a sensitivity of 83.8% and specificity of 63.9% for adenomyosis[7,8]. The overall diagnostic accuracy of the use of transvaginal ultrasound with color Doppler for adenomyosis is 93.8%[8]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure.

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- MRI of the pelvis is a second-line examination in the diagnosis of adenomyosis, mainly after an inconclusive US evaluation[2]. MRI pelvis is useful in individuals with coexisting leiomyoma[2]. A meta-analysis comparing the diagnostic performance of MRI and transvaginal ultrasound reported that MRI had a pooled sensitivity of 77% and a specificity of 89%[2]. The authors concluded that MRI performs more favorably than transvaginal ultrasound in the presence of associated uterine leiomyomas[2].

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Adnexal Mass/ Ovarian Cysts (PV-5)

Guideline

Suspected Adnexal Mass – Initial Evaluation (PV-5.1)
Simple Cysts (PV-5.2)
Complex Adnexal Masses (PV-5.3)
Screening for Ovarian Cancer/Suspected Ovary Cancer (PV-5.4)
References (PV-5)

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Suspected Adnexal Mass – Initial Evaluation (PV-5.1)

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- A potential mass is found on exam and/or found incidentally on other imaging
- Transvaginal (TV) ultrasound imaging (CPTCPT® 76830) is the medically necessary initial study of choice.
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) can be performed if requested as a complimentary study to the TV ultrasound.
  - Once confirmed, Color Doppler ultrasonography (CPT® 93975 or CPT® 93976) may be medically necessary to evaluate the vascular characteristics of adnexal masses.
- MRI Pelvis without contrast (CPT® 72195), OR without and with contrast (CPT® 72197; CPT® 72195 if pregnant) is medically necessary if ultrasound does not identify the origin of the pelvic mass (adnexal, uterine, or other in etiology).
  - If the mass is unrelated to female pelvic organ anatomy, see **Abdominal Mass (AB-13)** in the Abdomen Imaging Guidelines.
- For O-RADS™ 0 (incomplete evaluation) either repeat ultrasound OR MRI Pelvis without contrast (CPT® 72195), OR without and with contrast (CPT® 72197)is medically necessary
  - If pregnant, MRI pelvis without contrast CPT® 72195 is medically necessary to avoid gadolinium exposure to the fetus.

Background and Supporting Information

- Consultation with or referral to a gynecologic oncologist is recommended for females assigned at birth with an adnexal mass who meet one or more of the following criteria:[7]
  - Postmenopausal with elevated CA-125 level, ultrasound findings suggestive of malignancy, ascites, a nodular or fixed pelvic mass, or evidence of abdominal or distant metastasis.[7]
  - Premenopausal with very elevated CA-125 level, ultrasound findings suggestive of malignancy, ascites, a nodular or fixed pelvic mass, or evidence of abdominal or distant metastasis.[7]
  - Premenopausal or postmenopausal with an elevated score on a formal risk assessment test such as the multivariate index assay, risk of malignancy index, or the Risk of Ovarian Malignancy Algorithm or one of the ultrasound-based scoring systems from the International Ovarian Tumor Analysis group.[7]

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- O-RADS™ 0 should not be used for masses that are completely visualized but the diagnosis is unclear.
- Simple and Complex Adnexal Cysts
  - Simple cysts are smooth walled and clear without debris.
  - Complex cysts can have solid areas or excrescences, and/or debris in them, greater than 3mm irregular septations, mural nodules with Doppler-detected blood flow, and/or free abdominal/pelvic fluid.
- Suspected Adnexal Mass – Tumor Markers
  - The adnexa include the ovaries, Fallopian tubes, and ligaments that hold the uterus in place.
  - CA-125 is a tumor marker that is useful for the evaluation of adnexal mass:
    - Elevation occurs with both malignant (epithelial cancer) and benign entities (leiomyoma, endometriosis, PID, inflammatory disease such as lupus, and inflammatory bowel disease).
    - Increase in the markers over time occurs with malignancy only
    - Consider tumor markers in individuals with an abnormal ultrasound that is not a simple cyst
  - Other markers include Beta hCG, LDH, and AFP (germ cell tumors) and Inhibin A and B (granulosa cell tumor).

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Simple Cysts (PV-5.2)

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- Simple cysts are smooth, thin walled, anechoic and clear without debris. Simple cysts up to 10 cm in diameter as measured by ultrasound are almost universally benign.
  - Repeat TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary
    - Follow up according to the below schedule if ≤10 cm
  - Routine use of 3D rendering (CPT® 76376/CPT® 76377) for evaluation of simple ovarian cysts is not medically necessary.

Simple Cyst Follow-Up

Size	Pre-Menopausal	Post-Menopausal
≤3 cm	• None	• None
>3 cm to 5 cm	• None	• Follow-up in 12 months with TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary: ◦ If smaller (≥10-15% decrease) no further surveillance. ◦ If stable follow-up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) at 24 months from initial exam ◦ If enlarging (≥10%-15% increase) follow-up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) at 12 and 24 months from initial exam • If there is a change in morphology on follow imaging see **Complex Adnexal Masses (PV 5.3)**

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Size	Pre-Menopausal	Post-Menopausal
>5 cm to ≤10 cm	• Follow up in 8-12 weeks (proliferative phase if possible) with TV ultrasound (CPT® 76830) and/ or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary; further follow-up intervals may be adjusted on basis of degree of cyst change	• Follow-up in 3-6 months with TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary; further follow-up intervals may be adjusted on basis of degree of cyst change. • Subsequent follow up with TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary annually and if stable for 2 years or decreasing in size, no further imaging follow-up is needed.

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Size	Pre-Menopausal	Post-Menopausal
>10 cm	• If not excised, US follow up within 6 months with TV Ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary • If stable, follow up Ultrasound can be done at 12 and 24 months from initial exam • If solid component, MRI Pelvis without and with contrast (CPT® 72197) is medically necessary • If ultrasound equivocal for Simple cyst, MRI Pelvis without and with contrast (CPT® 72197) is medically necessary • If follow up ultrasound imaging shows changing morphology and/or a vascular component then MRI Pelvis without and with contrast (CPT® 72197)	• If not excised, US follow up within 6 months with TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary • If stable, follow up Ultrasound can be done at 12 and 24 months from initial exam • If solid component, MRI Pelvis without and with contrast (CPT® 72197) is medically necessary • If ultrasound equivocal for Simple cyst, MRI Pelvis without and with contrast (CPT® 72197) is medically necessary • If follow up ultrasound imaging shows changing morphology and/or a vascular component then MRI Pelvis without and with contrast (CPT® 72197) is medically necessary

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Complex Adnexal Masses (PV-5.3)

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- Ultrasound imaging should provide characteristics of the cyst/mass prior to consideration of advanced imaging.
- Complex cysts found on ultrasound have characteristics that include: solid areas or excrescences, and/or debris, may have greater than 3mm irregular septations, and/or mural nodules with Doppler-detected blood flow, and/or free abdominal/pelvic fluid. Complex cysts have an O-RADS™ score of 2 or higher.
- Routine use of 3D rendering (CPT® 76376/CPT® 76377) for evaluation of complex ovarian cysts is not medically necessary unless otherwise mentioned in the table below.

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Follow up Complex Adnexal Masses

Condition	Pre-Menopausal	Post-Menopausal
Typical hemorrhagic cyst < 10 cm (O-RADS™ 2)	• If initial ultrasound imaging confirms hemorrhagic cyst ≤5 cm no further imaging is medically necessary • If initial ultrasound imaging confirms hemorrhagic cyst >5 cm but <10 cm, follow up with Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) in 8-12 weeks is medically necessary. Duplex (Doppler) scan (CPT® 93975 complete; CPT® 93976 limited) may be approved as an add-on to TV ultrasound (CPT® 76830). ◦ If follow-up imaging confirms a hemorrhagic cyst that has not completely resolved or has enlarged, MRI Pelvis without and with contrast (CPT® 72197) is medically necessary. ◦ If stable, follow up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) is medically necessary at 24 months from initial exam	• Early postmenopausal (<5 years) either of the following is medically necessary: ◦ follow-up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) in 2-3 months OR ◦ MRI Pelvis without and with contrast (CPT® 72197) • Late postmenopausal (≥ 5 years) hemorrhagic cyst should not occur ◦ MRI Pelvis without and with contrast (CPT® 72197) is medically necessary

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Condition	Pre-Menopausal	Post-Menopausal
Hemorrhagic cyst ≥10cm (O-RADS™ 3)	• If ultrasound equivocal for Hemorrhagic cyst, MRI Pelvis without and with contrast (CPT 72197) • If initial ultrasound imaging confirms a Typical Hemorrhagic cyst ≥10cm ◦ If not excised consider TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) follow up within 6 months ▪ If stable, follow up Ultrasound can be done at 12 and 24 months from initial exam ◦ If solid component, MRI Pelvis without and with contrast (CPT® 72197) may be approved ◦ If follow up ultrasound imaging shows changing morphology and/or a vascular component then consider MRI Pelvis without and with contrast (CPT® 72197)	• MRI Pelvis without and with contrast (CPT® 72197) is supported

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Condition	Pre-Menopausal	Post-Menopausal
Typical Endometriomas < 10cm (O-RADS™ 2)	• If ultrasound equivocal for Endometriomas, MRI Pelvis without and with contrast (CPT 72197) • If initial imaging confirms a Typical Endometrioma, follow-up Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830); duplex (Doppler) scan (CPT® 93975 complete; CPT® 93976 limited) may be approved as an add-on to TV ultrasound (CPT® 76830) ◦ If <10cm and not surgically excised follow-up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) in 12 months ▪ If stable follow up Ultrasound can be done at 24 months from initial exam ◦ If follow up ultrasound imaging shows changing morphology and/or a vascular component then consider MRI Pelvis without and with contrast (CPT® 72197)	• If initial ultrasound imaging confirms a typical endometrioma < 10cm then either the following is medically necessary: ◦ Follow-up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) in 2-3 months OR ◦ MRI Pelvis without and with contrast (CPT® 72197)
Typical Endometriomas ≥10cm (O-RADS™ 3)	• If initial ultrasound imaging confirms a Typical Endometrioma ≥10cm the following is medically necessary: ◦ If not excised, TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) follow up within 6 months ▪ If stable follow up Ultrasound can be done at 12 and 24 months from initial exam ◦ If solid component, MRI Pelvis without and with contrast (CPT® 72197) may be approved ◦ CPT® 72197) ◦ If follow up ultrasound imaging shows changing morphology and/or a vascular component then consider MRI Pelvis without and with contrast (CPT® 72197)	• MRI Pelvis without and with contrast (CPT® 72197) is medically necessary

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Condition	Pre-Menopausal	Post-Menopausal
Typical Dermoid < 10cm (O-RADS™ 2)	• If initial features are only suggestive for, or if assessment is uncertain follow-up Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) within 3 months is medically necessary • OR • If ultrasound equivocal for Dermoid, MRI Pelvis without and with contrast (CPT 72197) is medically necessary • If initial ultrasound imaging confirms a Dermoid, follow-up Pelvic ultrasound (CPT® 76856 or CPT® 76857); and/or TV ultrasound (CPT® 76830); duplex (Doppler) scan (CPT® 93975 complete; CPT® 93976 limited) may be medically necessary as an add-on to TV ultrasound (CPT® 76830). ◦ If ≤10 cm, may consider follow-up TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) in 12 months if not surgically excised ▪ If stable follow up Ultrasound can be done at 24 months from initial exam ◦ If follow up ultrasound imaging shows changing morphology and/or a vascular component then consider MRI Pelvis without and with contrast (CPT® 72197)	• Same as Pre-Menopausal

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Condition	Pre-Menopausal	Post-Menopausal
Typical Dermoid ≥ 10cm (O-RADS™ 3)	• If ultrasound equivocal for Dermoid, MRI Pelvis without and with contrast (CPT 72197) • If initial ultrasound imaging confirms a Typical Dermoid ≥10cm ◦ If not excised consider TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856) follow up within 6 months ▪ If stable follow up Ultrasound can be done at 12 and 24 months from initial exam ◦ If solid component, MRI Pelvis without and with contrast (CPT® 72197) may be approved ◦ If follow up ultrasound imaging shows changing morphology and/or a vascular component then consider MRI Pelvis without and with contrast (CPT® 72197)	• Same as Pre-Menopausal
Typical benign extraovarian lesions Hydrosalpinges (Hydrosalpinx) or Peritoneal cysts (ORADS™ 2)	• If initial imaging confirms hydrosalpinx or peritoneal cysts, follow up imaging is not medically necessary	• If initial imaging confirms hydrosalpinx or peritoneal cysts, follow up imaging is not medically necessary

Complex and/or solid adnexal mass incompletely evaluated by ultrasound

- Generally a repeat ultrasound is medically necessary (see table above for appropriate time intervals): TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76857 or CPT® 76856)
- MRI Pelvis without and with contrast (CPT® 72197, CPT® 72195 if pregnant) one time is medically necessary as follows:
  - To follow masses when they cannot be optimally visualized by ultrasound (e.g. suboptimal sonography due to large mass or obese individual)
  - Unexplained change of appearance during ultrasound follow-up

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- Other Individual-driven indications (e.g. the application of established risk prediction models (e.g., family history of ovarian cancer), correlation with abnormal serum biomarkers, and/or pelvic symptoms)
- Differentiate the origin of pelvic masses that are not clearly of ovarian origin
- O-RADS™ score of 3 with a solid component
- O-RADS™ score of 4 or 5

- Concern for metastatic ovarian malignancy, see **Ovarian Cancer (ONC-21)** in the Oncology Imaging Guidelines

Background and Supporting Information

O-RADS™ Classification

O-RADS	
O-RADS™ 0	Incomplete Evaluation
O-RADS™ 1	Normal Ovary • No ovarian lesion • Physiologic cyst: follicle ≤3cm or corpus luteum typically ≤3cm
O-RADS™ 2	Almost Certainly Benign • Simple cyst less than 10 cm • Bilocular, smooth cyst • Unilocular, smooth, non-simple cysts (internal echos and/or incomplete septations) • Typical benign ovarian lesions <10cm (hemorrhagic cyst, dermoid cyst, endometrioma) • Typical benign extraovarian lesions (paraovarian cyst, peritoneal inclusion cysts, hydrosalpinx)
O-RADS™ 3	Low Risk • Typical benign ovarian lesions ≥10cm • Uni- or bilocular cyst, smooth, ≥10cm • Unilocular cyst, irregular, any size • Multilocular cyst, smooth, <10cm, Color Score (CS) <4 • Solid lesion, ± shadowing, smooth, any size, CS =1 • Solid lesion, shadowing, smooth, any size, CS 2-3

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O-RADS	
ORADS™ 4	Intermediate Risk • Bilocular cysts without solid component(s), Irregular, any size, any color score • Multilocular cysts without solid component(s) ◦ Smooth, 10 cm, CS <4 ◦ Smooth, any size, CS 4 ◦ Irregular, any size, any CS • Unilocular cyst with solid component(s) ◦ <4 papillary projections or any solid component(s) not considered a papillary projection, any size • Bl- or multilocular cyst with solid component(s), any size, CS 1-2 • Solid lesion, non-shadowing, smooth, any size, CS 2-3
ORADS™ 5	High Risk • Unilocular cyst, ≥4 papillary projections, any size, and CS • Bi- or multilocular cyst with solid component(s), any size, CS 3-4 • Solid lesion, ± shadowing, smooth, any size, CS 4 • Solid lesion, irregular, any size, any CS • Ascites and/or peritoneal nodules

Pre-Menopausal – see table above

- For females assigned at birth of reproductive age (Pre-Menopausal), evaluation may include a pregnancy test (a quantitative hCG may be necessary if an ectopic pregnancy is suspected), CBC, serial hematocrit measurements, and appropriate cultures.
- Symptomatic individuals often require immediate interventions (antibiotics, surgery, and/or expectant management).
- Ultrasound characteristics usually suggest the diagnosis (ectopic pregnancy, functional cysts, tubo-ovarian abscess (See **Pelvic Inflammatory Disease (PV-7.1)**), hydrosalpinx, dermoid, endometrioma, hemorrhagic cyst and pedunculated fibroids (See **Leiomyoma/Uterine Fibroids (PV-12.1)**) and direct the treatment.
- An ovarian mass suspicious for metastatic disease (e.g. from breast, uterine, colorectal or gastric cancer) should be evaluated based on the appropriate Oncology Imaging Guidelines.

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Post-Menopausal – see table above

- For post-menopausal females assigned at birth, most pelvic complex cysts or solid masses should be evaluated for surgical intervention and have tumor markers (i.e. CA-125) measured.
- Some for whom the usual management of a pelvic mass would include surgery may be at increased risk for perioperative morbidity and mortality. In such cases, repeat imaging may be a safer alternative than immediate surgery, although the frequency of follow-up imaging has not been determined.
- An ovarian mass suspicious for metastatic disease (e.g. from breast, uterine, colorectal or gastric cancer) should be evaluated based on the appropriate Oncology Imaging Guidelines.

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Screening for Ovarian Cancer/Suspected Ovary Cancer (PV-5.4)

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- See **Ovarian Cancer (ONC-21)** in the Oncology Imaging Guidelines

Evidence Discussion (PV-5)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for female reproductive organs[3,14]. Ultrasound has high sensitivity (>90%) for adnexal pathology[14]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow and differentiate solid components[2]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access and lack of ionizing radiation exposure. MRI is accepted as an adjunct modality to ultrasound in cases where ultrasound may not fully characterize a soft tissue abnormality due to its superior signal to noise ratio[14,15,16]. CT is of limited use in the evaluation of adnexal masses given its suboptimal delineation of adnexal soft tissue[14].
- Accurate diagnosis of adnexal pathology is imperative in order to limit invasive interventions for benign lesions[15] and improve preoperative triage to a gynecologic oncologist for high-risk lesions. In order to standardize reporting of adnexal lesions, the American College of Radiology (ACR) has created the Ovarian-Adnexal Reporting and Data-System (O-RADS)[7]. A meta-analysis of 26 studies demonstrated that O-RADS has high sensitivity for detection of malignancy (95%)[4]. A classification of O-RADS US Category 2 has an extremely low risk of malignancy (<1%), while a Category 5 has a high risk of malignancy (≥50%)[16]. For an indeterminate lesion on ultrasound or features concerning for malignancy, adjunct imaging with MRI is supported to aid in preoperative triage.

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Endometriosis (PV-6)

Guideline

Endometriosis (PV-6.1)
References (PV-6)

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Endometriosis (PV-6.1)

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- TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857) is medically necessary as the first line diagnostic exam for suspected endometriosis.
- MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) is medically necessary as follows:
  - Prior to planned surgery for suspected deep pelvic endometriosis such as rectovaginal endometriosis, deeply infiltrative bladder endometriosis, and cul-de-sac obliteration.
  - To characterize complex adnexal masses as endometrioma if ultrasound equivocal See **Complex Adnexal Masses (PV-5.3)**
    - If known or suspected thoracic endometriosis, see **Pneumothorax/Hemothorax (CH-19.1)**in the Chest Imaging Guidelines.

Evidence Discussion (PV-6.1)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound (TVUS) are widely accepted as the initial imaging modality of choice for evaluation of endometriosis[2,4,5,7]. A meta-analysis by Hudelist et al found transvaginal ultrasound was found to have a sensitivity and specificity of 91 and 98%, respectively, with a positive predictive value of 98% and negative predictive value of 95%[2]. A study by Goncalves et al compared TVUS done preoperatively to diagnostic laparoscopy for deep and ovarian endometriosis[7]. This study found TVUS to be accurate in identifying all sites of ovarian and deep endometriosis, with significantly higher sensitivity than diagnostic laparoscopy in detecting rectosigmoid endometriosis. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure.
- MRI of the pelvis can be useful for cases of suspected deep pelvic endometriosis[5,3]. A study by Macario et al found MRI of the pelvis prior to laparoscopy to have an overall sensitivity of 91.9% and specificity of 91.2% in the preoperative diagnosis of deep pelvic endometriosis with cul-de-sac obliteration[3]. MRI is also indicated for further evaluation of suspected endometrioma of the ovary if ultrasound is equivocal[11,12]. The American College of Radiology (ACR) Appropriateness Criteria for adnexal mass states, "When an adnexal mass is indeterminate on US, either the organ of origin is uncertain or it is unclear whether the mass is benign or malignant, then MRI with intravenous (IV) contrast (if feasible) becomes the modality of choice.[12]" Per the ACR Appropriateness Criteria, "MRI can readily diagnose typical endometriomas." A study by Dirrichs et al found MRI to improve sensitivity and specificity of diagnosis for indeterminate adnexal masses found with TVUS[13]. In this study, MRI changed the management decision in 34% of patients[13].

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Pelvic Inflammatory Disease (PID) (PV-7)

Guideline

Pelvic Inflammatory Disease (PV-7.1)
References (PV-7)

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Pelvic Inflammatory Disease (PV-7.1)

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- Clinical examination alone is usually sufficient for confirming the diagnosis of pelvic inflammatory disease. See **Pelvic Pain/Dyspareunia, Female (PV-11.1)** if other causes of pelvic pain are suspected.
- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) is medically necessary as the initial study for imaging of suspected pelvic inflammatory disease (PID) if diagnosis is uncertain following bimanual pelvic examination and laboratory testing (such as WBC, CRP and ESR, Microscopy of the vaginal secretions, and testing for Neisseria gonorrhoeae and Chlamydia trachomatis) OR for suspected Tubo-Ovarian Abscess (TOA). Color Doppler ultrasonography (CPT® 93975 or CPT® 93976) may be added.
- CT Pelvis with contrast (CPT® 72193) or MRI Pelvis with and without contrast (CPT® 72197) is medically necessary:
  - If diagnosis is uncertain following examination, laboratory testing and ultrasound
  - Ultrasound shows extensive abscess formation and further imaging is needed for treatment planning
  - Suspected TOA with inconclusive ultrasound
- If suspected abdominal abscess see **Abdominal Sepsis (Suspected Abdominal Abscess) (AB-3.1)**in the Abdomen Imaging Guidelines.

Background and Supporting Information

PID may be clinically suspected based on findings of abdominal and/or pelvic pain, cervical or vaginal mucopurulent discharge, dyspareunia, inter-menstrual and/or post coital bleeding, fever, low back pain, nausea/vomiting, urinary frequency, cervical motion tenderness, uterine and/or adnexal tenderness on exam.

Laboratory findings may include elevated erythrocyte sedimentation rate, elevated C-reactive protein, lab documentation of cervical infection with N. gonorrheae or C. trachomatis, WBC on saline microscopy of vaginal fluid, and/or endometrial biopsy with endometritis.

Evidence Discussion (PV-7.1)

- Clinical examination and laboratory testing are appropriate in the initial diagnostic testing for suspected pelvic inflammatory disease (PID)[2,7,10]. Imaging studies can be helpful when further evaluation is needed and to rule out other differential diagnoses such as ovarian cysts or gastrointestinal disease[2,7,10].

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- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for evaluation of pelvic inflammatory disease[2,7,10,8,9]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure. The addition of Power Doppler to ultrasonography has been found to increase sensitivity in the diagnosis of PID[2,7,8,9].
- CT Pelvis or MRI Pelvis can be considered if further imaging is needed following inconclusive ultrasound for diagnosis of PID, suspected tubo-ovarian abscess, or to evaluate for the extent of PID abscess formation for treatment planning[2,7,8,9,6,4].

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Polycystic Ovary Syndrome (PV-8)

Guideline

Polycystic Ovary Syndrome (PCOS) (PV-8.1)
References (PV-8)

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Polycystic Ovary Syndrome (PCOS) (PV-8.1)

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- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) is medically necessary when history, exam, and/or laboratory findings are suspicious for PCOS.
- Laboratory testing to be done prior to advanced imaging: Virilizing hormone levels (Testosterone and DHEAS). Disorders that mimic the clinical features of Polycystic ovary syndrome (PCOS) should be excluded by measuring: TSH, Prolactin, and 17-OHP (hydroxyprogesterone) levels. Others to consider based on the clinical presentation: Cortisol levels, ACTH, dexamethasone suppression testing, IGF-1, FSH, LH, estradiol.
- If elevated serum levels of androgens are found and an adrenal etiology is suspected - see **Adrenal Cortical Lesions (AB-16.1)** in the Abdomen Imaging Guidelines.

Background and Supporting Information

- Polycystic ovary syndrome is the most common hormonal disorder among females of reproductive age, and is one of the leading causes of infertility.
- Diagnostic criteria of polycystic ovary syndrome (Two of the following three criteria are required):
  - Oligo/anovulation
  - Hyperandrogenism
    - Clinical (hirsutism or less commonly male pattern alopecia) or
    - Biochemical (raised FAI (free androgen index) or free testosterone)
  - Polycystic ovaries on ultrasound
    - Defined as an ovary containing 12 or more follicles (or 25 or more follicles using new ultrasound technology) measuring 2 to 9 mm in diameter or an ovary that has a volume of greater than 10 mL on ultrasonography. A single ovary meeting either or both of these definitions is sufficient for diagnosis of polycystic ovaries.
- Clinical Features of PCOS
  - Hirsutism and male pattern balding consistent with hyperandrogenism
  - Irregular or absent menstrual cycles
  - Subfertility or infertility
  - Psychological symptoms – anxiety, depression, psychosexual dysfunction, eating disorders
  - Metabolic features – obesity, dyslipidaemia, diabetes

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Evidence Discussion (PV-8)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the modality of choice for evaluation of the ovaries in patients with suspected polycystic ovarian syndrome (PCOS)[1,2,3,6]. Ultrasound allows for real-time evaluation of the pelvic anatomy, has wide availability, fast access, and lack of ionizing radiation exposure. It also allows for follicular count which will help establish the diagnosis of PCOS[1,2,3,6].
- Laboratory testing may point to other etiology of symptoms and may better direct additional imaging[1,2,3,6].
- Imaging for suspected adrenal pathology is addressed in the Abdominal Section of these guidelines.

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Initial Infertility Evaluation, Female (PV-9)

Guideline

Initial Infertility Evaluation, Female (PV-9.1)
References (PV-9)

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Initial Infertility Evaluation, Female (PV-9.1)

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This guideline is not intended for fertility treatment follow-up and management. See individual fertility coverage policy for imaging during active fertility treatment.

- A one-time Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) is medically necessary for initial infertility workup.
  - Repeat ultrasounds or serial ultrasounds are not medically necessary for initial infertility workup
- To evaluate for tubal patency:
  - Hysterosalpingography (HSG) (CPT® 74740) **or** Sonohysterosalpingography (CPT® 76831) is medically necessary
- If ultrasound is indeterminate or there is clinical suspicion for intra-cavitary lesion (such as polyp or fibroid), hydrosalpinx, uterine synechia, adenomyosis or uterine anomalies either of the following are medically necessary:
  - 3D US imaging (add-on CPT® 76376 or CPT® 76377)
  - US Color Doppler (CPT® 93975 or CPT® 93976)

Background and Supporting Information

Some payors do not provide coverage for infertility evaluation and/or treatment.

Evidence Discussion (PV-9)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for evaluation of the female pelvic organs[1,2,5,6,8]. Ultrasound allows for real-time evaluation, has wide availability, fast access, and lack of ionizing radiation exposure[1]. The addition of 3D ultrasound is beneficial in cases when intrauterine abnormalities are suspected[5,3,4]. The diagnostic accuracy of 3D ultrasound is 90% to 95% for uterine anomalies[5]. Adding Doppler evaluation provides information about vascularity and tissue perfusion[5,7].
- Hysterosalpingography (HSG) or Sonohysterosalpingography can be utilized in assessing tubal patency. Sonohysterosalpingography is more operator dependent than HSG, however, both procedures benefit patients in that they can help avoid the more invasive laparoscopy and chromotubation, which carry the risks of surgery and anesthesia[6,8].

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Intrauterine Device (IUD) and Tubal Occlusion (PV-10)

Guideline

Intrauterine Device (PV-10.1)
Hysteroscopically Placed Tubal Occlusion Device (PV-10.2)
Implantable Contraceptive Devices (PV-10.3)
References (PV-10)

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Intrauterine Device (PV-10.1)

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- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) is medically necessary if:
  - Abnormal pelvic exam prior to IUD insertion, such as pelvic mass, irregularly shaped uterus, or enlarged uterus
  - Suspected IUD complication:
    - Abnormal IUD position
    - Uterine perforation
    - Severe pain
    - Excessive bleeding
    - Suspected infection

"Lost" IUD inability to palpate IUD string on pelvic exam, and/or see IUD on speculum exam:

- If unable to palpate, see or retrieve IUD string on pelvic exam and/or speculum exam an attempt to retrieve IUD strings with cytobrush or instrumentation of the cervix should be undertaken prior to imaging.:
  - TV ultrasound CPT® 76830 and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857); with or without 3-D Rendering (CPT® 76377 or CPT® 76376) is medically necessary:
    - If TV and/or Pelvic ultrasound is negative or non-diagnostic, plain x-ray should be performed if pregnancy test is negative
      - If IUD is not visualized on x-ray a diagnosis of expulsion can be made
    - CT Pelvis without contrast (CPT® 72192) or CT Abdomen and Pelvis without contrast (CPT® 74176) or MRI Pelvis without contrast (CPT® 72195) is medically necessary when both ultrasound and plain x-ray are equivocal or non-diagnostic as it may be useful to delineate IUD position and relationship to other abdominal organs.
- If pregnancy test is positive:
  - The use of gynecology CPT codes for pregnant females is not supported. Therefore, transvaginal ultrasound (CPT® 76830) and pelvic ultrasound (CPT® 76856 or CPT® 76857) are not medically necessary for those with a positive pregnancy test or known pregnancy. If a pregnancy test is positive, then obstetrical CPT codes are indicated. **(General Guidelines (PV-1.0)**).

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Hysteroscopically Placed Tubal Occlusion Device (PV-10.2)

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- TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857) is medically necessary if:
  - Suspected complication of hysteroscopically placed tubal occlusion device:
    - Abnormal tubal occlusion device position
    - Uterine perforation
    - Severe pain
    - Excessive bleeding

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Implantable Contraceptive Devices (PV-10.3)

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- If implant is unable to be palpated
  - If implant is radiopaque (contains barium sulphate)
    - Initial imaging should include either Ultrasound or X-ray of arm
    - If thoracic implant migration is suspected Chest X-ray should be considered
    - If Chest X-ray is equivocal CT Chest without or with contrast (CPT® 71250 or CPT® 71260) or CTA Chest (CPT® 71275) is medically necessary
  - If implant is radiolucent
    - Initial imaging should include Ultrasound of the arm
    - MRI Upper Extremity without contrast (CPT® 73218) is medically necessary if ultrasound is equivocal
    - If thoracic implant migration is suspected MRI Chest without or without and with contrast (CPT® 71550 or CPT® 71552) is medically necessary

Background and Supporting Information

- As of 2019, neither the Essure nor the Adiana tubal occlusion device is in production.
- Currently the only implant available in the United States is an etonogesterl containing implant. The original version of this implant (Implanon) was released in 2001. This was replaced by an updated implant in 2011 (Nexplanon) which contains barium sulphate, making it radiopaque and easily visualized on X-ray.
- A rare complication of the implant is distant vascular migration to the pulmonary vasculature.

Evidence Discussion (PV-10)

- Transabdominal and transvaginal ultrasounds are the initial imaging methods for locating a malpositioned IUD[12,14,11,5,2,3]. Ultrasound has the benefits of being widely available, accurate, and free from exposure to ionizing radiation. The addition of 3D image processing to ultrasound is advantageous as it allows for the visualization of the complete IUD, including the shaft and arms, and demonstrates its relationship to the endometrial cavity[8,2,3].
- In cases where the ultrasound is non-diagnostic and the pregnancy test is negative, an X-ray should be performed. X-rays are useful as IUDs are radiopaque; if the IUD is not visualized on an X-ray, a diagnosis of expulsion can be made[11].

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- If both ultrasound and X-ray results are equivocal, CT or MRI may be useful to delineate the IUD's position and its relationship to other abdominal organs[1,6].

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Pelvic Pain/Dyspareunia, Female (PV-11)

Guideline

Pelvic Pain/Dyspareunia, Female (PV-11.1)
References (PV-11)

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Pelvic Pain/Dyspareunia, Female (PV-11.1)

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- This guideline is intended for the workup of pelvic pain in relation to the female pelvic organs
  - If there is clinical concern for a non-gynecological cause of pelvic pain please see the most applicable condition-specific guideline (e.g., General Abdomen, General, General Peripheral Vascular Disease, General Nerve and Neuromuscular Disorder Imaging, etc.)
  - History, physical examination, and laboratory data can guide subsequent workup in individuals presenting with pelvic pain.
    - A pregnancy test should be performed in premenopausal females
  - Ultrasound is the initial work up for pelvic pain suspected to originate from the female pelvic organs
    - Premenopausal pelvic pain and the pregnancy test is positive,
      - See the applicable obstetrical imaging policy
    - Premenopausal pelvic pain and the pregnancy test is negative or the patient is postmenopausal
      - Ultrasound – transvaginal (CPT® 76830) and/or pelvic (CPT® 76856 or CPT® 76857)
      - Duplex Doppler (CPT® 93975 or CPT® 93976) is medically necessary if there is an ovarian mass and/or suspicion of ovarian torsion on the initial ultrasound.
      - Duplex Doppler (CPT® 93975 or CPT® 93976) is medically necessary for chronic pelvic pain (pelvic pain for 6 months or greater)
  - If ultrasound is indicative of pelvic organ disease, etc.)
    - Further imaging as per appropriate section of guidelines (i.e.-ovarian mass/ torsion **Adnexal Mass/Ovarian Cysts (PV-5)**, PID **Pelvic Inflammatory Disease (PV-7.1)**, etc.)
  - If initial ultrasound is normal
    - Further evaluation depending on the clinical suspicion may include urological work-up, gastroenterology work-up, laparoscopic evaluation(s)

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- If the initial ultrasound is equivocal for unexplained chronic pelvic pain (pelvic pain for 6 months or greater) and/or above evaluations are non-diagnostic the following is medically necessary:
  - CT Pelvis with contrast (CPT® 72193) OR
  - MRI Pelvis without contrast or with and without contrast (CPT® 72195 or CPT® 72197)

- Dysmenorrhea
  - Primary Dysmenorrhea
    - Empiric treatment with Nonsteroidal Anti-inflammatories, Hormonal Therapies or non-pharmacologic strategies
    - If failure of empiric treatment – Secondary Dysmenorrhea should be suspected
  - Secondary Dysmenorrhea
    - Initial evaluation is with Ultrasound – transvaginal (CPT® 76830) and/or pelvic (CPT® 76856 or CPT® 76857)
    - Additional work up depends on the pelvic pathology suspected
    - For suspected **Endometriosis (PV-6)**
    - For suspected **Adenomyosis (PV-4)**
    - For suspected **Fibroids (PV-12)**
- Evaluation for Pelvic Adhesive Disease
  - Initial imaging is an Abdominal (CPT® 76700 or CPT® 76705) and/or Pelvic Ultrasound (CPT® 76856 or CPT® 76857) to complete assessment for surgical planning
    - If ultrasound is inconclusive then MRI Pelvis without contrast or with and without contrast (CPT® 72195 or CPT® 72197) is medically necessary for surgical planning
- Persistent Genital Arousal Disorder/Genito-Pelvic Dysesthesia (PGAD/GPD)
  - Work-up should include detailed history, physical exam, and appropriate laboratory exams and neurogenital testing
  - Further work-up is dependent on the suspected etiology
    - For Urethral Diverticula – See **Urethral Diverticula (PV-13.2)**
    - For pelvic floor dysfunction – See **Urinary Incontinence/Pelvis Prolapse/Fecal Incontinence (PV-22)**
    - For Tarlov Cyst – See **Miscellaneous Spinal Lesions (SP 2.8)**
    - For Pudendal Neuropathy – See
    - For Pelvic Congestion Syndrome – See **Pelvic Congestion Syndrome (PVD-13.4)**

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- Work-up of interstitial cystitis/bladder pain syndrome (IC/BPS) should include history, physical exam, laboratory exam (urinalysis and urine culture), cystoscopy, and measurement of post void residual urine by bladder catheterization.
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) is then medically necessary.
    - CT Pelvis with contrast (CPT® 72193) is medically necessary if ultrasound is equivocal for complicated interstitial cystitis/bladder pain syndrome.
- Proctalgia Syndromes
  - Prior to advanced imaging, the evaluation of rectal/perineal pain should include:
    - Digital rectal examination (assess for mass, fissures, hemorrhoids, etc.)
    - Pelvic examination in females to exclude PID
    - Recent flexible sigmoidoscopy or colonoscopy subsequent to the start of reported symptoms to exclude inflammatory conditions or malignancy.
    - Endoanal ultrasound (CPT® 76872), MRI Pelvis with and without contrast (CPT® 72197), or CT Pelvis with contrast (CPT® 72193) are medically necessary after the above studies have been performed or if laboratory or clinical information suggest infection, abscess, or inflammation
- MRI (MRI Pelvis without contrast CPT® 72195) for Defecography is considered investigational/experimental by UHC.

Background and Supporting Information

- Dysmenorrhea is pain with the menstrual cycle. It can be divided into primary dysmenorrhea (pain in the absence of pelvic pathology) and secondary dysmenorrhea (pain due to pelvic pathology or recognized medical condition).
- Multiple meta-analyses have been performed regarding the detection of intra-abdominal adhesions by various imaging modalities. Limperg et al found that ultrasound has a sensitivity of 95.9%, specificity of 93.1%, a PPV of 60.4% and a NPV of 99.2% for the identification for assessment of periumbilical bowel adhesion. In addition, Gerner-Rasmussen found an overall accuracy for detecting intra-abdominal adhesions of 76-100% for ultrasound, 79-90% for MRI and 66% for CT. Ultrasound has the benefit of being widely available and lacks exposure to contrast agents. MRI is able to visualize adhesions between viscera, but has a higher false positive rate and over diagnoses adhesions.
- Interstitial Cystitis/Bladder Pain Syndrome (IC/BPS) has an unpleasant sensation (pain, pressure, discomfort), perceived to be related to the urinary bladder. It is associated with lower urinary tract symptoms of more than six weeks duration, in the absence of infection or other identifiable causes.
- Proctalgia syndromes are characterized by recurrent episodes of rectal/perineal pain, and may be due to sustained contractions of the pelvic floor musculature.

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Evidence Discussion (PV-11)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for pelvic pain of gynecologic origin[3,9,10,11]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow and distinguish fluid and cysts from soft tissue[9,10,11]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure.
- MRI is accepted as an adjunct modality to ultrasound in cases where ultrasound may not fully characterize a soft tissue abnormality due to its superior signal to noise ratio[9,10,11].

  CT of the pelvis may demonstrate engorged veins, pelvic fulid, peritoneal thickening, hydrosalpinx or pyosalpinx and tubo-ovarian abscess[9,10].
- MRI pelvis, CT pelvis or endoanal ultrasound are appropriate for the evaluation of proctalgia after digital rectal examination, pelvic examination in females and recent endoscopy to exclude inflammatory conditions or malignancy[4].
- Often, the history, physical examination, and laboratory data can guide subsequent workup in individuals presenting with pelvic pain. If initial ultrasound is normal, further evaluation may include urological work-up, gastroenterology work-up, or laparoscopic evaluation(s)[7].
  - The differential diagnosis for chronic pelvic pain is extensive. Determining the etiology of pelvic pain is important to plan treatment[5,6,7].

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Leiomyoma/Uterine Fibroids and other Uterine Masses (PV-12)

Guideline

Suspected Uterine Mass (PV-12.0)
Leiomyoma/Uterine Fibroids (PV-12.1)
Endometrial Polyp (PV-12.2)
Asymptomatic Thickened Endometrium (PV-12.3)
References (PV-12)

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Suspected Uterine Mass (PV-12.0)

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- A potential mass found on exam and/or found incidentally on other imaging
  - Transvaginal (TV) ultrasound imaging (CPT® 76830) is the medically necessary initial study
    - Pelvic ultrasound (CPT® 76856 or CPT® 76857) is medically necessary if requested as a complimentary study to the TV ultrasound
  - Further imaging as per appropriate section of guidelines (i.e., ovarian mass/torsion **Adnexal Mass/Ovarian Cysts (PV-5)**, **Adenomyosis (PV-4.1)**, etc.)

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Leiomyoma/Uterine Fibroids (PV-12.1)

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Leiomyomata are also known as “fibroids.”

- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) are medically necessary for any of the following:
  - Suspected leiomyoma with symptoms of pelvic pain, suspected ureteral obstruction secondary to inability to void urine, pelvic pressure and/or abnormal uterine bleeding and/or an enlarged uterus found on physical exam with a negative pregnancy test (if pre-menopausal).
  - Pre-operative prior to myomectomy
  - Recurrent symptoms such as abnormal bleeding, pain, or pelvic pressure
  - 3-D Rendering (CPT® 76377 or CPT® 76376) and/or Duplex (Doppler) scan (CPT® 93975 complete; CPT® 93976 limited) if ultrasound is equivocal and intracavitary lesion is suspected, or for surgical planning for myomectomy
  - There is no current evidence to support 3-D Rendering (CPT® 76377 or CPT® 76376) for planning for uterine artery embolization.
- MRI Pelvis and/or Abdomen to determine surgical approach for hysterectomy is not medically necessary.
- MRI Pelvis without and with contrast (CPT® 72197), or without contrast (CPT® 72195) in the evaluation of leiomyomas is medically necessary for the following:
  - Guide the treatment of leiomyoma/fibroid in an enlarged uterus with multiple leiomyoma/fibroid following indeterminate ultrasound when myomectomy is planned.
  - Equivocal sonohysterography or panoramic hysteroscopy with suspected submucous leiomyoma and imaging is needed to plan for myomectomy
  - Leiomyoma necrosis is suspected
  - Guide the treatment of leiomyoma/fibroid in an enlarged uterus with multiple leiomyoma/fibroid following indeterminate ultrasound when Radiofrequency Ablation of Leiomyomas is planned
  - Uterine artery embolization is being considered
    - If MRI is equivocal, MRA Pelvis (CPT® 72198) or CTA Pelvis (CPT® 72191) if requested by or in consultation with the interventional radiologist planning the uterine artery embolization
    - There is no evidence to support interval MRI after embolization unless persistent or recurrent symptoms

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        - The vascular embolization code (CPT® 37243) is inclusive of all radiological supervision and interpretation, intraprocedural roadmapping, and imaging guidance necessary to complete the intervention.
            - Additional advanced imaging codes are not medically necessary during the procedure to complete the intervention.
- If malignancy is suspected, See **Oncology Imaging Guidelines**.
    - MRI Pelvis with and without (CPT® 72197) for suspected leiomyosarcoma is medically necessary if one or more of the following ultrasound features AND symptoms are present;
        - Ultrasound features suggestive of leiomyosarcoma are:
            - Large sized (greater than 8 cm)
            - Irregular borders
            - Areas of cystic change or necrosis
            - Increase in central and peripheral vascularity
            - Rapid change in size
        - Symptoms suggestive of leiomyosarcoma would include postmenopausal woman with an new or rapidly enlarging myometrial mass or rapid growth of a uterine mass in a premenopausal patient (increase of 6 weeks gestation size within 1 year)
- CT is not medically necessary for evaluating female pelvic anatomy due to its limited soft tissue contrast resolution

Evidence Discussion (PV-12)

- Transabdominal pelvic and/or transvaginal pelvic ultrasound are widely accepted as the initial imaging modality of choice for uterine fibroids[1,2]. Ultrasound also allows for real-time evaluation with color and power Doppler which can help identify vascular flow and distinguish fluid and cysts from soft tissue. 3-D rendering is useful for further evaluation of intracavitary lesions and for surgical planning for myomectomy[3]. Additional benefits to ultrasound as a first line imaging modality include wide availability, fast access, and lack of ionizing radiation exposure.
- MRI is accepted as an adjunct modality to ultrasound in cases where ultrasound may not fully characterize a soft tissue abnormality due to its superior signal to noise ratio. MRI can be useful for surgical planning for myomectomy, determining degeneration or necrosis of fibroids, and to plan uterine artery embolization or radiofrequency ablation[4,5,7,9].
- MRI may be considered for suspected leiomyosarcoma in cases where ultrasound features and symptoms are suggestive of this diagnosis[8]. The reported prevalence of unsuspected sarcoma at surgery for symptomatic leiomyoma ranges widely, from 0.01% (one in 10 000) to 0.28% (one in 352)[8].

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- MRI offers the highest accuracy for characterization of uterine masses before intervention due to improved soft-tissue contrast, larger field of view, diffusion sequences, and multiplanar sequences. For procedural planning, MRI offers better localization of fibroid position in the uterus and can be used to assess viability and arterial supply of fibroids. In the context of preprocedural planning, MRI features have been evaluated for performance in separating leiomyosarcoma from leiomyomas or atypical leiomyomas. MRI features noted in multiple studies as associated with leiomyosarcoma include the following features: intermediate to high signal intensity of the mass at T2-weighted imaging, irregular margins of the uterine mass with the adjacent myometrium, and high signal intensity at high–b value diffusion-weighted imaging and corresponding low signal intensity on apparent diffusion coefficient maps[8].
- MRA or CTA may be used to determine vascular flow to uterine fibroids for embolization planning in cases where MRI is insufficient. Knowledge of the vascular supply for fibroids is crucial for successful embolization of target arteries[11].
- CT is of limited use in the evaluation of pelvic anatomy due to its limited soft tissue contrast resolution[10].

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Endometrial Polyp (PV-12.2)

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- Transvaginal (TV) ultrasound imaging (CPT® 76830) is the medically necessary initial study
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) is medically necessary if requested as a complimentary study to the TV ultrasound
  - 3-D Rendering (CPT® 76377 or CPT® 76376) is medically necessary for further delineation of the intracavitary lesion
- Hysteroscopy remains the gold standard for diagnosis and treatment of endometrial polyps
  - Allows for direct visualization, histologic diagnosis and concurrent treatment of the polyp
  - If biopsy confirms a malignancy, then see the appropriate oncology guideline

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Asymptomatic Thickened Endometrium (PV-12.3)

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- Thickened endometrium (>4mm) incidentally discovered in a postmenopausal individual without bleeding
  - Need not routinely trigger evaluation, but individualized assessment, including endometrial sampling, based on patients risk factors is medically necessary
    - If biopsy confirms a malignancy, then see the appropriate oncology guideline
    - Advanced imaging is not medically necessary for Endometrial Intraepithelial Neoplasia or Atypical Endometrial Hyperplasia (EIN-AEH) as imaging has not been found to improve outcomes for this condition.

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Periurethral Cysts, Urethral Diverticula, and Vaginal Masses (PV-13)

Guideline

Periurethral cysts, Skene duct cyst, Bartholin's gland cysts and Gartner’s duct cyst (PV-13.1)
Urethral Diverticula (PV-13.2)
Vaginal Masses (PV-13.3)
References (PV-13)

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Periurethral cysts, Skene duct cyst, Bartholin's gland cysts and Gartner's duct cyst (PV-13.1)

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- Initial evaluation includes any of the following:
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or Transvaginal ultrasound (CPT® 76830) and/or Transperineal ultrasound (CPT® 76872) is medically necessary
    - MRI Pelvis without and with contrast (CPT® 72197) is medically necessary for surgical planning when ultrasound equivocal

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Urethral Diverticula (PV-13.2)

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- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or Transvaginal ultrasound (CPT® 76830) and/or Transperineal ultrasound (CPT® 76872) are medically appropriate for initial evaluation but are not required prior to advanced imaging.
- Urethrography or CT Urethrography (CT Pelvis without and with contrast CPT® 72194 or CT Pelvis with contrast CPT® 72193) is medically necessary to evaluate any urethral abnormalities.
- MRI Pelvis without and with contrast (CPT® 72197) is medically necessary for surgical planning

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Vaginal Masses (PV-13.3)

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- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or Transvaginal ultrasound (CPT® 76830) and/or Transperineal ultrasound (CPT® 76872) is medically necessary for initial evaluation
- MRI Pelvis without and with contrast (CPT® 72197) is medically necessary for surgical planning

Background and Supporting Information

Symptomatic infection of congenital periurethral glands can result in urethral diverticula. Symptoms include pain, urinary urgency, frequency of urination, recurrent urinary tract infection, dribbling after urination, or incontinence.

Evidence Discussion (PV-13)

- Transabdominal, transvaginal and transperineal ultrasound are often utilized as initial imaging for female pelvic anatomy[5,6,10,11]. Ultrasound has the benefit of being widely available, accurate and does not have exposure to ionizing radiation. MRI is useful in cases of equivocal ultrasound imaging or for surgical planning[5,6,11].
- Multiple modalities can be used for the detection of urethral diverticula. Transperineal and transvaginal ultrasound can be utilized in detecting urethral diverticula. Ultrasound has the advantage of being readily available, does not require catheterization and lacks exposure to ionizing radiation. However ultrasound is operator dependent and the reported sensitivity for detection of urethral diverticula ranges from <50 to 100%[4]. Urethrography can also be used to detect urethral diverticula with a sensitivity of 67-95% but carries the risk of radiation exposure[4]. MRI has excellent soft tissue resolution and has a reported sensitivity of 100% for urethral diverticula[2].

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Congenital (Mullerian) Uterine and Vaginal Anomalies (PV-14)

Guideline

Uterine Anomalies (PV-14.1)
Vaginal Anomalies (PV-14.2)
References (PV-14)

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Uterine Anomalies (PV-14.1)

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- This guideline is intended for congenital uterine anomalies.
- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) is medically necessary for initial evaluation. 3-D Rendering (CPT® 76377 or CPT® 76376) may be an add-on if uterine anomaly is suspected on ultrasound.
- If ultrasound is indeterminate:
  - Sonohysterosalpingography (CPT® 76831) is medically necessary
- Retroperitoneal ultrasound (CPT® 76770 or CPT® 76775) is medically necessary to evaluate for possible coexisting renal anomalies if a complex uterine anomaly is present.
  - MRI Abdomen without contrast or without and with contrast (CPT® 74181 or CPT® 74183) or CT urography (CT Abdomen and Pelvis without and with contrast CPT® 74178) is medically necessary for indeterminate renal anomaly on ultrasound.
- An arcuate uterus is considered a normal variant. Therefore, advanced imaging of a known arcuate uterus is not medically necessary.
- MRI Pelvis without and with contrast (CPT® 72197) is medically necessary when:
  - Ultrasound is indeterminate for a complex uterine anomaly, or
  - Requested for surgical planning of previously diagnosed uterine anomaly
    - If ultrasound confirms uterine septum, advanced imaging is not indicated for surgical planning

Background and Supporting Information

Mullerian anomalies are complex structural anomalies deriving from errors in the embryonic development of the mullerian duct. These may include uterine remnant or agenesis, cervical agenesis, unicornate utereus, bicornuate uterus, uterine didelphys, vaginal septum and/or other complex anomalies. Uterine septum is not a complex uterine anomaly.

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Vaginal Anomalies (PV-14.2)

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- This guideline is intended for congenital vaginal anomalies.
- Pelvic ultrasound (CPT® 76856 or CPT® 76857) and/or TV ultrasound (CPT® 76830) and/or Transperineal ultrasound (CPT® 76872) and/or Translabial ultrasound (CPT® 76857) are medically necessary for initial evaluation. 3-D Rendering (CPT® 76377 or CPT® 76376) is medically necessary as an add-on if vaginal anomaly is suspected on ultrasound.
- MRI Pelvis without and with contrast (CPT® 72197) is medically necessary when:
  - Ultrasound is indeterminate for a complex vaginal anomaly, or
  - Requested for surgical planning of previously diagnosed vaginal anomaly

Background and Supporting Information

- Mullerian anomalies are complex structural anomalies deriving from errors in the embryonic development of the mullerian duct. These may include uterine remnant or agenesis, cervical agenesis, unicornate utereus, bicornuate uterus, uterine didelphys, vaginal septum and/or other complex anomalies. Uterine septum is not a complex uterine anomaly.

Evidence Discussion (PV-14)

- Transabdominal and transvaginal ultrasound remain the preferred initial imaging for female pelvic anatomy[1]. Ultrasound has the benefit of being widely available, accurate and does not have exposure to ionizing radiation, making it an excellent first line modality for the evaluation of Müllerian anomalies. With the addition of 3D imaging, ultrasound has a reported sensitivity as high as 100% for the detection of uterine anomalies[2]. MRI is also highly sensitive for the detection of uterine anomalies and is useful in cases of equivocal ultrasound imaging or for surgical planning of known complex malformations[2].
- For detection of congenital anomalies of the kidney and upper urinary tract ultrasound is usually the first line imaging modality because of its wide availability, low cost and lack of ionizing radiation[6,7]. CT or MRI can be utilized for further delineation of the renal anatomy in cases where ultrasound is inconclusive.

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Fetal MRI and Other Pregnancy Imaging (PV-15)

Guideline

Fetal MRI (PV-15.1)
Placenta Accreta/Placenta Accreta Spectrum/Placenta Percreta (PV-15.2)
C-section, Cornual or Interstitial Ectopic Pregnancy (PV-15.3)
Pelvimetry (PV-15.4)
Abnormal Non-Invasive Prenatal Testing (NIPT) and Maternal Malignancy (PV-15.5)
References (PV-15)

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Fetal MRI (PV-15.1)

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CPT® Code Guidance
• Fetal MRI (CPT® 74712) [plus CPT® 74713 for each additional fetus]
• Do not report CPT® 74712 and CPT® 74713 in conjunction with CPT® 72195, CPT® 72196, CPT® 72197
• If only placenta or maternal pelvis is imaged without fetal imaging, use MRI Pelvis (CPT® 72195)

Indications for Fetal MRI

- Fetal MRI (CPT® 74712) [plus CPT® 74713 for each additional fetus] optimally performed after 18 to 22 weeks gestation, is medically necessary for assessment of known or suspected fetal abnormalities for counseling, surgical, or delivery planning.
  - There are cases when surgical planning may necessitate imaging earlier than 18 weeks. For those cases where surgery is to be performed prior to 18 weeks and they otherwise meet indications for imaging per this criteria, Fetal MRI may be approved.
- 3D-4D (CPT® 76376 or CPT® 76377) rendering can be added for surgical planning with diagnosis of complex CHD in the fetus or for surgical planning of other complex fetal malformations
- Repeat fetal MRI (CPT® 74712) [plus CPT® 74713 for each additional fetus] is medically necessary later in pregnancy for:
  - Delivery or perinatal surgical planning
- Fetal MRI indications include but may not be limited to the following:
  - Brain
    - Congenital anomalies
      - Ventriculomegaly
      - Agenesis of the corpus callosum
      - Abnormalities of the cavum septum pellucidum
      - Holoprosencephaly
      - Posterior fossa anomalies
      - Malformations of cerebral cortical development
      - Microcephaly
      - Solid or cystic masses
      - Cephalocele

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- Screening fetuses with a family risk for brain anomalies
  - Tuberous sclerosis
  - Corpus callosal dysgenesis
  - Malformations of cerebral cortical development
- Vascular abnormalities
  - Vascular malformations
  - Hydranencephaly
  - Intra-uterine cerebrovascular accident (CVA)

◦ Spine
- Congenital anomalies
  - Neural tube defects
  - Sacrococcygeal teratomas
  - Caudal regression/sacral agenesis
  - Syringomyelia
  - Vertebral anomalies

◦ Skull, face, and neck
- Masses of the face and neck
  - Vascular or lymphatic malformations
  - Hemangiomas
  - Goiter
  - Teratomas
  - Facial clefts
- Airway obstruction
  - Conditions that may impact parental counseling, prenatal management, delivery planning, and postnatal therapy

◦ Thorax
- Masses
  - Congenital pulmonary airway malformations (congenital cystic adenomatoid malformation; sequestration, and congenital lobar emphysema);
  - Congenital diaphragmatic hernia
  - Effusion
  - Mediastinal masses
  - Assessment for esophageal atresia
- Volumetric assessment of lung
  - Cases at risk for pulmonary hypoplasia secondary to oligohydramnios, chest mass, or skeletal dysplasias

◦ Abdomen, retroperitoneal and pelvis
- Bowel anomalies such as anorectal malformations, or complex bowel obstructions such as with megacystis microcolon hypoperistalsis syndrome

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        - Abdominal wall defect
        - Mass
            - Abdominal–pelvic cyst
            - Tumors (e.g. hemangiomas, neuroblastomas, sacrococcygeal teratomas, and suprarenal or renal masses)
        - Complex genitourinary anomalies (e.g. cloaca, prune belly syndrome)
    - Congenital Heart Disease (CHD)
    - Skeletal dysplasia
    - Multiple malformations
    - Complications of monochorionic twins/TTTS (e.g. Laser treatment of twins, demise of one twin, conjoined twins)
    - Any suspected fetal anomaly associated with severe oligohydramnios or anhydramnios.

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Placenta Accreta/Placenta Accreta Spectrum/Placenta Percreta (PV-15.2)

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- Obstetrical Ultrasound is medically necessary as the initial imaging modality, Color Doppler CPT® 93975 Duplex scan (complete) or CPT® 93976 Duplex scan (limited) is medically necessary as an add-on to evaluate vascularity for suspected or confirmed placenta accreta spectrum
- MRI Pelvis without contrast (CPT® 72195) is medically necessary if the ultrasound is indeterminate or advanced imaging is needed for surgical planning.
- MRI Pelvis without contrast (CPT® 72195) is medically necessary if only placenta or maternal pelvis is imaged without fetal imaging

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C-section, Cornual or Interstitial Ectopic Pregnancy (PV-15.3)

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- If a cornual (interstitial) ectopic or C-section scar ectopic pregnancy is suspected on ultrasound the following are medically necessary:
  - 3D rendering (CPT® 76376 or CPT® 76377), and/or Color Doppler (CPT® 93976) is medically necessary with ultrasound
  - MRI Pelvis without contrast (CPT® 72195) is medically necessary if ultrasound is inconclusive.

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Pelvimetry (PV-15.4)

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- Pelvimetry (CT or MRI Pelvimetry) lacks sufficient evidence to be clinically useful. Current recommendations are that further randomized control studies be performed before it is adapted into routine clinical practice.

Evidence Discussion (PV-15)

- Transabdominal and transvaginal obstetric ultrasound remain the preferred initial imaging for fetal evaluation of the fetus and maternal pelvic anatomy[1]. Ultrasound has the benefit of being widely available and does not have exposure to ionizing radiation.Paragraph
- Fetal MRI has emerged as an adjunct imaging to fetal ultrasound in cases where the initial ultrasound is unclear or additional information is needed for surgical or delivery planning. It has the benefits of not being limited by maternal body habitus, fetal position, ossification of fetal skull/bones, or oligohydramnios[1].
- There is much uncertainty surrounding the use of gadolinium in pregnancy. Gadolinium is water-soluble and can cross the placenta, reaching the amniotic fluid and fetal circulation. While the risk of fetal effects of gadolinium remains uncertain, it has been shown to be teratogenic in animal studies. Given these possible fetal risks, the use of gadolinium in pregnancy should be limited. Its use should only be in situations where the benefits clearly outweigh the risks[8].
- MRI can be used as an adjunct to ultrasound if there is suspicion for abnormal placentation. Sensitivity and specificity for placental invasion is comparable between ultrasound and MRI (sensitivity of 88% and sensitivity of 86% for ultrasound and 93% and 94% for MRI)[14]. MRI has also been associated with both false positive and false negative diagnoses. Hence, a stepwise approach to evaluation, starting with ultrasound, then followed by the use of MRI for equivocal or nondiagnostic ultrasound is supported.
- Ectopic pregnancy is the leading cause of maternal mortality in the first trimester[9]. Ultrasound remains the initial imaging modality for ectopic pregnancy, but MRI may add additional information, especially in cases of rare implantation-site ectopic pregnancy (e.g. Cesarean Section scar ectopic). MRI is indicated in cases where the ultrasound is nondiagnostic[9].
- There is currently insufficient evidence to support the use of imaging pelvimetry (x-ray, CT or MRI) in delivery planning[11].

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Abnormal Non-Invasive Prenatal Testing (NIPT) and Maternal Malignancy (PV-15.5)

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- Non-invasive Prenatal Testing (NIPT) is a common screening method to assess the risk of fetal aneuploidy in the antenatal period. Cell-free DNA (cfDNA) of placental origin circulating in the maternal blood is tested, typically by chromosome microarray analysis, for certain aneuploidies. The presence of genomic anomalies originating from a maternal source can confound these results and have also been linked to occult maternal malignancy.
- When found, maternally derived anomalies merit further work up.
  - Complete work up to rule out fetal source of anomalous results should be performed.
    - This would include further testing such as detailed fetal anatomy scan, amniocentesis or chorionic villus sampling (CVS) if applicable.
  - A targeted maternal work-up should be performed.
    - Complete history, physical and appropriate lab work up (i.e. CBC, peripheral smear, LDH, Liver and renal function testing, appropriate tumor markers) should be performed to better elucidate possible malignant source.
      - If clinical work up reveals a suspected source, targeted imaging depending on suspected source (i.e. mammogram, chest x-ray, liver ultrasound) should be undertaken
      - If clinical work up does not reveal potential source
        - Whole Body MRI (CPT® 76498) is medically necessary

Background and Supporting Information

Studies have show that the most common cancers found among those with abnormal NIPT were of hematologic origin, followed by breast and colon cancers.

Some tumor markers are less reliable in pregnancy. CA 125, CA 15.3, AFP and SCC Ag can be elevated in normal pregnancy. However, CA19-9, LDH, AMH, CEA and HE-4 are not commonly elevated in pregnancy. These tumor markers can aid in focusing clinical work up. NIPT is typically performed in the first and the early second trimesters. This is prior to physiologic elevation of inhibin B in the third trimester and development of

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hypertensive abnormalities of pregnancy (which can increase LDH), so these markers may be considered in early pregnancy.

Evidence Discussion (PV 15.5)

No prospective large-scale studies have evaluated NIPT and incidental detection on maternal malignancy, hence the specificity and sensitivity of this testing remains unknown. However, a series of retrospective studies report a positive predictive value of NIPT for detection of occult maternal malignancy of over 50%. There are currently no evidence-based guidelines by which to classify a NIPT result as suspicious for malignancy[1,2,3,4,5].

If initial work up determines a maternal source of the abnormal NIPT, targeted maternal work up is merited. If targeted work up fails to reveal possible source of abnormal NIPT then additional screening is merited[5,6]. Whole body MRI is preferred in pregnancy as it yields the highest diagnostic benefit. MRI also lacks ionizing radiation, making a preferred imaging modality to CT or PET. Gadolinium should be avoided during pregnancy secondary to possible risk to the fetus[7,8,9]. MRI, however, is susceptible to artifact and may not be achievable by pregnant patients, who may not be able to breath hold and lay supine for long durations[10,11].

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Molar Pregnancy and Gestational Trophoblastic Neoplasia (GTN) (PV-16)

Guideline

Molar Pregnancy and GTN (PV-16.1)
References (PV-16)

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Molar Pregnancy and GTN (PV-16.1)

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- Molar pregnancy –
  - Ultrasound is the initial medically necessary study
  - Once diagnosed on an Obstetrical Ultrasound treatment is usually evacuation.
- Individuals should undergo chest x-ray pre- and post-evacuation.
  - If chest x-ray is positive for metastases, management as per GTN guidelines, see **Gestational Trophoblastic Neoplasia (GTN) (ONC-22.5)** in the Oncology Imaging Guidelines.
- Serum hCG levels are obtained every 1-2 weeks after treatment of molar pregnancy until they normalize
- Individuals with a molar pregnancy and rising or plateauing hCG levels post evacuation and/or Gestational trophoblastic neoplasia
  - See **Gestational Trophoblastic Neoplasia (GTN) (ONC-22.5)** in the Oncology Imaging Guidelines.

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Impotence/Erectile Dysfunction (PV-17)

Guideline

Impotence/Erectile Dysfunction (PV-17.1)
References (PV-17)

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Impotence/Erectile Dysfunction (PV-17.1)

PV.ED.0017.1.A
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- Imaging depends on the suspected disease and is medically necessary as follows:
  - Penile Doppler ultrasound (CPT® 93980) if erectile dysfunction suspected[2]
  - CTA Pelvis with contrast (CPT® 72191) if large vessel vascular insufficiency is suspected following ultrasound.
  - Duplex ultrasound (CPT® 93980) to assess penile vasculature in Peyronie's disease[1]
  - If male hypogonadism is suspected, See **Pituitary (HD-19)** in the Head Imaging Guidelines
- Functional MRI or PET studies are not medically necessary for this indication.
- Priapism
  - Penile Doppler Ultrasound (CPT® 93980) is medically necessary if non-ishemic priapism is suspected
  - MRI likely does not have a role in the initial diagnosis of priapism given the time sensitive nature of diagnosis and management
  - In patients with persistent non-ischemic priapism where an embolization may be necessary CTA (CPT® 72191) or MRA Pelvis (CPT® 72198) is medically necessary
  - Penial Doppler Ultrasound (CPT® 93980) is medically necessary post procedure for ischemic priapism
  - If patient has priapism >24-48 hours or refractory to treatment, MRI Pelvis without and with contrast (CPT® 72197) or MRI Pelvis without contrast (CPT® 72195) is medically necessary

Evidence Discussion (PV-17)

- Erectile dysfunction (ED) may utilize penile Doppler ultrasound to assess penile vasculature. Ultrasound has the advantages of being able to provide robust information about both cavernous arterial inflow and the veno-occlusive capacity of the penis[7], is readily available, minimally invasive and tolerated well by patients[8]. Advanced imaging with CTA of the pelvis with contrast may be indicated if large vessel vascular insufficiency is suspected. A penile duplex ultrasound may be utilized in the workup of Peyronie's disease[1].
- Advanced imaging for ED or Peyronie's disease with either PET or functional MRI is considered investigational.
- A penile Doppler ultrasound may be utilized for workup of non-ischemic priapism or post procedure for ischemic priapism[5]. The sensitivity of Doppler ultrasound in localizing an anterior-cavernosal fistula is approximately 100%[4]. If embolization is planned, CTA or MRA of the pelvis may be indicated.

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Penis–Soft Tissue Mass (PV-18)

Guideline

Penis-Soft Tissue Mass (PV-18.1)
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Penis-Soft Tissue Mass (PV-18.1)

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- Penile ultrasound (CPT® 76857) is medically necessary for initial evaluation soft-tissue lesions of the penis, Duplex (Doppler) scan CPT® 93975 complete; CPT® 93976 limited) is medically necessary as an add-on.
- If primary penile cancer is suspected, biopsy is indicated
  - For further workup of biopsy confirmed penile cancer see **Cancers of External Genitalia – Initial Work-up/Staging (ONC-24.6)** in the Oncology Imaging Guidelines.
- Peyronie's Disease
  - Ultrasound (CPT® 76857) is medically necessary
  - MRI Pelvis without and with contrast (CPT® 72197) is medically necessary if ultrasound is equivocal and surgery or injection therapy is being contemplated

Evidence Discussion (PV-18)

- Soft tissue lesions of the penis can be evaluated with penile ultrasound with doppler imaging as an initial evaluation[1]. Ultrasound allows a readily available, non-invasive option for accurate assessment of the vascular and structural features of the penis while avoiding ionizing radiation. Advanced imaging with CT abdomen and pelvis and/or lymphoscintigraphy or SPECT/CT may be indicated for biopsy proven cancer depending on the stage, however is not necessary for the initial workup of a penile mass[2].
- Peyronie's disease can be initially assessed utilizing ultrasonography[5]. Advanced imaging with MR can be performed after equivocal ultrasound if necessary prior to surgical intervention or injection therapy[6].

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Male Pelvic Disorders (PV-19)

Guideline

Male Pelvic Disorders (PV-19.1)
References (PV-19)

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Male Pelvic Disorders (PV-19.1)

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- This guideline is intended for the workup of pelvic pain in relation to the male pelvic organs
  - If there is clinical concern for a non-genitourinary cause of pelvic pain please see the most applicable condition-specific guideline (e.g., General Abdomen, General, General Peripheral Vascular Disease, General Nerve and Neuromuscular Disorder Imaging, etc.)
- Chronic Pelvic Pain Syndrome (CPPS)
  - This may include males experiencing chronic perineal pain, bilateral scrotal content pain, penile pain, pelvic pain or pain with ejaculation. See: **Scrotal Pathology (PV 20.1)** for evaluation of scrotal pain as predominant symptom.
  - Workup includes comprehensive history, physical exam, and laboratory studies as well as cystoscopy, urodynamics, and/or imaging when the diagnosis is unclear
    - Transabdominal pelvic ultrasound (bladder and prostate [CPT® 76856 or CPT® 76857]) is medically necessary as initial imaging
    - CT Pelvis with contrast (CPT® 72193) OR MRI Pelvis without contrast (CPT® 72195) OR with and without contrast (CPT® 72197) is medically necessary if ultrasound is equivocal or nondiagnostic and alternative clinical conditions are being considered
- Prostate
  - Prostate Disorders
    - Suspected Benign Prostatic Hypertrophy with obstructive voiding symptoms:
      - Transrectal ultrasound (CPT® 76872) or Pelvis transabdominal ultrasound (bladder and prostate [CPT® 76856 or CPT® 76857]) is medically necessary.
    - For prostatitis with urinary retention or suspected abscess any of the following imaging studies are medically necessary:
      - Transrectal ultrasound (CPT® 76872) or Pelvis transabdominal ultrasound (bladder and prostate [CPT® 76856 or CPT® 76857])
      - CT Pelvis with contrast (CPT® 72193) or MRI Pelvis without contrast (CPT® 72195) or with and without contrast (CPT® 72197) if ultrasound is equivocal for abscess or mass
  - Prostate Artery Embolization (PAE)
    - MRA Pelvis (CPT® 72198) or CTA Pelvis (CPT® 72191) is medically necessary for evaluation of the pelvic vasculature if:
      - prostate artery embolization is planned

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        - The vascular embolization code (CPT® 37243) is inclusive of all radiological supervision and interpretation, intraprocedural roadmapping, and imaging guidance necessary to complete the intervention.
            - Additional advanced imaging codes are not medically necessary during the procedure to complete the intervention.
- Testicular
    - Hematospermia, transrectal ultrasound (TRUS) (CPT® 76872) is medically necessary as the initial imaging study in all cases.
        - MRI Pelvis without contrast (CPT® 72195) or MRI Pelvis without and with contrast (CPT® 72197) is medically necessary to evaluate:
            - Suspected hemorrhage within the seminal vesicles
            - Radiation injury, neoplasia
            - Failure of conservative treatment for 2 weeks
            - Abnormal findings on Transrectal ultrasound
- Rectal
    - Proctalgia Syndromes
        - Prior to advanced imaging, the evaluation of rectal/perineal pain should include:
            - Digital rectal examination (assess for mass, prostate, fissures, hemorrhoids, etc.)
            - Recent flexible sigmoidoscopy or colonoscopy subsequent to the start of reported symptoms to exclude inflammatory conditions or malignancy
        - Endoanal ultrasound (CPT® 76872), MRI Pelvis without and with contrast (CPT® 72197), or CT Pelvis with contrast (CPT® 72193)is medically necessary after the above studies have been performed or if laboratory or clinical information suggest infection, abscess, or inflammation
    - MRI (MRI Pelvis without contrast CPT® 72195) for Defecography is considered investigational/experimental by UHC
- Bladder
    - Work-up of interstitial cystitis/bladder pain syndrome (IC/BPS) may include history, physical exam, laboratory exam (urinalysis and urine culture), cystoscopy, and measurement of post void residual urine by bladder catheterization
        - Pelvic ultrasound (CPT® 76856 or CPT® 76857) is medically necessary
            - CT Pelvis with contrast (CPT® 72193) is medically necessary if ultrasound is equivocal for complicated interstitial cystitis/bladder pain syndrome

Background and Supporting Information

- The proctalgia syndromes are characterized by recurrent episodes of rectal/perineal pain, and may be due to sustained contractions of the pelvic floor musculature.

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Evidence Discussion (PV-19)

- For individuals with lower urinary tract symptoms suspected to be caused by Benign Prostatic Hypertrophy ultrasound is the modality of choice for evaluation. It allows for assessment of bladder volume and post-void residual as well as intravesical prostatic protrusion[6]. Ultrasound is advantageous as it is readily available, effective, and free of ionizing radiation.
- Prostate Artery Embolization is an excepted treatment for the management of lower urinary tract symptoms according to the American Urological Association[11]. Imaging is indicated for further delineation of the pelvic vasculature to aid in preprocedure surgical planning. The accuracy of CTA to identify the Prostate artery has been shown to approximately 97%[13]. MRI angiography has been shown to identify the prostate artery in 76% of cases, has been helpful in identifying malignancy when suspected and does not carry the risk of radiation exposure[13].
- Transrectal ultrasound is supported for the initial diagnostic imaging for hematospermia[2]. Ultrasound has high sensitivity for detecting abnormalities of the prostate and seminal tract, demonstrating abnormalities in 82-95% of men with hematospermia[2]. Ultrasound is advantageous as it is readily available, effective, and free of ionizing radiation. It also allows for simultaneous aspiration or biopsy of any lesions detected. MRI is a useful adjunct to ultrasound imaging. CT has limited value in the evaluation of hematospermia due to its limited ability to differentiate structural changes of the prostate and seminal tract and its lack of soft-tissue contrast[2].
- In individuals with suspected Proctalgia initial evaluation should include a through exam, including digital rectal exam and direct visualization with sigmoidoscopy or colonoscopy to exclude other causes of rectal pain. Clinical history and normal digital rectal exam is often sufficient to make a diagnosis of Proctalgia[7]. If infection, abscess or inflammation is suspected imaging is indicated[15].
- The work up for interstitial cystitis/bladder pain syndrome (IC/BPS) should include a careful history, physical and laboratory examination[16]. Additional testing such as radiologic imaging should be undertaken only when it will alter the treatment approach[16]. Ultrasound may be useful for adjunct diagnosis[17] and has the advantages of being widely available and without ionizing radiation. Additional testing with CT may be appropriate when ultrasound results are inconclusive but bears the risk of ionizing radiation.

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Scrotal Pathology (PV-20)

Guideline

Scrotal Pathology (PV-20.1)
Paratesticular and spermatic cord masses (PV-20.2)
Testicular Microlithiasis (PV-20.3)
References (PV-20)

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Scrotal Pathology (PV-20.1)

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- Scrotal pain or mass
  - Scrotal ultrasound (CPT® 76870) and/or Duplex (Doppler) ultrasound (CPT® 93975 or CPT® 93976) for initial evaluation
  - MRI Pelvis without and with contrast (CPT® 72197) or Tc-99m scrotal scintigraphy (CPT® 78761) is medically necessary if ultrasound is inconclusive.
- Scrotal pain and hydrocele with suspicion of hernia
  - See Inguinal or Femoral Hernia, or Indeterminate **Groin Pain (AB-12.1)** in the Abdomen guidelines
- Cryptorchidism/undescended testis in the adult.
  - Scrotal ultrasound (CPT 76870), MRI Pelvis without and with contrast (CPT® 72197), or CT Pelvis with contrast (CPT® 72193)
- Variocele evaluation
  - Scrotal ultrasound and/or Duplex (Doppler) ultrasound (CPT® 76870 and/or CPT® 93975 or CPT® 93976) of the scrotum with color flow mapping in supine and upright positions is medically necessary to assess venous reflux into plexus pampiniformis if varicocele suspected (for example, in inguinal hernia evaluation)
  - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for right-sided varicocele, when there is suspicion for intra-abdominal pathology

Background and Supporting Information

- The causes of scrotal pain may include torsion, epididymitis, strangulated hernia, segmental testicular infarction, trauma, testicular tumor, and idiopathic scrotal edema.[1]

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Paratesticular and spermatic cord masses (PV-20.2)

PV.SP.0020.2.A

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- Scrotal ultrasound (CPT® 76870) is medically necessary as the initial imaging procedure.
  - MRI Pelvis without and with contrast (CPT® 72197), exploration and biopsy are additional considerations if ultrasound is inconclusive.

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Testicular Microlithiasis (PV-20.3)

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- Scrotal ultrasound (CPT® 76870) is medically necessary for initial evaluation
- Annual Scrotal ultrasound (CPT® 76870) follow-up is medically necessary only if a risk factor is present which include:
  - Family history of germ cell tumor
  - Maldescent
  - Orchidopexy
  - Testicular atrophy
- For personal history of germ cell tumor See **Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)** in the Oncology Imaging Guidelines

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Fistulae, Abscess, and Pilonidal Cyst (PV-21)

Guideline

Fistula in Ano (PV-21.1)
Abscess (PV-21.2)
Pelvic Fistula (PV-21.3)
Pilonidal Cyst (PV-21.4)
References (PV-21)

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Fistula in Ano (PV-21.1)

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- MRI Pelvis without and with contrast (CPT® 72197) is medically necessary.
  - If MRI cannot be performed, endoscopic ultrasound is superior, and thus preferential, to CT imaging.
  - CT Pelvis with contrast (CPT® 72193) is an inferior study to either of the above (accuracy of endoscopic ultrasound vs. CT for perianal fistula is 82% vs. 24%) and it is medically necessary only in circumstances in which MRI and endoscopic ultrasound cannot be performed.

Evidence Discussion (PV-21.1)

- Anorectal fistulas most commonly arise from abscesses that originate in the anal crypts (90%). Physical exam will frequently identify these but advanced imaging is often needed to determine the course of the fistulous tract, its relationship with the sphincteric musculature and associated infection/abscess[11]. Because of its superior resolution, MRI is the preferred modality, followed by endoscopic ultrasound and then CT[1].
- Non-iatrogenic anal fistula located in atypical positions (lateral) suggest the possibility of Crohn's disease. See IBD – Perirectal/Perianal disease (AB-23.3).

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Abscess (PV-21.2)

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- Perirectal Abscess
  - MRI Pelvis without and with contrast (CPT® 72197) is the preferred study
    - CT Pelvis with contrast (CPT® 72193) is medically necessary as an alternative study if desired.
  - For the evaluation of Perianal and Perirectal Disease related to Crohn's Disease, See **Perirectal/Perianal Disease (AB-23.3**) in the Abdomen Imaging Guidelines.
- Perineal Abscess
  - Fournier Gangrene
    - Diagnosis commonly made on the basis of clinical and physical exam
    - CT Pelvis, contrast as requested, is medically necessary for evaluating extent of disease for surgical planning
- Vulvar Abscess
  - CT Pelvis, contrast as requested, is medically necessary to delineate the extent of a vulvar abscess and evaluate surrounding structures for surgical planning
  - MRI Pelvis without and with contrast (CPT® 72197) may also be medically necessary for surgical planning

Evidence Discussion (PV-21.2)

- Pelvic infections can take the form of intraperitoneal abscesses or perineal wall infections [12].
- Refer to Abdominal Sepsis (AB-3-1) for intraabdominal pelvic abscess.
- History and physical can usually identify perineal (perirectal and perianal) abscesses[12]. Due to a high rate of recurrence due to associated fistulous tracts, advanced imaging with MRI (preferred because of its improved resolution), endorectal ultrasound or CT scan[1,11]. Primary treatment is surgical drainage[1,12].

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Pelvic Fistula (PV-21.3)

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- History and physical exam (to include pelvic and/or anorectal examination):
  - Rectovesicular Fistula - the following are medically necessary:
    - MRI Pelvis with and without contrast (CPT® 72197) OR
    - CT Pelvis with contrast (CPT® 72193)
  - Vaginal Fistula:
    - Enterovaginal, Colovaginal, Rectovaginal or Anovaginal:
      - Anoscopy and/or proctoscopy
      - Endoanal ultrasound (rarely used)
      - MRI Pelvis with and without contrast (CPT® 72197) is medically necessary as the initial modality for suspected enterovaginal fistula
      - CT Pelvis with contrast (CPT® 72193) is medically necessary if:
        - MRI contraindicated OR urgent evaluation of acute diverticulitis OR early postoperative period
    - Urinary Vaginal Fistula (Ureterovaginal, Vesicovaginal, or Urethrovaginal) - the following are medically necessary:
      - Cystoscopy
      - CT urography (CT Abdomen and Pelvis without and with contrast CPT® 74178) and/or CT cystography (CT Pelvis without contrast CPT® 72192) or
      - MRI Pelvis with and without contrast (CPT® 72197)

Background and Supporting Information

- A vaginal fistula is an abnormal communication between the vagina and either a portion of the digestive system or the urinary tract
  - Causes of vaginal fistula may include IBD, endometriosis, infection, tumor, radiation, obstetrical trauma and surgical injuries.
  - Symptoms of vaginal fistula-Persistent vaginitis, dyspareunia, perineal dermatitis, foul-smelling vaginal discharge, and/or urinary or fecal incontinence.
- A rectovesicular fistula is an abnormal communication between the rectum and the bladder.
  - Causes of rectovesicular fistula may include chronic infection, cancer, diverticulitis, IBD, radiation and surgical injuries.
  - Symptoms of rectovesicular fistula-Bubbles in the urine, brown or cloudy urine, blood in the urine, painful urination, recurrent urinary tract infection, and/or abdominal pain

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Evidence Discussion (PV-21.3)

- MRI has been established as a method of delineating vaginal fistulas. This is secondary to its excellent soft tissue resolution, allowing identification of acute inflammatory changes, post-surgical fibrosis, neoplastic tissue and abscesses[3]. It also has the benefit of lacking ionizing radiation, but may have limited access as compared with CT. MRI is also contraindicated by the presence of metallic foreign body or MRI-incompatible devices, such as some pacemakers[3]. Studies have shown a positive predictive value of 92% for delineation of anorectal vaginal fistulas[4].
- CT can also be utilized in the visualization of fistulas[3,6]. It does have lower contrast resolution than MRI[3] and does carry the risk of ionizing radiation. It may be beneficial in emergent situations given the wide availability or in situations where an MRI is contraindicated. CT-urography/cystography is also a mainstay in evaluation of the urinary tract and can be utilized to evaluate urinary vaginal fistulas[3].

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Pilonidal Cyst (PV-21.4)

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- Advanced imaging is not medically necessary for pilonidal cyst disease[9].
  - History and physical exam are sufficient for diagnosis of pilonidal cyst.
- If other underlying etiologies are suspected, refer to appropriate guidelines:
  - For suspected osteomyelitis, see: **General Infection/Osteomyelitis (MS-9.1)** in the Musculoskeletal Imaging Guidelines
  - For abdominal fistulae, see: **Fistulae (AB-48)** in the Abdomen Imaging Guidelines
  - For suspected spinal dysraphism, see: **Cutaneous Indications to Suspect Occult Spinal Dysrpahism (PEDSP-4.2)** in the Pediatric Spine Imaging Guidelines

Evidence Discussion (PV-21.4)

- Pilonidal cysts most frequently arise in the natal cleft, the groove between the buttocks overlying the sacral area. Asymptomatic disease usually does not require any treatment. Acute and chronic infections can be evaluated sufficiently with history and physical alone[9]. Advanced imaging is limited to concern for complicated disease (See **General Infection/Osteomyelitis - MS-9.1**).

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Urinary Incontinence/ Pelvic Prolapse/Fecal Incontinence (PV-22)

Guideline

Urinary Incontinence – Initial Imaging (PV-22.1)
Urinary Incontinence – Further Imaging (PV-22.2)
Pelvic Prolapse (PV-22.3)
Fecal Incontinence (PV-22.4)
References (PV-22)

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Urinary Incontinence – Initial Imaging (PV-22.1)

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- Initial Imaging, associated with other evaluations, are medically necessary as follows:
  - Non-Neurogenic Incontinence
    - Measurements of post void residual urine by Bladder ultrasound (CPT® 51798) OR Bladder catheterization
    - In addition to post void residual volume determination, screening for UTI should be considered
  - Neurogenic Incontinence
    - Ultrasound urinary tract (CPT® 76770 or CPT® 76775)

Background and Supporting Information

Urinary incontinence can be “stress,” “urgency,” or mixed; neurogenic or non-neurogenic; and complicated or uncomplicated. Neurogenic incontinence can occur from cerebral, spinal or peripheral neurological diseases.

Evidence Discussion (PV-22.1)

- The workup of urinary incontinence involves a thorough history and physical examination[2,6]. For incontinence due to non-neurogenic causes, advanced imaging is rarely necessary in the initial evaluation[2]. Assessment of the urine post void residual may be completed either with bladder ultrasound or urethral catheterization[6].
- Baseline imaging should be obtained in the evaluation of neurogenic urinary incontinence with renal bladder ultrasound[14].

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Urinary Incontinence – Further Imaging (PV-22.2)

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- CT Abdomen and Pelvis, contrast as requested, or CT Pelvis, contrast as requested, is medically necessary for any of the following:
  - Abnormality on ultrasound that requires further evaluation
  - Complicated incontinence
    - Failed conservative treatment
    - Pain or dysuria
    - Hematuria
    - Recurrent infection
    - Previous radical pelvic surgery
    - Suspected fistula
    - Suspected mass
    - Previous pelvic or prostate irradiation
  - Suspected fistulae
  - Detecting ectopic ureters if ultrasound is non-diagnostic
  - Pre-operative planning for complicated incontinence when ordered by or in consultation with the operating physician
- For neurogenic urinary incontinence See **Red Flag Indications (SP-1.2)** and **Myelopathy (SP-7.1)** in the Spine Imaging Guidelines and **Dementia (HD-8.1)** and **Normal Pressure Hydrocephalus (NPH) (HD-8.4)** in the Head Imaging Guidelines.

Evidence Discussion (PV-22.2)

- Urinary incontinence that has failed a trial of conservative treatment may require advanced imaging with CT of the abdomen and/or pelvis with or without contrast[2]. Advanced imaging may also be ordered for pre-operative planning if requested by the surgeon or to follow up on an abnormality noted on previous ultrasound[6].
- Other clinical scenarios where advanced imaging may be medically necessary are incontinence occurring concomitantly with abdominal or pelvic pain, dysuria or hematuria, or in the setting of recurrent urinary tract infections[2]. Incontinence in the setting of previous radical pelvic surgery or radiation may also require advanced imaging[14].
- If there is suspicion of a fistula, mass, or ectopic ureters (and ultrasound is non-diagnostic), advanced imaging with CT may be indicated[6].

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Pelvic Prolapse (PV-22.3)

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- Transvaginal (TV) ultrasound (CPT® 76830) and/or Transperineal ultrasound (CPT® 76872) is medically necessary as the initial study of choice
  - Pelvic ultrasound (CPT® 76856 or CPT® 76857) is medically necessary if requested as a complimentary study.
- Urodynamic testing may be helpful if there is incontinence with a stage II or greater prolapse or voiding dysfunction
- MRI Pelvis (CPT® 72195 or CPT® 72197) is medically necessary for the following:
  - Pelvic floor anatomy and pelvic organ prolapse evaluations if exam and TV ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857) are equivocal; or
  - Pre-operative planning for complex organ prolapse when ordered by or in consultation with the operating physician; or
  - Persistent incontinence following surgery
- Mesh and Graft complications
  - Diagnostic evaluation for mesh and graft complications may include colonoscopy, cystoscopy, and/or urodynamics
  - Transvaginal (TV) ultrasound (CPT® 76830) and/or Pelvic ultrasound (CPT® 76856 or CPT® 76857), CT Abdomen and/or Pelvis, contrast as requested, MRI Pelvis without contrast or without and with contrast (CPT® 72195 or CPT® 72197) are medically necessary depending on the mesh and graft complication
- Sacral osteomyelitis may be a complication of sacrocolpopexy. MRI Pelvis with and without contrast (CPT® 72197) is medically necessary for lower back pain and/or suspected sacral osteomyelitis after this procedure.
- MRI (MRI Pelvis without contrast CPT® 72195) for Defecography is considered investigational/experimental by UHC.

Evidence Discussion (PV-22.3)

- The mainstay of evaluation of pelvic organ prolapse remains clinical pelvic examination. This allows for direct evaluation of prolapse and calculation prolapse quantification[3,15].
- Translabial, transperineal or transvaginal ultrasound have shown correlation with[3,15]. Ultrasound also allows for real-time evaluation, has wide availability, fast access, and lack of ionizing radiation exposure.
- MRI has been shown to have excellent soft tissue delineation. It circumstances were clinical exam and ultrasound are equivocal, MRI may provide additional information for conditions such as enterocele, sigmoidocele and intussusception[15].

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- Complications related to mesh and graft placement in pelvic floor surgery are diverse in nature[4]. Work up for suspected complication is complex and may include a diverse range of diagnostic procedures such as radiologic imaging, cystoscopy, and colonoscopy. Surgical meshes have variable visibility[12]. Given the varied nature of this these complications modality of imaging should be tailored to suspected complication.
- A known rare complication of sacrocolpopexy is sacral osteomyelitis. In cases of suspected osteomyelitis, MRI is the preferred imaging as it has a very high sensitivity for detection infection, especially in early stages[12].

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Fecal Incontinence (PV-22.4)

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The evaluation of fecal incontinence generally proceeds as follows:

- Determine the severity of the incontinence (Bristol Stool Scale, Fecal Incontinence Severity Index, etc.)
- History and Physical to include digital rectal examination and perianal pinprick (to assess for neurogenic causes)
- Trial of conservative management
- Diagnostic Testing if symptoms persist to include:
  - Ano-rectal Manometry
  - Balloon Expulsion Test
  - Endoanal ultrasound (CPT® 76872) to confirm sphincter defects in individuals with suspected sphincter injury (e.g. history of vaginal delivery or anorectal surgery)
  - MRI Pelvis (CPT® 72197) is medically necessary if:
    - Ano-rectal manometry suggests weak sphincter pressures AND/OR there is an abnormal balloon expulsion test AND
    - There has been a failure of a trial of conservative management since the onset of or change in symptoms AND
    - Surgery is being considered
- MRI (MRI Pelvis without contrast (CPT® 72195) for Defecography is considered investigational/experimental by UHC.

Background and Supporting Information

With regards to fecal incontinence ACG Guidelines stated that “the internal sphincter is visualized more clearly by endoanal ultrasound, whereas MRI is superior for discriminating between an external anal sphincter tear and a scar and for identifying external sphincter atrophy.

However, guidelines adopted by the American Society of Colon and Rectal Surgeons stated that “Endoanal ultrasound is a useful and sensitive tool in the evaluation of patients with FI (fecal incontinence), especially when there is a history of vaginal delivery or anorectal surgery. Ultrasound can reliably identify internal and external sphincter defects that may be associated with sphincter dysfunction.” In addition, the guidelines note “Other modalities (eg, MRI) have shown substantial interobserver variability and, at this point, are likely inferior to ultrasound imaging, but they may provide additional information where endoanal ultrasound is unavailable.”

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Evidence Discussion (PV-22.4)

- According to the American College of Gastroenterology, the American Society of Colon and Rectal Surgeons and the American College of Obstetrics and Gynecology, complete history and physical exam is essential for the evaluation of patient with fecal incontinence[1,5,10].
- For individulas that fail conservative measures, ano-rectal manometry and rectal balloon expulsion testing should be performed. This may help to guide additional treatment and diagnostic testing[1].
- Endoanal ultrasound (EAUS) can be considered in individuals with suspected sphincter injury[1,5]. Ultrasound is widely available and well tolerated, however it is operator-dependent[16]. EAUS shows very good interobserver agreement in the diagnosis of sphincter defects and the measurement of the internal anal sphincter[16].
- MRI has also emerged as an imaging modality for evaluation of fecal incontinence. While EAUS is superior for the evaluation of the internal anal sphincter, MRI shows better distinction between fat and muscle in the evaluation of the external anal sphincter[16]. MRI is limited by the fact that it is not as readily available and is unsuitable for patients with limiting conditions such as metal implants and claustrophobia[16]. MRI defecography also may play a role in the evaluation of fecal incontinence as it allows for insight into important functional disorders related to defecation[16].

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Patent Urachus (PV-23)

Guideline

Patent Urachus (PV-23.1)
References (PV-23)

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Patent Urachus (PV-23.1)

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- Drainage from the umbilicus, redness around umbilicus, abdominal pain, or urinary tract infection from persistent fetal connection between the bladder and the umbilicus:
  - Ultrasound (CPT® 76856 or CPT® 76857 and/or CPT® 76700 or CPT® 76705) or voiding cystourethrography (VCUG) (CPT® 74455) is medically necessary for suspected patent urachus
  - CT Pelvis with contrast (CPT® 72193) or MRI Pelvis without contrast (CPT® 72195) or with and without contrast (CPT® 72197) is medically necessary if the ultrasound is equivocal or if additional imaging is needed for surgical planning if there is a suspected urachal carcinoma or other urachal abnormality.

Evidence Discussion (PV-23)

- A patent urachus (connecting bladder to umbilicus) can manifest as redness around or drainage from the umbilicus, abdominal pain, or urinary tract infections[1].
- If suspected, ultrasound is indicated as the initial evaluation as it can be diagnostic without exposing the patient to radiation[2,3].
- Advanced imaging of the pelvis is indicated for inconclusive ultrasound or for surgical planning[1,3].

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Bladder Mass (PV-24)

Guideline

Bladder Mass (PV-24.1)
References (PV-24)

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Bladder Mass (PV-24.1)

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- Bladder masses incidentally found on other imaging (ultrasound, cystoscopy or KUB):
  - CT Pelvis without contrast (CPT® 72192) for suspected bladder stone is medically necessary if initial imaging is equivocal or if surgery is planned
  - CT Pelvis without (CPT® 72192) is medically necessary for suspected bladder diverticuli
- See **Oncology Imaging Guidelines** for biopsy confirmed or suspected malignancy

Background and Supporting Information

Symptoms of bladder mass may include hematuria, urgency, frequency, chronic urinary infection, obstruction or urinary retention.

Evidence Discussion (PV-24.1)

- Symptoms of bladder mass may include hematuria, urgency or frequency of urination, urinary infection or urinary retention[1,4,5].
- Bladder masses may be found incidentally on initial imaging such as ultrasound, cystoscopy or KUB (Kidney, Ureter and Bladder X-ray).
- Suspected bladder stone may be further evaluated with CT pelvis if initial imaging is inconclusive or for surgical planning[5]. CT has a higher sensitivity than ultrasound for bladder stones[5].
- Suspected bladder diverticuli can be further evaluated with CT pelvis[1,4].

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Ureteral and/or Bladder Trauma or Injury (PV-25)

Guideline

Ureteral and/or Bladder Trauma or Injury (PV-25.1)
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Ureteral and/or Bladder Trauma or Injury (PV-25.1)

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- Abdominal and/or Pelvic ultrasound (CPT® 76700 and/or CPT® 76856) is medically necessary if requested
- CT cystography (CT Pelvis without contrast CPT® 72192) is medically necessary for suspected bladder injury
- CT Abdomen and Pelvis with OR with and without contrast (CPT® 74177 or CPT® 74178) is medically necessary if:
  - Suspected iatrogenic/operative injury OR
  - Blunt trauma and suspected bladder or ureteral injury with one or more of the following (See **Blunt Abdominal Trauma (AB-10.1)** in the Abdomen Imaging Guidelines):
    - Abdominal pain or tenderness
    - Pelvic or femur fracture
    - Hematocrit <30%
    - Hematuria
    - Non-examinable individual (intoxicated, less than fully conscious, Glasgow Coma Scale Score >13, etc.)
    - Evidence of abdominal wall trauma or seat-belt sign
    - Rapid deceleration injury

Background and Supporting Information

Bladder trauma: CT cystography- CT Pelvis without contrast allowing the radiologist or Urologist to instill contrast to r/o bladder injury and/or perforation.

Ureteral injury: "*Iatrogenic ureteral injuries can occur during gynecologic, obstetric, urologic, colorectal, general, or vascular surgery; gynecologic surgery accounts for more than half of all iatrogenic injuries.*"[2]

Evidence Discussion (PV-25)

- Ultrasound can performed for suspected ureteral and/or bladder trauma. It may aid in triage of injuries and may lead to immediate surgical intervention rather than additional imaging. However it has lower sensitivity compared to CT, particularly in genitourinary injury[4].
- For individuals with suspected bladder injury retrograde cystography is appropriate[2]. CT cystography has a reported 95-100% sensitivity and specificity for the diagnosis of

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bladder rupture[3,4]. It has the benefits of being widely available and accurate, but does have exposure to ionizing radiation.

- Those presenting with suspected ureteral injury CT of the abdomen and pelvis is appropriate for evaluation of the complete urinary tract. Imaging with contrast is preferred for evaluation of as it has higher sensitivity for detecting concurrent visceral organ and vascular injuries[3]. Urogram is helpful in further evaluation of the ureters [2] as it may show contrast extravasation from the ureter or partial or complete ureteral obstruction[1].

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Gender Affirmation Surgery; Pelvic (PV-26)

Guideline

Gender Affirmation Surgery; Pelvic (PV-26.1)
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Gender Affirmation Surgery; Pelvic (PV-26.1)

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- Preoperative imaging is medically necessary as outlined below if the individual has a health plan benefit covering pelvic gender affirmation surgery. Preoperative imaging is not medically necessary if pelvic gender affirmation surgery is not a health plan covered benefit.
- Preoperative imaging:
  - Metoidioplasty
    - Preoperative imaging is not medically necessary
  - Phalloplasty
    - Muscular flaps used for neophallus creation are generally obtained from anterior lateral thigh (pedicled flap) or forearm (radial free flap)
      - For planned radial free flap, upper extremity CT angiography (CPT® 73206) of anticipated donor site (unilateral) for evaluation of perforator anatomy.
      - For planned anterior lateral thigh flap, bilateral lower extremity CT angiogram (CPT® 73706)
      - If iodinazed contrast allergy, MRA (contrast as requested)
  - Vaginoplasty
    - Preoperative imaging is not supported
- Postoperative complications:
  - Doppler ultrasound is medically necessary when:
    - Monitoring of flap perfusion after phalloplasty for suspected vascular insufficiency
  - CT Abdomen and Pelvis OR CT Pelvis (contrast as requested - CPT® 74176, CPT® 74177, CPT® 74178, CPT® 72192, CPT® 72193, or CPT® 72194) is medically necessary for suspected postoperative complications
    - Complications after surgery may include hematoma, seroma, abscesses, fistula, urinary tract injury, etc. (See **Ureteral and/or Bladder Trauma or Injury (PV-25.1)** for ureteral and/or bladder injury)
  - MRI Pelvis with and without contrast (CPT® 72197) is medically necessary for:
    - Suspected fistula
    - Non diagnostic CT scan AND further imaging is needed for treatment planning

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Background and Supporting Information

- Metoidioplasty-Metoidioplasty is a procedure using clitoral hypertrophy and clitoral release to form masculine-appearing external genitalia
- Phalloplasty-Phalloplasty includes the creation of a neophallus using muscular flaps
- Vaginoplasty-Vaginoplasty refers to the surgical creation of a vulva and vaginal canal

Evidence Discussion (PV-26)

- Routine preoperative imaging is not supported for metoidioplasty or Vaginoplasty[2,4,5].
- CT angiography is indicated for preoperative evaluation for phalloplasty in order to map size, location and course of the vasculature[4,7]. CTA has been found to have high accuracy in perforator detection (sensitivity of 96-100% and specificity of 95-100% in studies investigating abdominal perforators), short time for image acquisition and high reproducibility. It however does carry the risk of ionizing radiation and exposure to iodinated contrast[8].
- Doppler ultrasound allows for monitoring of vascular perfusion of the neophallus after phalloplasty[4,5]. Ultrasound is readily available and does not carry risk of ionizing radiation.
- Expert opinion has stated that CT would be indicated for postoperative complications of gender affirmation surgery[2,4,5,7]. CT allows for fast and accurate identification of common postoperative complications such as abscess, hematoma and seroma. CT angiography aids in the diagnosis of arterial or venous thrombosis as well as identification of arterial bleeding in the setting of hemorrhage. CT does carry the risk of ionizing radiation and iodinated contrast allergy.
- MRI is the preferred modality for suspected fistula given its superior soft tissue delineation in these cases[2].

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General Guidelines (PN-1)

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Abbreviations for Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines

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Abbreviations for Peripheral Nerve Disorders Imaging Guidelines	
AIDS	acquired immunodeficiency syndrome
ALS	amyotrophic lateral sclerosis
CIDP	chronic unflammatory demyelinating polyneuropathy
CNS	central nervous system
CPK	creatinine phosphokinase
CT	computed tomography
EMG	electromyogram
LEMS	Lambert-Eaton myasthenic syndrome
MG	myasthenia gravis
MRI	magnetic resonance imaging
MRN	magnetic resonance neurography
MRS	magnetic resonance spectroscopy
NCV	nerve conduction velocity
PET	positron emission tomography
PNS	peripheral nervous system

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Abbreviations for Peripheral Nerve Disorders Imaging Guidelines	
PNST	peripheral nerve sheath tumor
POEMS	polyneuropathy, organomegaly, endocrinopathy, M-protein, skin changes
TOS	thoracic outlet syndrome

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General Guidelines (PN-1.0)

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- A pertinent clinical evaluation is required before advanced imaging can be considered. The clinical evaluation should include a pertinent history and physical examination, including a neurological examination (since the onset or change in symptoms), appropriate laboratory studies, non-advanced imaging modalities, and electromyography/nerve conduction (EMG/NCV) studies. Other meaningful technological contact (telehealth visit, telephone call or video call, electronic mail or messaging) since the onset or change in symptoms, by an established individual can serve as a pertinent clinical evaluation.
- Nerve conduction studies are often normal early in the disease course with changes occurring from 1 to 4 weeks after symptom onset in the majority of individuals. This will be taken into consideration on a case-by-case basis in regards to the EMG/NCV requirement in each section requirement of the Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines.
- Due to the termination of the federal public health emergency declaration, the COVID-19 pandemic is no longer considered an indication to waive electrodiagnostic (EMG/NCV) study requirements within the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines.
- If imaging of peripheral nerves is medically necessary, ultrasound is the preferred modality for superficial peripheral nerves. MRI may be used for imaging deep nerves such as the lumbosacral plexus or nerves obscured by overlying bone such as the brachial plexus or for surgical planning. CT is limited to cases in which MRI is contraindicated.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

Evidence Discussion (PN-1.0)

- Electromyography (EMG) and nerve conduction velocity (NCV) studies are useful in establishing the origin of peripheral nerve pathology and in guiding further

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diagnostic evaluation. Needle EMG following traumatic nerve injury may detect denervation of muscles that do not seem clinically affected. The optimal time to search for denervation changes is 10 to 14 days after the injury. Needle EMG may show residual innervation to paralyzed muscles. Follow-up EMG and NCV studies may demonstrate early evidence of re-innervation or evolving abnormalities that objectively demonstrate the temporal course of peripheral nerve pathology.

- Deferring EMG due to COVID-19 is less relevant at this time.
- For superficial peripheral nerves, ultrasound has significantly higher resolution than MRI. In terms of expense, safety, and noninvasiveness, ultrasound has clear advantages over MRI and the few comparative reports available confirm the value of ultrasound as an initial imaging choice.
- Advantages of ultrasound over MRI for detecting peripheral nerve pathology include lower cost, rapidity of examination, higher spatial resolution, imaging of the nerve in continuity, and ease of side-to-side comparisons. Ultrasound may better detect subtle changes in nerve caliber. This is important because peripheral nerve pathology is often fusiform in shape and can extend along the length of the nerve without greatly altering its cross-section area. MRI frequently misses multifocal (71%) and occasionally single pathologies.
- Advantages of MRI over ultrasound include superior contrast between tissues, imaging of structures that are deep or surrounded by bone, and tissue characterization using multi-sequence analysis and IV contrast.
- There is greater accuracy (96%) of diagnoses in cases of peripheral nerve sheath tumor, traumatic neuroma or neuropathy, idiopathic mono-neuropathy or plexopathy, fibrosis of nerves, nerve compression caused by ganglion or synovial cysts or any other soft tissue structures, non-neural soft tissue tumors, intra-neural granulomas, and vasculitis with ultrasound than MRI.

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Focal Neuropathy (PN-2)

Guideline

Focal Neuropathy (PN-2.1)
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Focal Neuropathy (PN-2.1)

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Carpal Tunnel Syndrome	YES	• When EMG/NCV and clinical findings are equivocal **AND** only when requested for pre-operative planning, MRI Upper Extremity Joint (Wrist) without contrast (CPT® 73221) is medically necessary. • For radiculopathy, see **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3)** in the Spine Imaging Guidelines.
Ulnar Neuropathy	YES	After EMG/NCV, only **ONE** of the following is medically necessary if requested for surgical consideration: • MRI Upper Extremity Joint (Elbow or Wrist) without contrast (CPT® 73221) **OR** • MRI Upper Extremity Other Than Joint (Forearm or Hand) without contrast (CPT® 73218)
Radial Neuropathy	YES	• MRI Upper Extremity Other Than Joint (Arm or Forearm) without contrast (CPT® 73218) when surgery is being considered. • MRI Upper Extremity Other Than Joint (Arm or Forearm) without and with contrast (CPT® 73220) if there is a suspicion of a nerve tumor such as a neuroma.
Radial Neuropathy Notes: Leads to wrist drop with common sites of entrapment at the inferior aspect of the humerus (Saturday night palsy) or the forearm (posterior interosseous syndrome). Entrapment of the nerve at the wrist (Wartenberg syndrome or handcuff palsy) typically spares motor involvement and results only in sensory changes. Trauma or fractures of the humerus, radius, or ulna can damage the radial nerve.		

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Pudendal Neuropathy	NO	• **Documented concern specifically for pudendal neuropathy, pudendal neuralgia, or pudendal entrapment**: MRI Pelvis without contrast (CPT® 72195) **OR** MRI Pelvis without and with contrast (CPT® 72197) ◦ If there is a contraindication to MRI **and** the above documented concern is present, then **ONE** of the following is medically necessary: ▪ CT Pelvis without contrast (CPT® 72192) ▪ CT Pelvis with contrast (CPT® 72193) ▪ CT Pelvis without and with contrast (CPT® 72194) • For all other pelvic concerns, see the following Pelvic Imaging Guidelines (as indicated): ◦ **Pelvic Pain/Dyspareunia Female (PV-11.1)** ◦ **Impotence/Erectile Dysfunction (PV-17.1)** ◦ **Male Pelvic Disorders (PV-19.1)** ◦ **Scrotal Pathology (PV-20.1)**
Pudendal Neuropathy Notes: Causes pain, sexual dysfunction, or sensory change in the genitals, perineum, and perianal region. May be caused by trauma, recurrent injury from exercise such as cycling, pelvic mass, or after viral infection (e.g., post-herpetic neuralgia).		
Sciatic Neuropathy	YES	• MRI Pelvis without contrast (CPT® 72195) • CT Pelvis without contrast (CPT® 72192) is **NOT** routinely medically necessary due to lack of soft tissue contrast. ◦ It should only be performed in the rare circumstance of contrast allergy and/or contraindication to MRI such as pacemaking device.

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Sciatic Neuropathy Notes: May be caused by trauma to the gluteal area with hematoma, injection palsy, hip or pelvic fractures, or hip replacement (arthroplasty). **Piriformis Syndrome** involves entrapment of the sciatic nerve at the sciatic notch in the pelvis by a tight piriformis muscle band. Concerns for piriformis syndrome should be imaged according to the sciatic neuropathy criteria.		
Femoral Neuropathy	NO	• MRI Pelvis without contrast (CPT® 72195)
Femoral Neuropathy Notes: May occur as a complication of pelvic surgery in females or those on anticoagulants with retroperitoneal bleeding, or as a mononeuropathy in diabetics		
Meralgia Paresthetica	NO	• MRI Pelvis without contrast (CPT® 72195) is medically necessary for **ANY** of the following scenarios: ◦ Cases of diagnostic uncertainty ◦ Pre-operative planning • CT Pelvis without contrast (CPT® 72192) is **NOT** routinely medically necessary due to lack of soft tissue contrast. ◦ It should only be performed in the rare circumstance of contrast allergy and/ or contraindication to MRI such as pacemaking device.
Meralgia Paresthetica Notes: Sensory loss in the lateral femoral cutaneous nerve as it exits the pelvis under the inguinal ligament (lateral thigh without extension into lower leg), and is usually diagnosed based on a careful history and physical exam. EMG/NCV testing is often technically difficult and not required.		
Peroneal Neuropathy	YES	• MRI Lower Extremity Joint (Knee) without contrast (CPT® 73721) **OR** MRI Lower Extremity Other Than Joint without contrast (CPT® 73718) when surgery is considered or when ordered by or in consultation with a surgeon.

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Tarsal Tunnel Syndrome	N/A	• See **Soft Tissue Mass and Morton's Neuroma (MS-10.3)** in the Musculoskeletal Imaging Guidelines.

- For phrenic nerve concerns and evaluation of diaphragmatic weakness, refer to **Elevated Hemidiaphragm (CH-30)**.

Evidence Discussion (PN-2.1)

- Focal neuropathies are typically diagnosed by a combination of clinical history, thorough neurological examination, and electrodiagnostic testing with electromyography (EMG) and nerve conduction studies (NCS).
- When clinical evaluation and electrodiagnostic testing are inconclusive, MRI may allow for better identification and anatomic localization of lesions and is considered the gold standard for imaging of the peripheral nerve.
- The sensitivity and specificity of MRI findings for carpal tunnel syndrome are low (sensitivity, 23%–96%; specificity, 39%–87%), and for this reason MRI imaging does not play a role in the routine clinical assessment of carpal tunnel syndrome. However, MRI of the wrist can help identify surgical candidates when clinical and electrodiagnostic findings are inconclusive.
- When caused by nerve entrapment or compression, focal neuropathies may benefit from surgical release or decompression. MRI can provide visualization of the cause of compression, rule out other causes of nerve injury, and allow for a more focused operative approach, particularly when surgery is considered to decompress common entrapment neuropathies of the ulnar, radial, and peroneal nerves.
- Sciatic, femoral, and pudendal neuropathies often occur secondary to trauma, compression, or entrapment of the affected nerve. These are often diagnosed clinically or localized with electrodiagnostic testing. MRI imaging of the pelvis may be medically necessary to assess for sources of compression, including occult malignancy.
- Meralgia paresthetica is the common term describing pathology of the lateral femoral cutaneous nerve of the thigh. The nerve is prone to injury and compression but may have a variable anatomic course. Meralgia paresthetica is primarily diagnosed by clinical history and exam, as neuroimaging and electrodiagnostic testing results may be difficult to interpret. Neuroimaging is most useful in cases of diagnostic uncertainty, particularly when surgical exploration and treatment are considered.

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Polyneuropathy (PN-3)

Guideline

Polyneuropathy (PN-3.1)
References (PN-3)

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Polyneuropathy (PN-3.1)

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Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments
Polyneuropathies with Central Nervous System (CNS) Involvement	YES	If clinical findings point to abnormalities in those areas, then **ANY** of the following are medically necessary: • MRI Brain without and with contrast (CPT® 70553), **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157), **AND/OR** • MRI Lumbar Spine without and with contrast (CPT® 72158)	Examples: Guillain-Barré syndrome, inflammatory polyneuropathies unspecified, and Lyme disease
AIDS-Related Cytomegaloviral Neuropathy/ Radiculopathy	YES	• MRI Lumbar Spine without and with contrast (CPT® 72158) • If concern for myelopathy, **ANY** of the following imaging are **ALSO** medically necessary: ◦ MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** ◦ MRI Thoracic Spine without and with contrast (CPT® 72157)	• Often presents with urinary retention and a clinically confusing picture in the legs. • For myelopathic signs and symptoms, see **Myelopathy (SP-7.1)** .

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Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	YES	MRI Lumbar Spine without and with contrast (CPT® 72158) **AND/OR** MRI Cervical Spine without and with contrast (CPT® 72156) if diagnosis uncertain following EMG/NCV. • For imaging requests of the brachial or lumbosacral plexus or muscle: See **Brachial Plexus (PN-4.1)**, **Lumbar and Lumbosacral Plexus (PN-5.1)**, and **Muscle Diseases (PN-8.5)**	
Multifocal Motor Neuropathy	YES	If diagnosis is uncertain following EMG/NCV, MRI of the Brachial Plexus is medically necessary with **ONE** of the following: • MRI Upper Extremity Other Than Joint without and with contrast (CPT® 73220) • MRI Chest without and with contrast (CPT® 71552) • MRI Neck without and with contrast (CPT® 70543)	
POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes)	YES	Advanced imaging is for the non-neurological etiologies of this rare osteosclerotic plasmacytoma syndrome.	See **Multiple Myeloma and Plasmacytomas (ONC-25)** in the Oncology Imaging Guidelines.
Subacute Sensory Neuronopathy & Other Paraneoplastic Demyelinating Neuropathies	YES	• Advanced imaging should be guided by specific clinical concern (see relevant guideline). • For evaluation of suspected paraneoplastic syndromes, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.	

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Background and Supporting Information

- Central nervous system (CNS) imaging (brain and spinal cord) is not required for polyneuropathy without CNS signs/symptoms.
- Distal symmetric polyneuropathy is the most common pattern of generalized peripheral neuropathy. It is typically sensory predominant and may demonstrate neurological abnormalities including reduced or absent deep tendon reflexes (DTRs), reduced sensation to multiple testing modalities (vibration, proprioception, etc). In more advanced staging, mild motor weakness may be present. It is most often associated with diabetes and metabolic abnormalities. In the absence of atypical findings (such as asymmetrical presentation, significant weakness, or upper motor neuron exam findings such as hyperreflexia or spasticity), distal symmetric polyneuropathy does not require CNS imaging.

Evidence Discussion (PN-3.1)

- Polyneuropathies are typically diagnosed by a combination of clinical history, thorough neurological examination, lab work-up, and electrodiagnostic testing with electromyography (EMG) and nerve conduction studies (NCS).
- For systemic polyneuropathies with potential for CNS involvement, such as Lyme disease-related polyneuropathy and some inflammatory polyneuropathies, MRI imaging of the brain and/or spinal cord may be helpful to identify typical patterns of involvement or to rule out other pathologies when clinical findings suggest CNS involvement.
- Neuropathy is the most common neurological complication of human immunodeficiency virus (HIV) infection and, in its most common form, is treated with symptom management and anti-viral therapy. However, other acquired immunodeficiency syndrome (AIDS)-related neurological disorders may be difficult to clinically differentiate from common HIV polyneuropathy and may require more aggressive treatment. Accurate diagnosis of AIDS-related cytomegalovirus (CMV) polyradiculopathy, HIV vasculitis, or AIDS-related motor neuron disease is required for appropriate treatment. MRI imaging of the spinal cord or nerve roots may assist with diagnosis when medically necessary.
- Chronic acquired demyelinating polyneuropathies, including chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN), are diagnosed by clinical history and results of electromyography and nerve conduction studies. If the diagnosis remains uncertain after these studies, neuroimaging may help establish the diagnosis. Evidence of lumbar nerve root involvement on MRI Lumbar Spine is supportive of a CIDP diagnosis. T2-weighted signal change on MRI of the brachial plexus is often present in MMN individuals.
- Polyneuropathy, organomegaly, endocrinopathy, M-protein, skin changes (POEMS) syndrome is a disorder affecting multiple organ systems which occurs in the setting of a plasma cell disorder. Diagnosis is based on electrodiagnostic confirmation of polyneuropathy and work-up of the underlying oncologic condition.

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- Electrodiagnostic testing can provide valuable findings in the investigation of some paraneoplastic polyneuropathies; however, identification of the underlying malignancy and appropriate oncological management are key to management.

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Brachial Plexus (PN-4)

Guideline

Brachial Plexus (PN-4.1)
References (PN-4)

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Brachial Plexus (PN-4.1)

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- EMG/NCV examination is required prior to advanced imaging except in cases of malignant infiltration or radiation plexitis as detailed below.

Brachial Plexus Imaging		
Indication	**Imaging**	**Notes**
Malignant infiltration (EMG not required)	Any **ONE** of the following: • MRI Upper Extremity Other Than Joint without contrast (CPT® 73218) • MRI Upper Extremity Other Than Joint without and with contrast (CPT® 73220) • MRI Chest without contrast (CPT® 71550) • MRI Chest without and with contrast (CPT® 71552) • MRI Neck without contrast (CPT® 70540) • MRI Neck without and with contrast (CPT® 70543)	
Radiation plexitis to rule out malignant infiltration (EMG not required)		
Neurogenic thoracic outlet Syndrome (TOS)		
Preoperative work-up requiring evaluation of the brachial plexus		
Brachial plexitis (Parsonage-Turner syndrome or painful brachial amyotrophy)	• Any **ONE** of the above studies **AND** • If there is concern for radiculopathy in addition to plexopathy, MRI Cervical Spine without contrast (CPT® 72141)	• For concern for cervical radiculopathy, see **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3)** • For details of brachial plexitis (Parsonage-Turner syndrome), see **Background and Supporting Information**.
Traumatic injury		

- MRI Chest and Neck are inherently bilateral, whereas MRI Upper Extremity is unilateral.
- If MRI is not available or is contraindicated, CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy

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and injury. In this circumstance, when the above criteria are met, only **ONE** of the following studies is medically necessary:
  - **CT Neck Soft Tissue**: CT Neck without contrast (CPT® 70490), **or** CT Neck with contrast (CPT® 70491), **or** CT Neck without and with contrast (CPT® 70492)
  - **CT Upper Extremity**: CT Upper Extremity without contrast (CPT® 73200), **or** CT Upper Extremity with contrast (CPT® 73201), **or** CT Upper Extremity without and with contrast (CPT® 73202)
  - **CT Chest**: CT Chest without contrast (CPT® 71250), **or** CT Chest with contrast (CPT® 71260), **or** CT Chest without and with contrast (CPT® 71270)
- MRI should be performed prior to consideration of PET imaging.
  - For PET imaging, see **PET Imaging in Oncology (ONC-1.4)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Brachial plexitis (Parsonage-Turner syndrome or painful brachial amyotrophy) is a self-limited syndrome characterized by initial shoulder region pain followed by weakness of specific muscles in a pattern which does not conform to involvement of a single root or distal peripheral nerve.

Evidence Discussion (PN-4.1)

- MRI is the imaging study of choice to evaluate the brachial plexus due to superior soft-tissue contrast and good spatial resolution, providing detailed definition of intraneural anatomy as well as localizing pathologic lesions in conditions in which electrodiagnostic and physical findings are nonspecific. A variety of findings may be seen within the brachial plexus on MRI, including increased T2 signal intensity, focal or diffuse enhancement, or enlargement or edema of nerve segments. Furthermore, signal abnormalities or atrophy in muscles supplied by the brachial plexus can help support a plexopathy. MRI is more sensitive than CT at identifying subtle infiltrative lesions regions or areas of enhancement.
- Regarding fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT, there is no relevant literature to support the use of FDG-PET/CT in the evaluation of traumatic or nontraumatic brachial plexopathy in the absence of a known malignancy.

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Lumbar and Lumbosacral Plexus (PN-5)

Guideline

Lumbar and Lumbosacral Plexus (PN-5.1)
References (PN-5)

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Lumbar and Lumbosacral Plexus (PN-5.1)

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- EMG/NCV examination is required prior to advanced imaging.
  - EMG/NCV is **NOT** required if there is concern for malignant infiltration.
- For suspected lumbar and/or lumbosacral plexopathy, **ONE** of the following is medically necessary:
  - MRI Pelvis without contrast (CPT® 72195) with fat suppression imaging, **OR**
  - MRI Pelvis without and with contrast (CPT® 72197) with fat suppression imaging, **OR**
  - MRI Abdomen without contrast (CPT® 74181) and MRI Pelvis without contrast (CPT® 72195) with fat suppression imaging, **OR**
  - MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) with fat suppression imaging
- If suspected lumbar and/or lumbosacral plexopathy is due to a traumatic injury, then MRI Lumbar Spine without contrast (CPT® 72148) is **ALSO** medically necessary.
  - See **Low Back (Lumbar Spine) Trauma (SP-6.2)**
- If MRI is not available or is contraindicated, CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy and injury. In this circumstance, when requested for suspected lumbar and/or lumbosacral plexopathy, **EITHER** of the following is medically necessary:
  - CT Pelvis with contrast (CPT® 72193) **OR**
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
- For PET imaging, see **PET Imaging in Oncology (ONC-1.4)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Lumbar and lumbosacral plexopathy may be caused by any of the following:
  - Malignant infiltration
  - Radiation
  - Traumatic injury
  - Inflammation including sarcoidosis and infection
  - Toxicity, including iatrogenic during delivery (obstetric) or related to nerve blocks (e.g., Botox®)
  - Metabolic, including etiologies such as diabetes

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Evidence Discussion (PN-5.1)

- MRI is the imaging study of choice to evaluate the lumbosacral plexus due to superior soft-tissue contrast and good spatial resolution, providing detailed definition of intraneural anatomy as well as localizing pathologic lesions in conditions in which electrodiagnostic and physical findings are nonspecific. Abnormal MRI findings in lumbosacral plexopathies include increased T2 signal intensity, focal or diffuse enhancement, or enlargement or edema of nerve segments. MRI is more sensitive than CT at identifying subtle infiltrative lesions, although CT may be useful to assess for psoas hematoma.
- Regarding fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT, there is no relevant literature to support the use of FDG-PET/CT in the evaluation of traumatic or nontraumatic lumbosacral plexopathy in the absence of a known malignancy.

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Muscle Disorders (PN-6)

Guideline

Muscle Disorders (PN-6)

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Muscle Disorders (PN-6)

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- See **Neuromuscular Junction Disorders (PN-8.4)**
- See **Muscle Disease (PN-8.5)**
- See **Gaucher Disease (Storage Disorders) (PN-8.6)**

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Magnetic Resonance Neurography (MRN) (PN-7)

Guideline

Magnetic Resonance Neurography (MRN) (PN-7.1)
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Magnetic Resonance Neurography (MRN) (PN-7.1)

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- MRN is supported when **ALL** of the following criteria are met:
  - The study is to evaluate a traumatic or compressive focal neuropathy **or** a brachial plexus injury.
  - The study is requested by a neurosurgeon, orthopedic surgeon, neurologist, or podiatrist after an in-person clinical evaluation **AND** when surgery is being considered.
  - EMG/NCV has been performed and results provided.
  - The diagnosis remains unclear following prior imaging of the region with x-ray, ultrasound, or conventional imaging (CT or MRI).
    - For conventional imaging criteria, see **Focal Neuropathy (PN-2.1)** and **Brachial Plexus (PN-4.1)**.
- MRN is reported as **ONE** of the following:
  - Unlisted MRI procedure code (CPT® 76498) **OR**
  - MRI extremity with **ONE** of the following codes:
    - MRI Upper Extremity, Other Than Joint, without contrast (CPT® 73218)
    - MRI Upper Extremity, Other Than Joint, without and with contrast (CPT® 73220)
    - MRI Lower Extremity, Other Than Joint, without contrast (CPT® 73718)
    - MRI Lower Extremity, Other Than Joint, without and with contrast (CPT® 73720)
- MRN for **ANY** other indication is considered **NOT medically necessary** at this time, including for assessment of lumbosacral plexopathy, neuromuscular disease, and polyneuropathy.

Background and Supporting Information

Magnetic resonance neurography utilizes standard MRI equipment with sequences and technology that allow for optimized viewing of the peripheral nerve. MRN creates greater contrast between the nerve and other surrounding soft tissue to allow a detailed view of the nerve tissue and layers. This allows for more accurate diagnosis of the location and degree of nerve injury.

Evidence Discussion (PN-7.1)

- Magnetic resonance neurography (MRN) offers advantages over standard MRI imaging by utilizing sequences and technology that optimize viewing of the peripheral nerve. MRN presents no increased risk to safety over standard MRI.[1] MRN is a non-

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invasive, accurate, reliable method of demonstrating normal and abnormal nerve and assessing regional muscle denervation with good surgical correlation to findings.

- Efficacy and reliability of MRN have been clinically validated in the diagnosis and localization of traumatic and compressive focal neuropathies and brachial plexus injuries for the purpose of surgical consideration. A clinical study assessing the impact of MRN data on surgical planning noted that review of MRN altered the suspected nerve involvement in 23% and changed the nerve injury grade in 27% of individuals studied. Surgeons reported MRN altered their determination of the need for surgery in 63%, timing of surgery in 41%, length of skin incision in 27%, and time in the operating room in 30% of cases reviewed. This data suggests that MRN may improve the selection of candidates for surgical repair of these lesions and may narrow the focus of surgery.
- There is insufficient literature to support the role of MRN for evaluation of other pathologies, including, but not limited to, lumbosacral plexopathy, neuromuscular disease, and polyneuropathy. Thus, MRN is considered not medically necessary at this time for indications other than traumatic and compressive focal neuropathies and brachial plexus injuries.

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Neuromuscular Disorders (PN-8)

Guideline

Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)
Spinal Muscular Atrophy (PN-8.2)
Fasciculations (PN-8.3)
Neuromuscular Junction Disorders (PN-8.4)
Muscle Diseases (PN-8.5)
Gaucher Disease (Storage Disorders) (PN-8.6)
References (PN-8)

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Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)

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- A neurological examination is **NOT** required for an individual with established diagnosis of motor neuron disease/ALS **or** when diagnosis is suspected by a neurologist, geneticist, or a physical medicine and rehabilitation (PM&R) specialist.
- For initial evaluation of suspected motor neuron disease/ALS, **ANY** of the following are medically necessary:
  - **Brain**: MRI Brain without contrast (CPT® 70551) **or** MRI Brain without and with contrast (CPT® 70553), **AND/OR**
  - **Cervical Spine**: MRI Cervical Spine without contrast (CPT® 72141) **or** MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR**
  - **Thoracic Spine**: MRI Thoracic Spine without contrast (CPT® 72146) **or** MRI Thoracic Spine without and with contrast (CPT® 72157), **AND/OR**
  - **Lumbar Spine**: MRI Lumbar Spine without contrast (CPT® 72148) **or** MRI Lumbar Spine without and with contrast (CPT® 72158)
- Repeat imaging can be evaluated based on the appropriate **Spine Imaging Guidelines.**

Background and Supporting Information

- Evidence of lower motor neuron dysfunction in a muscle may include clinical examination of muscle weakness/wasting or EMG abnormalities to meet the criteria for the diagnosis of ALS.
- Motor neuron diseases (also known as anterior horn cell diseases) are heterogeneous and encompass either upper motor neurons, or lower motor neurons, or both. Upper motor neurons begin in the cerebral cortex and descend into the brainstem (corticobulbar), or spinal cord, where there is a connection to the lower motor neuron that exits the central nervous system and reaches out to the muscle.
  - The various types can be divided into the areas so affected:
    - Amyotrophic lateral sclerosis (Lou Gehrig's disease) – both upper and lower motor neurons
    - Primary lateral sclerosis – upper motor neurons
    - Progressive muscular atrophy – lower motor neurons
    - Progressive bulbar palsy – rare and limited to bulbar muscles (muscles innervated by the cranial nerves – dysarthria and dysphagia)
  - Other rare conditions:

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  - Monomelic amyotrophy (Hirayama disease)
  - Spinal bulbar muscular atrophy (Kennedy disease)
- Signs of lower motor neuron pathology include weakness, fasciculations, atrophy, decreased muscle tone, decreased reflexes, and a plantar extensor response (Babinski sign).
- Signs of upper motor neuron pathology include weakness, increased muscle tone, increased reflexes, and a plantar flexor response.

Evidence Discussion (PN-8.1)

MRI of the Brain and/or Spine is medically necessary to evaluate for amyotrophic lateral sclerosis (ALS)-associated changes as well as evaluation for disorders that may mimic ALS.

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Spinal Muscular Atrophy (PN-8.2)

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- Molecular genetic testing is the standard tool for diagnosis for the early consideration in any infant with weakness or hypotonia.
  - MRI is **NOT** supported for diagnosis in children unless other diseases are being considered. See **Spinal Muscular Atrophy (PEDPN-5.1)**.
- In individuals with adult-onset disease, the differential includes later-onset motor neuron disorders, such as ALS.
  - For these conditions, advanced imaging is medically necessary when upper and lower motor neuron findings are present. For imaging, see **Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)**.

Evidence Discussion (PN-8.2)

- Spinal muscular atrophy (SMA) is a genetic/hereditary disorder. Molecular genetic testing is the standard tool for diagnosis of SMA. MRI is NOT medically necessary for diagnosis of SMA unless other diseases are being considered.

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Fasciculations (PN-8.3)

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Fasciculations are involuntary, irregular movements of muscle caused by activation of a single motor unit that may be secondary to benign or non-benign etiologies.

- **ALL** of the following evaluations are required prior to advanced imaging:
  - **History and physical exam** should include documentation of the following: time course of symptoms, areas of involvement, weakness, and any associated symptoms such as pain, sensory loss, or bowel or bladder dysfunction.
  - **EMG/NCV evaluation**
  - **Laboratory evaluation** (e.g., complete blood count; comprehensive metabolic panel; serum calcium; thyroid function testing; vitamin B12 level; sed rate; ANA; rheumatoid factor; serum protein electrophoresis with immunofixation; Lyme testing; HIV testing; testing for heavy metals; etc.)

  In the setting of clinical concern for radiculopathy, neuromuscular disorders, or muscle disorders, see the following imaging guidelines:
  - **Neuromuscular Junction Disorders (PN-8.4)**
  - **Muscle Diseases (PN-8.5)**
  - **Neck (Cervical Spine) Pain without and with Neurological Features (Including Stenosis) (SP-3.1)**
  - **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)**
- In the presence of upper motor neuron signs (e.g., increased tone; hyperreflexia; presence of Babinski or Hoffman signs) when there is concern for motor neuron disease, including amyotrophic lateral sclerosis (ALS), **ANY** of the following CNS studies are medically necessary:
  - **Brain**: MRI Brain without contrast (CPT® 70551) **or** MRI Brain without and with contrast (CPT® 70553), **AND/OR**
  - **Cervical Spine**: MRI Cervical Spine without contrast (CPT® 72141) **or** MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR**
  - **Thoracic Spine**: MRI Thoracic Spine without contrast (CPT® 72146) **or** MRI Thoracic Spine without and with contrast (CPT® 72157)
  - See **Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)**
- **Lumbar Spine**: Lumbar spine imaging is **NOT** medically necessary unless there is sphincter involvement **or** there is a need to rule out lower motor neuron etiologies in the lower extremities (e.g., lumbar radiculopathy). See the following Spine Imaging Guidelines:

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- **Red Flag Indications (SP-1.2)**
- **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)**

Evidence Discussion (PN-8.3)

- Fasciculations in isolation are usually benign, especially when they occur repetitively for seconds at a single site and in a single muscle. Fasciculations are more likely to be pathologic if they occur simultaneously in multiple muscles or if they are associated with objective weakness, atrophy, or hyperreflexia.
- Although fasciculations are characteristic of motor neuron disease/amyotrophic lateral sclerosis (MND/ALS) and may occur in other neurological conditions, they are also a very common occurrence in the general population, being noticed by about 70% of normal healthy individuals.
- EMG/NCV evaluation may help differentiate individuals with benign fasciculations from those who warrant further investigation.
- Appropriate laboratory evaluation and imaging would depend on the suspected etiology.

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Neuromuscular Junction Disorders (PN-8.4)

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Myasthenia Gravis (MG)

- For imaging requests related to ptosis and ocular movements associated with MG, see **Eye Disorders and Visual Loss (HD-32.1)**
- After an established diagnosis of MG or when MG is suspected by a neurologist, rheumatologist, or ophthalmologist, ONE of the following is medically necessary to assess for MG-related thymic disease:
  - CT Chest with contrast (CPT® 71260), **OR**
  - CT Chest without contrast (CPT® 71250), **OR**
  - MRI Chest without and with contrast (CPT® 71552), **OR**
  - MRI Chest without contrast (CPT® 71550)
- Repeat of **ANY ONE** of the above imaging studies is medically necessary if the initial imaging study was negative for **ANY** of the following scenarios:
  - Symptoms of chest mass
  - Rising anti-striated muscle antibody titers
  - Need for pre-operative evaluation (clinical presentation, electro-diagnostic studies, and antibody titers)

Lambert–Eaton Myasthenic Syndrome (LEMS)

Lambert–Eaton myasthenic syndrome (LEMS) is associated with malignancies, especially small cell lung cancer.

- For a suspected diagnosis, **ANY** of the following are medically necessary: CT Chest with contrast (CPT® 71260) **AND/OR** CT Abdomen and Pelvis with contrast (CPT® 74177)
  - See **Paraneoplastic Syndromes (ONC-30.3)**
- If initial CT was negative and there is persistent suspicion, **ANY** of the above imaging studies are medically necessary every 6 months for 2 years from date of initial negative imaging.
  - See **Paraneoplastic Syndromes (ONC-30.3)**

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Stiff-Person Syndrome

Stiff-person syndrome is associated with cancers such as, but not limited to, small cell lung cancer, pancreatic neuroendocrine cancer, and breast cancer.

- If stiff-person syndrome is suspected based on clinical findings, **ANY** of the following are medically necessary:
  - **Abdomen/Pelvis**: CT Abdomen and Pelvis with contrast (CPT® 74177), **or** CT Abdomen and Pelvis without and with contrast (CPT® 74178), **OR** MRI Abdomen without and with contrast (CPT® 74183), **and** MRI Pelvis without and with contrast (CPT® 72197)
  - **Chest**: CT Chest with contrast (CPT® 71260), **or** CT Chest without contrast (CPT® 71250)
  - **Symptomatic Body Areas**: CT with contrast **or** MRI without and with contrast of any other symptomatic body areas
  - See **Paraneoplastic Syndromes (ONC-30.3)**

Background and Supporting Information

- Myasthenia gravis is an autoimmune disease of the neuromuscular junctions, manifested by fatigable weakness of the cranial nerves (examples - ocular: ptosis, diplopia; bulbar: dysphagia, dysarthria, dysphonia), as well as generalized limb weakness, depending on the severity of the disease. Associated antibodies: acetylcholine receptor (AChR), muscle specific kinase (MuSK).
- Lambert Eaton myasthenic syndrome (LEMS) is also an autoimmune disease affecting the neuromuscular junction presenting with ocular and bulbar symptoms and proximal limb weakness. Associated antibodies: P/Q voltage-gated calcium channel (VGCC).
- LEMS can occur as a paraneoplastic syndrome associated with malignancy (cancer-associated LEMS) or as an autoimmune phenomenon in the absence of malignancy (non-tumor LEMS). Between 50% and 60% of all LEMS cases are associated with malignancy, particularly small cell lung carcinoma (SCLC), although LEMS has been described in individuals with non–small cell and mixed-cell lung carcinomas, neuroendocrine tumors such as prostate cancer, thymoma, and lymphoproliferative disorders.
- Stiff-person syndrome is an autoimmune disease associated with muscle spasm and muscle rigidity affecting the trunk and limb muscles. Associated antibodies: Glutamic acid decarboxylase (GAD).

Evidence Discussion (PN-8.4)

- In individuals with myasthenia gravis, advanced chest imaging with CT or MRI is preferred over x-ray for the evaluation of thymic disease and for planned thymectomy.

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- Lambert-Eaton myasthenic syndrome has been associated with malignancies. Initial and repeat imaging with CT of Chest and/or Abdomen and Pelvis are supported to evaluate for associated cancers, especially small cell lung cancer and neuroendocrine tumors.
- Stiff-person syndrome has been associated with malignancies. Initial and repeat imaging of the chest and/or abdomen and/or pelvis and/or any symptomatic body area with CT Chest and/or CT Abdomen and Pelvis or MRI Abdomen and/or Pelvis and/or CT or MRI of any symptomatic body area are supported to evaluate for associated cancers, such as small cell lung cancer, pancreatic neuroendocrine cancer, and breast cancer.

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Muscle Diseases (PN-8.5)

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- MRI may be helpful in demonstrating abnormalities in muscles that are difficult to examine or not clinically weak and can help distinguish between different types of muscle disease. MRI is also useful in determining sites for muscle biopsy.

Imaging for Muscle Disease		
Disease	**Indication**	**Imaging**
Any Known or Suspected Muscle Disease	To plan muscle biopsy	Typically an affected muscle is imaged. ◦ Upper Extremity: MRI Upper Extremity Other Than Joint without contrast (CPT® 73218) **OR** MRI Upper Extremity Other Than Joint without and with contrast (CPT® 73220)* **AND/OR** ◦ Lower Extremity: MRI Lower Extremity Other Than Joint without contrast (CPT® 73718) **OR** MRI Lower Extremity Other Than Joint without and with contrast (CPT® 73720)* * When indication column criteria are met, bilateral studies are supported if requested
Myopathy or Myositis	Additional evaluation after clinical exam, EMG/NCV, **OR** labs	
Inflammatory Muscle Diseases ◦ Dermatomyositis ◦ Polymyositis ◦ Inclusion body myositis	◦ Evaluation of differential diagnosis ◦ Selection of biopsy site ◦ Clinical concern for progression ◦ Treatment monitoring ◦ Detection of occult malignancy	

- For interstitial lung disease associated with inflammatory myopathies, see **Interstitial Lung Disease (ILD)/Diffuse Lung Disease (DLD) (CH-11.1)** in the Chest Imaging Guidelines.
- For dermatomyositis and polymyositis with concern for occult neoplasm, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.

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Evidence Discussion (PN-8.5)

- MRI is supported in known or suspected muscle disease to identify involved muscle(s). MRI may highlight muscle edema and pathology at the potential biopsy site.[2] MRI is helpful to avoid a false-negative biopsy.
- The ordering of tests should be based on the differential diagnosis arrived at by the history and examination. Laboratory evaluation is often a critical initial step to guide further investigations. Nerve conduction studies and EMG aid in making the diagnosis of neuromuscular disorders and are best conceptualized as extensions of the history and neurologic examination.
- MRI of the affected muscle is supported in the evaluation of individuals with suspected inflammatory myopathy to help identify a reversible etiology such as immune-mediated necrotizing myopathy.
- MRI of affected muscle is supported in the diagnosis and follow-up of individuals with inflammatory myopathies, such as dermatomyositis, polymyositis and inclusion body myositis to identify disease-specific patterns and evaluate response to treatment.
- Inflammatory muscle diseases, including dermatomyositis and polymyositis, have been associated with malignancy. Initial and repeat imaging with CT Chest and/or Abdomen and Pelvis are supported to evaluate for associated cancers, such as adenocarcinomas.

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Gaucher Disease (Storage Disorders) (PN-8.6)

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Imaging for Gaucher Disease
Initial Imaging: Any or all of the following:
• MRI Lumbar Spine without contrast (CPT® 72148) • Bilateral Femurs with MRI Lower Extremity, Other Than Joint, without contrast (CPT® 73718) • MRI Abdomen without contrast (CPT® 74181) • CT Chest without contrast (CPT® 71250) for individuals with new or worsening pulmonary symptoms
Every 12 months: Any or all of the following:
• To assess treatment response for individuals on enzyme replacement therapy or assess disease progression for individuals in surveillance ◦ MRI Lumbar Spine without contrast (CPT® 72148) ◦ Bilateral Femurs with MRI Lower Extremity, Other Than Joint, without contrast (CPT® 73718) ◦ MRI Abdomen without contrast (CPT® 74181) ◦ CT Chest without contrast (CPT® 71250) for individuals with documented pulmonary involvement
New or worsening pulmonary symptoms
• CT Chest without contrast (CPT® 71250)
Acute bone pain
• An initial X-ray should be obtained ◦ MRI of affected areas with and without contrast if x-ray is non-diagnostic or indicates the need for further imaging, such as equivocal for osteonecrosis, infection, or malignancy

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- PET/CT imaging is considered not medically necessary in the evaluation of Gaucher disease. ^{18}F-FDG does not reliably detect Gaucher disease in the marrow, and other isotopes are not yet FDA-approved for clinical use.

Background and Supporting Information

- Gaucher disease is group of autosomal recessive inborn errors of metabolism characterized by lack of the enzyme acid ß-glucuronidase with destructive ceramide storage in various tissues. Gaucher disease is a treatable disorder (enzyme replacement) in which the liver, spleen, and bone marrow/bones are the most affected organs. Diagnosis is established by decreased enzyme activity or genetic testing.
- Three major types of Gaucher disease are recognized:
  - **Type I** (non-neuropathic form or adult form): progressive hepatomegaly, splenomegaly, anemia and thrombocytopenia, and marked skeletal involvement; lungs and kidneys may also be involved, but central nervous system is spared
  - **Type II** (acute neuropathic form or infantile form): severe progressive neurological involvement and death by 2 to 4 years of age; hepatomegaly and splenomegaly are also present (usually evident by 6 months of age)
  - **Type III**: type I with neurological involvement and slowly progressive disease. Onset may be present before 2 years of age with survival to the third or fourth decade of life.
- Additionally, there is a perinatal-lethal and a cardiovascular form. The cardiovascular form involves the heart, spleen and eyes. Note that cardiopulmonary complications may be present, with varying frequency and severity, in all subtypes.
- Individuals with Gaucher disease are at risk for osteonecrosis, osteomyelitis, and bony tumors

Evidence Discussion (PN-8.6)

- Initial imaging and lifelong re-imaging is supported due to Gaucher disease's progressive, multisystem involvement.
- Due to bone involvement, including increased risk for multiple myeloma, skeletal x-rays, MRI of Lumbar Spine and MRI Bilateral Femurs are supported. Delineating the extent of disease can have a positive impact on developing appropriate treatment strategies.
- CT is the preferred study for the evaluation of lung parenchyma and is supported to evaluate for pulmonary involvement.
- MRI Abdomen is supported to evaluate for associated visceral disease, such as hepatic, splenic and biliary disease. This modality has better signal-to noise-ratio and soft tissue contrast helping to make more precise diagnosis.
- The role of PET/CT imaging in Gaucher disease is yet to be established. In the absence of malignancy, PET/CT is not considered medically necessary in the

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evaluation of Gaucher disease. Unnecessary use of this study would expose the individual to excess radiation and noncontributory imaging.

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Peripheral Nerve Sheath Tumors (PNST) (PN-9)

Guideline

Peripheral Nerve Sheath Tumors (PNST) (PN-9.1)
References (PN-9)

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Peripheral Nerve Sheath Tumors (PNST) (PN-9.1)

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PNST such as Schwannomas or neurofibromas arise from Schwann cells or other connective tissue of the nerve. They can be located anywhere in the body.

When Peripheral Nerve Sheath Tumors (PNST) is suspected, the following advanced imaging is medically necessary:	
Suspected Lesion/Indication	**Imaging**
Vestibular Schwannoma	• MRI Brain without and with contrast (CPT® 70553) See **Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)** in the Head Imaging Guidelines
Paraspinal Neurofibroma	**ANY** of the following imaging: • MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157), **AND/OR** • MRI Lumbar Spine without and with contrast (CPT® 72158)
Neurofibroma of the Limb or Torso (other than Paraspinal)	• MRI without and with contrast or without contrast of the area of interest after plain x-ray* ◦ See **Soft Tissue Mass (MS-10.1)** in the Musculoskeletal Imaging Guidelines *Plain x-ray is not required in an individual with a cancer predisposition syndrome. • See **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)** in the Pediatric and Special Populations Oncology Imaging Guidelines

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Routine follow-up imaging is NOT indicated *except in the following scenarios:*	
Suspected Lesion/Indication	**Imaging**
New symptoms or neurological findings	• MRI without and with contrast of the known body area containing PNST
Post-operatively for ANY of the following scenarios: • At the discretion of or in consultation with the surgeon; • If the tumor was not completely removed and the imaging is requested to re-establish baseline	• MRI without and with contrast of the known body area containing PNST or from which PNST was removed
Request for metastatic work-up when malignant transformation is known or suspected	**ANY** of the following imaging: • CT Chest with contrast (CPT® 71260) **AND/OR** • CT Abdomen with contrast (CPT® 74160)

- For guidelines related to known malignancies in individuals with neurofibromatosis 1 (NF1), see the appropriate imaging guideline for the specific cancer type.

Background and Supporting Information

- The role of PET imaging in peripheral nerve sheath tumors is not yet well established.
- Malignant transformation may be present in approximately 5% of peripheral nerve sheath tumors.

Evidence Discussion (PN-9.1)

- Peripheral nerve sheath tumors (PNSTs) may arise from any body region. PNSTs are susceptible to malignant transformation. Therefore, MRI of the known or suspected body region is supported for evaluation.
- MRI is the preferred imaging modality for soft tissue tumors, such as PNSTs, and is a relatively safe imaging modality since radiation exposure is not involved. The role of PET imaging in peripheral nerve sheath tumors is not yet well established. Otherwise, PET imaging in this clinical scenario would not add any clinical value and would unnecessarily expose individuals to radiation.

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Nuclear Imaging (PN-10)

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Nuclear Imaging (PN-10.1)

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- Nuclear Medicine
  - Nuclear medicine studies are **NOT** medically necessary in the evaluation of peripheral nerve disorders.

Evidence Discussion (PN-10)

- Though PET has well established roles in disorders other than peripheral neuropathies, the resolution of PET is on the order of millimeters, which limits its usefulness in evaluation of peripheral nerve disorders. As a result, PET may be most useful for nerve injury in combination with higher resolution structural imaging such as MRI.
- Fusion PET/MRI's role in peripheral nerve injuries is less defined and still largely limited to animal studies as an alternative non-invasive diagnostic modality.
- At this time, the use of PET/MRI for peripheral nerve disease is considered not medically necessary.

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Abbreviations and Glossary for the PVD Imaging Guidelines

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(See also:Cardiac Imaging Guidelines Glossary)

AAA	Abdominal aortic aneurysm
ABI	Ankle brachial index: a noninvasive, non-imaging test for arterial insufficiency – (see toe-brachial index below). This testing can also be done after exercise if resting results are normal.
Claudication or intermittent claudication	usually a painful cramping sensation of the legs with walking or severe leg fatigue
CLI	Critical Limb Ischemia
CTA	Computed tomography angiography
CTV	Computed tomography venography
DLCO	Diffusion capacity: defined as the volume of carbon monoxide transferred into the blood per minute per mmHg of carbon monoxide partial pressure
DVT	Deep venous thrombosis
ECG	Electrocardiogram
ENT	Ears, Nose, Throat
EVAR	Endovascular Aneurysm Repair
HbA1C	Hemoglobin A1C: test used to determine blood sugar control for individuals with diabetes
MRA	Magnetic resonance angiography

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MRV	Magnetic resonance venography
PAD	Peripheral artery disease
PAH	Pulmonary artery hypertension
PFT	Pulmonary function tests
PVD	Peripheral vascular disease
PSV ratio	Peak systolic velocities
SVC	Superior vena cava
TEVAR	Thoracic Endovascular Aortic Repair
TIA	Transient ischemic attack
TTE	Transthoracic echocardiogram
Toe-Brachial Index	Useful in individuals with ABI above the normal range due to non-compressible posterior tibial or dorsalis pedis arteries
V/Q Scan	Ventilation and perfusion scan

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General Guidelines (PVD-1.0)

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General Guidelines

- Prior to considering advanced imaging, a pertinent clinical evaluation or meaningful technological contact performed since the symptoms began or worsened is required describing any relevant medical treatments and a vascular history and physical that includes (when applicable):
  - Palpation of pulses
  - Evaluation of lower extremities for presence of non-healing wounds or gangrene
  - Associated skin changes such as thickened nails, absence of hair in the feet or calves, cool extremities
  - Evaluation for the presence of arterial bruits
  - Appropriate laboratory studies
  - Non-advanced imaging modalities, such as recent ABIs, venous duplex or venous valvular insufficiency study (VVI) performed after symptoms started or worsened
  - The affected limb(s), the extent of the edema (calf and/or thigh), pitting or non-pitting. With regard to venous insufficiency, presence or absence of hyperpigmentation or other skin changes, ulcerations if applicable, size of varicosities if present as well as distribution
  - Arterial examination to rule out phlegmasia alba/cerulea dolens which is compromised arterial flow secondary to extensive DVT if applicable
  - Appropriate laboratory studies, for example d-dimer, if applicable
- Arterial - ABI should be measured first:
  - If normal, then further vascular studies are generally not indicated.
  - If clinical suspicion for PAD remains high with normal ABI's, exercise ABI's (CPT® 93924) can be performed on a treadmill to elicit ischemia
  - The TBI (toe-brachial index) is used to establish the diagnosis of PAD in the setting of non-compressible arteries (ABI ≥1.40) and may also be used to assess perfusion in individuals with suspected CLI (rest pain and/or non-healing wound)
- If a prior imaging study (Ultrasound, MRA, CTA, Catheter angiogram, etc.) has been completed for a condition, a follow-up, additional, or repeat study for the same condition is generally not medically necessary unless there has been a change in the individual's condition, previous imaging showed an indeterminate finding, or condition specific sections in these guidelines support routine follow-up imaging.

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- Runoff studies (CPT® 75635 for CTA or CPT® 74185, CPT® 73725, and CPT® 73725 for MRA) image from the umbilicus to the feet
  - CTA Abdomen and lower extremities should be reported as CPT® 75635, rather than using the individual CPT® codes for the abdomen, pelvis, and legs
  - MRA Abdomen, MRA Pelvis and MRA Lower extremities should be reported as CPT® 74185, CPT® 73725, and CPT® 73725. The CPT® code for MRA Pelvis (CPT® 72198) should not be included in this circumstance.
- Venous-imaging
  - Venous duplex (CPT® 93970, CPT® 93971) of the limb is the initial imaging of choice.
  - Follow-up duplex imaging is not medically necessary to document resolution and should only be obtained for new signs/symptoms or for concerns of propagation of thrombus when the treatment plan would change (insertion of IVC filter, change of anticoagulation, etc.)

Evidence Discussion

Venous duplex should visualize the veins, with demonstration of the presence or absence of compressibility and venous flow. Venous valvular insufficiency studies visualize the veins of the lower extremity, assess for reflux (reversal of venous antegrade flow after valve closure) and measure its duration. CTV or MRV Abdomen and Pelvis images with contrast involves taking images from the diaphragm to just below the inguinal ligament after a delay of a few minutes after IV contrast is administered to optimize filling and therefore visualization of the venous vasculature.

Venous disease can be classified into three categories:

- Veno-occlusive disease
  - Types of thrombotic disease
    - Superficial venous thrombosis
    - Deep venous thrombosis
    - Iliac vein obstruction, unilateral or bilateral
    - May-Thurner syndrome
  - Signs/Symptoms of veno-occlusive disease is generally sudden onset of pain and edema in the limb.
  - Risk factors include age >40, obesity, pregnancy, prolonged immobility, post-surgery, and malignancy among others.
  - Procedures related to veno-occlusive disease include:
    - Thrombolysis
    - Thrombectomy
    - Post-iliac vein stent/angioplasty
- Venous insufficiency

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  - Types of venous insufficiency:
    - Superficial and deep venous reflux
    - Varicose veins
    - Reticular and spider veins.
  - Signs/symptoms of venous insufficiency include:
    - Chronic swelling in the leg that is relieved with elevation
    - Chronic swelling in the leg that is worse in the evenings
    - Aching or sense of heaviness in the leg
    - Hyperpigmentation of the calf particularly around the ankle
    - Itchy skin on legs and feet
    - Leather appearance of the skin of the calves
    - Skin ulcers in the calf particularly around the medial malleolus
    - Varicose veins
    - Spider veins/reticular veins/telangiectasias
  - Procedures related to the venous insufficiency include:
    - Endovenous laser ablation utilizing either chemical, laser or radio-frequency
    - Saphenous vein high ligation and stripping
    - Phlebectomy, stab or powered
    - Sclerotherapy, liquid or foam
- Venous malformations
  - Types of venous malformations include:
    - Arterio-venous malformations which can occur throughout the body
    - See **Pulmonary AVM (CH-24)** in the Chest imaging guidelines
    - See **Aneurysm and AVM (HD-12)** in the Head imaging guidelines
    - See **Pelvic Pain/Dyspareunia, Female (PV-11)** in the Pelvic imaging guidelines
    - Klippel-Trenaunay which affects primarily the lower extremity venous circulation and is characterized by varicose veins, limb size discrepancies, and port-wine stains.
  - Treatment includes:
    - Primarily embolization
    - Sclerotherapy
      - Klippel-Trenaunay: treatment can include phlebectomy and sclerotherapy of symptomatic varicose veins provided they meet the criteria for intervention.

General Information

- Risk factors for vascular disease include:
  - Diabetes

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- Cigarette smoking
- Hypertension
- Hyperlipidemia
- Age >50, with at least one risk factor, are considered “at risk” for vascular disease
- See also: **Impotence/Erectile Dysfunction (PV-17)** in the Pelvis Imaging Guidelines.

- Signs and symptoms of peripheral arterial disease
  - Claudication (Cramping pain in the legs, most notably back of the calves but can involve hips or thighs, after walking which is relieved with rest but recurs at a predictable distance)
    - Symptoms that are not consistent with claudication include
      - Generalized leg pain
      - Nocturnal cramps
      - Pain that is not easily relieved after a few minutes of rest
      - Burning pain in feet
  - Critical limb ischemia
    - Rest pain: Pain in the foot (not leg) at rest, particularly at night when the leg is elevated. Pain is relieved by dangling the leg off the bed or moving to an upright position
    - Non-healing wounds: Wounds present for >2 weeks with little to no evidence of healing
  - Erectile dysfunction can be associated with vascular disease
- Claudication and critical limb ischemia have different natural histories. Claudication generally follows a benign indolent course. 70% of individuals with claudication will have the same symptoms after five years with no progression. Critical limb ischemia, on the other hand, is associated with a high rate of limb loss (25%) and death (35%) one year after presentation
- Simultaneous venous and arterial systems evaluation are unusual but are occasionally needed
- Post-angioplasty/reconstruction: follow-up imaging is principally guided by symptoms. See also:
  - **Post Aortic Endovascular/Open Surgery Surveillance Studies (PVD-6.8)**
  - **Post-procedure Surveillance Studies (PVD-7.3)**

General Guidelines – Imaging

- Imaging Studies:
  - Carotid studies MRA Neck (CPT® 70543) or CTA Neck (CPT® 70491) capture the area from the top of the aortic arch (includes the origin of the innominate artery,

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common carotid artery, and subclavian artery, which gives off the vertebral artery) to the base of the skull.
- CTA or MRA Abdomen (CPT® 74175 or CPT® 74185) images from the diaphragm to the umbilicus or iliac crest
- CTA or MRA Chest (CPT® 71275 or CPT® 71555) images from the base of the neck to the dome of the liver
- Runoff studies (CPT® 75635 for CTA or CPT® 74185, CPT® 73725, and CPT® 73725 for MRA) image from the umbilicus to the feet
  - CTA Abdomen and lower extremities should be reported as CPT® 75635, rather than using the individual CPT® codes for the abdomen, pelvis, and legs
  - MRA Abdomen, MRA Pelvis and MRA Lower extremities should be reported as CPT® 74185, CPT® 73725, and CPT® 73725. The CPT® code for MRA Pelvis (CPT® 72198) should not be included in this circumstance

Quantification of plaque morphology

CPT® 0710T, CPT® 0711T, CPT® 0712T, CPT® 0713T

There is insufficient evidence to support the routine use of data from non-coronary computed tomography angiography (CTA) to quantify plaque morphology in noncoronary vessels. These studies are considered experimental, investigational, or unproven.[1]

Procedure Coding

Non-invasive Physiologic Studies of Extremity Arteries	CPT®
• Limited bilateral noninvasive physiologic studies of upper or lower extremity arteries. • Non-invasive physiologic studies of upper or lower extremity arteries, single level, bilateral (e.g., ankle/brachial indices, Doppler waveform analysis, volume plethysmography, transcutaneous oxygen tension measurement).	93922

[1] Guo Z, Liu Y, Xu J, et al. A deep learning model for carotid plaques detection based on CTA images: a two stepwise early-stage clinical validation study. Front Neurol. 2025;15:1480792. doi:10.3389/fneur.2024.1480792.

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Non-invasive Physiologic Studies of Extremity Arteries	CPT®
• Complete bilateral noninvasive physiologic studies of upper or lower extremity arteries, 3 or more levels. • Non-invasive physiologic studies of upper or lower extremity arteries, multiple levels or with provocative functional maneuvers, complete bilateral study (e.g., segmental blood pressure measurements, segmental Doppler waveform analysis, segmental volume plethysmography, segmental transcutaneous oxygen tension measurements, measurements with postural provocative tests, measurements with reactive hyperemia).	93923

- CPT® 93922 and CPT® 93923 can be requested and reported only once for the upper extremities and once for the lower extremities.
- CPT® 93922 and CPT® 93923 should not be ordered on the same request nor billed together for the same date of service.
- CPT® 93924 and CPT® 93922 and/or CPT® 93923 should not be ordered on the same request and should not be billed together for the same date of service.
- ABI studies performed with handheld dopplers, where there is no hard copy output for evaluation of bidirectional blood flow, are not reportable by these codes.

Non-invasive Physiologic Studies of Extremity Arteries	CPT®
Non-invasive physiologic studies of lower extremity arteries, at rest and following treadmill stress testing, complete bilateral study.	93924

Arterial Duplex – Upper and Lower Extremities	CPT®
Duplex scan of lower extremity arteries or arterial bypass grafts; complete bilateral.	93925
• A complete duplex scan of the lower extremity arteries includes examination of the full length of the common femoral, superficial femoral and popliteal arteries. • The iliac, deep femoral, and tibioperoneal arteries may also be examined.	
Duplex scan of lower extremity arteries or arterial bypass grafts; unilateral or limited study.	93926
• The limited study is reported when only one extremity is examined or when less than a full examination is performed (e.g. only one or two vessels or follow-up).	

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Arterial Duplex – Upper and Lower Extremities	CPT®
Duplex scan of upper extremity arteries or arterial bypass grafts; complete bilateral.	93930
• A complete duplex of the upper extremity arteries includes examination of the subclavian, axillary, and brachial arteries. • The radial and ulnar arteries may also be included.	
Duplex scan of upper extremity arteries or arterial bypass grafts; unilateral or limited study.	93931
• The limited study is reported when only one extremity is examined or when less than a full examination is performed (e.g. only one or two vessels or follow-up).	

Cerebrovascular Artery Studies	CPT®
Duplex scan of extracranial arteries; complete bilateral study.	93880
Duplex scan of extracranial arteries; unilateral or limited study.	93882
• This study is often referred to as a “carotid ultrasound” or “carotid duplex”. • Typically, it includes evaluation of the common, internal, and external carotid arteries.	

Transcranial Doppler Studies	CPT®
Transcranial Doppler study of the intracranial arteries; complete study	93886
Transcranial Doppler study of the intracranial arteries; limited study	93888
Transcranial Doppler vasoreactivity study	93890
Transcranial Doppler study of the intracranial arteries; emboli detection without intravenous microbubble injection	93892
Transcranial Doppler study of the intracranial arteries; emboli detection with intravenous microbubble injection	93893

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Venous Studies - Extremities	CPT®
Duplex scan of extremity veins, including responses to compression and other maneuvers; complete bilateral study.	93970
Duplex scan of extremity veins, including responses to compression and other maneuvers; unilateral or limited study.	93971
• These codes are used to report studies of lower or upper extremity veins. • A complete bilateral study of the lower extremity veins includes examination of the common femoral, proximal deep femoral, great saphenous and popliteal veins. Calf veins may also be included. • A complete bilateral study of upper extremity veins includes examination of the subclavian, jugular, axillary, brachial, basilica, and cephalic veins. Forearm veins may also be included.	

Visceral Vascular Studies	CPT®
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study.	93975
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study	93976
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; complete study	93978
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; unilateral or limited study	93979

Duplex for Hemodialysis Access	CPT®
Duplex scan of hemodialysis access (including arterial inflow, body of access and venous outflow).	93990
Duplex scan of arterial inflow and venous outflow for preoperative vessel assessment prior to creation of hemodialysis access; complete bilateral study	93985

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Duplex for Hemodialysis Access	CPT®
Duplex scan of arterial inflow and venous outflow for preoperative vessel assessment prior to creation of hemodialysis access; complete unilateral study	93986

Nuclear Medicine Imaging Indications

- Nuclear medicine studies are rarely used in the evaluation of peripheral vascular disorders but are considered medically necessary in the following circumstances:
  - Lymphoscintigraphy (CPT® 78195) for evaluation of lower extremity lymphedema when recent Doppler ultrasound of the lower extremity and abdomen are negative for valvular insufficiency.
  - Indium 111 (^{111}In)–labeled white blood cell (WBC) or Gallium-67 citrate studies (CPT® 78800, CPT® 78801, CPT® 78802, or CPT® 78803) for evaluation of **any** of the following:
    - Mycotic aneurysms.
    - Vascular graft infection.
    - Infection of central venous catheter or other indwelling device.
  - PET/CT (CPT® 78815) for clinical suspicion of aortic infection (graft or native aorta) when **both** of the following apply:
    - CT-angiogram is equivocal/indeterminate AND
    - Indium-111 or Gallium-67 studies have not been performed **and** are not available or not technically feasible.

Non-indications

Evidence does not support routine use of these studies for peripheral vascular evaluation. MRA, CTA, and duplex ultrasonography provide superior diagnostic value more likely to impact clinical management. Therefore, these studies are considered not medically necessary for any peripheral vascular imaging indication:

- Venous thrombosis imaging (CPT® 78456, CPT® 78457, and CPT® 75458)
- Vascular flow imaging (CPT® 78445)

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants

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of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Genetic Predisposition to Arterial Disease

Guideline

Screening for Peripheral Artery /Aneurysmal Disease (PVD-2)
References

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Screening for Peripheral Artery / Aneurysmal Disease (PVD-2)

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Asymptomatic Screening (PVD-2.1)

- Routine screening of asymptomatic individuals for PAD is not medically necessary. Those with CVD risk factors should be placed on best medical management and should be questioned on symptoms of PAD at annual physicals.
- Currently, there is no evidence to demonstrate that screening all individuals with PAD for asymptomatic atherosclerosis in other arterial beds improves clinical outcome.
- Resting ABI's may be medically necessary in individuals with abnormal pulse exams.
- Screening in the presence of aortic aneurysms:
  - In an individual with a known TAA, screening for AAA is medically necessary with an abdominal duplex. See **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)**
  - In an individual with a known AAA, screening for TAA is not supported by sufficient evidence and is not considered medically necessary.

Evidence Discussion

Screening for Suspected Peripheral Artery Disease/Aneurysmal Disease

Generally, routine screening for peripheral arterial disease in asymptomatic patients is not cost-effective and has not been shown to improve patient outcomes. There are some familial and genetic conditions that may predispose individuals to aneurysmal disease and dissection. In these cases, surveillance imaging has been shown to improve early detection and intervention when indicated. Conditions that may have an elevated risk for vascular disease include:

- Familial Aneurysm Syndromes
- Fibromuscular Dysplasia
- Spontaneous Coronary Artery Dissection (SCAD)
- Ehlers-Danlos
- Marfan
- Loeys-Dietz

In the case of aneurysms detected in patients with SCAD, Marfan, Loeys-Dietz and Ehlers-Danlos Type IV syndromes, a more frequent surveillance pattern along with additional anatomic region imaging may be indicated as these syndromes

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demonstrate a higher incidence of aneurysm development and degeneration. Intervention recommendations follow general guidelines for thoracic aortic, abdominal aortic and visceral artery aneurysms once detected.

While duplex imaging remains an effective modality for abdominal aortic surveillance, these conditions often involve the thoracic aorta as well as cerebrovascular and visceral vessel abnormalities that may be technically limited with this approach. Due to the high incidence of aneurysm development in multiple anatomic locations, CT/MR imaging is recommended for surveillance in this population for cases with indeterminate ultrasound imaging.

There is an association between bicuspid aortic valve and thoracic aneurysm development. Individuals diagnosed with this condition should undergo screening and follow standard surveillance patterns using CT/MR of the chest as well as echocardiography should a thoracic aortic aneurysm be detected. The addition of cardiac-specific CT/MR has not shown benefit in these cases as the pathology is usually noted within the aorta.

Multisystemic Smooth Muscle Syndrome [MSMS], Smooth Muscle Dysfunction Syndrome [SMDS] and ACATA2 mutations have a high incidence of aneurysm development early in life and should undergo screening and routine surveillance after genetic confirmation. To minimize radiation exposure in this pediatric population, MR and ultrasound imaging are recommended when possible.

Screening for Vascular related genetic connective tissue Disorders (PVD-2.2)

Vascular related genetic connective tissue Disorders include:

- Familial Aneurysm Syndromes
- Fibromuscular Dysplasia
- Spontaneous Coronary Artery Dissection (SCAD)
- Ehlers-Danlos
- Marfan
- Loeys-Dietz

Table of Thoracic Aorta Imaging Options

Description	CPT®
CT Chest without contrast	71250
CT Chest with contrast	71260
CTA Chest	71275

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Description	CPT®
MRA Chest	71555
TTE	93306

Screening and initial diagnosis

- Screening for Familial Syndromes is medically necessary in individuals with a positive family history (1st degree relative with dissection/TAA) but no known genetic syndrome/mutation, otherwise known as Suspected Familial Aneurysm Syndrome as follows:
  - ECHO (CPT® 93306, CPT® 93307, or CPT® 93308) and chest x-ray for all First-degree relatives (parents, siblings, children) of individuals with TAA and/or dissection.
  - Any imaging listed in the **Table of Thoracic Aorta Imaging Options** is medically necessary if these studies identify a TAA or are equivocal or do not visualize the ascending or descending aorta adequately.
  - Studies can be repeated at 2 year intervals if prior results are negative
- Initial imaging for individuals with documented SCAD/fibromuscular dysplasia/Marfan/Loeys-Dietz/Ehlers-Danlos type IV is medically necessary as follows:
  - On initial diagnosis of Ehlers-Danlos, Loeys-Dietz, Marfan, SCAD, or suspicion of fibromuscular dysplasia, full vascular imaging from head to pelvis with:
    - CTA or MRA Head (CPT® 70496 or CPT® 70546 or CPT® 70545)
    - CTA or MRA Neck (CPT® 70498 or CPT® 70548 or CPT® 70549)
    - CTA or MRA Chest or CT Chest with contrast
    - CTA Abdomen/Pelvis or MRA Abdomen/Pelvis (CPT® 74174) or (CPT® 74185 and CPT® 72198)
  - If there are no identified aneurysms or dissections, repeat imaging can be obtained at two-year intervals

Surveillance

Surveillance imaging

- If an aneurysm is identified in individuals with fibromuscular dysplasia, then the aneurysm can be surveilled per the typical time-frame as described in **Thoracic Aortic Aneurysm (TAA) (PVD-6.2)**, **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)**, **Iliac Artery Aneurysm (IAA) (PVD-6.4)**, and **Visceral Artery Aneurysm (PVD-6.5)**.
- Follow-up of aneurysms in individuals with documented SCAD/Marfan/Loeys-Dietz/Ehlers-Danlos type IV
  - Imaging is medically necessary every 6 months once an aneurysm has been identified until a decision has been made to repair.
    - Intracranial aneurysm – CTA or MRA Head (CPT® 70496 or 70544)

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- Aneurysm of a cervical artery – Carotid duplex or CTA or MRA neck if unable to fully visualize with carotid duplex
- Thoracic aorta – CTA Chest (CPT® 71275) or CT Chest with (CPT® 71260) or without (CPT® 71250), MRA chest (CPT® 71555)
- Abdominal aneurysm – Abdominal duplex (CPT® 93975/93976/93978/93979/76770/76775)
- Visceral aneurysm – These can be difficult to visualize on duplex. If not visible on duplex, can obtain a CTA or MRA Abdomen and Pelvis.

Background and Supporting Information

Fibromuscular dysplasia and spontaneous coronary artery dissection is diagnosed radiographically. Loeys-Dietz, Marfan, Ehlers-Danlos type IV are diagnosed with genetic testing.

Evidence Discussion

Screening for Vascular Related Genetic Connective Tissue Disorders

Generally, routine screening for peripheral arterial disease in asymptomatic patients is not cost-effective and has not been shown to improve patient outcomes. There are some familial and genetic conditions that may predispose individuals to aneurysmal disease and dissection. In these cases, surveillance imaging has been shown to improve early detection and intervention when indicated. Conditions that may have an elevated risk for vascular disease include:

- Familial Aneurysm Syndromes
- Fibromuscular Dysplasia
- Spontaneous Coronary Artery Dissection (SCAD)
- Ehlers-Danlos
- Marfan
- Loeys-Dietz

In the case of aneurysms detected in patients with SCAD, Marfan, Loeys-Dietz and Ehlers-Danlos Type IV syndromes, a more frequent surveillance pattern along with additional anatomic region imaging may be indicated as these syndromes demonstrate a higher incidence of aneurysm development and degeneration.[4,5] Intervention recommendations follow general guidelines for thoracic aortic, abdominal aortic and visceral artery aneurysms once detected.

While duplex imaging remains an effective modality for abdominal aortic surveillance, these conditions often involve the thoracic aorta as well as cerebrovascular and visceral vessel abnormalities that may be technically limited with this approach. Due to the high incidence of aneurysm development in multiple anatomic locations,

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CT/MR imaging is recommended for surveillance in this population for cases with indeterminate ultrasound imaging.

Screening for TAA with bicuspid aortic valves (PVD-2.3)

Table of Thoracic Aorta Imaging Options

Description	CPT®
CT Chest without contrast	71250
CT Chest with contrast	71260
CTA Chest	71275
MRA Chest	71555
TTE	93306

Indications

Screening

- Screening is medically necessary in individuals with bicuspid aortic valve:
  - Screening, any requested imaging from the **Table of Thoracic Aorta Imaging Options** and/or ECHO (CPT® 93306, CPT® 93307, or CPT® 93308).
    - There is no evidence-based data to support screening relatives of individuals with bicuspid aortic valve for TAA except with echocardiogram.
  - Follow-up per TAA Follow-up guidelines in **Thoracic Aortic Aneurysm (TAA) (PVD-6.2)**

Surveillance

If no dilatation of the aortic root or ascending thoracic aorta is found, there is no evidence-based data to support the medical necessity of continued surveillance imaging

Evidence Discussion

Screening for TAA with Bicuspid Aortic Valves

There is an association between bicuspid aortic valve and thoracic aneurysm development. Individuals diagnosed with this condition should undergo screening and follow standard surveillance patterns using CT/MR of the chest as well as echocardiography should a thoracic aortic aneurysm be detected. The addition of cardiac-specific CT/MR has not shown benefit in these cases as the pathology is usually noted within the aorta. If negative for bicuspid valve pathology, additional surveillance imaging is not been supported.

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Screening for Vascular Related Disorders in ACTA2 Mutations (PVD 2.4)

Screening for Vascular Related Disorders in Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations

Initial imaging

Upon initial genetic confirmation, all of the following studies are medically necessary:

- Transthoracic echocardiogram (TTE) (CPT® 93306)
- MRA Chest (CPT® 71555) or CTA Chest and CT Chest with contrast (CPT® 71275 and 71260)
- MRA Abdomen and Pelvis (CPT® 74185 and 72198) or CTA Abdomen and Pelvis (CPT® 74174)
- MRI Perfusion study Brain CPT® 70553
- MRA Head and Neck (CPT® 70544 or 70545 AND 70548)
- Ultrasound Upper extremity(ies) (CPT® 93930 or 93931) or CTA Upper extremity (CPT® 73206) or MRA Upper extremity (CPT® 73225)

Repeat testing

Repeat testing with any of the studies listed under initial imaging is medically necessary when there is documentation of new signs or symptoms.

Surveillance imaging

Surveillance imaging with any of the studies listed in initial imaging is medically necessary according to the following:

- Transthoracic echocardiogram repeat every 6 months
- Chest imaging can be repeated every 12 months starting at age 10
- Abdomen and pelvis imaging can be repeated every 12 months starting age 10
- Upper extremity imaging can be repeated every 12 months starting age 10
- MRI perfusion study Brain see **Dysfunction Syndrome (SMDS)/ACTA2 Mutations (PEDHD-12.8)**
- MRA Head and Neck see **Dysfunction Syndrome (SMDS)/ACTA2 Mutations (PEDHD-12.8)**

Background and Supporting Information

Smooth Muscle Dysfunction Syndrome presents as congenital mydriasis, a patent ductus arteriosus (PDA), pulmonary arterial hypertension (PAH) during infancy.

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Patients go on to developed aortic, peripheral arterial, and cerebrovascular disorders in childhood.

- Caused by heterozygous mutation of ACTA2. P.Arg179His
- Cases mostly due to de novo mutations, so imaging screening based on family history without genetic confirmation is not supported.
- Because radiation is a known risk factor for moyamoya disease. MRI/MRA Head is recommended instead of Computed Tomography (CT)/CTA

Evidence Discussion

Screening for Vascular Related Disorders in Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations

Multisystemic Smooth Muscle Syndrome [MSMS], Smooth Muscle Dysfunction Syndrome [SMDS] and ACATA2 mutations have a high incidence of aneurysm development early in life and should undergo screening and routine surveillance after genetic confirmation. To minimize radiation exposure in this pediatric population, MR and ultrasound imaging are recommended when possible.

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Cerebrovascular Imaging

Guideline

Cerebrovascular and Carotid Disease - Initial Imaging (PVD-3.1)
Surveillance Imaging with NO History of Carotid Surgery or Intervention (PVD-3.2)
Surveillance Imaging WITH History of Carotid Surgery or Intervention (PVD-3.3)
References

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Cerebrovascular and Carotid Disease - Initial Imaging (PVD-3.1)

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- Duplex ultrasound (CPT® 93880 bilateral or CPT® 93882 unilateral) is medically necessary to evaluate possible carotid artery disease, prior to considering advanced imaging, when ANY of the following apply:
  - Known or suspected retinal arterial emboli or Hollenhorst plaque
  - Pulsatile neck masses
  - Carotid or cervical bruit
  - Abnormal findings on physical exam of the carotid arteries (e.g., absent carotid pulses)
  - Preoperative evaluation of individuals with evidence of severe diffuse atherosclerosis, scheduled for major cardiovascular surgical procedures
  - Preoperative evaluation of individuals prior to elective cardiovascular surgery in individuals older than 65 years of age and in those with peripheral artery disease, history of cigarette smoking, history of stroke or TIA, or carotid bruit
  - Suspected Subclavian Steal Syndrome for additional imaging see **Upper Extremity PVD – Imaging (PVD-4.1)**
  - Blunt neck trauma in the absence of focal neurologic symptoms
  - Neurologic complaints after chiropractic neck manipulation
  - Vasculitis potentially involving carotid arteries, such as Takayasu's arteritis and fibromuscular dysplasia (FMD)
  - Remote history of stroke or TIA (Greater than one month).
- CTA or MRA Neck is medically necessary to evaluate suspected internal carotid artery dissection, in individuals with **any** of the following mechanisms of injury or risk factors for arterial dissection:
  - Chiropractic manipulation of neck
  - Whiplash injury
  - Fibromuscular dysplasia/Marfan's
  - Stroke in the young (age ≤50)
- CTA or MRA Neck is medically necessary to evaluate suspected vertebrobasilar pathology:
  - Symptoms include:
    - Vertigo associated with nausea and vomiting
    - Diplopia
    - Loss of vision in one or both eyes

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        - Dysarthria
        - Bifacial numbness
        - Bilateral extremity weakness and/or numbness
        - Acute changes in mental status
        - Loss of consciousness
        - Ataxia
    - MRA or CTA of **both** Neck and Head are required to visualize the entire vertebral-basilar system for evaluation of posterior circulation disease.
    - Surveillance imaging, post-stenting or known vertebrobasilar disease, interval determined by Vascular Specialist, Neurologist, or Neurosurgeon or any provider in consultation with a vascular specialist, neurologist, or neurosurgeon for ANY of the following:
        - Asymptomatic
        - Unchanged symptoms
        - New or worsening symptoms
- After Intracranial Hemorrhage:
    - Initial Imaging see **Head Trauma (HD-13.1)** in the Head Imaging Guidelines
    - Surveillance Imaging
        - Interval determined by neurosurgeon or neurologist or any provider in consultation with a neurologist or neurosurgeon.
- Carotid ultrasound screening in asymptomatic individuals due only to risk factors is **not** indicated.
- Repeat imaging of the cervical vessels (regardless of when the previous carotid imaging was performed) is medically necessary for new signs and symptoms consistent with carotid artery disease (e.g., TIA, amaurosis fugax, change in nature of a carotid bruit) using one of the following:
    - Duplex ultrasound (CPT® 93880 bilateral study or CPT® 93882 unilateral study)
    - MRA Neck with contrast (CPT® 70548) or without and with contrast (CPT® 70549)
    - CTA Neck (CPT® 70498)

Evidence Discussion

Cerebrovascular and Carotid Disease- Initial Imaging

Indications for carotid artery imaging are suspicion for carotid stenosis, aneurysm, dissection or vasculitis. The signs and symptoms generally accepted for carotid imaging are listed in the guideline.

Standard first line imaging is the Duplex ultrasound (DU). This study obtains gray-scale pictures, as well as velocity and direction of blood flow in the vessels. The combination of B mode and Doppler techniques allows for detection of all of the above mentioned

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pathologies. DU has limitations in evaluation of the vertebral artery origins as well as extent of carotid dissection. Therefore, if there is concern for either vertebrobasilar insufficiency or carotid dissection, CT angiography (CTA) or MR angiography (MRA) is supported.

However, DU has become the first-line imaging modality for identifying patients with internal carotid artery stenosis. In part, this is because consensus ultrasound criteria have been developed to standardize carotid ultrasound examinations and categorize carotid artery stenosis severity. The rationale for use of DU is its low cost, availability, and high sensitivity and specificity. It avoids exposure to radiation and intravenous contrast agents as well.

CTA and MRA have more limited use for screening due to the risks associated with them. CTA risks include intravenous contrast and radiation exposure. Contrast complications include allergy and contrast induced nephropathy. MRA risk is related to use of gadolinium contrast, which confers the risk of nephrogenic systemic fibrosis in patients with renal insufficiency. MRA also is contraindicated in patients with metallic implants. Additionally, both CTA and MRA are not appropriate for screening purposes, due to their considerable costs.

It is well established that for carotid stenosis 70-99% in an asymptomatic patient, or 50-99% stenosis in a symptomatic patient, there is a role for carotid intervention for stroke prevention. Once duplex identifies this degree of stenosis, CTA/MRA is medically necessary for pre-procedure planning. Additionally, CTA/MRA is medically necessary for patients undergoing evaluation for carotid artery stenting (CAS) as delineation of relevant anatomy is necessary for procedural success.

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Surveillance Imaging with NO History of Carotid Surgery or Intervention (PVD-3.2)

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- Surveillance imaging is medically necessary once a year for individuals with fibromuscular dysplasia of the extracranial internal carotid arteries.
- Reporting standards for carotid stenosis varies widely. The most commonly used criteria, however, is noted in the chart below published by the Society of Radiology in 2003

Primary parameters			Additional Parameters	
% Stenosis	ICA PSV (cm/sec)	Plaque estimate (%)	ICA/CCA PSV ratio	ICA/EDV (cm/sec)
Normal	< 125	None	< 2.0	< 40
< 50	< 125	< 50	< 2.0	< 40
50-69	125-230	≥ 50	2.0-4.0	40-100
≥ 70 but less than near occlusion	> 230	> 50	> 4.0	> 100
Near occlusion	High, low, or undetectable	Visible	Variable	Variable
Total occlusion	undetectable	Visible- no detectable lesion	Not applicable	Not applicable

- If normal study, no routine follow-up imaging is medically necessary
- If <50% internal carotid stenosis
  - Duplex ultrasound (CPT® 93880 bilateral or CPT® 93882 unilateral)is medically necessary every **two** years
- Between 50% and 70% internal carotid stenosis

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- Duplex ultrasound (CPT® 93880 bilateral or CPT® 93882 unilateral) is medically necessary annually.
- A repeat duplex (CPT® 93880 bilateral or CPT® 93882 unilateral) is medically necessary in three to six months until stability is reached when **one** of the following occurs:
  - Change in the character of the bruit
  - Duplex demonstrates rapid progression, including:
    - Doubling of peak systolic velocities in the internal carotid arteries
    - Increase of the ICA/CCA ratio
    - Heavy calcification in the internal carotid arteries
    - Thrombus in the internal carotid arteries
    - Ulcerated plaque in the internal carotid arteries
    - Echolucent plaque in the internal carotid arteries
- A one-time CTA Neck (CPT® 70498) or MRA Neck (CPT® 70548) is medically necessary to confirm degree of stenosis in individuals with ulcerated plaque or heavy calcification of the internal carotid artery seen on duplex.

- Internal carotid stenosis ≥70% or ICA/CCA ratio >4
  - Duplex ultrasound (CPT® 93880 bilateral or CPT® 93882 unilateral) or MRA Neck with contrast (CPT® 70548) or CTA Neck (CPT® 70498) is medically necessary at the following intervals:
    - Every 6 months until one of the following occurs:
      - Intervention is performed
      - Decision is made to not intervene
- MRA Neck with contrast (CPT® 70548) or CTA Neck (CPT® 70498) is medically necessary if duplex Ultrasound shows ≥70% occlusion/stenosis of the internal carotid artery or the ICA/CCA ratio is >4.0, even with a lower percentage of stenosis.
  - If carotid stent is planned
    - MRA Head (CPT® 70544, or CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) can be added

Evidence Discussion

Surveillance Imaging with NO History of Carotid Surgery or Intervention

DU is established as the primary diagnostic test for carotid surveillance imaging. Consensus statements have established the appropriate time intervals for repeat imaging. If however, there are new symptoms or physical findings, repeat duplex imaging is supported, regardless of time interval. If at any time there is >70% re-stenosis, CTA or MRA is indicated.

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Surveillance Imaging WITH History of Carotid Surgery or Intervention (PVD-3.3)

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- Duplex ultrasound (CPT® 93880 bilateral or CPT® 93882 unilateral) is medically necessary post-carotid surgery or intervention at the following intervals:
  - 1 month after procedure
  - Every 6 months for 2 years after procedure
  - Then annually
- If ≥70% residual internal carotid stenosis is seen on duplex at 1 month after procedure
  - Duplex ultrasound (CPT® 93880 bilateral or CPT® 93882 unilateral) or CTA Neck (CPT® 70498) or MRA Neck (CPT® 70548) is medically necessary at the following intervals:
    - Every 3-6 months for one year
    - Then annually or until decision is made to re-intervene.
- If ≥70% residual internal carotid stenosis is seen on duplex at any time post-procedure, then
  - CTA Neck (CPT® 70498) or MRA Neck (CPT® 70548) is medically necessary for further evaluation and at six-month intervals until decision is made to re-intervene.

Background and Supporting Information

- MRA Neck (CPT® 70548) or CTA Neck (CPT® 70498) may be medically necessary if ultrasound is technically difficult or confirmation of the degree of stenosis on ultrasound is needed because an interventional procedure is being considered

Evidence Discussion

Surveillance Imaging with WITH History of Carotid Surgery or Intervention

As described in PVD 3.1, DU is established as the primary diagnostic test for carotid surveillance imaging. Consensus statements have established the appropriate time intervals for repeat imaging. If however, there are new symptoms or physical findings, repeat duplex imaging is supported, regardless of time interval. If at any time there is >70% re-stenosis, CTA or MRA is indicated.

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Aortic Imaging

Guideline

Aortic Disorders General Information (PVD-6.1)
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Aortic Disorders General Information (PVD-6.1)

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Duplex ultrasound for visceral vascular studies	CPT®
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study.	93975
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study.	93976
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; complete study.	93978
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; unilateral or limited study.	93979
Ultrasound, abdominal aorta, real time, with image documentation, screening study for abdominal aortic aneurysm (AAA) for AAA screening	76706

- In clinical practice, CT, CTA, MRA are medically necessary to evaluate for stenosis of these vessels rather than ultrasound (Exception: Duplex ultrasound is medically necessary to rule out testicular or ovarian torsion or to evaluate an abdominal bruit or a pulsatile abdominal mass).
- Mesenteric Ischemia
  - See **Mesenteric/Colonic Ischemia (AB-6)** in the Abdomen Imaging Guidelines.

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Thoracic Aortic Aneurysm (TAA) (PVD-6.2)

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TAA Imaging Indications

Table of Thoracic Aorta Imaging Options

Description	CPT®
CT Chest without contrast	71250
CT Chest with contrast	71260
CTA Chest	71275
MRA Chest	71555
TTE	93306

Imaging of the thoracic aorta listed in the table above is considered medically necessary as follows:

- For suspected TAA, any requested imaging from the **Table of Thoracic Aorta Imaging Options** above is medically necessary for the following:
  - Abnormalities identified on chest x-ray (abnormality including widened mediastinum, suspicious calcifications) or other imaging studies (fluoroscopy, MRI Spine, etc.) abnormality
- Imaging for known thoracic aortic aneurysm can be chosen from the Table of Thoracic Aorta Imaging Options above
- For screening of TAA, see **Screening for Vascular related genetic connective tissue Disorders (PVD-2.2)**
- For TAA associated with a dissection see **Aortic and Arterial Dissection and Other Aortic Conditions (PVD-6.7)**
- For known TAA accompanied with chest pain or back pain and suspicion of rupture, any requested imaging from the Table of Thoracic Aorta Imaging Options above.
- For thoracic endovascular aortic repair (TEVAR) of thoracic aorta disease either of the following is medically necessary:
  - CTA Chest, and/or Abdomen, and/or Pelvis (CPT® 71275, CPT® 74175, CPT® 72191, CPT® 74174)

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  - MRA Chest, and/or Abdomen, and/or Pelvis (CPT® 71555, CPT® 74185, CPT® 72198)
- For follow-up of ascending aortic aneurysms, any requested imaging from the Table of Thoracic Aorta Imaging Options above is medically necessary for the following:
  - For individuals with ascending aortic aneurysms <4.0 cm in diameter
    - Repeat imaging annually
  - For individuals with ascending aortic aneurysms ≥4.0 cm
    - Repeat imaging 6 months.
  - TEE is medically necessary **only** when imaging with CT or MR is contraindicated
- For follow-up of descending aortic aneurysms, any requested imaging from the **Table of Thoracic Aorta Imaging Options** above for the following:
  - Medically treated/observation.
    - 3.5cm to 4.4 cm TAA can be followed annually.
    - ≥4.5 cm TAA can be followed every 6 months.
    - ≥3.0 cm TAA when there is concern for growth can have a one-time 3-month interval advanced imaging.
  - TEE is medically necessary **only** when imaging with CT or MR is contraindicated
- Screening in the presence of other aortic aneurysms.
  - In an individual with a known TAA, screening for AAA is medically necessary with an abdominal duplex. See **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)**.
  - In an individual with a known AAA, screening for TAA is not supported by sufficient evidence and is not medically necessary.
- Screening in individuals with bicuspid aortic valve or familial TAA syndromes. See **Screening for TAA with bicuspid aortic valves (PVD-2.3)**. See **Screening for Vascular related genetic connective tissue Disorders (PVD-2.2)**

Evidence Discussion

Thoracic aortic aneurysms may enlarge over time. Once an aneurysm meets specific size criteria, its risk of rupture as well as the high mortality risk associated with rupture exceeds the risk of surgical intervention. Operative treatment is reasonable for asymptomatic individuals when the diameter of the arch exceeds 5.5 cm. Surveillance recommendations for ascending and descending thoracic aortic aneurysms have been addressed in several major studies.

The location of the thoracic aorta within the chest cavity limits the ability of noninvasive ultrasound to monitor aneurysm size. American College of Radiology recommendations are for CT/MR imaging to monitor the thoracic aorta diameter to determine when surgical intervention is needed. Additionally, the American Heart Association also supports the use of transthoracic echocardiography to monitor the thoracic aortic diameter. In select patients with a difficult echocardiographic imaging window, transesophageal echocardiography is an alternative.

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Thoracic aortic aneurysms may be isolated; however, they may extended below the diaphragm to include portions of the abdominal aorta. Abdominal aortic aneurysms should be followed according to their designated guidelines but may be included with thoracic imaging for certain planned interventions. Certain genetic and familial aneurysm syndromes may have a higher risk of TAA incidence and may warrant additional imaging for detection and surveillance. Additionally, most surgical approaches for thoracic aortic repair require CT/MR imaging for preoperative planning.

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Abdominal Aortic Aneurysm (AAA) (PVD-6.3)

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Duplex ultrasound for visceral vascular studies	CPT®
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study.	93975
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study.	93976
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; complete study.	93978
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; unilateral or limited study.	93979
Ultrasound, abdominal aorta, real time, with image documentation, screening study for abdominal aortic aneurysm (AAA) for AAA screening	76706

Screen for AAA

Ultrasound abdominal aorta with any of the studies from the table of Duplex ultrasound for visceral vascular studies is medically necessary to screen for AAA as follows:

- One-time screening recommendations for AAA (Ultrasound CPT® 76706)
  - Individuals 65 to 75 years of age with a history of tobacco use.
  - Individuals older than 75 years with a history of tobacco use and in otherwise good health who have not previously received a screening ultrasound examination.
  - All first-degree relatives of individuals who present with an AAA and are between 65 and 75, or in those older than 75 in good health.
- AAA screening is considered medically necessary with ultrasound (CPT® 76706, 93975, 93976, 93978, or 93979) when there is a documented thoracic aortic aneurysm

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- There is insufficient evidence to support the use of ultrasound or advanced imaging to screen for a thoracic aortic aneurysm in individuals with known abdominal aortic aneurysm.

Survey known AAA

Ultrasound abdominal aorta with any of the studies from the table of duplex ultrasound for visceral vascular studies is medically necessary to survey known AAA as follows:

- Surveillance of AAA with duplex ultrasound (CPT® 76706, 93975, 93976, 93978, 93979)
  - >2.5 cm but <3.0 cm: 5 years
  - 3.0 cm to 3.9 cm: 3 year intervals
  - 4.0 cm to 4.9 cm in a male: every 12 months OR 4.0-4.4cm in a female: every 12 months
  - ≥5.0 cm in a male OR ≥4.5cm in a female: every 6 months
  - >5.4 cm (or aortic diameter has increased in size by 0.5 cm in six months, or at least 1 cm in a year) may undergo more frequent monitoring and should be evaluated by a Vascular Specialist
- Additional imaging is considered medically necessary as follows:
  - CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen and Pelvis without contrast (CPT® 74176), or CTA Abdomen and Pelvis (CPT® 74174), or CTA Abdomen (CPT® 74175), or CTA Pelvis (CPT® 72191).
    - Suspected or known AAA with recent-onset abdominal or back pain, particularly in the presence of a pulsatile epigastric mass or significant risk factors for AAA
    - Pre-operative imaging for AAA repair
- A one-time CTA Chest (CPT® 71275) or CT Chest with contrast (CPT® 71260) is considered medically necessary prior to planned treatment of AAA.

Evaluate a pulsatile abdominal mass

Ultrasound abdominal aorta with any of the studies from the table of Duplex ultrasound for visceral vascular studies is medically necessary to evaluate a pulsatile abdominal mass:

- Additional Imaging with CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen and Pelvis without contrast (CPT® 74176), or CTA Abdomen and Pelvis (CPT® 74174), or CTA Abdomen (CPT® 74175), or CTA Pelvis (CPT® 72191) for either:
  - Suspected or known AAA with recent-onset abdominal or back pain, particularly in the presence of a pulsatile epigastric mass or significant risk factors for AAA

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- Pre-operative imaging for AAA repair

Obese Individual (BMI ≥35)

- CT Abdomen and Pelvis with contrast (CPT® 74177) or without contrast (CPT® 74176) is medically necessary in an obese individual in place of US, or after an uninterpretable US, using the same timeline as a non-obese individual. Ultrasound abdominal aorta should ideally first be attempted to see if the image quality is adequate.

Evidence Discussion

Abdominal Aortic Aneurysm (AAA) Abdominal aortic aneurysmal disease is usually asymptomatic and discovered incidentally. Symptomatic aortic aneurysms are at high risk for rupture with a significant mortality risk and should be treated emergently with imaging and intervention as indicated. Screening for AAA has some benefit for high risk populations including some long-term tobacco users, individuals with known thoracic or other aneurysms and individuals with certain genetic or familial syndromes. Individuals found to have a pulsatile mass on abdominal exam may also warrant imaging for suspected aneurysmal disease.

For chronic aneurysms, several major studies have been performed with recommendations on surveillance frequency based on rupture risk. Once the rupture risk of an aneurysm meets or exceeds the risk of surgical repair, intervention is recommended. Most repair approaches use endovascular techniques that require preoperative imaging with CT/MR to determine specific anatomic data for appropriate device selection.

The anatomic location of abdominal aortic aneurysms allows ultrasound to be used as a primary imaging modality in most cases for surveillance. Ultrasound uses no radiation or contrast, can be performed in an outpatient setting and is cost-effective. CT/MR imaging is usually reserved for intervention planning or for cases where ultrasound has been found to have technical limitations related to body habitus or other structural issues.

For surgical planning, CT/MR imaging is frequently used to determine anatomic parameters. As many abdominal procedures may require instrument placement within the thoracic aorta, preoperative chest imaging is often a necessary component to surgical planning.[2]

Time between surveillance US in individuals with non-aneurysmal, but dilated aortic diameters, has been shortened, because there is increasing evidence that a significant portion of these individuals will progress to a AAA diameter that may require repair. In addition, because female patients were found to have a significantly higher risk of

[2] Wanhainen A, Van Herzeele I, Goncalves FB, et al. Clinical Practice Guideline Document: Editor's Choice – European Society for Vascular Surgery (ESVS) 2024 Clinical Practice Guidelines on the Management of Abdominal Aorto-Iliac Artery Aneurysms. *Eur J Vasc Endovasc Surg* (2024); 67:192-331

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rupture, more frequent surveillance imaging is justified at AAA diameter of 4 cm and larger.[3]

National guidelines support repair of asymptomatic abdominal aortic aneurysm of ≥5.5cm in males and ≥5.0cm in females. Evidence shows sex related variations in aortic size contributing to a higher risk for rupture at a given aortic diameter in females.[4]

[3] Wanhainen A, Van Herzeele I, Bastos Goncalves F, et al. Editor's Choice -- European Society for Vascular Surgery (ESVS) 2024 Clinical Practice Guidelines on the Management of Abdominal Aorto-Iliac Artery Aneurysms. *Eur J Vasc Endovasc Surg*. 2024;67(2):192-331. doi:10.1016/j.ejvs.2023.11.002

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Dissection

Guideline

Aortic and Arterial Dissection and Other Aortic Conditions (PVD-6.7)
References

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Aortic and Arterial Dissection and Other Aortic Conditions (PVD-6.7)

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Coding

Imaging for Aortic conditions	CPT®
CT Chest, and/or Abdomen, and/or Pelvis	71260 74177 74160 72193
CTA Chest, and/or Abdomen, and/or Pelvis	71275 74175 72191 74174
MRA Chest, and/or Abdomen, and/or Pelvis	71555 74185 72198

- CTA or MRA of the entire aorta (including arch branches) and extending through the femoral arteries is medically necessary for suspected aortic dissection. Any of the following studies can be used if acute dissection is suspected:
  - CT Chest (CPT® 71260 or CPT® 71270) **and/or one of the following:**
    - CT Abdomen (CPT® 74160 or CPT® 74170) with or without and with contrast
    - CT Pelvis (CPT® 72193 or CPT® 72194) with or without and with contrast
    - CT Abdomen and Pelvis (CPT® 74177 or CPT® 74178) with or without and with contrast
  - CTA Chest (CPT® 71275) and/or **one** of the following:
    - CTA Abdomen (CPT® 74175)
    - CTA Pelvis (CPT® 72191)
    - CTA Abdomen and Pelvis (CPT® 74174)

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  - MRA Chest and/or Abdomen and/or Pelvis (CPT® 71555 and/or CPT® 74185 and/or CPT® 72198)
- Any aortic dissection, regardless of treatment modality (medical or surgical), imaging of the chest, abdomen and pelvis (according to the Imaging for Aortic Conditions table above) is medically necessary at the following intervals:
  - 1 month
  - 6 months
  - 12 months
  - If stable, annually
- In individuals with Marfan syndrome/Loeys-Dietz/Ehlers-Danlos and an aortic dissection, post-dissection imaging is medically necessary as follows:
  - 1 month
  - 3 months
  - 6 months
  - 12 months
  - yearly thereafter
- If the dissection extends into the head or neck, the following imaging is also medically necessary:
  - CTA or MRA Head (CPT® 70496 or CPT® 70544)
  - Carotid duplex or CTA Neck or MRA Neck (CPT® 93980, CPT® 70498, or CPT® 70547)
- Imaging of asymptomatic and/or incidentally found arterial dissections that do not affect the aorta (such as, iliac arteries, visceral arteries, extracranial arteries) is medically necessary according to the general schedule as follows:
  - Within one month of discovery
  - Six months
  - 12 months
  - No further imaging after 12 months if noted to be stable
- Imaging of asymptomatic penetrating aortic ulcers treated medically is medically necessary according to the following time intervals as follows:
  - One month after diagnosis
  - If stable, every 6 months for 2 years
  - Then at appropriate intervals thereafter (depending on patient age and PAU characteristics) as determined by the provider managing the condition

Evidence Discussion

Aortic and arterial dissection is usually a result of vessel wall damage due to uncontrolled hypertension or physical trauma. Some connective tissue and genetic disorders also carry a higher risk for vessel dissection. Emergent cases usually present

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with symptoms of impending vessel rupture, organ malperfusion or hemodynamic instability. For these cases, advanced imaging is required for surgical planning. Additional imaging of anatomic regions with suspected involvement are considered necessary in these cases. Acute dissection cases without these features may be observed with medical management until stable.

A chronic arterial dissection will often degenerate into an aneurysm over time. The risk is related to the dissected area of aorta, as well as non-dissected native aortic segments because of underlying medial degeneration. Multiple major studies have been conducted to recommend criteria for intervention based on size and anatomic considerations. Individuals with certain genetic syndromes including Marfan, Loeys-Dietz and Ehlers-Danlos Type IV may be at risk for accelerated vessel degeneration and should undergo more frequent surveillance.

Due to the complex anatomy associated with arterial dissection, CT/MR imaging is recommended over duplex imaging for thoracic, abdominal and visceral artery dissections. Duplex imaging may still be useful for carotid monitoring depending on the extent of vessel involvement.

Asymptomatic arterial dissections not associated with the aorta should undergo regular surveillance for the first year after detection. No further imaging has found to be of benefit if findings remain stable.

Penetrating aortic ulcers are frequently found in the setting of severe aortic atherosclerosis and may carry a significant rupture risk. Frequent surveillance for the first year after detection is medically necessary to determine if intervention is necessary. If the penetrating aortic ulcer remains stable after twelve months, the frequency of imaging may be reduced to annually until intervention is indicated.

Classic symptoms of sharp, severe acute onset of retrosternal or interscapular chest pain is seen in 96% and is best adapted to the emergent setting. Chest x-ray is imprecise; any suspicion should be considered since up to 10% of individuals with aortic dissection present without classic symptoms.

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Post-operative Imaging

Guideline

Post Aortic Endovascular/Open Surgery Surveillance Studies (PVD-6.8)
References
Post-procedure Surveillance Studies (PVD-7.3)
References

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Post Aortic Endovascular/Open Surgery Surveillance Studies (PVD-6.8)

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Open Procedures

- Aortic root/ascending aortic procedures for aneurysm/dissection (ex: aortic root repair, arch/hemi-arch repair, Elephant trunk repair). One of the following post-operative studies [Echocardiography or CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) or CTA Chest (CPT® 71275)], is medically necessary as follows:
  - Within 1 year post-operative
  - Then every 5 years
- Open descending thoracic aortic aneurysm repair- **One** of the following post-operative studies [CT Chest with contrast (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) **or** CTA Chest (CPT® 71275)], is medically necessary as follows:
  - Within 1 year post-operative
  - Then every 5 years
- Open Aortic Abdominal Aneurysm Repair - contrast and non-contrast enhanced CT of the entire aorta (CPT® 74176, CPT® 74177, CPT® 74174) is medically necessary as follows:
  - Within 1 year post-operative
  - Then every 5-years
  - As requested to assess for suspected infection of the graft (see **Vascular Graft Infection (PVD-21)** for additional imaging in vascular graft infection).

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Endovascular procedures

Post-operative surveillance after TEVAR for any indication (PVD-6.8.1)

Imaging for post-operative TEVAR	CPT®
CT Chest, and/or Abdomen, and/or Pelvis	71260 74177 74160 72193
CTA Chest, and/or Abdomen, and/or Pelvis	71275 74175 72191 74174
MRA Chest, and/or Abdomen, and/or Pelvis	71555 74185 72198

Note:

Abdomen and Pelvis imaging is medically necessary only if TEVAR performed for a dissection that extends into the abdomen or pelvis

- **Any** of the studies listed in the table above are medically necessary for post-operative surveillance imaging after TEVAR as follows:
  - One month
  - Twelve months
  - Then annually for life
- If an endoleak is identified at the 1-month study, more frequent imaging is considered medically necessary as requested.

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Post-operative surveillance after abdominal EVAR (endovascular aneurysm repair) (PVD-6.8.2)

Imaging for post-operative abdominal EVAR	CPT®
CT Abdomen and/or Pelvis with contrast	74160 72193 74177
CT Abdomen and/or Pelvis without and with contrast	74170 72194 74178
CTA Abdomen and/or Pelvis	74175 72191 74174
MRA Abdomen and/or Pelvis	74185 72198

Post-operative surveillance after abdominal EVAR is medically necessary as follows:

- CT (any contrast level) or CTA of the entire aorta at 5-year intervals (CPT® 74176)
- CT as per above coding as requested and color duplex ultrasound (CPT® 93975, CPT® 93976, CPT® 93978, or CPT® 93979) one month after EVAR
- If no endoleak, or sac enlargement, repeat either preferred CT or duplex ultrasound (but not both) at 12 months
- If a type II endoleak is observed 1 month after EVAR, may approve **both** at 6 months:
  - Any of the above CT with contrast
  - Color duplex US
- If no endoleak or AAA sac enlargement is detected at 1 year after EVAR annual surveillance with:
  - Color duplex US
  - If DGUS is not available, any of the above CT can be performed
- If a type II endoleak is associated with an aneurysm sac that is shrinking or stable in size:
  - Continue surveillance with color duplex US every 6 months for 2 years

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  - Annually thereafter.
- If US detects a new endoleak, graft migration, or aneurysm sac growth > 5mm:
  - Any of the above CT scan as requested

Post-endoleak intervention surveillance imaging

Surveillance imaging after EVAR, or any subsequent endoleak intervention, is based on the most recent intervention.

For any subsequent interventions for endoleak repair, imaging is medically necessary at 1 month with CT and then follow protocol as above.

Surveillance following Complex EVAR (i.e., a branched/fenestrated graft)

CT as per above coding as requested and/or color duplex ultrasound (CPT® 93975, CPT® 93976, CPT® 93978, or CPT® 93979) is medically necessary at 1-month post-operative, 6 months post-operative, 12 months post-operative, and annually thereafter.

A 3 month follow-up study is considered medically necessary if endoleak of aneurysm enlargement is identified at 1-month post-operative.

Endovascular (Stent) Iliac Repair (PVD-6.8.3)

Imaging for endovascular iliac repair (stent)	CPT®
CT Pelvis	72193 72194
CTA Pelvis	72191
MRA Pelvis	72198

- One of the above studies is medically necessary for endovascular iliac repair (stent)
- If performed in conjunction with EVAR, surveillance will follow the same schedule as EVAR.
- For isolated iliac artery aneurysm repair, surveillance is medically necessary with an arterial duplex (CPT® 93975, CPT® 93976, CPT® 93978, or CPT® 93979) or CT or MR as above if duplex unavailable:
  - Post-cooperatively within the first month
  - 6 months after endovascular treatment
  - Annually

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Evidence Discussion

Post Aortic Endovascular/Open Surgery Surveillance Studies

Surgical approaches to aortic repair have evolved over the past several decades. Open repair is still common for treatment of ascending aortic pathology as well as some thoracic/abdominal aortic disease with complex anatomy. In recent years, however, advances in endovascular technology have made this modality preferred for descending thoracic, abdominal and iliac artery repairs.

Multiple studies have been performed with several recommendations for aortic graft surveillance depending on anatomic location and repair type. Open repair surveillance tends to use longer frequency intervals due to the durability of the repair approach. While less invasive, endovascular repairs require more frequent monitoring as the graft components have a higher risk of technical complication with a need for repeat intervention.

When an endovascular graft system becomes displaced or does not adhere properly to the native vessel wall, an endoleak may develop. An endoleak increases the risk of rupture within a previously repaired aneurysm sac and may require repeat intervention. Some endoleak types may be monitored with regular surveillance if the aneurysm sac diameter remains stable. However, if the aneurysm sac increases in size or if a high risk endoleak type is detected, repeat intervention with preoperative imaging is medically necessary to prevent rupture. Post-endoleak repairs also require regular surveillance as these may also be at risk for technical failure over time.

Thoracic aortic surveillance imaging is usually performed with CT/MR due to the anatomic limitations of duplex in this region. Abdominal aortic surveillance after endovascular repair is initially performed with CT/MR to ensure proper alignment of all graft components. Once this has been established, surveillance with duplex ultrasound is medically necessary unless technical limitations are noted. If an endoleak is detected or if additional pathology such as graft infection is suspected, CT/MR imaging would be appropriate to provide additional anatomic detail. Similarly, if there is concern for repeat intervention, preoperative imaging with CT/MR is indicated.

Complex endovascular aneurysm repair refers to techniques that address branch vessels with branches, stents, or fenestration. These techniques allow for minimally invasive repair of complicated anatomy but need for secondary intervention is relatively high. While duplex ultrasound may be useful for surveillance in these cases, CT scan allows for more detailed imaging and may be useful in some cases.

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Post-procedure Surveillance Studies (PVD-7.3)

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- Scheduled Interval
  - ABI (CPT® 93922) following any revascularization procedure
    - ABI (CPT® 93922) or Duplex ultrasound (CPT® 93926 unilateral study) at each routine follow-up is medically necessary generally after a history/physical has been performed
    - Further imaging studies such as CTA or MRA are medically necessary for worsening symptoms, an abnormal duplex or a significant reduction (>0.15) in the ABI

Indication	Imaging
Suprainguinal Revascularization, both open and endovascular therapy, including Aortobifem/iliofem/fem-fem bypass/iliac angioplasty/stent	• Clinical examination and ABI (CPT® 93922) with arterial duplex (CPT® 93925 or CPT® 93926) is medically necessary at: ◦ Within 1 month ◦ 6 months ◦ 12 months ◦ Then annually
Infrainguinal Open Revascularization (Femoral-popliteal, femoral-tibial, femoral-distal bypass)	
• With vein or autologous conduit	• Clinical exam and ABI (CPT® 93922) with arterial duplex (CPT® 93925 or CPT® 93926) is medically necessary at: ◦ Post-operatively ◦ 3 months ◦ 6 months ◦ 12 months ◦ Then annually

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Indication	Imaging
• With Prosthetic conduit (PTFE/Dacron)	• Clinical exam and ABI (CPT® 93922) with arterial duplex (CPT® 93925 or CPT® 93926) is medically necessary at: ◦ Post-operatively ◦ 6 months ◦ 12 months ◦ Then annually
Infrainguinal Endovascular Revascularization Femoropopliteal angioplasty/stent	• Clinical exam and ABI (CPT® 93922) with arterial duplex (CPT® 93925 or CPT® 93926) is medically necessary at: ◦ Within 1 month ◦ 3 month ◦ Every 6 months for two years ◦ Then annually

After suprainguinal intervention (PVD-7.3.1)

- One of the following studies: Arterial duplex, CTA Abdomen and Pelvis, CT Abdomen and Pelvis with contrast, CTA Aorta with lower extremity runoff, MRI Abdomen and Pelvis, MRA Abdomen and Pelvis, or MRA Aorta with lower extremity runoff is medically necessary for any **one** of the following:
  - Worsening signs or symptoms
  - Reduction of ABI >0.15
  - Peak systolic velocities or PSV ratio suggestive of high grade stenosis or in-stent re-stenosis

After infrainguinal intervention (PVD-7.3.2)

- CTA Lower Extremity (CPT® 73706) or MRA Lower Extremity (CPT® 73725) or CTA aorta with lower extremity runoff CPT® 75635) is medically necessary for any **one** of the following:
  - Worsening signs or symptoms
  - Reduction of ABI >0.15
  - Duplex suggestive of threatened graft
- If intervention was performed for a non-healing wound and wound has gone on to heal, additional imaging is **not** medically necessary for surveillance.

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- Repeat arterial duplex imaging can be obtained for worsening clinical signs and symptoms such as the presence of a new wound or rest pain

Evidence Discussion

Post-procedure Surveillance Studies

Once an intervention (open or endovascular) has been performed, surveillance imaging is supported. The rationale for this is to maintain patency of the treated lesions to avoid further symptoms and/or amputation. Surveillance imaging with ABI and duplex is supported at various intervals depending on the type of intervention, stent vs. bypass, and by bypass material.

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Vasculitis

Guideline

Large Vessel Vasculitis (PVD-6.9)
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Medium Vessel Vasculitis (PVD-6.10)
References
Small Vessel Vasculitis (PVD-6.11)
References

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Large Vessel Vasculitis (PVD-6.9)

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Inflammatory Aortitis (PVD-6.9.1)

Imaging for Inflammatory Aortitis	CPT®
CTA Chest	71275
MRA Chest	71555
CTA Pelvis	72191
MRA Pelvis	72198
CTA Abdomen and Pelvis	74174
CTA Abdomen	74175
MRA Abdomen	74185

Initial imaging

- Initial imaging with CTA or MRA of the affected body region is considered medically necessary when clinical evaluation leads to the suspicion of inflammatory aortitis.

Repeat imaging

- Follow-up imaging with CTA or MRA of the affected body region is considered medically necessary for:
  - Change in signs/symptoms
  - Known aneurysm monitoring
    - See **Cerebral Aneurysm and AVM (HD-12)** in the Head Imaging Guidelines
    - See **TAA Imaging Indications**
    - See **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)**

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Giant Cell Arteritis (GCA) (PVD-6.9.2)

Imaging for Giant Cell Arteritis	CPT®
CTA Chest	71275
MRA Chest	71555
CTA Pelvis	72191
MRA Pelvis	72198
CTA Abdomen and Pelvis	74174
CTA Abdomen	74175
MRA Abdomen	74185
MRA Head without contrast	70544
MRA Head with contrast	70545
MRA Neck without contrast	70547
MRA Neck with contrast	70548
CTA Neck	70498
PET/CT whole body	78816

Initial imaging

Cranial GCA is the more common type with temporal artery involvement. For predominantly Cranial GCA the following studies are considered medically necessary for initial imaging:

- US (CPT® 93880 or CPT® 93882) of the temporal (and or axillary) arteries is the preferred modality where available.
- MRA Head and/or MRA Neck (CPT® 70544, or CPT® 70545, or CPT® 70547, or CPT® 70548) when **either** of the following apply:
  - Vascular trained ultrasonography is not available

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  - US is negative or equivocal with a clinical suspicion of GCA
- MRA of the Abdomen and/or Lower extremities (CPT® 74185 and/or CPT® 73725) when involvement suspected clinically
- PET (CPT® 78816) is considered medically necessary when MRA or US are equivocal and one of the following conditions exists:
  - temporal artery biopsy is negative and GCA is still suspected
  - where the result would change clinical management
- US of the neck vessels (CPT® 93880 or CPT® 93882) and MRA Chest (CPT® 71555)

Note:

For suspected cerebral vasculitis in individuals with neurologic symptoms, see **Cerebral Vasculitis (HD-22)** in the Head Imaging Guidelines

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Extra-cranial GCA: less commonly encountered. None of the “classic” clinical signs or symptoms of cranial GCA are present initially but may develop later.

- Extra-cranial GCA is characterized by at least two or more of the following:
  - Jaw and/or upper extremity claudication
  - Fever/weight loss or fever of unknown origin (FUO) symptoms
  - New murmurs
  - Pulse asymmetry
  - Abdominal pain
  - Pulsatile mass
  - High inflammatory markers such as CRP or ESR > 50 mm/h
- Medically necessary imaging for aortic root, arch or abdomen involvement:
  - MRA Chest (CPT® 71555), MRA Neck (CPT® 70547), MRA Abdomen (CPT® 74185), CTA Chest (CPT® 71275), CTA Neck (CPT® 70498) or CTA Abdomen (CPT® 74175)
  - PET (CPT® 78815) if MRA or CTA are non-diagnostic and there is still suspicion for aortic root, arch or abdomen involvement

Repeat imaging

- Follow-up imaging is medically necessary for **any** of the following:
  - One-time documentation of remission or disease control
  - Change in signs/symptoms suggesting progression of disease
  - Although individuals with GCA can develop aortic aneurysms over time screening in the absence of signs or symptoms is not medically necessary
  - In individuals with known thoracic or abdominal aortic aneurysm:
    - See **TAA Imaging Indications** for thoracic aneurysm surveillance
    - See **Abdominal Aortic Aneurysm (AAA) (PVD-6.3)** for abdominal aneurysm surveillance.

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- Follow-up imaging is not considered medically necessary for individuals in clinical and biochemical remission or without aneurysm/complication.

Takayasu Arteritis (PVD-6.9.3)

Imaging for Takayasu Arteritis	CPT®
CTA Chest	71275
CTA Pelvis	72191
CTA Abdomen and Pelvis	74174
CTA Abdomen	74175
MRA Chest	71555
MRA Pelvis	72198
MRA Abdomen	74185
PET/CT skull base to mid-thigh	78815

Initial imaging

- Initial imaging is medically necessary for signs and symptoms suggestive of disease such as absent radial pulse, difficulty obtaining BP in one arm, or unexplained hypertension.
- Initial staging PET-CT is considered medically necessary in place of MRA or CTA of multiple areas when diffuse large vessel involvement, or occult vessel involvement, is suspected.
- Any of the following modalities are medically necessary for evaluation of Takayasu arteritis:
  - MRA of the affected body area(s) (contrast as requested)
  - CTA of the affected body area(s) (contrast as requested)
  - Ultrasound with Doppler of the affected body area(s)

Repeat imaging

Repeat imaging with any CTA or MRA in the above table is medically necessary at the following intervals:

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- Every 3 months to monitor treatment response during active treatment with systemic therapy.
- Annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention

Evidence Discussion

Large Vessel Vasculitis

Large vessel vasculitis is generally sub-grouped into three areas

- Aortitis (Inflammatory Aortitis)
- Giant Cell Vasculitis
- Takayasu Arteritis

Initial diagnosis of large vessel vasculitis (Inflammatory aortitis, Giant cell, and Takayasu arteritis) should be made through history, physical exam and laboratory values including inflammatory markers. In cases of suspected large vessel disease, ultrasound, CT/ and MR imaging of the neck/chest/abdomen/pelvis are indicated. PET imaging should be reserved for cases where CT/MR are non-diagnostic and the likelihood of disease based on other factors is high.

PET imaging has recently been incorporated into the American College of Rheumatology GL for the Diagnosis of GCA, for research purposes and to exclude alternate diagnosis.[5] PET can be utilized in the rare instances MRA or US are equivocal, temporal artery biopsy is negative and GCA is still suspected, or where the result would change clinical management.[6,7]

Aortic arch involvement is common (see below); US of the neck vessels and MRA of the chest may also be considered in initial screening. Abdominal involvement is less common in GCA, but common in Takayasu's arteritis (see below). MRA of the abdomen and/or LE's, may also be considered in initial screening when involvement suspected clinically.[1,3]

Aortitis

Aortitis may be congenital (Marfan's, Hypermobility Syndromes, others) or acquired, including traumatic, atherosclerotic (dissecting aneurysm, other), infectious (syphilis, tuberculosis, other), neoplastic or inflammatory (Ankylosing Spondylitis, Giant

[5] Ponte C, Grayson PC, Robson JC, et al. 2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis. *Ann Rheum Dis*. 2022;81(12):1647–1653. doi: 10.1136/ard-2022-223480.

[6] American College of Radiology (ACR). ACR Appropriateness Criteria® Noncerebral Vasculitis. *J Am Coll Radiol*. 2021;18(11S):S380–S393. doi: 10.1016/j.jacr.2021.08.005.

[7] Dejaco C, Ramiro S, Bond M, et al. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice: 2023 update. *Ann Rheum Dis*. 2024;83(6):741–751. doi: 10.1136/ard-2023-224543.

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Cell Arteritis, Cogan's, Relapsing Polychondritis, Behcet's Syndrome, Polyarteritis Nodosa, Granulomatous Polyangiitis, Lupoid, idiopathic, other).

Giant Cell Arteritis

Historically, GCA had been be subdivided into two basic types; Cranial Predominant and Extra-cranial Currently, both GCA type vasculitis and Takayasu's arteritis have similar management requirements as variants of Large vessel vasculitis (LVV).

Initial diagnosis of large vessel vasculitis (Inflammatory aortitis, Giant cell, and Takayasu arteritis) should be made through history, physical exam and laboratory values including inflammatory markers. In cases of suspected large vessel disease, ultrasound, CT/ and MR imaging of the neck/chest/abdomen/pelvis are may be indicated. PET imaging should be reserved for cases where CT/MR are non-diagnostic and the likelihood of disease based on other factors is high. Follow-up imaging is medically necessary for individuals with known aneurysmal disease or who remain symptomatic on active therapy. PET imaging has recently been incorporated into the American College of Rheumatology GL for the Diagnosis of GCA, for research purposes and to exclude alternate diagnosis.[1]

Takayasu Arteritis

Instead of multiple MRA's, an initial staging PET-CT can be approved where diffuse large vessel involvement, or occult vessel involvement, is suspected in an LVV. Ordering both studies initially is discouraged. Follow-up MRA is preferred as PET-CT in some patients may never normalize. For Takayasu arteritis, annual surveillance in the absence of symptoms is recommended due to the high risk of progressive vascular damage.

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Medium Vessel Vasculitis (PVD-6.10)

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- There are two main types of medium-size vessel vasculitis:
  - Kawasaki disease
  - Polyarteritis nodosa

Kawasaki disease

- Imaging guidelines below for Kawasaki Disease can also be accessed in **Kawasaki Disease (PEDCD-6)** in the Pediatric Cardiac imaging guideline

Long term routine surveillance in asymptomatic imaging for Kawasaki disease

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
All			All risk levels 4-6 weeks after acute illness		
1	Normal	Normal	One echo 2-12 months after acute illness	none	none
2	Dilation	Dilation	6 months One year If dilation remains echo every 2-5 years until resolves	None	None
		Normal	After acute illness: 2-12 months One echocardiogram at one year No echocardiogram after one year		

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AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
3.1	Small	Small	6 months 12 months then yearly	2-3 years	3-5 years
3.2	Small	Normal or dilated	6 months 12 months Then yearly	3-5 years	none
4.1	Medium	Medium	3 months 6 months 12 months Every 6-12 months after that	1-3 years	2-5 years
4.2	Medium	Small	6 months and 12 months, every 1 year	2-3 years	3-5 years
4.3	Medium	Normal Or Dilated	Every 1-2 years	2-4 years	none
5.1	Large	Large	1 month 3 months 6 months 9 months 12 months Then every 3-6 months	6-12 months	at 2-6 months, every 1-5 years
5.2	Large	Medium	Every 6-12 months	yearly	2-5 years
5.3	Large	Small	6-12 month	1-2 years	2-5 years

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AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
5.4	Large	Normal Or Dilation	1-2 years	2-5 years	none

Polyarteritis Nodosa

Initial imaging

Any ONE of the following modalities is medically necessary for documented clinical suspicion of Polyarteritis Nodosa:

- MRA of the affected body area(s) (contrast as requested)
- CTA of the affected body area(s) (contrast as requested)
- Ultrasound (US) with Doppler of the affected body area(s)

Repeat imaging

Imaging with MRA, CTA, or US with Doppler of the affected body area(s) is medically necessary for established Polyarteritis Nodosa as follows:

- Every 3 months during active treatment with systemic therapy to evaluate treatment response
- Annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention

Background and Supporting Information

- Based on AHA recommendations, the following classifications are used in risk stratification of coronary artery abnormalities[8]:
  - Z-score classification accounts for the effects of body size and age through use of baseline coronary dimensions adjusted for body surface area. The z-score value represents the number of standard deviation above the mean. (e.g., z=0 pt. has coronary artery dimension value equal to mean, z=2 person has value 2 standard deviation above the mean, based on age, gender, BSA).
  - Coronary Artery Abnormalities Risk Classification based on z-score:
    - 1 - No involvement at any time point (z-score always <2)
    - 2 - Dilation only (z-score 2 to <2.5)
    - 3 - Small aneurysm (z-score ≥2.5 to <5)
      - 3.1 - Current or persistent

[8] Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. Circulation. 2017;135(17). doi:10.1161/cir.0000000000000484.

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    - 3.2 - Decreased to dilation only or normal luminal dimension
  - 4 - Medium aneurysm (z-score ≥5 to <10, and absolute dimension <8 mm)
    - 4.1 - Current or persistent
    - 4.2 - Decreased to small aneurysm
    - 4.3 - Decreased to dilation only or normal luminal dimension
  - 5 - Large and giant aneurysm (z-score ≥10, or absolute dimension ≥8 mm)
    - 5.1 - Current or persistent
    - 5.2 - Decreased to medium aneurysm
    - 5.3 - Decreased to small aneurysm
    - 5.4 - Decreased to dilation only or normal luminal dimension
- Additional clinical features that may increase the long-term risk of myocardial ischemia:
  - Greater length and distal location of aneurysms that increase the risk of flow stasis
  - Greater total number of aneurysms
  - Greater number of branches affected
  - Presence of luminal irregularities
  - Abnormal characterization of the vessel walls (calcification, luminal myofibroblastic proliferation)
  - Presence of functional abnormalities (impaired vasodilation, impaired flow reserve)
  - Absence or poor quality of collateral vessels
  - Previous revascularization performed
  - Previous coronary artery thrombosis
  - Previous myocardial infarction
  - Presence of ventricular dysfunction

Evidence Discussion

Medium Vessel Vasculitis

Medium vessel vasculitis includes multiple inflammatory processes that affect the major arteries the cerebrovascular, thoracic and abdominal regions. Cardiac involvement is also common. Pediatric populations are most often affected. Aneurysmal degeneration risk is high and does require regular surveillance. The two most common types include:

- Kawasaki disease
- Polyarteritis nodosa

Initial diagnosis should be made through history, physical exam and laboratory values including inflammatory markers. Aneurysmal degeneration of vessels may be seen in

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multiple anatomic regions. For individuals with suspected disease, ultrasound, CT and MR imaging of the neck/chest/abdomen/pelvis is indicated.

Annual surveillance in the absence of symptoms is still recommended due to the high risk of progressive vascular damage.

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Small Vessel Vasculitis (PVD-6.11)

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- Advanced imaging is not sensitive enough to detect changes in small vessels, and is not medically necessary for primary assessment of any small vessel vasculitis.
- End-organ damage can occur with several of the small vessel vasculitides and are addressed in this guideline:
  - Henoch-Schönlein Purpura (HSP)
  - Granulomatosis with polyangiitis (GPA, formerly known as Wegener's granulomatosis)
  - Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as ChurgStrauss Syndrome)
  - Immune complex associated small-vessel vasculitis [immunoglobulin A–associated vasculitis (IgAV)]

IgA Vasculitis Henoch-Schönlein Purpura (HSP)

Initial imaging

Ultrasound (US) Abdomen (CPT® 76700) is medically necessary to evaluate possible gastrointestinal complications of known or suspected HSP including **any** of the following:

- Bowel wall edema and hemorrhage
- Intussusception

CT Abdomen with contrast (CPT® 74160) is medically necessary if additional information is needed after ultrasound for management

Repeat imaging

US Abdomen (CPT® 76700) is medically necessary for known HSP to evaluate new or worsening gastrointestinal symptoms

Background and Supporting Information

Henoch-Schönlein Purpura (HSP) is the most common vasculitis of childhood, mainly involving small blood vessels.

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Granulomatosis with Polyangiitis (GPA) Formerly Wegener's Granulomatosis

Initial Imaging

Initial imaging as a baseline prior to starting immunosuppressive therapy is medically necessary in all individuals who are newly diagnosed or suspected of having antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitides (AAV) with either or both of the following to evaluate the extent of the disease:

- CT Sinuses (CPT® 70486) and/or
- CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260)

Note:

Preferred CT imaging in individuals with AAV should be performed without an iodinated contrast agent administered.

Repeat Imaging

CT Sinuses (CPT® 70486) and/or CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) are medically necessaryfor **any** of the following:

- New or worsening clinical symptoms affecting the body area requested
- Assess response to medical therapy when a change in treatment regimen is being considered
- Annually-to evaluate the extent of disease

Eosinophilic Granulomatosis with Polyangiitis (EGPA) Formerly Churg-Strauss Syndrome

Initial Imaging

- CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) is medically necessary in the initial evaluation of Eosinophilic granulomatosis with polyangiitis (EGPA) formerly known as Churg-Strauss Syndrome.

Repeat Imaging

- CT Chest without contrast (CPT® 71250) or CT Chest with contrast (CPT® 71260) is medically necessary for any of the following:
  - New or worsening clinical symptoms affecting the body area requested

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- Assess response to medical therapy when a change in treatment regimen is being considered
- Annually-to evaluate the extent of disease

Immune Complex Associated Small-vessel Vasculitis

Initial Imaging

- Doppler ultrasound (US) of the affected body part (most commonly abdomen) is medically necessary in the initial evaluation of immune complex associated small-vessel vasculitis

Repeat Imaging

- Doppler ultrasound (US) of the affected body part (most commonly abdomen) is medically necessary in the following circumstances
  - New or worsening clinical symptoms affecting the body area requested
  - To assess response to medical therapy when a change in treatment is being considered
  - Annually-to evaluate the extent of disease

Evidence Discussion

Small Vessel Vasculitis

Advanced imaging is not routinely needed with diagnosis of Small vessel vasculitis. In suspected cases, clinical assessment of end-organ damage is usually indicated in both the adult and pediatric population

Initial diagnostic workup should include history, physical exam, and laboratory data including inflammatory markers. End-organ damage may involve multiple organ systems based on the type of vasculitis. For individuals with suspected disease, ultrasound, CT and MR imaging of the affected anatomic regions may be indicated. Annual surveillance in the absence of symptoms is still recommended due to the high risk of progressive vascular damage.

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Peripheral Artery Aneurysms

Guideline

Visceral Artery Aneurysm (PVD-6.5)
References
Iliac Artery Aneurysm (IAA) (PVD-6.4)
References
Lower Extremity Artery Aneurysms (PVD-7.4)
References

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Visceral Artery Aneurysm (PVD-6.5)

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Treatment is generally indicated for visceral aneurysms ≥2cm. Imaging of visceral artery aneurysms for initial evaluation or for treatment planning is medically necessary as follows:

- Initial imaging of calcifications seen on plain film imaging suspicious for visceral artery aneurysms (spleen, kidney, liver or intestines) with ultrasound (CPT® 76700, 76705, 93975, 93976, 93978, or 93979), **or** CTA Abdomen (CPT® 74175), **or** CT Abdomen with contrast (CPT® 74160)
- MRA Abdomen (CPT® 74185) without contrast in place of CTA or CT Abdomen in pediatric individuals or when there is a contraindication to contrast materials (i.e., renal insufficiency, contrast allergy, pregnancy)
- Ultrasound (CPT® 76700,76705, 93975, 93976, 93978, or 93979) **or** CTA Abdomen (CPT® 74175) **or** CT Abdomen with contrast (CPT® 74160) is considered medically necessary for further monitoring based on the intervals below or as determined by a vascular specialist or any provider in consultation with a vascular specialist:
  - Splenic artery aneurysms:
    - <20mm can be imaged every three years
    - 20mm to 29mm can be imaged annually
    - If ≥30mm, they should be referred for treatment, either stent, excision or splenectomy
  - For all other visceral artery aneurysms:
    - Initial evaluation with six-month follow-up for one year
    - Further follow-up annually if no significant enlargement is seen
- CTA Abdomen (CPT® 74175), MRA Abdomen (CPT® 74185), or CT Abdomen with contrast (CPT® 74160) is considered medically necessary following stent placement at:
  - 1 month
  - 6 months
  - 12 months
  - Then every year

Evidence Discussion

Aneurysmal disease, besides primarily involving the large vessels, can also affect medium and smaller sized vessels. Visceral cases are uncommon and occasionally are associated with certain connective tissue and genetic disorders. They are often found incidentally on imaging. Ultrasound, CT, or CTA imaging may be indicated for

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surveillance in these cases. Due to the anatomic location of the visceral vessels, duplex ultrasound may have technical limitations. Consideration of best surveillance study should be decided on initial imaging and whether a certain modality is felt to provide diagnostic information. Example; some splenic arterial aneurysms may be diagnostic with US but others may be obscured by bowel gas requiring CT/CTA. Splenic artery aneurysms, the most common (60%), tend to exhibit very slow rates of growth, while the other visceral artery aneurysms are more unpredictable in their rate of growth with a greater tendency to rupture. Visceral Artery Aneurysms are defined by an increase of more than 50% of the original arterial diameter and include hepatic, renal and intestinal artery aneurysms.

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Iliac Artery Aneurysm (IAA) (PVD-6.4)

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- Ultrasound (CPT® 76882 or CPT® 93925) is medically necessary for evaluation of a suspected IAA
  - CT Pelvis with contrast (CPT® 72193) if ultrasound is equivocal.
  - Ultrasound for follow-up imaging annually if an aneurysm is ≥2cm
- Additional Imaging
  - CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen and Pelvis without and with contrast (CPT® 74178), or CTA Abdomen and Pelvis (CPT® 74174) is medically necessary for preoperative imaging if endovascular or open repair is being considered

Background and Supporting Information

- Isolated IAA's are rare and are typically associated with AAA
- Approximately one third to one half of isolated IAA's are bilateral at time of presentation
- Abdominal Aortic aneurysm rupture usually occurs at a diameter of 5 cm or larger, whereas common iliac aneurysms that are less than 3 cm in diameter almost never rupture

Evidence Discussion

Annual surveillance of iliac artery aneurysms is medically necessary to determine when intervention is necessary if an iliac aneurysm exceeds 2cm in diameter. Duplex ultrasound is the primary imaging modality for individuals without technical limitations related to body habitus. It has been demonstrated to be accurate, cost-effective and does not use ionizing radiation or contrast. CT imaging may be indicated for cases where ultrasound is equivocal or for surgical planning.

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Lower Extremity Artery Aneurysms (PVD-7.4)

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Imaging Indications

Iliac Artery Aneurysm

See **Iliac Artery Aneurysm (IAA) (PVD-6.4)**

Femoral Artery Aneurysm

- Initial imaging is medically necessary as follows:
  - Ultrasound (CPT® 93925 bilateral study or CPT® 93926 unilateral study).
- Surveillance imaging is medically necessary as follows:
  - Asymptomatic true femoral aneurysms smaller than 2.5 cm in diameter
    - Ultrasound (CPT® 93926 unilateral study) annually
  - Asymptomatic true femoral aneurysms larger than 2.5 cm
    - Ultrasound (CPT® 93926 unilateral study) every 6 months
- Other imaging is medically necessary as follows:
  - CTA or MRA Lower extremity (CPT® 73706 or 73725 or 74198 or 75635):
    - Preoperative study for individuals with no plans for invasive angiography
    - Technically limited or abnormal ultrasound results

Popliteal Artery Aneurysm

- Initial imaging is medically necessary as follows:
  - Ultrasound (CPT® 93925 bilateral study or CPT® 93926 unilateral study) and Ultrasound to assess for a contralateral popliteal aneurysm and abdominal aortic aneurysm (CPT® 76770 or CPT® 76775)
- Surveillance imaging is medically necessary as follows:
  - If no intervention: Ultrasound (CPT® 93926 unilateral study) annually
  - Post-intervention: (ABI (CPT® 93922) and Duplex ultrasound are indicated as follows:
    - 3 months post-operative
    - 6 months post-operative
    - 12 months post-operative
    - Then annually

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- Other imaging is medically necessary as follows:
  - CTA or MRA (CPT® 73706 or 73725 or 74185 or 75635) for:
    - Preoperative study
    - Technically limited or abnormal ultrasound results

Evidence Discussion

Duplex ultrasound is the preferred modality for surveillance of lower extremity arterial aneurysms. This modality uses no ionizing radiation or contrast, has a reasonable level of accuracy and is cost-effective. CT/MR imaging may be indicated for preoperative planning or in cases where ultrasound is technically limited.

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Peripheral Arterial Imaging

Guideline

Upper Extremity PVD – Imaging (PVD-4.1)
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Renovascular Hypertension/Renal Artery Stenosis (PVD-6.6)
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References
Claudication and Critical Limb Ischemia (PVD-7.1)
References
Popliteal Artery Entrapment Syndrome (PVD-7.2)
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Arterial Imaging for Free Flaps in Reconstructive Surgery (PVD-7.5)
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Upper Extremity PVD – Imaging (PVD-4.1)

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Initial Imaging

For arterial insufficiency, arterial ultrasound upper extremities (CPT® 93930 or CPT® 93931) is medically necessary for signs and symptoms of arterial insufficiency including:

- Arm or hand claudication
- Bluish discoloration of the hand or fingers
- Unilateral cold, painful, pulseless hand
- Non-healing wound (>2 weeks with no healing or evidence of healing) or frank gangrene
- CTA Upper extremity (CPT® 73206) or MRA of Upper extremity (CPT® 73225), **and/or** CTA Chest (CPT® 71275) or MRA Chest (CPT® 71555) is medically necessary for:
  - Abnormal arterial ultrasound results
  - Equivocal arterial ultrasound results
  - Pre-operative planning

For Subclavian Steal Syndrome, the following studies are considered medically necessary:

- Carotid duplex (CPT® 93882) is the initial and definitive imaging study.
- CTA chest (CPT® 71275) or MRA chest (CPT® 71555) for carotid duplex showing reversal of flow in one of the vertebral arteries.
- Surveillance of subclavian arterial disease is **not** medically necessary if there has not been any intervention such as a carotid-subclavian bypass or subclavian stent

Note:

If the carotid duplex is not diagnostic for reversal of flow in the ipsilateral vertebral artery, then neurological symptoms should be evaluated according to the Head Imaging guideline

For suspected Fibromuscular Dysplasia of the brachial artery, medically necessary studies include:

- MRA Upper extremity (CPT® 73225)
- CTA Upper extremity (CPT® 73206)

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- Arterial Ultrasound (CPT® 93930 bilateral study or CPT® 93931 unilateral study)

For arterial thoracic outlet syndrome, medically necessary studies include:

- CTA Chest (CPT® 71275) (preferred study) or MRA Chest (CPT® 71555) (preferred study) or CT Chest either without or with contrast (CPT® 71250 or CPT® 71260) or MRI Chest with contrast (CPT® 71551)

Post-revascularization

- Arterial Duplex (CPT® 93931) is medically necessary following upper extremity arterial revascularization at:
  - Baseline (within one month)
  - 6 months
  - Then annually if stable
  - Anytime for new or worsening symptoms of arterial insufficiency

Evidence Discussion

Upper extremity PVD

Duplex ultrasound (DU) is the initial imaging modality for assessment of patients with symptoms of upper extremity arterial occlusive disease including arm/hand claudication, non-healing wounds, blue discoloration, or a unilateral cold, painful, and pulseless hand.

DU is a non-invasive, cost-effective method for screening for arterial disease of the upper extremity. It has high sensitivity and specificity when compared to CT angiography (CTA) and MR angiography (MRA). CTA and MRA also evaluate anatomic location of disease but are reserved for confirmed vascular disease, suspected fibromuscular dysplasia of the brachial artery, and/or pre-operative planning. They are not first line imaging studies due to the higher risks associated with their use and the cost.

Post-revascularization surveillance with DU has been established as a reliable method to monitor the intervention for recurrence of disease. The time intervals for surveillance have been established at one and six months post-intervention, then yearly. If at any time, there are new signs or symptoms of disease, repeat duplex imaging is supported.

If initial duplex demonstrates high-grade stenosis or occlusion of the subclavian artery, advanced imaging is NOT indicated unless the individual is symptomatic with arm claudication or signs of hypo-perfusion of the vertebral artery with recurrent dizziness.

Subclavian Steal refers to a hemodynamically significant stenosis or occlusion of the subclavian/innominate artery which results in the reversal of blood flow in the vertebral artery (VA). Signs/Symptoms associated with this syndrome include:

- Physical examination with a >15mmHg discrepancy in blood pressure between two arms
- Supraclavicular bruit

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- Symptoms of vertebrobasilar insufficiency, including vertigo and limb paresthesia particularly with use of the ipsilateral arm.

Thoracic outlet syndrome (TOS) refers to compression of the neurovascular structures within the thoracic outlet as they pass from the neck and thorax to the axilla.

- There are three types of TOS, neurogenic, arterial and venous.
- Neurogenic TOS refer to Brachial Plexus (PN-4.1) in the peripheral nerve disorders imaging
- Venous TOS refer to **Upper Extremity Venous Imaging (PVD 4.2)**

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Renovascular Hypertension/Renal Artery Stenosis (PVD-6.6)

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Initial imaging

- MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) or US kidney retroperitoneal (CPT® 76775) and/or Doppler (CPT® 93975, 93976, 93978, or 93979 if expertise is available) are medically necessary when there is documentation of **any** of the following:
  - Individual is adherent to full doses of three blood pressure medications (including a diuretic) yet has still not achieved goal
  - Sudden and persistent worsening of previously controlled hypertension
  - Onset of hypertension younger than 30 years of age
  - Malignant hypertension with coexistent evidence of acute end-organ damage (acute renal failure, new visual or neurological disturbance and/or advanced retinopathy) or flash pulmonary edema
  - Individuals who develop hypertension (≥140/90) within the first 20 weeks of pregnancy when hypertension persists >12 weeks post-partum
  - New or worsening renal function/increasing creatinine (especially after the administration of an ACE inhibitor or with angiotensin receptor blocking agent)
- Carotid duplex (CPT® 93880) is medically necessary to screen for carotid involvement in individuals with documented or highly suspicious renal artery stenosis due to fibromuscular dysplasia (mostly women between 15 and 50 years of age).

Screening

CTA Abdomen (CPT® 74175) or MRA Abdomen (CPT® 74185) is medically necessary to screen for renovascular fibromuscular dysplasia in hypertensive individuals with documented cervicocephalic fibromuscular dysplasia. The assessment of other vascular beds should be considered if supported by suggestive symptoms or medical history.

Carotid duplex (CPT® 93880) is medically necessary to screen for carotid involvement in individuals with documented or highly suspicious renal artery stenosis due to fibromuscular dysplasia (mostly women between 15 and 50 years of age)

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Repeat imaging post revascularization

- CTA Abdomen (CPT® 74175), or MRA Abdomen (CPT® 74185), or CT Abdomen with contrast (CPT® 74160) is medically necessary after stent placement at the following intervals:
  - 1 month post-procedure
  - 6 months post-procedure
  - 12 months post-procedure
  - Then annually

Evidence Discussion

Renovascular Hypertension

The American Heart Association 2017 guidelines on hypertension estimate renovascular disease to be the source of 5-34% of all cases. Multiple studies have shown that intervention is not more effective than medical therapy in most cases as reviewed by the KDIGO conference in 2022; however, it is noted that these recommendations continue to evolve. While duplex ultrasound is the preferred imaging modality, patients with compromised renal function or technical limitations due to body habitus may require CT/MR imaging for monitoring.

Current recommendations for renal artery screening in the setting of hypertension refractory to medical therapy include the following:

- Individual is adherent to full doses of three blood pressure medications (including a diuretic) yet has still not achieved goal
- Sudden and persistent worsening of previously controlled hypertension
- Onset of hypertension younger than 30 years of age
- Malignant hypertension with coexistent evidence of acute end-organ damage (acute renal failure, new visual or neurological disturbance and/or advanced retinopathy) or flash pulmonary edema
- Individuals who develop hypertension (≥140/90) within the first 20 weeks of pregnancy when hypertension persists >12 weeks post-partum o New or worsening renal function/increasing creatinine (especially after the administration of an ACE inhibitor or with angiotensin receptor blocking agent)

Fibromuscular dysplasia carries a higher risk of concomitant renal and carotid artery involvement. Screening of both anatomic regions is medically necessary upon diagnosis.

Post-intervention imaging with CT/MR imaging at standard 1/6/12 month intervals followed by annual imaging to assess for stent patency follow standard recommendations for non-coronary interventions. Additional surveillance is not indicated in the absence of symptoms.

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Renal artery revascularization has not been shown to be more effective than medical therapy in most situations and should not be pursued except in extreme cases, or if there is concern for Takayasu arteritis or fibromuscular dysplasia. Gadolinium agents may be contraindicated in individuals with severe renal disease or on dialysis due to the risk of developing nephrogenic systemic sclerosis.

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Median Arcuate Ligament Syndrome, Nutcracker Syndrome and other Abdominal Vascular Compression Syndromes (PVD-18)

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Median Arcuate Ligament Syndrome

Codes included

CPT®	Description
74175	Computed tomographic angiography, abdomen, with contrast material(s), including noncontrast images, if performed, and image post-processing
74185	Magnetic resonance angiography, abdomen, with or without contrast material(s)
93975	Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study
93976	Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study

Initial Imaging

- Duplex Doppler Ultrasound of Mesenteric Arteries (CPT® 93975 or 93976) is medically necessary as the initial imaging
- CTA or MRA Abdomen (CPT® 74175 or 74185) is medically necessary for **either**:
  - US results are equivocal
  - Preoperative planning

Repeat Imaging

- Surveillance imaging is **not** medically necessary post-operatively in the absence of abdominal symptoms.

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Evidence Discussion

A non-atherosclerotic cause of chronic mesenteric ischemia.

Patients are typically younger with an average age range of 30-50 years old and a female to male ratio of 4:1

Patients may have symptoms of postprandial abdominal pain, nausea, vomiting, food aversion, weight loss. Symptoms are nonspecific, so MALS may be considered a diagnosis of exclusion.

Imaging will demonstrate compression of the celiac artery by fibers of the median arcuate ligament, but this is a nonspecific finding and may be seen in the asymptomatic population.

Pathophysiologically related to foregut ischemia from compression of the celiac artery; may also be related to neuropathic pain secondary to compression of the celiac ganglion.

Treatment is generally surgical, via a variety of approaches.

Left Renal Vein Compression ("Nutcracker") Syndrome

Codes included

CPT®	description
93975	Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study
93976	Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study
74174	Computed tomographic angiography, abdomen and pelvis, with contrast material(s), including noncontrast images, if performed, and image post-processing
72198	Magnetic resonance angiography, pelvis, with or without contrast material(s)
74185	Magnetic resonance angiography, abdomen, with or without contrast material(s)

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Initial Imaging

- Abdominal duplex (CPT® 93975 or 93976) is medically necessary as the initial imaging to confirm left renal vein compression syndrome.
- CTV or MRV Abdomen and Pelvis (CPT® 74174) or (CPT® 74185 and 72198) is medically necessary for preoperative planning when left renal vein compression syndrome is confirmed by duplex ultrasound.

Repeat imaging

- Postoperative follow-up imaging is **not** medically necessary in the absence of symptomatology.

Background and Supporting Information

Patients may present with symptoms related to compression of the left renal vein between the aorta and superior mesenteric artery. A less common presentation ("posterior nutcracker syndrome") is related to compression of a retroaortic (or circumaortic) left renal vein between the aorta and vertebral body.

The radiologic finding may be asymptomatic or considered a normal variant.

Signs/symptoms may include hematuria, proteinuria, flank pain, pelvic congestion/ associated pain in females, varicocele in males.

There may be an overlap in symptom complex with patients who have pelvic congestion syndrome, iliac vein compression (May-Thurner).

There is no single accepted treatment modality. Approaches will range from conservative/medical (esp in younger patients), to endovascular, to open/laparoscopic/ robotic surgical.

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Claudication and Critical Limb Ischemia (PVD-7.1)

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- Resting ABI for initial evaluation of suspected PAD. This can be accomplished at the bedside as part of the physical examination or requested as CPT® 93922 (limited Doppler ultrasound) or CPT® 93923 (multi-level complete Doppler ultrasound)
  - CPT® 93923 is medically necessary once
  - Follow-up studies are medically necessary with CPT® 93922
  - Post-exercise ABI (CPT® 93924) is medically necessary if the resting ABI is >0.89 and PAD is still highly suspected clinically.
- History and physical suggestive of PAD include:
  - History
    - Claudication- reproducible calf or thigh cramping with exertion that is relieved completely with rest
    - Critical limb ischemia
    - Rest pain suggestive of ischemia-pain in the ball of foot when the leg is in an elevated position particularly at night
    - Distal non-healing wound or punched out ulcer with sharply demarcated edges present for >2 weeks with no evidence of healing, i.e. presence of granulation tissue
  - Physical Examination
    - Abnormal lower extremity pulse examination
    - Vascular bruit
    - Non-healing lower extremity wound
    - Lower extremity gangrene
    - Other suggestive lower extremity physical findings (e.g., elevation pallor/ dependent rubor)
    - Atrophic nails, hair loss, shiny skin
- If resting ABI (CPT® 93922) is normal (0.9 to 1.3) and disease is still suspected:
  - Differentiate from “pseudoclaudication”. See **Lumbar Spinal Stenosis (SP-9)** in the Spine Imaging Guidelines
  - Re-measure ABI after exercise (CPT® 93924)
  - A TBI (toe-brachial index) may be used as further screening in individuals with ABI’s ≥1.4
  - Advanced imaging is medically necessary only if there is consideration for invasive therapy not to confirm diagnosis

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- Duplex ultrasound (CPT® 93925 bilateral study or CPT® 93926 unilateral study) and Doppler studies are adjuncts to abnormal ABI that are considered medically necessary to identify location and extent of disease once there has been a decision for revascularization.
- MRA Aorta and Pelvic vessels, and Lower extremities (CPT® 74185, CPT® 73725 and CPT® 73725), or CTA with run-off (CPT® 75635) are medically necessary to further evaluate the lower extremity arteries for ANY of the following:
  - Potentially limb-threatening vascular disease evidenced by:
    - Skin breakdown
    - Non-healing ischemic ulcers
    - Resting leg pain
    - Gangrene
  - Blue Toe Syndrome:
    - Emboli from aortic plaque or mural thrombus
    - Hyperviscosity syndrome
    - Hypercoagulable states
    - Vasculitis
  - Preoperative planning for Intermittent claudication (i.e., non-limb threatening ischemia) AND there is documentation of both of the following:
    - Failed 3-months' conservative medical therapy (physician supervised walking/ exercise program plus medical therapy)
    - Functional disability (e.g., exercise impairment sufficient to threaten the individual's employment or to require significant alterations in the individual's lifestyle)
  - CTA lower extremity (CPT® 73706) OR MRA lower extremity (CPT® 73725) is medically necessary for evaluation of PVD when aortoiliac disease is not a concern or the state of the aorta and iliac arteries is already known as documented in the clinical history

Note:

MRA Pelvis (CPT® 72198) should not be requested/billed with CPT® 74185, CPT® 73725 and CPT® 73725

- To evaluate for an embolic source, the following studies are medically necessary:
  - CTA chest (CPT® 71275) OR MRA Chest (CPT® 71555) AND
  - CTA Abdomen and Pelvis (CPT® 74174) OR MRA Abdomen and Pelvis (CPT® 74185) (if imaging of abdomen/pelvis not already obtained)

    See also Echocardiogram in the Cardiac imaging guideline

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Evidence Discussion

Peripheral arterial disease (PAD) is defined as chronic, atherosclerotic occlusive disease of the lower extremities. The vast majority of patients with PAD are asymptomatic. A much smaller group has symptomatic PAD, consisting of intermittent claudication(IC), rest pain or tissue loss.

The natural history of PAD for asymptomatic and IC patients is relatively benign. It is estimated that 7% (4%–11%) of asymptomatic patients deteriorate to IC over a 5-year period. Multiple studies have established that patients with IC are at very low risk of major amputation (<1% per year).

For these reasons, the first line of treatment for patient with IC is risk factor reduction/ modification and exercise therapy. A meta-analysis of 1200 patients determined that exercise therapy, compared with placebo or usual care, provides an overall improvement in walking ability of 50% to 200%, with improvements maintained for up to 2 years. Additionally, with intensive medical management, <5% of patients will develop symptoms of advanced ischemia, such as ischemic rest pain, tissue loss, or require amputation.

Diagnosis:

Measurement of the ankle-brachial index (ABI) is the primary method for establishing the diagnosis of PAD. An ABI of 0.90 has been demonstrated to have high sensitivity and specificity for the identification of PAD compared with the gold standard of conventional angiography (CA). It is a simple non-invasive method to assess for PAD. It avoids use of radiation, and contrast agents. It can be performed easily in an office setting and is cost-effective.

Further studies for evaluation of anatomic location of disease are warranted if PAD is proven. Arterial duplex combines Doppler spectral analysis and B-mode imaging to evaluate blood flow and anatomy. It has been shown to be effective in localizing arterial vascular disease with comparable sensitivity and specificity to CT angiography (CTA), MR angiography (MRA) and CA. It is also non-invasive, cost-effective, and avoids radiation and contrast exposure.

Due to their associated risks, CTA, MRA and CA should be reserved for patients in whom revascularization treatment is being considered. CTA risks include exposure to intravenous contrast and radiation. Contrast complications include allergy and contrast induced nephropathy. MRA risk includes exposure to gadolinium which confers the risk of nephrogenic systemic fibrosis in patients with renal insufficiency. MRA also is contraindicated in patients with metallic implants. CA risks include radiation and contrast exposure as well as access site complications. Since it is an invasive procedure, there is risk for arterial injury, embolization and thrombosis.

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Popliteal Artery Entrapment Syndrome (PVD-7.2)

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- Diagnosis of popliteal artery stenosis or occlusion due to compression by adjacent muscle and tendons all of the following are medically necessary:
  - Resting ABI (CPT® 93922 OR 93923) AND/OR Post-Exercise ABI (CPT® 93924) AND
  - Ultrasound (CPT® 93926 unilateral study) AND
  - Either CTA Lower extremity (CPT® 73706), or MRA Lower extremity (CPT® 73725).
  - CT or MRI Lower Extremity (contrast as requested) if requested by the operating surgeon

Evidence Discussion

Popliteal Artery Entrapment

Popliteal artery entrapment syndrome is a nonatheromatous cause of lower extremity ischemic symptoms. It is caused by abnormal embryologic development of the structures in the popliteal fossa. The symptoms are caused by compression of the popliteal artery by the muscles or fibrous bands. The typical presentation is claudication symptoms in physically active young men who have normal pulse examination.

Popliteal Artery Entrapment Syndrome is typically seen in young men (ages 20 to 40) but is not exclusive to this gender or age group.

Due to its unusual nature and presentation, ultrasound, CT Angiography or MR Angiography of the lower extremity is supported for diagnosis. Since it is critical to understand the anatomic relationships of the muscles to the vessels in the popliteal fossa, CT or MRI of the lower extremity is supported if requested by the surgeon for pre-operative planning.

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Arterial Imaging for Free Flaps in Reconstructive Surgery (PVD-7.5)

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Indications

- Breast reconstruction preoperative planning: See **Breast Reconstruction (BR-3)** in the Breast Imaging Guidelines
- Head and neck reconstruction: CTA or MRA unilateral lower extremity (CPT® 73706 or 73725) of the harvest site is medically necessary to evaluate perforator anatomy for planned fibular flap
  - Bilateral imaging is medically necessary when requested from the operating surgeon to select harvest site

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Arteriovenous Malformations (AVMs) (PVD-9.1)

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- See **Pulmonary AVMs (CH-24.1)** in the Chest Imaging Guidelines
- See **Arteriovenous Malformations (AVMs) and Related Lesions (HD 12.2)** in the Head Imaging Guidelines
- See **Arteriovenous Malformations (AVMs) and Fistulas (PEDPVD-2.5)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines
- See **Pelvic Pain/Dyspareunia, Female (PV-11.1)** in the Pelvis Imaging Guidelines

Initial imaging

- Ultrasound with Doppler is medically necessary as an initial examination for superficial lesions in the limbs.
  - Large lesion characterization may be limited by ultrasound imaging window.
  - Ultrasound is also limited in evaluating AVM relationship to airway or bony structures.

Evaluation and surveillance

- MRI without contrast or without and with contrast of the affected body part is the study of choice for abdominal AVMs and deep tissue (below the skin) AVM's in the limbs.
- MRA (contrast as requested) of the affected body part is medically necessary for evaluation and surveillance of known AVMs.
- It is unusual for both MRI and MRA to be necessary for routine treatment response or surveillance imaging of AVMs, but both may be approved for preoperative planning.
- CT and CTA can also be used to characterize AVMs and their relationship to normal structures but is generally not better than MRI and has associated radiation risks.
  - CT with contrast and/or CTA (contrast as requested) of the affected body part is medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Post-embolization

- Advanced imaging is medically necessary one-time post-embolization to evaluate for successful resolution of the AVM.
- Additional imaging (same study performed pre-procedure or as requested by the treating provider) is medically necessary for treatment planning purposes if resolution of the AVM was not achieved.

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Background and Supporting Information

Arteriovenous malformations are characterized by a network of multiple abnormal vascular channels interposed between enlarged feeding arteries and draining veins. The arteriovenous fistula has a single communication interposed between a feeding artery and a draining vein. The normal capillary bed is absent in both lesions. Both lesions may have an aggressive clinical course and are characterized by a reddish pulsatile mass which has a thrill or bruit. Though often recognized at birth, these lesions may grow and present near adolescence.

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Complications

Guideline

Vascular Graft Infection (PVD-21)
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Suspected Retroperitoneal Bleed (PVD-20)
References

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Vascular Graft Infection (PVD-21)

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Suspected or Known Vascular Graft Infection

Clinical concern for vascular graft infection

Imaging of the affected vascular segment is considered medically necessary for thediagnosis of suspected vascular graft infection and/or treatment planning for known vascular graft infection as follows:

- Duplex arterial ultrasound and/or CTA or MRA of the affected arterial segment.
- CTA or MRA of proximal and/or distal vascular beds of the affected segment for operative planning as requested by the surgeon (or provider in consultation with the performing surgeon).
- Indium 111 (111In)–labeled white blood cell (WBC) or Gallium-67 citrate studies (CPT® 78800, CPT® 78801, CPT® 78802, or CPT® 78803)
- PET/CT (CPT® 78815) for clinical suspicion of aortic infection (graft or native aorta) when **both** of the following apply:
  - CT-angiogram is equivocal/indeterminate AND
  - Indium-111 or Gallium-67 studies have not been performed **and** are not available or not technically feasible.

Evidence Discussion

Infection of vascular grafts is uncommon but serious and potentially life-threatening. Clinical presentations are variable, and imaging may be necessary to identify and/or define extent of infection, as well as for surgical planning.

Ultrasound is an inexpensive imaging modality that may identify findings associated with infection, such as fluid collections and pseudoaneurysm. This method is a considered a reasonable first-line modality to image extracavitary infection, but utility in evaluating intracavitary grafts and associated infection is limited.

CTA is a commonly used modality to evaluate intracavitary grafts, but is also useful in evaluating extracavitary graft, especially for surgical planning. Disadvantages include contrast exposure, and limited sensitivity/specificity.

MRI may also be useful in evaluating vascular grafts and allows for good soft tissue resolution. Disadvantages include generally higher cost than CT scan, longer acquisition

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times, and limitations in use when patients have intracardiac devices and/or renal disease.

WBC scintigraphy, as well as PET/CT scan, allow for more definitive identification of infection, compared with CT scans. WBC scintigraphy may be limited due to the time needed for the study; while PET/CT may be limited due to low specificity.

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Suspected Retroperitoneal Bleed (PVD-20)

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Imaging

Any of the following studies is considered medically necessary to evaluate for retroperitoneal bleed:

- CT Abdomen and Pelvis with IV contrast (CPT® 74177)
- CT Abdomen and Pelvis with and without IV contrast (CPT® 74178)
- CTA Abdomen and Pelvis (CPT® 74174)

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Venous Procedures

Guideline

Imaging for Hemodialysis Access (PVD-8)
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IVC filters – Treatment (PVD-16.2)
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Post iliac vein stenting/angioplasty (PVD-17.1)
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Imaging for Hemodialysis Access (PVD-8)

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Arterial Evaluation and Venous Mapping Prior to AV Fistula (PVD-8.1)

- Imaging prior to AV fistula creation is medically necessary:
  - For vessel mapping CPT® 93985 or 93986
  - MRA Upper Extremity (CPT® 73225) may be needed if duplex imaging is equivocal
- Arterial evaluation to assess arterial suitability (size, degree of stenosis and calcification) prior to AV fistula creation is medically necessary as follows:
  - CPT® 93930 or CPT® 93931 can be used to report upper extremity arterial evaluation
- Venous mapping (CPT® 93970 or CPT® 93971) is medically necessary to assess venous suitability prior to AV fistula creation

Hemodialysis access imaging (PVD-8.2)

- Indications for Duplex ultrasound (CPT® 93990) of hemodialysis access include but are not limited to:
  - Individuals with decreased flow rates during hemodialysis.
  - Development of arm swelling or discomfort after access placement surgery or a hemodialysis session.
  - Prolonged immaturity of a surgically created AV fistula.
  - Suspected pseudoaneurysm.
  - Suspected AV fistula or graft stenosis.
  - Known or suspected fluid collection adjacent to an AV fistula or graft.
  - One Duplex US (CPT® 93990) can be performed after a surgically created AV fistula for assessment, although it is not generally needed.
- Central venous stenosis can cause new dialysis access to fail to mature or cause the premature failure of existing fistulas/grafts.
- CT Chest with contrast (CPT® 71260), or CTA Chest (CPT® 71275), or MRA Chest (CPT® 71555) is medically necessary when there is documentation of either:
  - Signs and symptoms of central venous stenosis including:
    - Arm swelling
    - Presence of numerous collateral veins

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    - Prolonged bleeding from dialysis puncture sites
  - A history of pacemaker placement or previous tunneled dialysis graft, regardless of signs and symptoms.

Evidence Discussion

Hemodialysis Access for creation and maintenance

Hemodialysis access imaging is required to assess options for creation of hemodialysis access as well as to evaluate for maturation, failure and complications related to the access and outflow central veins.

Prior to creation of a native arteriovenous fistula (AVF), venous duplex ultrasound should be performed of both upper extremities to assess for adequate vein for fistula creation on all patients. Vessel mapping should include arterial inflow assessment to assess size, degree of stenosis and areas of calcification that may exclude access creation. Advanced imaging (CT or MR) of the chest or upper extremity may be indicated to further assess abnormalities identified on duplex imaging or evidence of central venous outflow obstruction.

Duplex evaluation of hemodialysis access should be performed for patients with evidence of failed maturation, poor function of access or complication related to the use of the hemodialysis access by history, physical exam or functional parameters during dialysis. These parameters may include: elevated venous pressures, inefficient dialysis, and recirculation greater than 10-15% or decreased flow.

CT or MR of the chest may be indicated for patients with a history of central venous catheters or pacemaker/ICD wires, signs and symptoms of ipsilateral central venous stenosis including arm swelling, venous collaterals or prolonged bleeding after dialysis.

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IVC filters – Treatment (PVD-16.2)

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- Prior to IVC filter insertion
  - An initial venous duplex is medically necessary to assess for the presence of thrombus in the femoral vein which would affect the approach (transjugular or transfemoral)
- For consideration of removal, CT or CTA Abdomen and Pelvis (CPT® 74177 or, 74174) is medically necessary for **any** of the following:
  - A KUB demonstrates tilting of the filter or malposition of one of the filter thongs
  - New bilateral lower extremity swelling (venous duplex should be performed first)
  - Filter present for >12 months, with documentation stating intent to remove

Background and Supporting Information

- IVC filters are placed in individuals with known DVT that cannot be anti-coagulated, individuals with poor pulmonary reserve and high risk for DVT, or prophylaxis in trauma and surgical individuals.
- Most IVC filters inserted are retrievable and should be removed as soon as clinically feasible. After 12 months, removal of IVC filters can become technically more difficult.

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Post iliac vein stenting/angioplasty (PVD-17.1)

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Iliac venous stents can be placed after thrombolysis for DVT associated with May-Thurner's syndrome, DVT associated with extrinsic compression or for post thrombotic iliac obstruction.

- Arterial duplex (CPT® 93975, 93976, 93978, 93979) are medically necessary for:
  - Surveillance of iliac venous stents
  - Worsening signs or symptoms including increased edema when stent malfunction is suspected
  - Post-operatively within the first month, at six months, twelve months and then annually
- CTV or MRV Abdomen and Pelvis are medically necessary for an abnormal or indeterminate duplex

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Venous Imaging

Guideline

Upper Extremity Venous – Imaging (PVD-4.2)
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Acute Limb Swelling (PVD-12)
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Upper Extremity Venous – Imaging (PVD-4.2)

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Venous Insufficiency

- For symptoms of venous insufficiency including but not limited to unilateral pain and swelling of the upper extremity
  - Venous duplex upper extremities (CPT® 93970 or CPT® 93971) is medically necessary initially
  - If duplex ultrasound is non-diagnostic, the following studies are medically necessary:
    - MRV Upper extremity (CPT® 73225) and/or MRV Chest (CPT® 71555), or
    - CTV Upper extremity (CPT® 73206) and/or CTV Chest (CPT® 71275)
      - For venous thoracic outlet syndrome, CTV Upper extremity (CPT® 73206) or MRV Upper extremity (CPT® 73225), and/or CTV Chest (CPT® 71275) or MRV Chest (CPT® 71555) .
- CT Chest with contrast (CPT® 71260) is medically necessary for Superior Vena Cava Syndrome (upper extremity and facial symptoms).
- **Either** of the following is medically necessary when stenting of the SVC is being considered:
  - MRV Chest (CPT® 71555)
  - CTV Chest (CPT® 71275)

Lymphedema of the Upper extremity

When initial imaging with venous duplex has ruled out DVT, **either** of the following studies are medically necessary to evaluate for suspected lymphedema of the upper extremity:

- Lymphoscintigraphy (CPT® 78195)
- MR lymphangiography (CPT® 75801)

Evidence Discussion

Upper Extremity Venous Imaging

Duplex ultrasound is the initial imaging modality for assessment of patients with symptoms of upper extremity venous occlusive disease or venous insufficiency including arm edema, pain and ulceration. Duplex imaging is limited in assessment of the proximal subclavian vein and central veins due to anatomic interference by the rib

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cage and lungs. MRV or CTV is medically necessary to assess these more proximal segments of the venous outflow and central venous structures including the innominate veins and SVC.

Advanced imaging is medically necessary for treatment planning in SVC syndrome and proximal venous intervention. Follow-up imaging after PTA and/or stenting for SVC syndrome or proximal vein occlusive disease may be approved assessment of stent patency and for recurrent symptoms.

SVC syndrome is caused by acute or subacute, intrinsic or extrinsic obstruction of the SVC, (ex: lung cancer, fibrosis, indwelling catheters/devices, thrombus). Other symptoms include dyspnea, headache and dizziness.

Thoracic Outlet Syndrome (TOS) refers to compression of the neurovascular structures within the thoracic outlet as they pass from the neck and thorax to the axilla. There are three types of TOS, neurogenic (see Brachial plexus-PN 4.1), venous and arterial (see Upper Extremity PVD-PVD 4.1). Venous TOS typically occurs in young athlete after a history of exertion of the limb, or in the presence of a central venous catheter which traverses the subclavian vein.

Lymphedema is caused by dysfunction of the lymphatic system. Primary lymphedema is a result of a congenital malformation of the lymphatic system, whereas secondary lymphedema is due to an exogenous cause that impairs the lymphatic system. Typical etiologies include trauma, chronic infection, cancer treatments including lymph node dissections and radiation therapy. Most cases of secondary lymphedema of the upper extremity are related to breast cancer and it's treatment. Due to the anatomic disruption of the lymphatic system, there is accumulation of protein-rich interstitial fluid, persistent swelling and later fibro-adipose hypertrophy.

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Acute Limb Swelling (PVD-12)

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Superficial venous thrombosis (SVT) (PVD-12.1)

- The diagnosis of superficial venous thrombosis is generally made on the basis of physical examination.
  - Duplex ultrasound (CPT® 93970, CPT® 93971) is medically necessary as the initial imaging if the diagnosis is equivocal
  - Follow-up duplex ultrasound (CPT® 93970, CPT® 93971) is medically necessary only if thrombus in the superficial systems is encroaching onto the deep venous system (saphenofemoral or saphenopopliteal junction)

Background and Supporting Information

Superficial venous thrombosis (SVT) refers to acute or chronic thrombosis of the superficial veins in both the upper (cephalic and basilic veins) and lower extremities (greater [great] saphenous vein, lesser [small] saphenous vein). Treatment: Elevation and warm compresses until pain and swelling subsides.

Acute deep venous thrombosis (DVT) (PVD-12.2)

- Duplex ultrasound (CPT® 93970 bilateral study or CPT® 93971 unilateral study) is medically necessary as the initial imaging study for any suspected DVT
  - Deep venous thrombosis can present as
    - Symptomatic
      - Swelling
      - Pain
      - Warmth
      - Erythema
      - Pain with dorsiflexion of the foot (Homan's Sign)
      - Or with progression, such as phlegmasia cerulean dolens
      - Risk factors for DVT include age >40, obesity, malignancy, prolonged immobilization, hypercoagulability as well as those outlined in **Pulmonary Embolism (PE) (CH-25)** in Chest Imaging Guidelines.
- CTA/CTV Abdomen and pelvis with contrast are medically necessary to rule out IVC thrombus secondary to the filter when there is acute bilateral lower extremity swelling in an individual with a history of an IVC filter in place.

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- When there is concern for proximal DVT (iliofemoral):
  - Focused abdominal duplex can generally visualize the iliac veins and IVC to determine the absence or presence of iliac vein thrombus in an individual. If the results are equivocal or indeterminate:
    - CTV or MRV Abdomen and Pelvis with contrast (CPT® 74174 or CPT® 74185 and 73725) is medically necessary.
- For request concerning abdominal vein thrombosis, see **Abdominal Veins other than Hepatic and Portal Veins (AB-43.2)** in the Abdomen Imaging Guidelines
- For proximal DVT's (iliac vein DVT's or in cases of phlegmasia (extensive DVT compromising arterial inflow), thrombectomy (rarely performed) or thrombolysis is medically necessary.
- If the cause of the DVT is found to be due to May-Thurner, see also **May-Thurner Syndrome (PVD-13.3)**

Background and Supporting Information

Deep venous thrombosis is characterized by thrombosis of a deep vein in either the upper (brachial, axillary, subclavian veins) or the lower extremity (soleus muscle veins, gastrocnemius muscle veins, peroneal, posterior tibial, popliteal, femoral or iliac veins).

Follow-up imaging of known DVT (PVD-12.3)

- A repeat duplex ultrasound (CPT® 93970 bilateral study or CPT® 93971 unilateral study) is medically necessary in order to rule out proximal extension of a calf vein DVT in individuals who cannot be anticoagulated as follows:
  - One week after the initial diagnosis.
  - Serial imaging (up to 3 studies) over the first three weeks if calf DVT is not treated.
- Evidence does not support the use of imaging during long-term anticoagulation therapy to determine venous recanalization or for the purpose of determining whether or not to continue anticoagulation therapy.

Follow-up imaging after venous surgery (PVD-12.4)

- Venous duplex (CPT® 93971 unilateral study) of the treated limb is medically necessary to rule out a DVT within seven days of endovenous ablation.
- Follow-up routine imaging is **not** medically necessary after other venous procedures including:
  - Saphenous vein ligation and stripping
  - Phlebectomy
  - Sclerotherapy

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Generalized bilateral lower extremity edema (PVD-12.5)

Bilateral lower extremity edema is multifactorial. Prior to any request for advanced imaging, a workup for causes of the edema should be instituted including echocardiogram to rule out congestive heart failure and laboratory studies to exclude renal insufficiency and liver disease. The following imaging is medically necessary based on the suspected cause of the edema:

- Suspected abdominal or pelvic pathology
  - Abdominal ultrasound or duplex is the initial imaging
  - CT Abdomen and Pelvis or CT pelvis either with or without contrast can be performed if abdominal US is equivocal or indeterminate
- Suspected chronic venous insufficiency
  - A venous duplex CPT® 93970 (bilateral) or CPT® 93971 (unilateral) is medically necessary to evaluate for venous reflux.
- Suspected lymphedema
  - When initial noninvasive studies, such as ultrasound, are negative for venous valvular insufficiency **either** of the following advanced imaging studies is indicated:
    - Lymphoscintigraphy (CPT® 78195)
    - MRI lymphangiography (CPT® 73718)

Unilateral lower extremity edema (PVD-12.6)

Initial imaging for unilateral lower extremity edema with duplex ultrasound (CPT® 93971, unilateral study) is medically necessary.

- If there is concern for proximal DVT:
  - focused abdominal duplex to evaluate the iliac veins and IVC
  - CTV or MRV Abdomen and Pelvis (CPT® 74174 or CPT® 74185 and CPT® 72198) for indeterminate or equivocal duplex results.
- If there is concern for abdominal or pelvic pathology:
  - Abdominal/pelvic ultrasound or duplex
  - CT Abdomen and Pelvis, or CT Pelvis with or without contrast if the abdominal ultrasound is indeterminate or inconclusive

Background and Supporting Information

Unilateral edema favors localized causes of venous or lymphatic compromise, rather than systemic etiologies which tend to result in bilateral edema. Initial imaging is duplex ultrasound. This can assess for vascular and non-vascular causes, including DVT, venous reflux, popliteal cysts, hematoma, mass.

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Chronic Limb Swelling

Guideline

Chronic limb swelling due to chronic deep venous thrombosis (DVT)/May-Thurner syndrome (PVD-13)
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Chronic limb swelling due to venous insufficiency/Venous stasis changes/Varicose veins (PVD-14)
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Chronic limb swelling due to chronic deep venous thrombosis (DVT)/May-Thurner syndrome (PVD-13)

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Chronic DVT with incompletely lysed or residual DVT (PVD-13.1)

Individuals with incompletely lysed or residual DVT can develop post-thrombotic syndrome that can be characterized as chronic edema, venous stasis changes, pain and in advanced cases venous stasis ulceration.

- Imaging is medically necessary to evaluate for iliac venous obstruction from incompletely lysed thrombus in individuals with a history of proximal (iliofemoral) DVT who have developed post thrombotic syndrome.
  - Initial imaging is duplex (CPT® 93970 bilateral study or CPT® 93971 unilateral study)
  - Either a CTV or MRV Abdomen and Pelvis (CPT® 74174) or (CPT® 74185 and 72198) OR CTV Pelvis (CPT® 72191) or MRV Pelvis (CPT® 72198), or venography for treatment planning purposes.
- Imaging for post-thrombotic syndrome is only medically necessary for **either**:
  - Signs and symptoms suggestive of a new acute DVT
  - Preoperative planning for iliac vein/stenting for suspected iliac vein stenosis or occlusion

Background and Supporting Information

- Chronic deep venous thrombosis is defined as an acute DVT that is greater than 14 days old.
- Incompletely lysed DVT can cause luminal narrowing of the vein restricting venous outflow leading to stenosis or occlusion and /or can lead to valve dysfunction resulting in reflux of venous blood retrograde towards gravity. Both pathologies ultimately lead to chronic edema which can cause chronic pain and venous stasis disease.
  - The mainstay of treatment for chronic deep venous thrombosis is compression stockings
  - Selected individuals may be a candidate for iliac vein angioplasty/stenting.

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Post-thrombotic syndrome (PVD-13.2)

- Imaging for post-thrombotic syndrome is medically necessary when:
  - There are signs and symptoms suggestive of a new acute DVT
  - For preoperative planning for iliac vein/stenting in the setting of known iliac venous obstruction in those with a history of a proximal (iliofemoral) DVT.
- Imaging for post-thrombotic syndrome is NOT medically necessary for chronic swelling that has not changed in severity or character

May-Thurner syndrome (PVD-13.3)

- CTV or MRV Abdomen and Pelvis (CPT® 74174, or 74185 and 72198) OR CTV Pelvis or MRV Pelvis (CPT® 72191 or 72198) is medically necessary in individuals with a history of **one** of the following:
  - Left lower extremity iliac DVT
  - Persistent left lower extremity edema OR varicose veins OR venous stasis ulcer despite treatment of superficial venous disease in that extremity
  - Persistent left lower extremity edema OR varicose vein OR venous stasis ulcer in the absence of saphenous vein reflux.
- Imaging and/or prophylactic treatment of May-Thurner syndrome, in the absence of acute or chronic DVT **or** chronic left lower extremity edema and its sequelae such as varicose veins or venous stasis ulcers, is **not** considered medically necessary

Background and Supporting Information

In approximately 25% of people, the right iliac artery overlies the left iliac vein over the fifth lumbar vertebrae and its pulsations can compress the vein increasing the risk of DVT in the left extremity.

- Treatment is with iliac vein angioplasty/stenting

Pelvic congestion syndrome (PVD-13.4)

- Signs and symptoms of pelvic congestion syndrome include:
  - Chronic pelvic pain OR post-coital discomfort >6 months.
  - Associated symptoms can include the presence of labial varicosities or heavy menstrual periods.
- Initial imaging is via transvaginal or pelvic ultrasound to exclude other pathologies of chronic pelvic pain
- CTV or MRV Abdomen and Pelvis (CPT® 74174, or 74185 and 72198) is medically necessary if initial ultrasound is inconclusive or non-diagnostic.

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Evidence Discussion

Chronic limb swelling due to chronic deep venous thrombosis (DVT)/ May-Thurner syndrome, post-thrombotic syndrome and pelvic congestion syndrome

For patients with isolated left leg edema, or left leg edema greater than right leg, duplex ultrasound of the extremity should be performed to assess for DVT. If femoral DVT is identified, duplex ultrasound of the pelvis should be performed to assess for iliocaval DVT or evidence of a mass that may be causing compression of the iliac vein or IVC.

In patients with acute iliofemoral DVT, advanced imaging with either CTV or MRV of the abdomen and or pelvis is medically necessary for cases of iliofemoral DVT identified on ultrasound for treatment planning, or cases where iliofemoral DVT or stenosis is suspected but pelvic ultrasound is indeterminate or limited due to body habitus or overlying bowel gas obscuring the iliac vein.

Advanced imaging with CTV or MRV of the abdomen and/or pelvis is medically necessary in patients without acute DVT but a prior history of left iliac DVT or leg edema, varicosities or venous stasis disease in the absence of underlying venous insufficiency or following treatment of superficial venous insufficiency to assess for iliac vein compression. Post-Thrombotic syndrome due to prior DVT and subsequent deep venous insufficiency may lead to recurrent or chronic lower extremity edema. Management is compression therapy and advanced imaging is only indicated for assessment of symptoms consistent with new iliofemoral DVT or for preoperative planning for identified iliac vein stenosis.

Advanced imaging with CTV or MRV is medically necessary in patients with recurrent symptoms and a history of iliocaval stenting for iliac vein DVT, or compression.

Pelvic congestion syndrome may be a cause of chronic pelvic pain or post-coital discomfort. Initial imaging is pelvic or transvaginal ultrasound to assess for other sources of pelvic pain. Advanced imaging such as CTV or MRV of the abdomen and pelvis is medically necessary if ultrasound is inconclusive or non-diagnostic or for treatment planning.

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Chronic limb swelling due to venous insufficiency/Venous stasis changes/ Varicose veins (PVD-14)

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Venous Reflux (PVD-14.2)

- A venous valvular insufficiency study (CPT® 93970 bilateral study or CPT® 93971 unilateral study) is medically necessary to diagnose presence of reflux in the greater saphenous vein as well as the size of the refluxing vein.
- A duplex ultrasound (CPT® 93970 bilateral study or CPT® 93971 unilateral study) is medically necessary within six months before treatment with intervention to demonstrate the presence of pathologic reflux (>500ms) within the greater and lesser saphenous veins and document vein size.
- A post-ablation venous ultrasound (CPT® 93970 bilateral study or CPT® 93971 unilateral study) is medically necessary within seven days post-procedure.
  - If thrombus is noted within the saphenofemoral junction, repeat imaging can be performed within seven days to assess for propagation into the deep system.
- Post-procedure assessment by imaging techniques is not medically necessary to confirm efficacy or outcome of the procedure.

Evidence Discussion

Venous insufficiency–– General information

- Venous insufficiency is characterized by failure of the venous blood to flow in its normal antegrade path of flow and instead reflux backwards by the force of gravity usually secondary to malfunction of the venous valves.
- Risk factors include previous DVT, obesity, female sex assigned at birth, hereditary, and environmental factors such as prolonged standing on a hard surface.
- Venous insufficiency loosely includes the diagnosis of venous reflux, varicose veins, venous stasis ulcers and spider/reticular veins.
- Diagnosis is made with a venous valvular insufficiency study which documents the presence of reflux (>500ms) in the greater saphenous vein as well as the size of the refluxing vein (3-15mm).
- Treatment of superficial venous reflux is amenable to intervention in selected individuals who are symptomatic and have failed conservative therapy.

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- Ultrasound mapping or monitoring techniques are considered medically necessary only to initially determine the extent and configuration of symptomatic varicosities or valvular insufficiency.

Venous reflux

- Symptoms of venous reflux include chronic edema, pain, and venous stasis ulcerations. Symptoms of venous reflux can be ameliorated with compression therapy with graded compression stockings, elevation, avoidance of prolonged standing and weight loss. Venous reflux can be seen in both the deep and superficial venous systems. Reflux within the deep system is not amenable to intervention.
- Treatment of deep venous reflux is via active compression with compression stocks, pneumatic pumps or specialized dressings such as Unna boots.
- Treatment of symptomatic superficial venous reflux is via endovenous laser radiofrequency ablation of the greater or lesser saphenous vein resulting in closure of the vein allowing for venous blood to be rerouted to the deep venous system.
- Treatment of symptomatic superficial venous reflux can also be treated via saphenous vein ligation and stripping which has fallen out of favor but can be performed for a tortuous or enlarged (>15mm) greater or lesser saphenous vein. One complication of endovenous ablation is deep venous thrombosis.

Varicose Veins

- If the varicosities remain symptomatic despite conservative therapy, varicose veins are treated with sclerotherapy or phlebectomy generally on the basis of size.
- Varicose veins are defined as enlarged, tortuous veins visible under the skin. Symptoms associated with varicose veins include achiness and heaviness of the legs and pain/discomfort over the varicosities. Varicose veins can exist both in the absence and presence of venous reflux.
- Treatment involves conservative therapy such as compression stockings, avoidance of prolonged standing, intermittent elevation, weight loss (if applicable) and exercise which relieves the distention of the varicose veins ameliorating the symptoms.

Spider veins/reticular veins

- Spider veins are formed by the dilation of a cluster of blood vessels within the dermis – generally <3mm in diameter. Diagnosis is via physical examination. Spider veins are usually asymptomatic but can cause aching, burning and tenderness in the area overlying the abnormal veins. Spider veins can exist in the absence or presence of venous reflux. The presence of spider veins should not be an indication for treatment of venous reflux.
- Treatment of spider veins is generally cosmetic except in certain cases and can be treated with sclerotherapy.

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General Guidelines (SP-1)

Guideline

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Procedure Codes Associated with Spine Imaging

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MRI/MRA	CPT®
MRI Cervical without contrast	72141
MRI Cervical with contrast	72142
MRI Cervical without and with contrast	72156
MRI Thoracic without contrast	72146
MRI Thoracic with contrast	72147
MRI Thoracic without and with contrast	72157
MRI Lumbar without contrast	72148
MRI Lumbar with contrast	72149
MRI Lumbar without and with contrast	72158
MRA Spinal Canal	72159
MRI Pelvis without contrast	72195
MRI Pelvis with contrast	72196
MRI Pelvis without and with contrast	72197
MR Spectroscopy	76390

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MRI/MRA	CPT®
Magnetic resonance spectroscopy, determination and localization of discogenic pain (cervical, thoracic, or lumbar); acquisition of single voxel data, per disc, on biomarkers (ie, lactic acid, carbohydrate, alanine, laal, propionic acid, proteoglycan, and collagen) in at least 3 discs	0609T
Magnetic resonance spectroscopy, determination and localization of discogenic pain (cervical, thoracic, or lumbar); transmission of biomarker data for software analysis	0610T
Magnetic resonance spectroscopy, determination and localization of discogenic pain (cervical, thoracic, or lumbar); postprocessing for algorithmic analysis of biomarker data for determination of relative chemical differences between discs	0611T
Magnetic resonance spectroscopy, determination and localization of discogenic pain (cervical, thoracic, or lumbar); interpretation and report	0612T

CT	CPT®
CT Cervical without contrast	72125
CT Cervical with contrast (Post-Myelography CT)	72126
CT Cervical without and with contrast	72127
CT Thoracic without contrast	72128
CT Thoracic with contrast (Post-Myelography CT)	72129
CT Thoracic without and with contrast	72130
CT Lumbar without contrast (Post-Discography CT)	72131
CT Lumbar with contrast (Post-Myelography CT)	72132
CT Lumbar without and with contrast	72133

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CT	CPT®
CT Pelvis without contrast	72192
CT Pelvis with contrast	72193
CT Pelvis without and with contrast	72194

Ultrasound	CPT®
Spinal canal ultrasound	76800

Nuclear Medicine	CPT®
Bone Marrow Imaging, Limited	78102
Bone Marrow Imaging, Multiple	78103
Bone Marrow Imaging, Whole Body	78104
Bone or Joint Imaging, Limited	78300
Bone or Joint Imaging, Multiple	78305
Bone Scan, Whole Body	78306
Bone Scan, 3 Phase Study	78315
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, single area (e.g., head, neck, chest, pelvis), single day imaging	78800
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, 2 or more areas (e.g., abdomen and pelvis, head, and chest), 1 or more days imaging or single area imaging over 2 or more days	78801

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Nuclear Medicine	CPT®
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, single day imaging	78802
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), single area (e.g., head, neck, chest, pelvis), single day imaging	78803
Radiopharmaceutical localization of a tumor, inflammatory process, or distribution of radiopharmaceutical agents, including vascular flow and blood pool imaging when performed, planar, whole body, requiring two or more days of imaging	78804
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (e.g., head, neck, chest, pelvis), single day imaging	78830
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), minimum 2 areas (e.g., pelvis and knees, abdomen and pelvis), single day imaging, or single area imaging over 2 or more days	78831
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired CT transmission scan for anatomical review, localization, and determination/detection of pathology, minimum 2 areas (e.g., pelvis and knees, abdomen and pelvis), single day imaging, or single area imaging over 2 or more days	78832
Radiopharmaceutical quantification measurement(s) single area (list separately in addition to code for primary procedure)	78835

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General Guidelines (SP-1.0)

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- Before advanced diagnostic imaging can be considered, there must be an in-person clinical evaluation as well as a clinical re-evaluation after a trial of failed conservative therapy; the clinical re-evaluation may consist of an in-person evaluation or other meaningful contact with the provider's office such as email, web, telephone communications, or clinical documentation from a provider.
- An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation.
- The in-person clinical evaluation should include a relevant history and physical examination (including a detailed neurological examination), appropriate laboratory studies, non-advanced imaging modalities, results of manual motor testing, the specific dermatomal distribution of altered sensation, reflex examination, and nerve root tension signs (e.g., straight leg raise test, slump test, femoral nerve tension test). *The clinical evaluation must be in-person; other forms of meaningful contact (telephone call, electronic mail, telemedicine, or messaging) are not acceptable as an in-person evaluation.*
  - For those spinal conditions/disorders for which the Spine Imaging Guidelines require a plain x-ray of the spine prior to consideration of an advanced imaging study, the plain x-ray must be performed after the current episode of symptoms started or changed and results need to be available to the requesting provider of the advanced imaging study (see: **Anatomic Guidelines [SP-2.1]**).
- Clinical re-evaluation is required prior to consideration of advanced diagnostic imaging to document failure of clinical improvement following a six-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed. Clinical re-evaluation can include documentation of an in-person encounter with a provider or documentation of other meaningful contact with a provider's office by the individual (e.g., telephone call, electronic mail, telemedicine, or messaging).
  - Provider-directed treatment may include education, activity modification, NSAIDs (non-steroidal anti-inflammatory drugs), narcotic and non-narcotic analgesic medications, oral or injectable corticosteroids, a provider-directed home exercise/stretching program, cross-training, avoidance of aggravating activities, physical/occupational therapy, spinal manipulation, interventional pain procedures and other pain management techniques.
- Any bowel/bladder abnormalities or emergent or urgent indications should be documented at the time of the initial clinical evaluation and clinical re-evaluation.

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- Altered sensation to pressure, pain, and temperature should be documented by the specific anatomic distribution (e.g., dermatomal, stocking/glove or mixed distribution).
- Motor deficits (weakness) should be defined by the specific myotomal distribution (e.g., weakness of toe flexion/extension, knee flexion/extension, ankle dorsi/plantar flexion, wrist dorsi/palmar flexion) and gradation of muscle testing should be documented as follows:

Grading of Manual Muscle Testing	
0	No muscle activation
1	Trace muscle activation, such as a twitch, without achieving full range of motion
2	Muscle activation with gravity eliminated, achieving full range of motion
3	Muscle activation against gravity, full range of motion
4	Muscle activation against some resistance, full range of motion
5	Muscle activation against examiner's full resistance, full range of motion

- Pathological reflexes (e.g., Hoffmann's, Babinski, and Chaddock sign) should be reported as positive or negative.
- Asymmetric reflexes and reflex examination should be documented as follows:

Grading of Reflex Testing	
0	No response
1+	A slight but definitely present response
2+	A brisk response
3+	A very brisk response without clonus
4+	A tap elicits a repeating reflex (clonus)

- Advanced diagnostic imaging is often urgently indicated and may be necessary if serious underlying spinal and/or non-spinal disease is suggested by the presence of certain individual factors referred to as "red flags." See: **Red Flag Indications (SP-1.2)**.

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- Spinal specialist evaluation can be helpful in determining the need for advanced diagnostic imaging, especially for individuals following spinal surgery.
- The need for repeat advanced diagnostic imaging should be carefully considered and may not be medically necessary if prior advanced diagnostic imaging has been performed. Requests for simultaneous, similar studies such as spinal MRI and CT need to be documented as required for pre-operative surgical planning. These studies may be helpful in the evaluation of complex failed spinal fusion cases or needed for pre-operative surgical planning when the determination of both soft tissue and bony anatomy is required.
- Serial advanced imaging, whether CT or MRI, for surveillance of healing or recovery from spinal disease is not supported by the currently available scientific evidence-based medicine for the majority of spinal disorders.
  - Requests for repeat imaging may be considered on a case-by-case basis (e.g., concern for delayed union or non-union of spinal fracture, pseudoarthrosis of fusion, etc.)
- Advanced imaging is generally unnecessary for resolved or improving spinal pain and/or radiculopathy.
- Advanced diagnostic imaging has not been shown to be of value in individuals with stable, longstanding spinal pain without neurological features or without clinically significant or relevant changes in symptoms or physical examination findings.
- Anatomic regions of the spine/pelvis that are included in the following MRI and CT advanced diagnostic imaging studies:
  - Cervical spine: from the skull base/foramen magnum through T1
  - Thoracic spine: from C7 through L1
  - Lumbar spine: from T12 through mid-sacrum
  - Pelvis: includes hips, sacroiliac joints, sacrum, coccyx
- CT or MRI of the cervical and thoracic spine will image the entire spinal cord since the end of the spinal cord or conus medullaris usually ends at L1 in adults. Therefore, lumbar spine imaging is not needed when the goal is to image only the spinal cord unless there is known or suspected low lying conus medullaris (e.g., tethered cord).

Health Equity Consideration

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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General Considerations (SP-1.1)

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- See: **General Guidelines (SP-1.0)**

Background and Supporting Information

Straight leg raise test (also known as the Lasegue's test) – With the individual in the supine position, the hip medially rotated and adducted, and the knee extended, the examiner flexes the hip until the individual complains of pain or tightness in the back or back of the leg. If the pain is primarily back pain, it is less specific whereas if the pain is primarily in the leg, it is more likely nerve root irritation/radiculopathy. Disc herniation or pathology causing pressure between the two extremes are more likely to cause pain in both areas. The examiner then slowly and carefully drops the leg back (extends it) slightly until the individual feels no pain or tightness. The individual is then asked to flex the neck so the chin is on the chest, or the examiner may dorsiflex the individual's foot, or both actions may be done simultaneously. Both maneuvers are considered to be provocative tests for neurological tissue.

Slump test – The individual is seated on the edge of the examination table with the legs supported, the hips in neutral position, and the hands behind the back. The examination is performed in sequential steps. First, the individual is asked to "slump" the back into thoracic and lumbar flexion. The examiner maintains the individual's chin in neutral position to prevent neck and head flexion. The examiner then uses one arm to apply overpressure across the shoulders to maintain flexion of the thoracic and lumbar spines. While this position is held, the individual is asked to actively flex the cervical spine and head as far as possible (i.e., chin to chest). The examiner then applies overpressure to maintain flexion of all three parts of the spine (cervical, thoracic, and lumbar) using the hand of the same arm to maintain overpressure in the cervical spine. With the other hand, the examiner then holds the individual's foot in maximum dorsiflexion. While the examiner holds these positions, the individual is asked to actively straighten the knee as much as possible. The test is repeated with the other leg and then with both legs at the same time. If the individual is unable to fully extend the knee because of pain, the examiner releases the overpressure to the cervical spine and the individual actively extends the neck. If the knee extends further, the symptoms decrease with neck extension, or the positioning of the individual increases the individual's symptoms, then the test is considered positive.

Femoral nerve tension test (also known as the prone knee bending test) – The individual lies prone while the examiner passively flexes the knee as far as possible so that the individual's heel rests against the buttock. At the same time, the examiner should ensure that the individual's hip is not rotated. If the examiner is unable to flex the

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individual's knee past 90 degrees because of a pathological condition in the hip, the test may be performed by passive extension of the hip while the knee is flexed as much as possible. The flexed knee position should be maintained for 45 to 60 seconds. Unilateral neurological pain in the lumbar area, buttock, and/or posterior thigh may indicate an L2 or L3 nerve root lesion. Pain in the anterior thigh indicates tight quadriceps muscles or stretching of the femoral nerve.

Hoffmann's sign – The examiner holds the individual's middle finger and briskly flicks the distal phalanx. A positive test is noted if the interphalangeal joint of the thumb of the same hand flexes.

Babinski's sign – The examiner runs a sharp instrument along the plantar surface of the foot from the calcaneus along the lateral border to the forefoot. A positive test occurs with extension of the great toe with flexion and splaying of the other toes. A negative test occurs with no movement of the toes at all or uniform bunching up of the toes.

Chaddock sign – The examiner strokes the lateral malleolus. A positive test occurs with extension of the great toe.

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Red Flag Indications (SP-1.2)

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Red Flag Indications are intended to represent the potential for life or limb threatening conditions. Red Flag Indications are clinical situations in which localized spine pain and associated neurological features are likely to reflect serious underlying spinal and/or non-spinal disease and warrant exception to the requirement for documented failure of six weeks of provider-directed treatment. Advanced diagnostic imaging of the symptomatic level is medically necessary and/or work-up for a non-spinal source of spine pain for Red Flag Indications.

- Red Flag Indications include:
  - Motor Weakness
  - Aortic Aneurysm or Dissection
  - Cancer
  - Cauda Equina Syndrome
  - Fracture
  - Infection
  - Severe Radicular Pain

Motor Weakness

(See: Grading of Manual Muscle Testing and Reflex Testing in **General Guidelines [SP-1.0]**)

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History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
Clinical presentation including one or more of the following: • New onset motor weakness of grade 3/5 or less of specified muscle(s) • New onset foot drop • New onset bilateral lower extremity weakness • Progressive objective motor/sensory/deep tendon reflex deficits on clinical re-evaluation	MRI of the relevant spinal level* without contrast **OR** MRI of the relevant spinal level* without and with contrast **The SI joint and sacrum are relevant levels of the spine and are captured with pelvic imaging*.

Aortic Aneurysm or Dissection

History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
• New onset of back and/or abdominal pain in an individual with a known AAA; ***or*** • Acute dissection is suspected.	Spine imaging is not medically necessary, see: **Aortic Disorders, Renal Vascular Disorders and Visceral Artery Aneurysms (PVD-6)** in the Peripheral Vascular Disease Imaging Guidelines

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Cancer

History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
There is clinical suspicion of spinal malignancy AND ONE or more of the following: • Night pain • Uncontrolled or unintended weight loss • Pain unrelieved by change in position • Age >70 years • Severe or worsening spinal pain	**ONE** of the following: • MRI of the relevant spinal level* without contrast **OR** • MRI of the relevant spinal level* without and with contrast **OR** • CT of the relevant spinal level* without contrast **OR** • CT Myelogram of the relevant spinal level* **The SI joint and sacrum are relevant levels of the spine and are captured with pelvic imaging*.
Individual with a known history of cancer with back pain raising suspicion of spinal malignancy	See: **Spinal/Vertebral Metastases (ONC-31.6)** in the Oncology Imaging Guidelines

Cauda Equina Syndrome

History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
Clinical presentation including one or more of the following: • Acute onset of bilateral sciatica • Perineal sensory loss (“saddle anesthesia”) • Decreased anal sphincter tone • New onset bowel/bladder incontinence • Otherwise unexplained acute urinary retention	MRI Lumbar Spine without contrast (CPT® 72148) **OR** MRI Lumbar Spine without and with contrast (CPT® 72158)

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Fracture

History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
• Clinical suspicion of a pathological spinal fracture. ◦ Advanced imaging is medically necessary after x-ray; no conservative treatment is needed.	See: **Spinal Compression Fractures (SP-11.1)** for appropriate imaging studies
• Clinical suspicion of a spinal fracture after trauma ◦ Advanced imaging is medically necessary after x-ray; no conservative treatment is needed.	See: **Neck (Cervical Spine) Trauma (SP-3.2)**, **Upper Back (Thoracic Spine) Trauma (SP-4.2)**, or **Low Back (Lumbar Spine) Trauma (SP-6.2)** for appropriate imaging studies
• Clinical suspicion of a spinal fracture related to ankylosing spondylitis or DISH ◦ Advanced imaging is medically necessary ***without*** x-ray or conservative treatment.	See: **Neck (Cervical Spine) Trauma (SP-3.2)**, **Upper Back (Thoracic Spine) Trauma (SP-4.2)**, **Low Back (Lumbar Spine) Trauma (SP-6.2)**, or **Inflammatory Spondylitis (SP-10.2)** for appropriate imaging studies

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Infection

History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
There is a clinical suspicion of spinal infection (e.g., disc space infection, epidural abscess, or spinal osteomyelitis) and one or more of the following: • Fever • History of IV drug use • Recent bacterial infection (UTIs, pyelonephritis, pneumonia) • Recent spinal intervention (e.g., surgery, pain injection, or stimulator implantation) • Immunocompromised states • Long term use of systemic glucocorticoids • Organ transplant recipient taking anti-rejection medication • Diabetes mellitus • HIV/AIDS • Chronic dialysis • Immunosuppressant therapy • Neoplastic involvement of the spine • Laboratory values indicative of infection (e.g., elevated WBC, ESR, CRP, positive cultures) • Decubitus ulcer or wound overlying spine • Abnormal x-ray or CT suspicious for infection	**ONE** of the following: • MRI of the relevant spinal level* without and with contrast **OR** • MRI of the relevant spinal level* without contrast **OR** • CT of the relevant spinal level* without IV contrast **OR** • CT of the relevant spinal level* with IV contrast **OR** • FDG-PET/CT whole-body when x-ray or CT are abnormal **AND** when MRI cannot be performed or is inconclusive **OR** • 3-phase bone scan complete spine **OR** • Bone SPECT or Bone SPECT/CT **OR** • Gallium scan whole body or limited spine **OR** • Gallium SPECT or SPECT/CT **OR** • Gallium scan whole body or limited spine** with SPECT or SPECT/CT **The SI joint and sacrum are relevant levels of the spine and are captured with pelvic imaging.* ***Any bone scan can be combined with any Gallium scan*

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History, Symptoms or Physical Exam Findings (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
There is a clinical suspicion of spinal infection (e.g., disc space infection, epidural abscess, or spinal osteomyelitis) and one or more of the following: • New neurologic deficit on physical examination • Cauda equina syndrome	**ONE** of the following: • MRI of the relevant spinal level* without and with contrast **OR** • MRI of the relevant spinal level* without contrast **OR** • CT of the relevant spine level* without IV contrast **OR** • CT of the relevant spinal level* with IV contrast **The SI joint and sacrum are relevant levels of the spine and are captured with pelvic imaging.*

Severe Radicular Pain

All the following must be present (In-person clinical evaluation for the current episode of the condition required)	Advanced Diagnostic Imaging
• Severe radicular pain in a specified spinal nerve root distribution (minimum 9/10 on the VAS); and • Documented significant functional loss at work or at home; and • Severity of pain unresponsive to a minimum of seven (7) days of provider-directed treatment; and • Treatment plan includes one of the following: ◦ Transforaminal epidural steroid injection (TFESI) at any level(s); or ◦ Interlaminar epidural steroid injection (ILESI) at the cervical or thoracic levels; or ◦ A plan for urgent/emergent spinal surgery; or ◦ A plan for an urgent/emergent referral to/consultation from a spine specialist (Interventional Pain physician or Spine Surgeon)	MRI of the relevant spinal level* without contrast **OR** MRI of the relevant spinal level* without and with contrast **The SI joint and sacrum are relevant levels of the spine and are captured with pelvic imaging.*

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Definitions (SP-1.3)

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- **Radiculopathy**, for the purpose of this policy, is defined as the presence of pain resulting in significant functional limitations (i.e., diminished quality of life and impaired, age-appropriate activities of daily living), dysaesthesia(s) or paraesthesia(s) reported by the individual in a specified dermatomal distribution of an involved named spinal root(s) and **ONE or MORE** of the following:
  - Loss of strength of specific named muscle(s) or myotomal distribution(s) or demonstrated on detailed neurologic examination (within the prior 3 months), concordant with nerve root compression of the involved named spinal nerve root(s).
  - Altered sensation to light touch, pressure, pin prick or temperature demonstrated on a detailed neurologic examination (within the prior 3 months) in the sensory distribution concordant with nerve root compression of the involved named spinal nerve root(s).
  - Diminished, absent, or asymmetric reflex(es) on a detailed neurologic examination (within the prior 3 months) concordant with nerve root compression of the involved named spinal nerve root(s).
  - Either of the following:
    - A concordant radiologist's interpretation of an advanced diagnostic imaging study (MRI or CT) of the spine demonstrating compression of the involved named spinal nerve root(s) or foraminal stenosis at the concordant level(s) (Performed within the prior 12 months).
    - Electrodiagnostic studies (EMG/NCV's) diagnostic of nerve root compression of the involved named spinal nerve root(s). (Performed within the prior 12 months).
- **Radicular pain** is pain which radiates to the upper or lower extremity along the course of a spinal nerve root, typically resulting from compression, inflammation and/or injury to the nerve root.
- **Radiculitis** is defined, for the purpose of this policy, as radicular pain without objective neurological findings.

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Evidence Discussion (SP-1)

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Prior to advanced imaging, it is critical to perform a detailed history and physical examination in the evaluation of an individual for spinal pathology.[22,23,24] Features of the clinical history and physical examination not only help in the formulation of a differential diagnosis but also influence decisions about diagnostic imaging.[3] Also, as the more common findings on imaging studies are often nonspecific and non-diagnostic, clinical history and exam findings play a crucial role.[32] These incidental findings may lead to unnecessary further diagnostic workup and additional negative downstream outcomes.

Multiple studies have shown most individuals with acute neck or back pain will improve with 6 weeks of conservative care;[9,25,26] however, conservative care would not be necessary for individuals with red flag indications.[15]

Risks associated with imaging include but are not limited to radiation exposure and contrast complications.[27,29] Studies have also linked the increase rate of imaging with the increase rate of surgery and also found early magnetic resonance imaging (MRI) had an eight-fold increased risk of surgery.[27,28] It should also be of note that routine repeat advanced imaging for many spinal conditions has been shown to have limited value.[4,30,31]

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Imaging Techniques (SP-2)

Guideline

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MRI of the Spine (SP-2.2)
CT of the Spine (SP-2.3)
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Anatomic Guidelines (SP-2.1)

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- Anatomic regions of the spine/pelvis that are included in the following MRI and CT advanced diagnostic imaging studies:
  - Cervical spine: from the skull base/foramen magnum through T1
  - Thoracic spine: from C7 through L1
  - Lumbar spine: from T12 through mid-sacrum
  - Pelvis: includes hips, sacroiliac joints, sacrum, coccyx
- CT or MRI cervical and thoracic spine will image the entire spinal cord since the end of the spinal cord or conus medullaris usually ends at L1 in adults. Therefore, lumbar spine imaging is not needed when the goal is to image only the spinal cord unless there is known or suspected low-lying conus medullaris (e.g., tethered cord).
- The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study for the following conditions:
  - See: **Spinal Compression Fractures (SP-11)**
  - See: **Lumbar Spine Spondylolysis/Spondylolisthesis (SP-8)**
  - See: **Inflammatory Spondylitis (SP-10.2)**
  - See: **Upper Back (Thoracic Spine) Pain without and with Neurological Features (Including Stenosis) (SP-4.1)**
  - See: **Neck (Cervical Spine) Trauma (SP-3.2)**
  - See: **Coccydynia without Neurological Features (SP-5.2)**
  - See: **Spinal Deformities (e.g., Scoliosis/Kyphosis) (SP-14)** and **Spinal Dysraphism (PEDSP-4)** in the Pediatric and Special Populations Spine Imaging Guidelines
  - See: **Sacro-Iliac (SI) Joint Pain, Inflammatory Spondylitis/Sacroiliitis and Fibromyalgia (SP-10)**
  - See: **Post-Operative Spinal Disorders (SP-15)**

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MRI of the Spine (SP-2.2)

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- See: **Procedure Codes Associated with Spine Imaging**
- For MR Spectroscopy, all spine uses are considered not medically necessary.
  - See: **Imaging of Intervertebral Discs (SP-2.5)**
- MRI Spine is performed either without contrast, with contrast *or* without and with contrast. A "with contrast" study alone is medically necessary only to complete a study begun without contrast. Contrast is generally not medically necessary for most disc and nerve root disorders, fractures, and degenerative disease.
- MRI Spine indications include:
  - Evaluation of disc disease, spinal cord and nerve root disorders and most other spinal conditions including evaluation of congenital anomalies of the spine and spinal cord
  - Suspicion for or surveillance of known spine/spinal canal/spinal cord neoplastic disease
  - Suspicion, diagnosis of or surveillance of spinal infections, multiple sclerosis, or other causes of myelitis, syringomyelia, cauda equina syndrome or other "red flag" indications. See: **Red Flag Indications (SP-1.2)**.
  - Pre-operative evaluation to define abnormal or variant spinal anatomy that could influence the outcome of a potential surgical procedure. See: **Prior to Spine Surgery (SP-16.1)**.
  - Spinal imaging for individuals having undergone recent spinal surgery e.g., laminectomy, discectomy, spinal decompression, when history and physical examination is suspicious for hematoma, post-surgical infection, or cerebrospinal fluid (CSF) leak.

Positional MRI:

- Positional MRI is also referred to as dynamic, weight-bearing or kinetic MRI. Currently, there is inadequate scientific evidence to support the medical necessity of this study. As such, it should be considered not medically necessary.

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CT of the Spine (SP-2.3)

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- See: **Procedure Codes Associated with Spine Imaging**
- CT Spine indications include:
  - Contraindication to MRI
    - CT (contrast as requested) is medically necessary when ANY of the following MRI contraindications are documented:
      - Implanted ferromagnetic materials
      - Electronically, magnetically, or mechanically activated implanted devices that are not determined by the manufacturer as MRI compatible/conditional
  - CT without contrast, or CT without and with contrast (even if MRI has already been performed), for any spinal trauma/fractures, especially spinal trauma/fractures that could result in spinal instability and spinal cord/spinal nerve compression
  - CT without contrast, or CT without and with contrast (even if MRI has already been performed), for spinal neoplastic disease – primary or metastatic
  - CT without contrast, or CT without and with contrast (even if MRI has already been performed), in conjunction with myelography or discography (see: **CT/ Myelography [SP-2.4]** and **Imaging of Intervertebral Discs [SP-2.5]**)
  - CT without contrast, or CT without and with contrast (even if MRI has already been performed), for pre-operative evaluation to define abnormal or variant bony spinal anatomy that could influence the outcome of a potential surgical procedure (see: **Prior to Spine Surgery [SP-16.1]**)
  - CT without contrast, or CT without and with contrast, (even if MRI has already been performed), to assess spinal fusions when pseudoarthrosis is suspected (not to be used for routine post-operative assessment where x-rays are sufficient and/or there are no concordant clinical signs or symptoms)
  - CT without contrast, or CT without and with contrast (even if MRI has already been performed), for congenital, developmental, or acquired spinal deformity (see: **Spinal Deformities [e.g., Scoliosis/Kyphosis] [SP-14]**)
  - CT without contrast, or CT without and with contrast, for spondylolysis when routine x-rays are negative and/or MRI is equivocal, indeterminate, or non-diagnostic (see: **Lumbar Spine Spondylolysis/Spondylolisthesis [SP-8]**)
  - CT without contrast, or CT without and with contrast, to evaluate calcified lesions, (e.g., osteophytes, ossification of the posterior longitudinal ligament [OPLL])

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CT/Myelography (SP-2.4)

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- See: **Procedure Codes Associated with Spine Imaging**
- CT/Myelography is generally unnecessary as an initial study when a diagnostic quality MRI has been obtained.
- CT/Myelography indications include:
  - To clarify equivocal, indeterminate, or non-diagnostic MRI findings or to further evaluate the significance of multiple spinal abnormalities
  - When an MRI is contraindicated (see: **CT of the Spine [SP-2.3]**)
  - Pre-operative planning for spine surgery, (e.g., multilevel spinal stenosis or when a previous MRI is insufficient, equivocal, indeterminate, or non-diagnostic) (see: **Prior to Spine Surgery (SP-16.1)**)
  - Evaluation after previous spinal surgery when an MRI without and with contrast is contraindicated or MRI results are equivocal, indeterminate, or non-diagnostic
  - The guidelines allow for the approval of the post-myelogram CT (i.e., CPT® 72126, CPT® 72129, and CPT® 72132) only and not any other myelogram-related procedure codes (i.e., CPT® 72265 or CPT® 62284).

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Imaging of Intervertebral Discs (SP-2.5)

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Post-lumbar Discography CT:

- The guidelines allow for the post-lumbar discography CT procedure codes only and do not include any other discography-related procedure codes. A post-lumbar discography CT is considered medically necessary following an approved discography and ALL the following apply:
  - A post-discography CT is coded as without contrast.
  - A CT Lumbar Spine without contrast (CPT® 72131) is medically necessary if verified to be performed as a post-discography CT.
  - When a post-discography CT is requested and the discography has already been approved, authorization will be issued for the post-discography CT procedure codes.

Magnetic Resonance Spectroscopy:

- Magnetic Resonance Spectroscopy (MRS) involves the analysis of the levels of certain chemicals in pre-selected voxels (small regions) on an MRI scan done at the same time.
  - MRS (CPT® 76390, 0609T, 0610T, 0611T, and 0612T) is considered not medically necessary for all spine imaging uses at this time.

Background and Supporting Information

- Provocative Discography/CT and MR Spectroscopy lumbar spine are procedures purported to diagnose (or rule-out) a discogenic “pain generator” i.e., the source of non-specific axial spinal pain. These diagnostic studies, when reported as positive, are often used as an indication for spinal fusion in individuals with non-specific axial back pain.
- The following uses of discography are considered controversial:
  - To identify a symptomatic pseudoarthrosis in a failed spinal fusion
  - To identify which of two herniated discs seen on MRI is symptomatic when not determined clinically or otherwise
  - To confirm the discogenic nature of pain in an individual with an abnormal disc seen on MRI and to rule out pain from an adjacent disc level
  - To confirm the presumptive diagnosis of “internal disc disruption”
  - Discography of the cervical and/or thoracic spine
- The following uses of MR Spectroscopy lumbar spine are considered controversial:

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- To identify which of two herniated discs seen on MRI is symptomatic when not determined clinically or otherwise
- To confirm the discogenic nature of pain in an individual with an abnormal disc seen on MRI and to rule out pain from an adjacent disc level
- To confirm the presumptive diagnosis of “internal disc disruption”

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Ultrasound of the Spinal Canal (SP-2.6)

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- Spinal canal ultrasound (CPT® 76800) describes the evaluation of the spinal cord (canal and contents) most often performed in newborns, infants, young children and intraoperatively.
- CPT® 76800 describes evaluation of the entire spine and should not be reported multiple times for imaging of different areas of the spinal canal.
- CPT® 76998, rather than CPT® 76800, should be used to report intraoperative spinal canal ultrasound (ultrasonic guidance). Intraoperative use of spinal ultrasound (CPT® 76998) would not require prior authorization.

Indications for spinal canal ultrasound (CPT® 76800):

- This study is generally limited to infants, newborns, and young children because of incomplete ossification of the vertebral segments surrounding the spinal cord, including the assessment of CSF in the spinal canal and for image-guided lumbar puncture.
- When ossification of the vertebral segments is incomplete for evaluation of suspected or known tethered cord (see: **Tethered Cord [PEDSP-5]** in the Pediatric and Special Populations Spine Imaging Guidelines).
- Evaluation of suspected occult and non-occult spinal dysraphism (see: **Spinal Dysraphism [PEDSP-4]** in the Pediatric and Special Populations Spine Imaging Guidelines).
- Evaluation of spinal cord tumors, vascular malformations, and cases of birth-related trauma.
- Contraindicated for use in the adult spine for the assessment of spinal pain, radiculopathy, facet inflammation, nerve root inflammation, disc herniation, and soft tissue conditions surrounding the adult spine other than for superficial masses.

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Limitations of Spinal Imaging in Degenerative Disorders (SP-2.7)

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- Non-specific axial spinal pain is ubiquitous. Advanced diagnostic imaging infrequently identifies the source of the spinal pain (pain generator).
- Incidental findings on MRI and CT, including bulging, protruding, extruding or herniated discs, are often non-concordant, asymptomatic and increase in incidence as the spine ages.
- In individuals with poorly defined clinical presentations, “abnormal” spinal advanced diagnostic imaging results are infrequently clinically concordant, significant, material or substantive and may even lead to inappropriate treatment.
- Performing advanced spinal imaging based only on the presence of spinal degenerative findings identified on x-rays is not generally medically necessary in individuals who are either asymptomatic or present with non-specific axial spinal pain.

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Miscellaneous Spinal Lesions (SP-2.8)

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Vertebral body hemangiomas:

- Vertebral body hemangiomas are common and are generally benign and incidental findings on plain x-rays and advanced diagnostic imaging studies.
- If the appearance of a vertebral body hemangioma is typical on plain x-ray, further spinal advanced diagnostic imaging is not usually required, unless there are associated neurologic symptoms or signs on physical examination.
- If the appearance of a vertebral body hemangioma is atypical on plain x-ray, with or without neurological signs or symptoms on physical exam, MRI without contrast or MRI without and with contrast is medically necessary.
- Occasionally, MRI may be equivocal, indeterminate, or non-diagnostic and CT without contrast of the spinal area is medically necessary to help clarify the diagnosis.
- No follow-up imaging is necessary once the diagnosis of a vertebral body hemangioma is established without neurological features.

Tarlov cysts:

- Tarlov cysts are most often cystic dilatations of nerve root sleeves in the lumbar spine and sacrum.
- Controversy exists as to whether Tarlov cysts can result in neurologic signs and symptoms but they can result in erosion of the adjacent bone.
- Usually Tarlov cysts are benign, incidental findings on advanced diagnostic imaging studies. Further evaluation of a known or suspected Tarlov cyst can be performed with an MRI Lumbar Spine without and with contrast study (CPT® 72158) or CT/ Myelography Lumbar Spine (CPT® 72132).

Other spinal lesions:

- MRI without and with contrast or a CT without contrast is medically necessary if:
  - Other spinal lesions are seen on routine x-rays or a non-contrast MRI; ***and***
  - These additional advanced imaging studies are recommended by a spine specialist or radiologist to further characterize or diagnose the lesion; ***or***
  - Required for surgical planning.

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MRA Spinal Canal (SP-2.9)

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- MR angiography (MRA) Spine (CPT® 72159) imaging is utilized infrequently.
- Cerebrospinal Fluid (CSF) flow studies using MRI are included in CPT® codes 70551, 70552, and 70553 and should not be coded or reported separately, (see **CSF Flow Imaging (HD-24.4)**)

Indications may include:

- Suspected spinal cord arteriovenous malformation (AVM) or arteriovenous fistula (AVF):
  - MRI Spine of the relevant spine region without and with contrast should be the initial imaging study.
  - If suspicion for a spinal AVM or AVF is high based upon the results of the MRI Spine, catheter angiography is recommended .
- Subarachnoid hemorrhage where no brain aneurysm has been previously identified
  - Catheter angiography should be performed and is the most definitive study to define possible spinal pathology resulting in a spinal canal subarachnoid hemorrhage.
  - See: **General Guidelines – CT and MR Angiography (CTV and MRV) (HD-1.5)** in the Head Imaging Guidelines
  - See: **Cerebral Aneurysms (HD-12.1)** in the Head Imaging Guidelines
- Pre-operative planning
  - MRA Spinal canal may be useful in identifying major intercostal feeder vessels to the spinal cord prior to surgical procedures that might interfere with this blood supply. However, catheter angiography is generally a more definitive study for this purpose.
- 3D Rendering (CPT® 76377 or CPT® 76376) is medically necessary with spinal angiography to define the presence, location, and anatomy of intraspinal vascular malformations.

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Spine PET/CT (SP-2.10)

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- At the present time there is controversy regarding spine PET/CT due to inadequate scientific evidence to support the medical necessity of PET/CT for the routine assessment of spinal disorders, other than for neoplastic disease.
- See: **Spinal/Vertebral Metastases (ONC-31.6)** in the Oncology Imaging Guidelines
- Spine PET/CT should be considered not medically necessary.

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Cone-beam CT (SP-2.11)

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- Cone-beam CT for imaging of the cervical spine is considered not medically necessary.

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3D Rendering (SP-2.12)

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- See: **3D Rendering (MS-3)** in the Musculoskeletal Imaging Guidelines

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Evidence Discussion (SP-2)

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- X-rays are first line imaging for suspected inflammatory spine disease[6], following trauma[13], concerns of spinal deformities[7], or post-operative spinal disorders[8]. Although x-rays may not confirm a definitive diagnosis, they provide information that can better direct advanced imaging modalities.
- MRI utilizes a magnetic field and radio waves with computer processing to produce detailed images which have excellent soft tissue characterization and is the primary modality for evaluating the spinal cord, intervertebral disc disease and other soft tissue pathology of the spine.[9,10,11] Positional MRI[2] and MR Spectroscopy[20] lacks sufficient scientific evidence to support its routine clinical use.[2]
- CT is medically necessary as an alternative to MRI when MRI is contraindicated or equivocal.[10,12,13,15] CT is also medically necessary for evaluation of bony pathology including but not limited to fractures[13], bony neoplastic disease[15], calcified lesions[16], post traumatic[13] and perioperative bony processes.[8,14] Following lumbar discography CT is medically necessary to evaluate disc anatomy.[19]
- CT/Myelography provides indirect visualization of the thecal sack in the spinal canal. MRI is the primary medically necessary advanced imaging for these indications as the cord, thecal sac, and spinal canal can be directly visualized. CT/Myelography may be medically necessary when MRI is indeterminate or contraindicated.[17,18]
- Ultrasound can be used to visualize the spinal canal in young children before the posterior elements ossify. Ultrasound will not penetrate ossified bone.[21]
- MRA has limited indications in spine imaging but may be appropriate for evaluating spinal vascular malformations.
- There is inadequate scientific evidence to support the medical necessity of PET/CT for the routine assessment of spinal disorders, other than for neoplastic disease.
- There is inadequate scientific evidence to support the medical necessity of cone beam CT for the routine assessment of spinal disorders outside of the operative setting which is outside the coverage of these guidelines.

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Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3)

Guideline

Neck (Cervical Spine) Pain without and with Neurological Features (Including Stenosis) (SP-3.1)
Neck (Cervical Spine) Trauma (SP-3.2)
Evidence Discussion (SP-3)
References (SP-3)

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Neck (Cervical Spine) Pain without and with Neurological Features (Including Stenosis) (SP-3.1)

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***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**) • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**)

Advanced Diagnostic Imaging:	MRI Cervical Spine, without contrast (CPT® 72141)
Comments:	CT Cervical Spine without contrast (CPT® 72125) **OR** CT Myelography (CPT® 72126) is medically necessary when MRI is contraindicated.

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Neck (Cervical Spine) Trauma (SP-3.2)

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***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**) • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**) • Results of plain x-rays of the cervical spine performed after the current episode of symptoms started or changed need to be available to the requesting provider (not required for high-risk mechanisms as below**)

Advanced Diagnostic Imaging:	**MRI Cervical Spine without contrast (CPT® 72141) OR CT Cervical Spine without contrast (CPT® 72125)** **For individuals with ankylosing spondylitis or DISH (diffuse idiopathic skeletal hyperostosis), both MRI of the entire spine (CPT® 72141, 72146, and/or 72148) AND CT of the entire spine (CPT® 72125, 72128, and/or 72131) are medically necessary** **For individuals with ankylosing spondylitis or DISH (diffuse idiopathic skeletal hyperostosis) plain x-rays and a 6-week trial of provider-directed treatment and clinical re-evaluation are NOT required.**
Comments:	Plain x-rays **ARE** required for suspected fracture in **non-high** risk injuries. Plain x-rays and a 6-week trial of provider-directed treatment and clinical re-evaluation are **NOT** required for individuals with a high risk factor(s) for suspected cervical spine injury within the last 3 months (See below**).

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**High-risk factors of suspected cervical spine injury may include:

- Long term use of systemic glucocorticoids
- History of prior low energy fractures
- History of low bone mineral density
- Age ≥65 years
- Head trauma and/or maxillofacial trauma
- Pedestrian in a motor vehicle accident
- Fall from elevation ≥3 feet/5 stairs
- Diving accident
- Head-on motor vehicle collision without/with airbag deployment
- Rollover motor vehicle collision
- Ejection from the vehicle in a motor vehicle collision
- High speed of the vehicle at the time of collision
- Not wearing a seatbelt/shoulder harness in a motor vehicle collision
- Minor direct/indirect trauma to the cervical spine/maxillofacial areas in individuals with ankylosing spondylitis or DISH

Background and Supporting Information

- Pain radiation patterns from the cervical spine area into the thoracic spine area do not necessarily justify the addition of thoracic spine advanced diagnostic imaging.
- Cervical radiculopathy is often confused with shoulder disorders, brachial plexopathy, peripheral nerve entrapment and/or motor/sensory neuropathies. Electrodiagnostic testing (EMGs/NCVs) is generally used to confirm, not establish, a diagnosis of peripheral nerve entrapment and/or a motor/sensory neuropathy based upon history and physical examination findings. Electrodiagnostic testing is often considered when advanced imaging of the spine does not reveal neurocompressive pathology and/or after 6 weeks of unimproved symptoms of extremity pain, weakness, numbness and/or tingling.
- Individuals with ankylosing spondylitis or DISH are at high risk of cervical spine fractures even with minor direct/indirect trauma to the cervical spine which can result in quadriparesis/quadriplegia

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Evidence Discussion (SP-3)

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X-rays provide critical information that cannot be obtained with advanced imaging modalities and remain central to providing optimal care for spine individuals.[8,15,17] Unnecessary CT scans increase individuals' radiation exposure,[18] however, is the initial imaging for indviduals involved in trauma with a high-risk factor for cervical spine injury.[7] Additionally, for individuals with diffuse idiopathic skeletal hyperostosis (DISH) or ankylosing spondylitis with a history of low-energy trauma, whole spine MRI or CT imaging is mandatory due to the high prevalence of acute fractures and the low specificity for fracture detection on radiographs.[10,12] The American College of Radiology (ACR) Appropriateness Criteria for Cervical Neck Pain or Cervical Radiculopathy (revised 2018) indicates that in the absence of red flag symptoms, early advanced imaging may not be required as abnormal findings are not uncommon in asymptomatic individuals and correlate poorly with the presence of neck pain.[8]

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Upper Back (Thoracic Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-4)

Guideline

Upper Back (Thoracic Spine) Pain without and with Neurological Features (Including Stenosis) (SP-4.1)
Upper Back (Thoracic Spine) Trauma (SP-4.2)
Evidence Discussion (SP-4)
References (SP-4)

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Upper Back (Thoracic Spine) Pain without and with Neurological Features (Including Stenosis) (SP-4.1)

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***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**). • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**). • Plain x-rays are required for thoracic back pain without neurological features (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**).

Advanced Diagnostic Imaging:	**MRI Thoracic Spine without contrast (CPT® 72146)**
Comments:	A CT Thoracic spine without contrast (CPT® 72128) **OR** CT Myelography (CPT® 72129) is medically necessary when MRI is contraindicated.

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Upper Back (Thoracic Spine) Trauma (SP-4.2)

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All the following are required prior to advanced imaging:
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**). • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**). • Conservative treatment and clinical re-evaluation are not required in high-risk individuals. ◦ High-risk individuals include those that have sustained an acute traumatic injury with midline thoracolumbar tenderness, high-energy injury mechanisms, or >60 years of age. • **After above criteria are met:** MRI Thoracic Spine without contrast (CPT® 72146) OR CT Thoracic Spine without contrast (CPT® 72128)

Indication:	Advanced Imaging
Ankylosing Spondylitis (AS) or Diffuse Idiopathic Skeletal Hyperostosis (DISH)	• MRI of the entire spine (CPT® 72141, 72146, and/or 72148) **AND** • CT of the entire spine (CPT® 72125, 72128, and/or 72131) For individuals with AS or DISH, a 6-week trial of provider-directed treatment and clinical re-evaluation are **NOT** required

Background and Supporting Information

- Thoracic radiculopathy presents with pain radiation from the thoracic spine around the trunk. At upper thoracic spine levels, the pain radiation is from the thoracic spine around the rib cage following the sensory distribution of an intercostal nerve.

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- Advanced diagnostic imaging is generally not medically necessary in evaluation of axial low back pain with radiation toward the thoracic region unless there are documented clinical features indicating a thoracic spine disorder.

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Evidence Discussion (SP-4)

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The precision in identifying thoracic spine diseases is dependent on a meticulous association with the individual's clinical examination and medical history because the usual observations from imaging studies are frequently ambiguous and non-conclusive.[5]

Wood et al. demonstrated that asymptomatic individuals may exhibit positive findings on thoracic spine MRI at a rate as high as 70%.[6] For individuals with atraumatic thoracic back pain, data from Linna, et al. supports initial conservative management followed by evaluation by a surgical specialist before ordering a thoracic spine MRI.[5] Red flag indications, however, obviate the need for conservative care.

Regarding the value of x-rays, Goodwin et al. stated that plain films provide critical information that cannot be obtained with other imaging modalities, and they remain central to providing optimal care for spine individuals.[7] When there is a history of blunt trauma and a high-risk factor for thoracic spine injury, however, CT imaging is appropriate for initial imaging.[8,2]

Additionally, for individuals with diffuse idiopathic skeletal hyperostosis (DISH) or ankylosing spondylitis with a history of low-energy trauma, whole spine MRI or CT imaging is mandatory due to the high prevalence of acute fractures and the low specificity for fracture detection on radiographs.[3,4]

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Low Back (Lumbar Spine) Pain/Coccydynia without Neurological Features (SP-5)

Guideline

Low Back (Lumbar Spine) Pain without Neurological Features (SP-5.1)
Coccydynia without Neurological Features (SP-5.2)
Evidence Discussion (SP-5)
References (SP-5)

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Low Back (Lumbar Spine) Pain without Neurological Features (SP-5.1)

SP.LB.0005.1.A
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***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**. • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**). • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**)

Advanced Diagnostic Imaging:	**MRI Lumbar Spine without contrast (CPT® 72148)**
Comments:	A CT Lumbar spine without contrast (CPT® 72131) **OR** CT Myelography (CPT® 72132) is medically necessary when MRI is contraindicated

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Coccydynia without Neurological Features (SP-5.2)

SP.LB.0005.2.A
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***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**). • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**) • Plain x-rays of the sacrum/coccyx are negative for fracture.

Advanced Diagnostic Imaging:	**MRI Pelvis without contrast (CPT® 72195)**
Comments:	A CT Pelvis without contrast (CPT® 72192) when MRI is contraindicated.

Background and Supporting Information

Coccydynia is often reported by individuals as “tailbone” pain that is usually idiopathic or post-traumatic and generally follows a benign course.

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Evidence Discussion (SP-5)

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Acute low back pain is usually a self-limited condition and improves with conservative treatment in 6 weeks. The American College of Radiology Appropriateness Criteria for low back pain (revised 2021) states that imaging may be considered in those individuals who have had up to 6 weeks of medical management and physical therapy that resulted in little or no improvement in their back pain.[29]

A meta-analysis by Chou et al. found no clinically significant difference in individual outcomes between those who had immediate lumbar imaging versus usual care.[33] It should also be noted that there are risks associated with imaging including but not limited to radiation exposure and contrast complications.[34] Studies have also linked the increase rate of imaging with the increase rate of surgery and also found early magnetic resonance imaging (MRI) had an eight-fold increased risk of surgery.[34,35]

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Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) With or Without Low Back (Lumbar Spine) Pain (SP-6)

Guideline

Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)
Low Back (Lumbar Spine) Trauma (SP-6.2)
Evidence Discussion (SP-6)
References (SP-6)

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Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)

SP.LE.0006.1.A
v1.0.2026

***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**). • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**).

Advanced Diagnostic Imaging:	**MRI Lumbar Spine without contrast (CPT® 72148)**
Comments:	A CT Lumbar spine without contrast (CPT® 72131) **OR** CT Myelography (CPT® 72132) is medically necessary when MRI is contraindicated. See also: **Lumbar Spinal Stenosis (SP-9.1)**

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Low Back (Lumbar Spine) Trauma (SP-6.2)

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***All* the following are required prior to advanced imaging:**
• An in-person clinical evaluation for the current episode of the condition is required to have been performed before advanced imaging is considered. This may have been either the initial clinical evaluation or a clinical re-evaluation (see also: **General Guidelines (SP-1.0)**). • Failure of a 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed (unless presence of a red flag as defined in **Red Flag Indications (SP-1.2)**). • Clinical re-evaluation after treatment period (may consist of an in-person evaluation or other meaningful contact (see also: **General Guidelines (SP-1.0)**). • Conservative treatment and clinical re-evaluation are not required in high-risk individuals. ◦ High-risk individuals include those that have sustained an acute traumatic injury with midline thoracolumbar tenderness, high-energy injury mechanisms, or >60 years of age. • **After above criteria are met:** MRI Lumbar Spine without contrast (CPT® 72148) **OR** CT Lumbar Spine without contrast (CPT® 72131)

Indication:	Advanced Diagnostic Imaging
Ankylosing Spondylitis (AS) or Diffuse Idiopathic Skeletal Hyperostosis (DISH)	• MRI of the entire spine (CPT® 72141, 72146, and/or 72148) **AND** • CT of the entire spine (CPT® 72125, 72128, and/or 72131) For individuals with AS or DISH, a 6-week trial of provider-directed treatment and clinical re-evaluation are **NOT** required.

- Definitions of radiculopathy, radiculitis, and radicular pain: See **Definitions (SP-1.3)**
- Sciatic Neuropathy, Femoral Neuropathy, Peroneal Neuropathy and Meralgia Paresthetica: See **Focal Neuropathy (PN-2)** in the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines

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- Lumbar and/or Lumbosacral Plexopathy: See **Lumbar and Lumbosacral Plexus (PN-5)** in the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines
- Advanced imaging of the hip or pelvis is not generally required in the evaluation of apparent lumbar radiculopathy unless a separate recognized indication for such studies is documented. See: **Hip (MS-24)** in the Musculoskeletal Imaging Guidelines.

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Evidence Discussion (SP-6)

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Acute low back pain is usually a self-limited condition and improves with conservative treatment in 6 weeks. The American College of Radiology Appropriateness Criteria® for low back pain (revised 2021) states that imaging may be considered in those individuals who have had up to 6 weeks of medical management and physical therapy that resulted in little or no improvement in their back pain.[26]

A meta-analysis by Chou et al. found no clinically significant difference in individual outcomes between those who had immediate lumbar imaging versus usual care.[31] It should also be noted that there are risks associated with imaging including but not limited to radiation exposure and contrast complications.[32] Studies have also linked the increase rate of imaging with the increase rate of surgery and also found early magnetic resonance imaging (MRI) had an eight-fold increased risk of surgery.[32,33] In typical individuals with low back pain or radiculopathy, MR imaging does not appear to have measurable value in terms of planning conservative care, that knowledge of imaging findings does not alter outcome, and that individual knowledge of imaging findings is associated with a lesser sense of well-being.[23]

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Myelopathy (SP-7)

Guideline

Myelopathy (SP-7.1)
Evidence Discussion (SP-7)
References (SP-7)

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Myelopathy (SP-7.1)

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- Myelopathy is the development of abnormal spinal cord function with long tract signs usually secondary to spinal cord compression, but also inflammation (transverse myelitis, MS, etc.), neoplastic disease or spinal cord infarction.
  - For imaging of transverse myelitis, see: **Transverse Myelitis (HD-16.4)** in the Head Imaging Guidelines
- Examination findings may include loss of manual dexterity, spastic legs, ataxia, hyperreflexia, upgoing toes (positive Babinski), Hoffmann's sign, sustained clonus, Lhermitte's sign, crossed radial reflex, inverted radial reflex, and/or finger escape sign. Sensory level and urinary incontinence/retention may be seen.
  - Advanced imaging is generally medically necessary in the initial evaluation of documented or reasonably suspected myelopathy.
- X-rays are not required for advanced imaging in individuals with potential myelopathy regardless of any history of spine surgery, trauma, or other reasons which may otherwise require x-rays (e.g., **Neck (Cervical Spine) Trauma (SP-3.2)**, **Upper Back (Thoracic Spine) Trauma (SP-4.2)**, **Post-Operative Spinal Disorders (SP-15)**).
- Conservative treatment is not a requirement for advanced imaging in individuals with potential myelopathy.
- MRI Cervical and Thoracic Spine without contrast, or without and with contrast, are medically necessary for:
  - Evaluation of reasonably suspected myelopathy
  - Post-traumatic syrinx with increased spinal pain or worsening neurological symptoms
  - Sustained, prominent, and unexplained Lhermitte's sign
  - Unexplained Babinski's or Hoffmann's signs
  - Unexplained hyperreflexia
  - Unexplained bilateral motor weakness
- MRI Cervical, Thoracic, and Lumbar Spine without contrast, or without and with contrast, are medically necessary for:
  - Suspected tethered cord and/or low-lying conus medullaris.
- CT without contrast, or CT with contrast (myelography), can also be considered for either of the following:
  - An alternative to MRI, when MRI is contraindicated
  - In addition to MRI, for surgical planning

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Background and Supporting Information

Lhermitte's sign – With the individual in the long leg sitting position on the examination table, the examiner passively flexes the individual's head and one hip simultaneously with the leg kept straight. A positive test occurs if there is sharp pain down the spine and into the upper or lower extremities.

Babinski's sign – The examiner runs a sharp instrument along the plantar surface of the foot from the calcaneus along the lateral border to the forefoot. A positive test occurs with extension of the great toe with flexion and splaying of the other toes. A negative test occurs with no movement of the toes at all or uniform bunching up of the toes.

Hoffman's sign – The examiner holds the individual's middle finger and briskly flicks the distal phalanx. A positive test is noted if the interphalangeal joint of the thumb of the same hand flexes.

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Evidence Discussion (SP-7)

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- MRI is the preferred imaging modality for evaluation of myelopathy. It provides superior soft tissue definition to other options and allows direct visualization of intramedullary cord signal changes which can affect prognosis and management.[10]
- CT Myelogram may be appropriate when MRI is contraindicated or for surgical planning. It may allow better visualization of bony neuroforaminal narrowing and may provide additional anatomic information when the MRI is ambiguous.[10,11]
- CT Can be useful in demonstrating bony encroachment on the cord, but MRI is superior in demonstrating bone marrow changes and intramedullary cord signal. It is of limited value in evaluation of non-compressive causes of myelopathy.[10]

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Lumbar Spine Spondylolysis/ Spondylolisthesis (SP-8)

Guideline

Spondylolysis (SP-8.1)
Spondylolisthesis (SP-8.2)
Evidence Discussion (SP-8)
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Spondylolysis (SP-8.1)

SP.SP.0008.1.A
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Results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider, unless otherwise specified below.

Indication	Imaging Study
• Clinical suspicion of spondylolysis is high	• X-ray is not required • Tomographic SPECT Planar (CPT® 78803 or 78831) • SPECT/CT Hybrid (CPT® 78830 or 78832)
• Negative SPECT bone scan	MRI Lumbar Spine without contrast (CPT® 72148) **OR** CT Lumbar Spine without contrast (CPT®72131)
• Evaluation of a lesion seen on SPECT bone scan	CT Lumbar Spine without contrast (CPT® 72131)
• Documented need for preoperative planning	MRI Lumbar Spine without contrast (CPT® 72148) **AND/OR** CT Lumbar Spine without contrast (CPT®72131)
• Failure of 6 weeks of provider-directed conservative treatment (which may include immobilization with a spinal orthosis) after the current set of symptoms or physical exam findings started or changed with clinical re-evaluation	MRI Lumbar Spine without contrast (CPT® 72148) **OR** CT Lumbar Spine without contrast (CPT®72131)
• Evaluation for stress reaction in bone, to visualize nerve roots	MRI Lumbar Spine without contrast (CPT® 72148)
• When an MRI is medically necessary, however, it is contraindicated	CT Lumbar Spine without contrast (CPT®72131)

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Indication	Imaging Study
• Evaluation of bony anatomy	CT Lumbar Spine without contrast (CPT®72131)
• Monitor healing of a pars interarticularis fracture that was determined to have healing potential on a prior CT (i.e., non-sclerotic lesion)	CT Lumbar Spine without contrast (CPT® 72131) of the symptomatic spinal level

- For pediatric spondylolysis, see: **Spondylolysis (PEDSP-2.4)** in the Pediatric and Special Populations Spine Imaging Guidelines
- Bony healing cannot be achieved non-surgically in an established well defined isthmic pars interarticularis defect whether it is developmental or the result of a pars interarticularis fracture non-union. Repeat advanced diagnostic imaging is not medically necessary in this setting.

Background and Supporting Information

- Spondylolysis is most often an incidental finding on plain x-rays, and advanced imaging is generally not medically necessary.
- MRI is not medically necessary in the early diagnosis of spondylolysis due to the potential for false negative results.

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Spondylolisthesis (SP-8.2)

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- CT Lumbar Spine without contrast (CPT® 72131) or MRI Lumbar Spine without contrast (CPT® 72148) can be considered after plain x-ray (results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider) for the following:
  - Failure of 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed and clinical re-evaluation (see also: **General Guidelines [SP-1.0]**); ***or***
  - Preoperative evaluation; ***or***
  - See: **Red Flag Indications (SP-1.2)**

Background and Supporting Information

- Stress reactions and stress fractures of the pars interarticularis are most common in athletes and others whose activities involve repetitive flexion/extension loading of the lumbar spine and may be acute or chronic and unilateral or bilateral. Pars interarticularis defects can be an incidental finding on plain x-rays and is frequently asymptomatic.
- Spondylolisthesis is the forward (anterolisthesis) or backward (retrolisthesis, usually not clinically significant) displacement of one vertebra in relation to an adjacent vertebra, most commonly at L4-5 and L5-S1, although other levels of the spine may be involved. Spondylolisthesis is often an incidental finding on plain x-ray and is frequently asymptomatic.

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Evidence Discussion (SP-8)

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Spondylolysis is a very common incidental finding on radiographs in the general population but majority will be asymptomatic.[7] Spondylolysis is one of the potential causes of back pain in gymnasts.[6] Symptomatic pars lesions are particularly a clinical problem in adolescent athletes.[7,8] Spondylolysis and spondylolisthesis are a common cause of low back pain especially in young athletes but is a less common cause of neurologic compromise.[4] Plain radiographs with particular views display the majority of defects.[5] MRI has sensitivity of 78% for detecting L4-L5 lumbar degenerative spondylolisthesis compared with 98% for lateral standing films.[3] MRI is less sensitive than CT for detecting pars defects but it is useful for evaluating bone marrow edema,nerve root compression, and stress reactions.[8]

Lumbar spondylolysis can heal with conservative treatment depending on the spinal level affected and stage of the defects. The site of defects in the pars, condition of contralateral pars, presence of spondylolisthesis, the degree of lumbar lordosis are among other factors significantly affecting union.[10]

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Lumbar Spinal Stenosis (SP-9)

Guideline

Lumbar Spinal Stenosis (SP-9.1)
Evidence Discussion (SP-9)
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Lumbar Spinal Stenosis (SP-9.1)

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- MRI Lumbar Spine without contrast (CPT® 72148) or CT Lumbar Spine without contrast (CPT® 72131) is medically necessary for those individuals with clinical suspicion of lumbar spinal stenosis if:
  - Failure of 6-week trial of provider-directed treatment after the current set of symptoms or physical exam findings started or changed and clinical re-evaluation (see also: **General Guidelines (SP-1.0)**); ***or***
  - Red Flag Indications (see: **Red Flag Indications (SP-1.2)**); ***or***
  - Severe symptoms of neurogenic claudication restricting normal activity or requiring the frequent use of narcotic analgesics
- A CT/Myelogram Lumbar Spine (CPT® 72132) may also be considered for individuals who have failed 6-weeks of provider-directed treatment if requested by the operating surgeon for surgical planning, especially for multi-level lumbar spinal stenosis.

Background and Supporting Information

Lumbar spinal stenosis refers to a decrease in the space available for the neural elements within the spinal canal that include spinal nerve roots and the cauda equina. It is usually a degenerative condition of the aging spine which can be asymptomatic or a common cause of buttock/low back and/or leg pain (neurogenic claudication) in this population. Neurogenic claudication is a common symptom of lumbar spinal stenosis that is aggravated by walking, especially down hills or stairs, with prolonged standing and is often relieved by sitting and bending forward. Neurogenic claudication should be differentiated from vascular claudication (leg/calf pain) that is often aggravated by walking and relieved fairly rapidly by stopping and rest. The differential diagnosis for lumbar spinal stenosis should include peripheral vascular disease, hip disorders, and peripheral neuropathy.

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Evidence Discussion (SP-9)

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A presumptive diagnosis of symptomatic lumbar stenosis can be made with the history and physical examination.[5] Imaging can help differentiate neurogenic claudication from vascular claudication. MRI or CT may confirm the presence of spinal stenosis. The American College of Radiology Appropriateness Criteria for low back pain (revised 2021) states MRI may be helpful when there is low back pain with radiculopathy or signs of spinal stenosis.[1] Bony findings can be seen better on CT and soft-tissue lesions are more detectable on MRI.[1] In individuals with subacute or chronic low back pain with or without radiculopathy that is a surgical or intervention candidate with persisting symptoms after six weeks of conservative treatment, CT lumbar spine without IV contrast can also be used to assess facets and neural foramina and is equal to MRI for predicting significant spinal stenosis.[1] For those not responsive to conservative treatment surgery should be considered. A prospective cohort study evaluating individual outcomes two years after spine surgery for spinal stenosis showed individuals had better outcomes than individuals who did not have surgery.[4]

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Sacro-Iliac (SI) Joint Pain, Inflammatory Spondylitis/Sacroiliitis and Fibromyalgia (SP-10)

Guideline

Sacro-Iliac (SI) Joint Pain/Sacroiliitis (SP-10.1)
Inflammatory Spondylitis (SP-10.2)
Fibromyalgia (SP-10.3)
Evidence Discussion (SP-10)
References (SP-10)

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Sacro-Iliac (SI) Joint Pain/Sacroiliitis (SP-10.1)

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- CT Pelvis without contrast (CPT® 72192) or MRI Pelvis without contrast (CPT® 72195) is medically necessary if:
  - Initial plain x-rays are equivocal or not diagnostic; ***and***
  - Failure of 6 weeks of provider-directed treatment after the current set of symptoms or physical exam findings started or changed and clinical re-evaluation (see also: **General Guidelines (SP-1.0)**); ***or***
  - Any ONE of the following:
    - Fractures of the sacrum or sacroiliac joint(s); ***or***
    - See: **Red Flag Indications (SP-1.2)**; ***or***
    - Pre-operative planning
- For suspected neoplastic or infectious disease, see **Red Flags (SP-1.2)**
- See: **Rheumatoid Arthritis (RA) and Inflammatory Arthritis (MS-15.1)** in the Musculoskeletal Imaging Guidelines

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Inflammatory Spondylitis (SP-10.2)

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- **Initial imaging**
  - Plain x-rays are required initially, then:
    - MRI Sacroiliac Joints (MRI Pelvis) without and with contrast (CPT® 72197) AND MRI Spine area of interest without and with contrast) **OR**
    - MRI Sacroiliac Joints (MRI Pelvis) without contrast (CPT® 72195) AND MRI Spine area of interest without contrast **OR**
    - MRI Sacroiliac Joints (MRI Pelvis) without and with contrast (CPT® 72197) **OR**
    - MRI Sacroiliac Joints (MRI pelvis) without contrast (CPT® 72195) **OR**
    - CT Sacroiliac Joints (CT Pelvis) without contrast (CPT® 72192) OR CT Sacroiliac Joints (CT Pelvis) without contrast (CPT® 72192) AND CT Spine area of interest without contrast if MRI is contraindicated
  - When plain x-rays **AND** MRI of the sacroiliac joints are negative, then:
    - MRI Spine area of interest without and with contrast **OR**
    - MRI Spine area of interest without contrast **OR**
    - CT Spine area of interest without contrast
- **Follow-up imaging for treatment response or disease progression**
  - Repeat plain x-rays show no progression of disease of the SI joints **OR** SI joints and spine area of interest then,
    - MRI Sacroiliac Joints (MRI Pelvis) without and with contrast (CPT® 72197) AND MRI Spine area of interest without and with contrast) **OR**
    - MRI Sacroiliac Joints (MRI Pelvis) without contrast (CPT® 72195) AND MRI Spine area of interest without contrast **OR**
    - MRI Sacroiliac Joints (MRI Pelvis) without and with contrast (CPT® 72197) **OR**
    - MRI Sacroiliac Joints (MRI Pelvis) without contrast (CPT® 72195)
- If there is documented ankylosing spondylitis or DISH (diffuse idiopathic skeletal hyperostosis) and spine pain following trauma, then:
  - See: **Neck Trauma (SP-3.2)**, **Upper Back Trauma (SP-4.2)**, **Low Back Trauma (SP-6.2)**

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Fibromyalgia (SP-10.3)

SP.DI.0010.3.A

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- Advanced diagnostic imaging is not supported by the scientific evidence for the evaluation and treatment of fibromyalgia.

Background and Supporting Information

- Sacroiliitis can present with pain localized to the SI joint or referred pain to the buttock and/or posterior thigh without neurologic signs or symptoms. Affected individuals can often point to the SI joint as the pain source. Provocative and/or therapeutic SI joint anesthetic/corticosteroid injections can have diagnostic value.
- There is no evidence demonstrating that advanced diagnostic imaging substantiates changes to individual management decisions in individuals with proven SI joint disorders when visible on routine plain x-rays.
- MRI has shown inflammatory changes in the SI joints prior to visible x-ray changes in several studies. However, the ability of MRI to characterize inflammation in early ankylosing spondylitis, the ability of MRI to predict erosive changes, and the value of monitoring treatment effects using serial MRI studies remains controversial in adults.

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Evidence Discussion (SP-10)

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For individuals with proven sacro-iliac joint disorders visible on routine plain x-rays, there is no evidence that advanced diagnostic imaging substantiates changes to individual management decisions.

X-rays are first line imaging for suspected inflammatory sacroiliitis but have a low sensitivity for detecting abnormalities in early disease, and x-ray findings may not be visible until several years after onset of symptoms.[12] SI Joint MRI is appropriate when X-ray is equivocal or non-diagnostic.[12] There is increase the diagnostic accuracy of MRI in sacroiliitis with use of contrast.[12] Contrast use benefits must be weighed against potential disadvantages of need for IV access, potential risk for nephrogenic systemic fibrosis or contrast reaction, and increased cost. If an individual is unable to undergo MRI, a non-contrast CT may be helpful as it has improved sensitivity over conventional radiography for detection of subtle erosions, although it lacks sensitivity for inflammatory changes of inflammatory sacroiliitis.[12]

With suspected inflammatory spondylitis, x-rays are useful to assess for structural changes of syndesmophytes, erosions, vertebral body squaring, and ankylosis.[9] When x-rays are non-diagnostic, MRI (with or with contrast) can demonstrate active inflammatory changes.[9] The value of monitoring treatment response using serial MRIs remains controversial and investigational in adults.

Plain x-rays are not required prior to advanced imaging in individuals with documented ankylosing spondylitis or DISH (diffuse idiopathic skeletal hyperostosis) and spine pain following trauma, due to high risk of spinal fractures even with low-energy trauma and the low specificity for fracture detection on x-ray in these individuals.[8]

Advanced diagnostic imaging is not supported by scientific evidence for the evaluation and treatment of fibromyalgia.

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Spinal Compression Fractures (SP-11)

Guideline

Spinal Compression Fractures (SP-11.1)
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Spinal Compression Fractures (SP-11.1)

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Individuals with no history of malignancy

- MRI without contrast, CT without contrast, or whole-body bone scan (CPT® 78306), SPECT (CPT® 78803), or SPECT/CT (CPT® 78830) of the affected spinal region is medically necessary after plain x-ray evaluation ***and*** the location of the individual's spinal pain is concordant with the spinal x-rays for any ONE of the following:
  - X-rays reveal a new spinal compression fracture ***or***
  - X-rays are non-diagnostic and severe spinal pain persists for more than one week in an individual already predisposed to low energy/insufficiency fractures ***or***
  - The age of the spinal compression fracture deformity on plain x-ray is indeterminate ***or***
  - Surgical planning following known insufficiency spinal compression fractures in individuals who are candidates for kyphoplasty, vertebroplasty or other spine surgical procedures

Individuals with a history of malignancy

- For individuals with new symptomatic or asymptomatic vertebral compression fractures on radiographs, please refer to the cancer-specific guidelines within the General Oncology Imaging Guidelines for appropriate imaging studies.
- See also: **Red Flag Indications (SP-1.2)**

Background and Supporting Information

Insufficiency/low energy spinal compression fractures of the spine occur due to the lack of structural integrity to withstand physiologic loads and minor spinal trauma. Low bone mineral density is the primary etiology for most of these fractures but could also occur in the setting of other bone disease and medical conditions, in addition to neoplastic disease and infection. Sudden localized back pain, with or without trauma, is a typical presentation of insufficiency/low energy spinal compression fractures and can often be an incidental finding on plain x-rays and can be asymptomatic.

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Evidence Discussion (SP-11)

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The diagnosis of a spinal compression fracture may be suspected based on history and physical examination. Plain anteroposterior and lateral radiographs should be the initial imaging study obtained for a suspected compression fracture.[3,7]

For individuals (without a known malignancy) with a new symptomatic vertebral compression fracture identified on radiographs, MRI without contrast, CT without contrast, whole-body bone scan, SPECT or SPECT/CT is supported by the American College of Radiology Appropriateness Criteria for Management of Vertebral Compression Fractures (revised 2022).[2] Advanced imaging can also be helpful for identifying a fracture that is not well visualized on plain films.[3,5]

It has been shown that bone marrow signal on MRI can help identify an acute fracture and distinguish ages of compression fractures.[8,9] Additionally, the benefits of advanced imaging prior to vertebral augmentation have been reported.[10,11,12]

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References (SP-11)

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Spinal Pain related to Cancer (SP-12)

Guideline

Spinal Pain related to Cancer (SP-12)

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Spinal Pain related to Cancer (SP-12)

SP.CA.0012.A

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- For guidelines regarding advanced diagnostic imaging in this clinical setting, See **Spinal/Vertebral Metastases (ONC-31.6)**

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Spinal Canal/Cord Disorders (e.g., Syringomyelia) (SP-13)

Guideline

Initial Imaging Pathway (SP-13.1)
Follow-up Imaging (SP-13.2)
Evidence Discussion (SP-13)
References (SP-13)

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Initial Imaging Pathway (SP-13.1)

SP.CD.0013.1.A

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- MRI Cervical Spine without contrast or without and with contrast (CPT® 72141 or CPT® 72156) and MRI Thoracic Spine without contrast or without and with contrast (CPT® 72146 or CPT® 72157) is medically necessary when syringomyelia is suspected.
- Once a syrinx is identified by prior imaging, the following are medically necessary if not already performed:
  - MRI Brain without contrast (CPT® 70551) to evaluate for syringobulbia **AND**
  - MRI Cervical Spine without contrast or without and with contrast (CPT® 72141 or CPT® 72156) **AND**
  - MRI Thoracic Spine without contrast or without and with contrast (CPT® 72146 or CPT® 72157) **AND**
  - MRI Lumbar Spine without contrast or without and with contrast (CPT® 72148 or CPT® 72158) to define the lower most extent of the syrinx or to identify a skip lesion

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Follow-up Imaging (SP-13.2)

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- MRI Cervical Spine without contrast (CPT® 72141) and MRI Brain without contrast (CPT® 70551) and/or MRI Thoracic Spine without contrast (CPT® 72146) when involved
  - If there is a concern for malignancy, imaging can be performed without and with contrast
  - Annual imaging until non-progression of the syringomyelia is established
  - Following surgical treatment (including posterior fossa decompression)
  - Advanced diagnostic imaging every three years for life can be performed once non-progression of the syringomyelia is established
  - Repeat advanced diagnostic imaging is medically necessary when there is evidence of neurologic deterioration

Background and Supporting Information

Syringomyelia may begin to form in childhood but rarely becomes symptomatic before the adult years.

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Evidence Discussion (SP-13)

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- MRI of the spinal cord is the modality of choice to characterize the size and extent of a syrinx both at time of original discovery and on follow up imaging.[4]
- MRI of the brain is the modality of choice to characterize syringobulbia in the hindbrain.[4,5]

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Spinal Deformities (e.g., Scoliosis/ Kyphosis) (SP-14)

Guideline

Spinal Deformities (e.g., Scoliosis/Kyphosis) (SP-14.1)
Revision Anterior Spinal Deformity Surgery (SP-14.2)
Evidence Discussion (SP-14)
References (SP-14)

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Spinal Deformities (e.g., Scoliosis/ Kyphosis) (SP-14.1)

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- MRI without contrast, MRI without and with contrast, or CT/Myelography if MRI is contraindicated of the affected spinal regions is medically necessary after plain x-rays (e.g., Cobb radiographs) of the affected spinal regions have been performed and results are available to the requesting provider:
  - For pre-operative evaluation; ***or***
  - For cases of congenital scoliosis and other atypical curves that may be associated with spinal canal/cord pathology such as tethered cord, syringomyelia, diastematomyelia, or tumors; ***or***
  - For cases of scoliosis and/or kyphosis when there are associated neurologic signs and symptoms on physical examination; ***or***
  - Scoliosis with a convex left thoracic curve due to a high association of a convex left thoracic curve with underlying spinal canal/cord pathology
- CT of the affected spinal regions (contrast as requested) is medically necessary in cases with a complex osseous deformity for pre-operative evaluation
- CT Angiography (CTA) or MR Angiography (MRA) is not medically necessary for pre-operative planning for initial anterior spinal surgery for surgical correction of spinal deformities

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Revision Anterior Spinal Deformity Surgery (SP-14.2)

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- If requested by the operating surgeon, the following studies can be performed for pre-operative planning for revision of anterior thoracic or lumbar spinal surgery:
  - CTA Pelvis (CPT® 72191) **OR**
  - CTA Abdomen (CPT® 74175) **OR**
  - CTA Abdomen and Pelvis (CPT® 74174) **OR**
  - MRA Pelvis (CPT® 72198) and/or MRA Abdomen (CPT® 74185)

Background and Supporting Information

- Scoliosis is defined as a curvature of the spine in the coronal plane. Scoliosis can involve any or all levels of the spine but generally involves the thoracic and/or lumbar spine. Scoliosis initially occurs in the pediatric and adolescent population and persists throughout life. If scoliosis begins in adulthood, it is usually secondary to neurologic disorders (e.g., post-traumatic paralysis) or degenerative spondylosis. Sagittal plane spinal deformity (e.g., kyphosis, hyperlordosis) may be associated with scoliosis.

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Evidence Discussion (SP-14)

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Plain radiography continues to be the primary imaging modality for the initial diagnosis of spinal deformity and for follow up of deformity progression.[13,14] Plain x-rays allow the easy measurement of Cobb angles which remains essential in the evaluation of scoliosis.[13]

Individuals with congenital scoliosis, atypical curves (for example, left thoracic) or abnormal neurological findings will benefit from MRI to help identify spinal cord abnormalities.[13,15,16,17] CT is considered the gold standard for the evaluation of osseous structures and can be useful in the evaluation of complex bony deformity.[13] Additionally, MRI and CT can be valuable for pre-operative evaluation.[18,19,20]

For revision anterior spinal deformity surgery, CT angiography or MR angiography may be medically necessary, however, concerns associated with these modalities are radiation exposure (CT), availability of the imaging modalities in close proximity to individuals, potential out of pocket costs to individuals and sensitivity to individual movement (MRI).[11,21]

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Post-Operative Spinal Disorders (SP-15)

Guideline

Greater than Six Months Post-Operative (SP-15.1)
Routine Post-Fusion Imaging (SP-15.2)
Prolonged Intractable Pain Following Spinal Surgery Within Six Months (SP-15.3)
Revision Anterior Fusion Surgery (SP-15.4)
Evidence Discussion (SP-15)
References (SP-15)

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Greater than Six Months Post-Operative (SP-15.1)

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- Following plain x-rays of the affected spinal regions post-surgical and performed after the current episode of symptoms started or changed with results available to the requesting provider, MRI without and with contrast, MRI without contrast, or CT without contrast of the affected spinal region(s) is medically necessary when:
  - Individual is more than six months post-operative ***AND***
  - Failure of a six-week trial of provider-directed treatment after the current set of symptoms started or changed with clinical re-evaluation ***OR***
  - See: **Red Flag Indications (SP-1.2)**
- See: **Nuclear Medicine (SP-17)** for nuclear medicine imaging when MRI/CT are non-diagnostic in back pain with suspected failed fusion surgery

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Routine Post-Fusion Imaging (SP-15.2)

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- Following a clinically successful spinal fusion, advanced diagnostic imaging is generally not medically necessary.
- **PET** is not currently medically necessary for the routine assessment of spinal fusions or unsuccessful spine surgery (see also: **Spine PET (SP-2.10)**).

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Prolonged Intractable Pain Following Spinal Surgery Within Six Months (SP-15.3)

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- Following plain x-rays of the affected spinal regions post-surgical with results available to the requesting provider, MRI without and with contrast of the affected spinal region(s) is medically necessary if there are residual, new, recurrent, or worsening symptoms related to the spinal region(s) for which surgery was performed within the last 6 months.
  - CT without contrast, or CT with contrast (Myelography) of the affected spinal region(s) if MRI is contraindicated.

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Revision Anterior Fusion Surgery (SP-15.4)

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- If requested by the operating surgeon, the following studies for pre-operative planning prior to surgical revision of a thoracic or lumbar anterior spinal arthrodesis:
  - CTA Pelvis (CPT® 72191) **OR**
  - CTA Abdomen (CPT® 74175) **OR**
  - CTA Abdomen and Pelvis (CPT® 74174) **OR**
  - MRA Pelvis (CPT® 72198) and/or MRA Abdomen (CPT® 74185)

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Evidence Discussion (SP-15)

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Despite advances made in high-resolution spinal imaging, plain films remain integral in providing optimal care for spine individuals and continue to provide critical information that cannot be obtained with other imaging modalities.[6] X-ray imaging with anteroposterior, lateral, oblique, and flexion-extension views is considered the primary imaging modality for post-operative evaluation and can provide complementary information to advanced imaging.[1,7] X-rays can provide information as to whether a concomitant instability is present which would further assist with pre-operative planning. Also, when prior surgery is a concern, x-ray provides additional clinical information as to the details of the hardware for which this detail can many times be obscured with advanced imaging techniques (MRI/CT). An x-ray often has a larger field-of-view than an MRI or CT and has the potential to identify more proximal or distal pathology in the spine that could ultimately assist in determining the individual's diagnosis.[6] X-rays can also determine whether an advanced diagnostic imaging study is actually needed, what specific advanced diagnostic imaging study is warranted and if contrast is required.

There are risks associated with advanced imaging including but not limited to radiation exposure, implanted device complications, metallic foreign body complications and contrast complications.[10] Studies have also linked the increase rate of imaging with the increase rate of surgery and also found early magnetic resonance imaging (MRI) had an eightfold increased risk of surgery.[11,12]

Although most individuals with acute neck or back pain will improve with 6 weeks of conservative care[7,8,9], conservative care would not be necessary for individuals with prolonged intractable pain present within 6 months of surgery or if a red flag indication was present. In general, initial plain x-rays and an initial course of conservative care can provide a significant clinical benefit that would outweigh the clinical harm from perhaps briefly delaying advanced imaging if needed. A course of conservative care or plain x-ray findings many times may obviate the need for advanced imaging which possess their own set of significant risks.

For revision thoracic or lumbar anterior spinal arthrodesis, CT angiography or MR angiography may be medically necessary, however, risks are present with these modalities including radiation exposure (CT), availability of the imaging modalities in close proximity to individuals, potential out-of-pocket costs to individuals, and sensitivity to individual movement (MRI).[5,10]

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Other Imaging Studies and Procedures Related to the Spine Imaging Guidelines (SP-16)

Guideline

Prior to Spine Surgery (SP-16.1)
Prior to Interventional Spinal Injections (SP-16.2)
Prior to Spinal Cord Stimulator (SCS) Placement/Removal (SP-16.3)
Following Vertebral Augmentation Procedures (SP-16.4)
Evidence Discussion (SP-16)
References (SP-16)

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Prior to Spine Surgery (SP-16.1)

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- Advanced imaging needed for surgical planning (e.g., MRI and/or CT) should be performed within the past six (6) months for pre-operative planning prior to spine surgery when the criteria for advanced imaging studies of the spine are met as otherwise stated in the Spine Imaging Guidelines. (See: **MRI of the Spine [SP-2.2]**, **CT of the Spine [SP-2.3]**, **CT/Myelography [SP-2.4]**)
- MR Angiography (MRA) and CT Angiography (CTA) are generally not medically necessary for pre-operative planning of initial anterior spinal surgery unless abnormal vasculature is known or reasonably anticipated.

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Prior to Interventional Spinal Injections (SP-16.2)

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- Advanced diagnostic imaging studies of the spine are not required prior to facet joint injections, medial branch blocks or radiofrequency ablations unless the criteria for advanced imaging studies of the spine are met as otherwise stated in the Spine Imaging Guidelines.
- Advanced diagnostic imaging studies of the cervical spine and/or thoracic spine are medically necessary within twenty-four (24) months prior to interlaminar or transforaminal epidural steroid injections of the cervical and/or thoracic spine when the criteria for advanced imaging studies of the spine are met as otherwise stated in the Spine Imaging Guidelines.
- Advanced diagnostic imaging studies of the lumbar spine are medically necessary prior to transforaminal epidural steroid injections of the lumbar spine when the criteria for advanced imaging studies of the spine are met as otherwise stated in the Spine Imaging Guidelines.
- Advanced diagnostic imaging studies of the lumbar spine are not required prior to lumbar spine interlaminar or caudal epidural steroid injections unless the criteria for advanced imaging studies of the spine are met as otherwise stated in the Spine Imaging Guidelines.
- For an individual with evidence of symptomatic spinal stenosis, MRI or CT with or without myelography demonstrating severe spinal stenosis at the level to be treated within the past twenty-four (24) months is required for an initial trial of a transforaminal, interlaminar or caudal epidural steroid injection when ALL of the following criteria are met:
  - Diagnostic evaluation has ruled out other potential causes of pain
  - Significant functional limitations resulting in diminished quality of life and impaired age-appropriate activities of daily living (ADLs)
  - Failure of at least four (4) weeks of conservative treatment (e.g., exercise, physical methods including physical therapy and/or chiropractic care, NSAIDs, and/or muscle relaxants)
- See: **Red Flag Indications (SP-1.2)** for severe radicular pain

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Prior to Spinal Cord Stimulator (SCS) Placement/Removal (SP-16.3)

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- MRI Thoracic Spine without contrast (CPT® 72146) is generally the study of choice prior to SCS placement. CT Thoracic Spine without contrast (CPT® 72128) **OR** CT/ Myelography Thoracic Spine (CPT® 72129) are acceptable alternatives.
- Imaging of the lumbar spine is not medically necessary for placement nor removal of spinal cord stimulators.

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Following Vertebral Augmentation Procedures (SP-16.4)

SP.OI.0016.4.A

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- CT without contrast of the affected spinal region(s) within 24 hours post-procedure to evaluate neurologic sequelae resulting from cement extravasation

Background and Supporting Information

- MRI has not been shown to change the outcome of interventional pain procedures in recent scientific evidence-based studies and without substantial change in the clinical picture or intervening surgery. Repeat advanced diagnostic imaging studies are not necessary with each spinal injection or series of spinal injections.

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Evidence Discussion (SP-16)

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- Advanced imaging needed for surgical planning (e.g., MRI and/or CT) should be performed for pre-operative planning prior to spine surgery.[9,10] MRA and CTA are generally not medically necessary for pre-operative planning of initial anterior spinal surgery unless abnormal vasculature is known or reasonably anticipated.
- Advanced diagnostic imaging studies of the spine are not required prior to facet joint injections, medial branch blocks or radiofrequency ablations unless the criteria for advanced imaging studies of the spine are met as otherwise stated in the Spine Imaging Guidelines.[5,7]
- MRI Thoracic Spine without contrast is generally the study of choice prior to SCS placement, however, CT Thoracic Spine without contrast or CT/ Myelography Thoracic Spine are acceptable alternatives. Imaging of the lumbar spine is not medically necessary for placement nor removal of spinal cord stimulators.[11]
- CT without contrast of the affected spinal region(s) within 24 hours post-procedure to evaluate neurologic sequelae resulting from cement extravasation.[12]

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Nuclear Medicine (SP-17)

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Nuclear Medicine (SP-17)

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- For evaluation of suspected loosening of orthopedic implants, when recent plain x-ray is non-diagnostic:
  - Bone scan (CPT® 78315) **OR**
  - Distribution of Radiopharmaceutical Agent SPECT (CPT® 78803, or 78831) **OR**
  - SPECT/CT (CPT® 78830)
- Back pain with suspected failed fusion surgery, with suspected painful pseudoarthrosis and MRI/CT are non-diagnostic:
  - Radiopharmaceutical Localization SPECT (CPT® 78803, or 78831) **OR**
  - SPECT/CT (CPT® 78830)
- Any of the following studies are medically necessary for initial evaluation of suspected spine osteomyelitis and for follow-up imaging for evaluation of response to treatment in established spine osteomyelitis:
  - 3-phase bone scan (CPT® 78315) of the spine **OR**
  - Bone SPECT (CPT® 78803 or 78831) or Bone SPECT/CT (CPT® 78830 or 78832) **OR**
    - SPECT CPT® codes include flow and blood pool and delayed SPECT imaging
  - Gallium scan whole-body (CPT® 78804) or Limited spine (CPT® 78801) **OR**
  - Gallium SPECT (CPT® 78803 or 78831) or SPECT/CT (CPT® 78830 or 78832) **OR**
  - Gallium scan whole-body (CPT® 78804) or Limited spine (CPT® 78801) with SPECT (CPT® 78803 or 78831) or SPECT/CT (CPT® 78830 or 78832)
  - **NOTE:** Any bone scan can be combined with any Gallium scan
- Suspected spine infection (such as epidural abscess or discitis osteomyelitis) next imaging study
  - FDG-PET/CT whole-body when x-ray or CT are abnormal **AND** when MRI cannot be performed or is inconclusive[7]
- For evaluation of facet arthropathy in individuals with ankylosing spondylitis, osteoarthritis, or rheumatoid arthritis:
  - Radiopharmaceutical Localization Inflammatory Imaging (one of CPT® codes: 78800, 78801, 78802, or 78803) **OR**
  - Distribution Of Radiopharmaceutical Agent SPECT (CPT® 78803) **OR**
  - SPECT/CT (CPT® 78830)
- For the evaluation of back pain and suspected spondylolysis:
  - Radiopharmaceutical Agent SPECT (CPT® 78803 or 78831) **OR**
  - SPECT/CT (CPT® 78830 or 78832)

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- For the evaluation of a new symptomatic compression fracture identified on radiographs or CT, and no known malignancy:
  - Whole body bone scan with add on SPECT (CPT® 78803) or SPECT/CT (CPT® 78830)
  - See also: **Spinal Compression Fractures (SP-11.1)**

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Evidence Discussion (SP-17)

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The American College of Radiology Appropriateness Criteria for low back pain (revised 2021)[1] states that CT or MRI imaging may be considered for individuals with spinal issues who have had failed at least 6 weeks of medical management or physical therapy. For most individuals with spine issues, MRI and or CT is the study of choice after failed conservative treatments. There is no relevant literature to support the use of bone scan with single-photon emission CT (SPECT) or SPECT/CT in the initial evaluation of acute uncomplicated LBP.

Given the risk of radiation from nuclear medicine imaging it is important to carefully select the proper individual indication. Based on American College of Radiology Appropriateness Criteria for low back pain (revised 2021) and also supported by literature nuclear medicine is used infrequently but is supported for the following indication:

- Structures with abnormal morphology on conventional imaging may not be the cause of LBP. Evidence suggests possible utility of bone scan with SPECT or SPECT/CT as a functional modality to localize the source of LBP, particularly for facet arthropathy.[5]
- SPECT bone scan is the reference standard for detection of radiographically occult active spondylolysis in the young individual.[6]
- Bone scan with SPECT/CT is usually not used for initial imaging but can be useful for radiographically occult fractures and can be used to evaluate acuity of vertebral fracture.
- Bone scan can be used for suspected loosening of orthopedic implants and failed fusion surgery when recent plain x-ray is non-diagnostic. MRI evaluation of these individuals can be significantly limited due to metal artifact from the implants.
- Bone scan and WBS labeled scans can be used for suspected spinal osteomyelitis.

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General Guidelines (PEDAB-1)

Guideline

Procedure Codes Associated with Abdomen Imaging
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Pediatric Abdominal Imaging Age Considerations (PEDAB-1.1)
Pediatric Abdomen Imaging Appropriate Clinical Evaluation and Conservative Treatment (PEDAB-1.2)
Pediatric Abdomen Imaging Modality General Considerations (PEDAB-1.3)
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Procedure Codes Associated with Abdomen Imaging

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MRI	**CPT®**
MRI Abdomen without contrast	74181
MRI Abdomen with contrast (rarely used)	74182
MRI Abdomen without and with contrast	74183
Unlisted MRI procedure (for radiation planning or surgical software)	76498
MRA	**CPT®**
MRA Abdomen	74185
CT	**CPT®**
CT Abdomen without contrast	74150
CT Abdomen with contrast	74160
CT Abdomen without and with contrast	74170
CT Abdomen and Pelvis without contrast	74176
CT Abdomen and Pelvis with contrast	74177
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CT Guidance for Needle Placement (Biopsy, Aspiration, Injection, etc.)	77012
CT Guidance for and monitoring of Visceral Tissue Ablation	77013
CT Guidance for Placement of Radiation Therapy Fields	77014

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Unlisted CT procedure (for radiation planning or surgical software)	76497
CTA	**CPT®**
CTA Abdomen	74175
CTA Abdomen and Pelvis	74174
Nuclear Medicine	**CPT®**
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging: skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging: whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
Adrenal Nuclear Imaging Cortex and/or Medulla	78075
Spleen Imaging Only with or without Vascular Flow	78185
Liver Imaging Static	78201
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Kidney Imaging (Nuclear) Static	78700
Kidney Imaging (Nuclear) with Vascular Flow	78701
Kidney Flow and Function, Single Study without Pharmacologic Intervention	78707
Kidney Imaging with Vascular Flow and Function with Pharmacological Intervention, Single	78708
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Nuclear Non-imaging Renal Function	78725
Ureteral Reflux Study (Radiopharmaceutical Voiding Cystogram)	78740
Radiopharmaceutical Localization Imaging Limited area	78800
Radiopharmaceutical Localization Imaging Whole Body	78802
Radiopharmaceutical Localization Imaging SPECT	78803
Ultrasound	**CPT®**
Ultrasound, abdomen; complete	76700

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Ultrasound, abdomen; limited	76705
Ultrasound, abdominal wall	76705
Ultrasound, retroperitoneal; complete	76770
Ultrasound, retroperitoneal; limited	76775
Ultrasound, transplanted kidney (with duplex Doppler)	76776
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; complete study	93975
Duplex scan of arterial inflow and venous outflow of abdominal, pelvic, scrotal contents and/or retroperitoneal organs; limited study	93976
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; complete	93978
Duplex scan of aorta, inferior vena cava, iliac vasculature, or bypass grafts; limited	93979

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General Guidelines (PEDAB-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms including a detailed history, physical examination, appropriate laboratory studies and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.
- These guidelines are based upon using advanced imaging to answer specific clinical questions that will affect individual management. Imaging is not indicated if the results will not affect individual management decisions. Standard medical practice would dictate continuing conservative therapy prior to advanced imaging in individuals who are improving on current treatment programs.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the abdomen is not supported. Advanced imaging should only be approved in individuals who have documented active clinical signs or symptoms of disease.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the same body area are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.
- Ultrasound should be the initial imaging study of choice in most children with abdominal conditions and should be done prior to advanced imaging.
  - For those individuals who do require advanced imaging after ultrasound, ultrasound can be very beneficial in selecting the proper modality, body area, image sequences, and contrast level that will provide the most definitive information for the individual.
  - CPT® codes vary by body area and presence or absence of Doppler imaging and are included in the table at the beginning of this guideline.

Red Flags

- Children with abdominal pain AND ANY of the following red flag signs or symptoms require additional investigation. The initial ultrasound is not required. Additional labs may be helpful but are not required.
  - Pain that wakes the child from sleep
  - Unexplained fever (T >100.4°F)
  - History of malignancy with a likelihood or propensity to metastasize to abdomen

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  - Dysphagia
  - GI bleeding
  - Significant vomiting
  - Elevated WBC per the testing laboratory's range
  - Low WBC (absolute neutrophil count <1000)
  - Guarding, rebound tenderness, or other peritoneal signs
  - Severe chronic diarrhea or nocturnal diarrhea in a toilet-trained child
  - Failure to thrive, involuntary weight loss, or delay in linear growth or pubertal development
  - Family history of inflammatory bowel disease, familial polyposis syndrome, celiac disease, or peptic ulcer disease
  - Abdominal mass, hepatomegaly, and/or splenomegaly on exam
  - Jaundice
  - Perianal disease
  - Suspected or confirmed COVID-19 infection with concern for multisystem inflammatory syndrome in children (MIS-C)
  - Failure to respond to 4 weeks of recent (within 60 days) provider directed conservative care
- See the condition-specific sections for when the above list of exclusionary criteria apply and lead directly to advanced imaging.
- The appropriate advanced imaging for the condition is listed in the condition-specific section.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and, language and literacy skills.

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Pediatric Abdominal Imaging Age Considerations (PEDAB-1.1)

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- Many conditions affecting the abdomen in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases that occur in both pediatric and adult populations, differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals age 18 years old and younger should be imaged according to the Pediatric Abdomen Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Abdomen Imaging Guidelines should be imaged according to the General Abdomen Imaging Guidelines. Individuals age >18 years old should be imaged according to the General Abdomen Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Pediatric Abdomen Imaging Appropriate Clinical Evaluation and Conservative Treatment (PEDAB-1.2)

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- See: **General Guidelines (PEDAB-1.0)**

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Pediatric Abdomen Imaging Modality General Considerations (PEDAB-1.3)

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- Ultrasound
  - See: **General Guidelines (PEDAB-1.0)**
- MRI
  - MRI Abdomen is generally performed without and with contrast (CPT® 74183) unless the individual has a documented contraindication to gadolinium or otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize individual movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition-based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Recent evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported by these guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same session.
  - The presence of surgical hardware or implanted devices may preclude MRI.

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        - The selection of best examination may require coordination between the provider and the imaging service. CT may be the procedure of choice in these cases.
- CT
    - CT Abdomen typically extends from the dome of the diaphragm to the upper margin of the sacroiliac joints, and CT Abdomen and Pelvis extends from the dome of the diaphragm through the ischial tuberosities.
        - In general, CT Abdomen is appropriate when evaluating solid abdominal organs.
        - In general, CT Abdomen and Pelvis is appropriate when evaluating inflammatory or infectious processes, hematuria, or conditions that appear to involve both the abdomen and the pelvis.
        - In some cases, especially in follow-up of a known finding, it may be appropriate to limit the exam to the region of concern to reduce radiation exposure.
    - The contrast level in pediatric CT imaging is specific to the clinical indication, as listed in the specific guideline sections.
    - CT Abdomen or Abdomen and Pelvis may be indicated for further evaluation of abnormalities suggested on prior US or MRI studies.
    - CT may be indicated without prior MRI or US as indicated in specific sections of these guidelines.
    - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
    - The selection of the best examination may require coordination between the provider and the imaging service.
- Nuclear Medicine
    - Nuclear medicine studies are commonly used in evaluation of the pediatric kidney and gallbladder. Other less common indications exist as well:
        - Esophageal motility study (CPT® 78258) and/or Gastroesophageal reflux study (CPT® 78262) is indicated in the evaluation of gastroesophageal reflux.
    - Nuclear intestinal imaging (preferred code for Meckel's Scan, CPT® 78290) or Gastric mucosa imaging (alternate code for Meckel's scan, CPT® 78261) is indicated for the following:
        - Suspected Meckel's diverticulum
        - Gastric mucosa imaging (CPT® 78261) is also indicated for:
            - Barrett's esophagus
            - Thoracic masses suspected of containing gastric mucosa
    - Gastric emptying study (CPT® 78264) is indicated for evaluation of either suspected delayed or rapid gastric emptying.
    - Gastric emptying study with small bowel transit (CPT® 78265) is indicated for evaluation of suspected abnormalities in both total and regional times for gastrointestinal transit of the small bowel.

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  - Gastric emptying study with small bowel and colon transit (CPT® 78266) is indicated for evaluation of suspected abnormalities in both total and regional times for gastrointestinal transit to the colon.
  - Gastrointestinal bleeding scintigraphy (CPT® 78278) is indicated for evaluation of brisk active GI bleeding with indeterminate endoscopy.
  - Gastrointestinal protein loss study (CPT® 78282) is indicated for decreased serum albumin or globulins and no evidence of GI bleeding.
  - Peritoneal-venous shunt patency study (CPT® 78291) is indicated for evaluation of shunt patency and function in an individual with ascites.
  - Nuclear renal imaging (CPT® 78701, CPT® 78707, CPT® 78708, or CPT® 78709) is indicated for evaluation of the following:
    - Renal transplant follow-up
    - Kidney salvage vs. nephrectomy surgical decisions
    - Acute renal failure with no evidence of obstruction on recent ultrasound
    - Chronic renal failure to estimate prognosis for recovery
- 3D Rendering
  - 3D Rendering indications in pediatric abdomen imaging are identical to those in the general imaging guidelines. See: **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Generalized Abdominal Pain (PEDAB-2)

Guideline

Generalized Abdominal Pain (PEDAB-2)
References (PEDAB-2)

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Generalized Abdominal Pain (PEDAB-2)

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- Chronic abdominal pain is a common complaint among children and adolescents. Chronic is defined as abdominal pain lasting for over 2 months.
- Functional abdominal pain disorders, including functional abdominal pain and functional dyspepsia (including symptoms of epigastric pain, burning, postprandial fullness, early satiety), are conditions in which there is no structural or organic disease.
- Ultrasound Abdomen (CPT® 76700 or CPT® 76705) is medically necessary as the initial imaging for children with no red flag signs or symptoms, normal physical examination, and normal laboratory studies (preliminary labs may include CBC, electrolytes, lipase, amylase, urinalysis, ESR, CRP, LFTs, and/or stool for blood and stool culture if diarrhea)
  - Gastroenterology (GI) specialist evaluation or consultation is helpful in determining the need for advanced imaging in these cases.
  - CT Abdomen (CPT® 74160) with contrast or CT Abdomen and Pelvis (CPT® 74177) with contrast is medically necessary if there are any red flag signs or symptoms (as listed in **General Guidelines (PEDAB-1.0)**).
- Children with abdominal pain that can be localized to a particular area of the abdomen should be imaged according to the relevant guideline section:
  - **Right Lower Quadrant Pain (PEDAB-3)**
  - **Flank Pain, Renal Stone (PEDAB-4)**
  - **Right Upper Quadrant Pain (PEDAB-8)**
  - **Left Upper Quadrant Pain (PEDAB-25)**
  - **Left Lower Quadrant Pain (PEDAB-29)**

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Right Lower Quadrant Pain (PEDAB-3)

Guideline

Right Lower Quadrant Pain (PEDAB-3)
References (PEDAB-3)

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Right Lower Quadrant Pain (PEDAB-3)

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- The presence of any red flag findings per **General Guidelines (PEDAB-1.0)** precludes adjudication based on any other criteria.
- Ultrasound (CPT® 76700 or CPT® 76705) is medically necessary as the initial examination. If positive or negative for uncomplicated appendicitis, no further imaging is necessary.
  - If the appendix is not visualized on ultrasound and the white blood cell count is not elevated, no further imaging is necessary in nearly all cases, although the referring physician should make the final determination of the need for advanced imaging.
- CT Abdomen and Pelvis with contrast (CPT® 74177), MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195), or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary for any of the following:
  - Individuals who are overweight (BMI ≥85th percentile for age)
  - Insufficient local ultrasound expertise exists
  - Ultrasound findings are inconclusive
  - Clinical suspicion of or initial imaging suggestive of complication (e.g., abscess, bowel obstruction)
- If the appendix is absent, follow guidelines in: **Generalized Abdominal Pain (PEDAB-2)**

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Flank Pain, Renal Stone (PEDAB-4)

Guideline

Flank Pain, Renal Stone (PEDAB-4)
References (PEDAB-4)

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Flank Pain, Renal Stone (PEDAB-4)

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- The presence of any red flag findings per **General Guidelines (PEDAB-1.0)** precludes adjudication based on any other criteria.
- Flank Pain imaging indications in pediatric individuals are very similar to those for adult individuals. See: **Flank Pain, Rule Out or Known Renal/Ureteral Stone (AB-4)** in the Abdomen Imaging Guidelines.
  - Ultrasound (CPT® 76770 or CPT® 76775) is the medically necessary initial study in children.
  - CT Abdomen and Pelvis without contrast (CPT® 74176) is medically necessary if ultrasound is inconclusive.
  - MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast is medically necessary if CT is inconclusive or if significant concern for radiation exposure from frequent CT use for a particular individual.
  - If hematuria is present, see: **Hematuria (PEDAB-7)** for imaging guidelines.
- Nuclear kidney imaging (CPT® 78707, CPT® 78708, CPT® 78709, or CPT® 78803) is medically necessary for evaluation of recurrent flank pain when CT and ultrasound are non-diagnostic, or for suspected obstructive uropathy.

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Urinary Tract Infection (UTI) (PEDAB-5)

Guideline

Upper Urinary Tract (PEDAB-5.1)
Lower Urinary Tract (PEDAB-5.2)
References (PEDAB-5)

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Upper Urinary Tract (PEDAB-5.1)

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- Ultrasound evaluation (CPT® 76770 or CPT® 76775) is the medically necessary initial imaging for all children with first-time upper urinary tract infection to diagnose hydronephrosis, pyelonephritis, or congenital renal anomaly.
  - If hydronephrosis is present, this should be further evaluated with voiding cystourethrography (VCUG), to evaluate for vesicoureteral reflux.
    - In boys, this is generally accomplished using fluoroscopic imaging and iodinated contrast to exclude urethral abnormalities.
    - In girls, Ureteral Reflux Study (Radiopharmaceutical Voiding Cystogram) (CPT® 78740) or fluoroscopic VCUG may be performed.
    - Contrast Enhanced Voiding Urosonography (CeVUS) may also be utilized at institutions with expertise in this modality.
- Diuretic renography using Tc-99m MAG 3 (CPT® 78707, CPT® 78708, or CPT® 78709) is medically necessary for:
  - differentiating a dilated non-obstructed urinary system from a true stenosis (e.g., UPJ obstruction; ureteral-vesical junction [UVJ] obstruction)
  - quantifying renal parenchymal function
  - ultrasound findings that are compatible with a multicystic dysplastic kidney to evaluate function of the affected kidney or a ureteral-pelvic junction (UPJ) obstruction of the contralateral kidney
  - diagnostic evaluation of upper tract dilatation when VCUG is negative
  - renal function evaluation in individuals with hydronephrosis
- CT is sensitive in diagnosing pyelonephritis and has a role in evaluation of renal abscess or unusual complications such as xanthogranulomatous pyelonephritis but has no role in the routine evaluation of UTI.
  - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary to evaluate the entire genitourinary tract for congenital abnormalities of distal tracts in complicated pyelonephritis.
- Magnetic resonance urography (MRU) (CPT® 74183 and CPT® 72197), is not a first-line test for the routine evaluation of a UTI but is medically necessary (where available) for investigation of a dilated upper urinary tract.
  - NOTE: MRU requires sedation in young children.
  - MRU can also quantitate renal function.
- Technetium-99m-dimercaptosuccinic acid (Tc-99m DMSA) scintigraphy (CPT® 78700, CPT® 78701, or CPT® 78803), is sensitive for evaluation of renal cortical damage.
  - DMSA scintigraphy is medically necessary for:
    - individuals with atypical or recurrent febrile acute urinary tract infections

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    - Atypical findings may include poor response to antibiotics, elevated creatinine, poor urine stream, or non E-coli organism.
  - individuals with febrile urinary tract infections older than 5 years of age with known vesicoureteral reflux
  - detection of post-pyelonephritic renal scarring at least 6 months after the documented upper tract UTI in high-risk individuals with recurrent UTIs
- Radiopharmaceutical nuclear medicine imaging (CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, CPT® 78830, CPT® 78831, or CPT® 78832) is medically necessary for evaluation of suspected pyelonephritis or diffuse interstitial nephritis.
- Nuclear non-imaging renal function study (CPT® 78725) is a quantitative study that can be used to evaluate renal function.
- US Kidneys and bladder (CPT® 76770 or CPT® 76775) (preferred) and/or Voiding Cystourethrography (CPT® 78740) are medically necessary for children with atypical (poor response to antibiotics within 48 hours, sepsis, poor urine stream, raised creatinine, or non-E. coli UTI) or recurrent febrile UTI.

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Lower Urinary Tract (PEDAB-5.2)

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- Ultrasound evaluation (CPT® 76770 or CPT® 76775) is medically necessary as the initial imaging for all children with first time UTI to diagnose hydronephrosis, pyelonephritis, or congenital renal anomaly.
  - Fluoroscopic Voiding cystourethrography (VCUG) is medically necessary for detection of possible vesico-ureteral reflux (VUR) in neonates or young children when hydronephrosis is seen on ultrasound.
- The American Academy of Pediatrics clinical practice guidelines no longer recommend routine VCUG for females, 2 to 24 months of age, after the first febrile UTI.
  - The current recommendation is to postpone the VCUG until the second febrile UTI UNLESS there are:
    - atypical or complex clinical circumstances
    - renal/bladder ultrasound findings that reveal hydronephrosis, scarring, or obstructive uropathy
- Vesicoureteral Reflux (VUR)
  - Fluoroscopic VCUG is typically performed for diagnosis and grading of VUR, and is the medically necessary first modality used for diagnosis.
  - Ureteral Reflux Study (Radiopharmaceutical Voiding Cystogram) (CPT® 78740), fluoroscopic VCUG, or CeVUS are medically necessary for follow up imaging of VUR.
- Male individuals with first UTI should be evaluated with fluoroscopic VCUG studies rather than radionuclide cystography, to visualize the male urethra for possible abnormalities such as posterior urethral valves, strictures, or diverticula.
- For female individuals, radionuclide cystography (CPT® 78740) or fluoroscopic VCUG are medically necessary as the initial study.
- MR urography is medically necessary for evaluation of ectopic distal ureteral insertion, or other complex lower urinary tract anatomy.
- Siblings of individuals with known vesicoureteral reflux can undergo Ureteral Reflux Study (Radiopharmaceutical Voiding Cystogram) (CPT® 78740) if they have renal scarring on ultrasound or history of UTI and no prior evaluation for VUR.

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Pediatric Acute Gastroenteritis (PEDAB-6)

Guideline

Pediatric Acute Gastroenteritis (PEDAB-6)
References (PEDAB-6)

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Pediatric Acute Gastroenteritis (PEDAB-6)

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- Advanced imaging is not medically necessary in pediatric acute gastroenteritis unless there is a concern for diagnosis other than acute gastroenteritis. See specific symptom/diagnosis sections listed below.
- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if abdominal red flag symptoms are present as listed in **General Guidelines (PEDAB-1.0)**.
- Additional imaging studies will depend on the specific symptoms. See the following sections for additional imaging guidelines:
  - **Generalized Abdominal Pain (PEDAB-2)** in the Pediatric Abdomen Imaging Guidelines
  - **Right Lower Quadrant Pain (PEDAB-3)** in the Pediatric Abdomen Imaging Guidelines
  - **Right Upper Quadrant Pain (PEDAB-8)** in the Pediatric Abdomen Imaging Guidelines
  - **Inflammatory Bowel Disease, Crohn Disease, or Ulcerative Colitis (PEDAB-9)** in the Pediatric Abdomen Imaging Guidelines
  - **Constipation, Diarrhea, and Irritable Bowel Syndrome (PEDAB-12)** in the Pediatric Abdomen Imaging Guidelines
  - **Abdominal Mass (PEDAB-13)** in the Pediatric Abdomen Imaging Guidelines
  - **Left Upper Quadrant (PEDAB-25)** in the Pediatric Abdomen Imaging Guidelines
  - **Intussusception (PEDAB-27)** in the Pediatric Abdomen Imaging Guidelines
  - **Bowel Obstruction (PEDAB-28)** in the Pediatric Abdomen Imaging Guidelines
  - **Left Lower Quadrant Pain (PEDAB-29)** in the Pediatric Abdomen Imaging Guidelines

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Hematuria (PEDAB-7)

Guideline

Hematuria (PEDAB-7)
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Hematuria (PEDAB-7)

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Hematuria is a relatively common complaint in pediatric individuals, and the imaging considerations are different from those occurring in adult individuals.

- Ultrasound kidneys (CPT® 76770 or CPT® 76775) and bladder (CPT® 76856 or CPT® 76857) are medically necessary for asymptomatic gross hematuria or microscopic hematuria with proteinuria present.
- Imaging is not considered medically necessary for asymptomatic microscopic hematuria without proteinuria.
- For painful hematuria and no recent trauma, ANY of the following studies are medically necessary:
  - CT Abdomen and Pelvis without contrast (CPT® 74176)
  - Ultrasound kidneys (CPT® 76770 or CPT® 76775)
  - Ultrasound bladder (CPT® 76856 or CPT® 76857)
- For hematuria and recent trauma, the following studies are medically necessary:
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
- CT Cystography (CT Pelvis with bladder contrast – CPT® 72193) is medically necessary if gross hematuria is present and pelvic fracture or traumatic bladder injury is suspected.
- 3-D reconstruction (CPT® 76377 or CPT® 76376) is medically necessary, if requested, for surgical planning.

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References (PEDAB-7)

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Right Upper Quadrant Pain (PEDAB-8)

Guideline

Right Upper Quadrant Pain (PEDAB-8)
References (PEDAB-8)

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Right Upper Quadrant Pain (PEDAB-8)

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- The presence of any red flag findings per **General Guidelines (PEDAB-1.0)** precludes adjudication based on any other criteria.
- Right upper quadrant pain imaging indications in pediatric individuals are very similar to those for adult individuals. See: **Abdominal Pain (AB-2)** in the Abdomen Imaging Guidelines.
  - US Abdomen (CPT® 76700) and/or Nuclear medicine imaging of the hepatobiliary system (HIDA scan, CPT® 78226 or CPT® 78227) is medically necessary for initial diagnosis of:
    - suspected acute cholecystitis or acalculous cholecystitis (symptoms may include RUQ pain with fever, elevated white blood cell count, positive Murphy sign).
    - suspected gallstones and bile duct obstruction (symptoms may include RUQ pain, no fever, normal white blood cell count).
    - Ultrasound results are not needed prior to nuclear medicine imaging of the hepatobiliary system (HIDA scan, CPT® 78226 or CPT® 78227).
  - MRI Abdomen with and without contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160) is medically necessary when either US or NM is equivocal.
  - In individuals with complaints of RUQ pain with no fever and an ultrasound shows only gallstones, MRI Abdomen without contrast (CPT® 74181), MRI Abdomen without and with contrast (CPT® 74183), or Nuclear medicine imaging of the hepatobiliary system (HIDA scan, CPT® 78226 or CPT® 78227) is medically necessary to exclude other sources of pain.

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Inflammatory Bowel Disease, Crohn Disease, or Ulcerative Colitis (PEDAB-9)

Guideline

Inflammatory Bowel Disease, Crohn Disease, or Ulcerative Colitis (PEDAB-9)
References (PEDAB-9)

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Inflammatory Bowel Disease, Crohn Disease, or Ulcerative Colitis (PEDAB-9)

ABP.IB.0009.A

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- Enterography is the medically necessary advanced imaging study for individuals with inflammatory bowel disease (IBD).
  - MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195) is generally preferred over CT when possible to avoid radiation exposure for children.

Children with Suspected Crohn Disease

Clinical features including weight loss, loose stools, vomiting, and intermittent abdominal pain. Small bowel imaging can provide important information to guide treatment relating to presence, severity, and extent of Crohn's disease and its complications. Initial evaluation typically includes laboratory evaluation and upper and lower endoscopy.

- MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195), CT Enterography (CPT® 74177), or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary for ANY of the following:
  - To detect severity and distribution of inflammatory changes
  - To identify complications (such as fistulizing disease or abscess formation)
- MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195) or CT Enterography (CPT® 74177) is medically necessary for evaluation of chronic abdominal pain associated with diarrhea due to concern for inflammatory bowel disease if:
  - there is a positive family history of inflammatory bowel disease, **OR**
  - there are endoscopy or colonoscopy findings suggestive of inflammatory bowel disease, **OR**
  - elevated inflammatory markers (fecal lactoferrin ≥4.0ug/g, CRP >0.5mg/dL, or fecal calprotectin ≥50ug/g), **OR**
  - diagnosis is still in doubt after colonoscopy and evaluation of inflammatory markers, and Crohn's disease is suspected.
- MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195) or CT Enterography (CPT® 74177) is medically necessary prior to endoscopy if requested by or in consultation with the physician who will be performing the endoscopy.

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- MRI Pelvis with contrast (CPT® 72196) or MRI Pelvis without and with contrast (CPT® 72197) is medically necessary for the following:
  - Concern for perianal fistula or abscess
- See: **IBD (Crohn's Disease or Ulcerative Colitis) (AB-23.1)** in the Abdomen Imaging Guidelines for additional information regarding serologic markers

Children with Established IBD

- MR Enterography (CPT® 74183 or CPT® 74181 and CPT® 72197 or CPT® 72195), CT Enterography (CPT® 74177), or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197), is medically necessary for ANY of the following:
  - Monitoring response to disease-modifying treatment on an annual basis or when treatment change is being considered
  - Monitoring post-operative recurrence
  - Individuals with new or worsening symptoms or suspected complications including abscess, perforation, fistula, or obstruction
- CT Abdomen and Pelvis with contrast (CPT 74177) is medically necessary if requested (instead of CTE or MRE) for ANY of the following:
  - New or worsening symptoms
  - Suspected complications including abscess, perforation, fistula, or obstruction
- MRI Pelvis with contrast (CPT® 72196) or MRI Pelvis without and with contrast (CPT® 72197) is medically necessary for the following:
  - Concern for perianal fistula or abscess

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Abdominal Sepsis (Suspected Abdominal Abscess) (PEDAB-10)

Guideline

Abdominal Sepsis (Suspected Abdominal Abscess) (PEDAB-10)

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Abdominal Sepsis (Suspected Abdominal Abscess) (PEDAB-10)

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- Abdominal sepsis imaging indications in pediatric individuals are identical to those for adult individuals.
  - See: **Abdominal Sepsis (Suspected Abdominal Abscess) (AB-3)** in the Abdomen Imaging Guidelines.

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Postoperative Pain within 60 Days Following Abdominal Surgery (PEDAB-11)

Guideline

Postoperative Pain within 60 Days Following Abdominal Surgery (PEDAB-11)
References (PEDAB-11)

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Postoperative Pain within 60 Days Following Abdominal Surgery (PEDAB-11)

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- Any of the following are medically necessary in individuals with suspected postoperative complications (e.g., bowel obstruction, abscess, anastomotic leak, etc.):
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
  - Ultrasound Abdomen (CPT® 76700 or CPT® 76705)
  - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
  - MRI Abdomen and Pelvis without contrast (CPT® 74181 and CPT® 72195)
    - Because MRI may not be practical for the timely evaluation of post-operative abscesses, MRI should only replace CT when the study can be completed in a similar time frame as CT.
- Radiopharmaceutical nuclear medicine imaging (CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, CPT® 78830, CPT® 78831, or CPT® 78832) is medically necessary for evaluation of any of the following:
  - Peritonitis
  - Postoperative fever without localizing signs or symptoms
- Beyond 60 days post-operatively, see: **Generalized Abdominal Pain (PEDAB-2)**.

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Constipation, Diarrhea, and Irritable Bowel Syndrome (PEDAB-12)

Guideline

Constipation, Diarrhea, and Irritable Bowel Syndrome (PEDAB-12)
References (PEDAB-12)

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Constipation, Diarrhea, and Irritable Bowel Syndrome (PEDAB-12)

ABP.DC.0012.A
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- Constipation and diarrhea are extremely common complaints in children. The overwhelming majority of individuals do not require advanced imaging for evaluation of constipation or diarrhea.
- Irritable bowel is rare in young children, but more common in adolescents. The overwhelming majority of individuals do not require advanced imaging for evaluation of irritable bowel syndrome.
  - In most cases, causes of constipation can be excluded based on a careful history and physical examination. Advanced Imaging should be performed if warning signs of other diseases are present.
- Constipation associated with additional signs or symptoms:
  - CT Abdomen (CPT® 74160) or Abdomen and Pelvis (CPT® 74177) with contrast is medically necessary if there are any red flag signs or symptoms (as listed in **General Guidelines (PEDAB-1.0)**)
  - Clinical suspicion of tethered cord (e.g., abnormal physical findings over the spine, abnormal neurological exam, or symptoms refractory to provider-directed treatment for at least 3 months), see: **Spinal Dysraphism and Tethered Cord (PEDSP-4)** in the Pediatric and Special Populations Spine Imaging Guidelines.
- Diarrhea associated with additional signs or symptoms:
  - CT Abdomen (CPT® 74160) with contrast or CT Abdomen and Pelvis (CPT® 74177) with contrast is medically necessary if there are any red flag signs or symptoms (as listed in **General Guidelines (PEDAB-1.0)**).
  - See: **Inflammatory Bowel Disease, Crohn Disease, or Ulcerative Colitis (PEDAB-9)** for concerns regarding suspected inflammatory bowel disease.
- Irritable bowel syndrome associated with additional signs or symptoms:
  - CT Abdomen (CPT® 74160) with contrast or CT Abdomen and Pelvis (CPT® 74177) with contrast is medically necessary if there are any red flag signs or symptoms (as listed in **General Guidelines (PEDAB-1.0)**).
- Hirschsprung disease:
  - MRI Pelvis without and with contrast (CPT® 72197) is medically necessary post-operatively in individuals who have signs of complications related to treatment to assess the position of the pulled-through bowel, the sphincter muscles, and the area of the posterior urethra.

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Abdominal Mass (PEDAB-13)

Guideline

Abdominal Wall Mass (PEDAB-13.1)
Intra-Abdominal Mass (PEDAB-13.2)
References (PEDAB-13)

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Abdominal Wall Mass (PEDAB-13.1)

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- For initial imaging of a newly discovered abdominal wall mass, ANY of the following studies are medically necessary:
  - Ultrasound (CPT® 76700 or CPT® 76705)
  - MRI Abdomen without contrast (CPT® 74181) or without and with contrast (CPT® 74183)
  - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) may be added to MRI Abdomen if below the umbilicus.
- If ultrasound and/or MRI are inconclusive or insufficient for preoperative planning, ANY of the following studies are medically necessary:
  - CT Abdomen with contrast (CPT® 74160) or without contrast (CPT® 74150)
  - CT Abdomen and Pelvis with contrast (CPT® 74177) or without contrast (CPT® 74176) if below the umbilicus

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Intra-Abdominal Mass (PEDAB-13.2)

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- Ultrasound (CPT® 76700) is medically necessary as the initial imaging study for children with an intra-abdominal mass.
  - US with Doppler (CPT® 93975) is medically necessary to evaluate vascular supply.
- Additional imaging studies will be determined by the results of the ultrasound, and will depend on the location and organ involvement associated with the mass as well as history, physical exam, and laboratory findings. See the following sections for additional imaging guidelines:
  - **General Guidelines (PEDONC-1)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Pediatric Lymphomas (PEDONC-5)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Neuroblastoma (PEDONC-6)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Pediatric Renal Tumors (PEDONC-7)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Pediatric Germ Cell Tumors (PEDONC-10)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Pediatric Liver Tumors (PEDONC-11)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Pediatric Adrenocortical Carcinoma (PEDONC-14)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - **Liver Lesion Characterization (PEDAB-15)**
  - **Adrenal Lesions (PEDAB-17)**
  - **Indeterminate Renal Lesion (PEDAB-19)**
  - **Spleen (PEDAB-26)**

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Renovascular Hypertension and Other Secondary Causes of Hypertension (PEDAB-14)

Guideline

Renovascular Hypertension and Other Secondary Causes of Hypertension (PEDAB-14)
References (PEDAB-14)

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Renovascular Hypertension and Other Secondary Causes of Hypertension (PEDAB-14)

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- Clinical evaluation for suspected hypertension should include repeated blood pressure measurements (generally ≥3 measurements).
  - Trained health care professionals in the office setting should make a diagnosis of hypertension (HTN) if a child or adolescent has auscultatory-confirmed blood pressure (BP) readings ≥95th percentile at 3 different visits.
  - Blood pressure can be obtained in-clinic, at home, or by using a wearable ambulatory blood pressure measurement (ABPM) device that records blood pressure at frequent intervals during normal activities and is downloaded later for computer analysis.

For Children Aged 1-<13 years	For Children Aged ≥13 years
Normal BP: <90th Percentile	Normal BP: <120/<80 mm Hg
Elevated BP: ≥90th percentile to <95th percentile or 120/80 mm Hg to <95th percentile (whichever is lower)	Elevated BP: 120/<80 to 129/<80 mm Hg
Stage 1 HTN: ≥95th percentile to <95th percentile + 12 mmHg, or 130/80 to 139/89 mm Hg (whichever is lower)	Stage 1 HTN: 130/80 to 139/89 mm Hg
Stage 2 HTN: ≥95th percentile + 12 mm Hg, or ≥140/90 mm Hg (whichever is lower)	Stage 2 HTN: ≥140/90 mm Hg

- ANY of the following studies are medically necessary for initial evaluation of a pediatric individual with suspected secondary hypertension:
  - Doppler or Duplex Ultrasound (CPT® 93975 or CPT® 93976)
  - Complete retroperitoneal Ultrasound (CPT® 76770)

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- Captopril renography (CPT® 78709) has largely been abandoned in clinical practice, replaced by CTA and MRA Abdomen, but may be supported for unusual circumstances.

Other considerations for imaging evaluation:

- CTA Abdomen (CPT® 74175) or MRA Abdomen (CPT® 74185) is medically necessary for pediatric individuals with hypertension to exclude fibromuscular dysplasia or other blood-flow restricting lesions of the renal arteries and suprarenal aorta.
- Children with high clinical suspicion for renin-mediated hypertension should undergo additional imaging whether Doppler US findings are positive or negative due to poor sensitivity for detecting distal intrarenal or accessory renal artery stenosis. CTA (CPT® 74175) has high sensitivity and specificity, and is medically necessary for these individuals.
- Echocardiography (CPT® 93306) is medically necessary at initial evaluation to screen for cardiac abnormalities, coarctation of the aorta, and end-organ damage such as left ventricular hypertrophy.
- Nuclear renal imaging (CPT® 78707, CPT® 78708, or CPT® 78709) is medically necessary for evaluation of the following:
  - Severe hypertension with progressive renal insufficiency or failure to respond to three-drug therapy
  - Malignant or accelerated hypertension
  - Acute worsening of previously stable hypertension
  - Diastolic BP >100 in an individual <35 years old
  - New onset severe hypertension
  - Hypertension in presence of asymmetric kidneys
  - Hypertension in presence of acute elevation in creatinine either unexplained or after treatment with ACE inhibitor
  - Abdominal bruit
  - Recurrent acute pulmonary edema and hypertension
  - Hypokalemia with normal or elevated plasma renin level in absence of diuretic therapy
  - Hypertension with known neurofibromatosis

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Liver Lesion Characterization (PEDAB-15)

Guideline

Liver Lesion Characterization (PEDAB-15)
References (PEDAB-15)

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Liver Lesion Characterization (PEDAB-15)

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- *High-risk individuals:
  - Prematurity
  - Low birth weight
  - Underlying chronic liver disease
  - Beckwith-Weidman syndrome
  - Familial adenomatous polyposis
  - Trisomy 18
  - Portosystemic shunts
  - Aicardi syndrome
  - Hereditary tyrosinemia
  - Bile salt export pump deficiency
- Pediatric-specific imaging considerations includes:
  - US Abdomen (CPT® 76700 or CPT® 76705) is medically necessary as the initial study in children. MRI is preferred over CT when possible to reduce radiation exposure.
- Liver lesion discovered on US:
  - Any high-risk individual*
    - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160)
  - Indeterminate liver lesion <3cm
    - Contrast-Enhanced US (CEUS, CPT® 76978, CPT® 76979)
    - If after Contrast-Enhanced US the lesion remains indeterminate or not fully characterized, or if Contrast-Enhanced US is not available:
      - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160)
  - Indeterminate liver lesion >3cm
    - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160)
- Liver lesion discovered on CT (non-contrast or single-contrast) or non-contrast MRI
  - Indeterminate Liver Lesion <3cm
    - Contrast-Enhanced US (CEUS, CPT® 76978, CPT® 76979)
    - If, after Contrast-Enhanced US, the lesion remains indeterminate or not fully characterized, or if Contrast-Enhanced US is not available:

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        - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160)
    - Indeterminate Liver Lesion >3cm
        - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160)
- For the imaging of specific focal liver lesions:
    - Suspected hepatic adenoma:
        - CT Abdomen (CPT® 74160 or CPT® 74170) or MRI Abdomen (CPT® 74183) every 6 months for 2 years, and then annually, to establish any growth patterns and assess for malignant transformation.
    - Hepatic Hemangioma
        - Limited US Abdomen in 6-12 months
            - If stable or decreasing in size: Surveillance 6-12 months until 2 years. After 2 years, no further follow-up is considered medically necessary unless it becomes symptomatic
            - If increasing in size: follow-up 3-6 months or biopsy
            - See below for pre-operative considerations
    - Focal Nodular Hyperplasia (FNH):
        - MRI Abdomen (CPT® 74183) or CT Abdomen (CPT® 74160 or CPT® 74170) to confirm a diagnosis of FNH. The use of Eovist contrast is often diagnostic in differentiating FNH from other lesions seen on MRI or CT.
        - Additional follow-up is limited abdominal US in 6-12 months
            - If stable or decreasing in size: Surveillance 6-12 months until 2 years. After 2 years, no further follow-up is considered medically necessary unless it becomes symptomatic.
            - In adolescent females diagnosed with FNH who are continuing to use oral contraceptives: Additional follow-up is annual US for 2 to 3 years. Follow-up with CT or MRI is medically necessary if the lesion is not adequately visualized on US.
            - If increasing in size: follow-up 3-6 months or biopsy
    - Hepatic cysts:
        - Asymptomatic, simple cysts
            - Limited US Abdomen in 6-12 months
            - If stable or decreasing in size: Surveillance 6-12 months until 2 years. After 2 years, no further follow-up is considered medically necessary unless it becomes symptomatic
            - If increasing in size: follow-up 3-6 months or biopsy
        - For complicated cysts (US shows internal septations, fenestrations, calcifications, irregular walls, as well as the presence of daughter cysts):

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  - MRI Abdomen (CPT® 74183) or CT Abdomen (CPT® 74160 or CPT® 74170) is medically necessary
- Additional scenarios and follow-up imaging for an indeterminate lesion:
  - Indeterminate lesion <1cm in high-risk individuals* on US, CT, or unenhanced MRI not specifically dealt with in the above guidelines:
    - If **biopsy cannot be performed**, follow-up MRI is medically necessary at 3-6 months. Additional imaging in this setting can be considered on an individual basis. This timeframe would also apply if the lesion is indeterminate and an MRI with Eovist is requested for further evaluation in this setting.
- Nuclear medicine liver imaging (ONE of CPT® codes: CPT® 78201, CPT® 78202, CPT® 78803, CPT® 78215, or CPT® 78216) is medically necessary for the following when ultrasound, CT, and MRI are unavailable or contraindicated:
  - Evaluation of liver mass, trauma, or suspected focal nodular hyperplasia (FNH)
  - Differentiation of hepatic hemangioma from FNH
  - Diffuse hepatic disease or elevated liver function tests
  - Suspected accessory spleen (CPT® 78215 or CPT® 78216 only)

Background and Supporting Information

- For liver lesions in children, it is important to differentiate between benign incidental versus malignant lesions.

Risk factors for malignant liver tumors in children

Hepatoblastoma	Hepatocellular carcinoma
◦ Prematurity	◦ Liver cirrhosis
◦ Low birth weight	◦ Chronic hepatitis B and C
◦ BWS and other overgrowth syndromes	◦ Hereditary tyrosinemia
◦ Familial adenomatous polyposis	◦ Portosystemic shunts
◦ Trisomy 18	◦ Bile salt export pump deficiency
◦ Portosystemic shunts	
◦ Aicardi syndrome	

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Liver Disease (PEDAB-16)

Guideline

Pediatric Liver Failure and Cirrhosis (PEDAB-16.1)
References (PEDAB-16.1)
Biliary Disease (PEDAB-16.2)
References (PEDAB-16.2)
Liver Elastography (PEDAB-16.3)
References (PEDAB-16.3)

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Pediatric Liver Failure and Cirrhosis (PEDAB-16.1)

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- Elevated liver function testing imaging indications in pediatric individuals are very similar to those for adult individuals. See: **Abnormal Liver Chemistries (AB-30)** in the Abdomen Imaging Guidelines.
- Causes of liver failure or cirrhosis in pediatric individuals are different from adults, and are frequently idiopathic, but commonly due to ONE of the following:
  - Biliary dysfunction (biliary atresia, cystic fibrosis, etc.)
  - Metabolic disease
  - Post-infectious
- Liver ultrasound (CPT® 76700) with duplex Doppler (CPT® 93975) is medically necessary as an initial study for individuals prior to approving CT or MRI for pediatric individuals.
  - MRI Abdomen without and with contrast (CPT® 74183) is indicated for evaluation of ultrasound findings that are inconclusive or technically limited, and is preferred over CT when possible to reduce radiation exposure.
- Repeat liver ultrasound (CPT® 76705) with duplex Doppler (CPT® 93975 or CPT® 93976) is medically necessary in pediatric individuals in the following circumstances:
  - Annually, for known chronic liver dysfunction or cirrhosis of any cause
  - New or worsening findings on history, physical exam, or laboratory results that suggest progression of liver disease
  - Portal venous congestion or portal hypertension is suspected
- Nuclear medicine liver imaging (ONE of CPT® codes: CPT® 78201, CPT® 78202, CPT® 78803, CPT® 78215, or CPT® 78216) is rarely performed, but is medically necessary for the following when ultrasound, CT, and MRI are unavailable or contraindicated:
  - Diffuse hepatic disease or elevated liver function tests

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Biliary Disease (PEDAB-16.2)

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- The definition of conjugated hyperbilirubinemia is serum conjugated bilirubin >1mg/dL if total bilirubin <5.0 or greater than 20 percent of total bilirubin if total bilirubin >5.0mg/dL. Obstructive causes of liver disease need to be evaluated. Additional labs may include total and fractionated bilirubin, AST, ALT, Alk Phos, GGT, and/or urinalysis.
- Ultrasound Abdomen (CPT® 76700 or CPT® 76705) is medically necessary as the initial imaging study.
  - Advanced imaging such as CT, MRI is rarely medically necessary unless otherwise indicated below.
- After initial ultrasound:
  - If biliary atresia is suspected:
    - Hepatobiliary system imaging (HIDA scan) is medically necessary if requested by surgeon before liver biopsy.
  - If choledochal cyst is suspected:
    - CT Abdomen with contrast (CPT® 74160) or MRI/MRCP (CPT® 74183 or CPT® 74181) is medically necessary.
    - For preoperative assessment: MRI/MRCP (CPT® 74183 or CPT® 74181) is medically necessary.
  - If primary biliary disease such as primary sclerosing cholangitis or primary biliary cholangitis is suspected:
    - CT Abdomen with contrast (CPT® 74160) or MRI/MRCP (CPT® 74183 or CPT® 74181) is medically necessary.
- Follow-up of resected choledochal cysts (types I and IV, and type V not treated by transplantation):
  - Immediately after resection:
    - Ultrasound Abdomen (CPT® 76700 or CPT® 76705) AND MRI Abdomen with cholangiography (CPT® 74183) is medically necessary.
  - Years 1-20 post-resection:
    - Ultrasound Abdomen (CPT® 76700 or CPT® 76705), biannually
      - MRI Abdomen with cholangiography (CPT® 74183) is medically necessary on every 3-year schedule (in lieu of ultrasound) for any of the following:
        - Ultrasound is not feasible due to body habitus
        - Ultrasound is unavailable
  - Years 21-40 post-resection:
    - Ultrasound Abdomen (CPT® 76700 or CPT® 76705), every 3 years

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- MRI Abdomen with cholangiography (CPT® 74183) is medically necessary on every 4-year schedule (in lieu of ultrasound) for any of the following:
  - Ultrasound is not feasible due to body habitus
  - Ultrasound is unavailable

◦ Residual cystic tissue is present following resection:
  ▪ Ultrasound Abdomen (CPT® 76700 or CPT® 76705), annually

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Liver Elastography (PEDAB-16.3)

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- The following is medically necessary for initial staging of liver fibrosis in suspected chronic liver disease:
  - Transient Elastography or Vibration-Controlled Transient Elastography (VCTE, e.g. Fibroscan) (CPT® 91200) is the initial imaging modality.
  - Magnetic Resonance Elastography (MRE, CPT® 76391) is medically necessary for ANY of the following:
    - Transient Elastography failure despite use of an XL-probe, OR BMI ≥30
    - Conflict between clinical picture and transient elastography results

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Adrenal Lesions (PEDAB-17)

Guideline

Adrenal Lesions (PEDAB-17)
References (PEDAB-17)

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Adrenal Lesions (PEDAB-17)

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- Adrenal masses in infants and young children usually present as palpable abdominal masses or are detected on in utero US. In the neonates, the common masses are adrenal hemorrhage and neuroblastoma. Abdominal US is the medically necessary initial imaging study.
  - If an adrenal mass is detected, it can often be adequately evaluated with short interval follow-up retroperitoneal ultrasound (CPT® 76770) in 7 to 10 days.
    - If repeat ultrasound is concerning for neuroblastoma or there is high clinical concern for neuroblastoma, MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen without and with contrast (CPT® 74170) are medically necessary to confirm the diagnosis. MRI is preferred over CT when possible to reduce radiation exposure. If these studies, confirm neuroblastoma ^{123}I-Metaiodobenzylguanidine (MIBG) scintigraphy is medically necessary for staging.
  - Neuroblastoma is the most common primary adrenal tumor in pediatric individuals between day 1 and 5 years of age. See: **Neuroblastoma (PEDONC-6)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Additional adrenal imaging considerations include the following:
  - Adrenal Nuclear Imaging of the cortex and/or medulla (CPT® 78075) is medically necessary for the following:
    - Distinguishing adrenal adenoma from adrenal hyperplasia.
    - Evaluation of suspected pheochromocytoma or paraganglioma.
      - MIBG preferred (ONE of CPT® codes: CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, or CPT® 78804).
      - For known pheochromocytoma or paraganglioma, see: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines.
    - Evaluation of suspected neuroblastoma, ganglioneuroblastoma, or ganglioneuroma.
      - MIBG preferred (ONE of CPT® codes: CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, or CPT® 78804 or hybrid SPECT/CT CPT® 78830, CPT® 78831, or CPT® 78832), see: **Neuroblastoma (PEDONC-6)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
    - History of multiple endocrine neoplasia syndromes: See **Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)** in the Pediatric and Special Populations Oncology Imaging Guidelines

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- History of neurofibromatosis: See **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines
- History of von Hippel-Lindau disease: See **Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)** in the Pediatric and Special Populations Oncology Imaging Guidelines

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Hemochromatosis (PEDAB-18)

Guideline

Hereditary (Primary) Hemochromatosis (PEDAB-18.1)
Transfusion-Associated (Secondary) Hemochromatosis (PEDAB-18.2)
References (PEDAB-18)

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Hereditary (Primary) Hemochromatosis (PEDAB-18.1)

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- Hereditary hemochromatosis imaging indications in pediatric individuals are identical to those for adult individuals. See: **Hereditary (Primary) Hemochromatosis (HH) and Other Iron Storage Diseases (AB-11.2)** in the Abdomen Imaging Guidelines.

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Transfusion-Associated (Secondary) Hemochromatosis (PEDAB-18.2)

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- Transfusion-associated hemochromatosis is a common complication of exposure to repeated red blood cell transfusions. This can occur in any individual with exposure to >20 transfusion episodes, but is most common among sickle cell disease, thalassemia, bone marrow failure (aplastic anemia, Fanconi anemia, etc.), oncology patients, and hematopoietic stem cell transplant patients.
- T2* MRI has been well established in the determination of organ iron burden in transfusion-associated hemochromatosis. Contrast use is not necessary for evaluation of iron burden. The following studies are medically necessary for evaluation of transfusion-associated hemochromatosis:
  - MRI Abdomen without contrast (CPT® 74181) for liver iron evaluation.
  - MRI Cardiac without contrast (CPT® 75557) for cardiac iron evaluation.
  - MRI Chest without contrast (CPT® 71550) can be approved as a single study to evaluate both heart and liver iron burden.
  - CPT® 74181 and CPT® 75557 can be approved alone, or together.
  - If requested, CPT® 71550 will evaluate both heart and liver and should not be approved with any other codes.
- Screening MRI is medically necessary every 12 months for chronically transfused individuals at risk of hemochromatosis.
- Imaging is medically necessary every 3 months for treatment response in individuals receiving active treatment (chelation and/or phlebotomy).

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Indeterminate Renal Lesion (PEDAB-19)

Guideline

Indeterminate Renal Lesion (PEDAB-19)
References (PEDAB-19)

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Indeterminate Renal Lesion (PEDAB-19)

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- Indeterminate renal lesion characterization imaging indications in pediatric individuals are very similar to those for adult individuals. See: **Indeterminate Renal Lesion (AB-35)** in the Abdomen Imaging Guidelines.
- Indeterminate renal lesion imaging indications in pediatric individuals are uncommon and are usually cysts or congenital anomalies.
- Pediatric-specific imaging considerations include the following:
  - Pediatric renal cysts have a lower risk of malignant progression than do renal cysts in adults.
  - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen with contrast (CPT® 74160) is medically necessary for individuals who have simple cysts but are symptomatic and surgical intervention is being considered.
  - MRI Abdomen without and with contrast (CPT® 74183) or CT Abdomen without and with contrast (CPT® 74170) is medically necessary for pediatric individuals with complex renal cyst identified on ultrasound.
  - For individuals with congenital anomalies, nuclear medicine studies with diuretic renography (CPT® 78708 or CPT® 78709) are medically necessary to determine function and cystography to determine presence of associated reflux.
  - Individuals with solid renal masses, see: **Pediatric Renal Tumors (PEDONC-7)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
  - Individuals with renal tumor predisposition syndromes, see: **Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Hydronephrosis (PEDAB-20)

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Hydronephrosis (PEDAB-20)
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Hydronephrosis (PEDAB-20)

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Hydronephrosis is a relatively common finding in pediatric individuals, with the following imaging considerations:

- Evaluation of antenatal hydronephrosis (AHN) with retroperitoneal ultrasound (CPT® 76771)
  - Postnatal ultrasound within 24 hours of life is medically necessary for males with distended bladder or infants with moderate/severe antenatal hydronephrosis diagnosed prenatally.
  - Postnatal ultrasound after seven days of life is medically necessary for infants with mild prenatal hydronephrosis.
  - A second postnatal ultrasound, even if initial is normal, at 4-6 weeks of age is medically necessary for all infants with antenatal hydronephrosis.
  - After this, infants with known hydronephrosis may be followed by ultrasound every 3-12 months until resolution of hydronephrosis.
    - This imaging represents a guideline-supported, scheduled follow-up imaging evaluation, as described in **Clinical Information (Preface-3.1)** in the Preface Imaging Guidelines.
- Hydronephrosis associated with urinary tract infection or vesicoureteral reflux, see: **Urinary Tract Infection (UTI) (PEDAB-5)** for imaging guidelines.
- In individuals with obstructive uropathy (including ureteropelvic junction obstruction (UPJO), ureterovesical junction obstruction (UVJO), and bladder outlet obstruction), retroperitoneal ultrasound (CPT® 76770) and diuretic renography (CPT® 78707, CPT® 78708, or CPT® 78709) are medically necessary for preoperative planning and postoperatively at 3 to 12 months.
  - If hydronephrosis has resolved on postoperative imaging then no further routine imaging is indicated.
- Magnetic resonance urography (MRU) (CPT® 74183 and CPT® 72197) is medically necessary in individuals with inconclusive ultrasound and diuretic renography.
- CT Abdomen with contrast (CPT® 74160) is medically necessary in individuals with inconclusive ultrasound and a suspected vascular cause of UPJO.

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Polycystic Kidney Disease (PEDAB-21)

Guideline

Polycystic Kidney Disease (PEDAB-21)
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Polycystic Kidney Disease (PEDAB-21)

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- Abdominal ultrasound (CPT® 76700) or retroperitoneal ultrasound (CPT® 76770) is medically necessary for clinical concern of polycystic kidney disease, or for screening individuals who are at risk for autosomal dominant polycystic kidney disease (ADPKD).
- There are no established MRI-based diagnostic criteria for ADPKD in children younger than 15 years of age.
- For older children and teens, imaging indications for MRI Abdomen without contrast (CPT® 74181) would be identical to those for adult individuals. See: **Polycystic Kidney Disease (AB-38.1)** in the Abdomen Imaging Guidelines.

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Blunt Abdominal Trauma (PEDAB-22)

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Blunt Abdominal Trauma (PEDAB-22)

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- Blunt abdominal trauma imaging indications in pediatric individuals are identical to those for adult individuals. See: **Blunt Abdominal Trauma (AB-10.1)** in the Abdomen Imaging Guidelines.

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Hernias (PEDAB-23)

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Hernias (PEDAB-23)

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- Hernia imaging indications in pediatric individuals are identical to those for adult individuals. See: **Hernias (AB-12)** in the Abdomen Imaging Guidelines.

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Abdominal Lymphadenopathy (PEDAB-24)

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Abdominal Lymphadenopathy (PEDAB-24)

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Abdominal Lymphadenopathy (PEDAB-24)

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- Abdominal lymphadenopathy imaging indications in pediatric individuals are identical to those for adult individuals. See: **Abdominal Lymphadenopathy (AB-8)** in the Abdomen Imaging Guidelines.

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Left Upper Quadrant Pain (PEDAB-25)

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Left Upper Quadrant Pain (PEDAB-25)
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Left Upper Quadrant Pain (PEDAB-25)

ABP.LT.0025.A

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- Left upper quadrant pain imaging indications in pediatric individuals are identical to those for adult individuals. See: **Abdominal Pain (AB-2)** in the Abdomen Imaging Guidelines.
- Nuclear medicine spleen imaging (CPT® 78185) is medically necessary for left upper quadrant pain when neither ultrasound nor CT is available.

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Spleen (PEDAB-26)

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Spleen (PEDAB-26)
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Spleen (PEDAB-26)

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- Spleen imaging indications in pediatric individuals are very similar to those for adult individuals. See: **Spleen (AB-34)** in the Abdomen Imaging Guidelines.
- Nuclear medicine spleen imaging (CPT® 78185) is medically necessary for the following indications when CT is unavailable:
  - Splenic trauma
  - Evaluation of splenic function
  - Suspected splenic mass, cyst, abscess, infarct, or metastasis
  - Radiation treatment planning
  - Asplenia
  - Suspected functional accessory spleen:
    - CPT® 78215 or CPT® 78216 instead of CPT® 78185 is medically necessary, if requested
- Pediatric-specific imaging considerations include the following:
  - MRI is preferred over CT when possible to reduce radiation exposure.

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Intussusception (PEDAB-27)

Guideline

Intussusception (PEDAB-27)
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Intussusception (PEDAB-27)

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- Intussusception, telescoping of one bowel loop into another, is a frequent cause of abdominal pain in young children. It may be associated with bloody stool.
  - Ultrasound (CPT® 76700 or CPT® 76705) is medically necessary as an initial study if there is a strong suspicion for intussusception.
  - CT Abdomen and/or Pelvis with contrast (CPT® 74160 or CPT® 74177) is medically necessary to help characterize or diagnose pathologic lead points identified on ultrasound.

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Bowel Obstruction (PEDAB-28)

Guideline

Bowel Obstruction (PEDAB-28.1)
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Bowel Obstruction (PEDAB-28.1)

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- Suspected high-grade obstruction
  - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary (preferred to avoid unnecessary radiation exposure).
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if MRI is not readily available.
- Suspected intermittent or low-grade small bowel obstruction.
  - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary (preferred to avoid unnecessary radiation exposure).
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary if MRI is not readily available.
  - If the etiology or level of suspected intermittent or low-grade small bowel obstruction remains undetermined and additional imaging is needed after CT Abdomen and Pelvis, the following are medically necessary:
    - CT Enteroclysis (CPT® 74176 or 74177), or
    - CT Enterography (CPT® 74177), or
    - MR Enteroclysis (CPT® 74183 and CPT® 72197), or
    - MR Enterography (CPT® 74183 and CPT® 72197)
- Small bowel obstruction suspected to be secondary to Crohn's Disease
  - See: **Inflammatory Bowel Disease, Crohn Disease, or Ulcerative Colitis (PEDAB-9)**

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Left Lower Quadrant Pain (PEDAB-29)

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Left Lower Quadrant Pain (PEDAB-29)
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Left Lower Quadrant Pain (PEDAB-29)

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Diverticulitis is the most common cause of left lower quadrant pain in adults but is extremely rare in children.

- CT Abdomen and Pelvis (CPT® 74177) with contrast is medically necessary if there are any red flag signs or symptoms (as listed in **General Guidelines (PEDAB-1.0)**)
- In the absence of red flags, advanced imaging is rarely helpful in the initial evaluation of these individuals. Consultation with a gastroenterologist can be helpful in determining the appropriate diagnostic pathway.
  - Pelvic ultrasound (CPT® 76856) is the medically necessary initial imaging study of choice for children for detecting gynecologic abnormalities that may cause left lower quadrant pain.
  - For male individuals or if ultrasound is inconclusive, advanced imaging may be appropriate for management as directed by gastroenterological evaluation or consultation.

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Celiac Disease (Sprue) (PEDAB-30)

Guideline

Celiac Disease (Sprue) (PEDAB-30)
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Celiac Disease (Sprue) (PEDAB-30)

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- Celiac disease imaging indications in pediatric individuals are identical to those for adult individuals. See: **Celiac Disease (Sprue) (AB-24)** in the Abdomen Imaging Guidelines.

***Background and Supporting Information*:**

There is no need for radiological small bowel imaging in uncomplicated celiac disease.

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References (PEDAB-30)

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Transplant (PEDAB-31)

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Transplant (PEDAB-31)

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- Liver and kidney transplant imaging indications in pediatric individuals are identical to those for adult individuals. See: **Transplant (AB-42)** in the Abdomen Imaging Guidelines.
- For post-transplant lymphoproliferative disorder in pediatric individuals, see: **Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Gaucher Disease (PEDAB-32)

Guideline

Gaucher Disease (PEDAB-32)

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Gaucher Disease (PEDAB-32)

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See: **Gaucher Disease (PEDPN-4)** in the Pediatric Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines.

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Vomiting Infant, Malrotation, and Hypertrophic Pyloric Stenosis (PEDAB-33)

Guideline

Vomiting Infant, Malrotation, and Hypertrophic Pyloric Stenosis (PEDAB-33)
References (PEDAB-33)

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Vomiting Infant, Malrotation, and Hypertrophic Pyloric Stenosis (PEDAB-33)

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- Vomiting in infants is generally classified as either bilious (implying obstruction distal to the Sphincter of Oddi) or non-bilious.
- Bilious vomiting may be a true emergency, as some of the conditions causing this could result in compromise of blood supply to the intestines, a potentially life-threatening situation.
- Suspected malrotation is an indication for emergent imaging. If malrotation with mid-gut volvulus is suspected, Ultrasound abdomen, limited (CPT® 76705) is medically necessary.
- Hypertrophic Pyloric Stenosis is an idiopathic condition wherein the circular muscle controlling emptying of the stomach thickens causing a relative obstruction of the gastric outlet. The condition can occur at any age (including occasionally in adults), but the typical child is male, aged 2 to 6 weeks. Projectile non-bilious vomiting is the most common presenting complaint, but the description of projectile vomiting is subjective. The differential diagnosis for non-bilious vomiting includes common conditions such as viral gastroenteritis and gastro-esophageal reflux.
  - US Abdomen, limited (CPT® 76705) is medically necessary for infants with persistent projectile non-bilious vomiting. If initial studies are not diagnostic, repeat studies are medically necessary (as frequently as daily) until the vomiting resolves or the diagnosis is made.

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Pancreatitis (PEDAB-34)

Guideline

Acute Pancreatitis (PEDAB-34.1)
Chronic Pancreatitis (PEDAB-34.2)
References (PEDAB-34)

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Acute Pancreatitis (PEDAB-34.1)

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- The presence of any red flag findings per **General Guidelines (PEDAB-1.0)** precludes adjudication based on any other criteria.
  - If red flag is present (as per **General Guidelines (PEDAB-1.0)**), then CT Abdomen and Pelvis with contrast (CPT® 74177), CT Abdomen with contrast (CPT® 74160), or MRI/MRCP (CPT® 74183 or CPT® 74181) is medically necessary.
- Initial imaging
  - US Abdomen (CPT® 76700 or CPT® 76705) is medically necessary
  - If ultrasound performed and is nondiagnostic due to technical limitation (obesity, overlying gas, etc.), or if ultrasound is negative and there is continued clinical suspicion of acute pancreatitis, MRI/MRCP (CPT® 74183 or CPT® 74181) is medically necessary.
    - CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen with contrast (CPT® 74160) is medically necessary if MRI/MRCP cannot be performed.
  - CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for management of acute pancreatitis in the following situations:
    - Evaluation of known or suspected complications of acute pancreatitis
    - To characterize degree of organization of collections before intervention
- Abdominal US (CPT® 76700 or CPT® 76705) is medically necessary to follow known fluid collections for resolution or progression.
- Acute Recurrent Pancreatitis (ARP)
  - MRI/MRCP (CPT® 74183 or CPT® 74181) is medically necessary for the following:
    - To identify structural or obstructive causes
    - To assess for progression to chronic pancreatitis
  - In a child who requires sedation for imaging, it is reasonable to alternate MRI/MRCP with Abdominal US (CPT® 76700 or CPT® 76705) or CT Abdomen with contrast (CPT® 74160) for serial monitoring of acute recurrent pancreatitis as recommended by or in consultation with a gastroenterologist or pancreatic specialist.

Background and Supporting Information

- The role of imaging is to identify findings at diagnosis, assess for local complications, identify potential etiologies, monitor evolution of local complications, plan and guide interventions.

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Chronic Pancreatitis (PEDAB-34.2)

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- The role of imaging is to contribute to or establish initial diagnosis, stage/monitor disease, assess for superimposed acute pancreatitis, identify potential etiologies of chronic pancreatitis, characterize secretory function, and/or plan for surgical intervention.
- If chronic pancreatitis is suspected, the following are medically necessary:
  - MRI Abdomen without and with contrast (CPT® 74183)
    - CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen with contrast (CPT® 74160) if MRI cannot be performed
- To evaluate suspected or known episode of acute pancreatitis in a child with chronic pancreatitis, the following is medically necessary:
  - Abdominal US (CPT® 76700 or CPT® 76705)
  - CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen with contrast (CPT® 74160) or MRI/MRCP (CPT® 74183 or CPT® 74181) are medically necessary in the following situations:
    - If ultrasound is negative and imaging diagnosis of acute pancreatitis is needed
    - If planning endoscopic or surgical interventions

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References (PEDAB-34)

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General Guidelines (PEDCD-1.0)

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Heart disease in the pediatric population involves predominantly congenital lesions. Pediatric individuals can have acquired heart disease unique to children. For those diseases which occur in both pediatric and adult populations, differences exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.

Pediatric Cardiac Imaging Appropriate Clinical Evaluation

- Prior to considering advanced imaging (CT, MR, Nuclear Medicine) or echocardiogram, a pertinent clinical evaluation should be performed, including the following (both):
  - A detailed history, physical examination, or meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging)
  - A review of appropriate diagnostic studies (laboratory, EKG, echocardiogram, and other diagnostic imaging)
- A recent clinical evaluation is not needed prior to advanced imaging (CT, MR, Nuclear Medicine) or echocardiogram if **any** of the following apply:
  - Individual is undergoing guideline-supported scheduled imaging evaluation.
  - Echocardiogram is being performed at the first cardiology visit for an appropriate indication as stated in these guidelines.
  - Routine imaging is anticipated at the next visit (e.g., one year follow-up echocardiogram for a 10 year old with a VSD).
- Advanced imaging of the heart and echocardiogram are medically necessary in any of the following:
  - Individuals who have documented active clinical signs or symptoms of disease involving the heart
  - As follow-up for findings on echocardiograms
  - See **Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)** for indications for initial echocardiogram in asymptomatic individuals
- Repeat imaging studies of the heart are **not** medically necessary unless one of the following applies:
  - Repeat imaging is considered medically necessary in a specific guideline section.
  - There is evidence for progression of disease.
  - There is new onset of disease with documentation of how repeat imaging will affect individual management or treatment decisions.

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  - See **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** for indications for repeat echocardiogram in asymptomatic individuals
- Asymptomatic individuals with exposure to cardiotoxic drugs can have serial echocardiograms as per **Cardiotoxicity and Echocardiography (PEDONC-19.2)** in the Pediatric Oncology Imaging Guideline.
- Advanced imaging and echocardiogram is **not** medically necessary, in the absence of other appropriate indications listed in these guidelines, for **any** of the following:
  - Individuals starting ADHD medications
  - To screen asymptomatic individuals for disorders involving the heart

Pediatric Cardiac Imaging Modality General Considerations

- MRI
  - MRI and MRA studies are frequently medically necessary for evaluation of congenital heart defects not well visualized on echocardiography, thoracic arteries and veins not visualized on echocardiography, cardiomyopathies, and right ventricular disease, as well as in follow-up for these indications.
  - Due to the length of time for image acquisition and the need for the individual to be motionless during the acquisition, anesthesia is required for almost all infants and young children (age <7 years), as well as older children with delays in development or maturity. In this population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:
    - MRI is typically performed without and with contrast.
    - If multiple body areas are supported for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
- CT
  - CT is primarily used to evaluate the coronary and great vessels in congenital heart disease if cardiac MR is contraindicated.
  - Coding considerations are listed in **CT Heart and Coronary Computed Tomography Angiography (CCTA) General Considerations (PEDCD-10.1)**
- Ultrasound
  - Echocardiography is the primary modality used to evaluate the anatomy and function of the pediatric heart and is generally indicated before considering other imaging modalities.
  - Coding considerations are listed in **Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)**
- Nuclear Medicine

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- SPECT, PET stress may be medically necessary for individuals with anomalous CA, angina chest pain, and follow-up for Kawasaki and MIS-C. See specific sections for those indications.
- Multi Gated Acquisition (MUGA) studies (CPT® 78472, CPT® 78473, CPT® 78481, CPT® 78483, CPT® 78494, or CPT® 78496) are rarely performed in pediatrics but can be approved for the following:
  - Certain pediatric oncology individuals when echocardiography is insufficient: See: **Appropriate Clinical Evaluations (PEDONC-1.2)** for imaging guidelines.
  - Quantitation of left ventricular function when recent echocardiogram shows ejection fraction of <50% and MUGA results will impact acute patient care decisions.
- SPECT/CT fusion imaging involves SPECT (MPI) imaging and CT for optimizing location, accuracy, and attenuation correction combines functional and anatomic information.
  - There is currently no evidence-based data to formulate appropriateness criteria for SPECT/CT fusion imaging.
  - Combined use of nuclear imaging, including SPECT, along with diagnostic CT (fused SPECT/CT) is considered investigational.
- Central C-V Hemodynamics (CPT® 78414) is not an imaging study and is an outdated examination.
- Cardiac Shunt Detection (CPT® 78428) is rarely performed in pediatrics but is medically necessary for individuals in whom Cardiac MR is not diagnostic.
  - Calculation of left and right ventricular ejection fractions
  - Assessment of wall motion
  - Quantitation of right to left shunts
- Myocardial Tc-99m Pyrophosphate Imaging
  - Infarct Avid Myocardial Imaging studies (CPT® 78466, CPT® 78468, and CPT® 78469), or Myocardial Tc-99m Pyrophosphate Imaging, although still available, has variable sensitivity and specificity for identifying infarcted myocardial tissue, and the current use for this indication is limited
  - CPT® 78466, CPT® 78468, and CPT® 78469, CPT® 78800 or CPT® 78803 may be used for identification of myocardial ATTR (transthyretin) amyloidosis. See **Myocardial Tc-99m Pyrophosphate Imaging in Cardiac Amyloidosis (CD-3.8)**

Pediatric Age

- Age definitions for pediatric individuals (for purpose of these guidelines)
  - Infant (birth through 12 months)
    - Neonate or newborn (birth through 28 days)
  - Child 1-18 years

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- Pre-adolescent child 1-10 years
- Adolescent 11-18 years

◦ Children or Pediatric refers to all individuals ages birth-18 years

MUGA (Multi Gated Acquisition) – Blood Pool Imaging	CPT®
Myocardial Imaging, infarct avid, planar, qualitative or quantitative	78466
Myocardial Imaging, infarct avid, planar, qualitative or quantitative with ejection fraction by first pass technique	78468
Myocardial Imaging, infarct avid, planar, qualitative or quantitative tomographic SPECT with or without quantification	78469
Radiopharmaceutical Localization Imaging Limited area	78800
Radiopharmaceutical Localization Imaging SPECT Note: When reporting CPT® 78803, planar imaging of a limited area or multiple areas should be included with the SPECT	78803

Supporting Information

Individuals who are <18 years old should be imaged according to the Pediatric Cardiac Imaging Guidelines and the general Cardiac Imaging Guidelines. Individuals who are age ≥18 years should be imaged according to the Cardiac Imaging Guidelines, except where directed otherwise by a specific guideline section. Adult individuals who also have congenital heart disease should be imaged by **Congenital heart disease – General Information (CD-11.1)** in the general Cardiac Imaging Guidelines.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount, and variance from these guidelines may be appropriate and warranted for specific clinical situations.

Procedure Codes Associated with Cardiac or PVD Imaging

MRI/MRA	CPT®
Cardiac magnetic resonance imaging for morphology and function without contrast material	75557

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MRI/MRA	CPT®
Cardiac magnetic resonance imaging for morphology and function without contrast material; with stress imaging	75559
Cardiac magnetic resonance imaging for morphology and function without contrast material(s), followed by contrast material(s) and further sequences	75561
Cardiac magnetic resonance imaging for morphology and function without contrast material(s), followed by contrast material(s) and further sequences; with stress imaging	75563
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure)	75565

CT	CPT®
Computed tomography, heart, without contrast material, with quantitative evaluation of coronary calcium	75571
Computed tomography, heart, with contrast material, for evaluation of cardiac structure and morphology (including 3D image postprocessing, assessment of cardiac function, and evaluation of venous structures, if performed)	75572
Computed tomography, heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease (including 3D image postprocessing, assessment of LV cardiac function, RV structure and function and evaluation of venous structures, if performed)	75573

CTA	CPT®
Noninvasive estimate of coronary fractional flow reserve (FFR) derived from augmentative software analysis of the data set from a coronary computed tomography angiography, with interpretation and report by a physician or other qualified health care professional	75580

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CTA	CPT®
Computed tomographic angiography, heart, coronary arteries and bypass grafts (when present), with contrast material, including 3D image postprocessing (including evaluation of cardiac structure and morphology, assessment of cardiac function, and evaluation of venous structures, if performed)	75574
Computed tomographic angiography, abdominal aorta and bilateral iliofemoral lower extremity runoff, with contrast material(s), including noncontrast images, if performed, and image postprocessing	75635

Nuclear Medicine	CPT®
Determination of central c-v hemodynamics (non-imaging) (e.g., ejection fraction with probe technique) with or without pharmacologic intervention or exercise, single or multiple determinations	78414
Cardiac shunt detection	78428
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), single study; with concurrently acquired computed tomography transmission scan	78429
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); single study, at rest or stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78430
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); multiple studies at rest and stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78431

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Nuclear Medicine	CPT®
Myocardial imaging, positron emission tomography (PET), combined perfusion with metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), dual radiotracer (e.g., myocardial viability);	78432
Myocardial imaging, positron emission tomography (PET), combined perfusion with metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), dual radiotracer (e.g., myocardial viability); with concurrently acquired computed tomography transmission scan	78433
Absolute quantitation of myocardial blood flow (AQMBF), positron emission tomography (PET), rest and pharmacologic stress (List separately in addition to code for primary procedure)	78434
Myocardial perfusion imaging, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78451
Myocardial perfusion imaging, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78452
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78453
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78454

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Nuclear Medicine	CPT®
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion and/or ejection fraction, when performed), single study	78459
Myocardial imaging, infarct avid, planar; qualitative or quantitative	78466
Myocardial imaging, infarct avid, planar; with ejection fraction by first pass technique	78468
Myocardial imaging, infarct avid, planar; tomographic SPECT with or without quantification	78469
Cardiac blood pool imaging, gated equilibrium; planar, single study at rest or stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without additional quantitative processing	78472
Cardiac blood pool imaging, gated equilibrium; multiple studies, wall motion study plus ejection fraction, at rest and stress (exercise and/or pharmacologic), with or without additional quantification	78473
Cardiac blood pool imaging (planar), first pass technique; single study, at rest or with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78481
Cardiac blood pool imaging (planar), first pass technique; multiple studies, at rest and with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78483
Myocardial imaging, positron emission tomography (PET), perfusion (including ventricular wall motion and/or ejection fraction, when performed); single study at rest or stress (exercise or pharmacologic)	78491
Myocardial imaging, positron emission tomography (PET), perfusion (including ventricular wall motion and/or ejection fraction, when performed); multiple studies at rest and/or stress (exercise or pharmacologic)	78492

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Nuclear Medicine	CPT®
Cardiac blood pool imaging, gated equilibrium, SPECT, at rest, wall motion study plus ejection fraction, with or without quantitative processing	78494
Cardiac blood pool imaging, gated equilibrium, single study, at rest, with right ventricular ejection fraction by first pass technique (List separately in addition to code for primary procedure)	78496
Quantitative differential pulmonary perfusion, including imaging when performed	78597
Quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed	78598
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, single area (e.g., head, neck, chest, pelvis), single day imaging	78800
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, 2 or more areas (e.g., abdomen and pelvis, head and chest), 1 or more days imaging or single area imaging over 2 or more days	78801
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, single day imaging	78802
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Nuclear Medicine	CPT®
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (e.g., head, neck, chest, pelvis), single day imaging	78830
Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment	0331T
Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment; with tomographic SPECT	0332T

Ultrasound	CPT®
Transthoracic echocardiography for congenital cardiac anomalies; complete	93303
Transthoracic echocardiography for congenital cardiac anomalies; follow-up or limited study	93304
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, complete, with spectral Doppler echocardiography, and with color flow Doppler echocardiography	93306
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, complete, without spectral or color Doppler echocardiography	93307
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, follow-up or limited study	93308
Echocardiography, transesophageal, real-time with image documentation (2D) (with or without M-mode recording); including probe placement, image acquisition, interpretation and report	93312

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Ultrasound	CPT®
Echocardiography, transesophageal, real-time with image documentation (2D) (with or without M-mode recording); placement of transesophageal probe only	93313
Echocardiography, transesophageal, real-time with image documentation (2D) (with or without M-mode recording); image acquisition, interpretation and report only	93314
Transesophageal echocardiography for congenital cardiac anomalies; including probe placement, image acquisition, interpretation and report	93315
Transesophageal echocardiography (TEE) for congenital cardiac anomalies; placement of transesophageal probe only	93316
Transesophageal echocardiography for congenital cardiac anomalies; placement of transesophageal probe only	93317
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Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); complete	+93320
Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); follow-up or limited study (List separately in addition to codes for echocardiographic imaging)	+93321
Doppler echocardiography color flow velocity mapping (List separately in addition to codes for echocardiography)	+93325

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Ultrasound	CPT®
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report;	93350
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report; including performance of continuous electrocardiographic monitoring, with supervision by a physician or other qualified health care professional	93351
Use of echocardiographic contrast agent during stress echocardiography (List separately in addition to code for primary procedure)	+93352
Myocardial strain imaging using speckle tracking-derived assessment of myocardial mechanics (List separately in addition to codes for echocardiography imaging)	+93356
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, for congenital cardiac anomalies; complete	C8921
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, for congenital cardiac anomalies; follow-up or limited study	C8922
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, complete, without spectral or color doppler echocardiography	C8923
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording when performed, follow-up or limited study	C8924

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Ultrasound	CPT®
Transesophageal echocardiography (TEE) with contrast, or without contrast followed by with contrast, real time with image documentation (2D) (with or without M-mode recording); including probe placement, image acquisition, interpretation and report	C8925
Transesophageal echocardiography (TEE) with contrast, or without contrast followed by with contrast, for congenital cardiac anomalies; including probe placement, image acquisition, interpretation and report	C8926
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report	C8928
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, complete, with spectral doppler echocardiography, and with color flow doppler echocardiography	C8929
Transthoracic echocardiography, with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report; including performance of continuous electrocardiographic monitoring, with physician supervision	C8930
Myocardial contrast perfusion echocardiography, at rest or with stress, for assessment of myocardial ischemia or viability (List separately in addition to code for primary procedure)	+0439T

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social

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conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Congenital Heart Disease

Guideline

Congenital Heart Disease General Information (PEDCD-2.1)
Congenital Heart Disease Coding (PEDCD-2.2)
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AVSD (Atrioventricular Canal, Endocardial Cushion Defect) (PEDCD-2.4.3)
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Pulmonary Stenosis (PS) (PEDCD-2.4.7)
Pulmonary Atresia with Intact Septum (PAIVS) (PEDCD-2.4.8)
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Aortic Coarctation and IAA (Interrupted Aortic Arch) (PEDCD-2.4.11)
Coronary Anomalies (PEDCD-2.4.12)
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Double Outlet Right Ventricle (DORV) (PEDCD-2.4.14)
D-Loop Transposition of the Great Arteries (D-Loop TGA) (PEDCD-2.4.15)
Congenitally Corrected Transposition of the Great Arteries (ccTGA, LTGA) (PEDCD-2.4.16)
Truncus Arteriosus (PEDCD-2.4.17)
Single Ventricle (SV) (PEDCD-2.4.18)
Eisenmenger and PHT (with CHD)- (PEDCD-2.4.19)
References (PEDCD-2)

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Congenital Heart Disease General Information (PEDCD-2.1)

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- Congenital heart disease accounts for the majority of cardiac problems occurring in the pediatric population. Individuals may be diagnosed any time spanning prenatal evaluation to adolescence. For diagnostic and surveillance imaging of individuals over 18 years of age, see **Congenital heart disease – General Information (CD-11.1)** in the general Cardiac Imaging Guidelines.
- There are a number of variables that influence the modality and timing of imaging individuals with congenital heart disease, which results in a high degree of individuality in determining the schedule for imaging these individuals, including:
  - Gestational age
  - Individual's age
  - Physiologic effects of the defect
  - Status of interventions (catheterization and surgical)
  - Rate of individual's growth
  - Stability of the defect on serial imaging
  - Comorbid conditions
  - Activity level
- Age definitions for pediatric individuals (for purpose of these guidelines)
  - Infant (birth through 12 months)
    - Neonate or newborn (birth through 28 days)
  - Child 1-18 years
    - Pre-adolescent child 1-10 years
    - Adolescent 11-18 years
  - Children or Pediatric refers to all individuals ages birth-18 years
- Newborns (neonates) have special considerations as they have potentially rapidly changing physiology.
  - Newborns with any concerns for ductal dependent lesion can have echocardiograms at any frequency.
  - Newborns have changes in pulmonary vascular resistance that can affect clinical status rapidly and may require more frequent imaging.
  - Neonatal physiology can extend to the first couple of months of life.
  - Newborns can have one repeat echo, if prior echocardiogram is abnormal or equivocal (either in the hospital or as newborn outpatient).

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- Individuals can have an echocardiogram at that time for **Change in clinical status and/or new concerning signs or symptoms**. This can include:
  - Shortness of breath
  - Fatigue
  - Chest discomfort
  - Percentile weight loss
  - Weight gain
  - Poor feeding
  - Tachypnea
  - Tachycardia
  - CHF signs on exam
  - Change in EKG, Pulse ox, or laboratory values
- An additional study can be approved prior to the next routine interval to assess for more rapid change if the change in clinical status involves the echocardiogram itself, such as:
  - Increasing stenosis gradient
  - Increasing regurgitation amount
  - Increasing pulmonary vascular resistance
  - Decreased ventricular function
  - Change in ductal status
- In individuals that can have both Cardiac MR or Cardiac CT and/or MRA Chest or CTA Chest, this is abbreviated as CMR/CT-MRA/CCTA.
- Individuals with medication adjustments may require additional imaging at that time.
  - Pediatric dosing tends to be mg/kg or mcg/kg. Adjusting the dose to the same mg/kg would not be considering a dosing change for imaging.
  - More than one echocardiogram is considered medically necessary in infants due to rapid growth in this age group, changing physiology, pulmonary vascular resistance, ductal size and weight changes, because dose adjustments are done by weight, and dose response and may require more than one echocardiogram during a medication adjustment.
- Heart surgery
  - TTE is considered medically necessary one month prior to heart surgery. Depending on lesion, MR/CT Cardiac and/or Chest may also be included.
  - Can have an echocardiogram within one month post-operative
  - Cardiac MRI/CCT if prior echocardiogram is equivocal
- MRA/CTA Chest can be performed if prior echocardiogram is equivocal and there are issues regarding aortic arch or pulmonary arteries or veins.
- In individuals who have a documented equivocal echocardiogram due to a technical factor (i.e., poor acoustic windows due to body habitus) which will likely be present

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on subsequent echocardiograms, a Cardiac MR/CT, or MRA/CTA Chest may be done with the frequency of echoes, if done instead of an echo.

- MRA/CTA Chest if thoracic issue not seen on echo
- For routine non-invasive imaging for a specific lesion see **Imaging and Surveillance per Congenital Lesion (PEDCD-2.4)**
- For catheterizations see section **Cardiac Catheterization - Indications (PEDCD-11.2)**
- **Individuals with Pulmonary hypertension with CHD** should be reviewed for both their lesion and for PHT in section **Pediatric Pulmonary Hypertension General (PEDCD-7.1)**

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Congenital Heart Disease Coding (PEDCD-2.2)

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Congenital Heart Disease Echocardiography Coding (PEDCD-2.2.1)

- **Any** of the following echocardiography code combinations are appropriate for re-evaluation of individuals with known congenital heart disease:
  - CPT® 93303, CPT® 93320, and CPT® 93325
  - CPT® 93304, CPT® 93321, and CPT® 93325
  - CPT® 93303
  - CPT® 93304
- CPT® 93306 is not medically necessary in the evaluation of known congenital heart disease.

Congenital Heart Disease imaging per modality (PEDCD-2.2.2)

Echocardiogram

- Transthoracic echocardiogram (TTE)
  - TTE for congenital cardiac anomalies; complete (CPT® 93303)
  - TTE for congenital cardiac anomalies; limited study (CPT® 93304)
  - TTE (2D) m-mode recording, complete, with spectral and color flow doppler echocardiography (CPT® 93306)
  - TTE (2D) with or without m-mode recording; complete (CPT® 93307)
  - TTE (2D) with or without m-mode recording; limited study (CPT® 93308)
- Transesophageal echocardiogram (TEE)
  - TEE (2D) including probe placement, imaging, interpretation, and report (CPT® 93312)
  - TEE for congenital cardiac anomalies; including probe placement, imaging, interpretation, and report (CPT® 93315)

MRI

- Cardiac (CMR)
  - Cardiac MRI for morphology and function without contrast (CPT® 75557)
  - Cardiac MRI for morphology and function without and with contrast (CPT® 75561)
  - Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure) (CPT® 75565)
- MRI Chest

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- MRI Chest without contrast (CPT® 71550)
- MRI Chest with contrast (CPT® 71551)
- MRI Chest with & without contrast (CPT® 71552)

- MRI Angiography (MRA)
  - MRA Chest (excluding myocardium) with or without contrast (CPT® 71555)

CT

- Cardiac (CCT)
  - CT, Heart, with contrast material, for evaluation of cardiac structure and morphology (CPT® 75572)
  - CT, Heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease (CPT® 75573)
- CT Angiography-Cardiac (CCTA)
  - CTA Heart, coronary arteries and bypass grafts (when present), with contrast, including 3D image post processing (CPT® 75574)
- CT-Chest
  - CT Thorax without contrast (CPT® 71250)
  - CT Thorax with contrast (CPT® 71260)
  - CT Thorax without & with contrast (CPT® 71270)
- CT Angiography-Chest (CTA Chest)
  - CTA Chest without and with contrast (CPT® 71275)

Stress Imaging (echo, MRI, MPI)

- Stress echo
  - Echocardiography (TTE), (2D), with or without m-mode, during rest and cardiovascular stress, with interpretation and report (CPT® 93350)
  - Echocardiography (TTE), (2D), m-mode, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation (CPT® 93351)
- Stress MRI
  - Cardiac MRI for morphology and function without contrast, with stress imaging (CPT® 75559)
  - Cardiac MRI for morphology and function without and with contrast, with stress imaging (CPT® 75563)
- Myocardial perfusion imaging (MPI)
  - MPI, tomographic (SPECT) including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic) (CPT® 78451)

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- MPI, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection (CPT® 78452)

Pulmonary perfusion imaging

- Pulmonary perfusion imaging (e.g., particulate) (CPT® 78580)
- Pulmonary ventilation (e.g., aerosol or gas) and perfusion imaging (CPT® 78582)
- Quantitative differential pulmonary perfusion, including imaging when performed (CPT® 78597)
- Quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed (CPT® 78598)

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Congenital Heart Disease Modality Considerations (PEDCD-2.3)

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- Echocardiography is the primary imaging modality used for diagnosing and monitoring congenital heart disease and is generally required before other imaging modalities are considered medically necessary unless otherwise indicated in a specific guideline section.
- Cardiac MRI either without contrast (CPT® 75557) or without and with contrast (CPT® 75561) is considered medically necessary when a recent echocardiogram is inconclusive, needs confirmation of findings, or does not completely define the disease (for subsequent follow-up studies, a recent echocardiogram is not a requirement).
  - CPT® 75565 is also medically necessary for individuals with valvular disease or a need to evaluate intracardiac blood flow. These individuals will usually have CPT® 93320 and CPT® 93325 performed with their echocardiography studies.
  - MRA Chest (CPT® 71555) may be added if the aorta or pulmonary artery needs to be visualized beyond the root or if aortopulmonary collaterals, pulmonary veins, or systemic veins need to be visualized.
    - MRA Chest alone (CPT® 71555) should be performed if the individual cannot cooperate with full cardiac MRI exam.
- MRA Chest (CPT® 71555) is considered medically necessary for assessment of the great arteries, pulmonary veins, and systemic chest veins with inconclusive recent echocardiography findings, including the following:
  - Coarctation of the aorta
  - Tetralogy of Fallot
  - Anomalous pulmonary veins
  - Transposition of the great arteries
  - Truncus arteriosus
  - Vascular rings and other lesions of the great arteries, with inconclusive recent echocardiography findings
- CT imaging is considered medically necessary when recent echocardiogram is inconclusive:
  - Report CPT® 75574 for evaluating coronary artery anomalies
  - Report CPT® 75573 for congenital heart disease
  - CPT® 71275 Determination of vascular extra-cardiac anatomy in individuals with complex congenital heart disease

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  - Pulmonary artery (PA) and Pulmonary vein (PV) assessment
- CTA Chest (CPT® 71275) is considered medically necessary with inconclusive recent echocardiography findings to assess:
  - Coarctation of the aorta
  - Tetralogy of Fallot
  - Anomalous pulmonary veins and other lesions of the great arteries
  - Vascular rings
- Pulmonary perfusion imaging
  - Pulmonary perfusion imaging (e.g., particulate) (CPT® 78580)(CPT® 78582) (CPT® 78597)(CPT® 78598)
  - Individuals with congenital heart disease or suspected congenital heart diseases, who have clinical questions regarding relative pulmonary blood flow, can have perfusion imaging.

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Imaging and Surveillance per Congenital Lesion (PEDCD-2.4)

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- Echocardiography is often repeated frequently throughout a pediatric individuals life and can generally be approved regardless of symptoms based on the lesion and age of the individual. These are listed in sections in sections below.
  - Modifiers following guidelines
    - Some congenital conditions may require more frequent testing, especially with more complex heart disease, congestive heart failure, obstructive heart lesions, ductal dependent lesions, changes in clinical status, repeat interventions, and/or in neonates.
    - Any individual being treated for heart failure, with consideration for changing medical regimen can have an echocardiogram.
- Echocardiography is performed during the physician office visit, and these studies should not be denied because of lack of recent contact.
- Adults 18 years and older who also have congenital heart disease should be imaged according to **Congenital heart disease – General Information (CD-11.1)**

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Atrial Defects-Secundum ASD, PFO, and Partial Anomalous Pulmonary Venous Return (PAPVR), Sinus Venosus Defect (PEDCD-2.4.1)

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For imaging in AVSD see **AVSD (Atrioventricular canal, endocardial cushion defect) (PEDCD-2.4.3)**

PFO (Patent Foramen Ovale)

- Routine surveillance in an asymptomatic individual with PFO is not medically necessary.
  - PFO is a normal variant.
  - In infants, a PFO that is difficult to distinguish from an ASD can be imaged with the same guidelines as used in a small unrepaired ASD (with congenital echo).
  - Individuals with PFOs may have an additional indication for an echocardiogram and can be imaged according to the echocardiogram guidelines in **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** and **Congenital Heart Disease Modality Considerations (PEDCD-2.3)** in the general Cardiac Imaging Guidelines.
- Follow-up imaging with an echocardiogram is considered medically necessary when there is documentation of the following:
  - New cardiac symptoms
  - A concern that the last echocardiogram was equivocal for other cardiac issues
  - Question of a clot/embolism that has gone across the PFO
  - Prior echocardiogram did not differentiate the PFO from a secundum ASD
- TTE (CPT® 93306- non congenital echocardiogram) is considered medically necessary when an individual with a prior history of PFO requires an echocardiogram for any new reason.
- Preoperative for PFO closure
  - TTE or TEE
    - Closure is rare in children, TTE or TEE is medically necessary in individuals with transient ischemic attacks or strokes with suspected atrial level shunt.
    - CMR/CT-CMRA/CTA if unclear findings from echocardiogram
- Intra-procedural PFO

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- Intra-procedural TEE (CPT® 93355) is not in scope for this program.
- Post procedure PFO closure
  - Post-surgical imaging as follows (PFO generally requires less frequent monitoring post device than ASDs):
    - TTE one time within 30 days of closure
    - TTE one time within 6 months of PFO closure
  - TTE or TEE is considered medically necessary at any time post procedure when there is concern for **any** of the following:
    - Infection
    - Malposition
    - Embolization
    - Persistent shunt
      - For persistent shunt, TTE is considered medically necessary every 3 months until the finding is stable or there is a need for intervention
  -

Secundum ASD, Sinus Venosus ASD, Asymptomatic Isolated (ASD) and Unobstructed (PAPVR)

- Any surgical status
  - TTE is considered medically necessary for any of the following:
    - Initial evaluation of a change in clinical status and/or new concerning signs or symptoms
    - Prior to planned cardiac intervention
    - Repeat any time prior to next allowed study if concern for elevated pulmonary vascular resistance/Pulmonary hypertension
  - CMR/CT-CMRA/CTA
    - If anomalous vein or SV defect cannot be assessed on echo
    - To assess shunt or RV for considering of surgery, or if echocardiogram equivocal
- Unrepaired
  - Newborn with isolated ASD can have one repeat TTE within 2 months
  - Small asymptomatic isolated ASD with no pulmonary hypertension can have TTE as follows:
    - Infant <6 months every three months
    - Infant ≥6 months, repeat at one year.
    - Child Every 3 years
  - Routine surveillance for ≥moderate ASD or PAPVR >1 vein
    - Infant every 3 months
    - Echocardiogram (TTE) every 1 year

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- Prior to planned repair of ASD
  - TTE and/or TEE
  - MRI if any residual issues unanswered by echo
- Prior to planned SV defect or PAPVR
  - TTE and/or TEE
  - CMR/CT-CMRA/CTA
- Post- ASD closure with device
  - TTE post device closure
    - 1 week
    - 1 month
    - Every 3 months
    - 1 year
    - Every 2 years
  - May repeat TTE every 3 months until the finding is stable or there is a need for intervention if there is significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension
- Post-surgical closure of ASD
  - TTE
    - Within the first month
    - Within the 1st year
    - Every 2 years after the first year study
  - May repeat TTE every 3 months until the finding is stable or there is a need for intervention if significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension

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Ventricular Septal Defect (VSD) (PEDCD-2.4.2)

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All

TTE is considered medically necessary for any of the following:

- Change in clinical status and/or new concerning signs or symptoms
- Prior to planned cardiac intervention

Unrepaired

TTE

- Small muscular VSD, No Symptoms, No pulmonary hypertension
  - Newborn 1 repeat within 2 months
  - Infant every 6 months
  - Child every 3 years
- Small VSD in location other than muscular
  - Newborn 1 repeat TTE within 2 months
  - Infant TTE every 6 months
  - Child TTE every year
- Moderate or large VSD on medical management
  - Newborn TTE every 2 weeks
  - Infant every 1-month
  - Child <2 years old TTE every 3 months
  - Child >2 years old TTE every year

Post Repair VSD

TTE

- One study within one month of surgery
- One study within one year of surgery
- After first year of surgery, every 2 years
- Following surgical or device closure in an individual with significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension
  - Pre-adolescent Child (age 1-10)-TTE every 3 months
  - Adolescent (age 11-18)- TTE every 6 months

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AVSD (Atrioventricular Canal, Endocardial Cushion Defect) (PEDCD-2.4.3)

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Any surgical status

TTE is medically necessary for any of the following:

- Change in clinical status and/or new concerning signs or symptoms
- Prior to planned cardiac intervention

Unrepaired

- Partial/transitional Atrioventricular canal (AVC)
  - TTE is considered medically necessary as follows:
    - Newborn repeat once within the next 2 months
    - Infant every 3 months
    - Child <2 years every 6 months
    - Child ≥2 years, 1 year
- Complete AVC
  - TTE
    - Newborn, TTE repeat within first month
    - Infant <6 weeks, TTE every 2 weeks.
    - Infant ≥6 weeks, TTE monthly

Repaired (TTE)

- Within one month of surgery
- Within 1 year
- Then annually
- May repeat TTE every 3 months until the finding is stable or there is a need for intervention if residual shunt, valvular LV dysfunction, LVOT obstruction, arrhythmia, arrhythmia or PHT, symptoms of heart failure

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PDA (Patent Ductus Arteriosus) (PEDCD-2.4.4)

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Any surgical status

TTE is considered medically necessary for any of the following:

- Initial evaluation of a change in clinical status or new concerning signs or symptoms
- Prior to planned cardiac intervention

Unrepaired

- Newborn, one repeat TTE in newborn period
  - None, if spontaneously closed
- >1-year-old
  - No Routine surveillance in an asymptomatic individual with a trivial, silent PDA
- Infant
  - Small PDA: TTE every 3 months
  - ≥ Moderate PDA: TTE every month
- Pre-adolescent Child
  - Small PDA: every 1 year
  - Moderate PDA: every 6 months
- Adolescent every 3 years

Post PDA device

- Post procedure surveillance (TTE)
  - One echo in first 30 days
  - Annually for first 2 years
  - Every 5 years after first 2 years
- Post procedure LPA stenosis or aortic obstruction
  - Pre-adolescent Child
    - TTE annually
    - MRA/CTA Chest, or (lung perfusion for LPA stenosis) if questions remain unanswered after TTE
  - Adolescents
    - Every two years TTE and

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- MRA/CTA Chest, or (lung perfusion for LPA stenosis) if questions remain unanswered after TTE

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TAPVR Total Anomalous Pulmonary Venous Return (PEDCD-2.4.5)

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Any surgical status
- TTE, TEE, CMR/CT-CMRA/CTA, Lung perfusion scan are considered medically necessary for any of the following:
  - Change in clinical status and/or new concerning signs or symptoms
  - Prior to planned cardiac intervention

Unrepaired
- No restrictions

Repaired
- TTE one Post procedure evaluation first 30 days
- TTE every 3 months in infancy
- Pre-adolescent Child: every 1 year
- Adolescence: TTE every 2 years

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Ebstein Anomaly and TV Dysplasia (PEDCD-2.4.6)

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Any surgical status
- TTE, TEE, CMR/CT-CMRA/CTA are considered medically necessary for any of the following:
  - Change in clinical status and/or new concerning signs or symptoms
  - Prior to planned repair or intervention

Unrepaired
- Newborn repeat study within 30 days.
- Infant
  - Trivial TR is a normal finding
  - Mild TR- TTE every year
  - ≥ Moderate TR- TTE every 3 months
- Child
  - Mild TR every year TTE
  - ≥ Moderate every 6 months

Repaired (TTE)
- Post-op within 30 days
- TTE once a year
- TTE every 6 months if valvular or ventricular dysfunction, or arrhythmias
- Pre-adolescent Child: every year
- Adolescent: every 2 years
- Every 3 months if CHF or atrial arrhythmias

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Pulmonary Stenosis (PS) (PEDCD-2.4.7)

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Any surgical status
- TTE is considered medically necessary for any of the following:
  - Change in clinical status and/or new concerning signs or symptoms
  - Prior to planned cardiac procedure
  - If increasing gradient, 1 additional study prior to next allowed study
  - PS in Williams syndrome: See **LVOT Lesions (PEDCD-2.4.10)**

Unrepaired
- Neonate
  - TTE repeat study within 30 days
- Infant PS asymptotic (any severity)
  - TTE every 3 months
- Pre-adolescent Child
  - TTE every 1 year
- Adolescent
  - TTE every 2 years
  - MRA/CTA Chest if pulmonary artery dilation every 3 years

Post procedure (TTE)
- Within 30 days
- Infant
  - TTE every 3 months
- Pre-adolescent Child
  - TTE 1 year
  - Moderate or severe sequelae TTE every 6 months
- Adolescent
  - TTE every 2 years
- Any individual with heart failure, TTE every 3 months

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Pulmonary Atresia with Intact Septum (PAIVS) (PEDCD-2.4.8)

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Any surgical status

TTE is considered medically necessary for any of the following:

- Prior to planed repair
- Change in clinical status and/or new concerning signs or symptoms

Post procedural: Palliation

- TTE
  - 1 within 30 days
  - Every 1 month until repaired

Post procedural: Complete Repair

- TTE within 30 days post-op
- Any age
  - TTE every three months for CHF
- Infant
  - TTE at 3 months in asymptomatic infant
- Child (all ages)
  - TTE annually
  - Every 6 months if moderate sequelae
- Adolescent (in addition to TTE)
  - CMR/CT and/or CMRA/CTA every 3 years

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Mitral Valve Disease (PEDCD-2.4.9)

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Any surgical status
- TTE is considered medically necessary for any of the following:
  - Prior to planned surgery
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms

Unrepaired congenital mitral valve stenosis
- Infant in first three months of life
  - Weekly TTE
- After 3 months (TTE)
  - Every 3 months if mild MS
  - Every month if ≥ moderate MS
- Child (TTE)
  - With moderate MS every 3 months until a decision is made to intervene
  - Child with mild symptoms annually

Unrepaired: Congenital Mitral Regurgitation (MR) including Mitral Valve Prolapse
- Infant
  - TTE every 6 months an asymptomatic infant with mild MR
  - TTE every month in asymptomatic infant with ≥ moderate MR
- Child
  - TTE every 2 years with mild MR, normal LV size and systolic function
  - TTE every 6 months with ≥ moderate MR
  - TTE every 3 years in an asymptomatic with MVP and mild MR

Post procedure, surgical or catheter based
- TTE within 30 days
- Infant
  - TTE every 3 months, mild MS or MR, and no LV dysfunction
  - TTE every month in ≥ moderate MS or MR, dilated LV, and no LV dysfunction
- Child
  - TTE annually
    - In a child with normal prosthetic mitral valve function and no LV dysfunction
    - In a child with mild MS or MR, and no LV dysfunction

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- TTE every 3 months
  - In a child with ≥ moderate MS or MR, dilated LV, and no LV dysfunction
  - In a child with prosthetic mitral valve or ventricular dysfunction, and/or arrhythmias

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LVOT Lesions (PEDCD-2.4.10)

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Subvalvular Aortic stenosis

Any surgical status

- TTE, TEE, Cardiac MR/CT are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Preoperative
- If aortic dimension z-score >2
  - TTE or Chest CTA/MRA every 2 years if stable z-score
  - TTE or Chest CTA/MRA every 6 months if increasing z-score

Unrepaired

- Newborn- No restrictions
- Infant TTE
  - 1 monthly for any subvalvular AS, but ≤ mild AR
- Child
  - TTE one per year if mild AS and no AR
  - TTE every 6 months ≥ moderate subvalvular AS and/or mild AR
  - Routine surveillance (6–12 months) in an asymptomatic child with ≥ moderate AS and/or ≥ moderate AR

Repaired

- Infant
  - TTE within 30 days
  - TTE every 3 months ≤ mild MS and or AR
  - TTE every 1 month ≥ moderate AS or AR
- Child
  - TTE every 1 year ≤ Mild AS or AR
  - TTE every 6 months ≥ moderate AS or AR
  - TTE every 3 months if heart failure

Aortic Valve Stenosis and/or regurgitation/ BAV (Bicuspid Aortic Valve)

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Any surgical status

- TTE, TEE, Cardiac MR/CT are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Preoperative

Unrepaired

- Infant <3 months
  - TTE 1 per week
- Infant >3 months
  - TTE every 3 months
  - TTE every 1 month, if ≥ moderate AS or AR
- Child
  - TTE every 1 year with mild AS/AR and no aortic dilation
  - TTE every 6 months with moderate AS/AR, or aortic dilation.
  - TTE every 3 years if BAV with trivial or mild valvular dysfunction and no aortic root dilation
  - Every 6 months in any as with increasing z-score aortic root ascending aorta

Post procedural

- Within 30 days TTE
- Infant
  - Every 1 month following neonatal intervention with ≥ moderate AS or AR or LV dysfunction
  - Every 3 months ≤ mild AS/AR and no LV dysfunction
- Child (TTE)
  - 6 months echocardiogram if ≥ moderate AS or AR
  - 1-year echocardiogram if ≤ mild AS or AR, and/or normal prosthetic valve
  - Every 3 months if CHF or Ventricular dysfunction

Supravalvular AS

Any surgical status

- TTE, TEE, Cardiac MR/CT, Chest MRA/CTA are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Preoperative
  - Williams syndrome

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        - Individuals with Williams syndrome can be screened/evaluated for arch abnormalities and pulmonary artery abnormalities and coronary artery abnormalities with the same intervals as TTE referenced below.
    - Stress imaging can be done at initial evaluation and for cardiac symptoms, change in clinical status, and/or new concerning signs or symptoms

Unrepaired

- Infant
    - TTE every 3 months
- Child
    - TTE every 1 year
    - TTE every 6 months if moderate AS

Post-operative (TTE)

- Within 30 days
- Every 2 years in mild to moderate AS
- Every 6 months if ≥ moderate AS

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Aortic Coarctation and IAA (Interrupted Aortic Arch) (PEDCD-2.4.11)

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All individuals
- TTE, MRA/CTA Chest are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Prior to planned surgery/intervention
- Cardiac MR/CT is considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Prior to planned surgery/intervention if any issues remain not answered on echo

Screening for related intracranial aneurysms see **Cerebral Intracranial Aneurysms (HD-12.1)**

Unrepaired Aortic Coarctation
- Newborn, TTE weekly if assessing for ductal closure
- Infant with mild coarctation in absence of PDA
  - Echocardiogram every 3 months
- Child with mild coarctation
  - Echocardiogram every 1 year
  - MRA Chest, CTA Chest every 3 years

Post procedure: surgical or catheter based
- TTE
  - Within 30 days of procedure
  - Every 3 months if mild or no sequelae in first year
  - Every 6 months if mild or no sequelae in the second year
  - Every 1 year after the second year
  - Every 3 months at any time if CHF symptoms or ≥ moderate sequelae
- MRA/CTA Chest every 3 years (include Cardiac MR/CT if issues not clarified on echo)
- Screening for intracranial aneurysms see **Cerebral Intracranial Aneurysms (HD-12.1)**

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Coronary Anomalies (PEDCD-2.4.12)

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- CCTA or cardiac MRI is considered medically necessary for evaluating coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels.
  - CPT® 75574 for evaluating coronary artery anomalies
  - CPT® 75573 for congenital heart disease
  - CTA Chest (CPT® 71275) can be added to evaluate great vessels.
- Congenital anomalies of the coronary arteries are an important cause of sudden death in pediatric individuals. Coronary arteries may arise from the wrong coronary artery cusp leading to ischemic changes during exercise. These lesions may be found incidentally during a murmur evaluation. Anomalous coronary arteries may be seen on echocardiogram during an evaluation for chest pain or syncope or palpitations. In addition, individuals with no echocardiographic findings but symptoms concerning for angina chest pain may require stress testing.
  - Individuals who have positive echocardiographic findings, regardless of symptoms, and individuals who have classical typical angina chest pain regardless of echocardiographic findings, may require treadmill stress testing, stress imaging, or advanced imaging such as Cardiac MRI, Stress echocardiogram, PET, Cardiac CT, and/or cardiac catheterization.
- Congenital coronary anomalies include abnormal origin of a coronary artery from the PA, anomalous aortic origin of a coronary artery from a different aortic sinus of Valsalva (left coronary artery from the right sinus of Valsalva or right coronary artery from the left sinus of Valsalva), coronary arteriovenous fistula, and coronary artery ostial atresia, all in the setting of normal conotruncal anatomy.
  - Any surgical status
    - Prior to planned surgery, or change in clinical status and/or new concerning signs or symptoms
      - TTE
      - CMR or CCT
        - Can initially include MRA/CTA Chest
        - If the origin of the coronaries arteries is below the sinus of Valsalva then a chest study is not needed on subsequent imaging.
        - If the origin of the coronary artery is not at the level of the sinus of Valsalva, a MRA/CTA Chest can be included when MR/CT imaging is required
      - Stress imaging- to assess the need for surgery

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- Unrepaired
  - Routine surveillance every 2 years in an asymptomatic individual with anomalous right coronary artery from the left aortic sinus
    - TTE
    - Stress imaging
  - Although typically repaired, in the event that a repair is not completed, anomalous left coronary artery from the right coronary sinus can have imaging.
    - TTE annually
    - Stress imaging annually
  - Routine surveillance in an asymptomatic individual with small coronary fistula
    - TTE- every 2 years
  - Routine surveillance in an asymptomatic individual with moderate or large coronary fistula
    - TTE annually
- Post-procedural: surgical or catheter
  - TTE
    - Within 30 days of procedure
    - Monthly the first year following repair
    - Every 3 months after first year of surgery
    - Annually after the second year of surgery
    - Every 3 months if ventricular dysfunction
  - Stress testing
    - EKG stress testing without imaging may be medically necessary in the first post year and every 1-2 years depending on level of activity.
    - Stress testing with imaging
      - First postoperative year
      - If EKG stress test positive of equivocal
- Change in clinical status and/or new concerning signs or symptoms
- Individuals with congenital heart disease such as TOF, Truncus Arteriosus, and TGA have increased incidence of coronary artery anomalies
- Individuals with Williams syndrome can have coronary artery stenosis
- Individuals with confirmed coronary artery anomalies may require repeat imaging based on the clinical scenario
- CCTA to rule out anomalous coronary artery should be limited to one of the following:
  - Individuals who need to have an anomalous coronary artery mapped prior to an invasive procedure
  - Individuals who have not had a previous imaging study that clearly demonstrates an anomalous coronary artery

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- Individuals with a history that includes one or more of the indications in **Indications for CCTA (PEDCD-10.3)**

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Tetralogy of Fallot (TOF) (PEDCD-2.4.13)

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Any surgical status
- TTE, CMR/CT-CMRA/CTA
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Evaluation prior to planed pulmonary valve replacement, cardiac intervention, or surgery

Unrepaired
- Newborn-TTE no limits
- Infant
  - 1 per month

Post procedure palliation
- 1 per month following palliative procedure prior to complete repair, valvuloplasty, PDA and/or RVOT stenting, or shunt placement before complete repair

Post-operative TOF (initial repair)
- TTE
  - Within 30 days of repair
  - Pre-adolescent Child every 12 months
  - Adolescence every 24 months
  - Every 6 months in an individual with valvular dysfunction other than pulmonary valve, RVOT obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit
  - TTE every 3 months if CHF
- Cardiac MR/CT, MRA/CTA Chest every
  - Routine surveillance (36 months) in an individual with PR and preserved ventricular function
  - 12 months if moderate (≥150 mL/m2) or progressive (increase of >25 mL/m2) RV dilatation or dysfunction (RVEF ≤48% or ≥6% decrease in EF) or nearing imaging criteria for PVR.

Post-surgical or catheter based pulmonary valve replacement
- TTE

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  - Within 30 days follow-up
  - 1 and 6 months after replacement
  - One year post procedure
  - Annually after replacement
  - Every 6 months if RV-to-PA conduit dysfunction, valvular or ventricular dysfunction, branch pulmonary artery stenosis, or arrhythmias
  - Every 3 months if CHF symptoms
- CMR/CT-CMRA/CTA every 2 years

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Double Outlet Right Ventricle (DORV) (PEDCD-2.4.14)

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Any surgical status

- TTE, CMR/CT-CMRA/CTA are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Evaluation prior to repair

Unrepaired

- TTE
  - Newborn no limit
  - Monthly infant with balanced systemic and pulmonary circulation
  - Every 3 months child with balanced circulation

Postoperative

- TTE
  - Within 30 days
  - First year post-op every 6 months
  - After one year, TTE every 1 year
  - TTE 3 months in an individual with valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit, heart failure
- Cardiac MR/CT, MRA/CTA Chest
  - 3 years for asymptomatic individual

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D-Loop Transposition of the Great Arteries (D-Loop TGA) (PEDCD-2.4.15)

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Any surgical status

- **TTE, CMR/CT-CMRA/CTA**, s**tress imaging** are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Any time after procedure involving coronary arteries
- **CMR/CT-CMRA/CTA** every 5 years.

Unrepaired (TTE)

- No Limits

Post arterial switch

- TTE
  - Within 30 days of repair
  - Infant: every one month
  - Child: every 3 months
  - Child with ≥ moderate sequelae: TTE every three 3 months (moderate valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, branch pulmonary artery stenosis, or arrhythmias
- Routine CMR/CT
  - Every 3 years
  - Every year if neo AI
- MRA/CTA Chest
  - Every 3 years
  - Every year if neo AI or aortic dilation
- Stress imaging
  - 1 routine test after arterial switch at any time

Post Rastelli

- TTE
  - Within 30 days
  - Every three months following procedure for one year

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  - Pre-adolescent Child Every 6 months following the first year after repair if no or mild sequelae
  - Adolescent annually
  - Every three months if moderate valvular dysfunction, LVOT obstruction, presence of an RV-to-PA conduit, branch, pulmonary artery stenosis, or arrhythmias, or heart failure
- CMR/CT-CMRA/CTA every 3 years

Post atrial switch

- TTE Every 1 year if mild to no symptoms
  - Every 3 months TTE, and CMR MRA CCT CTA if ≥moderate systemic AV, valve regurgitation, systemic RV dysfunction, LVOT obstruction, or arrhythmias, or CHF
  - Routine **CMR/CT-CMRA/CTA** every 3 years

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Congenitally Corrected Transposition of the Great Arteries (ccTGA, LTGA) (PEDCD-2.4.16)

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Any surgical status

- TTE, TEE, **CMR/CT-CMRA/CTA** are considered medically necessary for any of the following:
  - Change clinical status and/or new concerning signs or symptoms
  - Preoperative evaluation (typically within one month)
- CMR/CT-CMRA/CTA every 3 years

Unrepaired

- TTE
  - Newborn-Weekly
  - Infant
    - Every 3 months if no cardiac symptoms and only mild findings
    - Every 1 month is cardiac symptoms and moderate findings
  - Child
    - <2 years every 3 months
    - >2 years every 1 year
    - Every 6 months if ≥moderate AV regurgitation
    - Every 3 months if CHF symptoms
  - CMR/CT-CMRA/CTA
    - Every 3 years

Postoperative: Anatomic Repair

- TTE
  - Post–operative evaluation (within 30 days)
  - Every 3 months within a year following repair in an asymptomatic individual with no or mild sequelae
  - Every 1 year after the first year following repair in an asymptomatic individual with no or mild sequelae
  - Every 6 months if valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, or presence of a RV-to-PA conduit

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  - Every 3 months if CHF symptoms
- CMR/CT-CMRA/CTA
  - Every 3 years

Postoperative: Physiological Repair with VSD Closure and/or LV-to-PA Conduit

- TTE
  - Postoperative evaluation (within 30 days)
  - Every 3 months within a year following repair in an asymptomatic individual with no or mild sequelae
  - Annually in an asymptomatic individual with no or mild sequelae
  - Every 3 months if in an individual with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction, or with CHF symptoms
- CMR/CT-CMRA/CTA every 3 months in an individual with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction, or with CHF symptoms

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Truncus Arteriosus (PEDCD-2.4.17)

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Any surgical status

- TTE, CMR/CT-CMRA/CTA are considered medically necessary for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Prior to planned intervention or surgery

Postoperative

- TTE
  - Within 30 days
  - Monthly in first year after surgery
  - After first year every 6 months
  - Every 3 months if
    - ≥ moderate truncal stenosis or regurgitation
    - Residual VSD or RV to PA conduit or Branch PA obstruction
    - Symptoms of CHF
- CMR/CT-CMRA/CTA
  - Annually if ≥ moderate Truncal stenosis or regurgitation

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Single Ventricle (SV) (PEDCD-2.4.18)

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SV references individuals not amenable to biventricular repair, including but not limited to hypoplastic left heart syndrome, tricuspid atresia, Double inlet left ventricle, mitral atresia, unbalanced AVSD, and forms of PA/IVS.

Any surgical status

- Any/All: **TTE, TEE, CMR/CT-CMRA/CTA, and cardiac catheterization** (CPT® 93593, 93594, 93595, or 93597) are considered medically necessary for any of the following:
  - Change clinical status and/or new concerning signs or symptoms
  - Preoperative evaluation (typically within one month)

Unrepaired SV

- TTE allowed one study per week

Stage 1 palliation (TTE)

- Often called Norwood or Sano, or hybrid catheterization procedure
- Routine weekly TTE

Stage 2 palliation (TTE)

- Often referred to as Glenn procedure
- Within 30 days after surgical or catheterization intervention
- 1 per month in infant or child

Stage III, also called Fontan

- TTE within 30 days
- TTE every three months within first post-op year
- Every 6 months after first year
- Every 3 years allow **CMR/CCT-MRA/CTA**
- TTE every 3 months until the finding is stable or there is a need for intervention if there is valvular dysfunction, arrhythmias, heart failure

Surveillance

Cardiac catheterization is considered medically necessary for all Single Ventricles once every 10 years.

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Eisenmenger and PHT (with CHD)-(PEDCD-2.4.19)

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PHT without CHD is covered in section **Pediatric Pulmonary Hypertension General (PEDCD-7.1)**

These are in addition to studies supported by lesion.

Any surgical status
- TTE, CMR/CT-CMRA/CTA are considered medically necessary for any of the following:
  - Change clinical status and/or new concerning signs or symptoms
  - Preoperative evaluation (typically within one month)

Initial evaluation (TTE)
- Change in clinical status and/or new concerning signs or symptoms
- Before and after PHT therapy

Eisenmenger Syndrome (ES) individual
- TTE every 6 months

PHT associate with CHD
- Unrepaired individuals with evidence of elevated pulmonary vascular resistance can have echocardiograms based on the frequency requested by the provider.
- TTE and Cardiac CMR/CCT for changes in change in pulmonary arterial hypertension-targeted therapy in an individual with postoperative PH
- TTE every 3 months in postoperative stable child with PHT

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Multisystem Inflammatory Syndrome in Children (MIS-C)

Guideline

Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)
References (PEDCD-12)

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Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)

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MIS-C Indications for Cardiac Imaging (PEDCD-12.2)

MIS-C Initial Cardiac Imaging (PEDCD-12.2.1)

- When there is concern for MIS-C, as in atypical or incomplete Kawasaki (see **Kawasaki Disease - Acute Phase (PEDCD-6.2)**), echocardiogram (TTE) can be approved
- A cardiac MRI can be approved at the time of diagnosis when there are issues that can affect treatment management not answered by other testing.
- Cardiac CCTA can be done if there is incomplete visualization of the coronary arteries.
- Repeat echocardiograms may be required and approved if either:
  - Treatment decisions will be affected by results (e.g., treating with IVIg).
  - There are new signs or symptoms.

MIS-C Repeat Cardiac Imaging (PEDCD-12.2.2)

The following imaging guidelines reference outpatient management of individuals who have been discharged from the hospital after stability for MIS-C has been established.

- An echocardiogram (TTE) can be approved at the time of presentation and followed by serial echocardiograms (TTE) until stabilization has been achieved for any of the following:
  - New cardiac signs, symptoms, or findings
  - Evidence of recurrence of MIS-C
  - Changes in medication
- Serial echocardiograms can be approved based on the ordering cardiologist's discretion or the treating medical provider in consultation with a cardiologist when there is documented cardiac dysfunction.
- Individuals who are discharged from the hospital after MIS-C and have stable findings can have an echocardiogram (TTE):
  - Within 1 week of discharge
  - 4 weeks post-discharge
  - At 6 months post-discharge
  - One year post-discharge

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- Cardiac CCTA can be done if there is incomplete visualization of the coronary arteries.
- A routine cardiac MRI can be done once after 3 months in an individual with evidence of cardiac involvement (e.g., symptoms, EKG, labs, or echocardiogram).
- Individuals with changes, or unanswered questions, on echocardiogram (TTE) may have a Cardiac MRI based on **Cardiac MRI and Cardiac Indications for MRA Chest (CD-5.2)** in the cardiac imaging guidelines.

Individuals with dilated coronary arteries can have imaging based on the AHA Kawasaki guidelines.

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echocardiogram (TTE)	Routine Stress Imaging	Routine Coronary Imaging
All			All risk levels 4-6 weeks after acute illness		
1	Normal	Normal	One TTE 2-12 months after acute illness	none	none
2	Dilation	Dilation	6 months One year If dilation remains TTE every 2-5 years until resolves	None	None
		Normal	After acute illness: 2-12 months One echocardiogram at one year No echocardiogram after one year		
3.1	Small	Small	6 months 12 months Then yearly	2-3 years	3-5 years

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AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echocardiogram (TTE)	Routine Stress Imaging	Routine Coronary Imaging
3.2	Small	Normal or dilated	6 months 12 months Then yearly	3-5 years	none
4.1	Medium	Medium	3 months 6 months 12 months Every 6-12 months after that	1-3 years	2-5 years
4.2	Medium	Small	6 months and 12 months, every 1 year	2-3 years	3-5 years
4.3	Medium	Normal Or Dilated	Every 1-2 years	2-4 years	none
5.1	Large	Large	1 month 3 months 6 months 9 months 12 months Then every 3-6 months	6-12 months	at 2-6 months, every 1-5 years
5.2	Large	Medium	Every 6-12 months	yearly	2-5 years
5.3	Large	Small	6-12 month	1-2 years	2-5 years

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AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echocardiogram (TTE)	Routine Stress Imaging	Routine Coronary Imaging
5.4	Large	Normal Or Dilation	1-2 years	2-5 years	none

Symptomatic individuals

- Echocardiogram can be performed at any time with new or progressing cardiac symptoms.
- Stress imaging when there are new or progressing symptoms of ischemia or ventricular dysfunction
- Invasive or coronary imaging coronary angiography (CT, MRI, invasive) when the above studies are positive, inconclusive, or otherwise lead to a conclusion that intervention is needed

Evidence Discussion

SARS-CoV-2 (COVID 19) is usually mild in children. Some children develop a severe inflammatory disease that can present in a similar way to Kawasaki disease or toxic shock syndrome. This syndrome has been defined by the US Centers for Disease Control and Prevention as multisystem inflammatory syndrome in children (MIS-C).

These guidelines are intended for use in the outpatient management of cardiac findings of MIS-C. Additional information can be found in **COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)** for the outpatient management of head imaging.

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Heart Murmur

Guideline

Heart Murmur (PEDCD-3.1)
References (PEDCD-3.1)

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Heart Murmur (PEDCD-3.1)

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When evaluating an individual with a murmur for the first time, it will not be known whether the individual has congenital heart disease or not.

Initial pediatric TTE combinations

Cardiologist only submits charges for the procedure actually performed
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- Any one of the above echocardiography code combinations is considered medically necessary for evaluation of either:
  - Any pathologic murmur
  - Any innocent murmur with associated cardiac signs or symptoms
- Repeat echocardiography is not medically necessary if the initial echocardiogram was normal and the murmur has not changed in quality.

Evidence Discussion

- Heart murmurs are extremely common in pediatric individuals. The thinner chest wall in children allows clearer auscultation of blood flowing through the chambers of the heart, which may result in a murmur on physical exam.
- The majority of murmurs are innocent and do not require further evaluation. More than 30% of children may have an innocent murmur detected during physical examination. Innocent murmurs are typically systolic ejection murmurs with a vibratory or musical quality and generally change in quality when the individual changes position.
- Other types of murmurs can be pathologic and require additional evaluation, usually by a pediatric cardiologist. Echocardiography is considered medically necessary and is performed as part of the office visit.

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Chest Pain

Guideline

Chest Pain General (PEDCD-4.1)
References (PEDCD-4.1)

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Chest Pain General (PEDCD-4.1)

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Initial pediatric TTE combinations

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- Any one of the TTE combinations above is considered medically necessary for pediatric individuals with chest pain and one or more of the following:
  - Exertional chest pain
  - Non-exertional chest pain with abnormal EKG
  - Chest pain with signs or symptoms of pericarditis
  - First-degree relative with sudden unexplained death or cardiomyopathy
  - Recent onset of fever
  - Recent illicit drug use
  - Other signs or symptoms of cardiovascular disease
- Repeat echocardiography is not medically necessary if the initial echocardiogram is normal unless one of the following conditions is present:
  - Increased severity or change in quality of the chest pain
  - New signs or symptoms of cardiovascular disease other than pain
  - New abnormality on EKG
- Cardiac MR or cardiac CT is considered medically necessary for chest pain if prior evaluation suggests:
  - Any coronary artery abnormalities
  - Cardiomyopathy
  - Myocarditis
- Chest MRA or CTA if pulmonary embolism or aortic dissection is suspected
- Stress imaging is considered medically necessary if other imaging suggests coronary artery abnormality, or ETT suggests ischemia. EKG is uninterpretable. Any indication in section **Stress Testing with Imaging (CD-1.4)** in the Cardiac Imaging Guidelines. This can include Stress SPECT, echo or MR.

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Evidence Discussion

Chest pain in pediatric individuals is caused by a cardiac etiology in <5% of cases, yet causes great anxiety for parents resulting in requests for testing.

Individuals with CP may undergo an exercise stress test without imaging.

Echocardiography is performed as part of the office visit. When evaluating an individual for the first time, it will not be known whether the individual has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.

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Syncope

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Syncope (PEDCD-5.1)

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Initial pediatric TTE combinations

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- Echocardiography is not medically necessary for most individuals with isolated syncope.
- Echocardiography is considered medically necessary for pediatric individuals with syncope and one or more of the following:
  - Exertional syncope
  - Unexplained post-exertional syncope
  - Abnormal EKG
  - Absence of prodromal symptoms
  - Presence of preceding palpitations within seconds of loss of consciousness
  - Lack of a prolonged upright posture
  - Syncope in response to auditory or emotional
  - First-degree relative with any of the following before age 50:
    - Sudden cardiac arrest or death
    - Pacemaker or implantable defibrillator placement
  - First-degree relative with cardiomyopathy
  - Known congenital heart disease
  - History of Kawasaki disease, or other coronary pathology
  - Pathologic murmur, irregular rhythm, gallop, or click on physical examination
- Repeat echocardiography is not medically necessary if the initial echocardiogram is normal unless one of the following conditions is present:
  - Increased severity or change in quality of the syncope
  - New signs or symptoms of cardiovascular disease other than syncope
  - Family of history of sudden death, cardiomyopathy
  - New abnormality on EKG

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- Cardiac MR or Cardiac CT is considered medically necessary for chest pain if prior evaluation suggests any coronary artery abnormalities, cardiomyopathy, myocarditis.
- MRA or CTA Chest if pulmonary embolism or aortic dissection is suspected
- Stress imaging (SPECT, echo or MR) is considered medically necessary (any):
  - If other imaging suggests coronary artery abnormality
  - ETT suggests ischemia
  - EKG is uninterpretable
  - Any indication in section **Stress Testing with Imaging (CD-1.4)** in the Cardiac Imaging Guidelines

Evidence Discussion

Syncope in pediatric individuals is common, with up to 15% of individuals experiencing at least one episode by age 21. Syncope is caused by neurocardiogenic syndrome (vasovagal syncope) in 75% to 80% of cases, which is a benign and self-limiting condition. Despite this, syncope causes great anxiety for parents resulting in requests for testing.

Individuals with CP may undergo an exercise stress test without imaging.

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Pediatric Pulmonary Hypertension General

Guideline

Pediatric Pulmonary Hypertension General (PEDCD-7.1)
References (PEDCD-7.1)

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Pediatric Pulmonary Hypertension General (PEDCD-7.1)

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- Pulmonary hypertension in children can be caused by cardiac, pulmonary, or systemic diseases, and idiopathic disease occurs as well.
- Chest x-ray, EKG, and echocardiography (CPT® 93306, or CPT® 93303, with CPT® 93320, and CPT® 93325, see **Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)** for echocardiography coding considerations) for initial evaluation if pulmonary hypertension is suspected.
- Repeat echocardiography intervals are variable depending on age of individual, etiology, and severity.
  - After a comprehensive initial evaluation, echocardiograms using PH-specific protocols may be performed every 4 to 6 months.
  - Echocardiography is considered medically necessary at any time for new or worsening symptoms or to evaluate a recent change in therapy.
  - Right heart and/or left heart catheterization may be utilized for PAH individuals, including before and after initiation of PH-targeted therapy, and for individuals with concomitant congenital heart disease.
- CT Chest (CPT® 71250) may be medically necessary in addition to CTA Chest (CPT® 71275) or MRA Chest (CPT® 71555) for initial evaluation of all pediatric individuals with pulmonary hypertension to evaluate for pulmonary vascular or interstitial disease, or other intrathoracic causes.
- Cardiac MRI without and with contrast (CPT® 75561) is considered medically necessary for evaluation of inconclusive echocardiogram findings or for monitoring right ventricular function during follow-up.
- Stress echocardiograms may be medically necessary(as in the general cardiac imaging guidelines) see **Stress echo – coding** in the Cardiac Imaging Guidelines.

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Pericardial Effusion or Pericardial Tamponade

Guideline

Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade (PEDCD-9.5)

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Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade (PEDCD-9.5)

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- Echocardiogram is the initial imaging study of choice to evaluate pericardial effusions or diagnose pericardial tamponade.
- If a specific clinical question is left unanswered by another recent imaging study, **and** the answer to the clinical question will affect the management of the individual's clinical condition, contrast-enhanced cardiac MRI is useful for evaluating:
  - Pericarditis
  - Neoplastic effusion
  - Tamponade
  - Myocardial infiltration
- Cancers that can metastasize to the pericardium or myocardium and can cause a malignant effusion include lung, breast, renal cell, lymphoma and melanoma.

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Kawasaki Disease

Guideline

Kawasaki Disease (PEDCD-6)
References

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Kawasaki Disease (PEDCD-6)

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Kawasaki Disease Initial Imaging (PEDCD-6.1)

- Echocardiography (CPT® 93306) is considered medically necessary for initial assessment for suspected or known Kawasaki disease.
  - Coronary CTA (CPT® 75574), Cardiac MRI without contrast (CPT® 75557), Cardiac MRI without and with contrast (CPT® 75561), or MRA Chest (CPT® 71555) are considered medically necessary for evaluation of inconclusive echocardiogram findings or significant coronary artery abnormalities.
  - Screening of other body areas for aneurysms is not routinely medically necessary in Kawasaki disease, but MRA or CTA (contrast as requested) of the affected body area can be approved for evaluation of signs or symptoms suggesting aneurysm development.
  - See acute and chronic phase for imaging

Background and Supporting Information

- Kawasaki disease (KD) is the leading cause of acquired pediatric cardiac disease in the developed world. It is an acute febrile illness characterized by a medium vessel vasculitis, which predominantly affects the coronary arteries.
  - Individuals who do not fulfill the diagnostic criteria for classic KD may be considered to have incomplete (atypical) KD.
  - If Kawasaki disease is strongly suspected, treatment will often begin even before cardiac evaluation, since early treatment is associated with a lower risk for coronary aneurysm development.

Kawasaki Disease - Acute Phase (PEDCD-6.2)

- Echocardiography is medically necessary when the diagnosis of KD is suspected
  - In uncomplicated presentations, repeat echocardiography is medically necessary at the following intervals after treatment:
    - Within 1 to 2 weeks
    - Within 4 to 6 weeks
  - In presentations with important and evolving coronary artery abnormalities (z-score >2.5) detected during the acute illness, repeat echocardiography (at least twice per

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week) to monitor for thrombosis is medically necessary until luminal dimensions have stopped progressing.
    - In presentations with expanding large or giant aneurysms, TTE is medically necessary at the following intervals:
        - Twice per week while dimensions are expanding rapidly
        - Once weekly after dimension is stabilized for the first 45 days of illness
        - Then, monthly until the third month after illness onset
- CCTA, or Cardiac MRI, or coronary angiography are considered medically necessary in the acute phase in severe cases to evaluate proximal coronary artery abnormalities when results will impact management decisions.
- Transesophageal echocardiography, Cardiac MRI, and CCTA are not routinely used in the acute phase of illness but are considered medically necessary when **both**:
    - Visualization of the coronary arteries with Transthoracic echocardiography (TTE) is inadequate **and**
    - Results will impact immediate management decisions
- Evaluation of potential aneurysmal involvement in other arterial beds can be assessed with CMRI, CTA, and, rarely, invasive angiography after recovery from the acute illness for individuals with severe coronary artery involvement or symptoms or signs, such as the presence of a pulsatile axillary mass.
- Atypical or incomplete Kawasaki. Echo is considered medically necessary when atypical KD is being considered, may require repeat echocardiograms if treatment decisions will be affected by results (e.g., treating with IVIg), if new signs or symptoms (such as typical peeling) develop.

Evidence Discussion

- The acute phase of Kawasaki disease (KD) can last up to 4-6 weeks from the onset of fever until acute systemic inflammation has resolved and coronary artery dimensions are no longer expanding.
- Based on AHA recommendations, the following classifications are used in risk stratification of coronary artery abnormalities[1]:
    - Z-score classification accounts for the effects of body size and age through use of baseline coronary dimensions adjusted for body surface area. The z-score value represents the number of standard deviation above the mean. (e.g., z=0 pt. has coronary artery dimension value equal to mean, z=2 person has value 2 standard deviation above the mean, based on age, gender, BSA).
    - Coronary Artery Abnormalities Risk Classification based on z-score:
        - 1 - No involvement at any time point (z-score always <2)
        - 2 - Dilation only (z-score 2 to <2.5)

[1] Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. Circulation. 2017;135(17). doi:10.1161/cir.0000000000000484.

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        - 3 - Small aneurysm (z-score ≥2.5 to <5)
            - 3.1 - Current or persistent
            - 3.2 - Decreased to dilation only or normal luminal dimension
        - 4 - Medium aneurysm (z-score ≥5 to <10, and absolute dimension <8 mm)
            - 4.1 - Current or persistent
            - 4.2 - Decreased to small aneurysm
            - 4.3 - Decreased to dilation only or normal luminal dimension
        - 5 - Large and giant aneurysm (z-score ≥10, or absolute dimension ≥8 mm)
            - 5.1 - Current or persistent
            - 5.2 - Decreased to medium aneurysm
            - 5.3 - Decreased to small aneurysm
            - 5.4 - Decreased to dilation only or normal luminal dimension
    - Additional clinical features that may increase the long-term risk of myocardial ischemia:
        - Greater length and distal location of aneurysms that increase the risk of flow stasis
        - Greater total number of aneurysms
        - Greater number of branches affected
        - Presence of luminal irregularities
        - Abnormal characterization of the vessel walls (calcification, luminal myofibroblastic proliferation)
        - Presence of functional abnormalities (impaired vasodilation, impaired flow reserve)
        - Absence or poor quality of collateral vessels
        - Previous revascularization performed
        - Previous coronary artery thrombosis
        - Previous myocardial infarction
        - Presence of ventricular dysfunction

Kawasaki Disease - Chronic Phase (PEDCD-6.3)

- Long-term management begins at the end of the acute illness, usually at 4 to 6 weeks after fever onset. Management is based on two pieces of data:
    - The dimensions of the largest aneurysm at any point during the disease
    - The dimensions of the largest current aneurysm
- Additional risk factors that may be considered for imaging:
    - Greater length and distal location of aneurysms that increase the risk of flow stasis
    - Greater total number of aneurysms
    - Greater number of branches affected

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- Presence of luminal irregularities
- Abnormal characterization of the vessel wall (calcification, luminal myofibroblastic proliferation)
- Presence of functional abnormalities (impaired vasodilation, impaired flow reserve)
- Absence or poor quality of collateral vessels
- Previous revascularization performed
- Previous coronary artery thrombosis
- Previous myocardial infarction
- Presence of ventricular dysfunction
- Long term routine surveillance in asymptomatic imaging for Kawasaki disease-see chart

Long term routine surveillance in asymptomatic imaging for Kawasaki disease

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
All			All risk levels 4-6 weeks after acute illness		
1	Normal	Normal	One echo 2-12 months after acute illness	none	none
2	Dilation	Dilation	6 months One year If dilation remains echo every 2-5 years until resolves	None	None
		Normal	After acute illness: 2-12 months One echocardiogram at one year No echocardiogram after one year		

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AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
3.1	Small	Small	6 months 12 months then yearly	2-3 years	3-5 years
3.2	Small	Normal or dilated	6 months 12 months Then yearly	3-5 years	none
4.1	Medium	Medium	3 months 6 months 12 months Every 6-12 months after that	1-3 years	2-5 years
4.2	Medium	Small	6 months and 12 months, every 1 year	2-3 years	3-5 years
4.3	Medium	Normal Or Dilated	Every 1-2 years	2-4 years	none
5.1	Large	Large	1 month 3 months 6 months 9 months 12 months Then every 3-6 months	6-12 months	at 2-6 months, every 1-5 years
5.2	Large	Medium	Every 6-12 months	yearly	2-5 years
5.3	Large	Small	6-12 month	1-2 years	2-5 years

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AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
5.4	Large	Normal Or Dilation	1-2 years	2-5 years	none

Symptomatic individuals

- Echocardiogram can be performed at any time with new or progressing cardiac symptoms.
- Stress imaging when there are new or progressing symptoms of ischemia or ventricular dysfunction
- Invasive or coronary imaging Coronary angiography (CT, MRI, invasive) when the above studies are positive, inconclusive, or otherwise lead to a conclusion that intervention is needed.

**Adapted from: Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. Circulation. 2017;135(17). doi:10.1161/cir.0000000000000484.

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Echocardiography

Guideline

Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)
Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)
Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)
Transesophageal Echocardiography (TEE) (PEDCD-8.4)
References (PEDCD-8)

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Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)

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- CPT® codes for echocardiography are listed in **General Guidelines (PEDCD-1.0)**

Echocardiogram coding notes	CPT®
• The most commonly performed study is a complete transthoracic echocardiogram with spectral and color flow Doppler (CPT® 93306). ◦ CPT® 93306 includes CPT® 93320 and CPT® 93325, so those codes should not be approved along with CPT® 93306.	93306
• Doppler codes (CPT® 93320, CPT® 93321, and CPT® 93325) are add-on codes and are assigned in addition to code for the primary procedure, and should not be approved alone.	+93320 +93321 +93325
• For a 2D transthoracic echocardiogram without Doppler, report CPT® 93307.	93307
• A limited transthoracic echocardiogram is reported with CPT® 93308. ◦ Limited transthoracic echocardiogram should be billed if the report does not "evaluate or document the attempt to evaluate" all of the required structures. ◦ Unlike CPT® 93306, the Doppler CPT® 93321 and CPT® 93325 are not included with CPT® 93308. ◦ CPT® 93321 (not CPT® 93320) should be reported with CPT® 93308 if Doppler is included in the study. ◦ CPT® 93325 should also be reported with CPT® 93308 if color flow Doppler is included in the study.	93308
• For individuals with known congenital heart disease, a limited transthoracic echocardiogram is reported with CPT® 93304, +/- CPT® 93321 and CPT® 93325.	93304

- Providers performing an **initial** echocardiogram on a pediatric individual will not know what procedure codes they will be reporting until the initial study is completed.

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- If congenital heart disease is found on the initial echo, a complete echocardiogram is reported with codes CPT® 93303, CPT® 93320, and CPT® 93325 because CPT® 93303 does NOT include Doppler and color flow mapping.
- If no congenital issue is discovered, then CPT® 93306 is reported alone and includes 2-D, Doppler and color flow mapping.

- Since providers may not know the appropriate code(s) that will be reported at the time of the pre-authorization request, they may request multiple codes.
  - The following echocardiography code combinations for any **initial** echocardiogram:
    - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
    - CPT® 93303, CPT® 93306
    - CPT® 93306
      - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
  - Post-service audits may be completed to ensure proper claims submission.
- Correct coding for subsequent echocardiograms
  - If an individual is being followed for known congenital heart disease, and an echocardiogram is considered medically necessary, the appropriate codes are (CPT® 93303 or 93304) in addition to appropriate doppler codes (CPT® 93320 or 93321) and CPT® 93325.
  - If an individual has documented normal anatomy or acquired heart disease, and an echocardiogram is considered medically necessary, non-congenital codes are appropriate CPT® 93306 (includes all Doppler codes) or CPT® 93308 with CPT® 93321 and CPT® 93325.
  - For individuals with newborn physiology (e.g., ASD versus PFO, or PDA), the final echocardiogram that documents normal anatomy can be coded as congenital. However, any subsequent echocardiograms after that, which would be completed for a new indication, (e.g., shortness of breath) would be coded as non-congenital.

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Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)

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- In addition to indications listed in previous guideline sections, initial TTE evaluation is considered medically necessary for any of the following:
  - Any signs/symptoms that are possibly cardiac in nature, including (but not limited to) central cyanosis, dyspnea, edema, poor peripheral pulses, feeding difficulty, decreased urine output, hepatomegaly, or desaturation on pulse oximetry.
  - Abnormal EKG or cardiac biomarkers
  - Abnormal chest x-ray suggesting cardiovascular disease
  - First-degree relative with any of the following before age 50:
    - Sudden cardiac arrest or death
    - Pacemaker or implantable defibrillator placement
    - First-degree relative with cardiomyopathy
  - Supraventricular Tachycardia (SVT), Ventricular Tachycardia, or Premature Ventricular Contractions (PVCs)
  - Known or suspected valvular dysfunction
  - Persistent systemic hypertension
  - Individuals with new onset hypertension
    - TTE indicated to assess for cardiac target organ damage (LV mass, geometry, and function) at the time of consideration of pharmacologic treatment of systemic hypertension
  - Obesity (BMI >30) with additional cardiovascular risk factors
  - Stroke
  - Renal failure
  - Preoperative evaluation of individuals with chest wall deformities or scoliosis
  - Known or suspected vascular ring
  - Planned administration of cardiotoxic chemotherapy
    - Generally anthracyclines (doxorubicin, daunorubicin, mitoxantrone, idarubicin, epirubicin)
  - Planned radiation therapy involving heart muscle or hematopoietic stem cell transplant
  - Sickle cell disease or other hemoglobinopathy causing chronic anemia
  - Known or suspected vasculitis, acute rheumatic fever, or other systemic autoimmune disease
  - Aortopathy (such as Marfan, Ehlers-Danlos, Loeys-Dietz)

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    - Positive personal diagnosis
    - First degree relative
    - Positive gene
    - Finding suggestive of aortopathy such as x-ray showing aortic dilation
  - Muscular dystrophy
    - Positive personal diagnosis
    - First degree relative
    - Positive gene
    - Any findings suggestive of MD, such as neurological exam
  - Cardiomyopathy
    - Diagnosed by other modality (such as cardiac MR)
    - First degree relative
    - Positive genetic testing
    - Findings suggestive of, such as cardiomegaly on x-ray
  - Metabolic, mitochondrial, and storage disorders
    - Positive personal diagnosis
    - First degree relative
    - Positive genetic testing
    - Findings suggestive of on exam or lab findings
  - Abnormalities of cardiac or other viscera situs
  - Signs, symptoms, or blood culture suggestive of endocarditis
  - Known or suspected mass lesion involving the heart or great vessels
  - Known or suspected clot in atrium or ventricle
  - Known or suspected pulmonary hypertension
  - Known or suspected pericardial effusion
  - Complications during prenatal development:
    - Known or suspected cardiovascular abnormality on fetal echocardiogram
    - Maternal phenylketonuria (PKU)
    - Maternal diabetes with no fetal echo
    - Maternal teratogen exposure
    - Maternal infection during pregnancy with potential cardiac sequelae
  - Genetic abnormality known to be associated with cardiovascular disease such as:
    - Down syndrome
    - Turner syndrome
    - 22q11 deletion syndrome
    - Williams syndrome
    - Noonan syndrome
  - First-degree relative family history of:

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- Unexplained sudden death before age 50
- Hypertrophic cardiomyopathy
- Non-ischemic dilated cardiomyopathy
- Genetic abnormality known to be associated with cardiovascular disease
- Congenital left-sided heart lesion
- Heritable pulmonary arterial hypertension

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Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)

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- Repeat echocardiograms may be required for individuals with no new symptoms.
- In addition to indications listed in previous guideline sections, repeat TTE evaluation is considered medically necessary for any of the following:
  - In an individual with known cardiac disease and a previously normal echocardiogram when there is documentation of any of the following:
    - New or worsening cardiac symptoms
    - New EKG abnormality
    - Newly recognized family history suggestive of heritable heart disease
  - In an individual with prior normal evaluation
    - New or worsening symptoms
    - New EKG finding
    - New murmur
    - New finding of inheritable disease in first-degree relative
  - Individuals with first-degree family history of cardiomyopathy (such as, hypertrophic, dilated, arrhythmogenic) or aortopathy
    - Repeat echocardiogram every 12 months
    - Repeat echocardiogram can be done at the additional intervals when the family history or gene mutation is associated with neonatal or fetal disease:
      - At birth
      - Within the first 6 weeks
      - At 3 months
      - At 6 months
      - At one year
      - Then yearly
    - Repeat imaging is **not** medically necessary in individuals with first degree relative with known mutation when **both** of the following apply:
      - Individual has been tested and does not have that mutation
      - Individual has a normal echocardiogram
    - If there are abnormal findings on screening/surveillance imaging, a repeat echocardiogram is allowed to assess stability of findings.
  - Individual with a phenotype positive cardiomyopathy (with or without a positive gene) can be imaged as follows:

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- Infants (under one year)
  - TTE is considered medically necessary at frequency requested by pediatric cardiology or provider in consultation with pediatric cardiology.
- Children (over one year) yearly testing is considered medically necessary as follows:
  - Repeat TTE every 12 months
  - Repeat TTE to assess stability at discretion of pediatric cardiology, or provider in consultation with pediatric cardiology, after any new or changed clinical finding.
  - TTE at any time with documented new or changing symptoms.

◦ Individual with a known mutation associated with cardiomyopathy or aortopathy and no previous abnormal imaging
- Repeat echocardiogram every 12 months
  - Individuals whose gene mutation is associated with neonatal or fetal disease or there is a family history of neonatal or fetal disease can have repeat echocardiogram at the following intervals:
    - At birth
    - Within the first 6 weeks
    - Then at 3 months
    - At 6 months
    - At one year
    - Then yearly
- If there are abnormal findings on screening/surveillance imaging, a repeat echocardiogram is allowed to assess stability of findings.

◦ Individuals who are status post heart transplant can have echocardiograms repeated as often as requested by heart transplant team.

◦ Every 12 months for individuals receiving active therapy for ventricular hypertrophy, valvular dysfunction, cardiomyopathy
- One time repeat TTE can be approved at 6 months to assess response to a change in therapy

◦ Every 12 months for individuals with chronic pericardial effusions

◦ Every 12 months routine surveillance in asymptomatic individuals with muscular dystrophy (may be replaced by cardiac MRI CPT® 75557 or 75561 at 6 years of life)

◦ Every 12 months for sickle cell disease or other hemoglobinopathy causing chronic anemia and one of the following:
- High-risk genotype (Hgb SS or Sß^0, severe thalassemia, etc.)
- History of acute chest syndrome or intrinsic lung disease
- History of stroke

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    - Receiving chronic transfusion therapy
- As needed for monitoring cardiotoxicity during chemotherapy administration
- After completion of chemotherapy and/or radiation therapy. See **Cardiotoxicity and Echocardiography (PEDONC-19.2)** for imaging guidelines.
- Aortopathies- see **Thoracic Aortic Disease (PEDPVD-4.1)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines
- TTE follow-up for systemic hypertension
    - Individuals with evidence of end organ damage (Includes LVH, or decreased EF) can have echocardiogram every 6 months until echocardiogram normalizes.
    - Individuals without LV target organ injury (no LVH, normal EF) at initial echocardiographic assessment, repeat echocardiography at yearly intervals may be considered in those with persistent hypertension. (stage 2 HTN, or chronic stage 1 HTN incompletely treated (noncompliance or drug resistance)

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Transesophageal Echocardiography (TEE) (PEDCD-8.4)

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- Transesophageal echocardiography imaging indications in pediatric individuals are identical to those for adult individuals. See **Transesophageal Echocardiography (TEE)(CD-2.5)** in the Cardiac Imaging Guidelines.

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Computed Tomography

Guideline

CT Heart and Coronary Computed Tomography Angiography (CCTA) General Considerations (PEDCD-10.1)
Indications for CCTA (PEDCD-10.3)
Indications for Cardiac CT (CPT® 75572) (PEDCD-10.4)
Indications for Chest CTA with Cardiac CT or CTA (PEDCD-10.5)
References (PEDCD-10)

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CT Heart and Coronary Computed Tomography Angiography (CCTA) General Considerations (PEDCD-10.1)

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- Metal artifact reduces the accuracy of CCTA. Devices that can cause this issue include but are not limited to:
  - Surgical clips
  - Pacemaker devices
  - Defibrillator devices
  - Tissue expanders
- Cardiac testing that does not involve exposure to ionizing radiation should be strongly considered.

Radiation Dose (PEDCD-10.2)

- ACR–NASCI–SPR Practice Parameter for the Performance and Interpretation of Cardiac Computed Tomography (CT) stated, “Cardiac CT should be performed only for a valid medical indication and with the minimum radiation exposure that provides diagnostic image quality.”
- ACR–NASCI–SPR Practice Parameter for the Performance of Quantification of Cardiovascular Computed Tomography (Ct) And Magnetic Resonance Imaging (MRI) stated, “In younger patients, MRI may be the preferred modality, particularly when functional assessment with CT would require retrospective ECG gating and relatively high radiation doses. Further, the use of time-resolved MRA and phase contrast MRI methods offer significant advantages whose relative importance will depend on the specific application.”
  - See table: Practice Estimate of Effective Radiation Dose chart for Selected Imaging Studies in **General Guidelines (PEDCD-1.0)** in the general Cardiac Imaging Guidelines

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Indications for CCTA (PEDCD-10.3)

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- In addition to indications listed in previous guideline sections, CCTA is considered medically necessary for any of the following, when a recent TTE and/or MRI is inconclusive:
  - Persistent exertional chest pain and normal stress test
  - Full sibling(s) with history of sudden death syndrome before age 30 or with documented anomalous coronary artery
  - Resuscitated sudden death and contraindication to conventional coronary angiography
  - Unexplained new onset of heart failure if CCTA will replace conventional invasive coronary angiography
  - Documented ventricular tachycardia (6 beat runs or greater) if CCTA will replace conventional invasive coronary angiography
  - Equivocal coronary artery anatomy on conventional cardiac catheterization
  - In infants: otherwise unexplained dyspnea, tachypnea, wheezing, episodic pallor, irritability, sweating, poor feeding, and/or failure to thrive
    - The presence of other congenital heart disease is not a separate indication for CCTA to rule out anomalous coronary artery (except when coronary artery surgery is pending, i.e., Transposition of the great arteries, Tetralogy of Fallot, Truncus arteriosis, aortic root surgery)
  - Evaluation of the arterial supply and venous drainage in children with bronchopulmonary sequestration
- See **Coronary Anomalies (PEDCD-2.4.12)**

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Indications for Cardiac CT (CPT® 75572) (PEDCD-10.4)

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- In addition to indications listed in previous guideline sections, CCT is considered medically necessary for any of the following, when a recent TTE and/or MRI is inconclusive:
  - Cardiac or pericardial mass
  - Pericarditis
  - Complications of cardiac surgery or evaluation of post-operative anatomy
  - Cardiac thrombus in individuals with technically limited TTE, TEE, or MRI
  - Clinical suspicion of arrhythmogenic right ventricular dysplasia (ARVD) or arrhythmogenic cardiomyopathy (ARVC)
  - Native aortic abnormalities if echocardiogram is indeterminate
  - Intracardiac anatomy unclear after TTE or CMRI
  - A CTA Chest is medically necessary during a Cardiac CT if there are issues regarding the chest vessels that are inconclusive after echocardiogram or Cardiac MRI

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Indications for Chest CTA with Cardiac CT or CTA (PEDCD-10.5)

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- A Chest CTA may be medically necessary in individuals who require Cardiac CT or Cardiac CTA when:
  - A TTE or MRI is inconclusive for issues regarding chest vasculature
  - Routine imaging is considered medically necessary based on **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)**

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Magnetic Resonance Imaging

Guideline

Cardiac MRI Coding (PEDCD-9.2)
Indications for Cardiac MRI (PEDCD-9.3)
Indications for Chest MRA for Congenital Heart Disease (PEDCD-9.4)
References (PEDCD-9)

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Cardiac MRI Coding (PEDCD-9.2)

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Cardiac Imaging Procedure Codes

Cardiac MRI	CPT®
Cardiac magnetic resonance imaging for morphology and function without contrast.	75557
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences.	75561
Cardiac magnetic resonance imaging for morphology and function without contrast; with stress imaging (rarely used in pediatrics).	75559
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences; with stress imaging (rarely used in pediatrics).	75563
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure).	+75565

- Only one procedure code from the set: CPT® 75557, CPT® 75559, CPT® 75561, and CPT® 75563 should be reported per session.
- Only one flow velocity measurement (CPT® +75565) should be reported per session.

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Indications for Cardiac MRI (PEDCD-9.3)

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- In addition to indications listed in previous guideline sections, Cardiac MRI (CPT® 75557 or 75561) evaluation is considered medically necessary for any of the following, when a recent TTE is inconclusive:
  - Assessment of global ventricular function and mass if a specific clinical question is left unanswered by recent TTE and the MRI results will affect management of the individual's condition
  - Individuals with complex congenital heart disease (e.g., Tetralogy of Fallot [TOF], single ventricle, truncus arteriosis, Transposition of the Great Arteries [TGA]) may require a baseline MRI, or routine Cardiac MRI, especially as they approach their teenage years, due to poor imaging windows on echocardiogram, and the need for specific clinical information not seen on prior echocardiograms due to these known limitations. Once these individuals reach age 18, they can be imaged by adult congenital heart disease guideline.
  - Clinical suspicion of arrhythmogenic right ventricular dysplasia (ARVD) or arrhythmogenic cardiomyopathy (ARVC).
  - For pericardial disease (including constrictive pericarditis, restrictive pericarditis, and perimyocarditis), MRI should not be utilized to diagnose pericarditis but only to answer the question regarding possible constriction or restriction suggested clinically or by other techniques (TTE, etc.).
    - MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
  - Evaluate cardiac tumor or mass
    - MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
  - Evaluate anomalous coronary artery
    - MRI without and with contrast (CPT® 75561) or CCTA (CPT® 75574) after echocardiogram is considered the optimal test for this disorder.
  - For Fabry disease, late enhancement MRI may predict the effect of enzyme replacement therapy on myocardial changes that occur with this disease.
    - MRI without and with contrast (CPT® 75561) is considered the preferred test for this disorder.
  - Cardiac MRI (CPT® 75557 or 75561) can be performed to evaluate individuals with congenital cardiomyopathy (muscular dystrophy, glycogen storage disease, fatty acid oxidation disorders, mitochondrial disorders, etc.) or unexplained cases of cardiomyopathy in order to characterize the myocardium.

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- Cardiac stress perfusion study (CPT® 75559 or CPT® 75563) can be considered on a case by case basis for individuals with any of the following:
  - Anomalous coronary artery
  - Kawasaki disease
  - TGA
  - Ross operation
  - Other disorder with the potential for coronary ischemia
  - Individuals in whom an exercise stress test (EST) without imaging is considered medically necessary but the individual is not able to perform an EST
  - Individuals in whom an exercise stress test (EST) is equivocal, positive, or concern for a false negative
- Assessment of cardiac iron overload such as in hemochromatosis, thalassemia, sickle cell (either CPT® 75557 or CPT® 71550, T2* MRI, contrast not necessary)
  - Screening imaging may be approved every 12 months
  - Imaging may be approved every 3 months for treatment response in individuals receiving active treatment (chelation +/- phlebotomy)
  - Frequently performed along with MRI Abdomen (CPT® 74181) to assess liver iron deposition See **Transfusion-associated (Secondary) Hemochromatosis (PEDAB-18.2)** in the Pediatric Abdomen Imaging Guidelines.

- Asymptomatic individuals with Duchenne Muscular Dystrophy (DMD)
  - MRI Cardiac for DMD **either** CPT® 75557 or CPT® 75561 is considered medically necessary for surveillance if done in place of TTE every year starting at 6 years of age.
  - Female carriers would not typically be imaged until ≥18 years of age and should be imaged according to general Cardiac Imaging Guidelines.
  - CPT® 75565 or CPT® 71555 would not be medically necessary unless there was an independent indication for either of those codes.

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Indications for Chest MRA for Congenital Heart Disease (PEDCD-9.4)

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- For Familial Aortopathies, see **Thoracic Aortic Disease (PEDPVD-4.1)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines
- For individuals with known CHD for routine imaging **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)**
- For individuals who have both cardiac and ascending aorta abnormalities (e.g., truncus arteriosus), the following studies may be medically necessary following an inconclusive TTE:
  - Cardiac MRI (CPT® 75557 or CPT® 75561)
  - And MRI Chest (CPT® 71552) or MRA Chest (CPT® 71555) if aorta is involved
- For individuals with aortic abnormalities without cardiac abnormalities (i.e. normal intracardiac anatomy, but coarctation or peripheral pulmonary artery stenosis), the following studies may be medically necessary following an inconclusive TTE:
  - MRI Chest (CPT® 71552)
  - MRA Chest (CPT® 71555)
- MRA Chest (CPT® 71555) is considered medically necessary for individuals with cardiomyopathy or isolated abnormal intracardiac anatomy, when there are inconclusive images on echocardiogram related to chest vessels (e.g. aortic arch, pulmonary arteries, pulmonary veins, systemic veins).

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Diagnostic Heart Catheterization

Guideline

Cardiac Catheterization General Information (PEDCD-11.1)
Cardiac Catheterization Indications (PEDCD-11.2)
References (PEDCD-11)

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Cardiac Catheterization General Information (PEDCD-11.1)

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Cardiac Catheterization Procedure Codes

Cardiac Catheterization Procedures	CPT®
Congenital Heart Disease Code “Set”	93593-93597
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; normal native connections	93593
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; abnormal native connections	93594
Left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone, normal or abnormal native connections	93595
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); normal native connections	93596
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); abnormal native connections	93597
Anomalous coronary arteries, patent foramen ovale, mitral valve prolapse, and bicuspid aortic valve	93451-93464, 93566-93568
RHC without LHC or coronaries	93451
LHC without RHC or coronaries	93452

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Cardiac Catheterization Procedures	CPT®
RHC and retrograde LHC without coronaries	93453
Native coronary artery catheterization;	93454
with bypass grafts	93455
with RHC	93456
with RHC and bypass grafts	93457
with LHC	93458
with LHC and bypass grafts	93459
with RHC and LHC	93460
with RHC and LHC and bypass grafts	93461
LHC by trans-septal or apical puncture	+93462
Angiography of noncoronary arteries and veins performed as a distinct service	Select appropriate codes from the Radiology and Vascular Injection Procedures sections.
CPT® 93593 to 93597 are appropriate for invasive evaluation of congenital heart disease	

- These guidelines apply to individuals with stable conditions and who are not in the acute setting. Individuals in acute settings or with unstable angina should be handled as medical emergencies.
- Pediatric catheterization is done for many purposes, including diagnosis and intervention of congenital and acquired heart disease.

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- When device placement is planned (ASD/VSD device, transcatheter valve implantation, PDA device), the procedure codes for those devices include all cardiac catheterization(s), intraprocedural contrast injection(s), fluoroscopic radiological supervision and interpretation, and imaging guidance performed to complete the procedure. A diagnostic catheterization may be considered on a case-by-case basis if there are unanswered issues via noninvasive imaging.
- A right heart catheterization can be approved for pulmonary artery interventions (e.g., stents, coils).

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Cardiac Catheterization Indications (PEDCD-11.2)

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- Diagnostic catheterization is considered medically necessary:
  - When other advanced imaging has failed to resolve a clinical issue and results will impact the individual's management
    - For example, a catheterization to assess ventricular pressures and shunt to determine if VSD surgery is required
  - For preoperative assessment in complex heart disease
    - Norwood procedure
    - Bidirectional Glenn shunt
    - Fontan procedure
    - Pulmonary atresia
  - Pulmonary hypertension
  - During some interventions such as:
    - Valvuloplasty
    - Pulmonary artery or vein stents
  - Assessment of individuals who are status post heart transplant
  - See **Kawasaki Disease Initial Imaging (PEDCD-6.1)** for specific intervals in Kawasaki Disease
  - On an individual who is having a device placed when:
    - A diagnostic catheterization, or stenting is needed in addition to the device
    - The diagnostic catheterization is considered medically necessary separate from the device placement
  - Individuals with anomalous coronary arteries, or with syndromes associated with abnormal coronary arteries (i.e., Williams syndrome) or acquired CAD (i.e., KD see **Kawasaki Disease Initial Imaging (PEDCD-6.1)**
    - When diagnostic images are not adequate or evaluation or treatment decision
    - Preoperative for cardiac surgery
    - New symptoms concerning for ischemia

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General Guidelines (PEDCH-1)

Guideline

Procedure Codes Associated with Chest Imaging
General Guidelines (PEDCH-1.0)
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Procedure Codes Associated with Chest Imaging

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MRI	CPT®
MRI Chest without contrast	71550
MRI Chest with contrast (rarely used)	71551
MRI Chest without and with contrast	71552
Unlisted MRI procedure (for radiation planning or surgical software)	76498

MRA	CPT®
MRA Chest (non-cardiac)	71555

CT	CPT®
CT Chest without contrast	71250
CT Chest with contrast	71260
CT Chest without and with contrast (rarely used)	71270
CT Guidance for Placement of Radiation Therapy Fields	77014
Unlisted CT procedure (for radiation planning or surgical software)	76497

CTA	CPT®
CTA Chest (non-coronary)	71275

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Nuclear Medicine	CPT®
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging: skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging: whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
Pulmonary Ventilation (e.g., Aerosol or Gas) Imaging	78579
Pulmonary Perfusion Imaging	78580
Pulmonary Ventilation (e.g., Aerosol or Gas) and Perfusion Imaging	78582
Quantitative Differential Pulmonary Perfusion, Including Imaging When Performed	78597
Quantitative Differential Pulmonary Perfusion and Ventilation (e.g., Aerosol or Gas), Including Imaging When Performed	78598

Ultrasound	CPT®
Ultrasound, chest (includes mediastinum, chest wall, and upper back)	76604
Ultrasound, axilla	76882
Ultrasound, breast; *unilateral*, including axilla when performed; complete	76641
Ultrasound, breast; *unilateral*, including axilla when performed; limited	76642

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General Guidelines (PEDCH-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms, including a detailed history, physical examination, and appropriate laboratory, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the chest is not supported. Advanced imaging of the chest is only supported in individuals who have documented active clinical signs or symptoms of disease involving the chest.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the chest are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Pediatric Chest Imaging Age Considerations (PEDCH-1.1)

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- Many conditions affecting the chest in the pediatric population have different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are 18 years old or younger should be imaged according to the Pediatric Chest Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Chest Imaging Guidelines should be imaged according to the General Chest Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Chest Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Pediatric Chest Imaging Modality General Considerations (PEDCH-1.3)

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- MRI
  - MRI Chest is generally performed without and with contrast (CPT® 71552) unless the individual has a documented contraindication to gadolinium or otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize individual movement, anesthesia is usually medically necessary for almost all infants (except neonate) and young individuals (age <7 years), as well as older individuals with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition-based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast is medically necessary if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
  - If multiple body areas are supported by the guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently.
  - The presence of surgical hardware or implanted devices may preclude MRI.
  - The selection of best examination may require coordination between the provider and the imaging service.
- CT

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  - CT Chest is generally performed either with contrast (CPT® 71260) or without contrast (CPT® 71250).
    - There are no generally accepted pediatric indications for CT Chest without and with contrast (CPT® 71270).
  - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
  - The selection of best examination may require coordination between the provider and the imaging service.
- Ultrasound
  - Ultrasound Chest (CPT® 76604) or Axilla (CPT® 76882) is medically necessary as an initial study for evaluating adenopathy, palpable chest wall lesions, pleural effusion or thickening, patency of thoracic vasculature, and diaphragm motion abnormalities.
  - For those individuals who do require advanced imaging, ultrasound can be very beneficial in selecting the proper modality, body area, image sequences, and contrast level that will provide the most definitive information for the individual.
- Nuclear Medicine
  - Nuclear medicine studies other than PET/CT are rarely used in evaluation of the pediatric chest.
  - Pulmonary Ventilation-Perfusion Imaging (CPT® 78582) has been replaced by CTA Chest (CPT® 71275) or CT Chest with contrast (CPT® 71260), but is medically necessary for evaluation of suspected pulmonary embolism if CT is unavailable.
    - See **Pulmonary Embolism (PE) (CH-25.1)** in the Chest Imaging Guidelines.
  - Pulmonary Perfusion Imaging (CPT® 78580) is not medically necessary in lieu of CPT® 78582 for initial evaluation of suspected pulmonary embolism, but is medically necessary for follow-up of an equivocal or positive ventilation-perfusion lung scan (CPT® 78582) to evaluate for interval change.
  - Pulmonary Ventilation Imaging (CPT® 78579) is not medically necessary in lieu of CPT® 78582 for evaluation of suspected pulmonary embolism, but is medically necessary for additional evaluation of an abnormal perfusion-only scan (CPT® 78580).
  - Pulmonary split crystal function study (CPT® 78597 or CPT® 78598), also known as Quantitative Differential Pulmonary Perfusion, is medically necessary for preoperative planning of segmental, lobar, or lung resection.
  - Quantitative Differential Pulmonary Perfusion Lung Scan (CPT® 78597 or CPT® 78598), can be performed post-lung transplant to detect regional perfusion abnormalities.
  - Radiopharmaceutical nuclear medicine imaging of an inflammatory process (CPT® 78800, CPT® 78801, CPT® 78802, or CPT® 78803) is rarely performed, but is medically necessary for evaluation of sarcoidosis or toxicity from drug toxicity (cyclophosphamide, busulfan, bleomycin, amiodarone, or nitrofurantoin).

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- 3D Rendering
  - 3D Rendering indications in pediatric chest imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive. Clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Lymphadenopathy (PEDCH-2)

Guideline

Lymphadenopathy (PEDCH-2.1)
Reference (PEDCH-2)

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Lymphadenopathy (PEDCH-2.1)

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- Axillary lymphadenopathy imaging indications in pediatric individuals are identical to those for adult individuals. See **Axillary Lymphadenopathy (and Mass) (CH-2.2)** in the Chest Imaging Guidelines.
- Supraclavicular adenopathy in pediatric individuals is almost always pathologic, and advanced imaging is medically necessary prior to excisional biopsy. Fine-needle aspiration, while common in adults prior to advanced imaging, is not medically necessary for evaluating lymphadenopathy in pediatric individuals. ANY combination of the following studies are medically necessary for evaluation of supraclavicular adenopathy:
  - CT Chest with contrast (CPT® 71260)
  - MRI Chest without and with contrast (CPT® 71552)
  - Ultrasound Chest (CPT® 76604)
- If malignancy is suspected, refer to the condition specific imaging guidelines as below:
  - Lymphoma: **Pediatric Lymphomas (PEDONC-5)** in the Pediatric Oncology Imaging Guidelines.
  - Soft tissue sarcoma: **Pediatric Soft Tissue Sarcomas (PEDONC-8)** in the Pediatric Oncology Imaging Guidelines.
  - Neuroblastoma: **Neuroblastoma (PEDONC-6)** in the Pediatric Oncology Imaging Guidelines.

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Mediastinal Mass (PEDCH-3)

Guideline

Mediastinal Mass (PEDCH-3.1)
References (PEDCH-3)

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Mediastinal Mass (PEDCH-3.1)

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- The causes of mediastinal masses in pediatric individuals are generally different than those in adults, and therefore imaging considerations are different. Up to half of all pediatric mediastinal masses are malignant.
- Chest x-ray is medically necessary as an initial study for all individuals with suspected mediastinal mass.
- CT Chest with contrast (CPT® 71260) is medically necessary for any mediastinal mass identified on chest x-ray.
  - Masses can be very large and anterior masses frequently cause compression of the trachea and/or mediastinal blood vessels.
- MRI Chest without and with contrast (CPT® 71552) is medically necessary to evaluate any of the following findings:
  - a posterior (paravertebral) mediastinal mass on CT Chest that invades the spinal canal
  - CT findings that are inconclusive regarding specific anatomy
  - Large anterior mediastinal masses only whenanesthesia is not used to complete the study.
- PET/CT (CPT® 78815) is medically necessary prior to biopsy in pediatric individuals if lymphoma is known or strongly suspected or there is evidence of tracheal compression on CT imaging. See **Pediatric Lymphoma (PEDONC-5)** in the Pediatric Oncology Imaging Guidelines.
- MIBG (CPT® 78800, CPT® 78802, CPT® 78803, or CPT® 78804) is medically necessary prior to biopsy in pediatric individuals if neuroblastoma is known or strongly suspected. See **Neuroblastoma (PEDONC-6)** in the Pediatric Oncology Imaging Guidelines.
- Ultrasound chest (CPT® 76604) is medically necessary in individuals younger than 5 years old to distinguish prominent but otherwise normal thymus from true mediastinal mass.
- A single repeat CT Chest with contrast (CPT® 71260) is medically necessary to confirm stability and avoid biopsy when NONE of the following features are present:
  - anterior mediastinal mass
  - enlarged lymph nodes anywhere in the imaging field
  - lymphopenia
  - pleural effusion

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Hemoptysis (PEDCH-4)

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Hemoptysis – Imaging (PEDCH-4.1)
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Hemoptysis – Imaging (PEDCH-4.1)

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- True hemoptysis is rare in pediatric individuals. A detailed history, physical examination, and appropriate laboratory studies should be performed prior to considering advanced imaging.
  - Aspirated blood from epistaxis or emesis frequently presents as hemoptysis, and history and physical examination will aid in this assessment.
- Chest x-ray is medically necessary as an initial study for stable individuals.
  - Advanced imaging is not medically necessary to evaluate individuals with epistaxis when there is a normal chest radiograph and no personal or family history of underlying lung disease or bleeding disorder.
  - CT Chest with contrast (CPT® 71260) or CTA Chest (CPT® 71275) is medically necessary for all other pediatric individuals with true hemoptysis.
    - CT Chest without contrast (CPT® 71250) is medically necessary for individuals with a documented allergy to CT contrast or significant renal dysfunction.
- MRI is not medically necessary in the evaluation of pediatric hemoptysis.

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Cystic Fibrosis and Bronchiectasis (PEDCH-5)

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Cystic Fibrosis (PEDCH-5.1)
Bronchiectasis Not Associated with Cystic Fibrosis (PEDCH-5.2)
References (PEDCH-5)

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Cystic Fibrosis (PEDCH-5.1)

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- Chest x-ray is medically necessary for initial evaluation of acute clinical symptoms in individuals with cystic fibrosis.
- CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is medically necessary in the setting of any of the following (chest x-ray not required):
  - hemoptysis
  - pneumonia worsening despite antibiotic therapy
  - pleural effusion or empyema
  - suspected fungal pneumonia
  - monitoring treatment changes on bronchiectasis
  - expiratory CT for evaluating small airways disease
  - pre- and post-lung transplant evaluation
  - uncooperative individual
- Low dose CT Chest without contrast (CPT® 71250) is medically necessary **every 2 years** for monitoring of bronchiectasis and small airways disease.
- Cystic fibrosis associated liver disease develops in 5-10% of individuals with cystic fibrosis. Advanced imaging may be medically necessary if concerned for liver disease. See **Liver Disease (PEDAB-16)** in the Pediatric Abdomen Imaging Guidelines.

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Bronchiectasis Not Associated with Cystic Fibrosis (PEDCH-5.2)

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- Bronchiectasis not associated with cystic fibrosis is rare in pediatric individuals, and imaging indications are identical to those for adult individuals. See **Bronchiectasis (CH-7.1)** in the Chest Imaging Guidelines.

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Bronchiolitis (PEDCH-6)

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Bronchiolitis (PEDCH-6.1)
References (PEDCH-6)

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Bronchiolitis (PEDCH-6.1)

CHP.BR.0006.1.A
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Bronchiolitis is a self-limiting viral infection causing inflammation of the small airways, most common in infants under 12 months of age.

- Chest x-ray or chest ultrasound (CPT®76604) is medically necessary when there is a clinical suspicion of pneumonia or other complications.
- Advanced imaging is not medically necessary for routine evaluation or monitoring of bronchiolitis.
- CT Chest with contrast (CPT® 71260) is medically necessary for the following:
  - pleural effusion or empyema on relevant chest x-ray or chest ultrasound (CPT®76604)
  - immunocompromised individual with acute pulmonary symptoms
  - abnormality on relevant chest x-ray suggesting condition other than bronchiolitis

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Pneumonia (PEDCH-7)

Guideline

Pneumonia (PEDCH-7.1)
Coronavirus Disease 2019 (COVID-19) (PEDCH-7.2)
References (PEDCH-7)

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Pneumonia (PEDCH-7.1)

CHP.PN.0007.1.A

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- Pneumonia imaging indications in pediatric individuals are very similar to those for adult individuals. See **Pneumonia (CH-13.1)** in the Chest Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - Chest x-ray and/or Ultrasound chest (CPT® 76604) is medically necessary when the individual's condition does not respond to standard therapy.
  - CT Chest with contrast (CPT® 71260) is medically necessary for:
    - immunocompromised individuals with acute pulmonary symptoms
    - diagnosis of an abscess
    - necrotizing pneumonia cannot be confirmed
    - recurring pneumonia
    - acute complications
  - CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is medically necessary for individuals with recurrent lower respiratory tract infections.
  - Ultrasound chest (CPT® 76604) is medically necessary as an alternative for evaluation of complicated or recurrent childhood pneumonia.

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Coronavirus Disease 2019 (COVID-19) (PEDCH-7.2)

CHP.PN.0007.2.A

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- Pediatric imaging for COVID-19 positive individuals are similar to those for adult individuals. See **Coronavirus Disease 2019 (COVID-19) (CH-13.2)** in the Chest Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - chest x-ray is the initial imaging test for all pediatric individuals
  - for concerns involving **Multisystem Inflammatory Syndrome in Children (MIS-C)** see **(PEDCD-12)**

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Solitary Pulmonary Nodule (PEDCH-8)

Guideline

Solitary Pulmonary Nodule (PEDCH-8.1)
References (PEDCH-8)

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Solitary Pulmonary Nodule (PEDCH-8.1)

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The Fleischner Society guidelines for solitary pulmonary nodule management do not apply to pediatric individuals. An incidental solitary pulmonary nodule in a individual representing a primary lung carcinoma has never been reported in the literature. Similarly, an extrathoracic malignancy presenting with an incidental solitary pulmonary nodule in an otherwise healthy individual is very rare.

- CT Chest with contrast (CPT® 71260) is medically necessary as a one-time evaluation of a pulmonary nodule incidentally discovered on other imaging.
- Follow-up imaging of an incidental solitary pulmonary nodule in an asymptomatic healthy individual is **not** medically necessary.
  - Follow-up imaging is medically necessary for the following:
    - immunocompromised individuals
    - malignancy (see below)
    - invasive infection
    - new or worsening pulmonary symptoms
- Individuals with a malignant solid tumor who have pulmonary nodules of any size should have imaging according to the guideline section for the specific cancer type. See **Pediatric Oncology Imaging Guidelines** for specific imaging indications.
- This guideline section does not apply to multiple pulmonary nodules, which are imaged according to the underlying disorder in pediatric individuals.

Background and Supporting Information

A **nodule** is any pulmonary or pleural lesion that is a discrete, spherical opacity 2-30 mm in diameter surrounded by normal lung tissue. A larger nodule is called a mass. Entities that are not nodules, and are considered benign, include non-spherical linear, sheet-like, two-dimensional or scarring opacities.

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Positive PPD or Tuberculosis (PEDCH-9)

Guideline

Positive PPD or Tuberculosis (PEDCH-9.1)
References (PEDCH-9)

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Positive PPD or Tuberculosis (PEDCH-9.1)

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- Positive PPD and tuberculosis imaging indications in pediatric individuals are similar to those for adult individuals.
  - See **PPD or TB (Mycobacterium tuberculosis and Nontuberculous Mycobacterial Pulmonary Disease [NTM-PD]) (CH-14.1)** in the Chest Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - MRI Spine with and without contrast of the symptomatic spine level is medically necessary in individuals with concern for spinal involvement of tuberculosis.
  - MRI Chest with and without contrast (CPT® 71552) can be considered as a radiation-free alternative to chest CT in pediatric individuals.

Background and Supporting Information

- Chest x-ray can be useful as the initial imaging study when TB is suspected.

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Asthma (PEDCH-10)

Guideline

Asthma (PEDCH-10.1)
References (PEDCH-10)

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Asthma (PEDCH-10.1)

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- Chest x-ray and/or Ultrasound chest (CPT® 76604) is medically necessary to further evaluate respiratory distress when the individual's condition does not respond to standard therapy.
- Advanced imaging is not medically necessary for routine evaluation or monitoring of asthma.
- CT Chest without (CPT® 71250) or with (CPT® 71260) contrast is medically necessary for the following:
  - pleural effusion or empyema on relevant chest x-ray
  - immunocompromised individual with acute pulmonary symptoms
  - abnormality on relevant chest x-ray suggesting condition other than asthma, including suspected foreign body
  - asthma and poor response to bronchodilators or conventional inhaled corticosteroid therapy in whom associated conditions, such as allergic bronchopulmonary aspergillosis and eosinophilic pneumonia, can mimic asthma

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Pectus Deformities (PEDCH-11)

Guideline

Pectus Deformities (PEDCH-11.1)
References (PEDCH-11)

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Pectus Deformities (PEDCH-11.1)

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- CT Chest without contrast (CPT® 71250), MRI Chest with and without contrast (CPT® 71552), or MRI Chest without contrast (CPT® 71550) is medically necessary in individuals with a pectus deformity for any of the following:
  - preoperative planning
  - significant cardiac depression after chest x-ray and echocardiography (CPT® 93306)
  - evidence of pulmonary impingement after chest x-ray and pulmonary function tests (PFTs) if there is increasing shortness of breath
    - Note: It may not be possible to obtain PFTs in individuals younger than 9 years old.
  - evaluation of congenital heart disease or Marfan's syndrome when suspected

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Breast Masses (PEDCH-12)

Guideline

Breast Masses (PEDCH-12.1)

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Breast Masses (PEDCH-12.1)

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See **Pediatric Breast Masses (PEDONC-17)** in the Pediatric Oncology Imaging Guidelines.

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Vascular Malformations (PEDCH-13)

Guideline

Vascular Ring (PEDCH-13.1)
Other Vascular Malformations (PEDCH-13.2)
References (PEDCH-13)

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Vascular Ring (PEDCH-13.1)

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- Chest x-ray is the recommended initial study in individuals with respiratory symptoms. A chest x-ray is not medically necessary for individuals diagnosed with a vascular ring on prenatal imaging studies.
- Barium esophagram is the recommended initial study in individuals with feeding difficulties.
- CT Chest with contrast (CPT® 71260), CTA Chest (CPT® 71275), MRA Chest (CPT® 71555), or MRI Chest without contrast (CPT® 71550) in individuals with known or suspected vascular ring after prenatal imaging studies, chest x-ray, or barium esophagram.
- Echocardiogram is medically necessary to rule out associated congenital heart disease.
  - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325 are medically necessary for initial evaluation of individuals with vascular ring and no prior echocardiograms.

Background and Supporting Information

Vascular rings generally present with either respiratory symptoms (stridor, wheezing, tachypnea, cough) or feeding difficulties (dysphagia, slow feeding, hyperextension of the head while feeding, weight loss, failure to thrive), but can also be discovered incidentally on imaging obtained for other purposes.

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Other Vascular Malformations (PEDCH-13.2)

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See **Pulmonary Arteriovenous Malformations (PEDCH-14.2)** for Pulmonary AVMs.

See **Vascular Anomalies (PEDPVD-2)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines.

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Congenital Chest Diseases (PEDCH-14)

Guideline

Congenital Cystic Lung Diseases (PEDCH-14.1)
Pulmonary Arteriovenous Malformations (PEDCH-14.2)
Congenital Diaphragmatic Hernia (PEDCH-14.3)
Pulmonary Sequestration (PEDCH-14.4)
References (PEDCH-14)

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Pulmonary Sequestration (PEDCH-14.4)

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- Pulmonary sequestration imaging indications in pediatric individuals are similar to those for adult individuals.
  - See **Pleural-Based Nodules and Other Abnormalities (CH-17.1)** in the Chest Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - CTA Chest (CPT® 71275) is the gold standard for diagnosing pulmonary lung abnormalities.
  - CT Chest with contrast (CPT® 71260) and MRI Chest without contrast (CPT® 71550) are alternative imaging modalities that can be used for diagnosis.

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Congenital Cystic Lung Diseases (PEDCH-14.1)

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- This section includes common congenital cystic lung lesions such as:
  - bronchogenic cyst
  - congenital pulmonary airway malformation (congenital cystic adenomatoid malformation)
  - congenital lobar overinflation
- CT Chest with contrast (CPT® 71260) is medically necessary when a cystic lung lesion is suspected.
- MRI Chest with and without contrast (CPT® 71552) is medically necessary if CT is inconclusive or if requested for pre-operative planning.

Background and Supporting Information

- Cystic lung disease may be first identified on prenatal ultrasound, or discovered incidentally on chest x-ray.

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Pulmonary Arteriovenous Malformations (PEDCH-14.2)

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- Pulmonary arteriovenous malformations (PAVMs) are vascular structures that most commonly result from abnormal communication between pulmonary arteries and pulmonary veins.
  - Chest x-ray is medically necessary as an initial imaging modality for individuals with known AVMs, or individuals presenting with hypoxemia and/or hemoptysis.
  - CTA or MRA is medically necessary in individuals with known AVM or abnormal chest x-ray suggesting AVM for treatment planning.

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Congenital Diaphragmatic Hernia (PEDCH-14.3)

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- Congenital Diaphragmatic hernia (CDH) is a defect in the diaphragm which may allow the abdominal organs to enter the chest cavity, and may lead to compromised pulmonary function or may be associated with congenital heart disease.
  - Over 90% of the hernias occur in the posterolateral diaphragm (Bochdalek hernia) typically on the left side.
  - Most of the rest of the hernias are in the anteromedial diaphragm (Morgagni hernia).
- The vast majority of CDH are diagnosed prenatally (see **Fetal MRI [PV-15.1]**) in the Pelvis Imaging Guidelines), or as an inpatient shortly after delivery.
- If there is clinical concern for CDH, chest x-ray and/or US Chest (CPT® 76604) is medically necessary as the initial imaging study.
- CT Chest with contrast (CPT® 71260) or MRI Chest with and without contrast (CPT® 71552) is medically necessary when Chest x-ray and/or US are inconclusive, or if requested for treatment planning.

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General Guidelines (PEDHD-1)

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Procedure Codes Associated with Head Imaging
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Procedure Codes Associated with Head Imaging

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Procedure Codes Associated with Head Imaging	
MRI	**CPT®**
MRI Brain without contrast	70551
MRI Brain with contrast (rarely used)	70552
MRI Brain without and with contrast	70553
MRI Orbit, Face, Neck without contrast	70540
MRI Orbit, Face, Neck with contrast (rarely used)	70542
MRI Orbit, Face, Neck without and with contrast	70543
MRI Temporomandibular Joint (TMJ)	70336
Functional MRI Brain not requiring physician or psychologist	70554
Functional MRI Brain requiring physician or psychologist	70555
MR Spectroscopy	76390
Unlisted MRI procedure (for radiation planning or surgical software)	76498
MRA	**CPT®**
MRA Head without contrast	70544
MRA Head with contrast	70545
MRA Head without and with contrast	70546

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Procedure Codes Associated with Head Imaging	
MRA Neck without contrast	70547
MRA Neck with contrast	70548
MRA Neck without and with contrast	70549
CT	**CPT®**
CT Head without contrast	70450
CT Head with contrast	70460
CT Head without and with contrast	70470
CT Orbits without contrast (includes temporal bone and mastoid)	70480
CT Orbits with contrast (includes temporal bone and mastoid)	70481
CT Orbits without and with contrast (includes temporal bone and mastoid)	70482
CT Maxillofacial without contrast (includes sinuses, jaw, and mandible)	70486
CT Maxillofacial with contrast (includes sinuses, jaw, and mandible)	70487
CT Maxillofacial without and with contrast (includes sinuses, jaw, and mandible)	70488
CT Neck without contrast (includes jaw, and mandible)	70490
CT Neck with contrast (includes jaw, and mandible)	70491
CT Neck without and with contrast (includes jaw, and mandible)	70492
CT Guidance for Stereotactic Localization (used for sinus surgery planning)	77011
CT Guidance for Placement of Radiation Therapy Fields	77014

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Procedure Codes Associated with Head Imaging	
Unlisted CT procedure (for radiation planning or surgical software)	76497
CTA	**CPT®**
CTA Head	70496
CTA Neck	70498

Nuclear Medicine	CPT®
PET Brain Metabolic Evaluation	78608
PET Brain Perfusion Evaluation	78609
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; whole body	78816
Brain Scintigraphy Static Limited	78600
Brain Scintigraphy Limited with Vascular Flow	78601
Brain Scintigraphy Complete Static	78605
Brain Scintigraphy Complete with Vascular Flow	78606
Brain Imaging Vascular Flow	78610
Cisternogram	78630
Cerebrospinal Ventriculography	78635
Shunt Evaluation	78645
CSF Leakage Detection	78650

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Radiopharmaceutical Dacryocystography	78660
Ultrasound	**CPT®**
Echoencephalography (Head or Cranial Ultrasound)	76506
Ophthalmic ultrasound, diagnostic; B-scan & quantitative A-scan performed same encounter	76510
Ophthalmic ultrasound, diagnostic; quantitative A-scan only	76511
Ophthalmic ultrasound, diagnostic; B-scan	76512
Ophthalmic ultrasound, diagnostic; anterior segment ultrasound, immersion (water bath) B-scan	76513
Ophthalmic ultrasound, diagnostic; corneal pachymetry, unilateral or bilateral	76514
Ophthalmic biometry by ultrasound, A-scan	76516
Ophthalmic biometry by ultrasound, A-scan, with lens power calculation	76519
Ophthalmic ultrasonic foreign body localization	76529
Soft tissues of head and neck Ultrasound (thyroid, parathyroid, parotid, etc.)	76536
Transcranial Doppler study of the intracranial arteries; complete study	93886
Transcranial Doppler study of the intracranial arteries; limited study	93888

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General Guidelines (PEDHD-1.0)

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- A pertinent clinical evaluation including a detailed history, physical examination with a thorough neurologic examination since the onset or change in signs and/or symptoms, appropriate laboratory studies and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the individual is undergoing a guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail, or messaging) since the onset or change in signs and/or symptoms, can serve as a pertinent clinical evaluation.
  - A detailed neurological exam is required prior to advanced imaging except in the following scenarios:
    - Individual is undergoing a guideline-supported scheduled follow-up imaging evaluation
    - Tinnitus, TMJ, sinus or mastoid disease, ear pain, hearing loss, eye disease, papilledema, dental requests and epistaxis. (A relevant physical exam is still required.)
    - The request is from a neurologist, neurosurgeon, endocrinologist, otolaryngologist, or ophthalmologist who has evaluated the individual since onset of symptoms.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the head is not medically necessary. Advanced imaging of the head is only medically necessary in individuals who have documented active clinical signs or symptoms of disease involving the head.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the head are not medically necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.

Health Equity Consideration

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence;

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education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Pediatric Head Imaging Age Considerations (PEDHD-1.1)

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- Many conditions affecting the head in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are 18 years old or younger and any conditions not specifically discussed in the General Head Imaging Guidelines should be imaged according to the Pediatric Head Imaging Guidelines. Any conditions not specifically discussed in the Pediatric Head Imaging Guidelines should be imaged according to the General Head Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Head Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Pediatric Head Imaging Appropriate Clinical Evaluation (PEDHD-1.2)

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Requests for Studies with Overlapping Fields

- There are many CPT® codes for imaging the head that have significantly overlapping fields. In the majority of cases where multiple head CPT® codes are requested, only one CPT® code is appropriate unless there is clear documentation of a need for the additional codes to cover all necessary body areas.
- See **General Guidelines - Anatomic Issues (HD-1.1)** in the Head Imaging Guidelines for the correct coding of these studies.

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Pediatric Head Imaging Modality General Considerations (PEDHD-1.3)

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- MRI
  - MRI is superior to CT imaging of the pediatric head unless otherwise stated in a specific guideline section.
  - Anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with delays in development or maturity.
  - MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI/MRA of all necessary body areas, when multiple body areas are supported by the guidelines for the clinical condition being evaluated, should be obtained concurrently in the same anesthesia session.
    - MRI without and with contrast is medically necessary if requested to avoid repetitive anesthesia administration that would be needed to perform an MRI with contrast, if the initial study without contrast is inconclusive when:
      - intravenous access will already be present for anesthesia administration **AND**
      - there is no contraindication for using contrast.
  - For MRI spine imaging with contrast to be performed following a contrast-enhanced MRI Brain, see **CNS Tumors General Considerations (PEDONC-4.1)** in the Pediatric and Special Populations Oncology Guidelines.
- CT
  - CT is generally inferior to MRI for imaging of the pediatric head, but has specific indications in which it is the preferred modality listed in specific sections of these guidelines.
    - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
  - CT Head without contrast (CPT® 70450) may be medically necessary for:
    - Mass effect
    - Blood/blood products
    - Urgent/emergent settings due to availability and speed of CT
    - Trauma
    - Recent hemorrhage, whether traumatic or spontaneous
    - Bony structures of the head evaluations including dystrophic calcifications
    - Hydrocephalus evaluation and follow-up

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  - Some centers use limited non-contrast “fast or rapid MRI” (CPT® 70551) to minimize radiation exposure in children - these requests are appropriate.
- Prior to lumbar puncture in individuals with cranial complaints
- Scenarios in which MRI is contraindicated (i.e., pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)
- Concern for calcifications

- CT and MR Angiography (CTA and MRA) Head and Neck
  - Contrast level
    - MRA Head may be performed without contrast (CPT® 70544), with contrast (CPT® 70545), or without and with contrast (CPT® 70546).
    - CTA Head is performed without and with contrast (CPT® 70496).
    - MRA Neck may be done either without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549), depending on facility preference and protocols and type of scanner.
    - CTA Neck is done without and with contrast (CPT® 70498).
  - Indications for CTA and MRA Head and Neck vessels include, but are not limited to the following:
    - Pulsatile tinnitus
    - Hemifacial spasm if consideration for surgical decompression
    - Evaluation of stroke or TIA (See **Pediatric Stroke Initial Imaging (PEDHD-12.2)**, **Pediatric Stroke Subsequent Imaging (PEDHD-12.3)**, **Moyamoya Disease (PEDHD-12.4)**, **Sickle Cell Disease (PEDHD-12.5),** and **CNS Vasculitis (PEDHD-12.6)** including collateral assessment)
    - Follow-up of known cerebral artery stenosis
    - Trigeminal neuralgia that has failed medical therapy
    - Cerebral sinus thrombosis suspected with increased intracranial pressure (refractory headaches, papilledema, diagnosis of pseudotumor cerebri)
    - Aneurysm suspected with acute "thunderclap" headache syndrome and appropriate screening or evaluation of known subarachnoid hemorrhage and pseudoaneurysms (may be appropriate to limit CTA to include only the head to avoid unnecessary radiation to the individual) (See **Pediatric Intracranial Aneurysms (PEDHD-10.1)**)
    - Noninflammatory vasculopathy, including radiation vasculopathy (See **Long Term Pediatric Cancer Survivors (PEDONC-19)** in the Pediatric and Special Populations Oncology Imaging Guidelines)
    - Traumatic vascular injuries
    - Vascular malformations, vascular anatomic variants, and fistulas (See **Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2)**)
    - Arterial, including carotid dissections
    - Tumors of vascular origin or involving vascular structures

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        - Surgical and radiation therapy localization, planning and neuronavigation
        - Evaluation for vascular intervention and follow-up including post-surgical/post-treatment vascular complications
        - Intra-cranial pre-operative planning if there is concern of possible vascular involvement or risk for vascular complication from procedure
        - Vasculitis and collagen vascular disease (See **CNS Vasculitis (PEDHD-12.6)**)
        - Sickle cell disease (See **Sickle Cell Disease (PEDHD-12.5)**)
        - Moyamoya disease (See **Moyamoya Disease (PEDHD-12.4)**)
        - Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebral-basilar system
    - For follow-up of aneurysm clipping or coiling procedures, see **Intracranial Aneurysms (HD-12.1)** in the Head Imaging Guidelines.
- CT and MR Venography (CTV and MRV)
    - CTV and MRV are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography).
    - If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures.
    - MRA without and with contrast with venous sinus thrombosis to differentiate total from subtotal occlusion
- Ultrasound
    - Cranial ultrasound (CPT® 76506) is a non-invasive means of evaluating for intracranial abnormalities in infants with an open anterior fontanelle.
    - Transcranial Doppler ultrasonography has some utility in select populations of older children with known or suspected intracranial vascular disease.
- Nuclear Medicine
    - Nuclear medicine studies other than metabolic PET imaging on the pediatric brain or head are rarely performed in an elective outpatient setting, but the following studies are medically necessary for the following indications:
        - Brain Scintigraphy with or without vascular flow (any one of CPT® codes: CPT® 78600, CPT® 78601, CPT® 78605, or CPT® 78606)
            - Establish brain death
        - Radiopharmaceutical Localization Imaging SPECT (CPT® 78803)
            - Immunocompromised individuals with mass lesion detected on CT or MRI for differentiation between lymphoma and infection
            - Radiopharmaceutical Localization Imaging SPECT (CPT® 78803) with vasodilating agent acetazolamide (Diamox) challenge when surgery or other vascular intervention is being considered (i.e., Moyamoya)
        - Brain Imaging Vascular Flow (CPT® 78610)
            - Cerebral ischemia
            - Establish brain death (rarely done in outpatient setting)

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        - CSF Leakage Detection (CPT® 78650)
            - Evaluation of CSF rhinorrhea or otorrhea, or refractory post-lumbar puncture headache
        - Radiopharmaceutical Dacryocystography (CPT® 78660)
            - Suspected obstruction of nasolacrimal duct due to excessive tearing
- 3D Rendering
    - CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT® 76376 (3D rendering not requiring image post-processing on an independent workstation) is medically necessary in the following clinical scenarios:
        - Bony conditions:
            - Evaluation of congenital skull abnormalities in newborns, infants, and toddlers (usually for pre-operative planning)
            - Complex joint fractures or pelvis fractures
            - Spine fractures (usually for pre-operative planning)
            - Complex facial fractures
        - Pre-operative planning for other complex surgical cases
        - Cerebral angiography
    - 3D Rendering (CPT® 76377 or CPT® 76376) is medically necessary for surgical planning and surgical follow-up after craniotomy when ordered by a surgical specialist.
    - 3D Rendering indications in pediatric head imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines.
- CT or MRI Perfusion (See **CT or MRI Perfusion (HD-24.5)** in the Head Imaging Guidelines.)

Background and Supporting Information

- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive. Clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.
- "The U.S. Food and Drug Administration (FDA) is warning that repeated or lengthy use of general anesthetic and sedation drugs during surgeries or procedures in children younger than 3 years or in pregnant women during their third trimester may affect the development of children's brains. … Published studies in pregnant animals and young animals have shown the use of general anesthetic and sedation drugs for more than 3 hours caused widespread loss of nerve cells in the brain. …All the studies in children had limitations, and it is unclear whether any negative effects seen in children's learning or behavior were due to the drugs or to other factors, such as the underlying medical condition that led to the need for the surgery or procedure."

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- Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
  - The FDA has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.

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General Guidelines-Other Imaging Situations (PEDHD-1.4)

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- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) can be performed for nausea and vomiting which is persistent and unexplained and there is a negative GI evaluation.
- Screening for metallic fragments before MRI should be done initially with plain x-ray.
- Plain x-ray is generally sufficient to screen for aneurysm clips.
- For neurosarcoidosis, see **Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)**.

Background and Supporting Information

- The use of CT Orbital to rule out orbital metallic fragments prior to MRI is rarely necessary.
- Plain x-rays are generally sufficient; x-ray detects fragments of 0.12 mm or more, and CT detects those of 0.07 mm or more.

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Specialized Imaging Techniques (PEDHD-2)

Guideline

Magnetic Resonance Spectroscopy (MRS, CPT® 76390) (PEDHD-2.1)
Functional Magnetic Resonance Imaging (fMRI, CPT® 70554 and CPT® 70555) (PEDHD-2.2)
PET Brain Imaging (CPT® 78608) (PEDHD-2.3)
References (PEDHD-2)

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Magnetic Resonance Spectroscopy (MRS, CPT® 76390) (PEDHD-2.1)

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- Magnetic Resonance Spectroscopy (MRS) involves the analysis of the levels of certain chemicals in pre-selected voxels (small regions) on an MRI scan done at the same time.
- Uses in pediatric neuro-oncology: See **Pediatric CNS Tumors (PEDONC-4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- MRS is medically necessary in individuals with neonatal hypoxic ischemic encephalopathy to help estimate the age of the injury.
- Uses in Metabolic Disorders:
  - See **Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)**.
- MRS is considered not medically necessary for all other pediatric indications at this time.

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Functional Magnetic Resonance Imaging (fMRI, CPT® 70554 and CPT® 70555) (PEDHD-2.2)

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- fMRI is medically necessary to define eloquent areas of the brain as part of pre-operative planning for epilepsy surgery or removal of a mass lesion.
  - The documentation should be clear that brain surgery is planned.
  - Can be performed concurrently with MRI Brain (CPT® 70551 or CPT® 70553) and/or PET Brain Metabolic (CPT® 78608 or CPT® 78609).
- fMRI is considered not medically necessary for all other pediatric indications at this time.

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PET Brain Imaging (CPT® 78608) (PEDHD-2.3)

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- Metabolic FDG PET (CPT® 78608)
  - For pre-operative planning for epilepsy surgery, see **Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)**.
  - For encephalitis including autoimmune encephalitis, see **Autoimmune/ Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD 14.3)**.
  - For uses in pediatric neuro-oncology, see **Pediatric CNS Tumors (PEDONC-4)**.
  - Metabolic FDG PET (CPT® 78608) is considered not medically necessary for all other pediatric indications at this time.
- For Metabolic PET/MRI, see **Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)**.

Background and Supporting Information

- FDG PET Brain is medically necessary to define active areas of the brain as part of pre-operative planning for epilepsy surgery.
- FDG PET Brain/MRI is generally not medically necessary for neurological conditions, except pre-operative planning for epilepsy surgery, due to lack of standardization in imaging technique and interpretation.

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Pediatric Headache (PEDHD-3)

Guideline

Pediatric Headache (PEDHD-3.1)
References (PEDHD-3)

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Pediatric Headache (PEDHD-3.1)

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- A pertinent clinical evaluation including a detailed history and physical examination with a thorough neurologic examination, since the onset or change in signs and/or symptoms, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Advanced imaging is not medically necessary for pediatric individuals with headache in the absence of supported indications.

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Indications	Medically Necessary Imaging Study
◦ Age less than or equal to 5 years ◦ Focal neurological complaints including acute hypertension or altered mental status ◦ Clumsiness (common description of gait or coordination problems in young children) ◦ Headaches awakening from sleep or always present in the morning ◦ Headaches associated with morning nausea/vomiting ◦ Seizures ◦ Progressive worsening in headache frequency and severity without period of temporary improvement ◦ Systemic symptoms such as persistent fever, weight loss, rash, or joint pain ◦ Immunocompromised individual ◦ Known history of cancer of any type ◦ Known autoimmune or rheumatologic disease ◦ Known genetic disorder with predisposition to intracranial mass lesions ◦ History of stable chronic headaches with recent significant change in frequency or severity ◦ Neurological signs and/or symptoms, including headache, after COVID-19 infection ◦ If a recent CT Head is inconclusive	◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553)
◦ Abnormality identified on MRI Brain without contrast (CPT® 70551)	◦ MRI Brain with contrast (CPT® 70552) **OR** ◦ MRI Brain without and with contrast (CPT® 70553)

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Indications	Medically Necessary Imaging Study
◦ Headache precipitated by coughing, sneezing, physical exertion, or Valsalva ◦ Thunderclap headache ◦ Individual with hypercoagulable state or bleeding disorder	◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553) **AND/OR** ◦ MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** ◦ CTA Head (CPT® 70496)
◦ Papilledema on physical exam ◦ Focal signs and/or symptoms of bruit, dissection, vertebrobasilar insufficiency, and/or positional changes	◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553) **AND/OR** ◦ MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** ◦ CTA Head (CPT® 70496) **OR** ◦ MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) For requests of MRA OR CTA AND MRV, see **Pediatric Head Imaging Modality General Considerations (PEDHD-1.3)**.

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Indications	Medically Necessary Imaging Study
◦ Urgent/Emergent settings ◦ Sudden severe headache including thunderclap headache ◦ Acute setting of suspected intracranial infection prior to lumbar puncture (CT Head with contrast CPT® 70460 if intracranial spread of disease is suspected, to detect suppurative fluid collections) (See **General Guidelines-Other Imaging Situations (PEDHD-1.4)**) ◦ Concern for new hemorrhage, significant mass effect, or hydrocephalus and/or rapid clinical deterioration ◦ Recent head trauma ◦ Suspected skull or other bony involvement ◦ If MRI is contraindicated ◦ Ventriculoperitoneal shunt with suspected shunt malfunction. See **Macrocephaly, Microcephaly, and Hydrocephalus (PEDHD-7)** for additional imaging.	◦ CT Head without contrast (CPT® 70450) *CT Head permitted even with prior MRI Brain imaging.*

- If concern for CNS infection, see **CNS Infection (PEDHD-29)**.

Background and Supporting Information

- Headache is a very common complaint in school aged children and adolescents. Many of these children have a family history of one of the primary headache disorders, such as migraine or tension headache.
- CT Head poorly visualizes the posterior fossa in children and is generally insufficient to evaluate pediatric headaches in the absence of supported indications. CT is not supported in lieu of MRI solely to avoid sedation. See **Pediatric Head Imaging Modality General Considerations (PEDHD-1.3)**.

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Pediatric Head and Face Trauma (PEDHD-4)

Guideline

Head Trauma (PEDHD-4.1)
Facial Trauma (PEDHD-4.2)
References (PEDHD-4)

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Head Trauma (PEDHD-4.1)

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- In individuals with recent head trauma, a history focused on the incident and careful examination of the head, neck, and neurological function should be performed since the onset or change in signs and/or symptoms prior to considering advanced imaging.
- Advanced imaging is medically necessary for children with head trauma with ANY of the following red flags:
  - Loss of consciousness
  - Altered mental status or abnormal behavior
  - Known or suspected skull fracture
  - Glasgow Coma Score <15
  - Age younger than 2 years
  - Vomiting
  - Severe mechanism of injury, including, but not limited to:
    - Motor vehicle crash with patient ejection
    - Motor vehicle crash with death of another passenger
    - Motor vehicle crash with rollover
    - Pedestrian or bicyclist without helmet struck by a motorized vehicle
    - Head struck by a high-impact object
    - Falls of more than 1.5m (5 feet) for children aged 2 years and older and more than 0.9m (3 feet) for those younger than 2 years
  - Severe or worsening headache
  - Amnesia
  - Non-frontal scalp hematoma
- CT Head without contrast (CPT® 70450) is the primary advanced imaging study in individuals with acute head trauma.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for the following:
  - Children with an abnormal neurological exam that is not explained by the CT findings
  - Subacute (8 days to one month after initial traumatic event) or chronic blunt head trauma with new or worsening neurological signs or cognitive symptoms
  - Children suspected of being the victims of physical abuse. See **Suspected Physical Child Abuse (PEDMS-7)** in the Pediatric Musculoskeletal Imaging Guidelines.
- For facial trauma, see **Facial Trauma (PEDHD 4.2)** in the Pediatric Head imaging Guidelines.

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- For neck injury, see **Spine Pain Related to Trauma and Painless Spine Trauma (PEDSP 2.6)** in Pediatric Spine Imaging Guidelines.
- Repeat and follow-up imaging can be done at the discretion of the ordering provider.

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Facial Trauma (PEDHD-4.2)

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- CT Maxillofacial without contrast (CPT® 70486)

AND/OR

- CT Orbital/Facial/Temporal Bone without contrast (CPT® 70480) **OR** MRI Orbits/Face/Neck (CPT® 70540)

AND/OR

- CT Head without contrast (CPT® 70450) **OR** MRI Brain without contrast (CPT® 70551)
- Repeat and follow-up imaging can be done at the discretion of the ordering provider.
- For additional concerns, see **Facial Trauma (HD-13.2)** in the Head Imaging Guidelines.

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Sinusitis and Allergic Rhinitis (PEDHD-5)

Guideline

Sinus And Facial Imaging General Considerations (PEDHD-5.1)
Imaging Indications in Sinusitis (PEDHD-5.2)
Stereotactic CT Localization (CPT® 77011) (PEDHD-5.3)
Requests for both Head and Sinus Imaging (PEDHD-5.4)
Allergic Rhinitis (PEDHD-5.5)
Other Indications for Sinus Imaging (PEDHD-5.6)
References (PEDHD-5)

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Sinus And Facial Imaging General Considerations (PEDHD-5.1)

HDP.AR.0005.1.A

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- Acute sinusitis is a clinical diagnosis, and imaging is not medically necessary to establish a diagnosis. Acute bacterial sinusitis can be presumptively diagnosed in a child with acute upper respiratory infection (URI) symptoms and any of the following:
  - Persistent symptoms lasting >10 days without improvement.
  - Worsening symptoms after initial period of improvement.
  - Severe symptoms including purulent nasal discharge and fever >102.2°F for at least 3 consecutive days.

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Imaging Indications in Sinusitis (PEDHD-5.2)

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- Mild mucosal thickening in the paranasal sinuses or mastoids is an extremely common incidental finding noted on head imaging studies done for other indications. If there are no other abnormalities of facial structures noted, this finding is not an indication for advanced imaging of the sinuses or temporal bone.
- CT Maxillofacial without contrast (CPT® 70486) is medically necessary if ANY of the following is present:
  - No improvement after 10 days of appropriate antibiotic treatment (generally this will be amoxicillin/clavulanate, amoxicillin, cefdinir, cefuroxime, cefpodoxime, or ceftriaxone)
  - Recurrence of a treated infection within 8 weeks of effective treatment
  - Chronic sinusitis (persistent residual URI symptoms, including nasal obstruction, facial pressure/pain or cough for >90 days)
  - Known or suspected fungal sinusitis
    - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary if requested instead of CT Maxillofacial.
  - Pre-operative evaluation to assess surgical candidacy
- CT Maxillofacial with contrast (CPT® 70487) is medically necessary if ANY of the following is present:
  - Orbital or facial cellulitis
  - Proptosis
  - Abnormal visual examination
  - Ophthalmoplegia
  - Immunocompromised individual
  - Fungal or vascular lesions visualized in nasal cavity
- CT Head with contrast (CPT® 70460) **OR** MRI Brain without and with contrast (CPT® 70553) **OR** MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary if ANY of the following are present:
  - Focal neurologic findings
  - Altered mental status
  - Seizures
  - Concern for orbital complications
  - Concern for invasive fungal sinusitis

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  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** CTA Head (CPT® 70496) is medically necessary with these findings as well if there is clinical concern for associated vascular complications including but not limited to mycotic aneurysm or venous sinus thrombosis.
- Repeat sinus imaging is generally not medically necessary for individuals who have responded satisfactorily to treatment but is medically necessary with clear documentation of the need for updated CT results to direct acute individual care decisions.

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Stereotactic CT Localization (CPT® 77011) (PEDHD-5.3)

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- Stereotactic CT localization is frequently obtained prior to sinus surgery. The dataset is then loaded into the navigational workstation in the operating room for use during the surgical procedure. The information provides exact positioning of surgical instruments with regard to the individual's 3D CT images. In most cases, the pre-operative CT is a technical-only service that does not require interpretation by a radiologist.
- For treatment planning for sinus surgery CPT® 77011: A stereotactic CT localization scan is frequently obtained prior to sinus surgery. The dataset is then loaded into the navigational workstation in the operating room for use during the surgical procedure. The imaging facility should report CPT® 77011 when performing a scan not requiring interpretation by a radiologist.
- If a diagnostic scan is performed and interpreted by a radiologist, the appropriate diagnostic CT code (e.g., CPT® 70486) should be used.
- It is not appropriate to report both CPT® 70486 and CPT® 77011 for the same CT stereotactic localization imaging session.
- 3D Rendering (CPT® 76376 or CPT® 76377) should not be reported in conjunction with CPT® 77011 (or CPT® 70486 if used). The procedure inherently generates a 3D dataset.
- Such operative studies are medically necessary when ordered by the operating surgeon for this purpose.

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Requests for both Head and Sinus Imaging (PEDHD-5.4)

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- CT Head does not visualize all of the sinuses.
- MRI Brain provides excellent visualization of the sinuses sufficient to recognize sinusitis, and addition of sinus CT for this purpose is unnecessary.
- In individuals being evaluated for potential sinus surgery, separate CT Sinus is often appropriate even after an MRI Brain in order to visualize obstructions to spontaneous mucus flow. See **Stereotactic CT Localization (CPT® 77011) (PEDHD-5.3)**.
- Separate head imaging is not medically necessary in individuals with a normal neurological examination who have headaches associated with sinus symptoms.
- CT or MRI Sinus is not medically necessary for the evaluation of headaches or neurological complaints without a more specific indication pointing to a sinus etiology.

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Allergic Rhinitis (PEDHD-5.5)

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- Advanced imaging is not medically necessary for diagnosis or management of individuals with uncomplicated allergic rhinitis.

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Other Indications for Sinus Imaging (PEDHD-5.6)

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- See **Facial Trauma (PEDHD-4.2)** for imaging guidelines in trauma.
- CT Maxillofacial without contrast (CPT® 70486) - Congenital anomalies of facial structures.
- Cleft lip and palate can be associated with brain malformations and abnormal brain development.
  - MRI Brain (CPT® 70551) is medically necessary in cases of cleft lip and/or palate.
  - MRI Orbits/Face/Neck without contrast (CPT® 70540) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is appropriate if requested by surgeon or any provider in consultation with the surgeon. See **Facial Anomalies (PEDHD-8.2)**.
- 3D CT reconstructed images (CPT® 76377 **OR** CPT® 76376) in conjunction with routine CT should be an integral part of the examination in evaluating craniofacial abnormalities.
- CT Maxillofacial without and with contrast (CPT® 70488) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) - Tumors or other disorders of facial structures.
- Obstructive sleep apnea - See **Pediatric Sleep Disorders (PEDHD-24.1)** for imaging guidelines.
- See **Sinus and Facial Imaging (HD-29)** for conditions not addressed in **Sinus and Facial Imaging (PEDHD-5)**.

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Epilepsy and Other Seizure Disorders (PEDHD-6)

Guideline

Epilepsy and Other Seizure Disorders (PEDHD-6.0)
Initial Imaging of Non-Febrile Seizures (PEDHD-6.1)
Repeat imaging indications (PEDHD-6.2)
Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)
Febrile Seizures (PEDHD-6.4)
References (PEDHD-6)

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Epilepsy and Other Seizure Disorders (PEDHD-6.0)

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- A pertinent evaluation including a detailed history, physical examination with a thorough neurologic examination, since the onset or change in signs and/or symptoms, and appropriate laboratory studies should be performed prior to considering the use of an advanced imaging (CT, MRI, Nuclear Medicine) procedure. An exception can be made if the individual is undergoing guideline-supported, scheduled follow-up imaging evaluation or request is from or in consultation with a neurologist or neurosurgeon who has seen the individual since onset of symptoms. This clinical evaluation should also include family history and (whenever possible) the accounts of eyewitnesses to the event(s).

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Initial Imaging of Non-Febrile Seizures (PEDHD-6.1)

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for the following:
  - First-time seizure in child that has no known cause and is not associated with fever
  - Focal onset seizures
  - New onset primary generalized epilepsy (e.g., absence epilepsy or juvenile myoclonic epilepsy) in those who are neurologically abnormal (e.g., developmental delay)
  - Focal neurologic deficits
  - Inconclusive findings on recent cranial ultrasound or CT Head
    - If an individual meets criteria for MRI imaging for initial imaging of non-febrile seizure, MRI is medically necessary even with a recent negative CT.
  - Concern for a mass lesion or demyelinating disease
- CT Head without contrast (CPT® 70450) is medically necessary for the following (and does not preclude MRI imaging when requested):
  - First-time seizure in child associated with recent head trauma
  - Individual cannot safely undergo MRI (avoidance of sedation is not an indication) or in urgent situations
  - Concern for blood and/or calcifications
- Cranial ultrasound (CPT® 76506) for the following:
  - First-time seizure in child <30 days of age that has no known cause and is not associated with fever if the infant has an open fontanelle.
  - Cranial ultrasound is not required before MRI Brain for hypoxic ischemic encephalopathy (HIE) and/or congenital malformations.

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Repeat imaging indications (PEDHD-6.2)

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- Repeat MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for the following:
  - Need to perform MRI using Epilepsy Protocol (typically 3T magnet with thin section angled slices through hippocampus and temporal lobes, either without or without and with contrast)
  - New or worsening focal neurologic deficits
  - Drug-resistant epilepsy (DRE) (See **Background and Supporting Information** below.)
  - Change in seizure type
  - Repeat imaging for persistent seizures as requested by a specialist or any provider in consultation with a specialist
  - MRI Brain with contrast (CPT® 70552) **OR** MRI Brain without and with contrast (CPT® 70553) to clarify an abnormality on non-contrast MRI or if considering infection or inflammation

Background and Supporting Information

- DRE occurs when there is failure of 2 antiseizure medications (ASMs).
- DRE can also be referred to as "refractory", "intractable," and/or "pharmacoresistant" epilepsy.

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Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)

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- For individuals with a previous MRI Brain and documentation of intractable epilepsy for which surgical treatment or another interventional modality is under active consideration, (see **Background and Supporting Information**), any or all of the following requests can be obtained concurrently and are medically necessary:
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast 3T/7T (CPT® 70553) **AND/OR**
  - Ictal SPECT (CPT® 78803 or CPT® 78830) **AND/OR**
  - Functional MRI (f-MRI) (CPT® 70555 or CPT® 70554) See **Functional MRI (fMRI) (HD-24.2)** in the Head Imaging Guidelines **AND/OR**
  - MR Spectroscopy (CPT® 76390)
  - Metabolic (FDG) PET Brain (CPT® 78608) **AND/OR**
  - CTA Head (CPT® 70496) OR MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) **AND/OR**
  - CTA Neck (CPT® 70498) OR MRA Neck without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549)
  - 3D rendering CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT® 76376 (3D rendering not requiring image post-processing on an independent workstation) is not necessary for epilepsy surgery alone, since 3D rendering can be obtained as part of the MRI Brain epilepsy protocol, unless complicated surgical repair considerations involving craniotomy are required.
  - Metabolic PET/MRI is MRI Brain without contrast (CPT® 70551) OR MRI Brain without and with contrast (CPT® 70553) co-registered with FDG-PET Brain (CPT® 78608) and is medically necessary for pre-surgical evaluation of refractory seizure when requested by neurosurgeon or neurologist or any provider in consultation with a neurosurgeon or neurologist.
- For additional imaging requests for surgery, see **Neurosurgical Imaging (HD-28)** in the Head Imaging Guidelines and/or **Primary Central Nervous System Tumors – General Considerations (ONC 2.1)** in the Oncology Imaging Guidelines.
- For intracranial monitoring with stereo-EEG or grids/strips and electrodes, additional imaging for neuronavigation is medically necessary with one each of the following, after consulting the health plan direction for unlisted codes:
  - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)

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  - CT Head, contrast as requested
  - Due to variances with techniques currently available for neuronavigation, the following are medically necessary:
    - CTA Head (CPT® 70496) **OR** MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546)
- Repeat diagnostic head imaging is medically necessary if previous head imaging is inadequate or additional sequences/protocols are required (including epilepsy protocol imaging) or previous imaging is greater than 6 months old.
- Post-operative imaging including after intracranial (EEG) monitoring is medically necessary per neurosurgeon's request.

Background and Supporting Information

Examples of epilepsy surgery and other interventional modalities include but are not limited to:

- Focal resection
- Temporal lobe resection
- Extratemporal resection
- Lesionectomy
- Multiple subpial transections
- Laser Interstitial Thermal Therapy (LITT)
- Anatomical or functional hemispherectomy and hemispherotomy
- Corpus callostomy
- Steretactic radiosurgery (SRS)
- Neurostimulation device implantations including the following:
  - Vagus nerve stimulation (VNS)
  - Responsive neurostimulation (RNS)
  - Deep brain stimulation (DBS)
- Magnetoencephalography (MEG) plays an important role in clarifying the significance of abnormalities seen on both structural and functional imaging, for the purpose of epileptogenic zone localization for surgical planning. When used in conjunction with other techniques, MEG plays a major role in the non-invasive epilepsy surgery evaluation. Currently, the guidelines only require review for the MRI co-registered with MEG. MEG followed by co-registration with MRI Brain is referred to as Magnetic Source Imaging (MSI).

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Febrile Seizures (PEDHD-6.4)

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- Neuroimaging should not be performed in the routine evaluation of children with simple febrile seizures.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for febrile seizures in the presence of one or more of the following:
  - Seizure lasting >15 minutes
  - Focal onset seizures
  - Focal neurologic deficits
  - Multiple seizures within 24 hours
  - Macrocephaly (head circumference that is greater than the 97th percentile for age and sex, established by use of measurements and CDC growth charts. See **Macrocephaly (PEDHD-7.1)**)
  - Signs and symptoms of increased intracranial pressure
  - Developmental delay
  - MRI Brain permitted even with prior CT Head imaging
- CT Head without contrast (CPT® 70450) **OR** CT Head without and with contrast (CPT® 70470) is medically necessary for:
  - Concern for calcifications
  - For additional indications for CT Head, see **Pediatric Head Imaging Modality General Considerations (PEDHD-1.3)**.

Background and Supporting Information

- A typical febrile seizure is a generalized seizure occurring in the presence of fever (T >100.4°F/38°C) and no central nervous system infection in a child between the age of 6 months and 5 years.

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Macrocephaly, Microcephaly, and Hydrocephalus (PEDHD-7)

Guideline

Macrocephaly (PEDHD-7.1)
Microcephaly (PEDHD-7.2)
Hydrocephalus, Shunts, and Endoscopic Third Ventriculostomy (ETV) (PEDHD-7.3)
References (PEDHD-7)

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Macrocephaly (PEDHD-7.1)

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Indications	Age or Condition	Medically Necessary Imaging Study
Head circumference that is greater than the 97th percentile for age and sex, or head circumference increasing in percentiles over two visits established by use of measurements and CDC growth charts (cdc.gov).	Birth to age 6 months or open fontanelle	◦ Ultrasound Head (CPT® 76506)
	Hydrocephalus or hemorrhage is present on ultrasound	◦ CT Head without contrast (CPT® 70450)
	Any abnormality seen on ultrasound	◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553) *MRI Brain permitted even with prior CT Head imaging*.
	Greater than 6 months old or closed fontanelle	◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553) *CT is not medically necessary in this age group since uncomplicated hydrocephalus is less likely after early infancy.*

Background and Supporting Information

- Macrocephaly is defined as head circumference that is greater than the 97th percentile for age and sex, or head circumference increasing in percentiles over two visits established by use of measurements and CDC growth charts (cdc.gov).

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Microcephaly (PEDHD-7.2)

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- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for all individuals with head circumference that is less than the 3rd percentile for age and sex, or head circumference decreasing in percentiles over two visits established by use of measurements and CDC growth charts (cdc.gov).
  - CT is generally not recommended as that modality lacks the sensitivity to detect the relevant anatomical abnormalities.
  - For CT Head indications, see **Pediatric Head Imaging Modality General Considerations (PEDHD-1.3)**.

Background and Supporting Information

- Microcephaly is defined as head circumference that is less than the 3rd percentile for age and sex, or head circumference decreasing in percentiles over two visits established by use of measurements and CDC growth charts (cdc.gov).

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Hydrocephalus, Shunts, and Endoscopic Third Ventriculostomy (ETV) (PEDHD-7.3)

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Initial Imaging

Indication	Medically Necessary Imaging Study
Age 0-6 months	• Screening Ultrasound Head examination (CPT® 76506) • If ultrasound shows hydrocephalus, MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
Greater than 6 months old	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
Siblings of individuals with aqueductal stenosis (familial screening for hydrocephalus is not otherwise medically necessary)	• One-time CT Head without contrast (CPT® 70450) **OR** • Rapid MRI Brain without contrast (CPT® 70551)

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Repeat Imaging in the Setting of VP Shunt and/or ETV

Indication	Medically Necessary Imaging Study
New signs or symptoms suggesting shunt malfunction or ETV malfunction, including (but not limited to): • Sepsis • Decreased level of consciousness • Protracted vomiting • Visual or neurologic deterioration • Decline of mentation after initial improvement • New or changing pattern of seizures	• Rapid MRI Brain without contrast (CPT® 70551) **OR** • CT Head without contrast (CPT® 70450)
• Post-operatively, or • After shunt setting adjustments, or • As ordered by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• Rapid MRI Brain without contrast (CPT® 70551) **OR** • CT Head without contrast (CPT® 70450)

Repeat Imaging of Ventriculomegaly

Indication	Medically Necessary Imaging Study
0-6 months old	• Serial US Head (CPT® 76506) to monitor ventricular size to determine need and timing of placement of a ventricular catheter, or performance of an endoscopic third ventriculostomy (ETV), timing as requested by the provider
Older than 6 months	• MRI Brain without and with contrast (CPT® 70553), timing as requested by the provider **OR** • MRI Brain without contrast (CPT® 70551), timing as requested by the provider

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Abdominal Imaging Indications in the Setting of VP Shunts

Indication	Medically Necessary Imaging Study
• Suspicion of CSF pseudocyst formation or distal shunt outlet obstruction • Surveillance imaging of the abdomen is medically necessary if ordered by a neurologist, neurosurgeon, or interventional radiologist, or in consultation with one of these providers	• Ultrasound Abdomen (CPT® 76700) **AND/OR** • CT Abdomen **OR** CT Abdomen and Pelvis, contrast as requested

Spine Imaging

Indication	Medically Necessary Imaging Study
Including, but not limited to the following: • Chiari malformation • Malignant infiltration of meninges	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar Spine without contrast (CPT® 72148)

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Indication	Medically Necessary Imaging Study
Dandy-Walker malformation	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141)

- For hydrocephalus due to choroid plexus papillomas, see **Choroid Plexus Tumors (PEDONC-4.13)** in the Pediatric and Special Populations Oncology Imaging Guidelines for those lesions.
- For CSF flow imaging, see **CSF Flow Imaging (HD-24.4)** in the Head Imaging Guidelines.

Additional Rarely Used Studies

- Cisternogram (CPT® 78630) is rarely done in children, but is medically necessary for the following:
  - Known hydrocephalus with worsening symptoms
  - Suspected obstructive hydrocephalus
  - Suspected normal pressure hydrocephalus with gait disturbance and either dementia or urinary incontinence
- Cerebrospinal Ventriculography (CPT® 78635) is rarely done in children, but is medically necessary for the following:
  - Evaluation of internal shunt, porencephalic cyst, or posterior fossa cyst
- Nuclear Medicine Shunt Evaluation (CPT® 78645) and CSF Flow SPECT (CPT® 78803) are rarely done in children, but are medically necessary for the following:
  - Suspected malfunction of ventriculoperitoneal, ventriculopleural, or ventriculovenous shunts

Background and Supporting Information

- Head ultrasound can be performed while the fontanelles are still open and has excellent spatial and anatomic resolution, particularly within the first 2 months of life. After 6 months, smaller acoustic windows due to closing fontanelles limit the sensitivity of the examination.
- Rapid MRI Brain without contrast (CPT® 70551) provides more anatomical detail and does not involve radiation exposure, but many providers use CT Head as rapid MRI is not universally available.
- Hydrocephalus is traditionally divided into non-communicating (the obstruction lies within the course of the brain's ventricular system) and communicating (the obstruction is distal to the ventricular system).

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- Ventriculomegaly refers to enlarged ventricular spaces. It is often initially found on fetal ultrasound. It can be from an obstructive cause or can be relative secondary to small brain volume.
- Benign enlargement of the subarachnoid space in infancy (BESSI), also called benign external hydrocephalus or benign extra-axial fluid collections is the rapid increase in head circumference in an infant with enlarged frontal subarachnoid spaces. See **Macrocephaly (PEDHD-7.1)** for initial imaging guidelines. It typically requires no intervention. Once diagnosed and confirmed with MRI Brain, no additional imaging is required unless new neurological symptoms appear, worsen, or persist beyond age 4 years. If developmental motor delay, see **Developmental Motor Delay (PEDHD 19.3)**.

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Craniofacial Anomalies (PEDHD-8)

Guideline

Craniosynostosis Imaging (PEDHD-8.1)
Facial Anomalies (PEDHD-8.2)
References (PEDHD-8)

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Craniosynostosis Imaging (PEDHD-8.1)

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- Skull x-rays and/or ultrasound should be obtained prior to considering advanced imaging. In cases of very strong consideration of craniosynostosis with surgical planning in progress, x-rays and/or ultrasound are not required.
- CT Head without contrast (CPT® 70450) is medically necessary in the diagnosis of craniosynostosis, with reported sensitivity near 100%. CT also detects associated intracranial pathology.
- 3D rendering (CPT® 76376 or CPT® 76377) is medically necessary with the initial diagnostic CT to evaluate the extent of synostosis and determine surgical candidacy or for preoperative planning.
- CT Maxillofacial without contrast (CPT® 70486) **AND** CT Orbits/Temporal Bone without contrast (CPT® 70480) are generally not medically necessary to evaluate individuals with craniosynostosis, but are medically necessary if the craniosynostosis is part of a larger congenital defect which also involves the bones of the face or orbit.
- CT Maxillofacial without contrast (CPT® 70486) **AND/OR** CT Orbits/Temporal Bone without contrast (CPT® 70480) is/are medically necessary in certain types of craniosynostosis and is/are medically necessary when ordered by surgical specialties or in consultation with surgical specialties during surgical evaluation and planning.
- Ultrasound Head (CPT® 76506) is medically necessary as an alternative method of assessing sutural patency in neonates and infants when radiographs are indeterminate. If inconclusive or for pre-operative planning, CT with 3D rendering is medically necessary as discussed previously in this section.
- CT Head without contrast (CPT® 70450) is medically necessary post-operatively at the discretion of or in consultation with the surgical specialist coordinating the individual's care.
- Positional plagiocephaly typically does NOT require advanced imaging.

Background and Supporting Information

- Craniosynostosis is the premature closure of one or more cranial sutures, usually during infancy.
- Craniosynostosis may be caused by a genetic condition, such as Apert, Pfeiffer, or Crouzon syndrome to name a few.
- Abnormal head shape is the common clinical feature.

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Facial Anomalies (PEDHD-8.2)

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- Congenital anomalies of facial structures
  - CT Maxillofacial without contrast (CPT® 70486) **OR** MRI Orbits/Face/Neck without contrast (CPT® 70540) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
  - MRI Brain without contrast (CPT® 70551) is medically necessary in individuals with cleft lip and/or palate and encephalocele.
- See also **Craniosynostosis Imaging (PEDHD-8.1)**.

Background and Supporting Information

- Facial anomalies (such as cleft lip and palate) can be associated with brain malformations and abnormal brain development.
- Congenital anomalies of facial structures can include, but are not limited to:
  - Cleft lip/palate
  - Choanal atresia
  - Facial clefting
  - CHARGE syndrome
  - Nasofrontal dermoid
  - Encephalocele

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Chiari and Skull Base Malformations (PEDHD-9)

Guideline

Chiari Malformations (PEDHD-9.1)
Basilar Impression/Basilar Invagination (PEDHD-9.4)
Platybasia (PEDHD-9.5)
References (PEDHD-9)

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Chiari Malformations (PEDHD-9.1)

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- Ultrasound can be utilized for initial examination in infants to determine ventricular size and associated anomalies and to provide a baseline for follow-up evaluation but is not required prior to MRI.

Indication	Medically Necessary Imaging Study
◦ Initial evaluation for suspected or known Chiari malformations	Any of the following sets of imaging: MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** MRI Cervical Spine without contrast (CPT® 72141) **OR** MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** MRI Thoracic Spine without contrast (CPT® 72146) **OR** MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** MRI Lumbar Spine without contrast (CPT® 72148) **OR** MRI Lumbar Spine without and with contrast (CPT® 72158)

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Indication	Medically Necessary Imaging Study
◦ Repeat imaging for any of the following: ▪ At the discretion of or in consultation with the neurologist and/or neurosurgeon coordinating the individual's care ▪ New or worsening signs or symptoms ▪ Surgical procedure is actively being considered	Any of the following sets of imaging: MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** MRI Cervical Spine without contrast (CPT® 72141) **OR** MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** MRI Thoracic Spine without contrast (CPT® 72146) **OR** MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** MRI Lumbar Spine without contrast (CPT® 72148) **OR** MRI Lumbar Spine without and with contrast (CPT® 72158)

- Familial screening is not medically necessary for Chiari I Malformations.
- For CSF flow imaging, see **CSF Flow Imaging (HD-24.4)** in the Head Imaging Guidelines.

Background and Supporting Information

- Chiari I malformations involve caudal displacement or herniation of the cerebellar tonsils. Chiari I may be associated with syringomyelia, and rarely with hydrocephalus. Most cases are asymptomatic and discovered incidentally on a head scan performed for another indication. When symptoms are present, they are usually nonspecific but can include headache, lower cranial nerve palsies, or sleep apnea.
- Chiari II malformations are more severe than Chiari I malformations. These individuals usually present at birth. Myelomeningocele is always present, and syringomyelia and hydrocephalus are extremely common.
- Chiari III malformations include cerebellar herniation into a high cervical myelomeningocele. Chiari IV malformations refer to complete cerebellar agenesis.

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Both Chiari III and IV malformations are noted at birth and are rarely compatible with life.

- Repeat brain and spine imaging in individuals with Chiari I malformations and known syringomyelia or hydromyelia is highly individualized.

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Basilar Impression/Basilar Invagination (PEDHD-9.4)

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- See **Basilar Impression/Basilar Invagination (HD-5.4)** in the Head Imaging Guidelines.

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Platybasia (PEDHD-9.5)

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- See **Platybasia (HD-5.5)** in the Head Imaging Guidelines.

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Intracranial Aneurysms and AVM (PEDHD-10)

Guideline

Pediatric Intracranial Aneurysms and Subarachnoid Hemorrhage (PEDHD-10.1)
Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2)
References (PEDHD-10)

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Pediatric Intracranial Aneurysms and Subarachnoid Hemorrhage (PEDHD-10.1)

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- A pertinent evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation or request is from a neurologist or neurosurgeon who has seen the individual since onset of symptoms.
- MRI Brain permitted even with prior CT Head imaging.
- MRI Perfusion is obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553).
  - No additional CPT® codes are necessary or appropriate to perform MRI perfusion.
- Initial study for suspected subarachnoid hemorrhage:
  - CT Head without contrast (CPT® 70450) **AND/OR**
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
- If subarachnoid hemorrhage is present on CT or MRI, or lumbar puncture findings suggest hemorrhage, then the following is/are medically necessary:
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - CT Brain Perfusion (CPT® 0042T) or MRI Brain Perfusion (CPT® 70553)
- Initial study for suspected intracranial aneurysym - individuals presenting with headache, increased intracranial pressure, seizures, or focal neurologic findings:
  - CT Head without contrast (CPT® 70450) **AND/OR**
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
- If findings suspicious for intracranial aneurysm on prior imaging, then the following is supported:
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR**
  - CTA Head (CPT® 70496)
- Imaging for individuals with known aneurysm presenting with headache, increased intracranial pressure, seizures, or focal neurologic findings:
  - CT Head without contrast (CPT® 70450) **AND/OR**
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR**

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  - CTA Head (CPT® 70496)
- CT Head without contrast (CPT® 70450) **AND/OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** CTA Head (CPT® 70496) (except in the case of multisystemic smooth muscle syndrome (MSMS)/smooth muscle dysfunction syndrome (SMDS)/ACTA2 mutations, (see **HD-21.7** in the Head Imaging Guidelines)) is/are medically necessary for individuals with headache, increased intracranial pressure, seizures, or focal neurologic findings **AND** any of the following conditions (including but not limited to):
  - Aicardi–Goutières syndrome
  - Alpha-1-antitrypsin deficiency
  - Alpha-glucosidase deficiency
  - Azygos anterior cerebral artery
  - Bicuspid aortic valve
  - Coarctation of the aorta (for screening of intracranial aneurysms, see **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)**)
  - Ehlers-Danlos Syndrome
  - Fibromuscular dysplasia
  - Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome)
  - Hyper-IgE syndrome
  - Kawasaki disease
  - Klinefelter syndrome
  - Klippel-Trenaunay-Weber syndrome
  - Loeys-Dietz syndrome (there are 4 forms)
  - Marfan syndrome
  - Microcephalic osteodysplastic primordial dwarfism type II
  - Moyamoya syndrome
  - Multisystemic smooth muscle syndrome (MSMS)/smooth muscle dysfunction syndrome (SMDS)/ACTA2 mutations (See **HD-21.7**)
  - Neurofibromatosis type 1
  - Noonan syndrome
  - Patent ductus arteriosus
  - Pheochromocytoma
  - Pseudoxanthoma elasticum
  - Polycystic kidney disease
  - Tuberous sclerosis
- The timing of follow-up imaging for intracranial aneurysms in children is similar to that in adults. See **Intracranial Aneurysms (HD-12.1)** in the Head Imaging Guidelines.

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- Screening for intracranial aneurysms is not medically necessary in individuals with coarctation of the aorta repaired before 3 years of age. See **Intracranial Aneurysms (HD-12.1)** in the Head Imaging Guidelines.
- Screening is not medically necessary in asymptomatic individuals under 20 years of age, unless there are 2 or more first-degree relatives with intracranial aneurysm. See **Intracranial Aneurysms (HD-12.1)** in the Head Imaging Guidelines.

Background and Supporting Information

- Unlike adults, the majority of pediatric aneurysms are caused by genetic or developmental defects rather than environmental or lifestyle factors.
- Pediatric aneurysms most commonly present with subarachnoid hemorrhage, headache, increased intracranial pressure, seizure activity, or focal neurologic findings.

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Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2)

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- A pertinent evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation or request is from or in consultation with a neurologist or neurosurgeon who has seen the individual since onset of symptoms.
- Use this guideline for cerebral vascular malformations including arteriovenous, venous, cavernous, and capillary malformations and vein of Galen malformations.

Disorders	Indications (any of the following)	Medically Necessary Imaging Study
• Any cerebral vascular and/or AVM Disorder listed in this guideline	• When MRI is contraindicated • Any emergency setting	• CT Head without contrast (CPT® 70450) **AND/OR** • CTA Head (CPT® 70496) **AND/OR** • CTA Neck (CPT® 70498)
• Suspected Vein of Galen malformation in the neonate	• Confirmation by Ultrasound Head (CPT® 76506)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • Catheter Angiography to precisely identify the feeding arteries and draining vein, especially if embolization is planned

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Disorders	Indications (any of the following)	Medically Necessary Imaging Study
• Low flow malformations	• When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** • CTA Head (CPT® 70496)
• Suspected AVM after the neonatal period	• When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
• Known AVM	• When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** • CTA Head (CPT® 70496)

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Disorders	Indications (any of the following)	Medically Necessary Imaging Study
• Hereditary Hemorrhagic Telangiectasia (HHT, Osler-Weber-Rendu syndrome)	• Suspected based on family history with at least one affected first-degree relative. (biological parent or sibling or child) • At diagnosis • Screening for confirmed HHT • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist or neurosurgeon or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR** • CTA Head (CPT® 70496)

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Disorders	Indications (any of the following)	Medically Necessary Imaging Study
• Capillary malformation-arteriovenous malformation (CM-AVM)	• Suspected based on family history with at least one affected first-degree relative (biological parent or sibling or child) • At diagnosis • Screening for confirmed CM-AVM • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist or neurosurgeon or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR** CTA Head (CPT® 70496) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** MRI Thoracic Spine without contrast (CPT® 72146)

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Disorders	Indications (any of the following)	Medically Necessary Imaging Study
• Cerebral cavernous malformations (CCM)	• Suspected based on family history with at least one affected first-degree relative (biological parent or sibling or child). • At diagnosis • Screening for confirmed CCM • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist or neurosurgeon or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

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Disorders	Indications (any of the following)	Medically Necessary Imaging Study
• Microcephalic osteodysplastic primordial dwarfism, type II (MOPDII)	• Suspected based on family history with at least one affected first-degree relative (biological parent or sibling or child) • At diagnosis • Screening for confirmed MOPDII, repeated annually • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist or neurosurgeon or geneticist	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck without contrast (CPT® 70547, CPT® 70548 **OR** CPT® 70549) **OR** • CTA Neck (CPT® 70498)
• Sturge-Weber syndrome	• At diagnosis • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** • MRI Orbits/Face/Neck without contrast (CPT® 70540)

Additional indications

- CT Head without contrast (CPT® 70450) is medically necessary for ANY one of the following:
  - Mass effect

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  - Urgent/emergent settings due to availability and speed of CT
  - Trauma
  - Recent hemorrhage, whether traumatic or spontaneous
  - Prior to lumbar puncture in individuals with cranial complaints
  - Scenarios in which MRI is contraindicated (i.e., pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)
- Prior CT Head (CPT® 70450) does not exclude indication for MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551).

Background and Supporting Information

- Most intracranial AVMs are congenital, vary widely in their location and type, and are discovered at birth due to associated clinical findings or incidentally later in life. Certain hereditary conditions are associated with an increased risk for AVM development.
- Vascular malformations include arteriovenous, venous, cavernous, and capillary malformations. The vein of Galen malformation is the most common arteriovenous malformation, presenting in neonates with signs of high output congestive heart failure or later in infancy of childhood with signs of hydrocephalus. Low flow venous, cavernous, and capillary malformations may be asymptomatic and discovered incidentally, or they may present in childhood with seizures or neurologic findings secondary to intracranial hemorrhage.
- Hereditary AVMs usually have an autosomal dominant pattern of inheritance.

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Syncope (PEDHD-11)

Guideline

Syncope (PEDHD-11.1)
References (PEDHD-11)

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Syncope (PEDHD-11.1)

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- Advanced imaging of the brain is not medically necessary for individuals with isolated syncope without focal neurologic findings. See **Syncope (PEDCD-5)** in the Pediatric Cardiac Imaging Guidelines and **Epilepsy and Other Seizure Disorders (PEDHD-6)** for additional imaging considerations.

Background and Supporting Information

- Syncope in children is almost always neurocardiogenic (vasovagal) in nature. Intracranial mass lesions do not cause isolated syncope. Syncope and seizure activity can often be challenging to distinguish for unwitnessed syncope.
- There is no role for advanced neuroimaging for Postural Orthostatic Tachycardia Syndrome (POTS).

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Pediatric Stroke (PEDHD-12)

Guideline

Pediatric Stroke General Considerations (PEDHD-12.1)
Pediatric Stroke Initial Imaging (PEDHD-12.2)
Pediatric Stroke Subsequent Imaging (PEDHD-12.3)
Moyamoya Syndrome/Disease (PEDHD-12.4)
Sickle Cell Disease (PEDHD-12.5)
CNS Vasculitis (PEDHD-12.6)
COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)
Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations (PEDHD-12.8)
References (PEDHD-12)

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Pediatric Stroke General Considerations (PEDHD-12.1)

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- Imaging indications are the same for neonates as for older children.
- Children may not present with typical stroke findings.

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Pediatric Stroke Initial Imaging (PEDHD-12.2)

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- In the presence of neurological signs and/or symptoms with concern for stroke, **ANY** of the following studies are medically necessary:
  - CT Head without contrast (CPT® 70450) **OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553)
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** CTA Head (CPT® 70496)
  - For suspected carotid or vertebral artery dissection, CTA Neck (CPT® 70498) **OR** MRA Neck (CPT® 70547, CPT® 70548, **OR** CPT® 70549)
  - Note: Both MRA **OR** CTA Head **AND** Neck are needed to visualize the posterior vertebrobasilar circulation for evaluation of the vertebrobasilar stroke/TIA (vertigo associated with diplopia, dysarthria, bifacial numbness or ataxia) or concern for arterial dissection (risks may include premature stroke [under age 50], head or neck trauma, fibromuscular dysplasia, Ehlers-Danlos syndrome, and chiropractic neck manipulation)
- In individuals with COVID-19, See **COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)**

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Pediatric Stroke Subsequent Imaging (PEDHD-12.3)

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for any new or worsening neurological findings or seizure activity.
- Repeat imaging for follow-up and resolution of stroke or hemorrhage as determined by a neurologist, neurosurgeon, hematologist, or physiatrist (PM&R), or any provider in consultation with a neurologist, neurosurgeon, hematologist, or physiatrist (PM&R).
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553)
  - MRA Head/MRV Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR** CTA Head/CTV Head (CPT® 70496) for follow-up of known cerebral artery stenosis or thrombosis
  - Other surveillance imaging indications after stroke are listed in the disease-specific sections.

Background and Supporting Information

- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography):
  - If arterial and venous CT or MR studies are both performed in the same session, only **one** CPT® code is used to report both procedures.
  - If an arterial CTA **OR** MRA study has been performed and subsequently a repeat study is needed to evaluate the venous anatomy, then this study is medically necessary.
  - If a venous CTV **OR** MRV study has been performed and subsequently a repeat study is needed to evaluate the arterial anatomy, then this study is medically necessary.
  - MRA without and with contrast with venous sinus thrombosis to differentiate total from subtotal occlusion

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Moyamoya Syndrome/Disease (PEDHD-12.4)

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- See **Moyamoya Syndrome/Disease (HD-21.5)** in the Head Imaging Guidelines.

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Sickle Cell Disease (PEDHD-12.5)

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- The following imaging is medically necessary for all sickle cell individuals with a severe phenotype (Hgb SS or Hgb $S\beta^0$):
  - Transcranial Doppler (TCD) Ultrasound (CPT® 93886 or CPT® 93888) annually for all individuals aged 2 to 16:
    - A short interval repeat study is medically necessary for individuals with conditional (170-199cm/sec) flow results, or with individuals undergoing transfusion therapy.
  - Transcranial Doppler (TCD) Ultrasound (CPT® 93886 or CPT® 93888) for children aged 17 years old may be appropriate on a case-by-case basis.
    - See also **Stroke/TIA (HD-21.1)** in the Head Imaging Guidelines.
  - After 17 years old, for individuals with a history of abnormal TCDs, TCDs may be repeated every 3 months.
  - TCD is not medically necessary for individuals with other phenotypes (Hgb SC, Hgb $S\beta^+$).
  - See indications below for advanced imaging with MR or CT.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for any of the following:
  - Two non-imaging Transcranial Doppler (TCD) measurements of ≥200cm/sec or a single measurement of >220cm/sec or 2 assessment TCD measurements ≥185cm/sec or a single measurement >205cm/sec.
  - Persistently abnormal TCD velocities
    - For more regarding TCD, see **Background and Supporting Information**.
  - Screening to detect silent cerebral infarcts
  - New symptoms or cognitive impairment occurs or a change in academic performance
  - After an infarct-like lesion is identified, repeat every 12-24 months to assess for cerebral infarct progression
  - Prior to any change in therapy

Background and Supporting Information

- TCD is used to screen for overt and silent infarctions and monitor response to transfusion therapy.
- Individuals with sickle cell disease are at significantly increased risk for stroke and silent infarction, beginning at a very young age. Recent advances allow physicians to identify individuals at high risk for stroke and begin a primary stroke prevention program.

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- Identification of silent cerebral infarction is important because treatment with prophylactic red cell transfusions to maintain hemoglobin S levels at <30% of total hemoglobin may reduce recurrent stroke and extent of neurologic damage.

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CNS Vasculitis (PEDHD-12.6)

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- Medically necessary initial imaging for concern for primary CNS vasculitis, or for individuals with systemic vasculitis and suspected CNS involvement include the following:
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - CTA Neck (CPT® 70498)
- If MRI findings are suggestive of vasculitis, the following is medically necessary:
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - CTA Neck (CPT® 70498)
- The following are medically necessary for treatment response and/or surveillance imaging as ordered by a neurologist, rheumatologist, or neurosurgeon, or in consultation with one of these providers:
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - CTA Neck (CPT® 70498)

Background and Supporting Information

- A normal MRI Brain almost always completely excludes intracranial vasculitis.

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COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)

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- If concern for CNS infection, see **CNS Infection (PEDHD-29.1)**.
- For cardiac imaging, see **Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)**

  in the Pediatric Cardiac Imaging Guidelines.
- Symptoms of MIS-C may include some or all of the following:
  - Headache **AND/OR** irritability
  - Mucocutaneous changes similar to Kawasaki disease (i.e., strawberry tongue, red cracked lips, rash of hands and/or feet)
  - Polymorphus **AND/OR** vasculitic rash
  - Non-exudative conjunctivitis
  - Tachycardia **AND/OR** hypotension
  - Cough **AND/OR** shortness of breath
  - Abdominal pain, vomiting, **AND/OR** diarrhea
  - Lymphadenopathy, joint pain, **AND/OR** sore throat

Indication	Medically Necessary Imaging Study
◦ Imaging for neurological signs and/or symptoms, including headache, after known or presumed COVID-19 infection	◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553) **AND/OR** ◦ MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** ◦ CTA Head (CPT® 70496) **AND/OR** ◦ MRA Neck (CPT® 70547, CPT® 70548, **OR** CPT® 70549) **OR** ◦ CTA Neck (CPT® 70498)

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Background and Supporting Information

- COVID-19 infections in children are generally mild in comparison to that of adults, however a post-viral syndrome in children has become increasingly noted.
- Multisystem Inflammatory Syndrome in Children (MIS-C) can cause an inflammatory vasculopathy, prothrombotic state and/or post-viral myocarditis in children who have had a COVID-19 infection caused by SARS-CoV-2. The child may have had minor symptoms or been asymptomatic at the time of COVID-19 infection but the virus can trigger endothelial injury and activation of the IL-1 pathway similar to that in Kawasaki disease and acute rheumatic fever.

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Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations (PEDHD-12.8)

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- See **Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations (HD-21.7)** in the Head Imaging Guidelines.

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Benign Brain Lesions (PEDHD-13)

Guideline

Arachnoid Cysts (PEDHD-13.1)
Pineal/Colloid Cysts (PEDHD-13.2)
Acoustic Neuromas (PEDHD-13.3)

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Arachnoid Cysts (PEDHD-13.1)

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- See **Arachnoid Cysts (HD-35.1)** in the Head Imaging Guidelines.

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Pineal/Colloid Cysts (PEDHD-13.2)

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- See **Pineal/Colloid Cysts (HD-34.1)** in the Head Imaging Guidelines.

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Acoustic Neuromas (PEDHD-13.3)

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- See **Neurofibromatosis 2 (PEDPND-2.2)** in the Pediatric Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines.

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Pediatric Demyelinating Diseases (PEDHD-14)

Guideline

Pediatric Demyelinating Disease General Considerations (PEDHD-14.1)
Multiple Sclerosis (MS) (PEDHD-14.2)
Acute Disseminated Encephalomyelitis (ADEM) and Other Pediatric Demyelinating Disorders (PEDHD-14.3)
Transverse Myelitis (PEDHD-14.4)
References (PEDHD-14)

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Pediatric Demyelinating Disease General Considerations (PEDHD-14.1)

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- For evaluation of pediatric demyelinating disease, the following imaging is medically necessary:
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR**
  - MRI Thoracic Spine without and with contrast (CPT® 72157)
- MRI Lumbar Spine without and with contrast (CPT® 72158) is not medically necessary unless the individual has a tethered cord or other anatomic abnormality causing caudal displacement of the filum terminalis.
- CT imaging is generally **NOT** medically necessary in the evaluation of demyelinating disease.
- Metabolic (FDG) PET Brain (CPT® 78608) and MR Spectroscopy (CPT® 76390) are considered not medically necessary for evaluation of pediatric demyelinating diseases.
- For metabolic or genetic white matter disorders, including leukodystrophies and mitochondrial disorders, see **Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)**.

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Multiple Sclerosis (MS) (PEDHD-14.2)

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Indication	Medically Necessary Imaging Study
Initial diagnosis in individuals with clinical signs and/or symptoms suggestive of MS	• MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) If there is a contraindication to gadolinium administration, then • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146)

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Indication	Medically Necessary Imaging Study
Disease monitoring whether or not receiving treatment	Every 6 months or for new signs/ symptoms: • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) Every 12 months or for new signs/ symptoms: • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)
Optic neuritis suspected	• MRI Orbits/Face/Neck without and with contrast (CPT® 70543) may be added If there is a contraindication to gadolinium administration, then • MRI Orbits/Face/Neck without contrast (CPT® 70540) may be added
Symptoms suggestive of Progressive multifocal leukoencephalopathy (PML) during natalizumab (Tysabri®) therapy (or other medications with similar risk)	• MRI Brain without and with contrast (CPT® 70553) If there is a contraindication to gadolinium administration, then ◦ MRI Brain without contrast (CPT® 70551)

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Indication	Medically Necessary Imaging Study
Screening for individuals on natalizumab (Tysabri®) or other drugs with risk of progressive multifocal leukoencephalopathy (PML)	Every 6 months while on treatment: • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) Every 3-6 months for high-risk individuals positive for serum JC virus antibody and >18 months natalizumab exposure: • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
If MRI Brain without contrast (CPT® 70551) shows incidental evidence of possible demyelinating disease	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain with contrast (CPT® 70552)
After an MRI Brain without contrast (CPT® 70551), a follow-up MRI brain may be performed at the discretion of a neurologist, a neurosurgeon, or a neuro-ophthalmologist, or any provider in consultation with a neurologist, neurosurgeon, or neuro-ophthalmologist, and/or at the recommendation of the radiologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain with contrast (CPT® 70552)

Background and Supporting Information

- Multiple sclerosis is less common in children. About 4% of MS cases are diagnosed before age 18, and only ~0.7% of all MS cases begin before age of 10 years.
- Common presentations of MS in children include ataxia, optic neuritis, diplopia, transverse myelitis or as an acute encephalitis-like illness.
- Among children with suspected demyelinating diseases, the principal differential diagnosis is often between MS, Acute Disseminated Encephalomyelitis (See **Acute Disseminated Encephalomyelitis (ADEM) and Other Pediatric Demyelinating Disorders (PEDHD-14.3)** or MOG Antibody-Associated Disease (MOGAD). (See **MOG Antibody-Associated Disease (MOGAD) (HD-16.3)**).
- Medications with similar risks of Progressive Multifocal Leukoencephalopathy (PML) as Tysabri® include: dimethyl fumarate (Tecfidera®), fingolimod (Gilenya®),

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ocrelizumab (Ocrevus®), clardribine (Mavenclad®), diroximel fumarate (Vumerity®), eculizaumab (Soliris®), ozanimod (Zeposia®), alemtuzumab (Lemtrada®), monomethyl fumarate (Bafiertam®), rituximab (Rituxan®).

- If a non-contrast study shows incidental evidence of possible demyelinating disease, repeat imaging is appropriate as the presence of enhancing lesions may be helpful in confirming the diagnosis.
- 3D imaging in the evaluation of Multiple Sclerosis is not supported as a separate code. Most scanners are capable of 3D acquisitions or other imaging sequences.

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Acute Disseminated Encephalomyelitis (ADEM) and Other Pediatric Demyelinating Disorders (PEDHD-14.3)

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- MRI Brain without and with contrast (CPT® 70553) **AND/OR** MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** MRI Thoracic Spine without and with contrast (CPT® 72157) are medically necessary as the initial imaging in individuals with clinical signs and/or symptoms suggestive of ADEM OR for follow-up imaging when ordered by a neurologist or any provider in consultation with a neurologist.
  - MRI Brain without contrast (CPT® 70551) **AND/OR** MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** MRI Thoracic Spine without contrast (CPT® 72146) are medically necessary if there is a contraindication to gadolinium.
- Other pediatric demyelinating disorders that have clinical overlap with multiple sclerosis and ADEM include, but are not limited to:
  - **Neuromyelitis Optica Spectrum Disorders (HD-16.2)**
  - **Anti-MOG Syndromes (HD-16.3)**
  - **Traverse myelitis (HD-16.4)**
- For metabolic or genetic white matter disorders, including leukodystrophies and mitochondrial disorders, see **Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)**.

Background and Supporting Information

- ADEM has an acute onset, and is more common among younger children than MS, but the signs and symptoms overlap significantly and distinguishing between MS and ADEM can be challenging based on clinical examination alone.
- Most individuals will have complete clinical recovery by 12 months, while stable MRI abnormalities (gliosis) may persist. These findings do not require additional imaging unless the individual develops new neurologic symptoms. Prolonged symptoms or return of symptoms may represent a different demyelinating disorder.

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Transverse Myelitis (PEDHD-14.4)

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- See **Transverse Myelitis (HD-16.4)** in the Head Imaging Guidelines.

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Pituitary Dysfunction (PEDHD-15)

Guideline

Pituitary Dysfunction General Considerations (PEDHD-15.1)
Panhypopituitarism (PEDHD-15.2)
Isolated Growth Hormone Deficiency (PEDHD-15.3)
Diabetes Insipidus (DI) and Other Disorders of Anti-Diuretic Hormone (PEDHD-15.4)
Precocious Puberty (PEDHD-15.5)
Benign Pituitary Tumors (PEDHD-15.6)
Pituitary Malignancies (PEDHD-15.7)
References (PEDHD-15)

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Pituitary Dysfunction General Considerations (PEDHD-15.1)

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- The initial step in the evaluation of all potential pituitary masses is a detailed history, recent physical examination, and thorough neurological exam, including evaluation of the visual fields.
- Endocrine laboratory studies should be performed prior to considering initial advanced imaging.
- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) when pituitary imaging is medically necessary.
  - Pituitary Gland: one study (either MRI Brain without and with contrast [CPT® 70553] **OR** MRI Orbits/Face/Neck [CPT® 70543]) is adequate to report the imaging of the pituitary. The reporting of two CPT® codes to image the pituitary is not medically necessary.
- If a previous MRI Brain was reported to show a possible pituitary tumor with supporting evidence of pituitary disease or is inconclusive, a repeat MRI with dedicated pituitary protocol may be performed. If a prior MRI Brain was without contrast, a follow-up scan either MRI Brain with contrast (CPT® 70552) **OR** MRI Brain without and with contrast (CPT® 70553) is appropriate.
- For association between pituitary dysfunction and optic nerve issues, see **Eye Disorders and Visual Loss (HD- 32.1)** in the Head Imaging Guidelines.
- For repeat imaging, see **Pituitary, Sella, Hypothalamus (HD-19.1)** in the Head Imaging Guidelines.

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Panhypopituitarism (PEDHD-15.2)

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- Endocrine testing should be performed initially.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) with special attention to the pituitary is medically necessary for newly diagnosed Panhypopituitarism.
- Individuals with a normal pituitary on initial MRI do not need routine follow-up imaging.
- Individuals with mass lesions should have follow-up imaging according to the guidelines for the specific diagnosis.

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Isolated Growth Hormone Deficiency (PEDHD-15.3)

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- Clinical features include: height below the normal range (<3rd percentile), subnormal growth velocity or the child's height is below the range expected based on parental height.
- Risk factors include: a history of brain tumor, cranial irradiation, or other congenital/ organic hypothalamic-pituitary abnormality as well as an incidental finding of a hypothalamic-pituitary abnormality on MRI.
- Endocrine testing should be performed initially.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) with special attention to the pituitary is medically necessary for any of the following:
  - Both IGF1 and IGFBP3 are below the laboratory reference range for age/sex or Tanner stage.
  - 2 measurements of growth hormone stimulation with different stimulation agents (glucagon, clonidine, arginine, insulin, levodopa) performed on the same day or separate days produce maximal GH levels <10ng/mL. See **Background and Supporting Information.**
- Individuals with a normal pituitary on initial MRI do not need routine follow-up imaging.
- Individuals with mass lesions should have follow-up imaging according to the guidelines for the specific diagnosis.

Background and Supporting Information

- Growth hormone stimulation testing is limited by poor specificity and requires failure on 2 tests to diagnose growth hormone deficiency.
- Controversy exists as to the cutoff level which differentiates a normal response from a deficient response on provocative testing. Some experts support GH <7ng/mL however many pediatric endocrinologists consider a peak GH level <10ng/mL to be indicative of growth hormone deficiency and may identify children with partial GHD.

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Diabetes Insipidus (DI) and Other Disorders of Anti-Diuretic Hormone (PEDHD-15.4)

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- Laboratory testing should be performed initially. Diabetes insipidus is characterized by serum osmolality >300mOsm/kg and urine osmolality <300mOsm/kg.
- Central diabetes insipidus (CDI) is caused by diminished synthesis or secretion of vasopressin in the hypothalamus and nephrogenic diabetes insipidus (NDI) is caused by renal resistance to vasopressin.

Central Diabetes Insipidus (DI)

- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for newly diagnosed central DI.
- Individuals with a normal pituitary on initial MRI can have repeat MRI Brain without and with contrast (CPT® 70553) every 3-6 months for the first 2 years as germinomas may cause central DI while still too small to detect on imaging.
  - Serial measurement of β -hCG is also medically necessary for these individuals, and MRI should be repeated if a significant rise in β -hCG is detected on screening.
- Individuals with mass lesions should have follow-up imaging according to the guidelines for the specific diagnosis.

Nephrogenic DI

- Once this diagnosis is firmly established, further advanced imaging is usually not medically necessary.

Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH)

- Laboratory studies should be obtained prior to considering advanced imaging—urine osmolality should be high and serum osmolality low.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for initial evaluation of unexplained central (hypothalamic pituitary) SIADH.
- Individuals with a normal pituitary on initial MRI do not need routine follow-up imaging.
- Individuals with mass lesions should have follow-up imaging according to the guidelines for the specific diagnosis.

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Background and Supporting Information

- See **Small Cell Lung Cancer-Suspected/Diagnosis (ONC-7.1)** and **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.
- Pulmonary diseases including infection (tuberculosis, viral/bacterial pneumonia), acute respiratory infections, mechanical ventilation and others can cause SIADH although the mechanism is unclear. Individuals with lung disease should have chest imaging according to the guidelines for the specific diagnosis.

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Precocious Puberty (PEDHD-15.5)

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- Defined as the appearance of secondary sexual characteristics before age 8 in females and before age 9 in males. The diagnosis is made clinically using Tanner staging and often will include a bone age assessment (hand/wrist radiographs) and/or abdominal and/or pelvic ultrasound (See Peripheral Precocious Puberty below).
- Endocrine laboratory studies (baseline LH, FSH and either estradiol or testosterone) are used to determine if the etiology of precocious puberty is central (gonadotropin dependent) or peripheral (gonadotropin independent). Estradiol and testosterone levels will often be elevated to a pubertal range.

Central Precocious Puberty (CPP)

- An LH >0.3U/L on a random blood sample is the most reliable screening test for central precocious puberty. If LH <0.3U/L and CPP is suspected, a stimulation test with a GnRH analog is the gold standard.
- Neuroimaging should always follow hormonal studies that suggest a central origin of precocious puberty.
- MRI Brain without and with contrast (CPT® 70553, preferred study) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for evaluation of any child with documented central precocious puberty.
- MRI is appropriate irrespective of age and gender in individuals with precocious puberty and concurrent CNS symptoms of severe headache, visual changes, or seizures.
- Individuals with a normal pituitary on initial MRI do not need routine follow-up imaging.
- Individuals with mass lesions should have follow-up imaging according to the guidelines for the specific diagnosis. (**Benign Pituitary Tumors (PEDHD-15.6**) and **Pituitary Malignancies (PEDHD-15.7)**)

Peripheral Precocious Puberty

- The differential diagnosis of peripheral precocious puberty (LH suppressed or in the pre-pubertal range with elevated estradiol, testosterone, and/or adrenal androgens) is broad and may include ovarian, testicular, adrenal, and other sources of excessive hormonal production.
- Ultrasound Abdomen (CPT® 76700) in both genders and Ultrasound Pelvis (CPT® 76856) in females and Scrotal ultrasound (CPT 76870) in males depending on the suspected source of hormonal excess for initial imaging.

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- See **CNS Germinomas and Non-Germinomatous Germ Cell Tumors (PEDONC-4.7)** in the Pediatric and Special Populations Oncology Imaging Guidelines for evaluation of HCG secreting CNS tumors.
- See **Hepatoblastoma (PEDONC-11.2)** in the Pediatric and Special Populations Oncology Imaging Guidelines for evaluation of HCG secreting hepatic tumors.
- See **Pediatric Germ Cell Tumors (PEDONC-10)** in the Pediatric and Special Populations Oncology Imaging Guidelines and **Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)** in the Oncology Imaging Guidelines for evaluation of Leydig Cell tumors.
- See **Adrenal Cortical Lesions (AB-16.1)** in the Abdomen Imaging Guidelines for evaluation of adrenal virilizing tumors.

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Benign Pituitary Tumors (PEDHD-15.6)

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- Benign pituitary tumor indications in pediatric individuals are identical to those for adult individuals. See **Pituitary, Sella, Hypothalamus (HD-19)** in the Head Imaging Guidelines.

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Pituitary Malignancies (PEDHD-15.7)

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- See **Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (PEDONC-4.10)** or **Histiocytic Disorders (PEDONC-18)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Pediatric Ear Disorders (PEDHD-16)

Guideline

Hearing Loss (PEDHD-16.1)
Ear Pain (PEDHD-16.2)
Cholesteatoma (PEDHD-16.3)
Vertigo (PEDHD-16.4)
Tinnitus (PEDHD-16.5)
References (PEDHD-16)

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Hearing Loss (PEDHD-16.1)

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- A pertinent evaluation including a detailed history, physical examination (including otoscopic examination), and age-appropriate audiology testing should be performed on any child with known or suspected hearing loss prior to considering advanced imaging. The selection of imaging testing will depend on the age of the child and type of hearing loss.
- CT Orbits/Temporal Bone without contrast (CPT® 70480) is medically necessary for the following:
  - Conductive hearing loss of any cause
  - Pre-operative planning for resection of mass lesion or cochlear implant placement
  - Sensorineural hearing loss in individuals who cannot safely undergo MRI
  - Mixed conductive and sensorineural hearing loss
  - Congenital hearing loss
  - Total deafness
- MRI Brain without and with contrast (CPT® 70553) with attention to internal auditory canals (included in CPT® 70553 and does not require a separate CPT® code) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for the following:
  - Conductive hearing loss secondary to known or suspected mass lesion
  - Pre-operative planning for resection of mass lesion or cochlear implant placement
  - Sensorineural hearing loss of any cause
  - Mixed conductive and sensorineural hearing loss
  - Congenital hearing loss
  - Total deafness
  - For hearing loss associated with tinnitus, see **Tinnitus (PEDHD-16.5)**.
- Both modalities (CT and MRI) are supported simultaneously for evaluation and surgical planning if ordered by or in consultation with an ENT or neurosurgical specialist.
- Limited MRI Brain with attention to internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) is medically necessary when requested by the provider in place of a complete MRI Brain. Note: Limited MRI codes should not be used in addition to MRI Brain codes; IAC views are performed as additional sequences as part of the brain study. (See **General Guidelines – Anatomic Issues (HD-1.1)** in the Head Imaging Guidelines)

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Ear Pain (PEDHD-16.2)

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- A pertinent evaluation including a detailed history, physical examination (including otoscopic examination), should be performed on any child with ear pain prior to considering advanced imaging. Common causes of ear pain include external and middle ear infections, dental problems, sinus infection, neck problems, and referred pain from the oral pharynx, tonsillitis, and pharyngitis.
- Advanced imaging is not medically necessary in the overwhelming majority of pediatric individuals with ear pain.

Indications	Medically Necessary Imaging Study
◦ Any of the following ▪ Persistent ear pain without obvious cause ▪ Clinical suspicion for complicated or invasive infection such as mastoiditis ▪ Clinical suspicion of mass lesion causing ear pain ▪ Significant trauma with concern for hematoma formation ▪ Pre-operative planning	◦ ONE of the following: ▪ CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR** ▪ CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR** ▪ MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR** ▪ MRI Orbits/Face/Neck without and with contrast (CPT® 70543)

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Cholesteatoma (PEDHD-16.3)

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- ONE of the following studies is medically necessary for pre-operative evaluation in children with cholesteatoma:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR**
  - MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), **OR**
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
- ONE of the following studies is medically necessary one time post-operatively to exclude residual or regrown cholesteatoma to avoid the need for a second-look surgery:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR**
  - MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR**
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)

Background and Supporting Information

- Cholesteatomas are expansive cysts of the middle ear filled with cellular debris. They can be congenital or arise from recurrent middle ear infections or trauma to the tympanic membrane. Hearing loss is usually conductive, although if the lesion is large enough combined conductive and sensorineural hearing loss may be present. Otoscopic exam findings and symptoms may include a white mass in the middle ear cleft, painless drainage from the ear or chronic/recurrent ear infections. Advanced imaging for the diagnosis and management of suspected cholestatoma, in particular, should be reserved for the otolaryngologist or provider in consultation with the otolaryngologist.

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Vertigo (PEDHD-16.4)

HDP.ED.0016.4.A

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- A pertinent evaluation including a detailed history, physical examination (including otoscopic examination), should be performed on any child with vertigo prior to considering advanced imaging.
- If physical examination is otherwise normal and the vertigo responds to treatment, advanced imaging is not medically necessary.
- MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) is medically necessary for the following:
  - Vertigo with associated headache or ataxia
  - Vertigo associated with tinnitus
  - Vertigo that does not respond to vestibular treatment

Background and Supporting Information

Isolated vertigo is an uncommon complaint during childhood. Middle ear/Eustachian tube problems are the most common cause of isolated vertigo in children.

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Tinnitus (PEDHD-16.5)

HDP.ED.0016.5.A
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- Tinnitus without hearing loss is a less common complaint during childhood.
- Children with hearing loss and tinnitus should be imaged according to **Hearing Loss (PEDHD-16.1)**. A pertinent evaluation including a detailed history, physical examination (including otoscopic examination), and age-appropriate audiology testing should be performed on any child with known or suspected tinnitus prior to considering advanced imaging.
- Advanced imaging is not medically necessary in the overwhelming majority of pediatric individuals with isolated tinnitus and normal hearing.

Indications	Medically Necessary Imaging Study
• ANY of the following ◦ Clinical suspicion of mass lesion causing tinnitus ◦ Persistent tinnitus after recent significant trauma	• ONE of the following ◦ CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR** ◦ CT Orbits/Temporal Bone without and with contrast (CPT® 70482), **OR** ◦ MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR** ◦ MRI Brain without contrast with attention to internal auditory canals (CPT® 70553) **OR** ◦ MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
• ANY of the following: ◦ Pulsatile tinnitus ◦ Suspicion for vascular lesions	• MRA Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548 **OR** CPT® 70549) **OR** • CTA Neck (CPT® 70498)

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Autism Spectrum Disorders (PEDHD-17)

Guideline

Autism Spectrum Disorders (PEDHD-17.1)
References (PEDHD-17)

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Autism Spectrum Disorders (PEDHD-17.1)

HDP.AS.0017.1.A

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- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for:
  - new or worsening focal neurologic findings documented on a pertinent physical **AND/OR**
  - loss of developmental milestones and/or regression in two or more areas of development.
- PET imaging is considered not medically necessary in the evaluation of individuals with autism spectrum disorders.

Background and Supporting Information

- The group of diagnoses are classified as pervasive development disorders (PDD). These diagnoses are established on clinical criteria, and no imaging study can confirm the diagnosis.
- Comprehensive evaluation for autism might include history, physical exam, audiology evaluation, speech, language, and communication assessment, cognitive and behavioral assessments, and academic assessment.

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Behavioral and Psychiatric Disorders (PEDHD-18)

Guideline

Behavioral and Psychiatric Disorders (PEDHD-18.1)
Reference (PEDHD-18)

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Behavioral and Psychiatric Disorders (PEDHD-18.1)

HDP.BD.0018.1.A

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- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450) is medically necessary for:
  - acute onset mental status change.
  - new or worsening focal neurologic findings.
  - presentation of acute psychiatric symptoms with comorbid serious medical illness.
  - non-auditory hallucinations (e.g., visual, tactile, olfactory) with no known etiology.
  - non-response to adequate medication trials.
  - symptoms of an organic brain disorder (e.g., focal deficits, severe headache, or seizures).
- For concerns of PANS (Pediatric acute-onset neuropsychiatric syndrome) and PANDAS (Pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection), see **Movement Disorders including Tourette Syndrome (PEDHD-26)**.

Background and Supporting Information

- Routine brain imaging is not routinely recommended for obsessive compulsive disorder (OCD), attention deficit hyperactivity disorder (ADHD), and autism spectrum disorder (ASD).

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Developmental Disorders (PEDHD-19)

Guideline

Intellectual Disability (PEDHD-19.1)
Cerebral Palsy (PEDHD-19.2)
Developmental Motor Delay (PEDHD-19.3)
Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)
References (PEDHD-19)

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Intellectual Disability (PEDHD-19.1)

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- Intellectual disability may be primary or secondary to a variety of heterogeneous disorders. See **Background and Supporting Information**.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for new or worsening focal neurologic findings and/or new or worsening cognitive decline.

Background and Supporting Information

- Intellectual disability is a condition characterized by significant limitations in both intellectual functioning and adaptive behavior that originates before the age of 22. One way to measure intellectual functioning is an IQ test. Generally, an IQ test score of around 70 or as high as 75 indicates a significant limitation in intellectual functioning.

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Cerebral Palsy (PEDHD-19.2)

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- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is medically necessary for:
  - initial evaluation of newly diagnosed cerebral palsy.
  - new or worsening focal neurologic findings documented on a physical examination, including the presence of developmental delay.
  - re-evaluation after 24 months of age due to rapid myelination during the first 2 years of life.
- For spinal imaging requests, see **Myelopathy (SP-7.1)** in the Spine Imaging Guidelines.

Background and Supporting Information

- Many individuals with intellectual disability also have cerebral palsy, but not all individuals with cerebral palsy have intellectual disability.
- Cerebral palsy is a static motor encephalopathy caused by a variety of entities spanning developmental, metabolic, genetic, infectious, ischemic, and other acquired etiologies.

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Developmental Motor Delay (PEDHD-19.3)

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary for:
  - initial evaluation of newly diagnosed developmental motor delay with abnormal muscle tone.
  - toe walking, when associated with upper motor neuron signs including hyperreflexia, abnormal tone (spasticity/hypotonia), or positive Babinski sign.
  - new or worsening focal neurologic findings.
  - repeat brain imaging one time after 24 months of age due to rapid myelination during the first 2 years of life.
- For spinal imaging requests, see **Myelopathy (SP-7.1)** in the Spine Imaging Guidelines and **Spinal Dysraphism and Tethered Spinal Cord (PEDSP-4)** in the Pediatric and Special Populations Spine Imaging Guidelines.

Background and Supporting information

- There are many causes for developmental motor delay. Individuals with motor delay can have decreased, normal, or increased muscular tone. Individuals with normal tone do not require imaging unless they have focal neurologic findings.

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Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)

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Medically Necessary Imaging Study	Indications	Suspected or Known Neurometabolic and/or Neurogenetic Disorders Including, but not limited to
MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **AND/OR** Magnetic Resonance Spectroscopy (MRS, CPT® 76390)	Requested by a neurologist or geneticist, or any provider in consultation with a neurologist or geneticist, for: • Initial evaluation **AND/OR** • Disease monitoring **AND/OR** • New or worsening symptoms **AND/OR** • Change in therapy is being considered	• X-linked adrenoleukodystrophy (X-ALD, CALD) • Alexander disease (ALX, AXD, dysmyelinogenic leukodystrophy) • Canavan disease • Creatine deficiency • Globoid cell leukodystrophy (Krabbe disease) • Hypomyelination and congenital cataract • Maple syrup urine disease • Megalencephalic leukoencephalopathy with subcortical cysts

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Medically Necessary Imaging Study	Indications	Suspected or Known Neurometabolic and/or Neurogenetic Disorders Including, but not limited to
MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **AND/OR** Magnetic Resonance Spectroscopy (MRS, CPT® 76390)	Requested by a neurologist or geneticist, or any provider in consultation with a neurologist or geneticist, for: • Initial evaluation **AND/OR** • Disease monitoring **AND/OR** • New or worsening symptoms **AND/OR** • Change in therapy is being considered	• Metachromatic leukodystrophy (MCL) • Mitochondrial disorders (such as, but not limited to, Leigh's syndrome, Kearns-Sayre syndrome, Mitochondrial encephalopathy with lactic acidosis and stroke-like episodes (MELAS)) • Nonketotic hyperglycinemia • Pelizaeus-Merzbacher disease (PMD) • Vanishing white matter (VWM) disease (Leukoencephalopathy with VWM, childhood ataxia with CNS hypomyelination (CACH) syndrome)

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Ataxia (PEDHD-20)

Guideline

Ataxia (PEDHD-20.1)
References (PEDHD-20)

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Ataxia (PEDHD-20.1)

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- See also **Developmental Disorders (PEDHD-19)**.
- A pertinent evaluation, including a detailed history and physical examination with a thorough neurologic examination, should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation or request is from or in consultation with a neurologist or neurosurgeon who has seen the individual since onset of symptoms.

Indications	Medically Necessary Imaging Study
• ANY of the following: ◦ Ataxia ◦ Hereditary ataxia ◦ Slowly progressive ataxia	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **OR** • CT Head without contrast (CPT® 70450) *If the initial CT or MRI study is unable to answer the clinical question at hand, an exam of the other modality is medically necessary if needed to direct clinical management.*
• Suspected spinal etiology	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) **OR** • MRI Lumbar Spine without and with contrast (CPT® 72158)

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Indications	Medically Necessary Imaging Study
• Acute ataxia following significant head trauma	• CT Head without contrast (CPT® 70450) **OR** • CT Head without and with contrast (CPT® 70470) **OR** • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)
• Contraindication to MRI	• CT Head without and with contrast (CPT® 70470) **OR** • CT Head with contrast (CPT® 70460
• BOTH of the following: ◦ Contraindication to MRI ◦ Suspected spinal etiology	• CT Cervical Spine without contrast (CPT® 72125) **OR** • CT Cervical Spine without and with contrast (CPT® 72127) **AND/OR** • CT Thoracic Spine without contrast (CPT® 72128) **OR** • CT Thoracic Spine without and with contrast (CPT® 72130) **AND/OR** • CT Lumbar Spine without contrast (CPT® 72131) **OR** • CT Lumbar Spine without and with contrast (CPT® 72133)

- Repeat imaging is medically necessary when ordered by a neurologist or neurosurgeon, or any provider in consultation with one of these providers, or if there are new signs or symptoms.

Background and Supporting Information

- Ataxia refers to an abnormally ill-coordinated or unsteady gait for age. "Limb ataxia" refers to impaired coordination (for age) of limbs, especially arms. Developmental failure to acquire the ability to walk is a form of developmental delay, not ataxia.
- CT should not be used in place of MRI solely to avoid sedation in young children because MRI is superior for imaging the posterior fossa.

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Epistaxis (PEDHD-21)

Guideline

Epistaxis Imaging (PEDHD-21.1)
References (PEDHD-21)

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Epistaxis Imaging (PEDHD-21.1)

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- Initial evaluation of epistaxis (nosebleed), including recurrent epistaxis that is refractory to medical management is by direct or endoscopic visualization of the relevant portions of the upper airway.
- If a mass lesion is detected on direct visualization, the following imaging studies are medically necessary:
  - CT Maxillofacial without contrast (CPT® 70486) **OR** CT Maxillofacial without and with contrast (CPT® 70488) **AND/OR**
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)

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Papilledema/Pseudotumor Cerebri (PEDHD-22)

Guideline

Papilledema/Pseudotumor Cerebri (PEDHD-22.1)

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Papilledema/Pseudotumor Cerebri (PEDHD-22.1)

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- Papilledema/Pseudotumor cerebri indications in pediatric individuals are identical to those for adult individuals. See **Papilledema/Pseudotumor Cerebri (HD-17.1)** in the Head Imaging Guidelines.

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Cranial Neuropathies (PEDHD-23)

Guideline

Cranial Neuropathies (PEDHD-23.1)

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Cranial Neuropathies (PEDHD-23.1)

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- See **Cranial Neuropathies (HD-31.1)** in the Head Imaging Guidelines.

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Pediatric Sleep Disorders (PEDHD-24)

Guideline

Pediatric Sleep Disorders (PEDHD-24.1)
References (PEDHD-24)

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Pediatric Sleep Disorders (PEDHD-24.1)

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- See **Pediatric Sleep Guidelines (SL-3)** in the Sleep Apnea and Treatment Clinical Guidelines.
- For over 18 years of age OR regarding oral appliance OR hypersomnolence, see **Sleep-Related Imaging Guidelines (HD-37)** in the Head Imaging Guidelines.
- For Obstructive Sleep Apnea:
  - Imaging prior to tonsillectomy (with or without adenoidectomy) for obstructive sleep apnea is not medically necessary.
  - Initially, endoscopic examination of the upper airway should be performed.
    - CT Maxillofacial without contrast (CPT® 70486) is medically necessary for evaluation of obstructive anatomy if operative intervention, other than a tonsillectomy (with or without adenoidectomy), is being considered.
- For suspected central sleep apnea, the following is medically necessary:
  - MRI Brain without contrast (CPT® 70551) OR
  - MRI Brain without and with contrast (CPT® 70553)
- Advanced imaging is NOT medically necessary for the following:
  - Confusional arousals
  - Sleep terrors
  - Nightmare disorder
  - Sleepwalking (somnambulism)
  - Bed wetting (enuresis)
  - Insomnia
  - Narcolepsy (without or with cataplexy)
  - Restless leg syndrome/periodic limb movement disorder
- For suspected sleep-related seizure imaging, see **Epilepsy and Other Seizure Disorders (PEDHD-6)**.

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Temporomandibular Joint (TMJ)/Dental/ Maxillofacial Imaging in Children (PEDHD-25)

Guideline

Temporomandibular Joint Imaging (PEDHD-25.1)
Dental/Periodontal/Maxillofacial Imaging (PEDHD-25.2)
References (PEDHD-25)

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Temporomandibular Joint Imaging (PEDHD-25.1)

HDP.TJ.0025.1.A

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- Temporomandibular Joint (TMJ) imaging indications in pediatric individuals are very similar to those for adult individuals. See **Temporomandibular Joint Disease (TMJ) (HD-30.1)** in the Head Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - There is a paucity of clinical symptoms and poor sensitivity of conventional x-rays in diagnosing TMJ arthritis in pediatric individuals with arthritis.
- MRI TMJ (CPT® 70336) is medically necessary annually for detecting silent TMJ arthritis in children with juvenile idiopathic arthritis (JIA) as requested by a rheumatologist and/or oral/maxillofacial surgeon (OMS) and/or any provider in consultation with a rheumatologist or OMS.
- Repeat imaging with MRI TMJ (CPT® 70336) in individuals with JIA is medically necessary for any of the following:
  - Change in signs or symptoms suggesting progression of disease
  - To monitor the effects of treatment
- Bone Scintigraphy/Bone Scan 3 Phase Study (CPT® 78315) in individuals over 12 years of age is medically necessary in anticipation or consideration of surgery.
- Unilateral condylar hyperplasia is manifested by slow growth in areas of the mandible causing facial asymmetry. It is usually a self-limiting condition seen predominantly in 12-30 year-olds.

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Dental/Periodontal/Maxillofacial Imaging (PEDHD-25.2)

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- See **Dental/Periodontal/Maxillofacial Imaging (HD 30.2)** in the Head Imaging Guidelines.

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Movement Disorders including Tourette Syndrome (PEDHD-26)

Guideline

Tourette Syndrome (PEDHD-26.1)
Movement Disorders (PEDHD-26.2)
References (PEDHD-26)

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Tourette Syndrome (PEDHD-26.1)

HDP.MD.0026.1.A

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- The diagnosis of Tourette syndrome is made clinically and advanced neuroimaging is not medically necessary for either diagnosis or management.

Background and Supporting Information

- Tourette syndrome (TS) is a neurological disorder characterized by repetitive, stereotyped, involuntary movements and vocalizations called tics. The first symptoms of TS are almost always noticed in childhood. Some of the more common tics include eye blinking and other vision irregularities, facial grimacing, shoulder shrugging, and head or shoulder jerking. Perhaps the most dramatic and disabling tics are those that result in self-harm such as punching oneself in the face, or vocal tics including coprolalia (uttering swear words) or echolalia (repeating the words or phrases of others). Many with TS experience additional neurobehavioral problems including inattention, hyperactivity and impulsivity, and obsessive-compulsive symptoms such as intrusive thoughts/worries and repetitive behaviors.

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Movement Disorders (PEDHD-26.2)

HDP.MD.0026.2.A

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- MRI Brain without contrast (CPT® 70551), **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary in the following clinical scenarios:
  - Atypical clinical features for example, movements that persist in sleep, onset outside of typical age at onset (4-6 years for tics), rapid progression, incomplete or uncertain medication responsiveness, clinical diagnostic uncertainty, limbic encephalitis
  - Dystonia, intermittent involuntary muscle contractions
  - Chorea, continual irregular movements
  - Ballism, involuntary high amplitude movements
  - Athetosis, slow writhing continuous movements
  - Myoclonus, involuntary muscle jerks (not sleep myoclonus)
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is medically necessary only after a complete medical workup including labs, acute infection, and other comorbid psychiatric disorders (examples, such as obsessive compulsive disorder (OCD), attention deficit hyperactivity disorder (ADHD), and autism spectrum disorder (ASD) have been investigated for concerns of:
  - PANS (pediatric acute-onset neuropsychiatric syndrome) **AND/OR**
  - PANDAS (pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection)
- See **Movement Disorders (HD-15.1)** in the Head Imaging Guidelines for the following:
  - Suspected Huntington disease
  - Evaluation for surgical treatment of essential tremor or Parkinson's disease, including deep brain stimulator (DBS) placement

Background and Supporting Information

- Movement disorders are hyperkinetic and hypokinetic movements that are involuntary. The majority are diagnosed based on a clinical diagnosis and do not require imaging.
- Typically, benign movement disorders include:
  - Stereotypies, repetitive rhythmic movements
  - Tics that are vocal or motor with typical onset and course
  - Tourette syndrome
  - Essential tremor or tremors of anxiety or weakness
  - Restless leg syndrome

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- Routine brain imaging is not routinely recommended for OCD, ADHD, or ASD.
- There is little evidence to support the use of MRA/CTA and PET in the evaluation of movement disorders.

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Tuberous Sclerosis (PEDHD-27)

Guideline

Tuberous Sclerosis (PEDHD-27.1)

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Tuberous Sclerosis (PEDHD-27.1)

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- See **Tuberous Sclerosis Complex (TSC) (PEDONC-2.9)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Von Hippel-Lindau Syndrome (VHL) (PEDHD-28)

Guideline

Von Hippel-Lindau Syndrome (VHL) (PEDHD-28.1)

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Von Hippel-Lindau Syndrome (VHL) (PEDHD-28.1)

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- See **Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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CNS Infection (PEDHD-29)

Guideline

CNS Infection (PEDHD-29.1)
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CNS Infection (PEDHD-29.1)

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- CNS infection imaging indications in pediatric individuals are similar to those for adult individuals. See **CNS and Head Infection (HD-14.1)** in the Head Imaging Guidelines.
- For COVID-19 infection, see **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD 14.2)** in the Head Imaging Guidelines.
- For Multisystem Inflammatory Syndrome in Children (MIS-C), see **COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)**.
- For autoimmune/paraneoplastic encephalitis, see **Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)** in the Head Imaging Guidelines.
- For pediatric-specific imaging considerations, including suspected congenital brain infection and neonatal meningitis, the following is medically necessary:
  - Ultrasound Head (CPT© 76506) **AND/OR**
  - CT Head without contrast (CPT® 70450) **OR** CT Head without and with contrast (CPT® 70470)

    AND/OR
  - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
- Newborn infants with microcephaly should be evaluated as discussed in **Macrocephaly, Microcephaly, and Hydrocephalus (PEDHD-7).**

Background and Supporting Information

- Neonatal meningitis most often is caused by bacterial pathogens and usually occurs as a complication of sepsis in the first week of life. In older infants and children, meningeal inoculation occurs secondary to hematogenous spread or penetrating trauma.
- The common causes of prenatal infections of the central nervous system are cytomegalovirus, *Toxoplasma gondii*, herpes simplex type 2 virus, and most recently, zika virus. The findings suggesting prenatal brain infection include microcephaly, microphthalmia, chorioretinitis, cataracts, hypotonia, and seizures.

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Scalp and Skull Lesions (PEDHD-30)

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Scalp and Skull Lesions (PEDHD-30.1)

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- Scalp and skull lesion imaging indications (including Pott Puffy Tumor) in pediatric individuals are identical to those for adult individuals with the exception of neonates. See **Scalp and Skull Lesions (HD-20.1)** in the Head Imaging Guidelines.
- For neonates with palpable scalp and skull lesions, the following is medically necessary:
  - Ultrasound Head (CPT® 76506) **AND/OR**
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - CT Head without and with contrast (CPT® 70470)

Background and Supporting Information

- In neonates and young infants, scalp masses include the following:
  - Congenital lesions (cephalocele, dermoid cyst, epidermoid cyst)
  - Vascular lesions (hemangioma, sinus pericranii)
  - Extracranial hemorrhage related to birth trauma (caput succedaneum, cephalohematoma, subgaleal hematoma)
  - After the first year of life, malignant tumors, such as Langerhans cell histiocytosis, as well as metastases from neuroblastoma and rhabdomyosarcoma are additional causes of a scalp mass.

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Eye Disorders (PEDHD-31)

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Eye Disorders (PEDHD-31.1)
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Eye Disorders (PEDHD-31.1)

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- Eye disorder imaging indications in pediatric individuals are close to identical to those for adult individuals. See **Eye Disorders and Visual Loss (HD-32.1)** in the Head Imaging Guidelines.

Indication	Medically Necessary Imaging Study
◦ Evaluation for congenital disorders or disorders that begin early in life, such as, but not limited to the following: ▪ Optic nerve hypoplasia ▪ Septo-optic dysplasia ▪ Infantile nystagmus syndrome	◦ MRI Orbits/Face/Neck without contrast (CPT® 70540) **OR** ◦ MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** ◦ CT Orbits/Temporal Bone with contrast (CPT® 70481) **OR** ◦ CT Orbits/Temporal Bone without contrast (CPT® 70480) **AND/OR** ◦ MRI Brain without contrast (CPT® 70551) **OR** ◦ MRI Brain without and with contrast (CPT® 70553)

- Repeat imaging is medically necessary if requested by a neurologist, ophthalmologist, neuro-ophthalmologist, or neurosurgeon, or in consultation with one of these providers.
- For traumatic retinal hemorrhages as seen in suspected shaken baby syndrome, see **Head Trauma (PEDHD-4.1)**.
- For suspected and/or confirmed neuroblastoma, see **Neuroblastoma (PEDONC-6)**.
- For orbital cellulitis, see **Eye Disorders and Visual Loss (HD-32.1)** and/or **Imaging in Sinusitis (PEDHD-5.2)**.

Background and Supporting Information

- Neuroblastoma may be suspected in the setting of disorders, such as, but not limited to, Opsoclonus-Myoclonus syndrome, Horner's syndrome, and/or choroidal metastases.

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General Guidelines (PEDMS-1.0)

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Procedure Codes Associated with Musculoskeletal Imaging (PEDMS)

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MRA	CPT®
MRA Upper Extremity	73225
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Nuclear Medicine	CPT®
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Nuclear Medicine	CPT®
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Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (eg, head, neck, chest, pelvis), single day imaging	78830
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Complete bilateral noninvasive physiologic studies of upper or lower extremity arteries	93923
Duplex scan of upper extremity arteries or arterial bypass grafts; complete bilateral	93930
Duplex scan of upper extremity arteries or arterial bypass grafts; unilateral or limited	93931
Duplex scan of extremity veins including responses to compression and other maneuvers; complete bilateral study	93970
Duplex scan of extremity veins including responses to compression and other maneuvers; unilateral or limited study	93971
Duplex scan of hemodialysis access (including arterial inflow, body of access and venous outflow)	93990

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General Guidelines (PEDMS-1.0)

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- A pertinent clinical evaluation including a detailed history, physical examination, appropriate laboratory studies and basic imaging such as plain x-ray or ultrasound should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) can serve as a pertinent clinical evaluation.
- Plain x-ray should be done prior to advanced imaging. The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study. X-ray can rule out those situations that do not require advanced imaging, such as acute/healing fracture, osteomyelitis, and tumors of bone amenable to biopsy or radiation therapy (in known metastatic disease), etc.
  - Even in soft tissue masses, plain x-rays are helpful in evaluating for calcium/bony deposits, e.g. myositis ossificans and invasion of bone.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the same body area are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.
- Provider-directed conservative care may include any or all of the following: R.I.C.E (rest, ice, compression, and elevation), NSAIDs (non-steroidal anti-inflammatory drugs), narcotic and non-narcotic analgesic medications, oral or injectable corticosteroids, viscosupplementation injections, a provider-directed home exercise program, cross-training, physical medicine, or immobilization by splinting/casting/bracing.
- These guidelines are based upon using advanced imaging to answer specific clinical questions that will affect patient management. Imaging is not indicated if the results will not affect individual management decisions. Standard medical practice would dictate continuing conservative therapy prior to advanced imaging in individuals who are improving on current treatment programs.

Health Equity Consideration

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence;

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education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Age Considerations (PEDMS-1.1)

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- Many conditions affecting the musculoskeletal system in the pediatric population have different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are ≤18 years old should be imaged according to the Pediatric Musculoskeletal Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Musculoskeletal Imaging Guidelines should be imaged according to the General Musculoskeletal Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Musculoskeletal Imaging Guidelines except where directed otherwise by a specific guideline section.

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Appropriate Clinical Evaluation and Conservative Treatment (PEDMS-1.2)

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- See: **General Guidelines (PEDMS-1.0)**

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Modality General Considerations (PEDMS-1.3)

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- MRI
  - MRI without contrast is the preferred modality for pediatric musculoskeletal imaging unless otherwise stated in a specific guideline section, as it is superior in imaging the soft tissues and can also define physiological processes in some instances, e.g. edema, loss of circulation (AVN), and increased vascularity (tumors).
  - MRI without and with contrast is frequently recommended for evaluation of tumors, infection, post-operative evaluation, arthrography, and juvenile idiopathic arthritis, as described in the disease-specific guideline sections.
  - Due to the length of time required for MRI acquisition and the need to minimize movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years), as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous route. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast as supported by these condition-based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported by these guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same imaging session.

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- The presence of surgical hardware or implanted devices may preclude MRI, as magnetic field distortion may limit detail in adjacent structures. CT may be the procedure of choice in these cases.
- The selection of best examination may require coordination between the provider and the imaging service.

- CT
  - CT without contrast is generally superior to MRI for imaging bone and joint anatomy; thus it is useful for studying complex fractures (particularly of the joints, dislocations, and assessing delayed union or non-union of fractures, integration of bone graft material, if plain x-rays are equivocal.
    - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
  - CT beam attenuation can result in streak artifact which can obscure adjacent details. This can occur with radiopaque material such as metal objects or dense bones.
  - The selection of best examination may require coordination between the requesting provider and the rendering imaging facility.
- Ultrasound
  - Ultrasound is frequently used to evaluate infants for hip dysplasia, to detect and/or aspirate joint effusion, and as an initial evaluation of extremity soft tissue masses.
  - CPT® codes vary by body area and the use of Doppler imaging. These CPT® codes are included in the table at the beginning of this guideline.
- Nuclear Medicine
  - Nuclear medicine studies are commonly used in evaluation of the peripheral musculoskeletal system, and other rare indications exist as well:
    - Bone scan (CPT® 78315), Distribution of Radiopharmaceutical Agent SPECT (CPT® 78803, or 78831), or SPECT/CT (CPT® 78830) is indicated for evaluation of suspected loosening of orthopedic prostheses when recent plain x-ray is nondiagnostic.
    - Nuclear medicine bone marrow imaging (CPT® codes: CPT® 78102, CPT® 78103, or CPT® 78104), SPECT (CPT® code: 78803), or SPECT/CT (CPT® 78830) is indicated for detection of ischemic or infarcted regions in sickle cell disease.
    - Triple phase bone scan (CPT® 78315) is indicated for evaluation of complex regional pain syndrome or reflex sympathetic dystrophy.
- 3D Rendering
  - 3D Rendering indications in pediatric musculoskeletal imaging are identical to those in the general imaging guidelines. See: **3D Rendering (MS-3)** for imaging guidelines.
- Bilateral Imaging

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- Coding for bilateral imaging may vary from health care plan to health care plan. Not all coding options may be available for all health care plans.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Fracture and Dislocation (PEDMS-2)

Guideline

Fracture and Dislocation (PEDMS-2)
Acute Fracture (PEDMS-2.1)
Joint-Adjacent Fracture (PEDMS-2.2)
Growth Plate Injuries (Salter-Harris Fractures) (PEDMS-2.3)
Osteochondral or Chondral Fractures, Including Osteochondritis Dissecans (PEDMS-2.4)
Stress/Occult Fracture (PEDMS-2.5)
Compartment Syndrome (PEDMS-2.6)
Physical Child Abuse (PEDMS-2.7)
References (PEDMS-2)

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Fracture and Dislocation (PEDMS-2)

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- A pertinent clinical evaluation including a detailed history, physical examination, and plain x-ray should be performed prior to considering advanced imaging.

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Acute Fracture (PEDMS-2.1)

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- Plain x-rays should be performed initially in any obvious or suspected acute fracture or dislocation.
  - If plain x-rays are positive, no further imaging is medically necessary except in complex (comminuted or displaced) joint fractures where MRI or CT without contrast is medically necessary for preoperative planning.
  - 3D Rendering may sometimes be medically necessary for complex fracture repairs. See: **3D Rendering (MS-3)** in the Musculoskeletal Imaging Guidelines.
- Ultrasound (CPT® 76881 or CPT® 76882) is medically necessary for evaluation of fracture, but is not required to allow for other advanced imaging, especially in infants. Ultrasound is typically a secondary method of fracture detection when used, though some centers use it as the sole imaging modality for skull and clavicle fractures.
- CT or MRI without contrast is medically necessary if plain x-rays are negative or equivocal for fracture, and fracture or bone marrow edema is still clinically suspected, and if the results will determine immediate treatment decisions as documented by the treating physician.
- Bone scan is medically necessary for evaluation of suspected fracture when two x-rays are negative at least 10 days apart, using any of the following CPT® code combinations:
  - CPT® 78300, CPT® 78305, or CPT® 78306 as a single study
  - See: **Stress/Occult Fracture (PEDMS-2.5)** for bone scan indications

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Joint-Adjacent Fracture (PEDMS-2.2)

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- CT without contrast is medically necessary in complex (comminuted or displaced) fractures seen on plain x-ray involving a joint for preoperative planning.
- CT without contrast is medically necessary when there is clinical concern for delayed union or non-union of fracture or joint fusions on follow-up plain x-ray.

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Growth Plate Injuries (Salter-Harris Fractures) (PEDMS-2.3)

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- These fractures can generally be diagnosed and managed adequately with plain x-ray.
- In case of severe injury with displacement of bone fractures seen on plain x-ray, CT without contrast is medically necessary prior to surgical intervention.
- If there is concern for delayed union or non-union of the bone seen on plain x-ray, CT without contrast is medically necessary.
- MRI without contrast is medically necessary for the evaluation of a suspected physeal bar in a healing fracture or other complication of a fracture involving the growth plate seen on plain x-ray or CT which may result in abnormal growth. While physeal bars may be seen on CT, some fibrous physeal bars can be missed on CT. As such, MRI is the preferred imaging modality.
- Compressive injuries of the growth plate (Salter-Harris V) injuries may be difficult to identify on plain films, and MRI without contrast is medically necessary for confirmation.

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Osteochondral or Chondral Fractures, Including Osteochondritis Dissecans (PEDMS-2.4)

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An osteochondral fracture is a tear of the cartilage which covers the end of a bone, within a joint. It is also known as Osteochondritis Dissecans. In both disorders, the osteochondral fragment may separate from the articular surface and form loose bone fragments in a joint.

- If x-rays are negative and an osteochondral fracture is still suspected, or if x-ray or clinical exam suggests an unstable osteochondral injury, either MRI without contrast, MR arthrogram, or CT arthrogram of the involved joint is medically necessary.
- If plain x-rays show a non-displaced osteochondral fragment, follow up imaging should be with plain x-rays. Advanced imaging is not medically necessary.
- MRI without contrast or CT without contrast is medically necessary when healing cannot be adequately assessed on follow up plain x-rays.

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Stress/Occult Fracture (PEDMS-2.5)

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- These fractures can usually be adequately evaluated by history, physical exam, and x-ray. Advanced imaging may be medically necessary as discussed below if the initial evaluation of history, physical exam, and plain x-ray fails to establish a definitive diagnosis.
- Plain x-rays should be performed before advanced imaging. Plain x-rays are often negative initially, but may become positive after 14 days.
- If stress or occult fracture is suspected involving the pelvis, sacrum, hip, femur, tibia, tarsal navicular, proximal 5th metatarsal, or scaphoid, and initial plain x-ray fails to establish a definitive diagnosis:
  - MRI or CT without contrast is medically necessary, without conservative care or follow-up plain x-rays OR
  - Bone scan (CPT® 78315, 78306, or 78300), SPECT/CT (CPT® 78830), or Distribution Of Radiopharmaceutical Agent SPECT (CPT® 78803) is medically necessary in place of MRI or CT if provider requests
- For all other suspected stress or occult fractures, if follow-up plain x-rays are negative after 10 days of conservative care, or initial non-diagnostic x-ray is obtained a minimum of 14 days after the onset of symptoms:
  - MRI or CT without contrast is medically necessary OR
  - Bone scan (CPT® 78315, 78306, or 78300), SPECT/CT (CPT® 78830), or Distribution Of Radiopharmaceutical Agent SPECT (CPT® 78803) is medically necessary in place of MRI or CT if provider requests
- Periodic follow-up plain x-rays will usually show progressive healing.
  - CT without contrast is medically necessary when there is clinical concern for non-union.

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Compartment Syndrome (PEDMS-2.6)

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- Acute compartment syndrome is a clinical diagnosis made by direct measurement of compartment pressure and is a surgical emergency. Advanced imaging is not medically necessary.
- See: Compartment syndrome within the **Muscle and Tendon Injuries (MS-11.0)** section of the Musculoskeletal Imaging Guidelines.

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Physical Child Abuse (PEDMS-2.7)

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- See: **Suspected Physical Child Abuse (PEDMS-7)** for imaging guidelines.

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Soft Tissue and Bone Masses (PEDMS-3)

Guideline

Soft Tissue and Bone Masses – General Considerations (PEDMS-3.1)
Soft Tissue Mass with Negative X-ray and Abnormal Ultrasound (PEDMS-3.2)
Soft Tissue Mass with Calcification/Ossification on X-ray (PEDMS-3.3)
Mass Involving Bone (Including Suspected Lytic and Blastic Metastatic Disease) (PEDMS-3.4)
References (PEDMS-3)

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Soft Tissue and Bone Masses – General Considerations (PEDMS-3.1)

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- A pertinent clinical evaluation including a detailed history and physical examination should be performed prior to considering advanced imaging.
  - History and physical exam of any palpable soft tissue mass should include documentation of any of the following pertinent clinical features: location, size, and any association with pain.
- Plain x-rays should be performed as initial imaging. This is true even for soft tissue masses that are clearly not directly associated with osseous structures. Details such as soft tissue calcification, presence or absence of phleboliths, radiographic density, and any effect on adjacent bone are all potentially significant plain film findings that may help better identify the etiology of the mass and determine the optimal modality and contrast level when advanced imaging is indicated.
- Evaluation by a surgical specialist or oncologist is strongly recommended to help determine the most helpful advanced imaging studies for an individual.
- Ultrasound (CPT® 76881 or CPT® 76882) is medically necessary if initial plain x-ray is negative to evaluate:
  - ill-defined masses or areas of swelling
  - hematomas
  - subcutaneous lipomas with inconclusive clinical examination
  - lipomas in other locations
  - masses that have been present and stable for ≥1 year
  - vascular malformations (see: **Vascular Anomalies (PEDPVD-2)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines)
- MRI without and with contrast or without contrast is medically necessary for any of the following:
  - soft tissue mass >5 cm in diameter
  - soft tissue mass increasing in size
  - painful soft tissue mass
  - deep soft tissue mass or subfascial location
- Advanced imaging is not medically necessary for the following entities:
  - ganglion cysts
  - sebaceous cysts
  - hematomas
  - subcutaneous lipomas
    - MRI without or without and with contrast can be performed if surgery is planned.

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- MRI without and with contrast, or ultrasound (CPT® 76881 or CPT® 76882) is medically necessary for lipomas in other locations (not subcutaneous).

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Soft Tissue Mass with Negative X-ray and Abnormal Ultrasound (PEDMS-3.2)

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- MRI without and with contrast is medically necessary when plain x-ray is negative and ultrasound is abnormal.
  - CT without or with contrast is medically necessary if MRI is contraindicated.

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Soft Tissue Mass with Calcification/ Ossification on X-ray (PEDMS-3.3)

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- MRI without and with contrast is medically necessary when calcification/ossification is noted on plain x-ray.
  - CT without or with contrast is medically necessary if MRI is contraindicated.

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Mass Involving Bone (Including Suspected Lytic and Blastic Metastatic Disease) (PEDMS-3.4)

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- Plain x-rays of the entire bone containing the lesion are required prior to consideration of advanced imaging. Many benign bone tumors have a characteristic appearance on plain x-ray and advanced imaging is not medically necessary unless one of the following applies:
  - MRI without and with contrast and/or CT without is medically necessary for preoperative planning.
  - MRI without and with contrast is medically necessary when the diagnosis is uncertain based on plain x-ray appearance.
    - CT without or with contrast is medically necessary if MRI is contraindicated.
- Surveillance of benign bony lesions is with plain x-ray
  - MRI without and with contrast is medically necessary for new findings on x-ray, or new or worsening clinical symptoms not explained by recent x-ray.
- Osteochondroma, osteoid osteoma, osteogenic sarcoma, and Ewing sarcoma family of tumors should be imaged according to **Bone Tumors (PEDONC-9)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- If there is concern for metastatic disease in an individual with a known malignancy, refer to the appropriate Pediatric and Special Populations Oncology Imaging Guideline.

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Limping Child (PEDMS-4)

Guideline

General Evaluation of the Limping Child (PEDMS-4.1)
Limping Child with Suspected Trauma (PEDMS-4.2)
Limping Child with Suspected Infection (PEDMS-4.3)
Limping Child with No Evidence of Trauma or Infection (PEDMS-4.4)
References (PEDMS-4)

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General Evaluation of the Limping Child (PEDMS-4.1)

MSP.LC.0004.1.A

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- This guideline primarily applies to children under the age of 6 years. It may also be applied to older children with pre-existing conditions who may not be able to communicate, such as a child with severe intellectual disability. Many of these cases will be urgent, because of the risk of adverse outcomes in delay of diagnosis.
- A pertinent clinical evaluation, including a detailed history and physical examination, should be performed, which will help determine any indication for advanced imaging. Based on this clinical evaluation, the most likely etiology should be determined, usually trauma, infection, or neither trauma nor infection.
- X-ray should be obtained if there are no localized findings on physical examination.

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Limping Child with Suspected Trauma (PEDMS-4.2)

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- Plain x-rays are indicated for detection of fractures, destructive lesions, and avascular necrosis. For children under age 4 this may require x-rays of the entire leg from hip to foot. If clinical suspicion is high for “toddler fracture” imaging may start with tibia/fibula x-rays, and if a fracture is demonstrated, additional imaging may not be required.
- If initial x-rays are negative, but limping symptoms or avoidance of weight-bearing persist, follow-up x-rays in 7 to 10 days are indicated.
  - If plain films are negative and suspicion remains high for stress fractures or soft tissue injury, the following are medically necessary:
    - MRI without contrast of the affected body area OR
    - Radionuclide bone scan (CPT® 78300, CPT® 78305, CPT® 78306, or CPT® 78315), SPECT/CT (CPT® 78830), or SPECT (CPT® 78803) if implanted hardware or devices precluding MRI are present.
- CT use is limited in the evaluation of the limping child with suspected trauma.

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Limping Child with Suspected Infection (PEDMS-4.3)

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- Pain localized to hip:
  - It is essential to exclude septic arthritis. Ultrasound of the hip (CPT® 76881 or 76882) is used to exclude hip joint effusion.
    - Hip joint fluid aspiration to distinguish infection from non-infectious etiologies If hip joint effusion is demonstrated.
    - Plain x-rays should be obtained if no hip joint effusion is demonstrated.
    - MRI without contrast (CPT® 73721) or without and with contrast (CPT® 73723) is medically necessary if plain films are not diagnostic.
- Pain localized distal to hip:
  - MRI without contrast or without and with contrast of the affected body part is medically necessary if plain x-rays are not diagnostic.
- Non-localized pain:
  - Plain x-rays of the spine, pelvis, and lower extremities may be necessary to localize the abnormality.
  - If plain x-ray is not diagnostic and suspicion for infection remains high, one of the following:
    - Whole-body bone scan (CPT® 78306) OR
    - SPECT (CPT® 78803) OR
    - SPECT/CT (CPT® 78830) OR
    - MRI without contrast or without and with contrast of the affected body area

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Limping Child with No Evidence of Trauma or Infection (PEDMS-4.4)

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- This differential diagnosis is quite broad.
  - Transient (or toxic) synovitis of the hip:
    - Ultrasound of the hip (CPT® 76881 or CPT® 76882) is the medically necessary initial exam.
      - Plain x-rays if no hip effusion is demonstrated.
      - Hip joint fluid aspiration is indicated if a hip joint effusion is demonstrated. This is usually performed with US guidance, though fluoroscopic guidance or blind aspiration may be required.
  - Avascular Necrosis, see: **Avascular Necrosis (AVN)/ Legg-Calvé-Perthes Disease (PEDMS-6)**
  - Juvenile Idiopathic Arthritis, see: **Juvenile Idiopathic Arthritis (PEDMS-10.1)**
  - Histiocytic Disorders, see: **Histiocytic Disorders (PEDONC-18)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - Neoplasm, see: **General Guidelines (PEDONC-1)**, **Pediatric Leukemias (PEDONC-3)**, **Neuroblastoma (PEDONC-6)**, **Pediatric Soft Tissue Sarcomas (PEDONC-8)**, or **Bone Tumors (PEDONC-9)** in the Pediatric and Special Populations Oncology Imaging Guidelines
  - Child abuse, see: **Suspected Physical Child Abuse (PEDMS-7)**

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Developmental Dysplasia of the Hip (PEDMS-5)

Guideline

Developmental Dysplasia of the Hip (PEDMS-5)
References (PEDMS-5)

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Developmental Dysplasia of the Hip (PEDMS-5)

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Screening studies

- The routine use of ultrasound in screening neonates and infants without risk factors for developmental dysplasia of the hip (DDH) is not recommended by the American Academy of Pediatrics and the American Academy of Orthopedic Surgeons.
- Screening ultrasound (CPT® 76885 or CPT® 76886) is medically necessary for infants between 4 weeks of age and 4 months of age with one or more of the following risk factors:
  - Breech presentation
  - Family history of DDH
  - Abnormal hip exam (e.g., positive Ortolani or Barlow maneuvers, asymmetric thigh folds, shortening of the thigh observed on the dislocated side, limitation of hip abduction)
- For children between 4 and 6 months of age plain x-ray is the initial imaging modality as femoral head ossification is often seen on x-ray in normal individuals.
  - If x-ray is inconclusive, ultrasound (CPT® 76885 or CPT® 76886) is medically necessary.

Follow-Up/Diagnostic Studies

- Ultrasound is medically necessary earlier than 4 weeks of age in individuals with unstable/dislocated hip(s) undergoing abduction brace treatment.
- Follow-up hip ultrasound (CPT® 76885 or CPT® 76886) is medically necessary for the following:
  - Graf type IIA hip with an alpha angle (bony angle) between 50 to 59 degrees in a child less than 3 months of age and follow up hip ultrasound is requested to confirm normal development
  - Subluxation or dislocation was diagnosed on previous hip ultrasound using the dynamic Harke imaging method
  - Prior ultrasound demonstrates abnormal hip and treatment has been applied (such as a Pavlik harness or other device) to document effectiveness of treatment, to ensure the femoral head remains located in the acetabulum, or to identify treatment failure.
    - The usual interval for follow-up sonography is monthly, but earlier imaging is medically necessary for clinical suspicion of treatment failure, subluxation or dislocation of the hip.

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- MRI without and with contrast (CPT® 73723), MRI without contrast (CPT® 73721), or CT without contrast (CPT® 73700) is medically necessary to evaluate alignment following reduction. Children in casts or following surgery may require repeated advanced imaging to ensure the reduction remains satisfactory, or to assess incorporation of bone graft material.
- Hip ultrasound is NOT medically necessary for the following:
  - Infants older than 6 months of age as plain x-ray of the hips become more reliable due to femoral head ossification and should be used in infants over 6 months of age.
  - Type I, IIB, IIC, IID, and III hips diagnosed on a previous hip ultrasound using the Graf method. Type I hip is normal, and Type IIB, IIC, IID, and III will have imaging as directed by treating provider.
  - Plain x-ray of the hips should be performed rather than ultrasound if there is a clinical suspicion for teratogenic dysplasia.

Background and Supporting Information

- Developmental dysplasia of the hip (DDH) was formerly known as congenital dislocation of the hip. DDH includes a spectrum of abnormalities including abnormal acetabular shape (dysplasia) and malposition of the femoral head ranging from reducible subluxation to irreducible subluxation or dislocation of the femoral head. 60 to 80% of abnormalities are identified by physical exam, and more than 90% are identified by ultrasound. Treatment may involve placement in a Pavlik harness, casting, or surgery in extreme or refractory cases.
- Hip laxity is normal after birth and usually resolves spontaneously.
- There are two sonographic methods of evaluating the hip: the dynamic stress (Harcke) technique and the static (Graf) technique
- The overwhelming majority of Graf type IIA hips mature spontaneously but follow up may be required to ensure that maturation has occurred.

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Avascular Necrosis (AVN) / Legg-Calvé-Perthes Disease / Idiopathic Osteonecrosis (PEDMS-6)

Guideline

Avascular Necrosis and Legg-Calvé-Perthes Disease (PEDMS-6.1)
Osteonecrosis (PEDMS-6.2)
References (PEDMS-6)

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Avascular Necrosis and Legg-Calvé-Perthes Disease (PEDMS-6.1)

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- Plain x-ray is the initial imaging study and may be all that is necessary for follow-up.
- MRI Hip without contrast (CPT® 73721) or MRI Hip without and with contrast (CPT® 73723) is medically necessary if the diagnosis is uncertain on plain x-ray, or for preoperative planning.
  - If MRI is contraindicated or unavailable, any one of the following studies is medically necessary in lieu of MRI:
    - CT scan without contrast, OR
    - Nuclear bone scan (CPT® codes: 78300, 78305, 78306, or 78803) OR
    - SPECT/CT (CPT® 78830)

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Osteonecrosis (PEDMS-6.2)

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- Osteonecrosis can occur in a number of conditions, including during treatment for developmental dysplasia of the hip.
- Individuals with acute lymphoblastic leukemia, lymphoblastic lymphoma, or other conditions with recurrent exposure to high dose corticosteroids and known or suspected osteonecrosis should be imaged according to guidelines in: **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- Known or suspected osteonecrosis in long-term cancer survivors should be imaged according to guidelines in: **Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)** in the Pediatric and Special Populations Oncology Imaging Guidelines.
- X-ray is indicated as initial imaging study
- MRI either without contrast or without and with contrast is medically necessary in other individuals with concern for osteonecrosis and negative or inconclusive recent x-ray, if imaging results will change current individual management. Early phase of osteonecrosis may be seen on MR with normal x-ray findings.
  - CT scan without contrast is medically necessary for surgical planning.

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Suspected Physical Child Abuse (PEDMS-7)

Guideline

Suspected Physical Child Abuse (PEDMS-7)
References (PEDMS-7)

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Suspected Physical Child Abuse (PEDMS-7)

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The suspicion of physical abuse of a child often requires imaging, both for clinical management and for forensic purposes. Every effort should be made to support reasonable requests for imaging in these children.

Child abuse injuries may affect any organ or system. Fractures are common, but injuries may also involve solid and hollow visceral organs, and/or superficial and deep soft tissue injuries. Some fracture patterns are highly correlated with non-accidental mechanisms, such as the classic metaphyseal lesion, also known as a corner fracture or bucket handle fracture, but fractures may occur in any bone. Unsuspected fractures, multiple fractures at various stages of healing, or fractures of a configuration or distribution inconsistent with the history provided, may raise the suspicion for physical abuse.

Skeletal Injury

- The x-ray skeletal survey is the primary imaging procedure for detecting fractures, especially in children age 24 months or younger. In older children, skeletal survey may be indicated, but more tailored x-ray evaluation based on history and physical examination may be preferable to skeletal survey.
- When skeletal survey is negative, but clinical suspicion remains high, either of the following are medically necessary:
  - Bone scan (CPT® codes: CPT® 78300, 78305, 78306, 78315, or 78830) OR
  - Distribution of Radiopharmaceutical Agent SPECT (CPT® 78803)
- Suspected injury to the spine should usually first be evaluated with plain x-rays. CT without contrast and/or MRI without contrast or without and with contrast may be required for complete evaluation of osseous and soft tissue spine injuries. If requested for suspected or known physical abuse, both CT without contrast and/or MRI without contrast or without and with contrast of suspected sites are medically necessary.
- CT Chest without contrast (CPT® 71250) is medically necessary in individuals with a negative skeletal survey and a high clinical suspicion for rib fracture associated with child abuse.
- A repeat skeletal survey performed approximately 2 weeks after the initial examination can provide additional information on the presence and age of child abuse fractures and should be performed when abnormal or equivocal findings are found on the initial study and when abuse is suspected on clinical grounds

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Head Injury

- CT Head without contrast (CPT® 70450) is medically necessary when there is clinical evidence of head injury or when skull fracture of any age is detected on survey skull x-ray.
  - CT Head without contrast (CPT® 70450) is also medically necessary when known or suspected cervical trauma is present in a pediatric individual.
  - CT Head without contrast (CPT® 70450) is medically necessary in individuals less than 1 year of age, even if no neurologic symptoms are detected due to the great potential morbidity of abuse head trauma. MRI Brain without contrast (CPT® 70551) is also medically necessary.
  - MRI Spine without contrast (CPT® 72141, 72146, 72148) or without and with contrast (CPT® 72156, 72157, 72158) is medically necessary when there is clinical evidence of head injury or when skull fracture of any age is detected on survey skull x-ray. CT Spine (CPT® 72125, CPT® 72128, CPT® 72131) is medically necessary if MRI is not readily available.
- MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is medically necessary to evaluate brain parenchymal injury, or in a child where the clinical signs of brain injury are not sufficiently explained by CT findings.

Other Body Area Injuries

- CT should be performed with contrast unless an absolute contraindication exists.
- ANY of the following imaging studies are medically necessary for suspected injury to the abdomen or pelvis:
  - Abdominal ultrasound (CPT® 76700)
  - Pelvic ultrasound (CPT® 76856)
  - CT Abdomen with contrast (CPT® 74160)
  - CT Pelvis with contrast (CPT® 72193)
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
- ANY of the following imaging studies are medically necessary for suspected injury to the chest:
  - CT Chest without contrast (CPT® 71250)
  - CT Chest with contrast (CPT® 71260)

Screening of other children

- Contacts are defined as the asymptomatic siblings, cohabiting children, or children under the same care as an index child with suspected child physical abuse. All contact children should undergo a thorough physical examination and a history elicited prior to imaging. Contact children younger than 12 months should have neuroimaging, and skeletal survey. CT Head without contrast (CPT® 70450) or MRI Brain without contrast (CPT® 70551) is medically necessary. Contact children aged

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12 to 24 months should undergo skeletal survey. No routine imaging is medically necessary in asymptomatic children older than 24 months.

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Infection/Osteomyelitis (PEDMS-8)

Guideline

Infection/Osteomyelitis (PEDMS-8)
References (PEDMS-8)

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Infection/Osteomyelitis (PEDMS-8)

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- Infection and osteomyelitis imaging indications in pediatric individuals are similar to those for adult individuals other than the limping child.
  - See: **General Infection/Osteomyelitis (MS-9.1)** and **Charcot and Septic Joint (MS-9.2)** in the Musculoskeletal Imaging Guidelines for circumstances other than in the limping child.
  - See: **Limping Child with Suspected Infection (PEDMS-4.3)** for imaging guidelines when limping is present.
  - See: **Inflammatory Musculoskeletal Disease (PEDMS-10)** for imaging guidelines for chronic recurrent multifocal osteomyelitis (CRMO, which is an autoimmune disease).
- Ultrasound of the involved extremity (CPT® 76881 or CPT® 76882) is medically necessary to evaluate for effusion or soft tissue fluid collection
  - Ultrasound is not a prerequisite for other advanced imaging studies
- Bone scan (CPT® 78300, 78305, 78306, or 78315), SPECT/CT (CPT® 78830, or 78832), or SPECT (CPT® 78803, or 78831) is medically necessary for evaluation of suspected bone infection if MRI cannot be done and when infection is multifocal, or when the infection is associated with orthopedic hardware or chronic bone alterations from trauma or surgery. Combining bone scintigraphy with a labeled leukocyte scan enhances sensitivity. A labeled leukocyte scan (radiopharmaceutical localization of tumor, inflammatory process, or distribution of radiopharmaceutical agent(s) imaging) - one of the following CPT® codes: CPT® 78800, CPT® 78801, 78802, or CPT® 78803 in concert with Tc-99m sulfur colloid marrow imaging (one of CPT® codes: CPT® 78102, CPT® 78103, or CPT® 78104) or SPECT/CT (CPT® 78830) is medically necessary in cases with altered bone marrow distribution, such as joint prosthesis.

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Foreign Body (PEDMS-9)

Guideline

Foreign Body (PEDMS-9)
Reference (PEDMS-9)

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Foreign Body (PEDMS-9)

MSP.FB.0009.A

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- Foreign body imaging indications in pediatric individuals are similar to those for adult individuals. See: **Foreign and Loose Bodies (MS-6.0)** for imaging guidelines.
- The common soft tissue foreign bodies in children are wood, glass, and metal slivers. The latter two elements are radiopaque and visible to some degree on plain x-rays, whereas wood is usually radiolucent and nearly always imperceptible on x-rays. When a radiolucent foreign body is suspected, ultrasound (CPT® 76881 or 76882) is medically necessary to identify the foreign body.

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Inflammatory Musculoskeletal Disease (PEDMS-10)

Guideline

Inflammatory Musculoskeletal Disease (PEDMS-10.0)
Juvenile Idiopathic Arthritis (PEDMS-10.1)
Chronic Recurrent Multifocal Osteomyelitis (PEDMS-10.2)
Inflammatory Muscle Diseases (PEDMS-10.3)
References (PEDMS-10)

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Inflammatory Musculoskeletal Disease (PEDMS-10.0)

MSP.MD.0010.0.A

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- A pertinent clinical evaluation including a detailed history, physical examination, and plain x-rays should be performed prior to considering advanced imaging.
- Inflammatory arthritis imaging indications in pediatric individuals are very similar to those for adult individuals. See: **Inflammatory Arthritis (MS-12.1)** in the Musculoskeletal Imaging Guidelines. Specific pediatric considerations are included below.

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Juvenile Idiopathic Arthritis (PEDMS-10.1)

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- Ultrasound (CPT® 76881 or 76882) is medically necessary for assessment of: size and characteristics of joint effusions, extent of synovial hypertrophy (which is the hallmark of juvenile idiopathic arthritis), and involvement of tendinous structures.
  - Repeat imaging is medically necessary for monitoring treatment or with planned treatment change
  - MRI of the most symptomatic joint without contrast or without and with contrast is medically necessary if ultrasound is inconclusive and MRI findings would alter individual management
- Distribution of Radiopharmaceutical Agent SPECT (CPT® 78802, or 78803), or SPECT/CT (CPT® 78830), is medically necessary for evaluation of facet arthropathy in patients with ankylosing spondylitis, osteoarthritis, or rheumatoid arthritis.
- MRI TMJ (CPT® 70336) is medically necessary annually for detecting silent TMJ arthritis in children with juvenile idiopathic arthritis (JIA).
- MRI without or without and with of the most involved joint is medically necessary to evaluate involved or symptomatic joints in the following situations:
  - When diagnosis is uncertain prior to initiation of drug therapy
  - To study the effects of treatment with disease modifying anti-rheumatic drug (DMARD) therapy
  - To determine a change in treatment
- MRI (with the exception of the annual screening MRI of the TMJ discussed above) is not medically necessary for routine follow-up of treatment.

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Chronic Recurrent Multifocal Osteomyelitis (PEDMS-10.2)

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Chronic recurrent multifocal osteomyelitis (CRMO) is a rare autoimmune disease affecting multiple bones, arising most commonly during the second decade of life. Treatment consists of anti-inflammatory and immunomodulatory therapies, and is directed predominantly by status of clinical symptoms (most commonly pain).

- The following imaging is medically necessary for undividuals with CRMO for evaluation of new or worsening pain, or response to treatment in individuals without complete clinical resolution of pain symptoms, when plain x-rays are non-diagnostic:
  - Bone scan (CPT® codes: 78300, 78305, 78306, 78315) OR
  - SPECT (CPT® codes: 78803, or 78831), OR
  - Nuclear Bone Marrow imaging (CPT® codes: 78102, 78103, or 78104), OR
  - Radiopharmaceutical localization of tumor, inflammatory process, or distribution of radiopharmaceutical agent imaging (CPT® codes: 78800, 78801, 78802, or 78803), OR
  - SPECT/CT (CPT® codes: 78830, or 78832)
  - MRI without contrast of specific painful body areas when plain x-ray and bone scan are insufficient to direct acute individual care decisions.
- Literature suggests MRI may have greater sensitivity for clinically occult lesions than bone scan. Whole-body MRI (CPT® 76498) is medically necessary for CRMO in the following situations:
  - Individuals suspected of having CRMO if characteristic MR findings of CRMO would preclude the need for a biopsy.
    - Characteristic finding include multiple lesions most commonly involving the juxtaphyseal/peri-physeal portions of the tibia and femur, the clavicle and thoracolumbar spine.
  - Every 6-12 months in individuals with an established diagnosis of CRMO to monitor treatment or to evaluate for clinically occult but radiographically active lesions.
  - See: **Whole Body MR Imaging (Preface-5.2)** for additional details.

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Inflammatory Muscle Diseases (PEDMS-10.3)

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- A pertinent clinical evaluation including a detailed history, physical examination, and plain x-rays should be performed prior to considering advanced imaging.

Inflammatory Muscle Diseases:

These include but are not limited to dermatomyositis, polymyositis, and sporadic inclusion body myositis. MRI without contrast of a single site is medically necessary in these disorders for the following purposes:

- Selection of biopsy site
- Clinical concern for progression
- Treatment monitoring
- Detection of occult malignancy

Juvenile Dermatomyositis:

Contrary to adult dermatomyositis, juvenile dermatomyositis is very rarely paraneoplastic in nature, and routine screening for occult neoplasm is not medically necessary.

The following are medically necessary for juvenile dermatomyositis:

- MRI without contrast to confirm the diagnosis and thus avoid a biopsy.
- CT without contrast (CPT® 73700) or MRI (CPT® 73718) to follow progressive calcification in muscles
  - Both CT and MRI are rarely indicated concurrently.
- CT Chest (CPT® 71260) and Abdomen and Pelvis (CPT® 74177) with contrast for individuals with palpable lymphadenopathy or hepatosplenomegaly.

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Muscle/Tendon Unit Injuries (PEDMS-11)

Guideline

Muscle/Tendon Unit Injuries (PEDMS-11)

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Muscle/Tendon Unit Injuries (PEDMS-11)

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- Muscle and tendon unit injury imaging indications in pediatric individuals are identical to those in the general imaging guidelines. See: **Muscle and Tendon Injuries (MS-11.0)** in the Musculoskeletal Imaging Guidelines.

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Osgood-Schlatter Disease (PEDMS-12)

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Osgood-Schlatter Disease (PEDMS-12)
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Osgood-Schlatter Disease (PEDMS-12)

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- Osgood-Schlatter Disease is defined as traction apophysitis of the tibial tubercle in skeletally immature individuals. Diagnosis is by clinical examination and x-ray, and treatment is conservative.
- Advanced imaging is not indicated in this disorder.

Background and Supporting Information

- The condition is self-limited and is secondary to repetitive extensor mechanism stress activities, such as jumping and sprinting.

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Popliteal (Baker) Cyst (PEDMS-13)

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Popliteal (Baker) Cyst (PEDMS-13)

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Popliteal or Baker cyst in children is a different clinical entity than in adults and is almost never due to intra-articular pathology. These lesions are usually treated conservatively and rarely require surgery.

- Ultrasound (CPT® 76881 or 76882) is the medically necessary initial imaging study.
- MRI without contrast (CPT® 73721) is medically necessary for preoperative planning or if ultrasound is non-diagnostic.

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Slipped Capital Femoral Epiphysis (SCFE) (PEDMS-14)

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Slipped Capital Femoral Epiphysis (SCFE) (PEDMS-14)
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Slipped Capital Femoral Epiphysis (SCFE) (PEDMS-14)

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Slipped capital femoral epiphysis (SCFE) should be considered in young adolescents or preadolescents with groin, anterior thigh, or atraumatic knee pain. Symptoms often include a history of intermittent limp and pain for several weeks or months that are often poorly localized to the thigh, groin, or knee. Any obese adolescent or preadolescent presenting with a history of a limp and thigh, knee, or groin pain for several weeks to one month should be presumed to have a slipped capital femoral epiphysis (SCFE).

Imaging studies:

- Anteroposterior and lateral x-rays (frog leg or cross table lateral) of both hips will confirm or exclude the diagnosis.
  - If clinical suspicion remains after negative plain films, MRI without contrast (CPT® 73721) or without and with contrast (CPT® 73723) is medically necessary to detect widening of the physis before the femoral head is displaced (pre-slip).
- Because a significant percentage of SCFE is bilateral at presentation, it is medically necessary to evaluate the contralateral hip if requested, as some surgeons advocate surgical treatment of pre-slip.
- If MRI was not completed for diagnosis, MRI without contrast (CPT® 73721) is medically necessary for preoperative planning.

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Limb Length Discrepancy (PEDMS-15)

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Limb Length Discrepancy (PEDMS-15)

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Limb Length Discrepancy (PEDMS-15)

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- Limb length discrepancy imaging indications in pediatric individuals are identical to those in the general imaging guidelines. See: **Limb Length Discrepancy (MS-17.0)** in the Musculoskeletal Imaging Guidelines.

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Congenital Anomalies of the Foot and Lower Extremity (PEDMS-16)

Guideline

Tarsal Coalition (Calcaneonavicular Bar/Rigid Flat Foot) (PEDMS-16.1)
Club Foot (PEDMS-16.2)
Vertical Talus (PEDMS-16.3)
Femoral Anteversion and Tibial Torsion (PEDMS-16.4)
References (PEDMS-16)

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Tarsal Coalition (Calcaneonavicular Bar/ Rigid Flat Foot) (PEDMS-16.1)

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- Plain x-rays should be performed initially since the calcaneonavicular bar is readily visible in older children and adults.
  - Talocalcaneal coalition is more difficult to evaluate on plain x-rays.
- CT without contrast (CPT® 73700) or MRI without contrast (CPT® 73718) is medically necessary if tarsal coalition is suspected (because of restricted hindfoot motion on physical exam), and plain x-rays are inconclusive.

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Club Foot (PEDMS-16.2)

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Club Foot is a congenital foot contracture with foot in equinus (plantar flexion) and heel and forefoot in varus/adduction (turned in). Immediate diagnosis and specialty evaluation in the first week of life provide the best chance for successful correction.

- Plain x-rays should be performed initially since the anomaly is readily visible in older children and adults.
- Ultrasound (CPT® 76881 or 76882) is medically necessary to characterize the cartilaginous tarsal bones and demonstrate tarsal bone alignment in infants with non-ossified tarsal bones.
- MRI (CPT® 73718) or CT (CPT® 73700) is medically necessary to determine residual deficits following repair.
  - Ultrasound is not required prior to MRI or CT if those studies are appropriate.

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Vertical Talus (PEDMS-16.3)

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- Congenital vertical talus (also known as congenital rocker-bottom foot) is a fixed foot deformity characterized by irreducible talonavicular dislocation. The talus is plantar flexed and does not articulate with the navicular bone.
- Plain x-rays should be performed initially since the anomaly is readily visible in older children and adults.
- MRI (CPT® 73718) or CT (CPT® 73700) are medically necessary to determine residual deficits following repair.

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Femoral Anteversion and Tibial Torsion (PEDMS-16.4)

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- Femoral anteversion is a rotational deformity of the femur, which may lead to an in-toeing gait.
- Tibial torsion is a rotational deformity of the tibia that may lead to in-toeing or out-toeing gait, and can be associated with the foot deformities already discussed in **Tarsal Coalition (Calcaneonavicular Bar/Rigid Flat Foot) (PEDMS-16.1)**, **Club Foot (PEDMS-16.2)**, and **Vertical Talus (PEDMS-16.3)**.
- Both deformities are typically diagnosed on clinical examination, but CT Lower Extremity without contrast (CPT® 73700) OR MRI Lower Extremity without contrast (CPT® 73718) is medically necessary for preoperative evaluation.

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General Guidelines (PEDNECK-1)

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Procedure Codes Associated with Neck Imaging

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MRI	CPT®
MRI Orbit, Face, Neck without contrast	70540
MRI Orbit, Face, Neck with contrast (rarely used)	70542
MRI Orbit, Face, Neck without and with contrast	70543
MRI Temporomandibular Joint (TMJ)	70336
Unlisted MRI procedure (for radiation planning or surgical software)	76498

MRA	CPT®
MRA Neck without contrast	70547
MRA Neck with contrast	70548
MRA Neck without and with contrast	70549

CT	CPT®
CT Maxillofacial without contrast (includes sinuses, jaw, and mandible)	70486
CT Maxillofacial with contrast (includes sinuses, jaw, and mandible)	70487
CT Maxillofacial without and with contrast (includes sinuses, jaw, and mandible)	70488
CT Neck without contrast (includes jaw, and mandible)	70490

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Unlisted CT procedure (for radiation planning or surgical software)	76497

CTA	CPT®
CTA Neck	70498

Nuclear Medicine	CPT®
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging: skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging: whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
Thyroid Uptake, Single or Multiple Quantitative Measurement(s) (Including Stimulation, Suppression, or Discharge, When Performed)	78012
Thyroid Imaging (Including Vascular Flow, When Performed)	78013
Thyroid Imaging (Including Vascular Flow, When Performed); with Single or Multiple Uptake(s) Quantitative Measurement(s) (Including Stimulation, Suppression, or Discharge, When Performed)	78014
Thyroid Carcinoma Metastases Imaging Limited Area	78015

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Nuclear Medicine	CPT®
Thyroid Carcinoma Metastases Imaging with Additional Studies	78016
Thyroid Carcinoma Metastases Imaging Whole Body	78018
Thyroid Carcinoma Metastases Uptake (Add-on Code)	78020
Parathyroid Planar Imaging (Including Subtraction, When Performed)	78070
Parathyroid Planar Imaging (Including Subtraction, When Performed); with Tomographic (SPECT)	78071
Parathyroid Planar Imaging (Including Subtraction, When Performed); with Tomographic (SPECT), and Concurrently Acquired Computed Tomography (CT) for Anatomical Localization	78072
Salivary Gland Nuclear Imaging	78230
Salivary Gland Nuclear Imaging with Serial Imaging	78231
Salivary Gland Function Study	78232
Esophageal Motility Study	78258
Radiopharmaceutical Localization Imaging Limited area	78800
Radiopharmaceutical Localization Imaging Whole Body	78802
Radiopharmaceutical Localization Imaging SPECT	78803

Ultrasound	CPT®
Soft tissues of head and neck Ultrasound (thyroid, parathyroid, parotid, etc.)	76536
Duplex scan of extracranial arteries; complete bilateral study	93880
Duplex scan of extracranial arteries; unilateral or limited study	93882
Non-invasive physiologic studies of extracranial arteries, complete bilateral study	93875
Ultrasound guidance for needle placement	76942

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General Guidelines (PEDNECK-1.0)

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- A pertinent clinical evaluation including a detailed history, physical examination, since the onset or change in symptoms, and appropriate laboratory studies should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the neck is not medically necessary. Advanced imaging of the neck is only medically necessary in individuals who have documented active clinical signs or symptoms of disease involving the neck.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the neck are not medically necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Age Considerations (PEDNECK-1.1)

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- Many conditions affecting the neck in the pediatric population have different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are 18 years old or younger should be imaged according to the Pediatric Neck Imaging Guidelines. Any conditions not specifically discussed in the Pediatric Neck Imaging Guidelines should be imaged according to the General Neck Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Neck Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Modality General Considerations (PEDNECK-1.3)

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- MRI
  - MRI Orbit/Face/Neck is generally performed without and with contrast (CPT® 70543) unless the individual has a documented contraindication to gadolinium or otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize individual movement, sedation is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with developmental delays. Sedation may be administered by oral, intravenous, and/or inhalational routes. In order to limit sedation time in this subdivision of pediatric individuals, the following should be considered:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition-based guidelines. If intravenous access is medically necessary for sedation and there is no contraindication for using contrast, imaging without and with contrast may avoid repeating a study with sedation to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If studies of multiple body areas are medically necessary by these clinical guidelines for the clinical condition being evaluated, MRI studies of all necessary body areas should be obtained concurrently in the same sedation session.
  - The presence of surgical hardware or implanted devices may preclude the use of MRI.
  - Coordination between provider and the imaging service can result in providing the best choice of radiologic studies for the pediatric individual.
- CT
  - CT Neck typically extends from the base of the skull to the upper thorax.

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    - A separate CPT® code for head imaging in order to visualize the skull base is not medically necessary.
    - In some cases, especially in follow-up of a known finding, it is medically necessary to limit the exam to the region of concern to reduce radiation exposure.
  - CT Neck is generally performed with contrast (CPT® 70491) unless the individual has a documented contraindication to CT contrast or otherwise stated in a specific guideline section.
  - CT Neck may be medically necessary for further evaluation of abnormalities suggested on prior US or MRI Procedures.
  - In general, CT Neck is medically necessary when evaluating trauma, malignancy, and for preoperative planning.
  - CTA Neck (CPT® 70498) is medically necessary for evaluation of the vessels of the neck, especially with concern for dissection.
  - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
  - Coordination between the provider and the imaging service can result in the best choice of radiologic studies for the pediatric individual.
- Ultrasound
  - Ultrasound soft tissues of the neck (CPT® 76536) is medically necessary as an initial study for evaluating thyroid, parathyroid, parotid and other salivary gland lesions. Ultrasound is also used to further characterize adenopathy, palpable superficial masses, or swelling.
  - For those individuals who do require additional advanced imaging after ultrasound, ultrasound can be very beneficial in selecting the proper modality, body area, image sequences, and contrast level that will provide the most definitive information for the pediatric individual.
- Nuclear Medicine
  - Nuclear medicine studies of the neck in pediatric individuals are most commonly used to evaluate neck masses, or thyroid and parathyroid disease following initial studies with anatomic imaging, such as ultrasound, CT, or MRI. See **Neck Masses (Pediatric) (PEDNECK-2.1)** and **Thyroid and Parathyroid (PEDNECK-6)** for imaging guidelines.
  - Evaluation of salivary gland function in individuals with dry mouth (xerostomia) and **ONE** of the following:
    - Sjögren syndrome **OR**
    - Sialadenitis **OR**
    - History of head or neck radiation therapy, one of the following is medically necessary:
      - Salivary Gland Nuclear Imaging (CPT® 78230) **OR**

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  - Salivary Gland Nuclear Imaging with Serial Imaging (CPT® 78231) **OR**
  - Salivary Gland Function Study (CPT® 78232)

- 3D Rendering
  - 3D Rendering indications in pediatric neck imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Neck Masses (Pediatric) (PEDNECK-2)

Guideline

Neck Masses (Pediatric) (PEDNECK-2.1)
References (PEDNECK-2)

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Neck Masses (Pediatric) (PEDNECK-2.1)

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- Evaluation of neck masses in pediatric individuals involves careful consideration of clinical history and accurate physical examination. The individual's age and knowledge of the anatomy and common lesions of the neck are very important in narrowing the differential diagnosis.
- Congenital lesions (vascular lesions, thyroglossal duct cysts, branchial cleft cysts, teratomas, dermoids, and thymic cysts) and lymphadenopathy account for nearly all benign neck lesions. Primary malignant tumors in children are usually neuroblastoma, lymphoma, or rhabdomyosarcoma.
- Initial imaging of choice:
  - Ultrasound Neck (CPT® 76536)
    - Color Doppler ultrasound bilateral study of carotid arteries (CPT® 93880) **OR** Duplex unilateral study (CPT® 93882) is medically necessary to evaluate the vasculature.
- For inconclusive ultrasound or to further delineate abnormalities on ultrasound:
  - MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) **OR**
  - CT Neck with contrast (CPT® 70491)
- For concern for malignancy:
  - Ultrasound Neck is not medically necessary prior to imaging.
  - MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) **OR**
  - CT Neck with contrast (CPT® 70491)
- For lymphadenopathy persisting for more than 4-weeks of treatment **OR** suspicion of complications such as abscess formation:
  - Ultrasound Neck (CPT® 76536) is medically necessary. See **Cervical Lymphadenopathy (PEDNECK-3.1)**.
- Congenital cervical cysts:
  - Ultrasound Neck (CPT® 76536) is medically necessary for suspected cystic neck mass.
- Congenital cervical sinus, fistula, cyst, or for preoperative planning:
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) **OR**
  - CT Neck with contrast (CPT® 70491)
    - For fourth branchial cleft cyst/sinus/fistula- barium swallow is medically necessary in addition to the above conventional imaging.

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- Salivary gland nuclear imaging: **ONE** of the following is medically necessary for evaluation of parotid masses to allow preoperative diagnosis of Warthin's tumor:
  - Salivary Gland Nuclear Imaging (CPT® 78230) **OR**
  - Salivary Gland Nuclear Imaging with Serial Imaging (CPT® 78231) **OR**
  - Salivary Gland Function Study (CPT® 78232)
- Ranula (a cystic structure on the floor of the mouth):
  - CT Neck with contrast (CPT®70491) **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary, especially when there is concern for a "plunging" ranula (lesion extending into the submandibular space).

Background and Supporting Information

- Cervical lymphadenitis is common in children and follows most viral or bacterial infections of the ears, nose, and throat. No advanced imaging is necessary with uncomplicated lymph node enlargement.
- Congenital cervical cysts frequently present in children and include thyroglossal duct cyst (55% of cases), cystic hygroma (25%), branchial cleft cysts (16%), bronchogenic cyst (0.91%), and thymic cyst (0.3%).
- The most common malignant neck tumors in children are lymphoma, rhabdomyosarcoma, and neuroblastoma. Many neuroblastomas arise in the neck.

Differential Diagnosis of Neck Lesions by Anatomic Region:

- Subcutaneous tissues:
  - Teratoma (includes dermoid cysts)
    - Cervical teratomas are typically large bulky masses discovered at birth or in the first year of life.
    - Large lesions may cause stridor, dyspnea, or dysphagia.
    - Most teratomas arise in the anterior suprahyoid neck and may be midline or off midline in location and adjacent to or within a thyroid lobe.
  - Vascular lesions
  - Lipoma
  - Cellulitis
  - Plexiform neurofibromas
  - Keloid
  - Scar
  - Pilomatrixoma
  - Subcutaneous fat fibrosis (in neonates)
- Retropharyngeal space:
  - Abscess, cellulitis, adenitis
    - Usually involves children under age 6.

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        - Individuals have history of upper respiratory tract infection followed by high fever, dysphagia, and neck pain.
    - Lymphadenopathy
    - Extension of goiter
    - Extension of pharyngeal tumor
- Retrovisceral space (posterior to the cervical esophagus):
    - Gastrointestinal duplication cysts (usually are diagnosed in first year of life).
- Pretracheal space (contains trachea, larynx, cervical esophagus, recurrent laryngeal nerves, and thyroid and parathyroid glands):
    - Thyroglossal duct cyst
        - Thyroglossal duct cyst most commonly presents before the age of 20; 75% present as a midline mass and 43% of individuals present with an infected mass.
        - Usually presents as an enlarging, painless midline mass.
        - Thyroid carcinoma occurs in 1% of thyroglossal duct cysts.
    - Goiter
    - Laryngocele
    - Lymphadenopathy
    - Teratoma
    - Abscess
    - Ectopic thymus or cervical extension of normal thymus
- Danger space (closed space lying between the skull base and the posterior mediastinum and between the alar and prevertebral fasciae in a sagittal plane):
    - Cellulitis
    - Abscess
- Prevertebral space:
    - Neurenteric cyst
    - Cellulitis
    - Abscess
    - Spondylodiskitis
    - Lymphadenopathy
    - Paraganglioma
- Carotid sheath space:
    - Jugular vein thrombosis or phlebitis
    - Lymphadenopathy
    - Cellulitis
    - Abscess
    - Paraganglioma
- Parotid gland space:

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- Parotid lymphadenopathy
- Retromandibular vein thrombosis
- Parotiditis
- Sialodochitis (inflammation of the salivary gland duct)
- Salivary duct stone
- Abscess

- Submandibular and sublingual spaces:
  - Cellulitis
  - Abscess
  - Sialadenitis
  - Thyroglossal duct cyst
  - Branchial cleft cyst
    - 90% of branchial abnormalities arise from the second branchial apparatus.
    - Second branchial cleft cysts are the most common branchial cleft cyst and usually present in individuals between 10 and 40 years as painless fluctuant masses.
    - They typically present as slowly growing, non-tender masses in the upper neck.
    - Most second branchial cleft cysts are located in the submandibular space, at the anteromedial border of the sternocleidomastoid muscle, lateral to the carotid space, or posterior to the submandibular gland.
    - Ranula – typically cystic masses in the floor of the mouth
- Masticator space (includes masseter and pterygoid muscles):
  - Venous or lymphatic malformation
  - Cellulitis
  - Abscess
  - Rhabdomyosarcoma
- Parapharyngeal space:
  - Cellulitis
  - Abscess
  - Neurogenic tumors (CN V, IX, XI and XII)
  - Paragangliomas
  - Neurofibromas
  - Lymphoma
  - Rhabdomyosarcoma
- Paravertebral space:
  - Cervical dermal sinus (epithelium lined dural tubes that connect the skin with the central nervous system or its covering)
  - Meningocele
  - Rhabdomyosarcoma

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- Lymphoma
- Neuroblastoma
- Neurofibroma

- Posterior cervical space:
  - Lymphadenopathy
  - Lymphatic malformation

Congenital Neck Masses:

- Anterior neck masses
  - Branchial anomolies
    - Sinus: with either an internal (to the pharynx) or external (to the skin) opening
    - Fistula: with both an internal and external opening
    - Cyst: closed sac with no openings
  - First branchial anomalies
    - Typically sinus or cyst
    - Located anywhere from the external auditory canal to the region of the parotid gland, down to the level of the hyoid; may communicate with the preauricular soft tissue/parotid, parapharyngeal space or anterior triangle of the neck
      - Type I tract parallels the external auditory meatus.
      - Type II tract courses over the angle of mandible through the parotid ending near/within the external auditory canal bony cartilaginous junction.
  - Second branchial anomalies
    - The most common
    - Located from anterior neck in the region of the middle to lower two thirds of the sternocleidomastoid and the great vessels to the pharyngeal mucosa (tonsil)- a tract and/or cyst may occur anywhere along this path
  - Third branchial anomalies
    - Typically located from the low anterior neck to the base of the pyriform sinus
  - Fourth branchial anomalies
    - Potential tract from the low anterior neck to the thyroid gland or mediastinum
  - Cervical thymus
    - Ectopic thymic remnants can be found in the anterior neck (left more commonly than right) and extend deep (near the carotid sheath)- can connect to mediastinum or have cystic components.
- Midline
  - Ectopic Thyroid/Thyroglossal Duct Cysts
    - Anywhere from the tongue base to the mediastinum (a result of the normal embryologic pathway of the thyroid that fails to obliterate or reach its normal location in the lower neck)

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- Just off the midline
  - Thyroid/Thyroglossal Duct Cysts (cysts below the hyoid bone may be lateral to the midline in position)
    - Laryngocele
      - An abnormal dilation of the saccule of the larynx
        - Internal: within the thyroid cartilage
        - External: beyond the thyrohyoid membrane into the neck
- Anywhere within the neck
  - Teratomas
    - Tissue from all three germ cell layers (ectodermal, mesodermal and endodermal components) typically present as a firm mass, can have calcifications seen on imaging
  - Dermoid cysts
    - Cysts with ectodermal and mesodermal structures (commonly lined by epidermis and containing epidermal appendages) typically in the midline/ submental region, but can be anywhere in the head and neck including orbit
  - Epidermoid cysts
    - Cysts with only ectodermal components (with squamous material)
  - Pilomatixoma
    - Lesion derived from hair matrix/follicles
  - Vascular anomalies
    - Hemangiomas (most common)
      - Congenital: present at birth typically involute
      - Infantile: noted to have a rapid/proliferative phase followed by involution
    - High flow
      - Arteriovenous malformations (AVM) and arteriovenous fistuas (AVF) tangle of vessels
    - Low flow
      - Venous lymphatic and capillary malformations
  - Lymphatic malformations (lymphangiomas)
    - Result from a failure of lymph spaces to connect to the rest of the lymphatic system
      - Macrocystic: comprised of large cysts
      - Microcystic: comprised of smaller cysts typically more infiltrative, lending to difficult excision

Evidence Discussion

- Ultrasound, although operator dependent, is utilized for the initial evaluation of pediatric neck masses, secondary to its availability, expense, lack of radiation, and

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avoidance of sedation.[1] Ultrasound is limited by poor depth of penetration when evaluating deeper abnormalities.[2]

- Contrast enhanced CT is easily accessible and available; however, it exposes the individual to ionizing radiation.[1]
- MRI provides excellent soft tissue detail without radiation; however, due to the time needed for complete imaging, anesthesia or sedation may be required.[1] Children under 6 years of age, children with developmental delays, or children with anxiety/ claustrophobia may require deep sedation in order to successfully perform a complete radiologic study.[3]

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Cervical Lymphadenopathy (PEDNECK-3)

Guideline

Imaging (PEDNECK-3.1)
References (PEDNECK-3)

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Imaging (PEDNECK-3.1)

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- Painful acute lymphadenopathy and other painful neck masses (including neck "swelling") should be treated with a trial of conservative therapy for at least 4-weeks, including antibiotics if appropriate.
  - If there is improvement with conservative treatment, advanced imaging is not medically necessary.
  - Ultrasound (CPT® 76536) is medically necessary for any of the following:
    - Initial evaluation of persistent lymphadenopathy following 4-weeks of treatment/ observation **OR**
    - Unexplained fever (temperature ≥100.4°F) and there is clinical concern for suppurative lymphadenopathy/neck abscess
- For inconclusive ultrasound/to further characterize abnormalities found on ultrasound:
  - MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) **OR**
  - CT Neck with contrast (CPT® 70491)
- If systemic symptoms or other clinical findings suggest malignancy, see **Pediatric Lymphomas (PEDONC-5)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

Background and Supporting Information

- Both MRI and CT are superior to ultrasound for defining the relationship of an abscess to adjacent structures, particularly the airway, and for detecting posterior cervical, mediastinal and intracranial extension.
- Inflammatory lymph nodes from acute lymphadenitis are usually painful, tender, and mobile, frequently associated with upper respiratory infection, pharyngitis or dental infection.
- Occasionally, sarcoidosis or toxoplasmosis and human immunodeficiency virus (HIV) can cause inflammatory lymphadenopathy as well.

Evidence Discussion

- Ultrasound can be used not only as an initial imaging modality, but also as a tool for serial examinations in cases without pathologic features.[1]
- Ultrasound has the benefit of being able to be used to serially evaluate the pediatric individual with cervical adenopathy without the need for sedation/anesthesia and without exposure to radiation.[1]

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- Both CT and MRI can provide additional information that may assist with surgical planning. CT is low risk, but involves radiation exposure, while MRI does not involve radiation, but requires more time to complete the study and possibly sedation.[1]

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Dystonia/Torticollis (PEDNECK-4)

Guideline

Dystonia/Torticollis (PEDNECK-4.1)
References (PEDNECK-4)

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Dystonia/Torticollis (PEDNECK-4.1)

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Infants under 12 Months of Age (Congenital Muscular Torticollis/Fibromatosis Colli)

- Ultrasound Neck (CPT® 76536) is medically necessary as the initial study.
  - If Ultrasound is Positive→ No further imaging is medically necessary as the diagnosis is defined.
  - If Ultrasound is Negative or to further evaluate for other structural causes:
    - CT Neck with contrast (CPT® 70491) **OR**
    - MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR**
    - MRI Orbit/Face/Neck without and with contrast (CPT® 70543)

Background and Supporting Information

- Individuals usually present by 2-weeks of life with an anterior neck mass, which is commonly right sided (75% of cases). A history of a traumatic breech or forceps delivery is common.
- Congenital muscular torticollis is more common in male infants.

Children and Adults (Acquired Torticollis)

- Initial evaluation with history of trauma, and low suspicion of injury:
  - Plain radiographs of the cervical spine are medically necessary
    - To identify fracture or malalignment if plain radiographs are inconclusive or in individuals with a high-risk mechanism of cervical spine injury within the last 3 months (see below**):
      - CT Neck with contrast (CPT® 70491) **AND/OR**
      - CT Cervical Spine without contrast (CPT® 72125) is medically necessary.
- In the clinical setting of cervical spine trauma with an associated neurologic deficit:
  - MRI Cervical Spine without contrast (CPT® 72141) is medically necessary.
- In the absence of trauma, to identify underlying abscess, bony, muscular, vascular, or neurologic causes, ONE of the following is medically necessary:
  - CT Neck with contrast (CPT® 70491), **OR**
  - CT Cervical Spine without contrast (CPT® 72125), **OR**
  - MRI Cervical Spine without contrast (CPT® 72141), **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543), **OR**
  - MRA Neck without and with contrast (CPT® 70549)

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  - Positive→ Further advanced imaging is not medically necessary if a local cause has been identified.
  - Negative→ MRI Brain without and with contrast (CPT® 70553) is medically necessary to exclude CNS cause.

**High-risk mechanisms of cervical spine injury may include:

- Head trauma and/or maxillofacial trauma
- Pedestrian in a motor vehicle accident
- Fall from above standing height
- Diving accident
- Head-on motor vehicle collision without/with airbag deployment
- Rollover motor vehicle collision
- Ejection from the vehicle in a motor vehicle collision
- High speed of the vehicle at the time of collision
- Not wearing a seatbelt/shoulder harness in a motor vehicle collision
- Individuals with ankylosing spondylitis are at high-risk of cervical spine fractures even with minor direct/indirect trauma to the cervical spine which can result in quadriparesis/quadriplegia

Background and Supporting Information

- Injury or inflammation involving the sternocleidomastoid or trapezius muscles is the most common cause of acquired torticollis in children.
- Torticollis or cervical dystonia is an abnormal twisting of the neck in which the head is rotated or twisted. Acute causes are most common. Children with deep space neck infections present with torticollis approximately 50% of the time. Other causes are variable and may be congenital, acquired (caused by trauma, juvenile idiopathic arthritis, or neoplasm), or idiopathic.

Evidence Discussion

- Ultrasound is the initial method of evaluation of suspected fibromatosis colli. Ultrasound has a sensitivity of 100% and has the advantages of being readily available as well as avoiding sedation and radiation.[1]
- Standard x-rays are the initial imaging modality after trauma, but can be technically challenging due to head position.[2,3]
- Both CT and MRI assist in diagnosing non traumatic torticollis as a result of atlantoaxial subluxation; CT has the advantage of being readily available; however, it exposes the child to radiation, while MRI provides better soft tissue detail such as spinal cord compression, but may require sedation/anesthesia due to the length of the study.[3]

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Dysphagia (PEDNECK-5)

Guideline

Dysphagia (PEDNECK-5.1)
References (PEDNECK-5)

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Dysphagia (PEDNECK-5.1)

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- Dysphagia imaging indications in pediatric individuals are very similar to those for adult individuals. See **Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)** in the Neck Imaging Guidelines.
- For concern of foreign body ingestion as the etiology of dysphagia initial imaging:
  - X-rays of the neck and chest are medically necessary.
- For dysphagia associated with chest pain and difficulty swallowing both solids and liquids or gastroesphageal reflux:
  - Esophageal motility study (CPT® 78258) is medically necessary.
- For a suspected anatomical variant such as a vascular ring, right sided aortic arch, or double arch noted on chest radiography (which can be associated with dysphagia):
  - CTA Chest (CPT® 71275) **OR**
  - MRA Chest (CPT® 71555) is medically necessary.

Evidence Discussion

- X-rays are medically necessary as the initial evaluation of foreign body ingestion in children. Plain films provide the advantage of being readily available to detect radiopaque foreign bodies or other pathologic findings.[1]
- Chest CT and MRI provide information regarding anatomy and possible external compression resulting in dysphagia, but require the use of contrast and possible sedation.[1]

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Thyroid and Parathyroid (PEDNECK-6)

Guideline

Thyroid Masses or Nodules (PEDNECK-6.1)
Hyperthyroidism (PEDNECK-6.2)
Hypothyroidism (PEDNECK-6.3)
Parathyroid Imaging (PEDNECK-6.4)
References (PEDNECK-6)

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Thyroid Masses or Nodules (PEDNECK-6.1)

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- For the initial study for evaluation of thyroid masses, diffuse thyroid enlargement, or nodules in pediatric individuals:
  - Ultrasound Neck (CPT® 76536) is medically necessary.
- **For a normal or elevated TSH** with any solitary or suspicious thyroid nodule noted on imaging or physical exam:
  - Fine needle aspiration (FNA) under ultrasound guidance (CPT® 76942) is medically necessary.
- **For a low TSH:**
  - Nuclear thyroid scintigraphy (either CPT® 78013 or CPT® 78014) is medically necessary.
    - Hyperfunctioning nodules should be treated surgically but may also undergo FNA under ultrasound guidance (CPT® 76942) if suspicious in appearance and not being treated surgically.
    - Hypofunctioning nodules should undergo FNA under ultrasound guidance (CPT® 76942).
- For lymph node assessment if cervical lymph node imaging was not performed at the time of the initial diagnostic thyroid ultrasound:
  - Repeat imaging with Ultrasound Neck (CPT® 76536) is medically necessary.
- For preoperative planning in individuals with large or fixed masses, vocal cord paralysis, or bulky cervical or supraclavicular adenopathy:
  - CT Neck without contrast (CPT® 70490) **OR**
  - CT Neck with contrast (CPT® 70491) **OR**
  - MRI Orbit/Face/Neck without contrast (CPT® 70540) **OR**
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is medically necessary.
  - In addition, for individuals with substernal extension of the thyroid, pulmonary symptoms or abnormalities on chest x-ray:
    - CT Chest without contrast (CPT® 71250) **OR**
    - CT Chest with contrast (CPT® 71260) is medically necessary.
- If any biopsy reveals thyroid carcinoma, See **Thyroid Cancer (ONC-6)** in the Oncology Imaging Guidelines.

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- Repeat ultrasound (CPT® 76536) and/or FNA under ultrasound guidance (CPT® 76942) is medically necessary 3-6 months following initial biopsy if the initial biopsy shows inadequate, or non-diagnostic findings.
  - Repeat ultrasound (CPT® 76536) is medically necessary in 6-12 months if the nodule is stable and/or FNA is benign.
  - The nodule should be treated surgically if growing or the FNA is not benign.
- Repeat ultrasound (CPT® 76536) is medically necessary 6-12 months following initial biopsy if the initial biopsy shows benign findings.
  - Repeat ultrasound (CPT® 76536) is medically necessary every 1-2 years if the nodule is stable.
  - Repeat FNA under ultrasound guidance (CPT® 76942) or manage surgically if the nodule is growing or concerning new findings are present.
  - Benign nodules that have been surgically resected do not require routine imaging follow up in the absence of clinical or laboratory changes suggesting recurrence.
- If the initial biopsy shows indeterminate or suspicious findings, surgery is recommended.

Evidence Discussion

- Ultrasound is used as the initial evaluation for thyroid abnormalities and follow up as it is noninvasive, radiation free, inexpensive, very sensitive, and does not require sedation to complete.[1,2,25,26]
- Fine needle biopsy in pediatric thyroid nodules has a sensitivity of 94% and specificity of 100%.[3]
- Both CT and MRI can be valuable in preoperative evaluation of thyroid disease for determining involvement of lymph nodes or adjacent structures. However, iodine based contrasts can lead to suppression of radioactive iodine thyroid uptake, thus postponing radioiodine treatment and favoring the use of MRI.[1]

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Hyperthyroidism (PEDNECK-6.2)

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- For the initial study for evaluation of hyperthyroidism:
  - Ultrasound Neck (CPT® 76536) is medically necessary.
    - If a nodule or mass is discovered on ultrasound, the individual should be imaged according to **Thyroid Masses or Nodules (PEDNECK-6.1)**.
- For all other individuals with documented hyperthyroidism:
  - Thyroid uptake nuclear imaging (either CPT® 78012 or CPT® 78014) is medically necessary.

Background and Supporting Information

- Common causes are Graves' disease and autoimmune disorders (lupus, rheumatoid arthritis, and Sjögren syndrome).

Evidence Discussion

- Ultrasound is used as the initial evaluation for thyroid abnormalities and follow up as it is noninvasive, radiation free, inexpensive, very sensitive, and does not require sedation to complete.[1,2]
- The use of a radioactive iodine uptake scan (although involves ionizing radiation exposure) is medically necessary if the diagnosis is unclear after ultrasound and laboratory evaluation have been completed.[4,5]

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Hypothyroidism (PEDNECK-6.3)

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- For the initial study for evaluation of hypothyroidism:
  - Ultrasound Neck (CPT® 76536) is medically necessary.
    - If a nodule or mass is discovered on ultrasound, the individual should be imaged according to **Thyroid Masses or Nodules (PEDNECK-6.1)**.
- For individuals with documented congenital hypothyroidism, thyroid uptake nuclear imaging (either CPT® 78012 or CPT® 78014) is medically necessary.

Background and Supporting Information

- Causes of pediatric hypothyroidism include thyroid congenital dysgenesis, dyshormonogenesis autoimmune thyroiditis, Hashimoto thyroiditis, subacute thyroiditis, and abnormality in the pituitary gland or hypothalamus. Congenital hypothyroidism is usually diagnosed in the neonate on a routine perinatal screening examination.

Evidence Discussion

- Ultrasound is used as the initial evaluation for thyroid abnormalities and follow up as it is noninvasive, radiation free, inexpensive, very sensitive, and does not require sedation to complete.[1,2]
- The use of a radioactive iodine uptake scan (although involves ionizing radiation exposure) is medically necessary if the diagnosis is unclear after ultrasound and laboratory evaluation have been completed.[4,5]

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Parathyroid Imaging (PEDNECK-6.4)

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- Parathyroid imaging indications in pediatric individuals are the same as those for adult individuals. See **Parathyroid Imaging (Neck-8.3)** in the Neck Imaging Guidelines.

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Esophagus (PEDNECK-7)

Guideline

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References (PEDNECK-7)

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- Esophagus imaging indications in pediatric individuals are very similar to those for adult individuals. See **Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)** in the Neck Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - For suspected foreign body ingestion or impaction:
    - Plain x-rays are medically necessary for initial imaging.
    - Ultrasound Neck (CPT® 76536) is medically necessary for evaluation of upper esophageal foreign bodies.
    - See **Dysphagia and Upper Digestive Tract Disorders (Neck-3.1)**.
  - For evaluating congenital atresia with associated tracheoesophageal fistula:
    - Esophagram is medically necessary.
  - For evaluation of suspected congenital malformations with inconclusive x-rays or esophagram:
    - CT Neck with contrast (CPT® 70491) **AND/OR**
    - CT Chest with contrast (CPT® 71260)
      - 3D rendering (CPT® 76376 or CPT® 76377) is medically necessary for preoperative planning in complex cases.
  - Plain radiographs alone usually suffice for the diagnosis of esophageal atresia and a contrast examination of the esophagus is not warranted but may be medically necessary for post-operative evaluation.

Evidence Discussion

- The initial evaluation for possible foreign body ingestion is plain x-rays, because of their availability and affordability.[1,8]
- Esophagram, in the setting of esophageal atresia with tracheoesophageal fistula, allows for diagnosis of the condition through the use of a nasogastric tube being withdrawn while instilling contrast material.[1]
- Chest x-ray is used as the initial evaluation of the mediastinum when concerns are present for a congenital abnormality because of availability and affordability.[1] Evaluation of vascular ring or sling includes an esophagram. For further evaluation, CT is favored over MRI secondary to availability, expedited time, and cost.[2,3]

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Esophagus Health Equity Considerations

Health Equity Considerations

- Higher levels of housing instability and material deprivation are associated with esophageal foreign body ingestion and complications.

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Trachea (PEDNECK-8)

Guideline

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Trachea (PEDNECK-8.1)

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- Trachea imaging indications in pediatric individuals are similar to those for adult individuals. See **Imaging of the Larynx, Trachea, and Bronchus (Neck-9.1)** in the Neck Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - For evaluation of suspected congenital malformations if x-rays are inconclusive:
    - CT Neck with contrast (CPT® 70491) **AND/OR**
    - CT Chest with contrast (CPT® 71260) are medically necessary.
      - If requested, 3D rendering (CPT® 76376 or CPT® 76377) is medically necessary for preoperative planning in complex cases.
  - CT Chest (either without contrast CPT® 71250 or with contrast CPT® 71260) is medically necessary in complicated cases or when bronchoscopy is being considered.

Evidence Discussion

- Chest x-ray, because of its availability and affordability, is generally regarded as the initial imaging study in evaluating possible foreign body aspiration. Chest x-ray has a sensitivity of 62-88% and a specificity of 30-97% of detecting airway foreign bodies.[1,2,12]
- CT may play a role in diagnosing foreign body aspiration in children with non-specific complaints such as fever and cough. CT of the chest has greater than 92% sensitivity and specificity and a diagnostic accuracy of 98% in detecting airway foreign bodies and can play a role in the individual with an unclear or atypical clinical picture.[1,10,11]
- Chest x-ray is used as the initial evaluation of the mediastinum when concerns are present for a congenital abnormality. For further evaluation, CT is favored over MRI secondary to availability, expedited time, and cost.[4]

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General Guidelines (PEDONC-1)

Guideline

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines	
AFP	alpha-fetoprotein (tumor marker)
ALCL	anaplastic large cell lymphoma
ALL	acute lymphoblastic leukemia
AML	acute myelogenous leukemia
ß-hCG	human chorionic gonadotropin beta-subunit (tumor marker)
BKL	Burkitt's lymphoma
BWT	bilateral Wilms tumor
CCSK	clear cell sarcoma of the kidney
CNS	central nervous system
COG	Children's Oncology group
CPT®	current procedural terminology; trademark of the American Medical Association
CSF	cerebrospinal fluid
CT	computed tomography
CXR	chest x-ray

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines	
DAWT	diffuse anaplasia Wilms tumor
ESFT	Ewing sarcoma family of tumors
FAWT	focal anaplasia Wilms tumor
FHWT	favorable histology Wilms tumor
HL	Hodgkin lymphoma
HSCT	hematopoietic stem cell transplant (bone marrow or peripheral blood)
HVA	homovanillic acid
LL	lymphoblastic lymphoma
MIBG	metaiodobenzylguanidine (nuclear scan using ^{123}i or ^{131}i)
MPNST	malignant peripheral nerve sheath tumor
MRI	magnetic resonance imaging
NBL	neuroblastoma
NED	no evidence of disease
NHL	non-Hodgkin lymphoma
NPC	nasopharyngeal carcinoma
NRSTS	nonrhabdomyosarcomatous soft tissue sarcomas
OS	osteosarcoma
PET	positron emission tomography
PMBCL	primary mediastinal b-cell lymphoma

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Abbreviations for Pediatric and Special Populations Oncology Imaging Guidelines	
PNET	primitive neuroectodermal tumor
RCC	renal cell carcinoma
RMS	rhabdomyosarcoma
US	ultrasound
VMA	vannilylmandelic acid
WBC	white blood cell count
XRT	radiation therapy

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General Guidelines (PEDONC-1.0)

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- A relevant clinical evaluation or meaningful contact (telephone call, electronic mail or messaging) should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled off therapy surveillance evaluation or cancer screening. The clinical evaluation may include a relevant history and physical examination, including biopsy, appropriate laboratory studies, and results of non-advanced or advanced imaging modalities.
  - Because of the relatively small number of childhood cancer treatment centers, it is common to combine off-therapy visits with imaging and other sub-specialist visits to accommodate families traveling long distances for their child's care.
- Unless otherwise stated in the disease-specific guideline, a histological confirmation of malignancy (or recurrence) and the stage of disease is required to perform a medical necessity review of the requested imaging.
- Unless otherwise stated in the disease-specific guideline, advanced imaging of asymptomatic individuals is not routinely medically necessary without signs or symptoms of systemic involvement of cancer.
- Conventional imaging performed prior to diagnosis should not be repeated unless there is a delay of at least 6 weeks since previous imaging and treatment initiation or there are new or significantly worsening clinical signs or symptoms
- Generally, the studies listed in the disease-specific sections reflect the studies supported by current literature and research for that condition. If a study is not listed, then it is not medically necessary.
- Routine imaging of brain, spine, neck, chest, abdomen, pelvis, bones, or other body areas is not medically necessary except where explicitly stated in a diagnosis-specific guideline section, or if one of the following applies:
  - Known prior disease involving the requested body area
  - New or worsening symptoms or physical exam findings involving the requested body area (including non-specific findings such as ascites or pleural effusion)
  - New finding on basic imaging study such as plain x-ray or ultrasound
  - New finding on adjacent body area CT/MRI study (i.e., pleural effusion observed on CT abdomen)
  - Unless otherwise stated in the disease-specific guideline, advanced imaging of asymptomatic individuals is not routinely supported without signs or symptoms of systemic involvement of cancer.

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- Repeat imaging studies are not generally medically necessary unless there is evidence of disease progression, recurrence of disease, and/or the repeat imaging will affect an individual's clinical management.

- Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is medically necessary after every 2 cycles, which is usually ~6 weeks of therapy for solid tumors and usually ~8 to 12 weeks for CNS tumors
- Unless otherwise specified for a specific cancer type, once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not medically necessary for continued disease monitoring or surveillance unless one of the following applies:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive or suspicious for recurrence
  - Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging
  - PET avidity in a residual mass at the end of planned therapy is not an indication for PET imaging during surveillance

Clarification of phases of therapy

Phases of Oncology Imaging	Definition
Screening	• Imaging requested for individuals at increased risk for a particular cancer in the absence of known clinical signs or symptoms • Screening using advancing imaging is only supported for conditions listed in **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)**
Suspected/Initial Staging	• All imaging studies requested from the time cancer is first clinically suspected until the initiation of specific treatment ◦ CT Chest prior to anesthesia for biopsy or resection of solid tumors and CTs of other involved body areas are generally indicated and should be performed concurrently ◦ Metastatic CNS imaging and nuclear medicine imaging are generally deferred until after a histologic diagnosis is made, unless otherwise indicated by diagnosis-specific guideline

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Phases of Oncology Imaging	Definition
Treatment Response	• Imaging performed during any type of active treatment (chemotherapy or other medications, radiation therapy, or surgery), including evaluation at the end of planned active treatment ◦ Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is medically necessary after every 2 cycles, which is usually ~6 weeks of therapy for solid tumors and usually ~8 to 12 weeks for CNS tumors
Surveillance	• Imaging performed in individuals who are asymptomatic or have chronic stable symptoms and not receiving any active treatment, even if residual imaging abnormalities are present ◦ PET imaging is not medically necessary for surveillance imaging unless specifically stated elsewhere in the diagnosis-specific guideline sections
Recurrence	• All imaging studies completed at the time a recurrence or progression of a known cancer is strongly suspected or documented based on clinical signs or symptoms, laboratory findings, or results of basic imaging studies such as plain radiography or ultrasound • Following documented recurrence of childhood cancer, any studies recommended for initial staging of that cancer type in the diagnosis-specific imaging guideline section should be approved ◦ During active treatment for recurrent pediatric cancer, conventional imaging evaluation (CT or MRI, should use the same modality for ongoing monitoring as much as possible) of previously involved areas is medically necessary according to the treatment response imaging in the diagnosis-specific guideline section ▪ Always refer to the diagnosis specific guideline for PET indications in recurrence.

- Brain imaging is performed for signs or symptoms of brain disease.
  - MRI Brain without and with contrast (CPT® 70553) is the medically necessary study for evaluation of suspected or known brain metastases. If a non-contrast CT head shows suspicious lesion, MRI Brain is medically necessary to further characterize the lesion.

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- CT Head without and with contrast (CPT® 70470) is medically necessary when MRI is contraindicated or not available, or if there is skull bone involvement.
- Certain malignancies including, but not limited to, melanoma, lung cancer, and renal cell cancer have indications for brain imaging for asymptomatic individuals,
- If stage IV disease is demonstrated elsewhere or if systemic disease progression is noted, refer to disease specific guidelines
- Initiation of angiogenesis therapy is not an indication for advanced imaging of the brain in asymptomatic individuals (Avastin/Bevacizumab; <3% risk of bleeding and <1% risk of serious bleeding)

- Bone Scan:
  - Primarily used for evaluation of bone metastases in individuals with solid malignancies.
  - Indications for bone scan in individuals with history of malignancy include – bone pain, rising tumor markers, elevated alkaline phosphatase, or in individuals with primary bone tumor.
  - For evaluation of suspected or known bony metastases, CPT® 78306 (Nuclear bone scan whole-body) is medically necessary.
  - Radiopharmaceutical Localization scan SPECT (CPT® 78803 or CPT® 78831) or SPECT/CT (CPT® 78830 or CPT® 78832) is medically necessary as an add-on test for further evaluation of a specific area of interest.
  - CPT® codes 78300 (Nuclear bone scan limited), 78305 (Nuclear bone scan multiple areas), or 78315 do not have any indications in oncology nuclear medicine imaging.
- Bone scan supplemented by plain x-rays are the initial imaging modalities for suspected malignant bone pain. For specific imaging indications, see also:
  - **Nuclear Medicine (NM) Imaging in Oncology (ONC-1.3)**
  - **Bone (Non-Vertebral) Metastases (ONC-31.5)**
  - **Spinal/Vertebral Metastases (ONC-31.6)**
  - **Carcinoma of Unknown Primary Site (ONC-31.7)**
- Delay PET/CT for at least 12 weeks after completion of radiation treatment, unless required sooner for imminent surgical resection.
- PET/CT may be considered prior to biopsy in order to determine a more favorable site for biopsy when a prior biopsy was nondiagnostic or a relatively inaccessible site is contemplated which would require invasive surgical intervention for biopsy attempt.
- PET/CT may be indicated if:
  - Conventional imaging (CT, MRI, or bone scan) reveals findings that are inconclusive or negative, with continued suspicion for recurrence
- **Unless specified in diagnosis-specific guideline section PET/CT Imaging is NOT medically necessary for**:

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- Infection, inflammation, trauma, post-operative healing, granulomatous disease, rheumatological conditions
- Concomitantly with separate diagnostic CT studies
- Conclusive evidence of distant or diffuse metastatic disease on recent conventional imaging studies
- Metastatic disease in the central nervous system (CNS)
- Lesions less than 8 mm in size
- Follow up after localized therapy (i.e., radiofrequency ablation, embolization, stereotactic radiation, etc.)
- Rare malignancies, due to lack of available evidence regarding the diagnostic accuracy of PET in rare cancers
- Surveillance
  - Serial monitoring of individuals who are not currently receiving anti-tumor treatment or are receiving maintenance treatment
  - Serial monitoring of FDG avidity until resolution.
  - PET/CT avidity in a residual mass at the end of planned therapy is not an indication for PET/CT imaging during surveillance.
  - Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging

- Please refer to general guidelines section **PET Imaging in Oncology (ONC-1.4)** and **PET Imaging in Pediatric Oncology (PEDONC-1.4)** for further guidance regarding PET. Those guidelines should be applied with regard to radiotracer coverage.
- Please refer to general guidelines in **Unlisted Procedure Codes in Oncology (ONC-1.5)** for unlisted procedures in pediatric oncology.

Clinical Trials

- Similar to investigational and experimental studies, clinical trial imaging requests will be considered to determine whether they meet these evidence-based guidelines.
- Imaging studies which are inconsistent with established clinical standards, or are requested for data collection and not used in direct clinical management are not medically necessary.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence;

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education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Age Considerations (PEDONC-1.1)

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- The majority of malignancies occurring in the pediatric population are different diagnoses than those occurring in the adult population.
- For those diseases which occur in both pediatric and adult populations, minor differences may exist in management between pediatric and adult medical oncologists due to the following:
  - Age of the individual
  - Comorbidities
  - Differences in disease natural history between children and adults

AGE APPROPRIATE GUIDELINES	
Age of Individual	**Appropriate Imaging Guidelines**
≥18 years old at initial diagnosis	• General Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section
<18 years old at initial diagnosis	• Pediatric Oncology Imaging Guidelines, except where directed otherwise by a specific guideline section
15 to 39 years old at initial diagnosis (defined as Adolescent and Young Adult (AYA) oncology individuals)	• When unique guidelines for a specific cancer type exist only in either Oncology or Pediatric Oncology, AYA individuals should be imaged according to the guideline section for their specific cancer type, regardless of the individual's age • When unique guidelines for a specific cancer type exist in both Oncology and Pediatric Oncology, AYA individuals should be imaged according to the age rule in the previous bullet

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Appropriate Clinical Evaluations (PEDONC-1.2)

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- The majority of pediatric oncology imaging indications are listed in the diagnosis-specific guideline sections
  - Rare malignancies and other circumstances not specifically addressed elsewhere in the Pediatric Oncology guidelines, the following general principles apply:
    - Routine imaging of brain, spine, neck, chest, abdomen, pelvis, bones, or other body areas is not medically necessary in the absence of:
      - Localizing symptoms or
      - Abnormalities on plain radiography or ultrasound
- The overwhelming majority of pediatric oncology individuals treated in the United States will be enrolled on or treated according to recent Children's Oncology Group (COG) protocols
  - COG is a National Cancer Institute (NCI) supported clinical trials group
  - These imaging guidelines are generally consistent with evaluations recommended by COG protocols commonly used for direct individual care (whether formally enrolled on study or not)
    - Requests for exception to guidelines based on COG protocol should be accompanied by the COG road map or COG details in the medical record for case-by-case consideration.
  - For individuals enrolled in a COG study, imaging recommended by COG protocols is generally medically necessary unless the imaging is being performed solely to address a study objective and would not be indicated in usual clinical care.
    - Requests for exception to guidelines based on COG protocol should be accompanied by the COG road map or COG details in the medical record for case-by-case consideration.

Phases of Pediatric Oncology Imaging:

- Pediatric malignancies, in general, behave more aggressively than adult cancers, and the time from initial suspicion of cancer to specific therapy initiation can be measured in hours to days for most pediatric cancers
- It is recommended that children with pediatric solid tumors undergo CT evaluation of the chest prior to general anesthesia for biopsy or resection due to the risk of post-operative atelectasis mimicking pulmonary metastasis resulting in inaccurate staging and/or delay in therapy initiation

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- Unlike adult cancers, in most pediatric cancers surveillance does not begin until all planned multi-modal therapy is completed
- Pediatric cancers where surgical resection is considered curative are listed in the diagnosis-specific guideline sections
- Certain tumor types do not require surveillance with advanced imaging as individual outcomes following relapse are not improved by surveillance imaging. See diagnosis-specific guideline sections for details
- Imaging may be indicated more frequently than recommended by guidelines with clinical documentation that the imaging results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance

Phases of Oncology Imaging	Definition
Screening	• Imaging requested for individuals at increased risk for a particular cancer in the absence of known clinical signs or symptoms • Screening using advancing imaging is only supported for conditions listed in **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)**
Suspected/Initial Staging	• All imaging studies requested from the time cancer is first clinically suspected until the initiation of specific treatment ◦ CT Chest prior to anesthesia for biopsy or resection of solid tumors and CTs of other involved body areas are generally indicated and should be performed concurrently ◦ Metastatic CNS imaging and nuclear medicine imaging are generally deferred until after a histologic diagnosis is made, unless otherwise indicated by diagnosis-specific guideline
Treatment Response	• Imaging performed during any type of active treatment (chemotherapy or other medications, radiation therapy, or surgery), including evaluation at the end of planned active treatment ◦ Unless otherwise stated in the diagnosis-specific guidelines, imaging for treatment response is medically necessary after every 2 cycles, which is usually ~6 weeks of therapy for solid tumors and usually ~8 to 12 weeks for CNS tumors

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Phases of Oncology Imaging	Definition
Surveillance	• Imaging performed in individuals who are asymptomatic or have chronic stable symptoms and not receiving any active treatment, even if residual imaging abnormalities are present ◦ PET imaging is not medically necessary for surveillance imaging unless specifically stated in elsewhere in the diagnosis-specific guideline sections
Recurrence	• All imaging studies completed at the time a recurrence or progression of a known cancer is strongly suspected or documented based on clinical signs or symptoms, laboratory findings, or results of basic imaging studies such as plain radiography or ultrasound • Following documented recurrence of childhood cancer, any studies recommended for initial staging of that cancer type in the diagnosis-specific imaging guideline section should be approved ◦ During active treatment for recurrent pediatric cancer, conventional imaging evaluation (CT or MRI, should use the same modality for ongoing monitoring as much as possible) of previously involved areas is medically necessary according to the treatment response imaging in the diagnosis-specific guideline section ▪ Always refer to the diagnosis specific guideline for PET indications in recurrence.

Radiation Treatment Planning In Pediatric Oncology

- Imaging performed in support of radiation therapy treatment planning should follow guidelines outlined in **Unlisted Procedure Codes in Oncology (ONC-1.5).**

Cardiac Function Assessment in Pediatric Oncology During Active Treatment:

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Indication	Medically Necessary Imaging Study
• Evaluation of cardiac function prior to cardiotoxic chemotherapy, and for monitoring while on active therapy or at end of therapy at the discretion of the treating pediatric oncologist based on: ◦ Cumulative cardiotoxic therapy received to date ◦ Individual's age and gender ◦ Most recent echocardiogram results ◦ New or worsening cardiac symptoms	• Echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308)
• For either of the following: ◦ Echocardiography yielded a borderline shortening fraction (<30%) and additional left ventricular function data are necessary to make a chemotherapy decision OR ◦ Echocardiography windowing is suboptimal due to body habitus or tumor location	• Multi-gated acquisition (MUGA, CPT® 78472) blood pool nuclear medicine scanning

Background and Supporting Information

Immunosuppression During Pediatric Cancer Therapy and Imaging Ramifications:

- Individuals may be severely immunocompromised during active chemotherapy treatment and any conventional imaging request to evaluate for infectious complications during this time frame should be approved immediately
  - Imaging requests for infectious disease concerns for all individuals with absolute neutrophil count (ANC) <500 **or**
  - Inconclusive findings on chest x-ray or US at any ANC during active treatment should be approved as requested
  - Individuals may have therapy-induced hypogammaglobulinemia which requires supplemental intravenous immune globulin (IVIG) during maintenance therapy. These individuals receiving supplemental IVIG should be treated similarly to individuals with ANC <500 with regards to imaging for infectious disease

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- Some individuals are treated with very intensive chemotherapy regimens (including autologous stem cell transplantation) and spend the majority of their chemotherapy treatment phase in the hospital. See: **Hematopoietic Stem Cell Transplantation (ONC-29).**
- Due to the high risk of invasive infections, frequent CT may be indicated to evaluate known sites of invasive fungal infection, and in general these should be approved as requested
  - Surveillance imaging of asymptomatic individuals to detect invasive fungal infection has not been shown to impact individual outcomes
    - Imaging requests are indicated when acute clinical decisions will be made based on the imaging

Hematopoietic Stem Cell Transplant (HSCT) in Pediatric Oncology:

- Transplantation of hematopoietic stem cells from bone marrow, peripheral blood, or cord blood is commonly used in the following clinical situations:
  - High-risk or recurrent leukemia (allogeneic)
  - Recurrent lymphoma (allogeneic or autologous)
  - Hemophagocytic lymphohistiocytosis (allogeneic)
  - High-risk sickle cell disease (allogeneic)
  - High-risk neuroblastoma (autologous)
  - High-risk CNS tumors (autologous)
  - Recurrent Ewing sarcoma family of tumors (autologous, rarely allogeneic)
- Imaging considerations for HSCT should follow guidelines in: **Hematopoietic Stem Cell Transplantation (ONC-29).**

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Modality General Considerations (PEDONC-1.3)

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- Plain radiography
  - Chest x-ray (CXR) can provide a prompt means to evaluate primary intrathoracic tumors and continues to be the initial imaging study recommended to detect complications, such as suspected infection, in symptomatic individuals undergoing treatment.
  - CXR continues to be the initial imaging study recommended for pulmonary surveillance for some pediatric cancers. See diagnosis-specific guideline sections for details.
  - Plain radiography continues to be the initial imaging study recommended for evaluation of lesions involving the appendicular skeleton, both during and after completion of treatment. See diagnosis-specific guideline sections for details.
  - Plain abdominal radiographs have largely been replaced by ultrasound, CT, or MRI.
- Ultrasound
  - Ultrasound is not widely used in pediatric oncology for staging but is frequently used for surveillance in individuals who have successfully treated (primarily abdominal or pelvic) tumors with little or no residual disease.
  - See diagnosis-specific guideline sections for details.
- CT
  - CT with contrast is the imaging study of choice in pediatric individuals with lymphomas or solid tumors of the neck, thorax, abdomen, and/or pelvis
    - If CT contrast use is contraindicated due to allergy or impaired renal function, either CT without contrast or MRI without and with contrast may be substituted at the discretion of the ordering physician
- MRI
  - MRI without and with contrast is the study of choice for CNS tumors and musculoskeletal tumors
    - If MRI contrast use is contraindicated due to allergy or impaired renal function, MRI without contrast may be substituted at the discretion of the ordering physician
  - Due to the length of time for image acquisition and the need for stillness, anesthesia is required for almost all infants and young children (age <7 years), as well as older children with delays in development or maturity. In this population, MRI imaging sessions should be planned with a goal of avoiding a short-interval

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repeat anesthesia exposure due to insufficient information using the following considerations:

- MRI should generally be performed without and with contrast unless there is a specific contraindication to gadolinium use since the individual already has intravenous access for anesthesia
  - Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast
  - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAS) use is not warranted at this time
  - It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAS should be assessed
  - If requesting clinicians indicate that a non-contrast study is being requested due to concerns regarding the use of gadolinium, the exam can be approved

◦ In some instances, to reduce time under anesthesia for MRI in pediatric individuals, surveillance or restaging studies may be requested with contrast only when it is determined that repeat non-contrast imaging does not add to an individual clinical case. These may be approved on a case-by-case basis.

- If multiple body areas are supported by these guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session

• Nuclear medicine

◦ General PET imaging consideration can be found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**.

◦ Bone scan is frequently used for evaluation of bone metastases in pediatric oncology during initial treatment, treatment response, and surveillance

- CPT® 78315 has no specific indications for evaluation of malignant disease

◦ ^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy is the preferred metabolic imaging for neuroblastoma and is positive in 90% to 95% of neuroblastomas. MIBG is also used for evaluation of pheochromocytomas, paragangliomas, ganglioneuromas, and ganglioneuroblatomas, PET/CT indications are provided in the relevant sections.

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Study Type	Coding
Bone scan	• ANY of the following codes can be approved: ◦ CPT® 78300 ◦ CPT® 78305 ◦ CPT® 78306 ◦ CPT® 78803, 78830, or 78832 ▪ May be approved alone or in combination with: - CPT® 78305 - CPT® 78306
^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy	• ANY one of the following codes can be approved: ◦ CPT® 78801 ◦ CPT® 78802 ◦ CPT® 78804 • ANY one of the following codes may also be approved, individual or in combination with CPT® 78801, 78802, or 78804: ◦ CPT® 78803 ◦ CPT® 78830 ◦ CPT® 78831 ◦ CPT® 78832 ◦ CPT® 78800 may be approved for KNOWN neuroblastoma when only a single site follow up is desired but is not sufficient for the initial workup of suspected disease
Octreotide scan	Same coding as MIBG
Gallium scan	Same coding as MIBG

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PET Imaging in Pediatric Oncology (PEDONC-1.4)

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Throughout these guidelines, unless otherwise specified, the term "PET" refers specifically to ^{18}F-FDG-PET imaging and also applies to PET/CT fusion studies.

- PET imaging in pediatric oncology should use PET/CT fusion imaging (CPT® 78815 or CPT® 78816)
- The decision whether to use skull base to mid-femur ("eyes to thighs") procedure code for PET (CPT® 78815) or whole-body PET (CPT® 78816) is addressed in the diagnosis-specific guideline sections.
- PET imaging in oncology should use PET/CT fusion imaging (CPT® 78815 or CPT® 78816)
- PET/MRI is generally not supported for a vast majority of oncologic conditions due to lack of standardization in imaging technique and interpretation. However, it is medically necessary in select circumstances when all of the following criteria are met:
  - The individual meets condition-specific guidelines for PET/MRI OR
  - The individual meets ALL of the following:
    - The individual meets guideline criteria for PET/CT, AND
    - PET/CT is not available at the treating institution, AND
    - The provider requests PET/MRI in lieu of PET/CT
  - When the above criteria are met, PET/MRI is medically necessary using the code combination of PET Whole-Body (CPT® 78813) and MRI Unlisted (CPT® 76498). All other methods of reporting PET/MRI are inappropriate.
    - When clinically appropriate, diagnostic MRI codes may be medically necessaryat the same time as the PET/MRI code combination.
- Unbundling PET/CT imaging into separate PET (such as CPT® 78812 or CPT® 78813) and diagnostic CT codes is otherwise not medically necessary.
- PET imaging is not reliable for the detection of anatomic lesions smaller than 8 mm in size.
- Delay PET/CT for at least 12 weeks after completion of radiation treatment, unless required sooner for imminent surgical resection.
- PET imaging using isotopes other than ^{18}F-FDG, ^{68}Ga-DOTATATE, or ^{68}Ga-DOTATOC is considered not medically necessary at this time.
- PET has not been shown to be diagnostically useful in all forms of childhood cancer. PET is medically necessary for pediatric malignancies with significant published

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evidence regarding its diagnostic accuracy and importance in accurately directing individual care decisions. See diagnosis-specific guideline sections for details.

- PET imaging is not specific to cancer and has a high rate of false positivity. Inflammation, infection (especially granulomatous), trauma, and post-operative healing may show high levels of FDG uptake and be false-positive for malignant lesions.
- PET for rare malignancies not specifically addressed by these guidelines is generally not medically necessary due to lack of available evidence regarding diagnostic accuracy of PET in the majority of rare cancers. Conventional imaging studies should be used for initial staging and treatment response for these diagnoses.
- PET is medically necessary if **all** of the following apply:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are equivocal or suspicious
  - No other specific metabolic imaging (MIBG, octreotide, technetium, etc.) Is appropriate for the cancer type
  - The submitted clinical information describes a specific decision regarding the individual's care that will be made based on the PET results
- PET imaging for surveillance imaging **only** when specifically stated elsewhere in the diagnosis-specific guideline sections
- Unless otherwise specified for a specific cancer type, once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not medically necessary for continued disease monitoring or surveillance unless **one** of the following applies:
  - Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive or suspicious for recurrence
    - Residual mass that has not changed in size since the last conventional imaging does **not** justify PET imaging
    - PET avidity in a residual mass at the end of planned therapy is **not** an indication for PET imaging during surveillance.
  - Very rare circumstances where tumor markers or obvious clinical symptoms show strong evidence suggesting recurrence and PET would replace conventional imaging modalities
  - The individual is undergoing salvage treatment for a recurrent solid tumor with residual measurable disease on conventional imaging and confirmed repeat negative PET imaging will allow the individual to transition from active treatment to surveillance

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Diagnostic Radiation Exposure in Pediatric Oncology (PEDONC-1.5)

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- Young children are presumed to be at increased risk for malignancy from diagnostic radiation exposure, most commonly from CT and nuclear medicine imaging. They are more sensitive to radiation than adults and generally live longer after receiving radiation doses from medical procedures, resulting in a larger number of years during which to manifest a cancer.
- Because of this presumed increased risk in young children, requests to substitute MRI without and with contrast for CT with contrast to avoid radiation exposure are medically necessary if ALL of the following criteria apply:
  - The individual is presently a young child and the ordering physician has documented the reason for MRI, rather than CT, is to avoid radiation exposure.
  - The disease-specific guidelines do not list CT as superior to MRI for the current disease and time point, meaning the MRI will provide equivalent or superior information relative to CT.
  - The request is for a body area other than Chest as MRI is substantially inferior to CT for detection of small pulmonary metastases.
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)

Guideline

Screening Imaging in Cancer Predisposition Syndromes – General Considerations (PEDONC-2.1)
Li-Fraumeni Syndrome (LFS) (PEDONC-2.2)
Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)
Beckwith-Wiedemann Syndrome (BWS) (PEDONC-2.4)
Denys-Drash Syndrome (DDS) (PEDONC-2.5)
Wilms Tumor-Aniridia-Growth Retardation (WAGR) (PEDONC-2.6)
Familial Adenomatous Polyposis (FAP) and Related Conditions (PEDONC-2.7)
Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)
Tuberous Sclerosis Complex (TSC) (PEDONC-2.9)
Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)
Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)
Familial Retinoblastoma Syndrome (PEDONC-2.12)
Hereditary Paraganglioma-Pheochromocytoma (HPP) Syndromes (PEDONC-2.13)
Costello Syndrome (PEDONC-2.14)
Constitutional Mismatch Repair Deficiency (CMMRD or Turcot Syndrome) (PEDONC-2.15)
Hereditary Leiomyomatosis and Renal Cell Cancer (HLRCC) (PEDONC-2.16)
Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)
Infantile Myofibromatosis (PEDONC-2.18)
Bloom Syndrome (PEDONC-2.19)
References (PEDONC-2)

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Screening Imaging in Cancer Predisposition Syndromes – General Considerations (PEDONC-2.1)

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- This section is intended to give guidance for screening imaging prior to diagnosis with a specific malignancy. Once an individual with a cancer predisposition syndrome has been diagnosed with a malignant disease, future imaging decisions should be guided by the appropriate disease-specific guidelines except as explicitly stated elsewhere in this section.
- This section's guidelines are limited to cancer predisposition syndromes with screening imaging considerations. Syndromes requiring only clinical or laboratory screening are not discussed here.
- In general, a relevant detailed history and physical examination and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled screening evaluation identified in this section.
- Where MRI is indicated in these guidelines, CT may be approved only if MRI is contraindicated, given the risk of radiation exposure in these syndromes.
- Many of these cancer predisposition syndromes also affect adults as survival continues to improve for these individuals.
  - Adults with syndromes covered in this section may follow these imaging guidelines except where contradicted by specific statements in the general imaging guidelines.
- Documentation of genetic or molecular confirmation of the appropriate syndrome with increased cancer risk is preferred for any individual to qualify for screening imaging. There are a number of complex ethical, social, and financial issues involved in the decision to complete genetic testing in a pediatric individual.
  - From the 2013 AAP Policy Statement, "Predictive genetic testing for adult-onset conditions generally should be deferred unless an intervention initiated in childhood may reduce morbidity or mortality." **Imaging surveillance is one such intervention and should not be performed without justifiable cause**.
  - Genetic testing should be performed in conjunction with genetic counseling for appropriate communication of risks identified by testing.
  - When genetic testing is not possible, formal diagnosis after evaluation by a physician with significant training and/or experience in cancer predisposition

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syndromes (most commonly a geneticist or oncologist) is generally sufficient to confirm eligibility for screening imaging.

- Due to the length of time for image acquisition and the need for stillness, anesthesia is required for almost all infants and young children (age < 7 years), as well as older children with delays in development or maturity. In this individual population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:
  - MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use since the individual already has intravenous access for anesthesia.
    - Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
    - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If requesting clinicians indicate that a non-contrast study is being requested due to concerns regarding the use of gadolinium, the exam can be approved.
    - In some instances, to reduce time under anesthesia for MRI in pediatric individuals, surveillance or restaging studies may be requested with contrast only when it is determined that repeat non-contrast imaging does not add to an individual clinical case. These may be approved on a case-by-case basis.
  - If multiple body areas are supported by these guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session

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Li-Fraumeni Syndrome (LFS) (PEDONC-2.2)

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The following imaging studies should be considered medically necessary in individuals with LFS:

Indication	Medically Necessary Imaging Study
All individuals	• BOTH of the following, annually: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ Whole-body MRI (WBMRI, CPT® 76498) ▪ CPT® 76498 is the only code for whole-body MRI at this time. • Every 3 months from birth: ◦ Abdominal (CPT® 76700) and pelvic (CPT® 76856) ultrasound • Beginning at age 20: ◦ Annual Breast MRI (CPT® 77049), alternating every 6 months with breast ultrasound, in addition to the previously noted annual MRI studies.
Documented signs or symptoms suggestive of possible malignancy	• Targeted MRI without and with contrast of the involved body area(s)

Background and Supporting Information

- LFS - syndrome inherited in an autosomal dominant manner (50% risk to offspring) associated with germline mutations in *TP53* resulted in an increased susceptibility to a variety of cancers.
  - Eighty percent of individuals will have germline *TP53* mutation:
    - Tumor-specific *TP53* mutations are much more common than germline *TP53* mutations and are not associated with an increased risk for subsequent cancers.
    - If *TP53*-negative, formal diagnosis of LFS should be assigned by a physician with significant training and/or experience in LFS (most commonly a geneticist or oncologist) based on specified clinical criteria prior to beginning a screening imaging program.

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    - *TP53* mutations may be present in 50% to 80% of pediatric adrenocortical carcinoma, 10% of pediatric rhabdomyosarcoma, and 10% of pediatric osteosarcoma.
- Individuals with LFS have an increased sensitivity to ionizing radiation, so screening strategies resulting in significant radiation exposure are not appropriate (CT and nuclear medicine).
- When a specific malignancy is suspected, the individual should be imaged according to the imaging guideline specific to the suspected cancer type.
- Annual complete detailed physical examinations, complete blood counts, and urinalyses form the backbone of LFS cancer screening.

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Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)

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The following imaging studies should be considered medically necessary in individuals with NF1:

Indication	Medically Necessary Imaging Study
• Clarification of the diagnosis of NF1 if evaluation by a physician with significant training and/or experience in neurofibromatosis is inconclusive ◦ Most commonly a neurologist, geneticist, ophthalmologist, or oncologist	• One-time MRI Brain (CPT® 70553) and Orbits (CPT® 70543) without and with contrast
• New or worsening neurological or visual symptoms	• MRI Brain (CPT® 70553) and Orbits (CPT® 70543) without and with contrast
• Clinical symptoms suggestive of change in a known plexiform neurofibroma ◦ Examples include: pain, rapid growth, and neurologic dysfunction	• MRI without and with contrast
• Clinical symptoms concerning for malignant transformation of a known plexiform neurofibroma, and ALL of the following are met: ◦ Recent MRI is inconclusive regarding transformation or progression ◦ Negative PET will result in a decision to avoid biopsy in a difficult or morbid location	• PET/CT (CPT® 78815 or 78816)
• Baseline tumor burden at age 16 or older (one-time imaging)	• WBMBRI (CPT® 76498)

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Indication	Medically Necessary Imaging Study
• New soft tissue mass(es)	• See: Soft tissue mass information in **Soft Tissue Mass and Morton's Neuroma (MS 10.1)** in the Musculoskeletal Imaging Guidelines or **Soft Tissue and Bone Masses – General Considerations (PEDMS 3.1)** in the Pediatric Musculoskeletal Imaging Guidelines depending on the individual's age at the time the mass is discovered. Plain x-ray or ultrasound is not required prior to advanced imaging in these individuals.
• New bone mass(es)	• See: **Bone Tumors - General Considerations (PEDONC-9.1)**
• Documented optic pathway gliomas	• See: **Intracranial Low Grade Gliomas (PEDONC-4.2)**
• Known plexiform neurofibromas	• MRI without and with contrast of a known body area containing a neurofibroma is indicated for any of the following: ◦ Every 3 months for treatment response in individuals receiving active treatment ◦ New or worsening clinical symptoms suggesting progression ◦ Preoperative planning
• Biopsy-proven MPNST in individuals with known NF-1 or NF-2	• For adult individuals, see: **ONC-12 Sarcomas-bone, soft tissue and GIST** • For pediatric individuals, see: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)**

Background and Supporting Information

NF1:

- Common syndrome inherited in an autosomal dominant manner (50% risk to offspring) affecting 1 in 2500 people. The diagnosis is commonly made based on

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established clinical criteria including café-au-lait spots, Lisch nodules of the iris, axillary freckling, family history, and the presence of NF-associated tumors.
- Genetic testing is encouraged for children with possible NF1 and no family history prior to assigning a diagnosis, but will not identify a mutation for all individuals with NF1.
- The majority of tumors are benign in nature, but malignant degeneration can occur.
- NF1-affected persons have increased sensitivity to ionizing radiation, so CT and nuclear medicine imaging are not appropriate screening or surveillance studies for these individuals.
- CT and/or nuclear medicine studies may be indicated for acute clinical situations and should be judged on a case-by-case basis.
- Annual ophthalmology evaluation is strongly recommended beginning at the time of diagnosis of NF1 to evaluate for optic pathway abnormalities:
  - Screening MRIs Brain (CPT® 70553) and Orbits (CPT® 70543) for asymptomatic individuals are **not** generally recommended due to the ~60% rate of unidentified bright objects (UBOs, T2-weighted signal abnormalities) which mostly disappear by age 30
    - Routine follow up imaging of UBOs is not warranted in the absence of acute symptoms suggesting new or worsening intracranial disease
- Individuals with NF1 are at increased risk for plexiform neurofibromas (PN) and malignant peripheral nerve sheath tumors (MPNST— a high grade sarcoma)
  - Screening imaging of asymptomatic individuals for these tumors is not supported by evidence. PET imaging is not supported for PN surveillance in asymptomatic individuals at this time as the positive predictive value is only 60% to 65% even in symptomatic individuals.
- Although PET imaging has a positive predictive value of only 61% to 63% in NF1 with suspected transformation to MPNST, the negative predictive value is high (96% to 99%)
  - Considerations for PET/CT coding and indications/coding for PET/MRI are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
  - Inconclusive PET findings should lead to biopsy of the concerning lesion.
  - Repeat PET studies are not indicated due to the poor positive predictive value in this setting.
- 2017 AACR recommendations support a single baseline Whole-Body MRI to assess tumor burden in late adolescence or young adulthood. Further imaging should be based on focused MRI for symptomatic changes or pre-operative planning, and further surveillance WBMRI are not supported.

The following imaging studies should be considered medically necessary in individuals with NF2:

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Indication	Medically Necessary Imaging Study
• All individuals, beginning at age 10 years	• Annual MRI Brain without and with contrast (CPT® 70553)
• All individuals without a history of spinal tumors, beginning at age 10 years	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 3 years
• All individuals with a history of spinal tumors	• Annual MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
• Clinical symptoms of intracranial mass or vestibular disease	• MRI Brain without and with contrast (CPT® 70553)
• Any of the following: ◦ Clinical symptoms suggestive of new or progressive spinal or paraspinal tumors, including uncomplicated back pain or radiculopathy ◦ Recent diagnosis with a meningioma or vestibular schwannoma	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
• Surveillance for progression of unresected tumors in individuals with known vestibular schwannoma	• Annual MRI Brain without and with contrast (CPT® 70553)
• Known vestibular schwannomas and any of the following: ◦ New or worsening clinical symptoms including hearing loss ◦ Preoperative planning	• MRI Brain without and with contrast (CPT® 70553)
• Known meningioma	• See: **Meningiomas (ONC-2.8)** in the Oncology Imaging Guidelines
• Known ependymoma	• See: **Ependymoma (PEDONC-4.8)**

Background and Supporting Information

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- NF2 is substantially less common than NF1. It is inherited in an autosomal dominant manner (50% risk to offspring) affecting ~1 in 25000 people. NF2 is associated with increased risk for meningiomas (50% of affected individuals), vestibular schwannomas, and spinal tumors (75% of affected individuals).

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Beckwith-Wiedemann Syndrome (BWS) (PEDONC-2.4)

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The following imaging studies should be considered medically necessary in individuals with BWS:

Indication	Medically Necessary Imaging Study
• All individuals, from birth to the 8th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• Purely cystic adrenal mass found on screening ultrasound	• Continue screening ultrasound (CPT® 76700 or 76705) every 3 months without additional imaging (i.e., advanced imaging is not medically necessary for a purely cystic adrenal mass)
• Solid or mixed adrenal mass found on screening ultrasound AND: ◦ Individual age 0 months to 5 months, and ◦ Mass 0 cm to 3 cm in diameter	• MIBG imaging (See: **PEDONC-1.3** for coding) and either CT or MRI Abdomen (contrast as requested)
• Solid or mixed adrenal mass found on screening ultrasound AND: ◦ Individual age 0 to 5 months, and ◦ Mass >3 cm in diameter	• MIBG imaging (See: **PEDONC-1.3** for coding) and MRI Abdomen (contrast as requested)
• Solid or mixed adrenal masses on screening ultrasound AND: ◦ Individual age 6 months or greater	• MIBG imaging (See: **PEDONC-1.3** for coding) prior to biopsy or resection ◦ If no evidence of malignancy on biopsy or resection, resume screening abdominal ultrasound every 3 months
• Solid or mixed adrenal masses on screening ultrasound AND: ◦ No evidence of malignancy based on MIBG, CT or MRI, Urine HVA/ VMA, and serum ACTH	• Repeat abdominal ultrasound (CPT® 76700 or 76705) every 6 weeks for 2 years

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Indication	Medically Necessary Imaging Study
• Known renal tumors	• See: **Pediatric Renal Tumors (PEDONC-7)**
• Known hepatoblastoma	• See: **Hepatoblastoma (PEDONC-11.2)**
• Known neuroblastoma	• See: **Neuroblastoma (PEDONC-6)**
• Known adrenocortical carcinoma	• See: **Pediatric Adrenocortical Carcinoma (PEDONC-14)**
• Known pheochromocytoma	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

Background and Supporting Information

- Inherited syndrome characterized by macroglossia, hemihypertrophy, macrosomia, organomegaly, and neonatal hypoglycemia. Individuals with isolated hemihypertrophy are also imaged according to this guideline.
- Caused by mutation at chromosome 11p15, affected children are predisposed to Wilms tumor, hepatoblastoma, rhabdomyosarcoma, and adrenal tumors.

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Denys-Drash Syndrome (DDS) (PEDONC-2.5)

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The following imaging studies should be considered appropriate in individuals with DDS:

Indication	Medically Necessary Imaging Study
• All individuals, from birth to the 8th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• Known renal tumors	• See: **Pediatric Renal Tumors (PEDONC-7)**

Background and Supporting Information

- Characterized by pseudohermaphroditism, early renal failure, and >90% risk of Wilms tumor development in each kidney. Associated with mutations at 11p13, risk of renal failure after detection of symptomatic Wilms tumor is 62%, so early detection may allow for renal-sparing surgical approaches.

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Wilms Tumor-Aniridia-Growth Retardation (WAGR) (PEDONC-2.6)

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The following imaging studies should be considered medically necessary in individuals with WAGR:

Indication	Medically Necessary Imaging Study
• All individuals, from birth to the 8th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• Known renal tumors	• See: **Pediatric Renal Tumors (PEDONC-7)**

Background and Supporting Information

- Named for the components of the disorder, it is associated with mutations at 11p13. As the name suggests, individuals are predisposed to Wilms tumor, with 57% of individuals in one cohort developing Wilms tumor. Risk of renal failure after detection of symptomatic Wilms tumor is 38%, so early detection may allow for renal-sparing surgical approaches.

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Familial Adenomatous Polyposis (FAP) and Related Conditions (PEDONC-2.7)

- Individuals with Lynch, Gardner, and Turcot syndromes should also be imaged according to these guidelines.

The following imaging studies should be considered medically necessary in individuals with FAP and related conditions:

- For colonoscopy and endoscopy guidance for those with FAP and related conditions, See: *EGD-1.16*, *CAPEND-5*, or *COLON-17: Genetic Syndromes* for the endoscopic management of polyposis syndromes.

Indication	Medically Necessary Imaging Study
• All individuals, from birth to the 6th birthday	• Abdominal ultrasound (CPT® 76700) every 3 months
• All individuals, beginning at age 12 years	• Annual thyroid ultrasound (CPT® 76536)
• All individuals, beginning at age 30 years	• Annual pelvic ultrasound (CPT® 76856)
• Family history of desmoid tumors	• Abdominal ultrasound (CPT® 76700) annually for life after age 6
• Individuals with Spigelman Stage III or IV or if duodenectomy is being planned • See: *Genetic Syndromes (EGD-1.16)* for additional information regarding Spigelman staging.	• MR Enterography (MRI Abdomen without and with contrast, CPT® 74183 and MRI Pelvis without and with contrast, CPT® 72197)
• Known colorectal tumors	• See: **Colorectal Cancer (ONC-16)** in the Oncology Imaging Guidelines
• Known desmoid tumors	• See: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)**

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Background and Supporting Information

- Inherited in an autosomal dominant manner (50% risk to offspring), it is also known as Adenomatous Polyposis Coli (APC). It is associated with the development of thousands of colonic polyps by age 20 and >90% risk of colorectal carcinoma. Prophylactic total colectomy is recommended by age 20 for most individuals. FAP is also associated with hepatoblastoma, tumors of the pancreas and small bowel, medulloblastoma, and thyroid cancer.

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Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)

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The following imaging studies should be considered medically necessary in individuals with MEN1:

Indication	Medically Necessary Imaging Study
All individuals, beginning at age 5 years	• Annual MRI Brain without and with contrast (CPT® 70553) • Annual MRI Abdomen without and with contrast (CPT® 74183), CT Abdomen with contrast (CPT® 74160), or ultrasound (CPT® 76700) • Annual Octreotide study (see: **PEDONC-1.3** for coding)
All individuals, beginning at age 15 years	• Annual MRI Chest without and with contrast (CPT® 71552) or CT Chest with contrast (CPT® 71260)
Known thyroid cancer	• See: **Thyroid Cancer (ONC-6)** in the Oncology Imaging Guidelines
Known pheochromocytoma	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

The following imaging studies should be considered medically necessary in individuals with MEN2a and MEN2b:

Indication	Medically Necessary Imaging Study
All individuals, beginning at age 5 years	• MRI Abdomen without and with contrast (CPT® 74183) every 3 years
• One of the following: ◦ Elevated catecholamines ◦ Inconclusive adrenal mass on MRI	• ONE of the following: ◦ Octreotide study (see: **PEDONC-1.3** for coding) ◦ Adrenal Nuclear Imaging (CPT® 78075)

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Indication	Medically Necessary Imaging Study
• Known pheochromocytoma	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)**

Background and Supporting Information

- Inherited in an autosomal dominant manner (50% risk to offspring)
- MEN1 is characterized by parathyroid, pancreatic islet cell, and pituitary gland tumors (3 P's), as well as carcinoid tumors in the chest and abdomen, and 28% of individuals will develop at least one tumor by age 15.
- MEN2a is characterized by medullary thyroid carcinoma, parathyroid adenomas, and pheochromocytomas.
- MEN2b is characterized by ganglioneuromas of the GI tract and skeletal abnormalities presenting in infancy.
- Recommended cancer screening includes:
  - Individuals with MEN2a and MEN2b receive annual measurement of catecholamines for pheochromocytoma screening

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Tuberous Sclerosis Complex (TSC) (PEDONC-2.9)

- Abdominal MRI (CPT® 74183) is medically necessary for women of childbearing age planning pregnancy.

The following imaging studies should be considered medically necessary in individuals with TSC:

Indication	Medically Necessary Imaging Study
• All individuals, at the time of suspected diagnosis until age 25 years	• Annual Brain MRI without and with contrast (CPT® 70553)
• All individuals at diagnosis	• Single baseline MRI Abdomen without and with contrast (CPT® 74183)
• If no renal lesions seen on baseline MRI Abdomen	• From diagnosis through age 11: ◦ Annual Renal US (CPT® 76770) • Age 12 (or 10 years earlier than the youngest family member with renal cell carcinoma, whichever comes earlier): ◦ MRI Abdomen without and with contrast (CPT® 74183) annually in lieu of ultrasound
• For documented renal lesions on baseline MRI Abdomen or any ultrasound	• MRI Abdomen without and with contrast (CPT® 74183) annually
• All individuals, beginning at age 18 years	• CT Chest without contrast (CPT® 71250) every 5 years
• All individuals, for cardiac screening or follow up of known cardiac disease	• See: **Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)** and **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** in the Pediatric Cardiology Imaging Guidelines
• Individuals with documented abnormalities on baseline CT Chest	• Additional CT Chest without contrast or with contrast (CPT® 71250 or 71260) every 1 year

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Indication	Medically Necessary Imaging Study
• Any of the following: ◦ New pulmonary symptoms ◦ Worsening pulmonary function testing	• CT Chest without contrast (CPT® 71250)
• Known SEGA tumors	• See: **Intracranial Low Grade Gliomas (LGG) (PEDONC-4.2)**
• Known renal cell carcinoma	• See: **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)**

Background and Supporting Information

- Inherited in an autosomal dominant manner (50% risk to offspring), affecting ~1 in 6000 individuals, it is associated with benign tumors, hypopigmented skin macules (ash leaf spots), developmental delay, and epilepsy.
- Malignancies associated with this syndrome include:
  - Subependymal giant cell astrocytomas (SEGA tumors)
    - Historically, early surgery was important to reduce morbidity related to these tumors
    - More recently, everolimus has been successfully used to treat these tumors without surgery, and early detection remains an important feature for success
  - Renal cell carcinoma
  - Cardiac rhabdomyosarcoma
  - Pulmonary lymphangioleiomyomatosis
- Recommended cancer screening includes:
  - Ophthalmologic and dermatologic evaluation annually

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Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)

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The following imaging studies should be considered medically necessary in individuals with VHL:

Indication	Medically Necessary Imaging Study
• All individuals, beginning at age 11 years	• MRI Brain without and with contrast (CPT® 70553) and MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, and Lumbar-CPT® 72158) every 2 years
• Individuals with frequent ear infections, prior to age 8	• One-time MRI Brain without and with contrast (CPT® 70553)
• All individuals, beginning at age 5 years	• Annual Abdominal US (CPT® 76700)
• All individuals, beginning at age 15 years	• MRI Abdomen without and with contrast (CPT® 74183) every 2 years
• ONE of the following: ◦ Elevated catecholamines ◦ Inconclusive adrenal mass on MRI	• Octreotide study (CPT® 78800, CPT® 78801, CPT® 78802, CPT® 78803, or CPT® 78804) or Adrenal Nuclear imaging (CPT® 78075)
• Individuals with known hemangioblastoma anywhere in the body (based on imaging) that has not been resected	• Both of the following, every 1 year: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, and Lumbar-CPT® 72158)
• Known hemangioblastoma that has not been resected and new or worsening symptoms	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, and Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
• Known (based on imaging) CNS hemangioblastoma	• See: **Intracranial Low Grade Gliomas (LGG) (PEDONC-4.2)**
• Known renal cell carcinoma	• See: **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)**
• Known pheochromocytoma or other neuroendocrine tumor	• See: **Neuroendocrine Cancers And Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

Background and Supporting Information

- Inherited in an autosomal dominant manner (50% risk to offspring), it is associated with CNS hemangioblastomas, retinal angiomas, endolymphatic sac tumors (ELST), gastrointestinal stromal tumor (GIST), renal cell carcinoma (RCC), and pheochromocytomas and other neuroendocrine tumors (NETs). Pediatric individuals are at risk of developing hemangioblastomas and pheochromocytomas that can remain clinically occult until symptoms become severe. Historically, substantial mortality was attributable to RCC, pancreatic NET, and CNS hemangioblastoma.
- Recommended cancer screening includes:
  - Annual ophthalmologic evaluation beginning at birth
  - Annual measurement of catecholamines beginning at age 2
  - Audiology assessment every 2 years beginning at age 5

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Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)

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The following imaging studies should be considered medically necessary in individuals with Rhabdoid Tumor Predisposition Syndrome:

Indication	Medically Necessary Imaging Study
• All individuals, at diagnosis, as early as birth if requested	• Whole-body MRI (WBMRI) (CPT® 76498)
• All individuals, from birth to 6 months of age	• ALL of the following, monthly: ◦ US Head (CPT® 76506) ◦ US Abdomen (CPT® 76700) ◦ US Pelvis (CPT® 76856) ◦ US Neck (CPT® 76536) ◦ MRI with and without contrast of areas of concern found on baseline WBMRI
• All individuals, from age 7 months to 5 years	• ALL of the following, every 3 months: ◦ US Abdomen (CPT® 76700) ◦ US Pelvis (CPT® 76856) ◦ US Neck (CPT® 76536) ◦ MRI Brain (CPT® 70553) ◦ MRI Spine (CPT® 72156, CPT® 72157, and CPT® 72158) without and with contrast ◦ MRI with and without contrast of areas of concern found on baseline WBMRI
• All individuals, after age 5 years	• Annual WBMRI (CPT® 76498)
• Clinical symptoms or WBMRI findings suggesting malignancy	• Targeted advanced imaging of the suspected disease site (CT with or without contrast, or MRI without and with contrast)

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Indication	Medically Necessary Imaging Study
• Inconclusive findings on ultrasound	• MRI with and without contrast of the inconclusive US site ◦ MRI is then medically necessary in place of ultrasound for remainder of planned screening

Background and Supporting Information

- Inherited in an autosomal dominant manner (50% risk to offspring), it is associated with malignant rhabdoid tumors of the kidney and extrarenal locations, and atypical teratoid/rhabdoid tumors (ATRT) of the CNS. It is caused by a germline mutation in *INI1* or *SMARCB1*, and is associated with a more variable prognosis than de novo rhabdoid tumors.

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Familial Retinoblastoma Syndrome (PEDONC-2.12)

- Regular physical and ophthalmologic evaluations under anesthesia (EUA) are the hallmark of surveillance strategies for these individuals, and asymptomatic screening imaging does not have a defined role at this time.

The following imaging studies should be considered medically necessary in individuals with Familial Retinoblastoma Syndrome:

Indication	Medically Necessary Imaging Study
• Individuals with retinomas (premalignant retinal lesions)	• Annual MRI Orbits (CPT® 70543)
• Either of the following: ◦ Inconclusive EUA findings ◦ New symptoms	• US Orbits (CPT® 76512, 76510, or 76511) or MRI Orbits (CPT® 70543) ◦ These studies should be used if at all possible in lieu of CT or nuclear imaging to avoid radiation exposure

Background and Supporting Information

- This syndrome is inherited in an autosomal dominant manner (50% risk to offspring). As the name suggests, it is associated with retinoblastoma, as well as osteosarcoma, pediatric melanoma, and a significantly increased risk for radiation-related malignancies.

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Hereditary Paraganglioma-Pheochromocytoma (HPP) Syndromes (PEDONC-2.13)

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The following imaging studies should be considered medically necessary in individuals with HPP Syndromes:

Indication	Medically Necessary Imaging Study
• All individuals with SDHx, MAX, TMEM127 mutations, beginning at age 6	• Every 2 years, ONE of the following sets of imaging: ◦ Whole-body MRI (CPT® 76498) ▪ CPT® 76498 is the only code for whole-body MRI at this time OR ◦ MRI Neck without and with contrast (CPT® 70543), MRI Chest without and with contrast (CPT® 71552), MRI Abdomen without and with contrast (CPT® 74183), and MRI Pelvis without and with contrast (CPT® 72197) ▪ If MRI cannot be performed: CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177) ▪ MRI Neck imaging should include skull base, thus separate MRI Brain (CPT® 70553) is only medically necessary if there are CNS-specific symptoms.
• Initial screening for individuals diagnosed at age 18 or older	• One-time PET/CT (CPT® 78815 or 78816), with 68Ga-DOTA-SSAs ◦ FDG may be substituted if Dotatate radiotracers are not available
• Known pheochromocytoma or other neuroendocrine tumors	• See: **Neuroendocrine Cancers and Adrenal Tumors (ONC-15)** in the Oncology Imaging Guidelines

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Background and Supporting Information

- Caused by mutations in *SDHx* and related genes, this syndrome is inherited in an autosomal dominant manner (50% risk to offspring), and is associated with pheochromocytomas and paragangliomas.
- Individuals with multiple endocrine neoplasias should not use this guideline and should be imaged according to **Multiple Endocrine Neoplasias (MEN) (PEDONC-2.8)**.
- MRI is preferred to CT to minimize radiation exposure given these individuals' lifelong need for screening
- All individuals with HPP receive annual measurement of catecholamines
- EANM–SNMMI joint guidelines propose the use of 68Ga-DOTA-SSAs PET as the first-choice functional imaging modality in adult SDHx mutation carriers and the use of 18F-FDG when 68Ga-DOTA-SSAs PET is not available.

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Costello Syndrome (PEDONC-2.14)

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The following imaging studies should be considered medically necessary in individuals with Costello Syndrome:

Indication	Medically Necessary Imaging Study
• Following confirmation of gene mutation	• ANY or ALL of the following: ◦ Echocardiogram ▪ CPT® 93306 or ▪ CPT® 93308 with 93321 and 93325 ◦ MRI Brain (CPT® 70553) without and with contrast ◦ MRI Cervical (CPT® 72156) and Thoracic Spine (CPT® 72157) without and with contrast
• All individuals, from birth to 10th birthday	• Every 3 months: ◦ US Abdomen (CPT® 76700) and Pelvis (CPT® 76856)
• Known cardiac disease	• See: **Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)** and **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** in the Pediatric Cardiology Guidelines
• Known rhabdomyosarcoma	• See: **Rhabdomyosarcoma (RMS) (PEDONC-8.2)**
• Known neuroblastoma	• See: **Neuroblastoma (PEDONC-6)**

Background and Supporting Information

- Caused by mutations in *HRAS* genes, this syndrome is inherited in an autosomal dominant manner (50% risk to offspring), and is associated with rhabdomyosarcoma and neuroblastoma in early childhood, and transitional cell cancer of the bladder in older children and adults.

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Constitutional Mismatch Repair Deficiency (CMMRD or Turcot Syndrome) (PEDONC-2.15)

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The following imaging studies should be considered medically necessary in individuals with CMMRD/Turcot Syndrome:

Indication	Medically Necessary Imaging Study
• All individuals, after CMMRD diagnosis is confirmed by genetic mutation	• MRI Brain without and with contrast (CPT® 70553) every 6 months
• All individuals, beginning at age 4 years	• Annual esophagogastroduodenoscopy and colonoscopy
• All individuals, beginning at age 6 years	• Annual whole-body MRI (CPT® 76498) ◦ 76498 is the only code for whole-body MRI at this time

Background and Supporting Information

- A highly penetrant and aggressive cancer predisposing syndrome resulting from autosomal recessive inheritance of biallelic mutations in mismatch repair genes, CMMRD syndrome leads to substantial risk for several commonly fatal childhood malignancies - high-grade CNS tumors (glioma, PNET, medulloblastoma) and hematologic malignancies (non-Hodgkin lymphoma, acute lymphoblastic leukemia). Individuals with CMMRD are also at increased risk for gastrointestinal tumors.

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Hereditary Leiomyomatosis and Renal Cell Cancer (HLRCC) (PEDONC-2.16)

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The following imaging studies should be considered medically necessary in individuals with HLRCC:

Indication	Medically Necessary Imaging Study
• Beginning at age 8 years, individuals with at least ONE of the following: ◦ Documented fumarate hydratase (FH) gene mutation/variant consistent with HLRCC ◦ Histologically confirmed multiple cutaneous piloleiomyomas ◦ At least TWO of the following manifestations: ▪ Surgical treatment for symptomatic uterine leiomyomas before age 40 ▪ Type 2 papillary renal cell carcinoma or collecting duct renal carcinoma before age 40 ▪ A first-degree family member with either of the two above criteria	• MRI Abdomen with and without contrast (CPT® 74183), annually

Background and Supporting Information

- Hereditary leiomyomatosis and renal cell cancer (HLRCC) is a disorder in which affected individuals tend to develop benign tumors containing smooth muscle tissue (leiomyomas) in the skin and, in females, the uterus. Approximately 20 percent of people with HLRCC develop renal cell cancer. People with HLRCC are commonly diagnosed with kidney cancer in their forties but cases have been reported in individuals as young as 11.
- In 2019, Forde et. al. published the first large, prospective study of screening for HLRCC and showed that most symptomatic individuals present with stage 3 or 4 RCC with a high risk of death and one life is saved for every 5 individuals on an MRI screening protocol.

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Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)

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Birt-Hogg-Dube Syndrome (BHDS)

The following imaging studies should be considered medically necessary in individuals with BHDS:

Indication	Medically Necessary Imaging Study
• All individuals, beginning at age 20 years	• MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) annually ◦ If no family history of renal tumors AND after 2 consecutive normal MRIs, MRI is medically necessary every 2 years

Background and Supporting Information

- Birt-Hogg-Dube Syndrome (BHDS) in an autosomal dominant disorder with a heterozygous pathogenic variant in FLCN gene.
- These individuals have, or may have:
  - Cutaneous manifestations (fibrofolliculomas, acrochordons, angiofibromas, oral papules, cutaneous collagenomas, and epidermal cysts)
  - Pulmonary cysts/history of pneumothorax
  - A seven-fold increase in renal tumors
    - The most common renal tumors are oncocytoma, chromophobe, and a hybrid of these two.
    - Renal tumors may be multifocal and bilateral.
    - Median age of renal tumor diagnosis is 48 years.
- Recommended cancer screening includes:
  - Screening colonoscopy starting at age 40
    - Earlier colonoscopy may be considered for those with a family history of colorectal cancer earlier than age 40.

BAP1 Tumor Predisposition Syndrome

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The following imaging studies should be considered medically necessary in individuals with BAP1:

Indication	Medically Necessary Imaging Study
• All individuals, beginning at age 30 years	• Abdominal MRI with and without contrast (CPT® 74183) every 2 years

Background and Supporting Information

BAP1 tumor predisposition syndrome (TPDS) is an autosomal dominant condition with a predisposition to melanoma (ocular and cutaneous), mesothelioma, clear cell renal cancer, and chromophobe renal cancer.

Hereditary Papillary Renal Carcinoma (HPRC)

The following imaging studies should be considered medically necessary in individuals with HPRC:

Indication	Medically Necessary Imaging Study
• All individuals, beginning at age 30 years	• Abdominal MRI with and without contrast (CPT® 74183) annually

Background and Supporting Information

- Hereditary papillary renal carcinoma (HPRC) is an autosomal dominant condition involving the MET gene and predisposes individuals to multifocal, bilateral renal tumors.

DICER1

The following imaging studies should be considered medically necessary in individuals with DICER1:

Indication	Medically Necessary Imaging Study
• DICER1 mutation asymptomatic lung surveillance	• Once at age 3-6 months of age and again at 2.5-3 years of age: ◦ CT Chest (contrast as requested) • In addition, chest x-ray at birth and every 6 months until 8 years of age and annually until age 12

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Indication	Medically Necessary Imaging Study
• DICER1 mutation asymptomatic abdominal and pelvic surveillance	• Every 3 months, from birth to the 8th birthday, then every 12 months until 12 years of age: ◦ Abdominal US (CPT® 76700) • After age 12, it is medically necessary for females with DICER1 mutation to continue abdominal and pelvic ultrasounds (CPT® 76700 and CPT® 76856 or CPT® 76857) annually until age 40
• DICER1 mutation thyroid surveillance	• Baseline thyroid US (CPT® 76536) by 8 years of age then every 3 years
• Individuals with DICER1 mutation and new CNS symptoms	• MRI Brain without and with contrast (CPT® 70553)

Other Renal Predisposition Syndromes

The following imaging studies should be considered medically necessary in individuals with other renal predisposition syndromes:

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Indication	Medically Necessary Imaging Study
• Individuals with any of the following genetic variants or syndromes: ◦ REST, TRIM28, FBXW7, NYNRIN, KDM3B, XPO5, CHECK2, PALB2, CTNNB1, DROSHA, WT1 and 2, WTX, DGCR8, SIC1 and 2, BCORL1, MLLT1, MYCN ◦ Bloom Syndrome/BLM mutations ◦ Frasier Syndrome ◦ Trisomy 18 ◦ Perlman Syndrome ◦ Bohring-Opitz Syndrome (ASXL1) ◦ MULIBREY and Nanism Syndrome ◦ Congenital anomalies associated with Wilms Tumor ▪ Horseshoe kidney ▪ Renal ectopia, hypoplasia, or renal/ureteral duplication ▪ Congenital mesoblastic nephroma	• Every 3 months, from birth to the 8th birthday: ◦ Abdominal US (CPT® 76700)

Background and Supporting Information

- The list of syndromes and congenital anomalies associated with Wilms Tumor specifically is ever growing. The following genetic variants and syndromes, not otherwise listed in **PEDONC-2** for more specific imaging, may have imaging as shown above.

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Infantile Myofibromatosis (PEDONC-2.18)

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The following imaging studies should be considered medically necessary in individuals with Infantile Myofibromatosis:

Indication	Medically Necessary Imaging Study
• Assess for the presence of multifocal disease in children under two years of age	• WBMRI (CPT® 76498) once within 6 months of diagnosis
• Pre-operative planning or • Signs or symptoms suggesting progression that may require treatment with surgery or chemotherapy	• Imaging of cutaneous/muscular sites with MRI with and without contrast of symptomatic sites or sites requiring treatment.
• Unresected, known visceral sites of disease	• MRI with and without contrast of unresected visceral involved sites, every 6 months until the age of two years
• Post-operative evaluation and both of the following: ◦ Adequacy of resection is unclear and ◦ Re-excision is being considered	• One-time MRI with and without contrast
• Surveillance for emergence of visceral disease	• Every 6 months, until the age of two years: ◦ CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Abdomen and Pelvis (CPT® 74177) • CT should not duplicate simultaneous MRI imaging of involved sites
• Individuals requiring chemotherapy	• See: **PEDONC-8.3**

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Background and Supporting Information

- Infantile myofibromatosis is a benign condition characterized by soft tissue tumors, 90% of which present in the first 2 years of life. 75% of cases present as solitary lesions affecting the skin and/or muscles of the head, neck and trunk. Other patterns of inheritance include congenital multicentric disease limited to skin and muscle, congenital multicentric with a single visceral site, and congenital with multiple visceral site involvement.
- The condition is most commonly sporadic, though familial cases associated with *PDGFRB* and *NOTCH3* have been described
- The majority of cases with skin or muscle only involvement regress spontaneously.
- Visceral cases are lethal in the absence of therapy in 75% of cases due to organ compression, particularly with cardiopulmonary involvement.
- If vital structures are involved or significant symptoms occur, treatment is generally radical resection. Chemotherapy is sometimes utilized for progressive or multifocal life threatening lesions.
- Infants may present with a single lesion and develop further lesions in the first two years of life. This scenario, along with the inability of infants to readily express symptoms, impacts the imaging studies that may be approved.
- Routine surveillance of cutaneous/muscular sites is not supported as spontaneous regression is common.
- Surveillance beyond the age of two years is not supported.

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Bloom Syndrome (PEDONC-2.19)

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The following imaging studies should be considered medically necessary in individuals with Bloom Syndrome:

Indication	Medically Necessary Imaging Study
Malignancy screening particularly lymphomas	• Every 2 years, starting at age 13: ◦ Whole-Body MRI (CPT® 76498)
Wilms tumor screening	• See: **Other Renal Cell Cancer Predisposition Syndromes (PEDONC-2.17)**
Gastrointestinal tumor screening	• There is no strong data to support endoscopic screening
Breast cancer screening	• There is no strong data to support Breast MRI. See: **Breast MRI Indications (BR-5.1)** for any updates to imaging

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Pediatric Leukemias (PEDONC-3)

Guideline

Pediatric Leukemia General Considerations (PEDONC-3.1)
Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)
Acute Myeloid Leukemia (AML) (PEDONC-3.3)
References (PEDONC-3)

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Pediatric Leukemia General Considerations (PEDONC-3.1)

- MRI Brain without and with contrast (CPT® 70553) is medically necessary in individuals exhibiting CNS symptoms
  - CT Head without or with contrast (CPT® 70450 or CPT® 70460) is medically necessary for urgent concerns where MRI would delay care (i.e., suspected CNS bleeding)
  - Imaging due to CSF tumor burden has not been shown to improve the detection of CNS involvement compared with CSF alone
- See: **Hematopoietic Stem Cell Transplantation (ONC-29)** for imaging guidelines related to transplant

Background and Supporting Information

- The overwhelming majority of leukemias occurring in children are acute. Chronic myelogenous leukemia (CML) is rare in children, and the occurrence of chronic lymphocytic leukemia (CLL) appears to have only been reported once in pediatric individuals to date
- Routine advanced imaging is not indicated in the evaluation and management of chronic myeloid leukemia in the absence of specific localizing clinical symptoms or clearance for hematopoietic stem cell transplantation

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Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)

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- Individuals with B-precursor or T-cell lymphoblastic lymphoma without bone marrow involvement are treated similarly to individuals with leukemia of the same cell type and should be imaged according to this guideline section
- This section does not apply to individuals with mature B-cell histology (primarily Burkitt's in children). Please refer to **Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)** for guidelines for these individuals
- Scrotal Ultrasound CPT® 76870 and/or doppler ultrasound of the scrotum CPT® 93975 or 93976 is medically necessary for suspected testicular involvement.
- Chest x-ray should be performed to evaluate for mediastinal mass in suspected cases or upon initial diagnosis.
  - CT Chest with contrast (CPT® 71260) is medically necessary to immediately evaluate for airway compression and anesthesia safety prior to attempting histologic diagnosis if mediastinal widening is seen on chest x-ray
  - In individuals with known or strongly suspected T-cell histology or other suspected lymphoblastic lymphoma involvement, EITHER of the following is medically necessary for initial staging purposes:
    - CT Neck (CPT® 70491), CT Chest (CPT® 71260), and CT Abdomen and Pelvis (CPT® 74177) with contrast OR
    - PET/CT (CPT® 78816)
    - Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for individuals exhibiting CNS symptoms.
  - Imaging due to CSF tumor burden has not been shown to improve the detection of CNS involvement compared with CSF alone.
    - CT Head without or with contrast (CPT® 70450 or CPT® 70460) is medically necessary for urgent concerns where MRI would delay care (i.e., suspected CNS bleeding)

Additional imaging in lymphoblastic lymphoma/lymphomatous extramedullary disease:

- CT is medically necessary to assess response to therapy **only** for individuals with known bulky nodal disease (usually with T-cell histology) at the end of induction (4

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to 6 weeks). CT is medically necessary for individuals with residual masses every new therapy phase (Consolidation, Interim maintenance, etc., generally every 8 to 12 weeks) until disease resolution is seen

- PET/CT (CPT® 78815) is medically necessary when residual mass ≥8 mm in diameter is present on recent CT imaging **and** immediate radiation or chemotherapy plan will be based on results.
  - Residual mass of any size with no PET-avidity is considered a complete response at the extranodal/lymphomatous site.
- Chest x-ray or Abdominal ultrasound (CPT® 76700) **only**, as indicated by site(s) of bulky disease present at diagnosis, is medically necessary for further surveillance, once CT imaging shows no evidence of disease.
- CT of all involved bulky nodal areas is medically necessary in individuals with persistent residual masses performed as part of an end of therapy evaluation

Immunosuppression during ALL therapy:

- CT or MRI is medically necessary for infectious disease concerns for individuals with ALL with:
  - Absolute neutrophil count (ANC) <500 or
  - Inconclusive findings on chest x-ray **or** ultrasound at any ANC during active treatment
- Individuals with ALL are severely immunocompromised during the first 4 to 6 weeks of treatment (induction) and any conventional imaging request to evaluate for infectious complications during this time frame is medically necessary
- MRA/MRV of the head (CPT® 70544, 70545, or 70546) is medically necessary for the following:
  - To rule out bleeding associated with sinus venous thrombosis in individuals treated with asparaginase

Imaging during therapy for relapsed ALL:

- Frequent CT or MRI imaging is medically necessary to evaluate known or suspected new sites of invasive fungal or other aggressive infections
- Surveillance imaging of asymptomatic individuals to detect invasive fungal infection is medically necessary **only** when acute clinical decisions will be made based on the imaging

Imaging of known or suspected osteonecrosis in ALL:

- MRI without contrast or without and with contrast of the affected joint(s) is medically necessary for symptoms suggesting osteonecrosis
  - CT without contrast is medically necessary when MRI is contraindicated or unavailable, or for diagnosis of suspected subchondral fracture

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- MRI Bilateral Hips (CPT® 73721 or CPT® 73723 with modifier -50) is medically necessary once at 6 to 9 months after diagnosis for individuals age ≥11 years
- Repeat MRI without contrast of the affected joint(s) is medically necessary every 2 cycles of maintenance (every 6 months) if reintroduction of corticosteroids is being considered in individuals whose symptoms have resolved and are still receiving active treatment
- MRI without contrast of the affected joint(s) is medically necessary for preoperative planning for individuals undergoing core decompression
- See: **Osteonecrosis In Long Term Cancer Survivors (PEDONC-19.4)** for information on osteonecrosis in ALL individuals who have completed therapy

Background and Supporting Information

- The majority of individuals with ALL have B-precursor ALL and routine advanced imaging is not necessary.
- Individuals with ALL who relapse are treated with very intensive chemotherapy regimens and most spend the majority of their chemotherapy treatment phase in the hospital
- Individuals may have therapy-induced hypogammaglobulinemia which requires supplemental intravenous immune globulin (IVIG) during maintenance therapy. Those receiving supplemental IVIG should be treated similarly to individuals with ANC <500 with regards to imaging for infectious disease
- Osteonecrosis (ON) in individuals with ALL is a relatively common complication of ALL and its treatment, primary corticosteroids. Approximately 3% of younger children and 12 to 15% of adolescents are affected by ON at some point during therapy. The peak incidence occurs approximately one year from the time of diagnosis
- Screening MRI of asymptomatic individuals age ≤10 years to detect osteonecrosis has not been shown to impact individual outcomes, and it is not standard to alter treatment based on imaging findings alone without symptoms
- If osteonecrosis is detected on initial MRI, corticosteroids are often withheld during maintenance chemotherapy (but continued in earlier phases of therapy).

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Acute Myeloid Leukemia (AML) (PEDONC-3.3)

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- Frequent CT or MRI imaging is medically necessary to evaluate known sites of invasive fungal infection
- Surveillance imaging of asymptomatic individuals is medically necessary to detect invasive fungal infection **only** when acute clinical decisions will be made based on the imaging
- Advanced imaging may be approved on a case-by-case basis for rare individuals with bulky tumor masses (commonly referred to as chloromas, leukemic sarcomas, or myeloid sarcomas) noted on physical examination or other imaging such as plain film or ultrasound

Background and Supporting Information

- The majority of AML individuals do not have any bulky disease and routine advanced imaging is not necessary
- AML individuals are treated with very intensive chemotherapy regimens and spend the majority of their chemotherapy treatment phase in the hospital

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CNS Tumors (PEDONC-4)

Guideline

CNS Tumors General Considerations (PEDONC-4.1)
CNS Low Grade Gliomas (LGG) (PEDONC-4.2)
CNS High Grade Gliomas (HGG) (PEDONC-4.3)
Medulloblastoma (MDB), Other CNS Embryonal Tumors, and Pineoblastoma (PEDONC-4.4)
Atypical Teratoid/Rhabdoid Tumors (ATRT) (PEDONC-4.5)
Pineocytomas and Pineal Parenchymal Tumors (PEDONC-4.6)
CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)
Ependymal Tumors (Ependymoma) (PEDONC-4.8)
Malignant Tumors of the Spinal Cord (PEDONC-4.9)
Craniopharyngioma and Other Tumors of the Sellar Region (PEDONC-4.10)
Primary CNS Lymphoma (PEDONC-4.11)
Meningiomas (PEDONC-4.12)
Choroid Plexus Tumors (PEDONC-4.13)
References (PEDONC-4)

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CNS Tumors General Considerations (PEDONC-4.1)

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- The classification of pediatric central nervous tumors has recently been revised to incorporate molecular biomarkers in addition to histology, immunohistochemical results and ultrastructure characteristics. The changes in nomenclature are incorporated in these guidelines.
- Central nervous system tumors are the second most common form of childhood cancer, accounting for ~20% of all pediatric malignancies

Red Flag Symptoms Raising Suspicion for CNS Tumors Include:
Any headache complaint from a child age ≤5 years
Headaches awakening from sleep
Focal findings on neurologic exam
Clumsiness (common description of gait or coordination problems in young children)
Headaches associated with morning nausea/vomiting
New onset of seizure activity with focal features
Papilledema on physical exam
Loss of developmental milestones (infants and young children)

MRI Considerations

- MRI is the preferred imaging modality for all pediatric CNS tumors
  - MRI Brain without and with contrast (CPT® 70553) is the primary imaging study for pediatric brain tumors
  - MRI Brain without contrast (CPT® 70551) if requested for initial evaluation of suspected CNS tumor for children able to undergo MRI without sedation
  - Initial MRI should be performed without and with contrast in order to avoid a second anesthesia exposure in younger children requiring sedation for MRI

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- MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) can be substituted where MRI Spine without and with contrast is indicated, if being performed immediately following a contrast-enhanced MRI Brain
- Functional MRI (fMRI) (CPT® 70555 or CPT® 70554) is medically necessary to depict spatial relationships between eloquent cortex and neoplasms for preoperative planning and to promote safe resections (following baseline MRI Brain).
- Isotropic volumetric MRI (CPT® 76376, 76377, 0865T, or 0866T) is medically necessary for preoperative planning to accurately localize tumors.
- MRI Orbits without and with contrast (CPT® 70543) is medically necessary in individuals who present with papilledema, altered vision, strabismus, nystagmus, anisocoria, proptosis, ocular cranial nerve palsies, coloboma, or leukocoria.

CT Considerations

- CT for evaluation of ventriculomegaly or other operative considerations, or for children who cannot undergo MRI safely
  - CT for evaluation of headaches related to head trauma or evaluation of skull or facial bone abnormalities
  - Post-contrast CT is generally not indicated
- CT is not a recommended study for evaluation of pediatric headache when brain tumor is clinically suspected because of its limited diagnostic accuracy in this area. MRI should be used as first line imaging in these cases
- CT should not be used in place of MRI to avoid sedation in young children when red flag symptoms for CNS tumors are present

MRA/CTA and Perfusion Studies

- MRA or CTA **only** for preoperative planning or to clarify inconclusive findings on MRI or CT
- CT and MRI Perfusion
  - See: **CT or MRI Perfusion (HD-24.5)** in the Head Imaging guidelines

MR Spectroscopy (MRS, CPT® 76390)

- MRS is **only** medically necessary for use in brain tumors of specified histologies where diagnostic accuracy has been established in peer-reviewed literature
  - See diagnosis-specific guidelines for MRS indications
- MRS is considered not medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section
- MR spectroscopy is not medically necessary for routine surveillance

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PET Brain Imaging (CPT® 78608 and CPT® 78609)

- PET Brain Metabolic imaging (CPT® 78608) is **only** medically necessary for use in brain tumors of specified histologies where diagnostic accuracy has been established in peer-reviewed literature
  - See diagnosis-specific guidelines for PET indications
- PET Brain Metabolic is not medically necessary for routine surveillance
- PET Brain Metabolic imaging is considered not medically necessary for all other histologies and indications not listed in a diagnosis-specific guideline section
- PET Brain Perfusion imaging (CPT® 78609) is not medically necessary in the evaluation or management of primary CNS tumors
- Fusion PET/CT studies (CPT® 78814, CPT® 78815, or CPT® 78816) are not medically necessary in the evaluation or management of primary CNS tumors

Timing and Frequency of Imaging

- Definitive imaging should be completed prior to considering biopsy given the high degree of morbidity associated with operating on the CNS
  - Occasionally biopsy is not necessary because the imaging findings provide a definitive diagnosis
    - Examples include diffuse intrinsic pontine glioma and optic pathway gliomas in an individual with known neurofibromatosis
- Perioperative imaging frequency
  - Children may undergo very frequent imaging in the immediate perioperative period around resection or debulking of a CNS tumor due to the small anatomic spaces involved
    - Requests for imaging during this time period to specifically evaluate postoperative course or ventriculoperitoneal shunt functioning are medically necessary
  - MRI Brain without and with contrast (CPT® 70553) is medically necessary one time in the immediate preoperative period (even if another study has already been completed) to gain additional information which can be important in optimizing individual outcomes, such as:
    - Completion of additional specialized MRI sequences such as diffusion-tensor imaging
    - Perfusion imaging (CPT® 70553, 70551, OR 70552)
    - Tractography
    - Other sequences not reported under a separate CPT® code but not part of a routine MRI Brain series
  - Repeat MRI Brain that is being requested solely for loading into operative navigation software should not be requested as a diagnostic code but is medically necessary under a treatment planning code (CPT® 76498).

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- Clinical note: for all pediatric CNS tumors, cerebrospinal fluid pathology positive for malignancy is considered leptomeningeal/spinal disease.

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CNS Low Grade Gliomas (LGG) (PEDONC-4.2)

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- MR Spectroscopy and PET Brain Metabolic are not medically necessary for routine surveillance.
- MRI is generally superior to CT for staging and restaging CNS malignancies, but CT is medically necessary in accordance with these guidelines where MRI is contraindicated.

Indication	Medically Necessary Imaging Study
Initial staging of all LGG	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • MRI Orbits without and with contrast (CPT® 70543) in addition to the above studies for individuals presenting with visual signs or symptoms as listed in **PEDONC-4.1**
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
• At any time, for ANY of the following: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)

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Indication	Medically Necessary Imaging Study
• ANY of the following: ◦ Determining the need for biopsy when transformation to high grade glioma is suspected based on clinical symptoms or recent MRI findings ◦ Evaluation of a brain lesion of indeterminate nature when the PET findings will be used to determine whether biopsy/resection can be safely postponed	• PET Brain Metabolic imaging (CPT® 78608)
• ANY of the following: ◦ Distinguish low grade from high grade gliomas ◦ Evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed ◦ Distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy	• MR Spectroscopy (MRS, CPT® 76390)
Baseline imaging after resection, to assess degree of resection	• MRI Brain without and with contrast (CPT® 70553) • MRI with and without contrast at level of resected spinal site • If orbital resection, MRI Orbits without and with contrast (CPT® 70543)
Treatment response at the completion of radiotherapy	• MRI Brain without and with contrast (CPT® 70553) • MRI with and without contrast at level of irradiated spinal site • If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543)

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Indication	Medically Necessary Imaging Study
Treatment response on chemotherapy	• MRI Brain without and with contrast (CPT® 70553) approved every 2 cycles during active treatment and at the end of planned chemotherapy • If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543)
Additional treatment response imaging during induction chemotherapy for individuals with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles
Surveillance, including individuals with unresected disease on observation only	• For individuals with intracranial primary: ◦ MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter. ◦ If prior orbital involvement, MRI Orbits without and with contrast (CPT® 70543) ◦ MRI Spine is not medically necessary during surveillance in individuals without prior history of spinal involvement except to evaluate symptoms suspicious for spinal cord recurrence • For individuals with a history of spine primary tumor or metastatic spinal involvement: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter

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Indication	Medically Necessary Imaging Study
Surveillance imaging for individuals with optic pathway glioma and a history of NF1	• Every 3 months for 2 years, then every 6 months for 3 years, then annually thereafter: ◦ MRI Brain without and with contrast (CPT® 70553) and ◦ MRI Orbits without and with contrast (CPT® 70543)
Suspected intracranial or intraspinal recurrence	• All imaging supported in initial staging criteria

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Background and Supporting Information

Includes the following tumors:
• Pediatric-type diffuse low-grade gliomas ◦ Diffuse astrocytoma, MYB- or MYBL1-altered ◦ Angiocentric glioma ◦ Polymorphus low-grade neuroepithelial tumor of the young ◦ Diffuse low-grade glioma, MAPK pathway-altered • Circumscribed astrocytic gliomas ◦ Pilocytic astrocytoma ◦ High-grade astrocystoma with piloid features ◦ Pleomorphic xanthoastrocytoma ◦ Subependymal giant cell astrocystoma (SEGA) ◦ Choroid glioma ◦ Astroblastoma, MN • Glioneuronal and neuronal tumors ◦ Gangliоglioma ◦ Desmoplastic infantile gangliоglioma/desmoplastic infantile astrocystoma ◦ Dysembryoplastic neuroepithelial tumor ◦ Diffuse glioneuronal tumor with oligodendroglioma-like features and nuclear clusters ◦ Papillary glioneuronal tumor ◦ Rosette-forming glioneuronal tumor ◦ Myxoid glioneuronal tumor ◦ Diffuse leptomeningeal glioneuronal tumor ◦ Gangliocytoma ◦ Multinodular and vacuolating neuronal tumor ◦ Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease) ◦ Central neurocytoma ◦ Extraventricular neurocytoma ◦ Cerebellar liponeurocytoma • Any other glial tumor with a WHO grade I or II classification

General Considerations:

- Account for 40 to 60% of pediatric CNS tumors.
- These tumors are defined as having a WHO grade of I or II (out of IV), can occur anywhere in the CNS

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Treatment Considerations:

- Children with neurofibromatosis and small optic pathway tumors may not undergo biopsy or resection and will proceed directly to treatment or surveillance
- Children on observation without specific treatment should be imaged according to surveillance guidelines for LGG
- Individuals who undergo complete resection should be imaged according to surveillance guidelines after post-resection imaging
- Individuals age >10 years with incompletely resected tumors usually receive adjuvant radiation therapy
- Individuals age ≤10 years with incompletely resected tumors are commonly treated with chemotherapy

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CNS High Grade Gliomas (HGG) (PEDONC-4.3)

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Indication	Medically Necessary Imaging Study
Initial staging of all HGG	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
• At any time, for ANY of the following: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)

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Indication	Medically Necessary Imaging Study
• ANY of the following: ◦ Distinguishing radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy ◦ Evaluating inconclusive MRI findings when the PET findings will be used to determine need for biopsy or change in therapy, including a change from active therapy to surveillance ◦ Evaluation of a brain lesion of indeterminate nature when the PET findings will be used to determine whether biopsy/resection can be safely postponed	• PET Brain Metabolic Imaging (CPT® 78608)
• ANY of the following: ◦ To distinguish low grade from high grade gliomas ◦ To evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed ◦ To distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy	• MR Spectroscopy (MRS, CPT® 76390)
• To depict spatial relationships between eloquent cortex and tumor prior to resection	• Functional MRI (fMRI) (CPT® 70544 or CPT® 70555)
Baseline imaging following resection, to assess degree of resection	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast of resected area, or whole spine if requested (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Treatment response at the completion of radiotherapy	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast of irradiated area, or whole spine if requested (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response on chemotherapy	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment and at the end of planned chemotherapy
Additional treatment response imaging during induction chemotherapy for individuals with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Surveillance	• For individuals with intracranial primary: ◦ MRI Brain without and with contrast (CPT® 70553) every 2 months for 1 year, then every 3 months thereafter ◦ MRI Spine is not medically necessary during surveillance in individuals without prior history of spinal involvement except to evaluate symptoms suspicious for spinal cord recurrence • For individuals with a history of spine primary tumor or metastatic spinal involvement: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 months for 1 year, then every 3 months thereafter • For both brain and spine disease, imaging above may be complemented by MR perfusion and/or MR spectroscopy, and these studies are medically necessary if requested.
Suspected intracranial or intraspinal recurrence	• All imaging supported for initial staging
Suspected spinal cord recurrence	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Background and Supporting Information

Includes the following tumors:
• Diffuse midline glioma, H3 K27-altered • Diffuse hemispheric glioma, H3 G34-mutant • Diffuse pediatric-type high-grade glioma, H3-wildtype and IDH-wildtype • Infant-type hemispheric glioma • Any other glial tumor with a WHO grade of III or IV classification

General Considerations:

- Rare in children compared with the adult population, but represent 10% to 20% of pediatric CNS tumors
- Prognosis is very poor, and survival significantly beyond 3 years from diagnosis is rare, even with complete surgical resection at initial diagnosis
- These tumors are defined as having a WHO histologic grade of III or IV (out of IV) can occur anywhere in the CNS (though the majority occur in the brain)

Treatment Considerations:

- Individuals who undergo complete resection should be imaged according to surveillance guidelines after post-resection imaging
- Individuals with incompletely resected tumors are commonly treated with chemotherapy

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Medulloblastoma (MDB), Other CNS Embryonal Tumors, and Pineoblastoma (PEDONC-4.4)

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- MR Spectroscopy and PET Brain Metabolic are not medically necessary for routine surveillance.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals, preoperatively	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0867T, or 0866T) • Functional MRI (fMRI) (CPT®) 70555 or 70554)
• At any time, for ANY of the following: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)

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Indication	Medically Necessary Imaging Study
• ANY of the following: ◦ To distinguish radiation-induced tumor necrosis from progressive disease within 18 months of completing radiotherapy ◦ To evaluate inconclusive MRI findings when the PET findings will be used to determine need for biopsy or change in therapy, including a change from active therapy to surveillance ◦ To evaluate a brain lesion of indeterminate nature when the PET findings will be used to determine whether biopsy/resection can be safely postponed	• PET Brain Metabolic Imaging (CPT® 78608)
• ANY of the following: ◦ To evaluate a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Postoperative (preferably within 48 hours of surgery) to quantify residual tumor volume	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic (CPT® 72157, Lumbar-CPT® 72158) if spinal disease was resected
Within 28 days post-op, if spinal imaging was not performed preoperatively	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Treatment response	• At the start of adjuvant chemotherapy and every 2 cycles until therapy is completed: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic CPT® 72157, Lumbar-CPT® 72158)
Children age <3 years treated with multiple cycles of high dose chemotherapy with autologous stem cell rescue in lieu of radiotherapy	• Disease evaluations (imaging per treatment response guidelines) prior to each cycle (every 4 to 6 weeks) if needed for response determination.
End of treatment evaluation	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine with contrast (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) or MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Surveillance	For low- or average-risk individuals: • Every 3 months for 2 years, then every 6 months for 3 years: ◦ MRI Brain without and with contrast (CPT® 70553) • Every 6 months for 2 years: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) For high- or very high-risk individuals: • Every 3 months for 2 years, then every 6 months for 3 years: ◦ MRI Brain without and with contrast (CPT® 70553) • Every 3 months for 2 years, then every year for 3 years: ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For additional imaging guidelines for individuals in long-term follow-up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)**

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Background and Supporting Information

Includes the Following Tumors:
• Medulloblastoma ◦ Molecularly-defined Medulloblastoma ▪ WNT-activated Medulloblastoma ▪ SHH-activated and TP53-wildtype Medulloblastoma ▪ SHH-activated and TP53-mutant Medulloblastoma ▪ Non-WNT/Non-SHH ◦ Medulloblastoma, histologically defined • Other CNS Embryonal Tumors (previously supratentorial primitive neuro-ectodermal tumors) ◦ CNS embryonal tumor ◦ Cribriform neuroepithelial tumor ◦ Embryonal tumor with multilayered rosettes ◦ CNS neuroblastoma ◦ FOXR2-activated CNS tumor with BCOR internal tandem duplication • Pineoblastoma
Risk Assessment is Important in Determining Optimal Treatment **High-Risk Features Include the Following:**
• Spinal metastasis (including cytology positive only) • Multifocal intracranial tumors • Anaplastic histology • All other CNS embryonal tumors and pineoblastomas • >1.5 cm^2 residual tumor area on postoperative MRI and age <3 years
Individuals without any high-risk features are considered "Average Risk"

General Considerations:

- Account for 15% to 25% of pediatric CNS tumors
- Prognosis is generally favorable
- Leptomeningeal spread is common and can occur after initial diagnosis

Treatment Considerations:

- Individuals generally proceed to chemoradiotherapy within 31 days of surgical resection

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- All individuals receive adjuvant chemotherapy lasting 6 to 12 months that begins ~6 weeks after completion of chemoradiotherapy

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Atypical Teratoid/Rhabdoid Tumors (ATRT) (PEDONC-4.5)

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- PET Brain Metabolic does not have a defined role in the evaluation of ATRT at this time and is, therefore, not medically necessary.
- MR Spectroscopy is not medically necessary for routine surveillance.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals, preoperatively	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • Renal US (CPT® 76770) ◦ If renal US is abnormal, refer to: **Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)**
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
• At any time, for ANY of the following: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)

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Indication	Medically Necessary Imaging Study
Postoperative (preferably within 48 hours of surgery) to quantify residual tumor volume	• MRI Brain without and with contrast (CPT® 70553)
Within 28 days post-op, if spinal imaging was not performed preoperatively	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response to induction chemotherapy	• After every 2 cycles: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Individuals treated with consolidation chemotherapy and autologous stem cell rescue	• Disease evaluation is medically necessary following the end of the planned stem cell rescues but may occur prior to each cycle (every 4 to 6 weeks) if needed for response determination
End of treatment evaluation	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Surveillance	• Every 3 months for 2 years, then every 6 months for 3 years, then annually for 10 years: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)**

Background and Supporting Information:

General Considerations

- Highly aggressive tumor occurring primarily in very young children that has a clinical presentation very similar to medulloblastoma with a much higher rate of leptomeningeal spread.
- Metastases can occur outside the CNS, and associated tumors can also arise in the kidneys (Malignant Rhabdoid Tumor of the Kidney, MRT).
  - Rhabdoid malignancies occurring outside the CNS should be imaged according to **Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)**.
- Overall prognosis is poor, with <20% of individuals surviving beyond 2 years from diagnosis.
- Individuals generally proceed to induction chemotherapy shortly following surgical resection or biopsy.
- Following completion of chemotherapy some individuals will proceed to radiotherapy.
  - MRI performed at the end of consolidation therapy should serve as the diagnostic MRI prior to radiotherapy.

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Pineocytomas and Pineal Parenchymal Tumors (PEDONC-4.6)

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- PET Brain Metabolic imaging and MR Spectroscopy do not have a defined role in the evaluation of pineocytoma.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• MRI Brain without and with contrast (CPT® 70553)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
• Additional initial staging imaging for individuals with: ◦ Multicentric tumors ◦ Atypical histology including pineoblastoma-like elements (grade 2 or 3 pineal parenchymal tumor which have not been considered a pineoblastoma) ◦ Clinical signs or symptoms suggesting spinal cord involvement	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Baseline imaging following resection	• MRI Brain without and with contrast (CPT® 70553)
End of radiotherapy	• MRI Brain without and with contrast (CPT® 70553)
Additional imaging at end of radiotherapy for individuals with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Surveillance	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually thereafter
Additional surveillance imaging for individuals with cord involvement at diagnosis	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) after completion of therapy every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually thereafter
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553)
Suspected spinal cord recurrence or progression	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Background and Supporting Information

General Considerations:

- Low grade malignancy that is similar in presentation to low grade glioma (LGG)
- Surgical resection is curative for most individuals
- Individuals with a complete resection should then be imaged according to surveillance guidelines
- Individuals with incompletely resected tumors may receive adjuvant radiation therapy
- After end of radiotherapy imaging, these individuals should be imaged according to surveillance guidelines

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CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)

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- PET Metabolic Brain imaging does not have a defined role in the evaluation of CNS GCT and is, therefore, considered not medically necessary.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
• At any time, for ANY of the following: ◦ For rapid assessment in the acute setting ◦ Evaluation of acute intracranial hemorrhage ◦ Evaluation of ventriculomegaly ◦ Evaluation of shunt-related issues	• CT Head without contrast (CPT® 70450)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Treatment response to induction chemotherapy	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles

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Indication	Medically Necessary Imaging Study
Additional treatment response to induction chemotherapy for individuals with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles
End of induction chemotherapy for individuals with localized intracranial tumors	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Prior to second-look surgery	• MRI of all known sites of measurable disease
Prior to radiotherapy	• MRI of all known sites of measurable disease
At the end of all planned therapy	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine (with or without and with contrast)
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Surveillance	• Every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually until 5 years after completion of therapy: ◦ MRI Brain without and with contrast (CPT® 70553) ◦ MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)**

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Indication	Medically Necessary Imaging Study
Suspected recurrence - new or worsening neurologic symptoms (including worsening of diabetes insipidus)	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

Background and Supporting Information

Includes the following tumors:
• CNS Germinoma • Non-Germinomatous Germ Cell Tumors (NGGCT) ◦ Embryonal carcinoma ◦ Yolk sac tumor ◦ Choriocarcinoma ◦ Immature Teratoma ◦ Mature Teratoma ◦ Mixed germ cell tumor

General Considerations:

- More common in older school age children and younger adolescents, but can occur throughout the pediatric age range
- Although leptomeningeal spread is common, prognosis is excellent due to high sensitivity to chemotherapy and radiotherapy
- Individuals generally proceed to chemotherapy shortly following surgical resection or biopsy and will usually receive 2 to 4 cycles
- Following completion of chemotherapy, individuals with residual disease will proceed to second-look surgery and/or radiotherapy

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Ependymal Tumors (Ependymoma) (PEDONC-4.8)

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- PET Brain Metabolic imaging does not have a defined role in the evaluation of ependymal tumors and therefore is considered not medically necessary.
- MR Spectroscopy is not medically necessary for routine surveillance

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Baseline imaging following resection	• MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s)
Completion of radiotherapy	• MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s)
Prior to radiotherapy	• MRI of all known sites of measurable disease
Treatment response to induction chemotherapy	• MRI Brain without and with contrast (CPT® 70553) or MRI without and with contrast of involved spinal level(s) every 2 cycles

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Indication	Medically Necessary Imaging Study
End of induction chemotherapy and again at end of all therapy	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Prior to second-look surgery	• MRI of all known sites of measurable disease
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Surveillance, primary intra**cranial** ependymal tumor and NO history of spinal cord involvement	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) can be approved annually for 2 years

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Indication	Medically Necessary Imaging Study
Surveillance, primary intra**cranial** ependymal tumor AND metastatic cord involvement at diagnosis	• MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment
Surveillance, primary intra**spinal** ependymal tumor and NO history of intracranial involvement	• MRI without and with contrast of the involved spinal level(s) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment
Surveillance, primary intra**spinal** ependymal tumor AND metastatic intracranial involvement at diagnosis	• MRI of the involved spinal level(s) without and with contrast every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment • MRI Brain without and with contrast (CPT® 70553) every 3 months for 2 years, then every 4 months for 1 year, then every 6 months in years 4 and 5, then annually to 10 years post treatment

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Background and Supporting Information

Includes the following tumors:

- Ependymal tumors
  - Supratentorial ependymoma
  - ZFTA fusion-positive Supratentorial ependymoma
  - YAP1 fusion-positive Posterior fossa ependymoma
  - Posterior fossa ependymoma, group
  - PFA Posterior fossa ependymoma, group PFB

General Considerations:

- Occur primarily intracranially, roughly 2/3 in the posterior fossa
- Overall prognosis is very good, with supratentorial tumors faring better
- Primary spinal tumors can also occur, and are more common in adult individuals than pediatric individuals.
- Surgery is the primary treatment modality
- Individuals with a complete resection should then be imaged according to surveillance guidelines
- Radiotherapy +/- chemotherapy is used for:
  - Incompletely resected tumors
  - Anaplastic histology
  - Infratentorial location
    - Individuals with incomplete resection or high-risk histology that receive adjuvant radiation therapy should then be imaged according to surveillance guidelines after end of radiotherapy imaging.
- Individuals with gain of chromosome 1q have worse progression-free survival and overall survival outcomes
- RELA-fusion supratentorial ependymoma did not portend worse overall survival outcomes in recent Clinical Oncology Group studies, and is generally not considered an indication for more frequent surveillance imaging

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Malignant Tumors of the Spinal Cord (PEDONC-4.9)

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- If a disease-specific guideline exists, image according to the guidance found in that disease-specific guideline section.
- Common histologies of primary spinal cord tumor in children include:
  - Low Grade Glioma, see: **CNS Low Grade Glioma (LGG) (PEDONC-4.2)** for guidelines
  - High Grade Glioma, see: **CNS High-Grade Glioma (HGG) (PEDONC-4.3)** for guidelines
  - Ependymoma, see: **Ependymal Tumors (Ependymoma) (PEDONC-4.8)** for guidelines
  - NF 1 or 2, see **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** for guidelines
  - Any type of malignant spinal cord tumor can occur, but other histologies are rare.

For rare histologies that do not have a disease-specific guideline section, follow the imaging outlined in the table below.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
Treatment response, every 2 cycles	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • Additionally, MRI Brain without and with contrast (CPT® 70553) for known intracranial disease

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Indication	Medically Necessary Imaging Study
Signs or symptoms of recurrence or progression	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) • Additionally, MRI Brain without and with contrast (CPT® 70553) for known intracranial disease
Surveillance of rare histologies is highly individualized. Surveillance imaging after recent evaluation by a physician with significant training and/or experience in pediatric spinal cord tumors (most commonly a pediatric neurosurgeon or pediatric oncologist) may be indicated in these rare individuals.	• MRI Brain and/or MRI Spine may be considered

Background and Supporting Information

- Treatment principles are the same as tumors of the brain, and should follow imaging guidelines according to the specific histologic type
- Multiple spinal cord tumors should raise suspicion for neurofibromatosis
- Asymptomatic surveillance imaging should generally end at the time point appropriate for the specific tumor type

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Craniopharyngioma and Other Tumors of the Sellar Region (PEDONC-4.10)

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- Individuals of all ages should be imaged according to these guidelines.
- PET Brain Metabolic Imaging and MR Spectroscopy do not have a defined role in the evaluation of craniopharyngioma and are, therefore, considered not medically necessary.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• MRI Brain without and with contrast (CPT® 70553) ◦ Concurrent CT Head without contrast (CPT® 70450) in addition to MRI if craniopharyngioma is suspected
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
• Additional initial staging for individuals with: ◦ Multicentric tumors ◦ Clinical signs or symptoms suggesting spinal cord involvement	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Operative planning or image guidance	• MRA Head (CPT® 70544, 70545, or 70546) OR • CTA Head (CPT® 70496)
Baseline imaging following resection	• MRI Brain without and with contrast (CPT® 70553)
Completion of radiotherapy	• MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Treatment response to chemotherapy	• MRI Brain without and with contrast (CPT® 70553) approved every 2 cycles during active treatment and at the end of planned chemotherapy
Additional treatment response imaging during induction chemotherapy for individuals with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) ◦ Concurrent CT Head without contrast (CPT® 70450) in addition to MRI for craniopharyngioma • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) for signs or symptoms suggesting spinal cord involvement
Surveillance	• MRI Brain without and with contrast (CPT® 70553) can be approved every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually until 10 years after completion of therapy as late progressions can occur ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)**
Suspected spinal cord recurrence	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Background and Supporting Information

Includes the following tumors:
• Craniopharyngioma • Papillary craniopharyngioma • Pituicytoma, granular cell tumor of the sellar region, and spindle cell oncocytoma • PitNET • Pituitary blastoma

General Considerations:

- Imaging guidelines and treatment approaches for pediatric pituitary tumors other than those listed above are consistent with those used for adults with pituitary tumors
  - For these tumors follow guidelines in **Pituitary, Sella, Hypothalamus (HD-19)** in the Head Imaging Guidelines
- Craniopharyngiomas are less common, accounting for 6% to 8% of pediatric CNS tumors.
- Most commonly affects children in the pre-adolescent ages
- Several key imaging findings can be used to differentiate the tumors in this region including the presence of calcifications, cysts, and T1/T2 enhancement patterns in craniopharyngiomas
  - These are best evaluated using a COMBINATION of both MRI and CT modalities. Preoperative prediction is much more successful when BOTH modalities are obtained prior to biopsy.
- Other less common tumors in the optic chiasm, sella, and suprasella region may include Germ Cell Tumors (GCT, see: **PEDONC-4.7**) and Langerhans Cell Histiocytosis (LCH, see: **PEDONC-18**)

Treatment Considerations:

- Surgical resection is curative for many individuals
  - Those with a complete resection should then be imaged according to surveillance guidelines after post-resection imaging is completed
- Individuals with incomplete resection and receiving adjuvant radiation therapy can have a single MRI Brain (CPT® 70553) approved at completion of radiotherapy and should then be imaged according to surveillance guidelines

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Primary CNS Lymphoma (PEDONC-4.11)

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- Primary CNS lymphoma imaging indications in pediatric individuals are identical to those in the general imaging guidelines. See: **CNS Lymphoma (ONC-2.7)** in the Oncology Imaging Guidelines.
- CNS lymphomas also involving bone marrow and/or lymph nodes should be imaged according to: **Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)**.

Background and Supporting Information

- Primary CNS lymphoma is a solitary or multifocal mass occurring in the brain without evidence of systemic (bone marrow or lymph node) involvement.
- Usually associated with immunodeficiency, this is a very rare entity in pediatrics accounting for <0.1% of pediatric malignancies, so age-specific guidelines have not been established.

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Meningiomas (PEDONC-4.12)

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- Meningioma imaging indications in pediatric individuals are identical to those in the general imaging guidelines
  - See: **Meningiomas (Intracranial and Intraspinal) (ONC-2.8)** in the Oncology Imaging Guidelines

Background and Supporting Information

- Account for 1% to 3% of pediatric CNS tumors
- Usually associated with neurofibromatosis type 2 (NF-2) or prior therapeutic radiation exposure to the brain
  - Lifetime risk may be as high as 20% for young children receiving whole brain radiotherapy, most commonly occurring 15 to 20 years after radiation exposure.

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Choroid Plexus Tumors (PEDONC-4.13)

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- PET Metabolic Brain imaging does not have a defined role in the evaluation of choroid plexus tumors and is, therefore, considered not medically necessary.

Choroid Plexus Papilloma:

Indication	Medically Necessary Imaging Study
Suspected/Diagnosis	• MRI Brain without and with contrast (CPT® 70553)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Suspected return of hydrocephalus, or return of hydrocephalus seen on CT imaging	• MRI Brain without and with contrast (CPT® 70553)

Choroid Plexus Adenoma or Atypical Choroid Plexus Papilloma:

Indication	Medically Necessary Imaging Study
Suspected/Diagnosis	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T • Functional MRI (fMRI) (CPT® 70555 or 70554)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Suspected return of hydrocephalus, or return of hydrocephalus seen on CT imaging	• MRI Brain without and with contrast (CPT® 70553)

Choroid Plexus Carcinoma:

Indication	Medically Necessary Imaging Study
Initial staging of all individuals	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
In addition to the above imaging, for preoperative planning	Either or both of the following: • Isotropic volumetric MRI Brain (CPT® 76376, 76377, 0865T, or 0866T) • Functional MRI (fMRI) (CPT® 70555 or 70554)
Evaluation of a brain lesion of indeterminate nature when the MRS findings will be used to determine whether biopsy/resection can be safely postponed	• MR Spectroscopy (CPT® 76390)
Baseline imaging following resection	• MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Prior to radiotherapy	• MRI Brain without and with contrast (CPT® 70553) and MRI with and without contrast of all known sites with measurable disease prior to radiotherapy.
Completion of radiotherapy	• MRI Brain without and with contrast (CPT® 70553)
Treatment response to chemotherapy	• MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment
Additional treatment response to chemotherapy for individuals with measurable spinal cord disease on MRI	• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycles during active treatment
Prior to second-look surgery	• MRI of all known sites of measurable disease
End of all planned therapy	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Signs or symptoms of recurrence or progression	• MRI Brain without and with contrast (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Surveillance, no history of spinal cord involvement	• MRI Brain without and with contrast (CPT® 70553) every 4 months for 3 years, then every 6 months for 2 years after completion of therapy, then annually to 10 years after treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) at 12 and 24 months after completion of therapy
Surveillance, individuals with cord involvement at diagnosis	• MRI Brain without and with contrast (CPT® 70553) every 4 months for 3 years, then every 6 months for 2 years after completion of therapy, then annually to 10 years after treatment ◦ For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, see: **Second Malignant Neoplasms (SMN) (PEDONC-19.3)** • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 4 months for 3 years, then every 6 months for 2 years after completion of therapy, then annually to 10 years after treatment

Background and Supporting Information

- As a group these account for 1% to 4% of pediatric CNS tumors, and 70% of choroid plexus tumors present within the first 2 years of life
- Includes the following tumors:
  - Choroid plexus papilloma
  - Choroid plexus adenoma, or atypical choroid plexus papilloma
  - Choroid plexus carcinoma
- Choroid plexus papillomas
  - Choroid plexus papillomas outnumber other choroid plexus tumors by 4 to 5 times.

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- These ventricular tumors commonly present with hydrocephalus caused by increased CSF production, resulting in signs of increased intracranial pressure.
- Appearance on MRI is typical, and they are usually treated by excision.
- Regrowth is rare

- Choroid Plexus Adenoma or Atypical Choroid Plexus Papilloma
  - These are extremely rare tumors with features midway in the malignant spectrum between papillomas and carcinomas
  - They are more prone to local invasion but rarely to metastasis
  - Presenting symptoms are similar to papillomas
  - Appearance on MRI is typical, and they are usually treated by excision
  - Spinal imaging may be approved if requested at initial diagnosis
  - Regrowth is rare
- Choroid Plexus Carcinoma
  - This is a very aggressive malignancy, with high rates of metastasis to other parts of the CNS
    - Overall incidence of metastases in choroid plexus carcinoma is 12%–50%, which is associated with a worse outcome
  - Prognosis is significantly less favorable than for papillomas with overall survival rates of 35% to 40%
  - *TP53* mutations and alternative lengthening telomeres (ALT) are common in individuals with choroid plexus carcinoma
  - Surgical gross total resection is curative for many individuals
  - Individuals with confirmed gross total resection should then be imaged according to surveillance guidelines
  - Individuals with incomplete resection who receive adjuvant radiation therapy should be imaged according to surveillance guidelines after end of radiotherapy imaging
  - MR Spectroscopy is not indicated for routine surveillance

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Pediatric Lymphomas (PEDONC-5)

Guideline

Pediatric Lymphoma – General Considerations (PEDONC-5.1)
Pediatric Hodgkin Lymphoma (HL) (PEDONC-5.2)
Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)
Anaplastic Large Cell Lymphoma (ALCL) (PEDONC-5.4)
References (PEDONC-5)

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Pediatric Lymphoma – General Considerations (PEDONC-5.1)

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- Lymphoma mostly commonly involves the lymph nodes (LNs). However, lymphoma can also arise from primary lymphoid tissues (bone marrow or thymus) or various secondary lymphoid tissues (spleen, mucosa-associated lymphoid tissue) or non-lymphoid organs (skin, bone, brain, lungs, liver, salivary glands, etc.).
- Pediatric lymphomas are generally Hodgkin Lymphomas, Aggressive B-Cell Non-Hodgkin Lymphomas, Lymphoblastic Lymphomas, or Anaplastic Large Cell Lymphomas
- Individuals with Lymphoblastic Lymphoma (even those with bulky nodal disease) are treated using the leukemia treatment plan appropriate to the cell type (B or T cell).
  - These individuals should be imaged using guidelines in **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)**
- Other histologies are rare in pediatric individuals, and should be imaged according to the following guidelines:
  - Follicular lymphoma: **Follicular Lymphoma (ONC-27.3)** in the Oncology Imaging Guidelines
  - Marginal zone or MALT lymphomas: **Marginal Zone Lymphomas (ONC-27.4)** in the Oncology Imaging Guidelines
  - Mantle cell lymphomas: **Mantle Cell Lymphoma (ONC-27.5)** in the Oncology Imaging Guidelines
  - Cutaneous lymphomas: **Cutaneous Lymphomas and T Cell Lymphomas (ONC-27.8)** in the Oncology Imaging Guidelines
    - **Exception:** Cutaneous B-Lymphoblastic Lymphoma should be imaged using guidelines in **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)**
  - Castleman's Disease: **Castleman's Disease (Unicentric and Multicentric) (ONC-31.11)** in the Oncology Imaging Guidelines
- All CT imaging recommended in this section refers to CT with contrast only.
  - Noncontrast CT imaging has not been shown to be beneficial in the management of pediatric lymphomas
  - Given the limited utility of noncontrast CT imaging in pediatric lymphomas, MRI without **or** without and with contrast is recommended in place of CT for individuals who cannot tolerate CT contrast due to allergy or impaired renal function.
- MRI without and with contrast of symptomatic or previously involved bony areas is medically necessary in individuals with known lymphoma without prior plain x-ray or bone scan evaluation.

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  - Bone scan is inferior to MRI for evaluation of known or suspected bone metastases in lymphoma.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary as it is the preferred study for evaluation of suspected brain metastases in pediatric lymphoma.
  - CT Head with (CPT® 70460) or without and with contrast (CPT® 70470) is medically necessary when MRI is contraindicated.

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Pediatric Hodgkin Lymphoma (HL) (PEDONC-5.2)

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Indication	Medically Necessary Imaging Study
Initial staging	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) ◦ MRI Neck without and with contrast (CPT® 70543) as a substitute for CT to limit radiation exposure if requested • CT Chest with contrast (CPT® 71260) ◦ MRI is not a substitution for CT Chest • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast as substitute for CT to limit radiation exposure if requested • MRI Brain without and with contrast (CPT® 70553) for known CNS involvement or new signs or symptoms suggesting intracranial disease • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) ◦ Can be performed prior to biopsy if necessary for individual scheduling • CT or MRI of other body areas may be indicated for rare individuals based on physical findings or PET/CT results

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Indication	Medically Necessary Imaging Study
Restaging	• ALL of the following, as often as every 2 cycles of chemotherapy: ◦ CT of previously involved visceral areas ▪ In individuals on treatment for recurrent or refractory Hodgkin Lymphoma, all of the following: - CT Neck with contrast (CPT® 70491) - CT Chest with contrast (CPT® 71260) - CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Brain without and with contrast (CPT® 70553) for known CNS involvement AND • After cycles 2 and 4* of chemotherapy and at end of therapy: ◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (only using combination of CPT® 78813 and 76498) ▪ *For individuals with low-risk stage IA or IIA mixed cellularity Hodgkin lymphoma, this restaging PET/CT is medically necessary after cycles 1 and 3 (instead of cycles 2 and 4) if requested. End of therapy PET/CT or PET/MRI is medically necessary for all individuals with Hodgkin lymphoma. ◦ Diagnostic CT or MRI of previously involved areas may be combined with PET at end of therapy. • In addition to the above studies: ◦ If end of therapy PET/CT or PET/MRI done prior to radiation therapy documents Deauville 4 or 5 FDG avidity, one follow-up PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI is medically necessary >12 weeks after radiation therapy to confirm complete response.
Surveillance for individuals with no history of recurrent HL	• Imaging studies are only medically necessary when relapse is clinically suspected, because most individuals will clinically declare themselves and there is no survival advantage in pre-emptive imaging. • **Routine surveillance imaging is not medically necessary in individuals without a prior history of recurrent disease**

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Indication	Medically Necessary Imaging Study
Surveillance, individuals with **recurrent** HL and no evidence of disease following successful treatment	• ALL of the following, every 3 months for 2 years after completing therapy for recurrence: ◦ CT Neck with contrast (CPT® 70491) ◦ CT Chest with contrast (CPT® 71260) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177)
Clarify inconclusive findings on conventional imaging suspicious for recurrence AND considering biopsy to establish recurrence	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)
Clinical symptoms suggesting recurrence	• MRI Brain with and without contrast (CPT® 70553) for known CNS involvement or new signs or symptoms suggesting intracranial disease AND either of the following sets of imaging: • Conventional imaging: ◦ CT Neck with contrast (CPT® 70491) OR MRI Neck without and with (CPT® 70491) AND ◦ CT Chest with contrast (CPT® 71260) AND ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) OR MRI Abdomen and MRI Pelvis without and with contrast (CPT® 74183 and CPT® 72197) AND ◦ CT of other previously involved visceral areas or currently symptomatic areas OR • PET/CT (CPT® 78815 or 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

Background and Supporting Information

- Most individuals experiencing recurrence are detected based on physical findings, and frequent CT surveillance imaging of Hodgkin Lymphoma after completion of therapy does not improve post-recurrence overall survival. The primary determinant of survival at recurrence is time to relapse, regardless of whether relapse is detected

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clinically or via imaging. NCCN pediatric HL guidelines recommend no pre-emptive surveillance imaging.

- Pediatric individuals have a high rate of neck involvement with Hodgkin Lymphoma
- Early treatment response evaluations involve both PET and CT as decisions about chemotherapy drug selection and radiation treatment are frequently made based on both anatomic (CT-based) and metabolic (PET/CT-based) responses.
- Indications and coding for rare circumstances where PET/MRI may be approved are found in **PET imaging in Pediatric Oncology (PEDONC-1.4)**

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Pediatric Aggressive Mature B-Cell Non-Hodgkin Lymphomas (NHL) (PEDONC-5.3)

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Indication	Medically Necessary Imaging Study
Initial staging	ANY or ALL of the following: • CT Neck with contrast (CPT® 70491) ◦ MRI Neck without and with contrast (CPT® 70543) in place of CT Neck, if requested • CT Chest with contrast (CPT® 71260) • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) in place of CT Abdomen and Pelvis, if requested. ◦ Abdominal ultrasound (CPT® 76700 or 76705) at initial presentation if CT/MRI not available. • CT with contrast or MRI without and with contrast any other symptomatic body area • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498 [may be approved in addition to diagnostic CTs])
Additional initial staging if symptoms or extent of disease suggest intracranial extension or metastasis	• MRI Brain without and with contrast (CPT® 70553)
Additional initial staging if symptoms or extent of disease suggest intraspinal extension or metastasis (back pain)	• MRI of suspected spinal level involvement without and with contrast • MRI of whole spine without and with contrast if there is suspected leptomeningeal disease or if sedation will be required

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Indication	Medically Necessary Imaging Study
Restaging for treatment response (following initial response evaluation)	• ANY OR ALL of the following, as often as every cycle of chemotherapy (~every 3 weeks): ◦ CT with contrast or MRI without and with contrast (should be same modality as initial diagnosis if possible) of previously involved areas ◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) until a negative PET is obtained ▪ Whole-body PET/CT (CPT® 78816) may be approved if there is clinical suspicion of, or known, skull or distal lower extremity involvement. ▪ PET/CT may be approved in conjunction with diagnostic CTs/MRIs, PET/CT should not replace imaging with contrast-enhanced diagnostic-quality CT or MRI.
Restaging for all subsequent treatment response, including end of therapy evaluation, after negative PET/CT (either Deauville or Lugano 1, 2 or 3 as reported in formal radiology interpretation)	• CT with contrast or MRI without and with contrast (should be same modality as initial diagnosis if possible) of previously involved areas, as often as every 2 cycles of chemotherapy, and at the end of therapy
End of therapy PET with Deauville 4-5 avidity	ONCE, at least 6-8 weeks after end of therapy PET: • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498) OR • CT Neck, Chest, Abdomen, and Pelvis with contrast (CPT® 70491, CPT® 71260, CPT® 74177)
Assessment of disease activity in inconclusive residual masses seen on conventional imaging	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

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Indication	Medically Necessary Imaging Study
Individuals being treated with Rituximab who present with abdominal pain, due to risk of bowel perforation and obstruction	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ US, x-ray, or other red flags are not required prior to CT
Surveillance of asymptomatic individuals with residual masses in the chest or abdomen and pelvis	• Chest x-ray and Abdominal (CPT® 76700) and Pelvic (CPT® 76856) ultrasound 3 months after completion of therapy. If stable, no further imaging.
Clinical symptoms or laboratory findings suggesting recurrence	• MRI Brain without and with (CPT® 70553) for history of CNS involvement or new signs or symptoms suggesting intracranial disease And ANY OR ALL of the following: • CT Neck with contrast (CPT® 70491) **and** CT Chest with contrast (CPT® 71260) **and** CT Abdomen and Pelvis with contrast (CPT® 74177) **and** CT with contrast of other previously involved visceral areas or currently symptomatic areas (MRI with and without contrast in place of CT for all areas other than the chest if requested) **and** • PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)
Suspected PTLD recurrence with documentation of new palpable nodes, rising LDH, or rising quantitative EBV PCR	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

Background and Supporting Information

- Aggressive mature B-Cell NHL includes all of the following diagnoses, all of which should be imaged according to this section:
  - Burkitt's lymphoma/leukemia (BL)
  - Diffuse Large B-Cell Lymphoma (DLBCL)
  - Primary Mediastinal Large B-Cell Lymphoma (PMBCL)

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  - Post-transplant Lymphoproliferative Disorder (PTLD)
    - Most commonly occurs following solid organ or stem cell transplantation
  - Viral-associated lymphoproliferative disorders
    - Most commonly occurs following hematopoietic stem cell transplantation or in individuals with primary immunodeficiency
- Initial treatment is usually 7 days of low intensity therapy, with early response evaluation determining next steps in therapy using CT with contrast or MRI without and with contrast of previously involved areas performed around day 6
  - Individuals are customarily still inpatient for this evaluation so outpatient requests should be rare for this time point
- Routine asymptomatic surveillance with advanced imaging has not been found to impact individual outcomes as the majority of these individuals present clinically at relapse due to the highly aggressive nature of these lymphomas
- Indications and coding for rare circumstances where PET/MRI may be approved are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Anaplastic Large Cell Lymphoma (ALCL) (PEDONC-5.4)

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Indication	Medically Necessary Imaging Study
Initial staging	• ANY OR ALL of the following: ◦ CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI without and with contrast of affected area ▪ Substituted for CT in cases of paraspinal or soft tissue extremity primary tumors ◦ CT with contrast or MRI without and with contrast any other symptomatic body area ◦ PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and 76498)
Additional initial staging for individuals with bony primary tumors or metastatic disease	• Bone scan (See: **Modality General Considerations (PEDONC-1.3)**)
Restaging at the end of induction chemotherapy (commonly 4 to 6 weeks)	• CT with contrast or MRI without and with contrast of previously involved areas (should be same modality as initial diagnosis if possible)

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Indication	Medically Necessary Imaging Study
Treatment response in individuals treated with cytotoxic chemotherapy	• Every 2 cycles: ◦ CT of previously involved areas ▪ If CT is performed for primary treatment response, PET/CT (CPT® 78815 or CPT® 78816) to clarify inconclusive findings detected on conventional imaging OR ◦ PET/CT or PET/MRI (CPT® 78813 and CPT® 76498) until a negative PET is obtained ▪ If PET/CT (CPT® 78815 or CPT® 78816) is performed for primary treatment response, CT or MRI to clarify inconclusive findings detected on PET imaging
Restaging after negative PET/CT (either Deauville or Lugano 1, 2 or 3 as reported in formal radiology interpretation)	• CT with contrast of previously involved areas
Surveillance	• CT with contrast or MRI without and with contrast of all previously involved areas every 6 months for 2 years after completion of therapy.
Additional surveillance for individuals with bony primary tumors or metastatic disease	• Bone scan (See: **Modality General Considerations (PEDONC-1.3)** for coding) at 3, 6, 12, and 18 months after therapy is completed
Clinical symptoms suggesting recurrence	• CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), CT Abdomen and Pelvis with contrast (CPT® 74177), and CT with contrast of other previously involved or currently symptomatic areas
Clarify inconclusive findings on conventional imaging to evaluate the need for biopsy to establish recurrence	• PET/CT (CPT® 78815 or CPT® 78816) or PET/MRI (CPT® 78813 and CPT® 76498)

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Background and Supporting Information

- Similar in presentation to Hodgkin Lymphoma, and may be indistinguishable until immunocytology and molecular studies are complete.
- Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Neuroblastoma (PEDONC-6)

Guideline

Neuroblastoma – General Considerations (PEDONC-6.1)
Staging and Risk Grouping – Neuroblastoma (PEDONC-6.2)
Neuroblastoma – Initial Staging (PEDONC-6.3)
Neuroblastoma – Treatment Response Imaging (Risk Group Dependent) (PEDONC-6.4)
Neuroblastoma – Surveillance Imaging (Risk Group Dependent) (PEDONC-6.5)
References (PEDONC-6)

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Neuroblastoma – General Considerations (PEDONC-6.1)

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- PEDONC-6 should be used to review neuroblastoma, ganglioneuroblastoma, and ganglioglioma in individuals of all ages, with the exception of esthesioneuroblastoma in individuals of all ages, which should be reviewed using **Squamous Cell Carcinomas of the Head and Neck (ONC-3)** in the Oncology Imaging Guidelines.
- Neuroblastoma is divided into very low, low, intermediate, and high-risk disease based on International Neuroblastoma Risk Group (INRG) Staging System (see: **Staging and Risk Grouping (PEDONC-6.2)**). The treatment approaches for each risk group vary widely and have distinct imaging strategies. The risk group for a given individual should be provided by the ordering provider in the clinical information provided for review.
- For metabolic imaging in individuals who are MIBG positive at diagnosis and then become MIBG negative in response to treatment:
  - Continue to use MIBG (see: table below and **PEDONC-1.3** for coding)

Study Type	Coding
Bone scan	▪ Any of the following codes can be approved: - CPT® 78300 - CPT® 78305 - CPT® 78306 - CPT® 78803, 78830, or 78832 • May be approved alone or in combination with: ◦ CPT® 78305 ◦ CPT® 78306

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Study Type	Coding
^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy	▪ Any one of the following codes can be approved: - CPT® 78801 - CPT® 78802 - CPT® 78804 ▪ Any one of the following codes may also be approved, individual or in combination with CPT® 78801, 78802, or 78804 - CPT® 78803 - CPT® 78830 - CPT® 78831 - CPT® 78832 - CPT® 78800 may be approved for KNOWN neuroblastoma when only a single site follow up is desired but is not sufficient for the initial workup of suspected disease.
Octreotide scan	▪ Same coding as MIBG
Gallium scan	▪ Same coding as MIBG

- MIBG remains the standard of care metabolic imaging in neuroblastoma, 18F-FDG-PET-CT is not medically necessary unless one of the exceptions below is present. **All PET imaging in PEDONC-6 refers to 18F-FDG radiotracer.** All other radiotracers are considered not medically necessary at this time:

Indication	Medically Necessary Imaging Study
◦ Any of the following: ▪ Individuals with MIBG-negativity documented at initial diagnosis ▪ Individuals with discordant findings on MIBG and conventional imaging (i.e., it is suspected there is more active disease than is visible on MIBG) ▪ At major decision points (such as hematopoietic stem cell transplant or surgery), if MIBG and CT/MRI findings are inconclusive	◦ Whole-body 18F-FDG PET/CT (CPT® 78816) ◦ In scenarios where PET/CT is supported, it does not preclude the other diagnostic imaging studies supported throughout PEDONC-6. PET/CT is viewed as replacing MIBG in these scenarios. Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Indication	Medically Necessary Imaging Study
◦ Individuals currently receiving medications that may interfere with MIBG uptake that cannot be safely discontinued prior to imaging, including: ▪ Tricyclic antidepressants (amitriptyline, imipramine, etc.) ▪ Selective serotonin reuptake inhibitors (SSRI's, sertraline, paroxetine, escitolapram, etc.) ▪ Neuroleptics (risperidone, haloperidol, etc.) ▪ Antihypertensive drugs (alpha or beta blockers, calcium channel blockers) ▪ Decongestants (phenylephrine, ephedrine, pseudoephedrine) ▪ Stimulants (methylphenidate, dextroamphetamine, etc.)	◦ 18F-FDG PET/CT (CPT® 78816) is only medically necessary when specific documentation of the medication interaction is included with the current PET imaging request. ◦ Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Background and Supporting Information

- Neuroblastoma is the most common extracranial solid tumor of childhood, and generally arises from the adrenal gland or along the sympathetic chain. Neuroblastoma staging has recently changed to better incorporate the prognostic impact of biological and genetic characteristics, particularly segmental chromosome aberrations (SCA) as an additional genomic marker. SCAs of 1p or 11g are poor prognostic indicators.
- 90% to 95% of neuroblastomas secrete homovanillic acid (HVA) and vannilylmandelic acid (VMA) in the urine, and urine HVA/VMA should be performed at every disease evaluation for individuals with positive HVA or VMA at diagnosis
- 99mTc-MDP bone scan does not identify foci of disease that affect staging or clinical management and provides no advantage over MIBG scintigraphy and is not used for evaluation of most individuals with neuroblastoma

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Staging and Risk Grouping – Neuroblastoma (PEDONC-6.2)

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- Most recent treatment protocols are using the updated International Neuroblastoma Risk Group (INRG) staging system
  - L1: Localized tumor not involving vital structures as defined by the list of image-defined risk factors and confined to one body compartment
    - Image-defined risk factors include a list of specific imaging findings defining individuals less likely to be candidates for complete surgical resection
    - These risk factors involve the encasement of major blood vessels, airway, skull base, costovertebral junction, brachial plexus, spinal canal, or major organs or structures
  - L2: Locoregional tumor with presence of one or more image-defined risk factors
  - M: Distant metastatic disease (except stage MS)
  - MS: Metastatic disease in children younger than 18 months with metastases confined to skin, liver, and/or bone marrow with <10% involvement (MIBG must be negative in bone and bone marrow)
- The risk group for any given individual should be provided or documented by the requesting provider.

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Neuroblastoma – Initial Staging (PEDONC-6.3)

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The following imaging studies are medically necessary in the initial staging of individuals with neuroblastoma, ganglioneuroblastoma, or ganglioneuroma:

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• ^{123}I-metaiodobenzylguanidine (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) scintigraphy AND • ONE of the following sets of imaging: ◦ CT Neck with contrast (CPT® 70491), CT Chest with contrast (CPT® 71260), and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Neck without and with contrast (CPT® 70543), MRI Chest without and with contrast (CPT® 71552), and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
• All individuals with paraspinal tumors • Individuals with back pain or cord compression symptoms	In addition to the above imaging: • MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar (CPT® 72158) spine without and with contrast
Evaluation of suspected adrenal neuroblastoma, ganglioneuroblastoma, or ganglioneuroma when CT or MRI is inconclusive for an adrenal lesion.	• Adrenal nuclear imaging (CPT® 78075)

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Indication	Medically Necessary Imaging Study
Clinical signs/symptoms suggest brain and/or orbital involvement	Any or all of the following: • MRI Brain without and with contrast (CPT® 70553) • CT Head with contrast (CPT® 70470) • CT Orbits/Maxillofacial with contrast (CPT® 70487)

- PET should not be used unless one of the exceptions stated in section **Neuroblastoma – General Considerations (PEDONC-6.1)** is present.

Background and Supporting Information

- MIBG provides superior sensitivity and sensitivity for detecting viable osseous disease compared with bone scintigraphy so technetium bone scan is not necessary when MIBG is utilized. MIBG is positive in 90% to 95% of neuroblastomas.
  - Most MIBG imaging studies are SPECT/CT studies using CT for localization only. Separate diagnostic CT codes should not be approved for this purpose
  - Occasionally MIBG cannot be performed prior to initiation of therapy. In this circumstance, MIBG should be completed within 3 weeks of therapy initiation as the reduction in MIBG avidity in response to chemotherapy is not immediate. Inability to complete MIBG before starting therapy is not an indication to approve PET imaging

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Neuroblastoma – Treatment Response Imaging (Risk Group Dependent) (PEDONC-6.4)

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- Risk Grouping will not be known at the time of initial staging, but is critical for all imaging decisions after initial staging is complete. **The treating oncologist should always know the individual's risk grouping**. It is not possible to establish the appropriate imaging plan for a neuroblastoma individual without knowing his/her risk group.

All Very Low Risk and Low Risk Neuroblastoma Not Receiving Chemotherapy:

Indication	Medically Necessary Imaging Study
All individuals, no sooner than 4 weeks after diagnosis to determine if additional treatment is necessary	• CT with contrast or MRI without and with contrast of the primary tumor site. If primary tumor site is abdomen or pelvis, imaging of both sites is medically necessary. ◦ Ultrasound is medically necessary if requested in place of CT or MRI to avoid radiation and/or anesthesia exposure in low risk individuals

Background and Supporting Information

- Many individuals will be treated with surgical resection only without adjuvant therapy, and these individuals enter immediately into surveillance.

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All Intermediate Risk Neuroblastoma and Very Low Risk or Low Risk Neuroblastoma Receiving Chemotherapy:

Indication	Medically Necessary Imaging Study
Prior to surgical resection	• Restaging imaging (MIBG and CT or MRI, as performed at initial diagnosis) • PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met. • Additional imaging for individuals with paraspinal disease: ◦ MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158)
Treatment response, as often as every 2 cycles of chemotherapy (~every 6 weeks and at the end of planned treatment)	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Neck (CPT® 70543) without and with contrast, MRI Chest without and with contrast (CPT® 71552), and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) AND • CT or MRI of other sites with prior measurable disease • Additional imaging for individuals with paraspinal disease: ◦ MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158)
In addition to treatment response listed above, every 4 cycles, and at the end of planned chemotherapy treatment	• MIBG scan (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details) • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met

Background and Supporting Information

- Individuals generally receive 2 to 12 cycles of moderate-intensity chemotherapy depending on response to treatment.

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- Surgical resection may occur prior to or following chemotherapy depending on disease stage. Restaging prior to surgery is appropriate.

High-Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
• ANY of the following: ◦ Treatment response ▪ As often as every 2 cycles of chemotherapy, mAb, or biologic therapy (~every 6 weeks) ◦ Change in modality ▪ Prior to surgery, HSCT, XRT, or mAb therapy ◦ End of therapy	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) OR • MRI Neck (CPT® 70543) without and with contrast, MRI Chest without and with contrast (CPT® 71552), and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) AND • CT or MRI of other sites with prior measurable disease • Additional imaging for individuals with paraspinal disease: ◦ MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) • MIBG scan (see table in **Neuroblastoma - General consideration (PEDONC-6.1)** for MIBG coding details)
At completion of ^{131}I-MIBG therapy	• ^{123}I-MIBG scan ◦ FDG-PET cannot be used after ^{131}I-MIBG therapy
Preoperative planning	• More frequent imaging with any of the above modalities can be approved around the time of surgery if needed

Background and Supporting Information

- This group of individuals receives highly aggressive therapy using sequential chemotherapy, surgery, high dose chemotherapy with stem cell rescue, radiotherapy, monoclonal antibody (mAb) immunotherapy, and biologic therapy.

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Neuroblastoma – Surveillance Imaging (Risk Group Dependent) (PEDONC-6.5)

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Very Low Risk and Low Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
All individuals	• At 3, 6, 9, 12, 18, 24, 30 and 36 months after surgery: ◦ Ultrasound of involved areas OR • 3, 6, 9, 12, 18, 24, 30 and 36 months after surgery or to clarify unclear findings on ultrasound: ◦ CT with contrast or MRI without and with contrast of the primary tumor site
Clarification of findings on CT or MRI suspicious for disease recurrence	• MIBG (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details) • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.

Intermediate Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
All individuals	• Every 3 months for 1 year, then every 6 months for 1 year, and then at 36 months after surgery: ◦ CT with contrast or MRI without and with contrast of the primary tumor ▪ If the primary tumor is paraspinal: - MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) ◦ If primary site is abdomen or pelvis, both body sites are medically necessary (CPT® 74177 or the combination of CPT® 71297 and CPT® 74183) ◦ If negative at 36 months, ultrasound at 48 and 60 months after surgery to complete 5 years surveillance.

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Indication	Medically Necessary Imaging Study
• BOTH of the following: ◦ Individuals with stage 4, or M disease, or 4S, or MS disease AND ◦ Positive MIBG at completion of therapy	• Until a negative scan is achieved, every 3 months in year 1, then once in year 2 and year 3: ◦ MIBG scan (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) ◦ Once a negative MIBG is achieved, no further MIBG imaging is necessary. • MIBG is not medically necessary for all other intermediate risk individuals. • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.

High Risk Neuroblastoma:

Indication	Medically Necessary Imaging Study
All individuals	• Every 3 months for 1 year, then every 6 months for 1 year, and then annually to complete 6 years surveillance: ◦ CT with contrast or MRI without and with contrast of the primary tumor site AND ◦ MIBG scan (see table in **Neuroblastoma – General Considerations (PEDONC-6.1)** for MIBG coding details) ▪ 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met ◦ If primary site is abdomen or pelvis, both body sites are medically necessary (CPT® 74177 or the combination of CPT® 71297 and CPT® 74183) ◦ For history of paraspinal disease, MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) ◦ If negative at 6 years, no further advanced imaging.

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Indication	Medically Necessary Imaging Study
Suspected recurrence	• CT Chest/Abdomen/Pelvis with contrast (CPT® 71260, and CPT® 74177) or MRI without and with contrast, (CPT® 71552, CPT® 74183, and CPT® 72197) and other sites of prior measurable disease or current symptoms • MRI Brain with and without contrast (CPT® 70553) for signs or symptoms of brain involvement • MRI without and with contrast of the whole spine (CPT® 72156, CPT® 72157, and CPT® 72158) for history of paraspinal disease, back pain, or cord compression symptoms. • MIBG scan (see table in **Neuroblastoma - General Considerations (PEDONC-6.1)** for MIBG coding details) • 18F-FDG PET/CT or PET/MRI if exceptions noted in **PEDONC-6.1** are met.

Background and Supporting Information

- Very Low Risk and Low Risk Neuroblastoma:
  - CT Chest is not indicated in asymptomatic surveillance imaging of any stages of neuroblastoma individuals with no prior history of thoracic disease
- High-Risk Neuroblastoma:
  - Early detection of recurrence with ^{123}I-MIBG has been shown to improve post-relapse outcomes in high-risk neuroblastoma

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Pediatric Renal Tumors (PEDONC-7)

Guideline

Pediatric Renal Tumors – General Considerations (PEDONC-7.1)
Unilateral Wilms Tumor (UWT) (PEDONC-7.2)
Bilateral Wilms Tumor (BWT) (PEDONC-7.3)
Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)
Clear Cell Sarcoma of the Kidney (CCSK) (PEDONC-7.5)
Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)
Congenital Mesoblastic Nephroma (CMN) (PEDONC-7.7)
References (PEDONC-7)

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Pediatric Renal Tumors – General Considerations (PEDONC-7.1)

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- Ultrasound is considered first-line imaging for suspected pediatric renal mass, given the lack of radiation exposure and need for sedation. Advanced imaging for suspected Wilms Tumor should be done after initial assessment with ultrasound, which has often been completed prior to diagnosis.
- A variety of tumors can occur in the pediatric kidney, and include the following:
  - Wilms Tumor
    - Favorable Histology (FHWT)
    - Focal Anaplasia (FAWT)
    - Diffuse Anaplasia (DAWT)
    - Bilateral Wilms Tumor (BWT)
  - Renal Cell Carcinoma (RCC)
  - Clear Cell Sarcoma of the Kidney (CCSK)
  - Malignant Rhabdoid Tumor of the Kidney (MRT)
  - Congenital Mesoblastic Nephroma (CMN)
  - Other cancers occurring in the kidney:
    - Neuroblastoma
    - Primitive Neuroectodermal Tumor
    - Rhabdomyosarcoma
    - Non-Rhabdomyosarcoma Soft Tissue Sarcomas
    - These and other rare tumors have been reported occurring primarily in the kidney and should be imaged according to the guidelines for the specific histologic diagnosis.
- For suspected renal tumor, cell type unknown, image according to **Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)**
- PET is not routinely medically necessary for initial staging, treatment response, or surveillance of any pediatric renal tumor - rare circumstances for exceptions are listed in the relevant guideline sections.

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Unilateral Wilms Tumor (UWT) (PEDONC-7.2)

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Indication	Medically Necessary Imaging Study
Initial Staging	ONE of the following: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) OR • CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ Doppler ultrasound to evaluate for tumor thrombus is not medically necessary unless CT findings are inconclusive AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • PET is not routinely medically necessary in the initial staging of any pediatric renal tumor
Bilateral renal lesions noted on ultrasound or CT	• MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast
Additional initial staging imaging for any individual with neurologic signs or symptoms raising suspicion of CNS metastases	• MRI Brain without and with contrast (CPT® 70553)
Additional initial staging imaging for any individual with signs or symptoms raising suspicion of bony metastases	• Bone scan (see: **PEDONC-1.3** for coding)

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Indication	Medically Necessary Imaging Study
Treatment response ~every 2 cycles during treatment and at the end of planned therapy	• CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • CT Abdomen with contrast (CPT® 74160) or CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity	• PET/CT (CPT® 78815)
Surveillance	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250), or chest x-ray AND CT Abdomen with contrast (CPT® 74160) or Abdominal US (CPT® 76700) ◦ Every 3 months for 2 years, then every 6 months for 2 additional years, to complete 4 years surveillance • In addition, pelvic imaging (CT or ultrasound) for individuals with stage II or higher disease, for individuals treated with nephrectomy only, or for individuals with a history of tumor rupture, known pelvic involvement, diffuse anaplastic histology, or a history of disease recurrence.
• There are no data to support the use of PET imaging for routine surveillance in any individual with Wilms tumor.	

Background and Supporting Information

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.
- Only ~0.5% of individuals with Wilms tumor will ever develop brain metastases
- A very low risk subset of stage I FHWT will be observed after nephrectomy, and enter directly into surveillance.
- The majority of individuals will receive chemotherapy with or without XRT, beginning within 14 days of initial surgery.

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Bilateral Wilms Tumor (BWT) (PEDONC-7.3)

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Indication	Medically Necessary Imaging Study
Initial Staging	ONE of the following: • MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) OR • CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) is often performed prior to discovery of bilateral lesions and should not prevent MRI from being approved if requested ▪ Doppler ultrasound to evaluate for tumor thrombus is not medically necessary unless CT findings are inconclusive AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • PET is not routinely medically necessary in the initial staging of any pediatric renal tumor
Additional initial staging imaging for any individual with neurologic signs or symptoms raising suspicion of CNS metastases	• MRI Brain without and with contrast (CPT® 70553)

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Indication	Medically Necessary Imaging Study
Additional initial staging imaging for any individual with signs or symptoms raising suspicion of bony metastases	• Bone scan (see: **PEDONC-1.3** for coding)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for individuals with a contraindication to MRI • CT Chest with (CPT® 71260) or without contrast (CPT® 71250)
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity	• PET/CT (CPT® 78815)
Surveillance	• Every 3 months for 2 years, then every 6 months for 2 additional years, to complete 4 years surveillance: ◦ CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) or chest x-ray AND ◦ CT Abdomen with contrast (CPT® 74160) or Abdominal US (CPT® 76700) • Pelvic imaging (CT or ultrasound) for individuals with stage II or higher disease, individuals treated with nephrectomy only, or individuals with a history of tumor rupture, diffuse anaplastic histology, known pelvic involvement, or a history of disease recurrence.
• PET is not routinely medically necessary to assess treatment response or for surveillance in Wilms tumor.	

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Background and Supporting Information

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.
- Individuals with bilateral Wilms Tumor may begin therapy without a histologic diagnosis to preserve a localized disease stage and attempt to shrink the tumors to allow for renal-sparing surgical approaches.
- Only ~0.5% of Wilms tumor individuals will ever develop brain metastases
- If treating with chemotherapy without a biopsy, disease evaluation is indicated at week 6. If either tumor has not shrunk 50%, then open biopsy is indicated to confirm favorable histology.
- If partial nephrectomy still not feasible at week 6, the next disease evaluation is at week 12. Surgical resection should occur no later than week 12.

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Pediatric Renal Cell Carcinoma (RCC) (PEDONC-7.4)

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Indication	Medically Necessary Imaging Study
Initial Staging	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ If bilateral renal lesions are noted on ultrasound or CT, MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) should be strongly considered • CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • PET scan is not routinely medically necessary in the initial staging of any pediatric renal tumor
Additional initial staging for any individual with neurologic signs or symptoms raising suspicion of CNS metastases	• MRI Brain without and with contrast (CPT® 70553)
Additional initial staging for any individual with signs or symptoms raising suspicion of bony metastases	• Bone scan (See: **PEDONC-1.3** for coding)
Treatment response in individuals with residual measurable disease after initial surgery and receiving adjuvant medical therapy	• Every 2 cycles during active treatment: ◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) and ◦ CT Abdomen with contrast (CPT® 74160) ▪ Pelvic imaging is not medically necessary unless prior pelvic involvement has been documented

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Indication	Medically Necessary Imaging Study
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity	• PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
Surveillance in individuals with documented CNS metastases	• Every 6 months for 2 years after completion of all therapy: ◦ MRI Brain without and with contrast (CPT® 70553)
Surveillance in individuals with TFE3 or TFEB subtype	• Every 3 months for 2 years, then every 6 months for 2 years after completion of all therapy: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) ▪ Pelvic imaging is not medically necessary for surveillance unless prior pelvic involvement has been documented
Surveillance in all other histologies	• See: **Renal Cell Cancer (RCC) – Surveillance (ONC-17.4)**
New signs/symptoms suggestive of CNS recurrence	• MRI Brain without and with contrast (CPT® 70553)
• PET is not routinely utilized to assess treatment response in pediatric RCC.	

Background and Supporting Information

- A majority of pediatric cases have a novel subtype involving TFE3 or TFEB translocations, which have a different natural history than “adult type” RCC
- Individuals of any age with TFE3 or TFEB translocated RCC should be imaged according to this guideline section.
- 40% to 45% of pediatric RCC cases have similar histologies to adult RCC (clear cell, papillary, chromophobe, etc.) and imaging decisions will be similar to general oncology guidelines.
- Individuals with all other subtypes of RCC should be imaged according to **Renal Cell Cancer (RCC) (ONC-17)** in the Oncology Imaging Guidelines

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- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation. Doppler ultrasound to evaluate for tumor thrombus is no longer necessary unless CT findings are inconclusive, and should not be performed if CT is already completed.
- Other staging imaging than what is stated in the above table should be deferred until a histologic diagnosis is made, by complete nephrectomy for most unilateral renal tumors and biopsy for bilateral renal tumors or inoperable unilateral tumors
- Most individuals will have surgical resection of all disease at the time of diagnosis and will enter directly into surveillance

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Clear Cell Sarcoma of the Kidney (CCSK) (PEDONC-7.5)

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Be careful not to confuse the diagnosis with clear cell RCC. See: Renal Cell Cancer (RCC) (ONC-17) for imaging guidelines.

Indication	Medically Necessary Imaging Study
Initial Staging	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ Doppler ultrasound to evaluate for tumor thrombus is not medically necessary unless CT findings are inconclusive • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • Bone scan (see: **PEDONC-1.3** for coding) • MRI Brain without and with contrast (CPT® 70553) • PET is not routinely considered medically necessary in the initial staging of any pediatric renal tumor
Bilateral renal lesions are noted on ultrasound or CT in initial staging	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy	• CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) • Additionally, for individuals with CNS metastases at initial staging: ◦ MRI Brain without and with contrast (CPT® 70553) Bone scan (see: **Modality General Considerations (PEDONC-1.3)** for coding) at the end of planned therapy

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Indication	Medically Necessary Imaging Study
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity	• PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
Surveillance	• Every 3 months for 2 years after completion of all therapy: ◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) • Every 6 months for 3 years after completion of all therapy: ◦ MRI Brain without and with contrast (CPT® 70553) • Every 3 months for 1 year, then every 6 months for 2 years after completion of all therapy: ◦ Bone scan (see: **Modality General Considerations (PEDONC-1.3)** for coding) ◦ If negative at 36 months, no further advanced imaging. • Other surveillance imaging should be by Abdominal US (CPT® 76700) and chest x-ray

Background and Supporting Information

- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.
- Other staging imaging should be deferred until a histologic diagnosis is made, by complete nephrectomy for most unilateral renal tumors and biopsy for bilateral renal tumors or inoperable unilateral tumors

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Malignant Rhabdoid Tumor of the Kidney (MRT) and Other Extracranial Sites (PEDONC-7.6)

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Be careful not to confuse the diagnosis with rhabdomyosarcoma. See: **Rhabdomyosarcoma (RMS) (PEDONC-8.2)** for Imaging Guidelines

Indication	Medically Necessary Imaging Study
Initial Staging	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ Doppler ultrasound to evaluate for tumor thrombus is not medically necessary unless CT findings are inconclusive • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia exposure if possible • Bone scan (see: **PEDONC-1.3** for coding) • MRI Brain without and with contrast (CPT® 70553) • PET is not routinely medically necessary in the initial staging of any pediatric renal tumor
Bilateral renal lesions are noted on ultrasound or CT in initial staging	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)

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Indication	Medically Necessary Imaging Study
Treatment response ~every 2 cycles during treatment and at the end of planned therapy	• CT Chest with (CPT® 71260) *or* CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ◦ If primary site other than kidney, perform CT with contrast or MRI without and with contrast of primary site in place of abdominal and pelvic imaging • MRI Brain without and with contrast (CPT® 70553): ◦ Every 2 cycles during treatment for individuals with CNS metastases at initial staging ◦ At the end of planned therapy for all individuals • Bone scan (see: **PEDONC-1.3** for coding) at the end of planned therapy only if positive at initial diagnosis
Rare circumstances to establish the presence of active disease only when a major therapeutic decision depends on PET avidity	• PET/CT (CPT® 78815) • Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Indication	Medically Necessary Imaging Study
Surveillance	• Every 3 months for 2 years after completion of all therapy: ◦ CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • Every 3 months for 3 years after completion of all therapy: ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) or MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) ▪ If primary site other than kidney, perform CT with contrast or MRI without and with contrast of primary site in place of abdominal imaging • Every 3 months for 1 year, then every 6 months for 1 year after completion of all therapy: ◦ MRI Brain without and with contrast (CPT® 70553) • If bone scan positive at initial diagnosis: ◦ Every 3 months for 1 year, then every 6 months for 2 years: ▪ Bone scan (see: **PEDONC-1.3** for coding) ◦ If negative at 36 months, no further advanced imaging • The role of surveillance imaging beyond these timeframes in unclear. Abdominal US (CPT® 76700) and chest x-ray may be considered.
Continued surveillance of individuals with rhabdoid tumor predisposition syndrome	• See: **Rhabdoid Tumor Predisposition Syndrome (PEDONC-2.11)**

Background and Supporting Information

- MRT is a highly aggressive histologic variant that can also occur in other locations and all non-CNS sites should follow these guidelines.
- Primary CNS rhabdoid malignancies should be imaged according to **Atypical Teratoid/Rhabdoid Tumors (ATRT) PEDONC-4.5)**
- Many individuals will present with an asymptomatic abdominal mass, and will undergo ultrasound as a primary evaluation.
- Other staging imaging should be deferred until a histologic diagnosis is made, by complete nephrectomy for most unilateral renal tumors and biopsy for bilateral renal tumors or inoperable unilateral tumors

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Congenital Mesoblastic Nephroma (CMN) (PEDONC-7.7)

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Congenital Mesoblastic Nephroma Initial Staging

- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary in all individuals
- CT Chest with (CPT® 71260) is medically necessary to evaluate inconclusive findings on chest x-ray

Congenital Mesoblastic Nephroma Treatment Response

- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary once following resection to establish baseline imaging, and those with a complete resection should then be imaged according to surveillance guidelines
- CT Abdomen and Pelvis with contrast (CPT® 74177) is medically necessary for individuals receiving preoperative chemotherapy every 2 cycles of therapy until surgery and then should be imaged according to surveillance guidelines after their postoperative baseline imaging study

Congenital Mesoblastic Nephroma Surveillance Imaging

- Ultrasound is medically necessary as it is the preferred surveillance imaging modality to avoid radiation and anesthesia exposures
- CT Abdomen and Pelvis with contrast (CPT® 74177) **or** MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary for residual abnormalities present on post-operative imaging or inconclusive findings on ultrasound every 3 months for 1 year after completion of all therapy

Background and Supporting Information

- This is the most common primary renal tumor occurring in young infants, and the overall prognosis is very good
- Complete surgical removal is curative in most cases, and histologically confirmed metastatic disease or bilateral disease has never been reported
- Many individuals will present with an asymptomatic abdominal mass at the time of birth or abnormal prenatal ultrasound, and will undergo ultrasound as a primary evaluation.
- PET is not indicated in the initial staging of any pediatric renal tumor
- Recurrences are rare but most occur within 12 months of diagnosis

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Pediatric Soft Tissue Sarcomas (PEDONC-8)

Guideline

Pediatric Soft Tissue Sarcomas – General Considerations (PEDONC-8.1)
Rhabdomyosarcoma (RMS) (PEDONC-8.2)
Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)
References (PEDONC-8)

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Pediatric Soft Tissue Sarcomas – General Considerations (PEDONC-8.1)

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- Unless specified below, individuals age <18 years old should be imaged according to this guideline section. Exceptions include:
  - Rhabdomyosarcoma in individuals (except uterine rhabdomyosarcoma) of all ages should be imaged according to guidelines in **Rhabdomyosarcoma (RMS) (PEDONC-8.2)**
  - Uterine rhabdomyosarcoma individuals of all ages should be imaged according to guidelines in Uterine Cancer (ONC-22). See: **Uterine Cancer - General Considerations (ONC-22.0)** in the Oncology Imaging Guidelines
  - Kaposi's sarcoma in individuals of all ages should be imaged according to guidelines in **Kaposi's Sarcoma (ONC-31.10)** in the Oncology Imaging Guidelines
- Evaluation of soft tissue masses of uncertain nature prior to biopsy should follow general imaging guidelines:
  - Individuals age 0 to 17 years, see: **Soft Tissue and Bone Masses (PEDMS-3)** in the Pediatric Musculoskeletal Imaging Guidelines
  - Individuals age ≥18 years, see: **Bone Lesion (MS-10.2)** in the Musculoskeletal Imaging Guidelines
- Where there are indications for PET/CT in these guidelines, note that indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Rhabdomyosarcoma (RMS) (PEDONC-8.2)

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- Individuals of all ages are imaged according to this guideline, with exception of uterine rhabdomyosarcoma, which is imaged according to Uterine Cancer (ONC-22). See: **Uterine Cancer – General Considerations (ONC-22.0)** in the Oncology Imaging Guidelines.

Indication	Medically Necessary Imaging Study
Initial staging for all individuals	ALL of the following: • MRI without and with contrast of primary site ◦ CT with contrast if MRI is contraindicated • CT Chest with contrast or CT Chest without contrast ◦ Should be completed prior to anesthesia exposure if possible • Whole-Body PET/CT (CPT® 78816) ◦ Bone scan substituted for PET, if PET not available
Additional initial staging for ANY of the following: • Evaluation of inconclusive PET findings in the abdomen or pelvis • Primary site of abdomen or pelvis • Lower extremity primary site	• CT Abdomen and Pelvis with contrast (CPT 74177)

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Indication	Medically Necessary Imaging Study
Additional initial staging for ANY of the following: • Primary site involving the paraspinal or paravertebral region • PET or bone scan-avid lesions in skull, neck, vertebrae • Neurologic signs or symptoms raising suspicion of CNS metastases	ALL of the following: • MRI Brain (CPT® 70553) • MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)
Treatment response, every 2 cycles during treatment, prior to local control surgery, and at the end of planned therapy	ALL of the following: • CT Chest with (CPT® 71260) contrast or CT Chest without contrast (CPT® 71250) • MRI without and with contrast of primary site ◦ CT if used at initial imaging for MRI contraindication • CT with contrast or MRI without and with contrast of all known metastatic sites using the same conventional imaging modality as per initial staging
ANY of the following: • Response assessment prior to local control surgery or radiation therapy • Evaluation of residual mass visible on conventional imaging as part of end of therapy evaluation • Response assessment of disease visible on PET but not conventional imaging • PET results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance.	• Whole-body PET/CT (CPT® 78816) ◦ Once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not medically necessary for continued disease monitoring or surveillance unless one of the exceptions in section **General Guidelines (PEDONC-1.0)** applies.

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Indication	Medically Necessary Imaging Study
Restaging following local control surgery	ALL of the following: • MRI without and with contrast of primary site ◦ CT if used at initial imaging for MRI contraindication • CT with contrast or MRI without and with contrast of all known metastatic sites using the same conventional imaging modality as per initial staging
Surveillance, localized RMS	ALL of the following, every 3 months for 1 year, then every 4 months for 2 years, then every 6 months for 1 year after completion of all therapy: • MRI without or without and with contrast of primary tumor site ◦ CT if MRI contraindicated • Chest x-ray ◦ CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) for new or worsening clinical symptoms of chest disease or new findings on chest x-ray
Surveillance, metastatic RMS	ALL of the following, every 3 months for 1 year, then every 4 months for 2 years, then every 6 months for 1 year after completion of all therapy: • MRI without or without and with contrast of primary tumor site • CT if contraindication to MRI • CT Chest with (CPT® 71260) *or* without contrast (CPT® 71250) • CT with or without contrast of all known metastatic sites • Nuclear bone scan (see: **PEDONC-1.3** for coding) for surveillance of known bony metastases
Suspected recurrence	• Repeat conventional imaging as per initial staging for all individuals

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Indication	Medically Necessary Imaging Study
ONE of the following: • Biopsy-proven recurrence • Conventional imaging (CT, MRI, US, plain film) reveals findings that are inconclusive or suspicious for recurrence, and PET avidity will determine whether biopsy or continued observation is appropriate • Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence, and PET would replace conventional imaging modalities.	• Whole-body PET/CT (CPT® 78816)

Background and Supporting Information

- Soft tissue sarcomas occur in both adult and pediatric individuals, but some are more common in one age group than the other.
- Pediatric soft tissue sarcomas are divided into **two** groups:
  - Rhabdomyosarcoma (RMS) accounts for ~60% of soft tissue sarcomas in young individuals, but only ~25% of soft tissue sarcomas in adolescents
  - Non-rhabdomyosarcoma soft tissue sarcomas (NRSTS) which encompasses all other histologic subtypes
- Because RMS can arise from any muscle tissue, the presenting symptoms and primary tumor sites vary widely and strongly influence the appropriate imaging decisions
- CT with contrast is the preferred primary site imaging modality for RMS arising in the thoracic cavity (not the chest wall)
- MRI without and with contrast is the preferred primary site imaging modality for RMS occurring in all other anatomic locations, including the chest wall.
- PET/CT is superior to conventional imaging for detection of nodal and bony metastases in pediatric RMS and is indicated in the initial staging of all individuals after histologic diagnosis is established
- CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely indicated in the initial metastatic staging of pediatric RMS
- PET is not routinely utilized to assess treatment response in RMS

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- PET is generally not indicated during active treatment for recurrent pediatric cancer
- Residual mass that has not changed in size since the last conventional imaging does not justify PET imaging during surveillance
- PET avidity in a residual mass at the end of planned therapy is not an indication for PET imaging during surveillance

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Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)

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Indication	Medically Necessary Imaging Study
Initial staging of all individuals	ALL of the following: • MRI without and with contrast of primary site ◦ CT with contrast if contraindication to MRI • CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) ◦ Should be completed prior to anesthesia if possible • For primary tumor of the chest wall, both MRI Chest wall without and with contrast (CPT® 71552) and CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) to assess for pulmonary metastatic disease (indicated simultaneously) • PET/CT (CPT® 78815) ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement • Nuclear bone scan (see: **PEDONC-1.3** for coding) if PET is not available

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Indication	Medically Necessary Imaging Study
ANY of the following: • Additional initial staging for individuals with disease in the abdomen, pelvis, or lower extremities • Angiosarcoma, alveolar soft part sarcoma, clear cell sarcoma, epithelioid sarcoma, hemangiopericytoma, leiomyosarcoma, liposarcoma, retroperitoneal or intra-abdominal primary site (including pelvic primary site), other histologies documented to have propensity for lymphatic spread and deep-seated tumors • Inconclusive findings in the abdomen or pelvis on PET/CT	In addition to the above, one of the following combinations: • CT Abdomen and Pelvis with contrast (CPT® 74177) if not already performed • MRI Abdomen (CPT® 74183) and MRI Pelvis (CPT® 72197) without and with contrast • MRI Abdomen (CPT® 74183) with and without contrast and CT Pelvis (CPT® 72193) with contrast • CT Abdomen (CPT® 74160) with contrast and MRI Pelvis (CPT® 72197) without and with contrast
Further evaluation of ANY of the following: • PET or nuclear bone scan-avid lesions in skull, neck, or vertebrae seen on initial imaging • Neurologic signs or symptoms raising suspicion of CNS metastases • Myxoid round cell liposarcoma, angiosarcoma, alveolar soft part sarcoma, cardiac sarcoma • Additional staging for individuals with primary site arising in the paraspinal or paravertebral region	ALL of the following: • MRI Brain (CPT® 70553) • MRI Spine (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)

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Indication	Medically Necessary Imaging Study
Treatment response, every 2 cycles of treatment, prior to local control, and at the end of planned therapy	Either of the following: • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250), and • MRI without and with contrast of primary site ◦ CT if contraindication to MRI and • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging • Imaging of all known metastatic sites using the same conventional imaging modality as per initial staging OR • PET/CT (CPT® 78815) if disease visible on initial staging PET but not on conventional imaging ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement
ANY of the following, if PET positive at initial diagnosis: • Assessment prior to local control surgery or radiation therapy • Evaluation of residual mass visible on conventional imaging as part of end of therapy evaluation • Rare circumstances when PET results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance	• PET/CT (CPT® 78815) ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement ◦ Once PET has been documented to be negative for a given individual's cancer or all PET-avid disease has been surgically resected, PET is not medically necessary for continued disease monitoring or surveillance unless one of the exceptions in section **General Guidelines (PEDONC-1.0)** applies.

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Indication	Medically Necessary Imaging Study
Restaging following local control surgery	ALL of the following: • MRI without and with contrast of the primary site ◦ CT if contraindication to MRI • Imaging of all known metastatic sites using the same conventional imaging modality as per initial staging • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging
Surveillance, low grade, localized NRSTS of extremity or trunk treated with resection (with or without radiation) or chemotherapy	Every 3 months for the first 12 months, then every 4 months in years 2 and 3, and every 6 months in years 4 and 5, then annually thereafter: • MRI without and with contrast of the primary site • CT if MRI contraindicated AND Every 6 months for 3 years, then annually thereafter: • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250)
Surveillance all other NRSTS	Every 3 months for the first 2 years, then every 6 months in years 3 and 4, and then annually: • MRI without and with contrast of the primary site ◦ CT if MRI contraindicated ◦ Nuclear bone scan (see: **PEDONC-1.3** for coding) for surveillance of known bony metastases • CT Chest with (CPT® 71260) or CT Chest without contrast (CPT® 71250) • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging

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Indication	Medically Necessary Imaging Study
ANY of the following: • Angiosarcoma • Alveolar soft part sarcoma • Cardiac sarcoma	• In addition to the above studies: ◦ MRI Brain without and with contrast (CPT® 70553) annually For surveillance of individuals with known brain metastases, see: **Brain Metastases (ONC-31.3)**
Surveillance, recurrent NRSTS	ALL of the following after successful treatment for recurrent disease, every 3 months for 1 year (surveillance after year 1 should follow the standard timing listed in the appropriate surveillance section above): • CT Chest (CPT® 71260) with contrast • CT with contrast or MRI without and with contrast of the primary site • CT Abdomen and Pelvis with contrast (CPT® 74177) for the same indications as abdominal/ pelvic imaging for initial staging
ANY of the following: • Conventional imaging (CT, MRI, US, plain film) during surveillance reveals findings that are inconclusive or suspicious for recurrence, and PET avidity will determine whether biopsy or continued observation is appropriate • Obvious clinical symptoms show strong evidence suggesting recurrent and PET would replace conventional imaging modalities	• PET/CT (CPT® 78815) ◦ Whole-body PET/CT (CPT® 78816) if there is clinical suspicion of skull or distal lower extremity involvement

Background and Supporting Information

- All soft tissue sarcomas other than RMS fall into this category
- Because soft tissue sarcomas can arise from any soft tissue, the presenting symptoms and primary tumor sites vary widely and strongly influence the appropriate imaging decisions

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- CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely indicated in the initial metastatic staging of pediatric NRSTS
- Many individuals with NRSTS will be treated with surgical resection alone, and these individuals enter immediately into surveillance
- PET imaging is not routinely utilized to assess treatment response in NRSTS
- PET imaging is generally not indicated during active treatment for recurrent pediatric cancer.
- Residual mass that has not changed in size since the last conventional imaging does not justify PET
- PET avidity in a residual mass at the end of planned therapy is not an indication for PET imaging during surveillance
- Given that pediatric-specific resources are limited in regards to surveillance of NRSTS, this guideline combines published pediatric literature with NCCN recommendations for the same histologies.

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Bone Tumors (PEDONC-9)

Guideline

Bone Tumors – General Considerations (PEDONC-9.1)
Benign Bone Tumors (PEDONC-9.2)
Osteogenic Sarcoma (OS) (PEDONC-9.3)
Ewing Sarcoma Family of Tumors (ESFT), Including Primitive Neuroectodermal Tumors (PNET) (PEDONC-9.4)
References (PEDONC-9)

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Bone Tumors – General Considerations (PEDONC-9.1)

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These guidelines include both benign and malignant lesions.

- Unless specified below, individuals who are <18 years old should be imaged according to this guideline section. Exceptions include:
  - Osteogenic sarcoma individuals of all ages should be imaged according to guidelines in **Osteogenic Sarcoma (OS) (PEDONC-9.3)**
  - Ewing Sarcoma and Primitive Neuroectodermal Tumor individuals of all ages should be imaged according to guidelines in **Ewing Sarcoma and Primitive Neuroectodermal Tumors (ESFT) (PEDONC-9.4)**
  - Chondrosarcoma individuals of all ages should be imaged according to guidelines in **Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)**
  - Chordoma individuals of all ages should be imaged according to guidelines in **Bone Sarcomas – Initial Work-up/Staging (ONC-12.6)**
  - Giant cell tumor of bone and enchondroma individuals of all ages should be imaged according to guidelines in **Benign Bone Tumors – General Considerations (ONC-12.9)**
  - Other benign bone tumor individuals of all ages should be imaged according to guidelines in **Benign Bone Tumors (PEDONC-9.2)**
- Prosthetic devices for children after surgery for bony tumors are nearly all customized.
  - CT, contrast as requested in alignment with prosthetic manufacturer specifications, is medically necessary when requested by the operating surgeon for planning for customized-to-individual joint replacement or prosthetic surgery.
- All bone tumors should be evaluated by plain x-ray prior to any advanced imaging.
- PET does not reliably distinguish between benign and malignant bone tumors and should not be performed prior to biopsy.
- Where indications for PET/CT are noted, please note that indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**.
- CT Chest is superior to PET/CT for the detection of pulmonary metastases, and is medically necessary in the initial workup of all suspected malignant bone tumors.
  - CT Chest should be completed prior to anesthesia exposure, if possible.

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Benign Bone Tumors (PEDONC-9.2)

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- Osteochondroma

Indication	Medically Necessary Imaging Study
Preoperative planning	• MRI without and with contrast after evaluation by the operating surgeon • See: **General Considerations (PEDONC-9.1)** for requests related to prosthetic planning
• ANY of the following: ◦ Concern for malignant transformation ◦ New or worsening pain/symptoms ◦ Change on a recent plain x-ray	• MRI without contrast or without and with contrast

- Osteoid osteoma

Indication	Medically Necessary Imaging Study
Suspected, based on clinical history and plain film findings	• CT without contrast
• ANY of the following: ◦ CTs are not characteristic for diagnosis ◦ Individual has bone pain not localized to the area of findings on CT or x-ray	• ONE of the following: ◦ Triple phase bone scan (CPT® 78315) ◦ SPECT (CPT® 78803) ◦ Hybrid SPECT/CT (CPT® 78830)
• Individuals with new pain who have been previously treated with radiofrequency ablation or curettage	• Bone scan (CPT® code 78830, 78315, or 78803 – as requested)

- Other benign tumors
  - Refer to **Mass Involving Bone (Including suspected Lytic and Blastic Metastatic Disease) (PEDMS-3.4)**

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Background and Supporting Information

- Plain x-ray appearance is diagnostic for many benign bone tumors, and advanced imaging is generally unnecessary except for preoperative planning
- Plain x-ray appearance is diagnostic for osteochondroma for the majority of individuals and advanced imaging is generally unnecessary

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Osteogenic Sarcoma (OS) (PEDONC-9.3)

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Indication	Medically Necessary Imaging Study
Suspected or Biopsy Proven Diagnosis	• MRI without and with contrast of the primary site OR • CT, contrast as request, of the primary site if there is a contraindication to MRI AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • These studies are medically necessary even when PET/CT has already been authorized or performed
Initial Staging of Biopsy Proven Disease	• 18F-FDG PET/CT whole-body (CPT® 78816) ◦ Nuclear bone scan (see: **(PEDONC-1.3)** for coding) if PET not available ◦ PET/CT is medically necessary in addition to the conventional imaging listed in the suspected/ diagnosis sections • CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely medically necessary in the initial metastatic staging of pediatric OS but is medically necessary in the following situations: ◦ Evaluation of inconclusive PET findings ◦ Primary site of abdomen or pelvis
Suspected bony metastatic sites noted on PET or bone scan	• MRI without and with contrast of the suspected metastatic site OR • CT with contrast of the suspected metastatic site

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Indication	Medically Necessary Imaging Study
Restaging after 10 to 12 weeks of neoadjuvant chemotherapy prior to local control surgery	• MRI without and with contrast of primary site • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) • 18F-FDG PET/CT whole-body (CPT® 78816) or bone scan (see: **(PEDONC-1.3)** for coding)
Individuals with metastatic disease undergoing current chemotherapy	• CT Chest with (CPT®) or without contrast (CPT® 71250) can be performed every 2 cycles during treatment and at the end of planned chemotherapy • MRI without and with contrast of primary site can be performed every 2 cycles during treatment and at the end of planned chemotherapy • If previously positive for bony metastases, 18F-FDG PET/CT whole body (CPT® 78816) or bone scan (see: **(PEDONC-1.3)** for coding) every 2 cycles during treatment and at the end of planned chemotherapy • Imaging may be indicated more frequently around the time of surgical resection of primary or metastatic lesions to assess for resectability
Preoperative planning for local control surgery	• CT, contrast as requested • MRA and/or CTA may rarely be indicated for complicated surgical resections, and is medically necessary after evaluation by the operating surgeon to clarify inconclusive MRI findings for preoperative planning • See: **General Considerations (PEDONC-9.1)** for advancing imaging requests related to prosthetic planning

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Indication	Medically Necessary Imaging Study
Following local control surgery, until the end of planned adjuvant chemotherapy	• MRI without and with contrast of primary site ~6 weeks after surgical procedure, and at the end of planned chemotherapy • Plain x-rays of the primary site and chest immediately after local control and then every 2 months between the supported, listed advanced imaging studies. • CT Chest with (CPT® 71260) or without contrast (CPT® 71250): ◦ Measurable pulmonary metastases: every 6 weeks, and at the end of planned chemotherapy ◦ No measurable pulmonary metastases: every 4 months, and at the end of planned chemotherapy • Bone scan (see: **(PEDONC-1.3)** for coding) every 4 months, and at the end of planned chemotherapy ◦ 18F-FDG PET/CT whole-body (CPT® 78816) in place of bone scan, if positive for distant bone metastases at initial diagnosis
Recurrent metastatic or recurrent unresectable disease on treatment	• The following every 2 cycles of treatment, and at the end of planned chemotherapy: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ MRI without and with contrast of primary site ◦ PET is generally not medically necessary during active treatment for recurrent pediatric cancer. In rare circumstances, 18F-FDG PET/CT imaging with CPT® 78816 may be appropriate when results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance.

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Indication	Medically Necessary Imaging Study
Surveillance (all individuals regardless of primary site)	• Any or all of the following every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5 after completion of all therapy ◦ CT or MRI of primary site as performed during suspected or initial disease workup (provided the individual does not have an endoprosthesis that will cause MRI or CT artifact): ▪ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) • Bone scan (see: **(PEDONC-1.3)** for coding) every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually for 2 years after completion of all therapy
Suspected recurrence	• MRI without and with contrast of the primary site and / or site of suspected recurrence based on symptoms or other imaging OR • CT, contrast as request, of the primary site and/or site of suspected recurrence based on symptoms or other imaging if there is a contraindication to MRI AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely medically necessary in the metastatic staging of pediatric OS, but is medically necessary in the following situations: ◦ Evaluation of inconclusive PET findings ◦ Primary site of abdomen or pelvis • These studies are medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
• For suspected recurrence, ANY of the following: ◦ Conventional imaging reveals findings that are inconclusive or suspicious for recurrence and PET avidity will determine whether biopsy or continued observation is appropriate ◦ Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence and PET would replace conventional imaging modalities	• 18F-FDG PET/CT (CPT® 78816)
• Biopsy-proven recurrence	• PET/CT whole-body (CPT® 78816) for biopsy-proven recurrence ◦ If disease is considered potentially resectable, follow osteosarcoma treatment response as previous

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Ewing Sarcoma Family of Tumors (ESFT), Including Primitive Neuroectodermal Tumors (PNET) (PEDONC-9.4)

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Indication	Medically Necessary Imaging Study
Suspected/Diagnosis – ill-defined or non-discrete soft tissue mass without bony involvement	• US (CPT® 76881 or 76882) in addition to plain x-ray
Suspected or Biopsy Proven Diagnosis – Primary site	• MRI without and with contrast ◦ CT, contrast as requested, if there is a contraindication to MRI • MRA and/or CTA may rarely be indicated for complicated surgical resections, and is medically necessary after evaluation by the operating surgeon to clarify inconclusive MRI findings for preoperative planning • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is medically necessary in initial workup of all suspected or confirmed malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • These studies are medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
Suspected or Biopsy Proven Diagnosis – Chest wall primary	• MRI Chest without and with contrast AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is medically necessary in initial workup of all suspected or confirmed malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • These studies are medically necessary even when PET/CT has already been authorized or performed
Initial Staging (additional imaging after biopsy confirmed disease)	• PET/CT whole-body (CPT® 78816) ◦ Bone scan (see: **(PEDONC-1.3)** for coding) substituted for PET imaging if PET not available ◦ PET/CT is medically necessary in addition to the conventional imaging listed in the suspected/ diagnosis sections
• For ANY of the following: ◦ Evaluation of inconclusive PET findings ◦ Primary site involving the abdomen or pelvis	• CT Abdomen and Pelvis with contrast (CPT® 74177)
Restaging after 10 to 12 weeks of neoadjuvant chemotherapy prior to local control surgery	Imaging modality should be the same as used for initial staging, any or all from the list below as described: • MRI without and with contrast of the primary site • CT (contrast as requested) if requested per valuation by the operating surgeon for pre-operative planning ◦ See: **General Considerations (PEDONC-9.1)** for imaging requests related to prosthetic planning • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) • ONE of the following: ◦ PET/CT whole-body (CPT® 78816) ◦ Whole-body bone scan (see: **PEDONC-1.3** for coding) ◦ MRI bone marrow blood supply/diffusion-weighted MRI with ADC (CPT® 77084)

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Indication	Medically Necessary Imaging Study
Treatment response following local control surgery	• MRI without and with contrast of primary site 3 months after surgical procedure and at the end of planned chemotherapy • Plain x-rays of the primary site and chest immediately after local control then every 3 months • CT Chest with (CPT® 71260) or without contrast (CPT® 71250): ◦ Measurable pulmonary metastases: every 6 weeks and at the end of planned chemotherapy ◦ No measurable pulmonary metastases: every 3 months and at the end of planned chemotherapy • Whole-body bone scan (see: **PEDONC-1.3** for coding) at the end of planned chemotherapy • PET/CT whole-body (CPT® 78816) for clinical or imaging findings suggestive of local recurrence
Individuals with metastatic disease undergoing current chemotherapy	• CT Chest with (CPT® 71260) or without contrast (CPT® 71250) every 2 cycles during treatment and at the end of planned chemotherapy • MRI without and with contrast of primary site every 2 cycles during treatment and at the end of planned chemotherapy • If previously positive for bony metastases, PET/CT whole-body (CPT® 78816) or bone scan (see: **(PEDONC-1.3)** for coding) every 2 cycles during treatment and at the end of planned chemotherapy • Imaging may be indicated more frequently around the time of surgical resection of primary or metastatic lesions to assess for resectability

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Indication	Medically Necessary Imaging Study
Recurrent metastatic or recurrent unresectable disease on treatment	• Every 2 cycles of treatment, and at the end of planned chemotherapy: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ MRI without and with contrast of primary site ◦ PET is generally not medically necessary during active treatment for recurrent pediatric cancer. In rare circumstances, PET imaging may be appropriate when results are likely to result in a treatment change for the individual, including a change from active treatment to surveillance.
Surveillance for all disease **other than** low grade, stage I disease	• Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5, and annually for 5 years after completion of all therapy to complete 10 years of surveillance: ◦ CT or MRI of primary site as performed during suspected or initial disease workup (provided the individual does not have an endoprosthesis that will cause MRI or CT artifact): ◦ Bone scan (see: **(PEDONC-1.3)** for coding) • Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5: ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ After year 5, CXR should be used for surveillance

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Indication	Medically Necessary Imaging Study
Surveillance for low grade stage I disease only	• Every 3 months for year 1 and 2, then every 4 months in year 3, then every 6 months in year 4 and 5, and annually for 5 years after completion of all therapy to complete 10 years of surveillance: ◦ CT or MRI of primary site as performed during suspected or initial disease workup (provided the individual does not have an endoprosthesis that will cause MRI or CT artifact) ◦ Bone scan (see: **PEDONC-1.3** for coding) • Every 3 months for year 1 and 2 ◦ CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ After 24 months off therapy, CXR should be used for surveillance with CT supported for new or inconclusive findings on CXR
Suspected Recurrence	• MRI without and with contrast of the primary site and/or site of suspected recurrence based on symptoms or other imaging, OR • CT, contrast as request, of the primary site and/or site of suspected recurrence based on symptoms or other imaging if there is a contraindication to MRI AND • CT Chest with (CPT® 71260) or without contrast (CPT® 71250) ◦ CT Chest is medically necessary in initial workup of all suspected malignant bone tumors in children and should be completed prior to anesthesia exposure if possible • CT Abdomen and Pelvis with contrast (CPT® 74177) is not routinely medically necessary in the metastatic staging of pediatric EWS but is medically necessary in the following situations: ◦ Evaluation of inconclusive PET findings ◦ Primary site of abdomen or pelvis • These studies are medically necessary even when PET/CT has already been authorized or performed

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Indication	Medically Necessary Imaging Study
• For suspected recurrence, any of the following: ◦ Conventional imaging reveals findings that are inconclusive or suspicious for recurrence and PET avidity will determine whether biopsy or continued observation is appropriate ◦ Rare circumstances where obvious clinical symptoms show strong evidence suggesting recurrence and PET would replace conventional imaging modalities	PET/CT whole-body (CPT® 78816)
• Biopsy proven recurrence	• Refer to ESFT initial imaging for studies for suspected recurrence. • PET/CT whole-body (CPT® 78816) • If disease is considered potentially resectable, follow ESFT treatment response as previously noted

Background and Supporting Information

- Bone and bone marrow metastases can occur in ESFT, and cause a significant change in treatment approach.
- 18F-FDG PET/CT can replace bone scan and bone marrow biopsy in ESFT individuals and is indicated in the initial staging of all ESFT individuals after histologic diagnosis is established

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Pediatric Germ Cell Tumors (PEDONC-10)

Guideline

Pediatric Germ Cell Tumors (PEDONC-10)
References (PEDONC-10)

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Pediatric Germ Cell Tumors (PEDONC-10)

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Indication	Medically Necessary Imaging Study
Initial staging for all individuals	• CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ For testicular masses, MRI Abdomen without and with contrast (CPT® 74183) and CT Pelvis with contrast (CPT® 72193) are medically necessary instead, if requested • CT Chest with contrast (CPT® 71260) ◦ Should be completed prior to anesthesia exposure if possible
• Initial staging, in lieu of CT Abdomen and Pelvis with contrast (above), for ANY of the following: ◦ Proven or highly suspected ovarian neoplasm that was already imaged with ultrasound ◦ Immature sacrococcygeal teratoma suspected on other imaging ◦ Contraindication to CT contrast OR • To clarify inconclusive findings on initial staging CT	• MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
• Ovarian masses that are <10 cm in size, have minimal or no visible solid component on ultrasound, and have normal tumor markers	• These are almost universally benign teratomas or functional cysts; thus, advanced imaging is not medically necessary

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Indication	Medically Necessary Imaging Study
• Additional initial evaluation for individuals with ANY of the following: ◦ Symptoms suggesting CNS metastases ◦ Choriocarcinoma syndrome (hemorrhagic metastatic disease to lung with extremely elevated HCG) HCG >10,000miU/ml	• MRI Brain without and with contrast (CPT® 70553)
Additional initial evaluation for individuals with systemic symptoms or bone pain	• Nuclear bone scan (See: **PEDONC-1.3** for coding)
Restaging in individuals with disease not completely resected at initial diagnosis	• Every 2 cycles (~every 6 weeks) and at the end of planned therapy: ◦ CT Chest/Abdomen/Pelvis (CPT® 71260 and CPT® 74177) with contrast ▪ CT imaging may be indicated more frequently to assess for surgical resectability in individuals who have received more than 4 cycles of chemotherapy ▪ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT abdomen and pelvis for ovarian neoplasms if this modality was used for initial staging or for contraindication to CT contrast ◦ Imaging of any metastatic sites with the same modality used during initial staging
• Suspicious lesion seen on CT and both of the following: ◦ Relapse risk is determined to be low by the treating physician ◦ Biopsy would cause unnecessary morbidity for the individual	• Short-interval CT study of the involved area

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Indication	Medically Necessary Imaging Study
Stage I individuals age 0-10 years treated with surgery only	• Chest x-ray should be completed every 3 months for 1 year, then every 6 months in year two, to complete two years surveillance after completion of all therapy ◦ For those with primary mediastinal tumors at diagnosis, CT Chest with contrast (CPT® 71260) in lieu of CXR according to the above schedule. • Every 3 months for 1 year, then every 6 months in year two, to complete two years surveillance after completion of all therapy: ◦ CT Abdomen/Pelvis with contrast (CPT® 74177) ▪ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) substituted for surveillance for ovarian neoplasm if this modality was used for initial staging
Stage I individuals ages 11+ years treated with surgery only	• Every 4 months for 2 years, then every 6 months for 1 year, then every 12 months for 2 years to complete 5 years surveillance imaging after completion of all therapy: ◦ Chest x-ray ◦ For individuals with primary mediastinal tumors at diagnosis, CT Chest with contrast (CPT® 71260) in lieu of chest x-ray on the above schedule. • Every 4 months for 2 years, then every 6 months for 1 year, then every 12 months for two years to complete 5 years surveillance ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) ▪ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) substituted for surveillance for ovarian neoplasm if this modality was used for initial staging

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Indication	Medically Necessary Imaging Study
Stage II-IV individuals	• CT Chest with contrast (CPT® 71260) or CT Chest without contrast (CPT® 71250) and CT Abdomen and Pelvis with contrast (CPT® 74177) every 3 months for 1 year then every 6 months in year 2, then annually in years 3-5 after completion of all therapy. ◦ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) substituted for surveillance for ovarian neoplasm if this modality was used for initial staging • Individuals with brain or bone metastases should have surveillance imaging of those areas on the same schedule as the primary site imaging with the same modality used during initial staging
• Suspected Recurrence: ◦ Any clinically significant rise in tumor markers ◦ Symptoms suggesting recurrent disease ◦ Abnormal chest x-ray	• CT Chest with contrast (CPT® 71260) and CT Abdomen and Pelvis with contrast (CPT® 74177) ◦ MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) in lieu of CT for suspected recurrence of immature sacrococcygeal tumor or ovarian neoplasm or for contraindication to CT contrast • Whole-body bone scan (CPT® 78306) for individuals with a history of bone involvement or with bone pain
Clarify inconclusive findings on conventional imaging	PET/CT (CPT® 78815)

Background and Supporting Information

General Considerations:

- Malignant pediatric germ cell tumors commonly include one of four histologic subtypes:
  - Yolk sac tumor
  - Choriocarcinoma
  - Embryonal carcinoma
  - Mixed histology (including immature sacrococcygeal teratoma)

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- Tumors can occur in testicular, ovarian or extragonadal primary locations
- Sex cord stromal tumors (granulosa cell, theca, sertoli, and leydig tumors) are rare in pediatrics and should be imaged according to general guidelines in: **Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)** in the Oncology Imaging Guidelines
- This section applies to primary germ cell tumors occurring outside the central nervous system in children who are ≤15 years old at the time of initial diagnosis.
  - For individuals who are >15 years old at diagnosis, the overall prognosis is inferior and these individuals should be imaged according to general guidelines in: **Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20)** in the Oncology Imaging Guidelines.
- For CNS germ cell tumors, use the imaging guidelines in: **CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)**.

Treatment Considerations:

- Overall treatment strategies are similar for all malignant germ cell tumors.
- Individuals with localized GCT are often cured with surgery alone and do not receive adjuvant therapy.
  - These individuals should be imaged using surveillance guidelines after surgery is completed.
- Individuals receiving adjuvant chemotherapy are usually treated with 4 to 6 cycles of combination chemotherapy.

Imaging Modality Considerations:

- Initial evaluation of: Ovarian, testicular, and abdominal extragonadal suspected GCT should be completed by ultrasound and tumor markers (AFP, β-hCG)
  - Once a primary mass suspected to be GCT is discovered, initial staging is indicated prior to histologic confirmation
  - The degree of abdominal exploration and node sampling necessary for adequate staging is determined in part by imaging findings and is required for preoperative planning
- The primary method of response assessment is by tumor marker decrease
- The primary method of surveillance in pediatric GCT is frequent assessment of serum tumor markers, unless tumor markers were not elevated at diagnosis
- Surveillance imaging of the chest in disease stages I-IV should generally be performed using chest x-ray
  - See surveillance indications for specific imaging recommendations
- There has been no published evidence to date supporting the routine use of PET/CT in the evaluation of pediatric GCT

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  - Additionally, PET has been found to have similar efficacy to CT imaging in initial staging of adults with non-seminomatous GCT (the majority of pediatric GCT are non-seminomatous)
- PET as a marker of treatment response has been shown not to be predictive of individual outcomes in GCT and should not be approved
  - Suspicious lesions seen on conventional imaging should be biopsied to confirm active disease

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Pediatric Liver and Pancreatic Tumors (PEDONC-11)

Guideline

Pediatric Liver and Pancreatic Tumors – General Considerations (PEDONC-11.1)
Hepatoblastoma (PEDONC-11.2)
Pediatric Hepatocellular Carcinoma (HCC) (PEDONC-11.3)
Pediatric Pancreatic Carcinoma (PEDONC-11.4)
References (PEDONC-11)

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Pediatric Liver and Pancreatic Tumors – General Considerations (PEDONC-11.1)

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- Primary hepatic germ cell tumors should follow imaging guidelines in: **Pediatric Germ Cell Tumors (PEDONC-10)**.
- Primary hepatic sarcomas should follow imaging guidelines in: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS) (PEDONC-8.3)**.
- Imaging requests relating to liver transplant surgery and surveillance should follow guidelines in section **Transplant (AB-42)** in the Abdomen Imaging Guidelines.

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Hepatoblastoma (PEDONC-11.2)

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Hepatoblastoma Initial Staging:

- Ultrasound is medically necessary even after MRI or CT imaging in order to allow evaluation for tumor thrombus
- Once a primary liver mass is discovered, definitive imaging is medically necessary prior to histologic diagnosis, and may involve ANY of the following:
  - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) for evaluating tumor margins and vascular anatomy
    - Hepatobiliary-specific contrast agents (gadoxetate, gadobenate [MultiHance]) are preferred if available and are medically necessary if requested, whether or not a prior gadolinium-enhanced MRI has been previously performed
    - CT Abdomen and Pelvis with (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
      - Non-contrast imaging is not medically necessary due to the increased radiation exposure and limited additive benefit
  - MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) to evaluate vascular invasion
- CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250) is medically necessary in the initial work-up of all pediatric liver tumors and should be completed prior to anesthesia exposure if possible
- Bone scan (See: **Modality General Considerations (PEDONC-1.3)**) is medically necessary for initial evaluation of bony metastases only with systemic symptoms or bone pain.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary only for symptoms suggesting CNS metastases
- PET/CT is medically necessary only in very rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making.
  - Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

Hepatoblastoma Treatment Response:

- CT Chest with (CPT® 71260) **or** without contrast (CPT® 71250) is medically necessary every 2 cycles **and** at the end of planned therapy for individuals with incomplete resection at initial diagnosis

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- MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary every 2 cycles **and** at the end of planned therapy for individuals with incomplete resection at initial diagnosis
  - CT Abdomen and Pelvis with contrast (CPT® 74177) or CT Abdomen and Pelvis without and with contrast (CPT® 74178) is inferior to MRI for pediatric liver malignancies and is only medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
    - Non-contrast imaging is not medically necessary due to the increased radiation exposure and limited additive benefit
- While the majority of individuals will require abdomen and pelvis imaging at all time points, the pelvis imaging may be omitted at the discretion of the ordering physician
- MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) is medically necessary to evaluate vascular invasion
- Imaging of any metastatic sites with the same modality used during initial staging is medically necessary every 2 cycles and at the end of planned therapy for individuals with incomplete resection at initial diagnosis
- Imaging is medically necessary more frequently to assess for surgical resectability in individuals who have received more than 4 cycles of chemotherapy.
- Abdominal ultrasound is medically necessary if tumor thrombus was detected at initial diagnosis
  - If no tumor thrombus was present, continued ultrasound evaluations are not medically necessary without a specific reason documented in the clinical records
- PET/CT is medically necessary only in very rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision making.

Hepatoblastoma Surveillance Imaging:

- For surveillance in individuals with an AFP of >100 ng/ml, the following imaging is medically necessary:
  - CT Chest with contrast (CPT® 71260) and MRI Abdomen without and with contrast (CPT® 74183) for any clinically significant rise in tumor markers or symptoms suggesting recurrent disease.
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
      - Non-contrast imaging is not medically necessary due to the increased radiation exposure and limited additive benefit
- For individuals with AFP ≤100 ng/ml at diagnosis or recurrence, the following imaging is medically necessary:

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- MRI Abdomen with and without contrast (CPT® 74183) every 3 months for 2 years, then every 4 months for 2 years after completion of all therapy
- Chest x-ray or CT Chest with contrast (CPT® 71260) every 3 months for 2 years, then every 4 months for 2 years after completion of all therapy
- Individuals with brain or bone metastases should have surveillance imaging on the same schedule as the primary site imaging with the same modality used during initial staging

Background and Supporting Information

- Pediatric liver tumors primarily include hepatoblastoma and hepatocellular carcinoma, but hepatic germ cell tumors and primary hepatic sarcomas occur with some frequency. Tumor markers are useful for initial evaluation as well as treatment response, particularly in hepatoblastoma. Early consideration of liver transplant may be undertaken in children and adolescents with unresectable localized disease, provided that the disease remains confined to the liver.
- Hepatoblastoma occurs most commonly in very young children (median diagnosis age of 19 months). Most cases of hepatoblastoma are sporadic, but some are associated with genetic abnormalities, including Beckwith-Wiedemann syndrome, familial adenomatous polyposis, and trisomy 18. Most suspected liver tumors will have ultrasound and tumor markers (AFP, β-HCG, CEA) as part of the initial evaluation.
- There has been no published evidence to date supporting the routine use of PET/CT imaging in the evaluation of pediatric Hepatoblastoma during initial imaging, treatment response, or surveillance.
- PET/CT should not be approved in lieu of biopsy of suspicious lesions
- Individuals with localized hepatoblastoma of pure fetal histology are often cured with surgery alone and do not receive adjuvant therapy. These individuals should be imaged using surveillance guidelines after surgery is completed.
- Individuals receiving adjuvant chemotherapy are usually treated with 2 to 8 cycles of combination chemotherapy. Tumor marker decrease is important in response assessment but does not eliminate the need for advanced imaging in individuals with unresected hepatoblastoma.
- The primary method of surveillance in hepatoblastoma is frequent assessment of serum tumor markers (primarily AFP).
- No specific imaging for surveillance in individuals with an AFP of >100 ng/mL at diagnosis or recurrence.

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Pediatric Hepatocellular Carcinoma (HCC) (PEDONC-11.3)

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Pediatric HCC Initial Staging:

- Ultrasound is medically necessary even after MRI or CT imaging in order to allow evaluation for tumor thrombus
- Once a primary liver mass is discovered, definitive imaging prior to histologic diagnosis including ANY of the following medically necessary studies:
  - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
    - Hepatobiliary specific contrast agents (gadoxetate, gadobenate [MultiHance]) are preferred if available and are medically necessary if requested, whether or not a prior gadolinium-enhanced MRI has been previously performed.
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
      - Noncontrast imaging is not medically necessary due to the increased radiation exposure and limited additive benefit
  - MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) to evaluate vascular invasion
- CT Chest with (CPT® 71260) **or** without contrast (CPT® 71250) is medically necessary in the initial work-up of all pediatric liver tumors and should be completed prior to anesthesia exposure if possible
- MRI Brain without and with contrast (CPT® 70553) is medically necessary only for symptoms suggesting CNS metastases
- Nuclear bone scan (See **Modality General Considerations (PEDONC-1.3)**) is medically necessary for initial evaluation of bony metastases only in individuals with systemic symptoms or bone pain
- PET/CT is medically necessary in very rare circumstances for preoperative planning when MRI and CT are insufficient for surgical decision-making.
  - PET/CT should not be approved in lieu of biopsy of suspicious lesions
  - Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**

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Pediatric HCC Treatment Response:

- For individuals with disease not completely resected at initial diagnosis, the following imaging is medically necessary every 2 cycles (~6 weeks) **and** at the end of planned therapy:
  - CT Chest with (CPT® 71260) **or** CT Chest without contrast (CPT® 71250)
  - MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197)
    - While the majority of individuals will require abdomen and pelvis imaging at all time points, the pelvis imaging may be omitted at the discretion of the ordering physician
    - CT Abdomen and Pelvis with contrast (CPT® 74177) is inferior to MRI for pediatric liver malignancies and is only medically necessary if MRI is not available, contraindicated, or sedation is required but not available, or to clarify specific inconclusive areas on MRI.
      - Non-contrast imaging is not medically necessary due to the increased radiation exposure and limited additive benefit
      - MRA Abdomen (CPT® 74185) or CTA Abdomen (CPT® 74175) to evaluate vascular invasion
- Imaging of any metastatic sites with the same modality used during initial staging is medically necessary
- Abdominal ultrasound is medically necessary if tumor thrombus was detected at initial diagnosis
  - If no tumor thrombus was present, continued ultrasound evaluations are not medically necessary without a specific reason documented in the clinical records
- PET/CT is medically necessary in very rare circumstances for preoperative planning when MRI and CT scans are insufficient for surgical decision-making.

Pediatric HCC Surveillance Imaging:

- MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) is medically necessary every 3 months for 1 year, then every 6 months for 1 year, then annually for 3 years after completion of all therapy
- Chest x-ray or CT Chest with contrast (CPT® 71260) is medically necessary every 3 months for 1 year, then every 6 months for 1 year, then annually for 3 years after completion of all therapy

Background and Supporting Information

- Individuals with brain or bone metastases should have surveillance imaging on the same schedule as the primary site imaging with the same modality used during initial staging.

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- HCC, including its rare histologically distinct variant fibrolamellar hepatocellular carcinoma (FL-HCC), occurs mostly in older children and adolescents. Despite recent advances in treatment, overall survival of pediatric HCC diagnosed in advanced stages remains exceedingly poor, with five-year survival of only 17% to 22% for all stages of pediatric HCC (and FL-HCC). Most suspected liver tumors will have ultrasound and tumor markers (AFP, β-HCG, CEA) as initial evaluation.
- PET/CT should not be approved in lieu of biopsy of suspicious lesions
- The majority of hepatocellular carcinoma individuals are treated with surgery alone and do not receive adjuvant therapy. Individuals with successful upfront gross total resection should be imaged using surveillance guidelines after surgery is completed.
- PET/CT has no documented role in the surveillance evaluation of pediatric hepatocellular carcinoma.

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Pediatric Pancreatic Carcinoma (PEDONC-11.4)

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- This guideline applies to suspected or diagnosed pancreatic neoplasms in children.
- Beckwith-Wiedemann syndrome increases pancreatoblastoma risk. MEN1, VHL, neurofibromatosis, and tuberous sclerosis are also risk factors for pancreatic endocrine neoplasms.
  - Screening studies in these conditions are found in **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)**.

Indication	Medically Necessary Imaging Study
Initial Staging	▪ CT Abdomen and Pelvis with or without and with contrast (CPT® 74177 or CPT® 74178) OR MRI Abdomen without and with contrast plus MRI Pelvis with and without contrast (CPT® 74183 and CPT® 72917) ▪ CT Chest with (CPT® 71260) or without (CPT® 71250) contrast ▪ ^{68}Ga-Dotatate PET/CT whole-body (CPT® 78816) - Exception: for pediatric pancreatic tumor of non neuroendocrine origin (SPN, pancreatoblastoma and other exocrine origin) with equivocal conventional imaging, FDG PET/CT whole-body or skull to thighs (CPT® 78816 or 78815)
Treatment response ~every 2 cycles during treatment and at the end of planned therapy	▪ CT or MRI as used at time of initial imaging
If conventional imaging for treatment response is equivocal	▪ Neuroendocrine tumors: ^{68}Ga Dotatate PET/CT skull to thighs or whole-body (CPT® 78815 or 78816) ▪ Non-neuroendocrine tumors: FDG PET/CT skull to thighs or whole-body (CPT® 78815 or 78816)
Assess candidacy for PRRT therapy	▪ ^{68}Ga Dotatate PET/CT whole body CPT® 78816 to assess candidacy for PRRT therapy

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Indication	Medically Necessary Imaging Study
Surveillance	▪ MRI or CT, modality and contrast as used in initial imaging, every 3 months for 2 years, then every 4 months for 1 year, then every 6 months for 1 year, then in 12 months to complete 5 years surveillance.
Suspected recurrence	▪ Repeat all imaging as noted in initial staging section

Background and Supporting Information

- Pancreatic tumors in children are exceedingly rare. The most common are solid pseudopapillary neoplasm (SPN), a low-grade epithelial malignancy. Pancreatoblastoma is the second most common.

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Retinoblastoma (PEDONC-12)

Guideline

Retinoblastoma – General Considerations (PEDONC-12.1)
Retinoblastoma – Imaging (PEDONC-12.2)
References (PEDONC-12)

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Retinoblastoma – General Considerations (PEDONC-12.1)

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- Retinoblastoma (RB) is primarily a disease of the infant and young child, and presents with leukocoria (loss of red reflex).
- About 75% of individuals are diagnosed before the age of two years (bilateral RB presents at 12 months of age).
- Retinoblastoma can occur as heritable (25% of cases) or nonheritable (75%) disease.
  - Heritable RB is associated with a germline mutation in the RB1 gene often resulting typically in bilateral disease.
  - Individuals who carry the RB1 mutation also have increased risk of developing other cancers, such as osteosarcoma, soft tissue sarcomas, or melanoma. For more information on heritable retinoblastoma, see: **Familial Retinoblastoma Syndrome (PEDONC-2.12)**.
  - Whole-body MRI has shown poor sensitivity and specificity in individuals with predisposition to systemic malignancy due to germline RB1 mutations, and is not supported.
- Detailed evaluation by a physician with significant training and/or experience in retinoblastoma (most commonly a pediatric ophthalmologist or pediatric oncologist) is indicated prior to considering advanced imaging.
- Retinoblastoma can be unilateral, bilateral, or trilateral (involving the pineal gland). Extraocular spread of retinoblastoma is rare and generally confined to the brain.

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Retinoblastoma – Imaging (PEDONC-12.2)

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Retinoblastoma Medically Necessary Initial Staging Imaging:

- Tumor biopsy is NOT required prior to imaging
- MRI Orbits (CPT® 70543) and Brain (CPT® 70553) without and with contrast in the initial work-up
  - Brain imaging may be omitted or deferred at the discretion of the treating ophthalmologist or oncologist
- MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) if there is evidence of CNS metastasis on:
  - Ophthalmologic exam
  - MRI Brain
  - Lumbar CSF cytology
- CT Chest (CPT® 71260) and MRI Abdomen and Pelvis without and with contrast (CPT® 74183 and CPT® 72197) with clinical symptoms to suggest metastatic disease
- CT Orbital (contrast as requested) and orbital ultrasound if ordered by the treating ophthalmologist for a specified indication
- Nuclear bone scan (See: **Modality General Considerations (PEDONC-1.3)**) for individuals with systemic bone pain suggestive of bony metastases
- PET has no documented role in the evaluation of retinoblastoma

Retinoblastoma Medically Necessary Treatment Response Imaging:

- MRI Orbits (CPT® 70543) and/or Brain (CPT® 70553) every 2 cycles (~ every 6 weeks) and at the end of planned therapy
- For individuals with metastatic disease, imaging of known positive areas using the same modality at initial staging every 2 cycles (~6 to 8 weeks) and at the end of planned therapy

Retinoblastoma Medically Necessary Surveillance Imaging:

- Unilateral retinoblastoma
  - Surveillance using advanced imaging for unilateral retinoblastoma after enucleation or exenteration only for evaluation of specific clinical concerns.
- Bilateral retinoblastoma or individuals treated with ocular salvage approach

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- MRI Orbits (CPT® 70543) and Brain (CPT® 70553) for individuals undergoing ocular salvage treatment approaches every 6 months for 2 years following completion of therapy or until the age of 5 years, whichever is later

Background and Supporting Information

- CT should generally be avoided in retinoblastoma individuals under one year of age or with family history of retinoblastoma (heritable) due to substantially increased risks for secondary malignancy
- The primary method of surveillance in retinoblastoma is examination under anesthesia (EUA), although some older children can be sufficiently evaluated by exam without anesthesia (EWA).
- Surveillance using advanced imaging is generally not indicated for unilateral retinoblastoma after enucleation or exenteration but can be approved for evaluation of specific clinical concerns.
- Individuals with bilateral retinoblastoma or germline mutation in RB1 are at increased risk for subsequent pineoblastoma
- Routine MRI follow up for pineal disease is not currently supported by evidence in unilateral retinoblastoma individuals without germline RB1 mutations

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Pediatric Nasopharyngeal Carcinoma (PEDONC-13)

Guideline

Pediatric Nasopharyngeal Carcinoma – General Considerations (PEDONC-13.1)
Pediatric NPC – Imaging (PEDONC-13.2)
References (PEDONC-13)

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Pediatric Nasopharyngeal Carcinoma – General Considerations (PEDONC-13.1)

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- Pediatric nasopharyngeal carcinoma (NPC) is rare in comparison to adult NPC but is responsible for up to 50% of nasopharyngeal cancers in children and has higher rates of aggressive type III EBV-associated histology than adult NPC.
- Standard upfront treatment in pediatric NPC consists of 3 to 4 cycles of neoadjuvant chemotherapy followed by definitive chemoradiotherapy. Rare individuals with lower stage disease may be treated with radiotherapy alone.

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Pediatric NPC – Imaging (PEDONC-13.2)

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Pediatric NPC Medically Necessary Initial Staging Imaging:

- MRI Brain without and with contrast (CPT® 70553) **and** MRI Neck without and with contrast (CPT® 70543)
  - CT Head without and with contrast (CPT® 70470), CT Maxillofacial without and with contrast (CPT® 70488), **and/or** CT Neck with contrast (CPT® 70491) for individuals with documented contraindication to MRI imaging (avoidance of sedation should not be the sole reason)
- CT Chest with contrast (CPT® 71260) in initial staging
- Whole-body PET/CT (CPT® 78816) after histologic confirmation of NPC to evaluate for distant metastases
  - Bone scan when PET/CT is unavailable (See: **Modality General Considerations (PEDONC-1.3)**)
  - Indications and coding for rare circumstances where PET/MRI is medically necessary are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
  - CT Abdomen with contrast (CPT® 74160) for ANY of the following if PET/CT unavailable:
    - Initial EBV DNA load ≥4000 copies/mL
    - Signs and symptoms of liver disease (including abdominal pain and elevated LFTs)

Pediatric NPC Medically Necessary Treatment Response Imaging:

- MRI Brain without and with contrast (CPT® 70553) **and** MRI Neck without and with contrast (CPT® 70543) for response assessment at the following time points:
  - Following completion of neoadjuvant chemotherapy
  - Following completion of chemoradiotherapy
- CT Chest with contrast (CPT® 71260) **and** Whole-Body PET/CT (CPT® 78816) **or** bone scan (See: **Modality General Considerations (PEDONC-1.3)**) at the following time points:
  - Following completion of neoadjuvant chemotherapy only if positive at initial diagnosis
  - Following completion of chemoradiotherapy
- PET during active treatment for recurrent pediatric cancer in rare circumstances when results are likely to result in a treatment change, including a change from active treatment to surveillance.

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Pediatric NPC Medically Necessary Surveillance Imaging:

- MRI Brain without and with contrast (CPT® 70553) **and** MRI Neck without and with contrast (CPT® 70543) every 3 months for 1 year, then every 6 months for 2 years after completion of all planned therapy
- CT Chest with contrast (CPT® 71260) every 3 months for 1 year, then every 6 months for 2 years after completion of all planned therapy

Pediatric NPC Medically Necessary Suspected Recurrence Imaging:

- MRI Brain without and with contrast (CPT® 70553) and MRI Neck without and with contrast (CPT® 70543)
  - CT Head without and with contrast (CPT® 70470), CT Maxillofacial without and with contrast (CPT® 70488), and/or CT Neck with contrast (CPT® 70491) for individuals with documented contraindication to MRI imaging
- CT Chest with contrast (CPT® 71260)
- Whole-body PET/CT (CPT® 78816) or bone scan (See: **Modality General Considerations (PEDONC-1.3)**) for histologically confirmed recurrence of NPC. These studies are also medically necessary for:
  - Clarification of specified inconclusive findings seen on conventional imaging
  - Restaging to identify sites of disease when EBV PCR levels are abnormally high and conventional imaging is negative

Background and Supporting Information

- Metastasis frequently occurs in cervical lymph nodes and retropharyngeal space. Distal metastasis usually appears in bones, lungs, mediastinum, and rarely, in the liver. In many individuals, the initial presentation is a cervical adenopathy, and diagnosis is made with a lymph node biopsy.
- Quantitative EBV DNA PCR measured at initial diagnosis, as it can serve as an effective tumor marker if elevated at initial diagnosis.
- Skull base invasion is common in pediatric NPC and has a dramatic impact on prognosis, and is more easily recognized on MRI imaging
- PET is generally not indicated during active treatment for recurrent pediatric cancer
  - Whole-body PET/CT (CPT® 78816) or bone scan (See: **Modality General Considerations (PEDONC-1.3)**) are not indicated for routine surveillance in asymptomatic individuals

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Pediatric Adrenocortical Carcinoma (PEDONC-14)

Guideline

Pediatric Adrenocortical Carcinoma – General Considerations (PEDONC-14.1)
Pediatric ACC – Imaging (PEDONC-14.2)
References (PEDONC-14)

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Pediatric Adrenocortical Carcinoma – General Considerations (PEDONC-14.1)

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- Pediatric Adrenocortical Carcinoma (ACC) is a rare but aggressive tumor, with fewer than 25 cases diagnosed each year.
- Most individuals are diagnosed because of virilizing symptoms, Cushing syndrome, and rarely with feminization and hyperaldosteronism or detection on screening imaging recommended for specified cancer predisposition syndromes.
- See: **Li-Fraumeni Syndrome (LFS) (PEDONC-2.2)** and **Beckwith-Wiedemann Syndrome (BWS) (PEDONC-2.4)** for screening recommendations for individuals known to have these syndromes.

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Pediatric ACC – Imaging (PEDONC-14.2)

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- CT Abdomen without and with contrast increases radiation exposure and should not be routinely performed in a child with an adrenal lesion as washout criteria have not been validated in children

Indication	Medically Necessary Imaging Study
Initial staging	• CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) • Nuclear bone scan (see: **(PEDONC-1.3)** for coding)
• Solitary adrenal mass >4 cm on conventional imaging and plans for aggressive surgical resection • Inconclusive findings on conventional imaging	• FDG PET/CT scan (CPT® 78815)
After complete resection, with no plans for chemotherapy or radiotherapy	• See surveillance below
Restaging, for all unresected primary or metastatic disease on chemotherapy	• Every 2 cycles (~6 weeks) during chemotherapy, and following completion of all planned chemotherapy: ◦ CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183) • If positive for distant metastases at initial diagnosis: ◦ Nuclear bone scan (see: **(PEDONC-1.3)** for coding) every 2 cycles (~6 weeks) during chemotherapy and following completion of all planned chemotherapy

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Indication	Medically Necessary Imaging Study
Response assessment at the completion of radiotherapy	• CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Surveillance, individuals with only localized disease at diagnosis	• Every 3 months for 2 years, then every 6 months for 3 years: ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Surveillance, individuals with metastatic ACC	• Every 3 months for 2 years, then every 6 months for 3 years: ◦ CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast ◦ CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Recurrence	• CT Chest without contrast (CPT® 71250) or with (CPT® 71260) contrast • CT Abdomen with contrast (CPT® 74160) or MRI Abdomen without and with contrast (CPT® 74183)
Suspected bone recurrence	• Nuclear bone scan (see: **(PEDONC-1.3)** for coding)

Background and Supporting Information

- The mainstay of treatment is surgery.
  - Chemotherapy, adrenal suppression, and radiotherapy typically follow resection.
  - Many ACC individuals are treated with surgery alone and do not receive adjuvant therapy. These individuals should be imaged using surveillance guidelines after surgery is completed.

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Pediatric Melanoma and Other Skin Cancers (PEDONC-15)

Guideline

Pediatric Melanoma and Other Skin Cancers (PEDONC-15)
References (PEDONC-15)

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Pediatric Melanoma and Other Skin Cancers (PEDONC-15)

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- Pediatric melanoma is historically rare but has a steadily rising incidence, especially in adolescents and young adults (AYAs). Staging is assigned using the American Joint Committee on Cancer (AJCC) staging for adult melanoma. Most cases of melanoma arising in children and AYAs (~75%) are localized at diagnosis, and approximately 90% of individuals with pediatric melanoma are amenable to radical excision. The clinical management of adolescents and young adults with melanoma is still challenging and evolving because it is difficult to diagnose, and there is no standard treatment.
- Non-melanoma skin cancers (mostly basal cell carcinoma and squamous cell carcinoma) are extremely rare in pediatric individuals. In many cases, predisposing factors such as prolonged immunosuppression, radiation therapy, chemotherapy, voriconazole use, or a combination of the factors are present, and established age-specific guidelines for management of these skin tumors do not exist.
- Imaging guidelines and treatment approaches are consistent with those used for adults with melanoma and other skin cancers, and these individuals should follow the imaging guidelines in section **Melanomas and Other Skin Cancers (ONC-5)** in the Oncology Imaging Guidelines.

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Pediatric Salivary Gland Tumors and Thyroid Tumors (PEDONC-16)

Guideline

Pediatric Salivary Gland Tumors and Thyroid Tumors (PEDONC-16)
References (PEDONC-16)

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Pediatric Salivary Gland Tumors and Thyroid Tumors (PEDONC-16)

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Pediatric Salivary Gland Tumors

- The majority of pediatric salivary gland tumors arise in the parotid gland. Approximately 10% to 15% of tumors arise in the submandibular, sublingual, or minor salivary glands.
- Roughly 75% of pediatric salivary gland tumors are benign, most commonly pleomorphic adenoma.
- The most common malignant tumors occurring in the salivary glands are mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, undifferentiated carcinoma, and rarely adenocarcinoma.
- American Joint Committee on Cancer (AJCC) staging is used for pediatric as well as adult salivary gland tumors.
- Imaging and treatment guidelines for malignant pediatric salivary gland tumors are consistent with those used for adults with salivary gland tumors, and these individuals should follow the imaging guidelines in section **Salivary Gland Cancers (ONC-4)** in the Oncology Imaging Guidelines.

Pediatric Thyroid Tumors

- Differentiated thyroid cancers (DTC): Papillary, Follicular and Hürthle Cell are the most common childhood thyroid malignancy. Standard treatment is thyroidectomy and radioactive iodine (RAI).
- Imaging and treatment guidelines for malignant pediatric thyroid tumors are consistent with those used for adults with thyroid tumors, and these individuals should follow the imaging guidelines in section **Thyroid Cancers (ONC-6)** in the Oncology Imaging Guidelines.

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Pediatric Breast Masses (PEDONC-17)

Guideline

Pediatric Breast Masses (PEDONC-17)
References (PEDONC-17)

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Pediatric Breast Masses (PEDONC-17)

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- Ultrasound (CPT® 76641 and CPT® 76642) is the primary and medically necessary modality used for evaluation of pediatric breast masses.
- MRI has very limited utility in evaluation of pediatric breast masses prior to biopsy but may be indicated in rare cases for surgical planning when ultrasound is non-diagnostic.
- Pediatric individuals with confirmed breast cancer should be imaged according to section **Breast Cancer (ONC-11)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Less than 1% of pediatric breast lesions are malignant, and advanced imaging is generally not recommended without histological confirmation of malignancy.
- Mammography has limited utility in pediatric breast mass evaluation due to the high mammographic breast density in this age group, and the risk of the radiation exposure outweighs the benefit of this modality. As a result, mammography is NOT recommended for evaluation of pediatric or adolescent breast masses.
  - BI-RADS classification may overstate the risk of malignancy or need for biopsy in pediatric individuals.

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Histiocytic Disorders (PEDONC-18)

Guideline

Histiocytic Disorders – General Considerations (PEDONC-18.1)
Langerhans Cell Histiocytosis (LCH) (PEDONC-18.2)
Hemophagocytic Lymphohistiocytosis (HLH) (PEDONC-18.3)
Non-Langerhans Cell Histiocytoses (PEDONC-18.4)
References (PEDONC-18)

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Histiocytic Disorders – General Considerations (PEDONC-18.1)

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- The majority of histiocytic disorders occurring in the pediatric population are either Langerhans Cell Histiocytosis (LCH) or Hemophagocytic Lymphohistiocytosis (HLH).
- The Non-Langerhans cell histiocytoses encompass a variety of diseases and have limited imaging considerations except as specified later in this section.
- Where there are indications for PET/CT in these guidelines, please note that indications and coding for rare circumstances where PET/MRI may be approved are found in **PET Imaging in Pediatric Oncology (PEDONC-1.4)**
- PEDONC-18 applies to individuals of all ages.
- The use of PET in this guideline refers to Fluorodeoxyglucose (fluorine-18-2-fluoro-2-deoxy-D-glucose [FDG]) radiotracer only.

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Langerhans Cell Histiocytosis (LCH) (PEDONC-18.2)

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LCH Medically Necessary Initial Imaging:

- Whole-body PET/CT (CPT® 78816)
  - Whole-body Tc-99m bone scan (CPT® 78306) if PET is unavailable
- CT Chest with contrast (CPT® 71260) **or** high-resolution CT Chest without contrast (CPT® 71250) in addition to PET/CT for suspected pulmonary LCH based on ANY of the following:
  - Abnormal CXR
  - Symptoms of pulmonary involvement and normal CXR
  - Clarification of pulmonary findings on PET/CT
- MRI Brain without and with contrast (CPT® 70553) for ANY of the following:
  - Headaches or visual or neurologic disturbances
  - Polyuria/polydipsia or other endocrine abnormalities
  - Skull or craniofacial (including jaw) bone involvement
  - Otorrhea or hearing loss (CT Temporal Bone may be substituted if requested)
  - Other signs or symptoms suggesting intracranial involvement, including neurodegeneration syndrome
- CTA/MRA Head (CPT® 70496/70544) for operative planning or image guidance
- CT Abdomen and/or Pelvis with contrast (CPT® 74177, 74160, or 72193) for ANY of the following:
  - Abdominal and/or pelvic signs and symptoms **if PET/CT has not been performed** or to clarify abnormal abdominal/pelvic findings on PET/CT
  - Abdominal and/or pelvic findings on ultrasound if PET/CT has not been performed
- MRI Abdomen without and with contrast (CPT® 74183) for any of the following:
  - Elevated liver function tests (usually >5X upper limit of normal)
  - Abnormalities seen on abdominal ultrasound or CT
- MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) for ANY of the following **if PET/CT has not been performed**:
  - Vertebral lesions seen on skeletal survey
  - Clinical symptoms (including back pain) suggesting spinal involvement and negative skeletal survey

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LCH Medically Necessary Treatment Response Imaging:

- Both PET/CT and CT with contrast or MRI without and with contrast **only** for simultaneous treatment response evaluation with specific documentation showing that both are necessary (i.e., not for purpose of acquiring a PET/MRI)
- CT and/or MRI and/or PET/CT (if modality showed disease at initial diagnosis):
  - After 2-3 cycles of treatment
  - At completion of therapy (approximately 12 weeks) for individuals with persistent disease on cycle 2-3 imaging
  - After surgical curettage (CT or MRI of involved area, not repeat PET)
  - After radiation therapy
- Following the initial phase, treatment response evaluation of **involved sites** with CT with contrast or MRI with and without contrast, every ~3 months while receiving active treatment and at completion of therapy.
  - PET/CT for inconclusive conventional imaging, if disease was previously only measured/measurable on PET/CT, or for change from active treatment to surveillance
  - Shorter interval imaging for documented signs or symptoms concerning for disease progression

LCH Medically Necessary Surveillance Imaging:

- Surveillance imaging is determined by areas of disease involvement.
  - Bone involvement
    - Single site bone disease
      - Every 3 months for 1 year after completion of therapy:
        - PET/CT (CPT® 78815 or CPT® 78816)

          OR
        - CT or MRI contrast as requested of involved bony areas
    - Multifocal bone disease
      - Every 3 months for 2 years, then no more than annually:
        - PET/CT (CPT® 78815 or CPT® 78816)

          OR
        - CT or MRI contrast as requested of involved bony areas
    - Skull or craniofacial (including jaw) bone involvement should be imaged according to CNS involvement section below.

  Individuals with lung, CNS, and/or liver involvement are considered to have high-risk disease and warrant site-specific diagnostic imaging in addition to PET/CT as outlined below:
- Pulmonary involvement

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- CT Chest with (CPT® 71260) **or** without contrast (CPT® 71250) every 6 months for the first 2 years post completion of therapy for any of the following:
  - Individuals with a history of pulmonary involvement
  - Individuals with new respiratory or chest symptoms
  - New findings on CXR

  AND
- PET/CT (CPT® 78815 or CPT® 78816) every 6 months for 2 years, then no more than annually

- CNS involvement
  - PET/CT (CPT® 78815 or CPT® 78816) every 6 months for 2 years, then no more than annually

    AND
  - MRI Brain without and with contrast (CPT® 70553) at 6 weeks, and then every 6 months for 2 years after completion of all therapy for previously documented measurable intracranial lesions
    - If negative after two years, MRI Brain without and with contrast (CPT® 70553) at 4, 7, and 10 years after completion of all planned therapy
    - If residual measurable intracranial lesions are present at 6 months, imaging repeated every 3 months until negative or unchanged on two consecutive studies, at which time the schedule in the previous bullet should begin
  - MRI Brain without and with contrast (CPT® 70553) for documented hypothalamic-pituitary dysfunction every 6 months for 2 years and at 4, 7, and 10 years after completion of all planned therapy
    - MRI at any time for worsening neurologic symptoms
  - Intraspinal lesions imaged according to the same timeframes as brain imaging using MRI without and with contrast of all involved spine levels
- Liver involvement
  - For individuals with a history of liver involvement, ONE of the following every 6 months for 2 years after completion of all therapy:
    - Ultrasound Abdomen (CPT® 76700)
    - CT Abdomen with contrast (CPT® 74160)
    - MRI Abdomen without and with contrast (CPT® 74183)

    AND
  - PET/CT (CPT® 78815 or CPT® 78816) every 6 months for 2 years, then no more than annually
- Suspected recurrence or inconclusive findings on any surveillance imaging:
  - All imaging studies supported for initial workup including whole-body PET/CT (CPT® 78816)

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Background and Supporting Information

- This guideline may be used for all ages of individuals.
- LCH includes a heterogeneous group of disorders formerly known by other names, including histiocytosis X, eosinophilic granuloma, Letterer-Siwe Disease, Hand-Schuller-Christian Disease, and diffuse reticuloendotheliosis. LCH has a widely variable clinical presentation, ranging from single indolent lesions to disseminated multisystem disease.
- Most common sites of involvement are skin, bones, liver, lung, and pituitary, though other sites are possible.
- Individuals with localized or single site disease are often treated only with local therapies or observed, and should be imaged according to surveillance guidelines
- Individuals receiving systemic therapy will usually undergo treatment for ~12 months. Treatment response is assessed using any modalities showing disease at initial diagnosis after ~6 weeks of treatment.
- Skull or craniofacial (including jaw) bone involvement at diagnosis are at higher risk for CNS recurrence
- CNS LCH has a particularly high rate of refractory and recurrent disease, and requires longer imaging surveillance
- Liver involvement
  - Persistent liver involvement is rare, and imaging after completion of LCH therapy will be highly individualized depending on degree of liver dysfunction and plans for supportive therapy or liver transplant

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Hemophagocytic Lymphohistiocytosis (HLH) (PEDONC-18.3)

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- As imaging for this condition is usually done on an urgent basis, ANY or ALL of the following are medically necessary for the initial evaluation of HLH include:
  - Ultrasound Abdomen (CPT® 76700)
  - CT Abdomen and/or Pelvis (contrast as requested)
  - MRI Abdomen (CPT® 74183) and/or Pelvis (CPT® 72197) without and with contrast
  - Chest x-ray
  - CT Chest with contrast (CPT® 71260)
  - MRI Brain without and with contrast (CPT® 70553)
  - CTA/MRA Head (CPT® 70496/CPT® 70544) for operative planning or image guidance
  - CT Sinus without or with contrast (CPT® 70486 or CPT® 70487) if clinical suspicion for sinus disease
- Whole-body PET/CT (CPT® 78816) for the purpose of identifying a site for tissue diagnosis of a primary source of infection or malignancy if conventional imaging has been completed and is unrevealing
  - If a malignancy is identified as the inciting factor for HLH, additional imaging decisions for that malignancy should be based on the appropriate diagnosis-specific guidelines

Background and Supporting Information

- There are no standard imaging studies required for the diagnosis and initial evaluation of HLH. Most cases are diagnosed with a combination of physical findings, laboratory testing, and bone marrow evaluation. Advanced imaging studies may be necessary to assess organ dysfunction as HLH commonly affects the liver, spleen, and bone marrow, and less commonly the kidneys, lungs, and brain
- It is NOT required to perform ultrasound or plain film in a stepwise fashion if CT or MRI is planned as individuals with HLH can deteriorate rapidly
- There is no established standard role for PET in the diagnosis or treatment response evaluation of HLH
  - Secondary HLH is very difficult to treat if the primary cause is not concurrently treated

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Non-Langerhans Cell Histiocytoses (PEDONC-18.4)

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Juvenile Xanthogranuloma (JXG) medically necessary imaging:

- Skin and/or cervical nodes:
  - CT with contrast of appropriate area
- Systemic JXG with multi-organ involvement:
  - MRI Brain (CPT® 70553) **and/or** Orbits (CPT® 70543) without and with contrast
  - CT Neck (CPT® 70491), Chest (CPT® 71260), **and/or** Abdomen (CPT® 74160) with contrast
  - CTA/MRA Head (CPT® 70496/CPT® 70544) may be approved as part of operative planning or image guidance
- There is no established role for PET in the diagnosis or treatment of JXG

Rosai-Dorfman Disease (RDD) medically necessary imaging:

- RDD Initial Imaging Studies:
  - MRI Brain (CPT® 70553) and/or Orbits (CPT® 70543) without and with contrast
  - CTA/MRA Head (CPT® 70496/CPT® 70544) for operative planning or image guidance
  - CT Neck (CPT® 70491), Chest (CPT® 71260) and/or Abdomen/Pelvis (CPT® 74177) with contrast **if PET/CT not performed** or to follow up unclear findings in said body areas on PET/CT
  - CT Sinus without or with contrast (CPT® 70486 or CPT® 70487) if clinical suspicion for sinus disease and **PET/CT not performed** or to follow up unclear sinus findings on PET/CT
  - Whole-body PET/CT (CPT® 78816) after histologic confirmation of diagnosis
- RDD Treatment Response:
  - Treatment response imaging after 2-3 cycles of systemic therapy during active treatment using any modalities showing disease at diagnosis, including PET/CT.
    - Once PET/CT is negative, conventional imaging with other modalities that revealed disease at presentation for subsequent restaging at completion of chemotherapy and/or radiation and/or after surgical resection.
- RDD Surveillance Imaging:
  - PET/CT (CPT® 78816) every 3 months until stabilization of disease (two PET/CT with stable disease status).

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    - Further surveillance imaging of single-site bone disease:
        - Every 6 months using any modalities showing disease at initial diagnosis (excluding PET/CT) for 1 year
        - PET/CT is not medically necessary for routine surveillance of RDD but is medically necessary if conventional imaging is inconclusive for suspected recurrence.
    - Further surveillance imaging of multifocal bone disease:
        - Every 6 months using any modalities showing disease at initial diagnosis (excluding PET/CT) thereafter
        - PET/CT is not medically necessary for routine surveillance of RDD but is medically necessary if conventional imaging is inconclusive for suspected recurrence.

Erdheim-Chester Disease (ECD) medically necessary imaging:

- ECD Initial Imaging Studies:
    - PET/CT Whole Body (CPT® 78816)
        - Nuclear bone scan (See: **PEDONC-1.3: Modality General Considerations)** in lieu of PET if requested
    - MRI Brain without and with contrast (CPT® 70553) **and/or** MRI Orbits (CPT® 70543) without and with contrast for CNS symptoms, including diabetes insipidus
    - CTA/MRA Head (CPT® 70496/70544) for operative planning or image guidance
    - CT Neck (CPT® 70491), Chest (CPT® 71260) **and/or** Abdomen/Pelvis (CPT® 74177) with contrast **if PET/CT not performed** or if inconclusive findings in said body area on PET/CT
    - Cardiac MRI without and with contrast (CPT® 75561) for clinically suspected cardiac involvement
    - CT Sinus without or with contrast (CPT® 70486 or CPT® 70487) if clinical suspicion for sinus disease **if PET/CT not performed** or inconclusive sinus findings on PET/CT
- ECD Treatment Response:
    - Treatment response imaging every 3 months during active treatment using any modalities showing disease at initial diagnosis, including PET/CT
    - Once PET/CT shows no remaining FDG-avid lesions, additional PET imaging is only when conventional imaging studies are inconclusive and acute treatment decisions will be made based on PET results.
- ECD Surveillance Imaging:
    - CT **and/or** MRI **and/or** Nuclear bone scan **and/or** CTA **and/or** MRA **and/or** Cardiac MRI (if modality showed disease at initial diagnosis) every 3 months until the first year after completion of treatment and then every 6 months
    - PET/CT if conventional imaging is inconclusive for suspected recurrence.

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Background and Supporting Information

- Non-Langerhans Cell histiocytoses includes diagnoses such as juvenile xanthogranuloma (JXG), sinus histiocytosis with lymphadenopathy (Rosai-Dorfman Disease, RDD), and Erdheim-Chester Disease (ECD)
- In general, these are localized cutaneous or nodal disease without need for regular advanced imaging, but important exceptions are listed in this section.
- Juvenile Xanthogranuloma (JXG):
  - Generally involves only skin or cervical nodes, and involutes spontaneously
- Rosai-Dorfman Disease (RDD):
  - Characterized by bulky adenopathy (usually cervical) with frequent systemic involvement
  - There is no established role for routine surveillance imaging of asymptomatic individuals after treatment for RDD
- Erdheim-Chester Disease (ECD):
  - An aggressive histiocytic disorder with overall poor prognosis that is characterized by long bone involvement with frequent spread to multiple organs
  - Most individuals will receive systemic therapy
  - Once PET/CT shows no remaining FDG-avid lesions, additional PET imaging is not indicated unless conventional imaging studies are inconclusive and acute treatment decisions will be made based on PET results.

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Long Term Pediatric Cancer Survivors (PEDONC-19)

Guideline

Long Term Pediatric Cancer Survivors – General Considerations (PEDONC-19.1)
Cardiotoxicity and Echocardiography (PEDONC-19.2)
Second Malignant Neoplasms (SMN) (PEDONC-19.3)
Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)
CNS vascular changes in pediatric cancer survivors following CNS radiation (PEDONC-19.5)
References (PEDONC-19)

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Long Term Pediatric Cancer Survivors – General Considerations (PEDONC-19.1)

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- This section applies to individuals who have passed the end of the surveillance imaging period for their specific cancer, or 5 years after completion of therapy, whichever occurs first.
- As these are long-term survivors, many individuals falling under this guideline section will have reached adult age. However, these guidelines relate specifically to late effects of childhood cancer treatment and should be applied to all long-term childhood cancer survivors regardless of current age.
- The Children's Oncology Group has published comprehensive guidelines for the management of long-term childhood cancer survivors, and these are available at: http://www.survivorshipguidelines.org.
- A summary of cancer treatment should be available for all individuals in this category and should generally include, at minimum:
  - type of cancer and stage
  - dates of diagnosis, recurrence, cancer-related surgeries, beginning and end dates of chemotherapy, radiotherapy, and/or stem cell transplant
  - protocol number used for treatment and cumulative chemotherapy drug dose exposures
  - cumulative radiation dose, fraction number, modality, and field exposure
- Annual detailed history and complete physical examination is a critical component of cancer survivorship care and along with laboratory testing serves as the primary method of screening for the majority of late effects.
- Advanced imaging for asymptomatic screening is not routinely indicated except as specified in this section.
- Imaging requests related to new clinical signs or symptoms in a long term cancer survivor not explicitly covered in this section should be reviewed according to the guideline for the individual's cancer type or the relevant non-malignant clinical problem.

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Cardiotoxicity and Echocardiography (PEDONC-19.2)

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- Screening echocardiography (CPT® 93306, CPT® 93307, or CPT® 93308) for life after exposure to anthracycline chemotherapy, cardiotoxic immunotherapy, or cardiac exposure to radiotherapy

MEDICALLY NECESSARY SCREENING ECHOCARDIOGRAM INDICATIONS			
Age at Time of Exposure	**Cumulative Doxorubicin Equivalent Dose**	**Cumulative Radiation Dose to Cardiac Muscle**	**Echocardiogram Frequency**
All ages	None	None	None
0-0.99 years	≥250 mg/m2	None	Annual
	0-249 mg/m2	Any dose	Annual
	0-249 mg/m2	None	Every 2 years
1-4.99 years	≥250 mg/m2	Any dose	Annual
	0-249 mg/m2	15+ Gy	Annual
		0-14.99 Gy	Every 2 years
	None	35+ Gy	Annual
		15-34.99 Gy	Every 2 years
		0-14.99 Gy	Every 5 years
5+ years	≥250 mg/m2	Any dose	Every 2 years
	0-249 mg/m2	15+ Gy	Every 2 years

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MEDICALLY NECESSARY SCREENING ECHOCARDIOGRAM INDICATIONS			
Age at Time of Exposure	**Cumulative Doxorubicin Equivalent Dose**	**Cumulative Radiation Dose to Cardiac Muscle**	**Echocardiogram Frequency**
		0-14.99 Gy	Every 5 years
	None	35+ Gy	Every 2 years
		15-34.99 Gy	Every 5 years
		0-14.99 Gy	None
All ages with known ventricular dysfunction			Annual

- Stress echocardiography is not medically necessary as a screening study for anthracyclines cardiotoxicity in the absence of coronary artery disease symptoms. See: **Stress Testing with Imaging – Indications (CD-1.4)** for imaging guidelines.
- Female cancer survivors who are pregnant or planning to become pregnant:
  - If any of the following are present, echocardiogram is medically necessary as a baseline exam and in the 3rd trimester, and as clinically indicated for symptoms (see: **Pregnancy – Maternal Imaging (CD-11.4)** in the Cardiology Imaging Guidelines) if ANY of the following are present:
    - Anthracycline or cardiotoxic chemotherapy/immunotherapy exposure
    - Chest radiotherapy

Background and Supporting Information

- Exposure to cardiotoxic anthracycline chemotherapy agents is common in pediatric oncology due to the high success rate of this drug class in the treatment of pediatric cancers.
- Cardiac risk is assessed based on the age of the individual at the time of treatment initiation, the cumulative drug exposure expressed as doxorubicin equivalent mg/m^2, and the presence or absence of radiotherapy exposure to cardiac muscle.

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Second Malignant Neoplasms (SMN) (PEDONC-19.3)

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SMN—Breast Cancer

Clinical breast exam every 6 months supplemented with the following medically necessary imaging:

- MRI Breast (CPT® 77049) annually and annual mammogram beginning at age 25 or 8 years after completion of radiotherapy (whichever occurs later- screening breast MRI is not supported prior to age 25) for individuals who received therapeutic radiation exposure in the following fields while they were under 30 years of age:
  - Chest (thorax)
  - Whole lung
  - Mediastinal
  - Axilla
  - Mini-mantle, mantle, or extended mantle
  - Total (TLI) or subtotal (SLTI) lymphoid irradiation
  - Total body irradiation (TBI)

SMN – CNS Tumors

- Routine surveillance of asymptomatic individuals with normal neurologic exams is not supported by evidence and is therefore not medically necessary, with the exception of NF1 and NF2 below:
  - MRI Brain without and with contrast (CPT® 70553) is medically necessary every 2 years after completion of radiotherapy for individuals with NF1 or NF2.
- MRI Brain without and with contrast (CPT® 70553) is medically necessary for any individual with history of brain radiotherapy and new neurologic symptoms, including simple headache.
- MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar Spine (CPT® 72158) without and with contrast are medically necessary for any individual with history of spine radiotherapy and new neurologic symptoms including change in quality of pain.
  - MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) is medically necessary if being performed immediately following a contrast-enhanced MRI Brain.

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- MRI Brain without and with contrast (CPT® 70553) is medically necessary annually for individuals with history of brain radiotherapy and persistent neurologic symptoms.
- MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar Spine (CPT® 72158) without and with contrast is medically necessary annually for individuals with history of spine radiotherapy and persistent neurologic symptoms.
  - MRI Spine with contrast only (Cervical-CPT® 72142, Thoracic-CPT® 72147, Lumbar-CPT® 72149) is medically necessary if being performed immediately following a contrast-enhanced MRI Brain.

SMN—Colorectal Cancer

- Advanced imaging is not medically necessary. See: *Background and Supporting Information* section for recommended surveillance.

Background and Supporting Information

- SMN—Breast Cancer:
  - Clinical breast exam every 6 months supplemented with advanced imaging
- SMN-CNS Tumors:
  - These are associated with radiation exposure to the brain and with neurofibromatosis
  - Routine surveillance of asymptomatic individuals with normal neurologic exams is not supported by evidence, with the exception of NF1 and NF2 listed above
- SMN—Colorectal Cancer:
  - Colonoscopy is recommended every 5 years beginning at age 30 or 5 years after radiation exposure (whichever is later) for individuals with ≥30 Gy radiation exposure to the following fields:
    - Thoracic, Lumbar, Sacral, or Whole Spine
    - Abdomen
    - Pelvis
    - Total body irradiation (TBI)
- Colonoscopy is also recommended every 5 years beginning at age 30 or 5 years after radiation exposure (whichever is later) for individuals with:
  - Personal history of ulcerative colitis, GI malignancy, adenomatous polyps, or hepatoblastoma
  - Familial polyposis
    - Family history of colorectal cancer or polyps in a first degree (parent or sibling) relative
- While the American Cancer Society recently added computed tomographic colonography (CTC) (AKA “Virtual Colonoscopy”) as an acceptable option for colorectal cancer screening of average-risk adults, the National Comprehensive Cancer Network and United States Preventive Services Task Force concluded that

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data was too premature to warrant its use in screening. Colonoscopy remains the preferred screening modality for survivors at highest risk of colorectal cancer

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Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)

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Osteonecrosis is associated with corticosteroid, chemotherapy, and radiation exposure during treatment for ALL, NHL, and allogeneic HSCT in pediatrics. Osteonecrosis occurs primarily in hips, knees, and ankles and is frequently multifocal.

Osteoradionecrosis of the jaw can occur in individuals receiving radiotherapy to the mandible or maxilla; those receiving ≥40 Gy are at highest risk. Although unusual, it can also occur in any bone without symptoms. It is rare in other disease types.

- Plain films of symptomatic areas are indicated prior to advanced imaging.
- Routine bone density screening using DEXA or Quantitative CT screening has not been well normalized in the pediatric population, but imaging is medically necessary for those with symptoms to suggest bone density issues
  - DEXA or Quantitative CT screening is not medically necessary until age 18 unless a surgery, core decompression, or initiation of osteoporosis drugs will be planned based on the imaging results.
- Serial advanced imaging is not medically necessary in osteonecrosis without specific documentation regarding how the advanced imaging will change current individual management.
  - When advanced imaging is necessary for acute management decisions, MRI without contrast of the affected area(s) is medically necessary.
  - Surveillance imaging of asymptomatic individuals to detect osteonecrosis is not medically necessary as it has not been shown to impact individual outcomes, and it is not standard to alter treatment based on imaging findings alone without symptoms.
    - Follow-up MRI of incidentally discovered osteonecrosis findings in asymptomatic individuals has not been shown to impact individual outcomes and is not medically necessary.
    - For known osteonecrosis with articular collapse on other imaging, CT without contrast of area of interest is medically necessary for surgical planning.
- See: **Acute Lymphoblastic Leukemia (ALL) (PEDONC-3.2)** for information on imaging osteonecrosis in ALL during active treatment.

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CNS vascular changes in pediatric cancer survivors following CNS radiation (PEDONC-19.5)

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Children receiving cranial radiation are at increased risk of cerebrovascular complications, including: hemorrhagic and ischemic stroke, moyamoya, occlusive vasculopathy, and cavernomas. These complications may occur months to years after radiation exposure.

- MRI Brain without and with contrast (CPT® 70553) and MRA Head (CPT® 70544, 70545, or 70546) are medically necessary for all individuals of any age with new neurologic symptoms or headache and a history of cranial irradiation.
- MRI Brain without and with contrast (CPT® 70553) and MRA Head (CPT® 70544, 70545, or 70546) are medically necessary annually for 10 years post treatment in individuals with a history of cranial irradiation and any of the following additional risk factors:
  - Down syndrome, sickle cell disease, or neurofibromatosis 1 or 2
  - Parasellar or suprasellar tumors (i.e., craniopharyngioma)
  - Radiation dose >50 Gy
  - Radiation field involving Circle of Willis

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Hematopoietic Stem Cell Transplantation (HSCT) (PEDONC-20)

Guideline

General Considerations (PEDONC-20.0)
Pre-Transplant Imaging (PEDONC-20.1)
Post-Transplant Imaging (PEDONC-20.2)
References (PEDONC-20)

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General Considerations (PEDONC-20.0)

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Transplant Types:

- Allogeneic (“allo”): The donor and recipient are different people, and there are multiple types depending on the source of the stem cells and degree of match between donor and recipient. This is most commonly used in diseases originating in the hematopoietic system, such as leukemias and lymphomas, and bone marrow failure syndromes or metabolic disorders. **The goal is to replace the hematopoietic and immune system with healthy donor cells to treat the disease.** Common types are:
  - Matched sibling donor (MSD or MRD): Donor and recipient are full siblings and HLA-matched
  - Matched unrelated donor (MUD): Donor and recipient are HLA matched but not related to each other
  - Cord blood: Donor stem cells come from frozen umbilical cord blood not related to the recipient, sometimes from multiple different donors at once
  - Haploidentical transplant (haplo): Donor is a half-HLA match to the recipient, usually a parent
- Autologous (“auto”): The donor and recipient are the same person. The process involves delivery of high dose chemotherapy that is ablative to the bone marrow, followed by an infusion of one’s own harvested stem cells. This is primarily done in the context of solid tumors and the stem cells mainly rescue hematopoiesis to facilitate high-dose chemotherapy.
- Allogeneic HSCT results in a much greater degree of immunosuppression than autologous HSCT because of the need to allow the new immune system to chimerize with the recipient’s body. Immune reconstitution commonly takes more than a year for individuals who receive allogeneic HSCT, and individuals remain at high risk for invasive infections until that has occurred. In addition, patients may require prolonged immunosuppression for prevention and management of graft-vs-host disease.
- Recipients of autologous transplant are deeply immunosuppressed until complete count recovery, which may take several months. Graft-vs-host disease is not a consideration in auto transplants because the host is also the donor.

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Pre-Transplant Imaging (PEDONC-20.1)

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- Disease status assessment may be approved per individual disease guidelines if it has been 6 weeks or more between end of treatment imaging and HSCT or if there are signs and symptoms of disease progression.

Indication	Medically Necessary Imaging
Immediate pre-transplant period - screening for active or occult infection	• CT Chest with contrast or CT Chest without contrast (CPT® 71250 or CPT® 71260) ◦ CT Abdomen with contrast (CPT® 74160)for asymptomatic individuals if chest infection or disease is identified on CT Chest or for signs and symptoms of active abdominal infection including fever. ◦ CT Pelvis for pelvic signs and symptoms with CPT® 74177 or CPT® 72193 if abdominal imaging has already been performed. • CT Sinus (CPT® 70486) is not medically necessary for screening prior to HSCT, but is medically necessary for signs or symptoms of infectious or disease involvement of the sinuses
Individuals at risk of developing VOD, for pre-transplant baseline	• Abdominal ultrasound +/- Doppler (CPT® 76700 or 76705 +/- 93975)
Individuals with history of multiple blood transfusions at risk of iron overload, for iron quantification baseline	• CPT® codes as listed in **Transfusion-Associated (Secondary) Hemochromatosis (PEDAB-18.2)** in the immediate pre-transplant period even if done in the previous 12 months.
Suspected renal tubular dysfunction	• Nuclear renal imaging (CPT® 78700, CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709)

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Background and Supporting Information

- CT Sinus screening is not routinely recommended as two studies showed no utility and no effect on surgical consults.
- Changes from baseline abdominal ultrasound with regard to veno-occlusive disease (VOD) are more predictive than absolute measurements and thus baseline imaging is supported.
- Pre-transplant liver and cardiac iron levels are associated with adverse transplant outcomes and a baseline is supported for further management.

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Post-Transplant Imaging (PEDONC-20.2)

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Early post-transplant complication (<100 days post HSCT):

Indication	Medically Necessary Imaging
For signs and symptoms of pulmonary infection or pulmonary edema or new CXR abnormalities	• CT Chest without or with contrast (CPT® 71250 or CPT® 71260) ◦ For PET/CT requests to clarify infection vs malignancy, see disease-specific guidelines for equivocal conventional imaging
Suspected impending lung necrosis on other imaging	• MRI Chest without and with contrast (CPT® 71552)
• EITHER of the following: ◦ Suspected graft vs. host disease (GVHD) of chest ◦ Bronchiolitis Obliterans (BOOP/BOS)	• High-resolution CT Chest without contrast (CPT® 71250)
• ANY of the following: ◦ Suspected intra-abdominal and or pelvic infection (including cystitis or typhlitis) ◦ Suspected small bowel GVHD	• Abdominal Ultrasound (CPT® 76700 or CPT® 76705) +/- Pelvic Ultrasound (CPT® 76856 or CPT® 76857) ◦ CT Abdomen and Pelvis with contrast (CPT® 74177) for unclear findings on ultrasound
High clinical suspicion for intra-abdominal fungal infection	• CT Abdomen and Pelvis with contrast (CPT® 74177)
Suspected hepatic veno-occlusive disease (VOD)	• Ultrasound elastography (CPT® 91200) or Abdominal Ultrasound with Doppler (CPT® 76700 or CPT® 76705)

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Indication	Medically Necessary Imaging
Suspected CNS complication including infection, hemorrhage, thrombosis, or encephalopathy	• MRI Brain without contrast or MRI Brain without and with contrast (CPT® 70551 or CPT® 70553) ◦ CT Head without contrast (CPT® 70450) in the emergent setting or for suspected acute hemorrhage ▪ Acute CT should not preclude subsequent MRI imaging ◦ For additional imaging for suspected stroke, see: **Pediatric Stroke Initial Imaging (PEDHD-12.2)** in the Pediatric Head Imaging guidelines
Suspected musculoskeletal abscess or necrotizing fasciitis	• See: **Pediatric Infection/Osteomyelitis (PEDMS-8)** in the Pediatric Musculoskeletal Imaging guidelines
Fever of unknown origin (8 or more days of temperature 38.0 C/100.4 F or higher)	• PET/CT skull to thigh (CPT® 78815) if site-specific conventional imaging, microbiologic serologic studies, echocardiogram, urinalysis, and urine culture are all non-diagnostic

Late post-transplant complication imaging (100 or more days post HSCT):

Indication	Medically Necessary Imaging
CNS complications	Same imaging as for early post-transplant
Chest complications	Same imaging as for early post-transplant
Hepatic VOD	Same imaging as for early post-transplant
Chronic GI GVHD	• CT Abdomen and Pelvis (CPT® 74177) ◦ MR Enterography (CPT® 74183 and CPT® 72197) for suspected small bowel GVHD
Suspected renal tubular dysfunction	• Nuclear renal imaging (CPT® 78700, CPT® 78701, CPT® 78707, CPT® 78708, CPT® 78709)

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Indication	Medically Necessary Imaging
Suspected poor bone mineral density or osteonecrosis	See: **Osteonecrosis in Long Term Cancer Survivors (PEDONC-19.4)**
Suspected PTLD	See: **Post-Transplant Lymphoproliferative Disorder (PTLD) (PEDONC 5.3)**

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General Guidelines (PEDPV-1)

Guideline

Procedure Codes Associated with Pelvis Imaging
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Procedure Codes Associated with Pelvis Imaging

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General Guidelines (PEDPV-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms, including a detailed history, physical examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the pelvis is not supported. Advanced imaging of the pelvis is only medically necessary in individuals who have documented active clinical signs or symptoms of disease involving the pelvis.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the pelvis are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.
- Ultrasound
  - Ultrasound should be the initial imaging in most pelvic conditions to rule out those situations that do not require additional advanced imaging.
  - For those individuals who do require advanced imaging after ultrasound, ultrasound can be very beneficial in selecting the proper modality, body area, image sequences, and contrast level that will provide the most definitive information for the individual.
  - CPT® codes vary by body area and presence or absence of Doppler imaging and are included in the table at the beginning of this guideline.
  - Transabdominal ultrasound is appropriate in all pediatric individuals.
  - Transvaginal (TV) ultrasound is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Ultrasound (complete CPT® 76856 or, limited CPT® 76857) should substitute for TV in pediatric individuals or non-sexually active adult females

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following:

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safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Pediatric Pelvis Imaging Age Considerations (PEDPV-1.1)

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Many conditions affecting the pelvis in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.

- Individuals who are age 18 years or younger should be imaged according to the Pediatric Pelvis Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Pelvis Imaging Guidelines should be imaged according to the General Pelvis Imaging Guidelines. Individuals who are >19 years should be imaged according to the General Pelvis Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Pediatric Pelvis Imaging Modality General Considerations (PEDPV-1.3)

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- Ultrasound
  - See **General Guidelines (PEDPV-1.0)**
- MRI
  - MRI Pelvis is generally performed without and with contrast (CPT® 72197) unless the individual has a documented contraindication to gadolinium or otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize individual movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition-based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported by the guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
  - The presence of surgical hardware or implanted devices may preclude MRI.
  - The selection of best examination may require coordination between the provider and the imaging service.

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- CT
  - CT Pelvis typically extends from the iliac crest to the ischial tuberosities, and CT Abdomen and Pelvis extends from the dome of the diaphragm through the ischial tuberosities.
    - In general, CT Pelvis is appropriate when evaluating solid pelvic organs.
    - In general, CT Abdomen and Pelvis is appropriate when evaluating inflammatory or infections processes, hematuria, or conditions which appear to involve both the abdomen and the pelvis.
    - In some cases, especially in follow-up of a known finding, it may be appropriate to limit the exam to the region of concern to reduce radiation exposure.
  - The contrast level in pediatric CT imaging is specific to the clinical indication, as listed in the specific guideline sections.
  - CT Pelvis or Abdomen and Pelvis may be medically necessary for further evaluation of abnormalities suggested on prior US or MRI Procedures.
  - CT may be appropriate without prior MRI or US, as indicated in specific sections of these guidelines.
  - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
  - The selection of best examination may require coordination between the provider and the imaging service.
- Nuclear Medicine
  - Nuclear medicine studies are rarely used in imaging of the pediatric pelvis but are medically necessary in rare circumstances, including the following:
    - Lymph system mapping (CPT® 78195) is medically necessary for lower extremity lymphedema with recent negative Doppler ultrasound, or a history of Milroy's disease or prior pelvic lymph node dissection.
- 3D Rendering
  - 3D Rendering indications in pediatric pelvis imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Abnormal Uterine Bleeding (PEDPV-2)

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Abnormal Uterine Bleeding (PEDPV-2.1)
References (PEDPV-2)

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Abnormal Uterine Bleeding (PEDPV-2.1)

PVP.UB.0002.1.A

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- Abnormal uterine bleeding imaging indications in pediatric individuals are very similar to those for adult individuals. See **Abnormal Uterine Bleeding (AUB) (PV-2.1)** in the Pelvis Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - Transabdominal ultrasound is appropriate in all pediatric individuals.
  - Transvaginal (TV) ultrasound is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound is generally not appropriate in pediatric individuals or in individuals who have never been sexually active.
  - MRI Pelvis without contrast or without and with contrast (CPT® 72195 or CPT® 72197) is medically necessary if ultrasound is inconclusive.

Background and Supporting Information

The causes of vaginal bleeding in children differ from those in adolescents. Vaginal bleeding after the first week or so of life but before menarche is always abnormal and warrants evaluation. Common conditions before normal menarche include vaginal foreign bodies, infections, precocious puberty, and estrogen exposure. After menarche, pregnancy and excessive menstrual bleeding (ovulatory dysfunction) must be considered.

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Pelvic Inflammatory Disease (PID) (PEDPV-3)

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Pelvic Inflammatory Disease (PID) (PEDPV-3.1)
References (PEDPV-3)

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Pelvic Inflammatory Disease (PID) (PEDPV-3.1)

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- Pelvic inflammatory disease imaging indications in pediatric individuals are very similar to those for adult individuals. See **Pelvic Inflammatory Disease (PV-7.1)** in the Pelvis Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - Transabdominal ultrasound is appropriate in all pediatric individuals.
  - Transvaginal (TV) ultrasound is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound is generally not appropriate in individuals who are pre-pubescent or victims of abuse.
  - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) is medically necessary if ultrasound is inconclusive.
  - CT Pelvis with contrast (CPT® 72193) is medically necessary if MRI is not readily available.

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Amenorrhea (PEDPV-4)

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Amenorrhea (PEDPV-4.1)
References (PEDPV-4)

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Amenorrhea (PEDPV-4.1)

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- Females with primary amenorrhea and any of the following should be evaluated initially with pelvic ultrasound (CPT® 76856 or CPT® 76857):
  - Amenorrhea is usually primary and refers to absence of menstrual periods by age 16.
    - Normal pubertal development and negative pregnancy test.
    - Transabdominal ultrasound is appropriate in all pediatric individuals.
      - Transvaginal (TV) ultrasound is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound (CPT® 76830) is medically necessary for better view of genitourinary anomalies in sexually active females.
    - Delayed puberty with follicle-stimulating hormone (FSH) or luteinizing hormone (LH) that is elevated for the individual's age and Tanner stage.
- MRI Pelvis without contrast or without and with contrast (CPT® 72195 or CPT® 72197) and/or MRI Abdomen without contrast or without and with contrast (CPT® 74181 or CPT® 74183) are medically necessary for congenital anomalies or for pre-operative planning.

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Endometriosis (PEDPV-5)

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Endometriosis (PEDPV-5.1)
Reference (PEDPV-5)

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Endometriosis (PEDPV-5.1)

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- Endometriosis imaging indications in pediatric individuals are very similar to those for adult individuals. See **Endometriosis (PV-6.1)** in the Pelvis Imaging Guidelines.
- Pediatric-specific imaging considerations include:
  - Transabdominal ultrasound is appropriate in all pediatric individuals.
  - Transvaginal (TV) ultrasound is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound is generally not appropriate in individuals who are pre-pubescent or have never been sexually active.

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Suspected Adnexal Mass (PEDPV-6)

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Suspected Adnexal Mass (PEDPV-6.1)
References (PEDPV-6)

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Suspected Adnexal Mass (PEDPV-6.1)

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- Suspected adnexal mass imaging indications in pediatric individuals are very similar to those for adult individuals. See **Adnexal Mass/Ovarian Cysts (PV-5)** in the Pelvis Imaging Guidelines. Ultrasound is the first study for evaluation of a suspected adnexal mass.
- Pediatric-specific imaging considerations include the following:
  - Transabdominal ultrasound is appropriate in all pediatric individuals.
  - Transvaginal (TV) Ultrasound is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound is generally not appropriate in individuals who are pre-pubescent or have never been sexually active.
  - Adnexal masses with a solid component in individuals, age ≥15 years, should be imaged according to **Pediatric Germ Cell Tumors (PEDONC-10)** in the Pediatric and Special Populations Oncology Imaging Guidelines.

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Pelvic Pain/ Dyspareunia, and Ovarian Torsion (PEDPV-7)

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Pelvic Pain/Dyspareunia, and Ovarian Torsion (PEDPV-7.1)
References (PEDPV-7)

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Pelvic Pain/Dyspareunia, and Ovarian Torsion (PEDPV-7.1)

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- Pelvic Pain/Dyspareunia imaging indications in pediatric individuals are identical to those for adult individuals. See **Pelvic Pain/Dyspareunia, Female (PV-11.1)** in the Pelvis Imaging Guidelines.
- Ovarian torsion in children is typically associated with a normal ovary. Spontaneous torsion of a normal ovary is more common than torsion caused by a lead mass, such as a cyst or tumor. Torsion involves both the ovary and fallopian tube and typically presents with acute of onset of lower abdominal pain, often associated with nausea or vomiting.
  - Transabdominal ultrasound (CPT® 76856) with Doppler (CPT® 93975) is appropriate in all pediatric individuals.
  - Transvaginal (TV) ultrasound (CPT® 76830) with Doppler (CPT® 93975) is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound is generally not appropriate in individuals who are pre-pubescent or have never been sexually active.

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Polycystic Ovary Syndrome (PEDPV-8)

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Polycystic Ovary Syndrome (PEDPV-8.1)
References (PEDPV-8)

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Polycystic Ovary Syndrome (PEDPV-8.1)

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- Polycystic ovary syndrome imaging indications in pediatric individuals are very similar to those for adult individuals. See **Polycystic Ovary Syndrome (PCOS) (PV-8.1)** in the Pelvis Imaging Guidelines.
- Pediatric-specific imaging considerations include:
  - Transabdominal ultrasound (CPT® 76856) is appropriate in all pediatric individuals.
  - Transvaginal (TV) ultrasound (CPT® 76830) is appropriate in pediatric individuals who are sexually active or use a tampon and consent to the study. Transvaginal ultrasound is generally not appropriate in individuals who are pre-pubescent or have never been sexually active.

Background and Supporting Information

Adolescent girls may have multiple ovarian cysts as part of normal pubertal development. As such the diagnosis should not be based on morphology alone, but requires abnormal laboratory studies.

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Periurethral Cysts and Urethral Diverticula (PEDPV-9)

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Periurethral Cysts and Urethral Diverticula (PEDPV-9.1)

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Periurethral Cysts and Urethral Diverticula (PEDPV-9.1)

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- Periurethral cysts and urethral diverticula imaging indications in pediatric individuals are identical to those for adult individuals. See **Periurethral Cysts and Urethral Diverticula (PV-13)** in the Pelvis Imaging Guidelines.

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Fetal MRI (PEDPV-10)

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Fetal MRI (PEDPV-10.1)

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Fetal MRI (PEDPV-10.1)

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- Fetal MRI indications in pediatric individuals are identical to those for adult individuals. See **Fetal MRI (PV-15.1)** in the Pelvis Imaging Guidelines.

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Undescended Testis (PEDPV-11)

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Undescended Testis (PEDPV-11.1)
References (PEDPV-11)

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Undescended Testis (PEDPV-11.1)

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- Males with a history of cryptorchidism (undescended testis) have a several-fold risk increase of testicular cancer. It is important to diagnose and treat this condition either by bringing the undescended testis into the scrotum, or resecting the testis.
- Pediatric-specific imaging considerations include the following:
  - Suspected undescended testis is an indication for referral to a surgical subspecialist who should make the decision or be consulted on necessary imaging studies.
- After surgical evaluation or consultation, the following imaging is medically necessary for pre-operative evaluation:
  - Scrotal ultrasound (CPT® 76870) if testis not palpable in the scrotal sac and there is concern for retractile or inguinal testis. In general CT and MRI are not medically necessary to localize non-palpable testes, as the findings would typically not alter the surgical procedure.
    - If after ultrasound there is concern for associated urogenital abnormalities, or the surgical consultant or any provider in consultation with the surgical consultant indicates that advanced imaging results would significantly alter the surgical procedure either of the studies below are medically necessary:
      - MRI Abdomen (CPT® 74183) and Pelvis (CPT® 72197) without and with contrast
      - CT Abdomen and Pelvis with contrast (CPT® 74177)

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Scrotal Pathology (PEDPV-12)

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Scrotal Pathology (PEDPV-12.1)
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Scrotal Pathology (PEDPV-12.1)

PVP.SP.0012.1.A

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- Scrotal pathology imaging indications in pediatric individuals are very similar to those for adult individuals. See **Scrotal Pathology (PV-20.1)** in the Pelvis Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - Scrotal US (CPT® 76870) with Doppler (CPT® 93975 or CPT® 93976) is medically necessary for concerns of testicular torsion.
  - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) is medically necessary if torsion is unlikely on ultrasound and no surgical exploration is planned. MRI is not typically used for the acute scrotum due to the limited availability of equipment and the long examination time involved.
  - Since the acceptance of Doppler US as the primary imaging for evaluation of acute scrotum, scintigraphy is not medically necessary. The unavailability of nuclear medicine imaging in many practices and its use of ionizing radiation, its poor anatomical details, and the time required for imaging are other limiting factors.

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Penis-Soft Tissue Mass (PEDPV-13)

Guideline

Penis-Soft Tissue Mass (PEDPV-13.1)

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Penis-Soft Tissue Mass (PEDPV-13.1)

PVP.ST.0013.1.A

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- Penile soft tissue masses are very rare in pediatric individuals, and imaging indications are identical to those for adult individuals. See **Penis–Soft Tissue Mass (PV-18.1)** in the Pelvis Imaging Guidelines.

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Incontinence (PEDPV-14)

Guideline

Incontinence (PEDPV-14.1)
References (PEDPV-14)

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Incontinence (PEDPV-14.1)

PVP.IN.0014.1.A

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- Incontinence imaging indications in pediatric individuals are very similar to those for adult individuals. See **Urinary Incontinence/Pelvic Prolapse/Fecal Incontinence (PV-22)** in the Pelvis Imaging Guidelines.
- Most often incontinence in children is not due to a medical condition. Several uncommon disorders that can lead to urinary incontinence include a spinal cord defect such as spina bifida, ureteral duplication with ectopic insertion, and overactive bladder or dysfunctional voiding.
- No imaging is needed if primary enuresis is suspected; however, imaging evaluation may be warranted if ureteral duplication or overactive bladder or dysfunctional voiding is suspected. The physician should obtain a full medical history and urinalysis before imaging is done.
- Radiopharmaceutical urinary bladder residual study (CPT® 78730) is medically necessary for suspicion of urinary retention and a recent non-diagnostic ultrasound.
- Pediatric-specific imaging considerations include the following:
  - MRI Pelvis without and with contrast (CPT® 72197) is medically necessary if ultrasound is inconclusive or spinal abnormality is suspected.
  - CT Pelvis with contrast (CPT® 72193) is appropriate if MRI is not readily available.

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Patent Urachus (PEDPV-15)

Guideline

Patent Urachus (PEDPV-15.1)
References (PEDPV-15)

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Patent Urachus (PEDPV-15.1)

PVP.UR.0015.1.A

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- Ultrasound pelvis (CPT® 76856) is medically necessary as the initial evaluation for patent urachus.
  - ANY of the following are medically necessary if the ultrasound is inconclusive or insufficient for preoperative planning:
    - MRI Pelvis without contrast (CPT® 72195)
    - MRI Pelvis without and with contrast (CPT® 72197)
    - CT Pelvis with contrast (CPT® 72193)
- Repeat imaging of asymptomatic individuals is not generally necessary, but is medically necessary for the following:
  - New or worsening symptoms
  - Preoperative planning

Background and Supporting Information

The urachus is a “tube” connecting the fetal bladder to the umbilical cord. It is usually obliterated during fetal growth, but if it remains patent, there can be a complete or partial connection between the bladder and the umbilicus.

Ultrasound has an accuracy greater than 90%.

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General Guidelines (PEDPN-1)

Guideline

Procedure Codes Associated with Peripheral Nerve And Neuromuscular Disorders (PNND) Imaging
General Guidelines (PEDPN-1.0)
Age Considerations (PEDPN-1.1)
Modality General Considerations (PEDPN-1.3)
References (PEDPN-1)

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Procedure Codes Associated with Peripheral Nerve And Neuromuscular Disorders (PNND) Imaging

PNP.GG.ProcedureCodes.A
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MRI	CPT®
MRI Neck without contrast	70540
MRI Neck without and with contrast	70543
MRI Cervical without contrast	72141
MRI Cervical without and with contrast	72156
MRI Brachial Plexus without contrast (unilateral)	73218
MRI Brachial Plexus without and with contrast (unilateral)	73220
MRI Brachial Plexus without contrast (bilateral)	71550
MRI Brachial Plexus without and with contrast (bilateral)	71552
MRI Chest without contrast	71550
MRI Chest without and with contrast	71552
MRI Thoracic without contrast	72146
MRI Thoracic without and with contrast	72157
MRI Lumbar without contrast	72148
MRI Lumbar without and with contrast	72158
MRI Abdomen without contrast	74181

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MRI	CPT ®
MRI Abdomen without and with contrast	74183
MRI Pelvis without contrast	72195
MRI Pelvis without and with contrast	72197
MRI Upper Extremity Other Than Joint without contrast	73218
MRI Upper Extremity Other Than Joint with contrast	73219
MRI Upper Extremity Other Than Joint without and with contrast	73220
MRI Upper Extremity Joint without contrast	73221
MRI Upper Extremity Joint with contrast	73222
MRI Upper Extremity Joint without and with contrast	73223
MRI Lower Extremity Other Than Joint without contrast	73718
MRI Lower Extremity Other Than Joint with contrast	73719
MRI Lower Extremity Other Than Joint without and with contrast	73720
MRI Lower Extremity Joint without contrast	73721
MRI Lower Extremity Joint with contrast	73722
MRI Lower Extremity Joint without and with contrast	73723
Unlisted MRI procedure (for radiation planning or surgical software)	76498

MRA	CPT ®
MRA Upper Extremity	73225
MRA Lower Extremity	73725

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Nuclear Medicine	
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging: skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging: whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
Bone Marrow Imaging Limited Areas	78102
Bone Marrow Imaging Multiple Areas	78103
Bone Marrow Imaging Whole Body	78104
Nuclear Bone Scan Limited	78300
Nuclear Bone Scan Multiple Areas	78305
Nuclear Bone Scan Whole Body	78306
Bone Scan Three Phase	78315
Radiopharmaceutical Localization Imaging Limited Area	78800
Radiopharmaceutical Localization Imaging Whole Body	78802
Radiopharmaceutical Localization Imaging SPECT	78803

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General Guidelines (PEDPN-1.0)

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- A pertinent clinical evaluation including a detailed history, physical examination since the onset or change in symptoms with a thorough neurologic examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in symptoms can serve as a pertinent clinical evaluation.
  - EMG may not be of clinical utility or obtainable in infants or individuals with severe developmental delay
  - EMG/NCS results may not be abnormal until 10 days after injury.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the peripheral nervous system is not supported. Advanced imaging of the peripheral nervous system is only appropriate in individuals who have documented active clinical signs or symptoms of disease involving the peripheral nervous system.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the peripheral nervous system are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Age Considerations (PEDPN-1.1)

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- Many conditions affecting the peripheral nervous system in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases that occur in both pediatric and adult populations, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are 18 years old or younger should be imaged according to the Pediatric Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Peripheral Nerve and Neuromuscular Disorder Imaging Guidelines should be imaged according to the General Peripheral Nerve Disorder Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Modality General Considerations (PEDPN-1.3)

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- MRI
  - MRI without and with contrast is the preferred modality for pediatric peripheral nerve imaging unless otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize individual movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast as supported by these condition-based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Recent evidence based literature demonstrates the potential for gadolinium deposition in various organs including the brain after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported by the guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
- CT
  - CT is rarely used in the evaluation of pediatric peripheral nerve disorders. See specific guideline sections for indications.
- Ultrasound
  - Ultrasound is rarely used in the evaluation of pediatric peripheral nerve disorders. See specific guideline sections for indications.

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- Nuclear Medicine
  - Nuclear medicine studies are generally not medically necessary in the evaluation of peripheral nerve disorders. See for specific imaging guidelines regarding PET/CT in evaluation of peripheral nerve tumors.
- 3D Rendering
  - 3D Rendering indications in pediatric PND imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines.
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations

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Neurofibromatosis (PEDPN-2)

Guideline

Neurofibromatosis – General Information (PEDPN-2.0)
Neurofibromatosis 1 (PEDPN-2.1)
Neurofibromatosis 2 (PEDPN-2.2)

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Neurofibromatosis – General Information (PEDPN-2.0)

PNP.NF.0002.0.A
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- This guideline section includes imaging indications for individuals with neurofibromatosis and known benign lesions.
- For cancer screening guidelines, see **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric Oncology Imaging Guidelines.
- For Peripheral Nerve Sheath Tumors, see **Peripheral Nerve Sheath Tumors (PNST) (PND-9.1)** in the Peripheral Nerve and Neuromuscular Disorders (PND) Imaging Guidelines.
- For guidelines related to known malignancies in individuals with NF1, see the appropriate imaging guideline for the specific cancer type.

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Neurofibromatosis 1 (PEDPN-2.1)

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- See **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric Oncology Imaging Guidelines.
- For Peripheral Nerve Sheath Tumors, see **Peripheral Nerve Sheath Tumors (PNST) (PND-9.1)** in the Peripheral Nerve Disorders and Neuromuscular (PND) Imaging Guidelines.
- For guidelines related to known malignancies in individuals with NF1, see the appropriate imaging guideline for the specific cancer type.

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Neurofibromatosis 2 (PEDPN-2.2)

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- See **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric Oncology Imaging Guidelines.
- Individuals with NF2 and known meningioma should be imaged according to guidelines in **Meningiomas (Intracranial and Intraspinal) (ONC-2.8)** in the Oncology Imaging Guidelines.
- Individuals with NF2 and known ependymoma should be imaged according to guidelines in **Ependymoma (PEDONC-4.8)** in the Pediatric Oncology Imaging Guidelines.

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Brachial Plexus (PEDPN-3)

Guideline

Brachial Plexus (PEDPN-3.1)
References (PEDPN-3)

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Brachial Plexus (PEDPN-3.1)

PNP.BP.0003.1.A
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Disorders of the brachial plexus can generally be identified and distinguished from lesions in other locations by clinical and electromyography/nerve conduction (EMG/NCV) examination. If the diagnosis remains unclear, advanced imaging can be helpful as a pre-operative study to evaluate the anatomy of brachial plexus lesions that should have already been defined by clinical examination. However, due to the technical challenges, EMG/NCV is not required prior to imaging in children.

- MRI is the preferred modality for imaging the brachial plexus.
  - CT is not often useful and should not be used as a substitute for MRI.
  - MRI Upper Extremity Other Than Joint without contrast (CPT® 73218) or MRI Upper Extremity Other Than Joint without and with contrast (CPT® 73220) is medically necessary for unilateral brachial plexus.
  - MRI Chest without contrast (CPT® 71550) or MRI Chest without and with contrast (CPT® 71552) is medically necessary for bilateral brachial plexus studies.
  - MRI Neck without contrast (CPT® 70540) is indicated for upper trunk lesions.
  - It is rare for more than one CPT® code to be necessary to adequately image the brachial plexus area of interest.
  - MRI Shoulder without contrast (CPT® 73221) or MRI Shoulder without and with contrast (CPT® 73223) is medically necessary in infants with brachial plexopathy due to birth trauma if requested for preoperative planning. These individuals often have glenohumeral dysplasia and require shoulder surgery.
  - MRI Cervical Spine without contrast (CPT® 72141) is medically necessary if there is clinical suspicion for cervical nerve root avulsion.
  - PET/CT skull base to mid-thigh (CPT® 78815) is appropriate if there is a contraindication to MRI in individuals with a known malignancy or post-treatment syndrome.

Background and Supporting Information

The goal of imaging is to visualize the entire course of the neural network from the preganglionic to the postganglionic segments

The MRI Chest and Neck are inherently bilateral, whereas MRI Upper Extremity is unilateral.

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References (PEDPN-3)

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Gaucher Disease (PEDPN-4)

Guideline

Gaucher Disease (PEDPN-4.1)

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Gaucher Disease (PEDPN-4.1)

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- Gaucher Disease imaging indications in pediatric individuals are very similar to those for adult individuals. See **Gaucher Disease (Storage Disorders) (PN-6.3)** in the Peripheral Nerve and Neuromuscular Disorders(PND) Imaging Guidelines.

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Spinal Muscular Atrophy (PEDPN -5)

Guideline

Spinal Muscular Atrophy (PEDPN-5.1)
References (PEDPN-5)

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Spinal Muscular Atrophy (PEDPN-5.1)

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- Spinal Muscular Atrophy
  - Molecular genetic testing is the standard tool for diagnosis for the early consideration in any infant with weakness or hypotonia
    - MRI is usually not medically necessary
  - See **Developmental Motor Delay (PEDHD-19.3)** in the Pediatric Head Imaging Guidelines for presentation of weakness or a loss of skills.

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General Guidelines (PEDPVD-1)

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Procedure Codes Associated with PVD Imaging (PEDPVD)

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Description	CPT®
MRA	
Magnetic resonance angiography, head; without contrast material(s), followed by contrast material(s) and further sequence	70546
Magnetic resonance angiography, neck; without contrast material(s), followed by contrast material(s) and further sequences	70549
Magnetic resonance angiography, chest (excluding myocardium), with or without contrast material(s)	71555
Magnetic resonance angiography, pelvis, with or without contrast material(s)	72198
Magnetic resonance angiography, upper extremity, with or without contrast material(s)	73225
Magnetic resonance angiography, lower extremity, with or without contrast material(s)	73725
Magnetic resonance angiography, abdomen, with or without contrast material(s)	74185
CTA	
Computed tomographic angiography, head, with contrast material(s), including noncontrast images, if performed, and image postprocessing	70496
Computed tomographic angiography, neck, with contrast material(s), including noncontrast images, if performed, and image postprocessing	70498

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Description	CPT®
Computed tomographic angiography, chest (noncoronary), with contrast material(s), including noncontrast images, if performed, and image postprocessing	71275
Computed tomographic angiography, upper extremity, with contrast material(s), including noncontrast images, if performed, and image postprocessing	73206
Computed tomographic angiography, lower extremity, with contrast material(s), including noncontrast images, if performed, and image postprocessing	73706
Computed tomographic angiography, abdomen and pelvis, with contrast material(s), including noncontrast images, if performed, and image postprocessing	74174
Computed tomographic angiography, abdomen, with contrast material(s), including noncontrast images, if performed, and image postprocessing	74175
CTA Abdominal Aorta with Bilateral Iliofemoral Runoff	75635
Ultrasound	
Duplex scan of extracranial arteries; complete bilateral study	93880
Duplex scan of extracranial arteries; unilateral or limited study	93882
Non-invasive physiologic studies of extracranial arteries, complete bilateral study	93875
Limited bilateral noninvasive physiologic studies of upper or lower extremity arteries	93922
Complete bilateral noninvasive physiologic studies of upper or lower extremity arteries	93923
Duplex scan of upper extremity arteries or arterial bypass grafts; complete bilateral	93930
Duplex scan of upper extremity arteries or arterial bypass grafts; unilateral or limited	93931
Non-invasive physiologic studies of extremity veins, complete bilateral study	93965

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Description	CPT®
Duplex scan of extremity veins including responses to compression and other maneuvers; complete bilateral study	93970
Duplex scan of extremity veins including responses to compression and other maneuvers; unilateral or limited study	93971
Duplex scan of hemodialysis access (including arterial inflow, body of access, and venous outflow)	93990

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General Guidelines (PEDPVD-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms, including a detailed history, physical examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) can serve as a pertinent clinical evaluation.
- The use of advanced imaging to screen asymptomatic individuals for disorders involving the peripheral vascular system is not considered medically necessary unless otherwise stated in a specific guideline section.
- Advanced imaging of the peripheral vascular system is considered medically necessary in individuals who have documented active clinical signs or symptoms of disease involving the peripheral vascular system.
- Repeat imaging studies of the peripheral vascular system are not medically necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect the individual's management or treatment decisions unless otherwise stated in a specific guideline section.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Age Considerations (PEDPVD-1.1)

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Many conditions affecting the peripheral vascular system in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and general populations, differences may exist in management due to the individual's age, comorbidities, and differences in disease natural history between children and adults.

- Individuals who are 18 years old and younger should be imaged according to the Pediatric Peripheral Vascular Disease imaging guidelines if discussed. Any conditions not specifically discussed in the pediatric peripheral vascular disease imaging guidelines should be imaged according to the general peripheral vascular disease imaging guidelines. Individuals who are >18 years old should be imaged according to the general Peripheral Vascular Disease imaging guidelines, except where directed otherwise by a specific guideline section.

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Modality General Considerations (PEDPVD-1.3)

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- MRI
  - MRI is generally performed without and with contrast unless the individual has a documented contraindication to gadolinium or otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize the individual's movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years), as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this population, MRI sessions should be planned with a goal of minimizing anesthesia exposure adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition-based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast is considered medically necessary if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
      - Recent evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
  - The presence of surgical hardware or implanted devices may preclude MRI.
  - The selection of best examination may require coordination between the provider and the imaging service.
- CT

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  - CT or CTA is considered medically necessary for further evaluation of abnormalities suggested on prior US or MRI procedures.
  - CT is considered medically necessary without prior MRI or US, especially in the following (non-exhaustive list of) settings:
    - Lymphatic malformations
    - Vascular abnormalities (including vasculitis, thrombosis, narrowing, aneurysm, dissection, and varices)
    - For pre-operative planning or assessment of post-operative complications
  - In some cases, especially in follow-up of a known finding, it may be appropriate to limit the exam to the region of concern to reduce radiation exposure.
  - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.
  - The selection of the best examination may require coordination between the provider and the imaging service.
- Ultrasound
  - Ultrasound can be helpful in evaluating arterial, venous, and lymphatic malformations.
  - Ultrasound can be limited by the imaging window and the individual's body type.
  - CPT® codes vary by body area and presence or absence of Doppler imaging and are included in the table at the beginning of this guideline.
- 3D Rendering
  - 3D Rendering indications in pediatric imaging are identical to those in the general imaging guidelines. See **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be indicated and warranted for specific clinical situations.

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Vascular Anomalies (PEDPVD-2)

Guideline

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Lymphatic Malformations (PEDPVD-2.2)

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Initial imaging

- Ultrasound is medically necessary as an initial examination for superficial lesions.
- CT with contrast of the affected body part is medically necessary for lesions with acute enlargement and concerns for compression when MRI is contraindicated.
- MRI without contrast or without and with contrast of the affected body part is medically necessary for:
  - Lymphatic malformations involving deep tissues
  - Malformations too large to be completely imaged with ultrasound
  - Inconclusive ultrasound findings
  - Preoperative planning

Clinical changes and monitoring treatment

MRI without contrast or without and with contrast of the affected body part is medically necessary every 3 months during active treatment for individuals with aggressive lesions being treated with systemic therapy.

MRI without contrast or without and with contrast of the affected body part is medically necessary for:

- Preoperative planning
- Post-treatment evaluation

CT with contrast of the affected body part is considered medically necessary to evaluate lesions with acute enlargement and concerns for compression when MRI is contraindicated.

Surveillance

Annual surveillance imaging with MRI without contrast or without and with contrast is medically necessary when lymphatic malformations are located in body areas where growth could cause significant organ dysfunction or functional impairment (e.g. airway, intestine).

Evidence Discussion

Vascular and lymphatic malformations encompass a broad variety of conditions and have very heterogeneous natural history and treatment approaches. Lesions can be

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divided into low flow lesions (lymphatic, capillary and venous malformations), and high-flow lesions (arteriovenous malformations and fistulas).

Lymphatic malformations are composed of dilated lymphatic channels filled with proteinaceous fluid and do not connect to normal lymphatic channels. They are typically soft, non-pulsatile masses with normal overlying skin. MRI is often used for further characterization of lymphatic malformations, especially for deep or complex lesions, providing detailed images without ionizing radiation. Large lesion characterization may be limited by ultrasound imaging window. Ultrasound is the initial imaging of choice however it is limited in evaluating malformation relationship to airway or bony structures.[1]

[1] Snyder EJ, Sarma A, Borst AJ, Tekes A. Lymphatic Anomalies in Children: Update on Imaging Diagnosis, Genetics, and Treatment. *AJR Am J Roentgenol*. 2022;218(6):1089-1101. doi:10.2214/AJR.21.27200.

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Venous Malformations (PEDPVD-2.3)

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Venous Malformation Imaging Indications

Initial imaging

- Ultrasound with Doppler is medically necessary as an initial examination for superficial lesions.
- MRI without contrast or without and with contrast of the affected body part is medically necessary for venous malformations for preoperative assessment to evaluate the extent of malformation and their relationship to normal structures.
- MRA or CTA has a limited role in evaluating most venous malformations but is considered medically necessary (contrast as requested of the affected body part) if MRI or CT is equivocal and the results will impact acute management decisions.
- CT can also be used to characterize venous malformations and their relationship to normal structures but is generally not as accurate as MRI.
  - CT with contrast of the affected body part is considered medically necessary when MRI is inconclusive or contraindicated.

Clinical changes and monitoring treatment

MRI without contrast or without and with contrast of the affected body part is medically necessary for preoperative assessment of venous malformations.

CT Chest with contrast with PE protocol (CPT® 71260) or CTA Chest (CPT® 71275) is medically necessary to evaluate for suspected pulmonary embolism in individuals with Klippel-Trenaunay syndrome and CLOVES syndrome.

Surveillance

Annual surveillance imaging is medically necessary for venous malformations located in body areas where growth could cause significant organ dysfunction or functional impairment.

Evidence Discussion

Venous malformations are slow-flow lesions characterized by dilated venous spaces and a normal arterial component. They are soft, compressible, non-pulsatile lesions that are usually blue to deep purple in color. Lesions can range from very small to large

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infiltrating ones. Some may change size with Valsalva. These lesions can enlarge over time and become painful when associated with thrombophlebitis.[2]

Venous malformations are usually isolated, but they may be seen in multiple syndromes including Klippel-Trenaunay (KT) syndrome, Blue Rubber Bleb Nevus syndrome (BRBN), Maffucci syndrome, Proteus syndrome, Bannayan-Riley-Ruvalcaba syndrome, Parkes-Weber syndrome and congenital lipomatous overgrowth, vascular malformations, epidermal nevi and scoliosis/skeletal/spinal anomalies (CLOVES) syndrome. Both Klippel-Trenaunay syndrome and CLOVES syndrome have been found to have increased risk of venous thrombosis and pulmonary embolism, particularly after surgery or sclerotherapy.

[2] Bertino FJ, Hawkins CM. Contemporary management of extracranial vascular malformations. *Pediatr Radiol*. 2023;53:1600–1617. doi:10.1007/s00247-023-05670-1.

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Capillary Malformations (PEDPVD-2.4)

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Imaging Indications for Capillary Malformations

Initial Imaging

- MRI (without contrast or without and with contrast) is considered medically necessary to evaluate occult underlying neurologic structures associated with encephalocele, spinal dysraphism, or Sturge-Weber syndrome.

Evidence Discussion

Capillary malformations including Nevus simplex (NS) and Port wine birthmarks (PWBs) are characterized by a collection of small vascular channels in the dermis and generally do not require advanced imaging because the diagnosis is made clinically. However, MRI is considered medically necessary to evaluate underlying neurologic structures in specific cases, such as suspected Sturge-Weber syndrome.[3] Additional imaging is not considered medically necessary in the absence of other complex associated clinical findings such as Sturge-Weber syndrome (SWS) and Klippel-Trenaunay syndrome (KTS). [4]

[3] Bertino FJ, Hawkins CM. Contemporary management of extracranial vascular malformations. *Pediatr Radiol*. 2023;53:1600–1617. doi:10.1007/s00247-023-05670-1

[4] Paradiso MM, Shah SD. Infantile Hemangiomas and Vascular Anomalies. *Pediatr Ann*. 2024;53(4):e129-e137. doi:10.3928/19382359-20240205-04.

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Arteriovenous Malformations (AVMs) and Fistulas (PEDPVD-2.5)

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Imaging Indications for AVMs and Fistulas

Initial Imaging

- Ultrasound with Doppler is medically necessary as an initial examination for superficial lesions.
- MRI without contrast or without and with contrast of the affected body part is also medically necessary to evaluate the extent of AVMs and their relationship to normal structures.
- MRA (contrast as requested) of the affected body part is medically necessary for evaluation of known AVMs.
- It is unusual for both MRI and MRA to be necessary for routine treatment response or surveillance imaging of AVMs, but both are considered medically necessary for preoperative planning.
- CT and CTA can also be used to characterize AVMs and their relationship to normal structures but is generally not better than MRI and has associated radiation risks.
  - CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Clinical changes and monitoring treatment

MRI or MRA (contrast as requested) of the affected body part is medically necessary for evaluation of treatment response.

It is unusual for both MRI and MRA to be necessary for routine treatment response or surveillance imaging of AVMs, but both may be medically necessary for preoperative planning.

CT with contrast and/or CTA (contrast as requested) of the affected body part is medically necessary when MRI and/or MRA is inconclusive or contraindicated.

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Surveillance

MRI or MRA (contrast as requested) of the affected body part is medically necessary for annual surveillance of known AVMs located in body areas where growth could cause significant organ dysfunction or functional impairment.

Note:

For imaging indications specific to pulmonary AVM see **Pulmonary Arteriovenous Malformations (PEDCH-14.2)** in the Pediatric Chest Imaging guidelines, and for imaging indications specific to cerebral AVM see **Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2)** in the Pediatric Head Imaging guidelines.

Evidence Discussion

Arteriovenous malformations are characterized by a network of multiple abnormal vascular channels interposed between enlarged feeding arteries and draining veins. The arteriovenous fistula has a single communication interposed between a feeding artery and a draining vein. The normal capillary bed is absent in both lesions. Both lesions may have an aggressive clinical course and are characterized by a reddish pulsatile mass which has a thrill or bruit. Though often recognized at birth, these lesions may grow and present near adolescence.

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Vascular Tumors (PEDPVD-2.6)

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Imaging Vascular Tumors

Initial imaging

Any or all of the following imaging of the affected body part is considered medically necessary for the initial imaging and characterization of vascular tumors:

- Ultrasound with Doppler an initial examination
- MRI without contrast or without and with contrast to determine the extent of arteriovenous malformations and their relationship to normal structures
- MRA (contrast as requested) of the affected body part is considered medically necessary for evaluation.

CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Clinical changes and monitoring treatment

For changes in clinical status (symptoms or exam findings) related to the vascular tumor either of the following imaging is considered medically necessary:

- MRI without contrast or without and with contrast of the affected body to evaluate treatment response
- MRA (contrast as requested) of the affected body part

For preoperative planning both MRI and MRA (as requested) of the affected body part are considered medically necessary.

CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Surveillance imaging

- MRA (contrast as requested) of the affected body part is medically necessary for the surveillance of known vascular tumors.
- Imaging of vascular tumors with both MRI and MRA for routine treatment response or surveillance is not considered medically necessary.

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- CT with contrast and/or CTA (contrast as requested) of the affected body part is considered medically necessary when MRI and/or MRA is inconclusive or contraindicated.

Evidence Discussion

Vascular tumors include a variety of benign, borderline, and malignant tumors, which have variable clinical courses, including but not limited to Infantile hemangiomas see **Infantile Hemangiomas (PEDPVD-5)**, Epithelioid hemangioma, Kaposiform hemangioendothelioma, Kaposi sarcoma, Epithelioid hemangioendothelioma, and Angiosarcoma of soft tissue.

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Vasculitis (PEDPVD-3)

Guideline

Large Vessel Vasculitis (PEDPVD-3.2)
Medium Vessel Vasculitis (PEDPVD-3.3)
Small Vessel Vasculitis (PEDPVD-3.4)
References (PEDPVD-3)

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Large Vessel Vasculitis (PEDPVD-3.2)

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Imaging Indications

Initial imaging

- ANY of the following modalities is considered medically necessary for the initial evaluation of Takayasu arteritis:
  - MRA of the affected body area(s) (contrast as requested)
  - CTA of the affected body area(s) (contrast as requested)
  - Ultrasound with Doppler of the affected body area(s)

Clinical changes and monitoring treatment

- In individuals being treated with systemic therapy, imaging (MRA or CTA or US) of the affected areas is medically necessary every 3 months to monitor active treatment for response.

Surveillance

- Imaging with MRA or CTA or US of the affected areas is medically necessary annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention.

Evidence Discussion

Takayasu arteritis is the predominant large vessel vasculitis occurring in children.

Systemic vasculitis is much less common in pediatric individuals than in adults. The diagnostic pathways and treatment options are similar for both age groups. For additional information on vasculitis see **Large Vessel Vasculitis (PVD-6.9)** in the general Peripheral Vascular Disease Imaging Guidelines.

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Medium Vessel Vasculitis (PEDPVD-3.3)

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Imaging Indications

- Some children who have had COVID 19 develop a severe inflammatory disease that can present in a similar way to Kawasaki disease or toxic shock syndrome. This syndrome has been defined by the US Centers for Disease Control and Prevention as multisystem inflammatory syndrome in children (MIS-C). See **Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)** in the Pediatric Cardiac Imaging Guideline.
- Imaging guidelines for Kawasaki Disease- see **Kawasaki Disease (PEDCD-6)** in the Pediatric Cardiac Imaging Guideline.

Initial Imaging for Polyarteritis Nodosa

- For evaluation of polyarteritis nodosa:
  - ANY of the following modalities are considered medically necessary for the initial evaluation:
    - MRA of the affected body area(s) (contrast as requested)
    - CTA of the affected body area(s) (contrast as requested)
    - Ultrasound with Doppler of the affected body area(s)

Clinical Changes and Monitoring Treatment for Polyarteritis Nodosa

- For evaluation of polyarteritis nodosa:
  - MRA or CTA or US of the affected body part is medically necessary every 3 months during active treatment with systemic therapy for treatment response.

Surveillance Imaging for Polyarteritis Nodosa

- MRA or CTA or US of the affected body part is medically necessary annually for surveillance of known involved body areas to detect progressive vascular damage that may require intervention.

Evidence Discussion

Polyarteritis nodosa and Kawasaki Disease are the primary medium vessel vasculitides occurring in children.

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Small Vessel Vasculitis (PEDPVD-3.4)

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Imaging Indications Small Vessel Vasculitis

- Advanced imaging is not sensitive enough to detect changes in small vessels and is **not** considered medically necessary for primary assessment of any small vessel vasculitis.
- End-organ damage occurs with several of the small vessel vasculitides. Advanced imaging is considered medically necessary for the following:
  - Henoch-Schönlein Purpura (HSP) is the most common vasculitis of childhood, mainly involving small blood vessels. Ultrasound abdomen (CPT® 76700) is commonly used to evaluate possible gastrointestinal complications (including bowel wall edema and hemorrhage, and intussusception) in known or suspected HSP, and should be approved when requested for that indication.
  - Granulomatosis with polyangiitis (GPA, formerly known as Wegener's granulomatosis):
    - CT Sinuses (CPT® 70486) and/or CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is considered medically necessary in the following circumstances:
      - New or worsening clinical symptoms affecting the body area requested
      - To assess response to medical therapy when a change in treatment regimen is being considered
      - Annually to evaluate the extent of disease
  - Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg-Strauss Syndrome):
    - CT Chest without contrast (CPT® 71250) or with contrast (CPT® 71260) is considered medically necessary in the following circumstances:
      - New or worsening clinical symptoms affecting the body area requested
      - To assess response to medical therapy when a change in treatment regimen is being considered
      - Annually to evaluate the extent of disease
  - Immune complex associated small-vessel vasculitis [immunoglobulin A associated vasculitis (IgAV)]:
    - Doppler ultrasound of the affected body part (most commonly abdomen) is considered medically necessary in the following circumstances:
      - New or worsening clinical symptoms affecting the body area requested

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- To assess response to medical therapy when a change in treatment regimen is being considered
- Annually to evaluate the extent of disease

Evidence Discussion

Henoch-Schonlein Purpura (HSP) is the most common vasculitis of childhood, mainly involving small blood vessels. Imaging modalities such as ultrasound, CT, and Doppler ultrasound are used to evaluate gastrointestinal complications and monitor treatment response.

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Disorders of the Aorta and Visceral Arteries (PEDPVD-4)

Guideline

Thoracic Aortic Disease (PEDPVD-4.1)
Aortic Congenital Vascular Malformations (PEDPVD-4.2)
Visceral Artery Aneurysms (PEDPVD-4.3)
References (PEDPVD-4)

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Thoracic Aortic Disease (PEDPVD-4.1)

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Imaging Indications for Thoracic Aortic Disease

Familial Aortopathies

- For aortopathies such as the following:
  - Marfan
  - Ehlers-Danlos (EDS)- a genetic mutation known to predispose to aortic aneurysms/ dissections (TGFBR1, TGFBR2, FBN1, ACTA2, or MYH11)
  - Loeys-Dietz
  - Familial thoracic aneurysm and dissections
- Screening: for Family history with first-degree relative of aortopathy
  - Asymptomatic individuals with no signs or symptoms of disease, whose first-degree relative has no definitive gene defect, can have screening echo (TTE) annually.
- Initial workup: Individuals with suspected aortopathies (gene positive, physical exam positive, or other findings) or definite disease associated with aortopathy
  - Echocardiogram (TTE) at the time of evaluation.
  - If the consideration is for Loeys-Dietz any of the following are considered medically necessary in addition to the TTE at the time of work up:
    - MRA or CTA Head
    - MRA or CTA Neck
    - MRA or CTA Chest
    - MRA or CTA Abdomen and Pelvis
    - MRA or CTA of area of concern when there is an incidental finding on other imaging
- Surveillance: Suspected or known disease but **normal** aortic imaging:
  - Individuals with suspected genetic aortopathies but no disease can have an echocardiogram to assess for change:
    - At 6 months
    - Then annually
  - Individuals with Loeys-Dietz can be imaged with any of the following:
    - Echocardiogram
    - MRA or CTA of (any or all):
      - Head

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        - Neck
        - Chest
        - Abdomen
        - Pelvis
    - Individuals with Loeys-Dietz can be imaged with the above at the following intervals:
        - At 6 months
        - Then annually
- Surveillance: Suspected disease and **previous abnormal** imaging
    - Individuals with abnormal thoracic imaging can be imaged with (both):
        - Echocardiogram
        - CTA or MRA of (any):
            - Chest
            - Abdomen
            - Pelvis
            - Head (Loeys-Dietz)
            - Neck (Loeys-Dietz)
    - The above imaging is considered medically necessary as follows:
        - At the time of diagnosis
        - In 6 months after diagnosis (if older than 2 years)
        - Then as follows based on the individual's age:
            - Individual's age 0 to 2 years:
                - Every 3 months
            - Individual's age 3 to 12 years:
                - Every 6 months
            - Individual's age 13 years and older:
                - Every 12 months (if <4.5 or < 0.5 cm growth per year)
                - Every 6 months if ≥4.5 or ≥0.5 cm growth per year, or any Loeys-Dietz patient)
    - If the diameter z score is increased, then a repeat study can be done prior to the next allowed study, to assess for rate of change
- If there are symptoms of dissection any or all of the following are considered medically necessary:
    - Echo
    - CTA or MRA of (any or all):
        - Chest
        - Abdomen
        - Pelvis

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- For pediatric individual with dissection, imaging per vascular surgery and cardiology or any provider in consultation with vascular surgery at **any** interval.
- Miscellaneous syndromes with potential aortopathy as major feature of congenital heart disease
  - Individuals with Turner syndrome see section **Aortic disease in Turner Syndrome (CD-11.2.10)** in the Cardiac Imaging Guideline
  - Williams syndrome See section **LVOT lesions (PEDCD-2.4.10)** in the Pediatric Cardiology Imaging Guideline
  - Individuals with congenital heart disease would be managed based on **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)** in the Pediatric Cardiology Imaging Guideline
- Miscellaneous disorders that can affect the aorta such as osteogenesis imperfecta, homocystinuria, polycystic kidney disease, pseudoxanthoma elasticum, and Hurler syndrome.
  - Screening echocardiogram yearly.
  - If positive findings, follow protocol for aortic root dilatation.
- Follow-up of thoracic aortic abnormalities related to the following conditions are addressed in other imaging guidelines:
  - Coarctation of the Aorta- See **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)** in the Pediatric Cardiac Imaging Guidelines
  - Congenital rubella syndrome- See **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)** in the Pediatric Cardiac Imaging Guidelines
  - Kawasaki Syndrome- See **Kawasaki Disease (PEDCD-6)**
  - Neurofibromatosis- See **General Guidelines (PEDCD-1.0)** in the Pediatric Cardiac Imaging Guidelines

Evidence Discussion

Familial aortopathies such as Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, and familial thoracic aneurysm and dissections are included. Imaging modalities such as echocardiogram (TTE), magnetic resonance angiography (MRA), and computed tomography angiography (CTA) are used to evaluate the extent of aortic involvement and monitor disease progression.

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Aortic Congenital Vascular Malformations (PEDPVD-4.2)

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Imaging Indications for Aortic Congenital Vascular Malformations

Imaging for Aortic Congenital Vascular Malformation

Description	CPT®
CT Chest with contrast	71260
CTA Chest	71275
MRA Chest	71555
Cardiac MRI without contrast	75557
Cardiac MRI without and with contrast	75561

Any of the imaging in the **above table** is medically necessary for the initial diagnosis of Congenital Aortic Vascular Malformation.

Vascular rings may impact both the esophagus and trachea. CTA or MRA of the Neck is considered medically necessary to evaluate the relationship of the aortic arch to the trachea and esophagus for surgical planning.

Evidence Discussion

Congenital Aortic Vascular Malformations can be asymptomatic or can present with impact on breathing or swallowing when there is associated compression from vascular rings. Vascular rings may impact both the esophagus and trachea. Imaging modalities such as cardiac MRI, MRA, CT, and CTA are used to evaluate these malformations and their impact on surrounding structures.[5] 13.1 vascular ring

[5] Priya S, Thomas R, Nagpal P, et al. Congenital anomalies of the aortic arch. *Cardiovasc Diagn Ther*. 2018;8(Suppl 1):S26-S44. doi:10.21037/cdt.2017.10.15.

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Visceral Artery Aneurysms (PEDPVD-4.3)

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Imaging Indications for Visceral Artery Aneurysms

- Visceral artery imaging indications in pediatric individuals are identical to those for adult individuals. See **Visceral Artery Aneurysm (PVD-6.5)** in the General Peripheral Vascular Disease Imaging Guidelines.

Treatment is generally indicated for visceral aneurysms ≥2cm. Imaging of visceral artery aneurysms for initial evaluation or for treatment planning is medically necessary as follows:

- Initial imaging of calcifications seen on plain film imaging suspicious for visceral artery aneurysms (spleen, kidney, liver or intestines) with ultrasound (CPT® 76700, 76705, 93975, 93976, 93978, or 93979), **or** CTA Abdomen (CPT® 74175), **or** CT Abdomen with contrast (CPT® 74160)
- MRA Abdomen (CPT® 74185) without contrast in place of CTA or CT Abdomen in pediatric individuals or when there is a contraindication to contrast materials (i.e., renal insufficiency, contrast allergy, pregnancy)
- Ultrasound (CPT® 76700,76705, 93975, 93976, 93978, or 93979) **or** CTA Abdomen (CPT® 74175) **or** CT Abdomen with contrast (CPT® 74160) is considered medically necessary for further monitoring based on the intervals below or as determined by a vascular specialist or any provider in consultation with a vascular specialist:
  - Splenic artery aneurysms:
    - <20mm can be imaged every three years
    - 20mm to 29mm can be imaged annually
    - If ≥30mm, they should be referred for treatment, either stent, excision or splenectomy
  - For all other visceral artery aneurysms:
    - Initial evaluation with six-month follow-up for one year
    - Further follow-up annually if no significant enlargement is seen
- CTA Abdomen (CPT® 74175), MRA Abdomen (CPT® 74185), or CT Abdomen with contrast (CPT® 74160) is considered medically necessary following stent placement at:
  - 1 month
  - 6 months
  - 12 months
  - Then every year

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Evidence Discussion

Visceral artery aneurysms are rare but can be life-threatening if ruptured. Imaging indications in pediatric individuals are identical to those for adult individuals, with modalities such as MRA, CTA, and ultrasound being used for evaluation. Aneurysmal disease, besides primarily involving the large vessels, can also affect medium and smaller sized vessels. Visceral cases are uncommon and occasionally are associated with certain connective tissue and genetic disorders. They are often found incidentally on imaging. Ultrasound, CT, or CTA imaging may be indicated for surveillance in these cases. Due to the anatomic location of the visceral vessels, duplex ultrasound may have technical limitations. Consideration of best surveillance study should be decided on initial imaging and whether a certain modality is felt to provide diagnostic information. Example: some splenic arterial aneurysms may be diagnostic with US, but others may be obscured by bowel gas requiring CT/CTA.

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Infantile Hemangiomas (PEDPVD-5)

Guideline

General Considerations (PEDPVD-5.1)
Multiple Infantile Hemangiomas (PEDPVD-5.2)
PHACE(S) Syndrome (PEDPVD-5.3)
LUMBAR Syndrome (PEDPVD-5.4)
References (PEDPVD-5)

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General Considerations (PEDPVD-5.1)

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Most infantile hemangiomas do not require any imaging. Ultrasound with Doppler can be used when the diagnosis is uncertain, or with high-risk clinical considerations. Other general imaging considerations for other vascular neoplasms regarding MRI, MRA, CT, and CTA also apply to infantile hemangiomas. See **Imaging vascular tumors**.

- Multiple (5 or more) infantile hemangiomas can be associated with hepatic hemangiomas with risk potential for high-output cardiac failure and other risks see **Multiple Infantile Hemangiomas (PEDPVD-5.2)**.
- High-output cardiac failure can also be caused rarely by large cutaneous infantile hemangiomas. Affected infants may present with “failure-to-thrive”, a hyperdynamic precordium, tachycardia, bounding pulses with a widened pulse pressure, and a palpable thrill and/or audible bruit over the hemangioma. This is an indication for cardiac evaluation, including echocardiography (CPT® 93303 ordered with CPT® 93320 and CPT® 93325).
- Life threatening risk of airway obstruction is associated with infantile hemangiomas of the lower face (“beard distribution”), or of the anterior neck, or of the oral and/or pharyngeal mucosa.
- Location-associated functional impairment can be found with periocular infantile hemangiomas larger than 1 cm (impairing vision), or infantile hemangiomas involving lip(s) or oral cavity (impairing feeding).
- Ulceration can occur with profuse bleeding that can be life threatening.
- Disfigurement risk is increased with large (5 cm or larger) infantile hemangiomas, facial or scalp infantile hemangiomas, and breast infantile hemangiomas in female infants.
- An infantile hemangioma at least 2.5 cm in diameter overlying the lumbar spine or sacrum is an indication to do a spinal ultrasound (under 6 months of age) and/or MRI Lumbar Spine without contrast (CPT® 72148) or MRI Lumbar Spine without and with contrast (CPT® 72158).
- Infantile hemangiomas 5 cm or larger in size have an increased risk of extracutaneous structural abnormalities.
- Other high-risk indications include Syndromes or Associations with extracutaneous structural changes: for “PHACE(S) syndrome” See **Imaging Indications for PHACE(s) Syndrome**, and for “LUMBAR syndrome” See **Imaging Indications for LUMBAR Syndrome**.

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Evidence Discussion

Infantile Hemangiomas are the most common benign tumor of childhood, occurring in close to 5% of infants. Infantile Hemangiomas typically have a phase of rapid and significant growth between 1 month and 3 months of age; growth is usually completed by 5 months of age. Gradual involution then occurs, completed in 90% by age of 4 years but with residual skin changes frequently persisting. Though usually not needed for diagnosis, biopsy can be done when needed to identify unique markers not found on other vascular tumors.

When treatment is needed, imaging may be used to monitor response; consultation with a hemangioma specialist may be useful in guiding evaluation, treatment, and follow-up. The 2019 Clinical Practice Guideline of the American Academy of Pediatrics states "Unlike many diseases, management of IHs is not limited to 1 medical or surgical specialty. A hemangioma specialist may have expertise in dermatology, hematology-oncology, pediatrics, facial plastic and reconstructive surgery, ophthalmology, otolaryngology, pediatric surgery, and/or plastic surgery, and his or her practice is often focused primarily or exclusively on the pediatric age group."

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Multiple Infantile Hemangiomas (PEDPVD-5.2)

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- Multiple (5 or more) hemangiomas is an indication for Ultrasound with Doppler exam of the liver (CPT® 76700):
  - Initial imaging to look for hepatic hemangiomas
  - Repeat doppler ultrasound abdomen:
    - Monitor hepatic hemangiomas for progression
    - Monitor response to treatment.

Evidence Discussion

Multiple (5 or more) hemangiomas- though hepatic hemangiomas can be asymptomatic, they rarely can cause a high flow rate that can cause high-output cardiac failure and can be potentially fatal. "Diffuse" hepatic infantile hemangiomas are a rare subset of hepatic hemangiomas at high-risk for morbidity and mortality; affected infants usually present before 4 months of age with severe hepatomegaly, which can lead to lethal abdominal compartment syndrome with compromised ventilation, renal failure caused by renal vein compression, or compromise of inferior vena cava blood flow to the heart. Hepatic hemangiomas can also inactivate (via deiodination) thyroid hormones, causing risk of severe hypothyroidism.

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PHACE(S) Syndrome (PEDPVD-5.3)

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Imaging Indications for PHACE(s) Syndrome

Initial Imaging

- Initial diagnostic imaging is medically necessary when PHACE(S) syndrome is suspected by clinical findings including **any** of the following:
  - Infantile hemangioma ≥5 cm diameter on the face, scalp, and/or neck.
  - Infantile hemangioma <5cm on face, scalp, or neck in the setting of one or more major anomalies associated with PHACE(S) syndrome (i.e., coarctation of the aorta or midline ventral defect).
  - Infantile hemangioma on upper chest or proximal upper extremity ≥5 cm with no visible facial infantile hemangioma with PHACE(S) syndrome in the setting of one or more major anomalies associated with PHACE(S) syndrome (i.e., coarctation of the aorta or midline ventral defect).
  - Large intraorbital infantile hemangioma.
- Initial imaging for suspected PHACE(S) syndrome includes **any** of the following:
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
  - MRI Orbits without contrast (CPT® 70540) or MRI Orbits without and with contrast (CPT® 70543)
  - MRA Head without contrast (CPT® 70544) or MRA Head without and with contrast, (CPT® 70546)
  - MRA Neck may be done either without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549)
  - MRA Chest (CPT® 71555)
  - Transthoracic echocardiogram (CPT® 93303 with CPT® 93320 and CPT® 93325)
    - Cardiac MRI (CPT® 75557 or CPT® 75561) is medically necessary if abnormalities are identified on echocardiogram.
  - MRI Chest without contrast (CPT® 71550) or MRI Chest without and with contrast (CPT® 71552) is medically necessary if other clinical information or imaging shows involvement of the aorta.

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Surveillance

Repeat imaging is medically necessary when results will impact clinical management of the individual based on the results of the initial clinical and imaging assessment and any subsequent clinical changes, or when high-risk findings have been identified by clinical evaluation such as any of the following:

- Evidence of past arterial stroke
- Arterial stenosis or occlusions, with or without moyamoya-like vascular changes
- Structural brain changes, with neurosurgical evaluation clarifying the need for follow-up.
- Changes in the aortic arch, coarctation of the aorta, and congenital cardiac anomalies, with pediatric cardiology evaluation clarifying the need for follow-up see **Imaging and Surveillance per Congenital Lesion (PEDCD-2.4)** in the Pediatric Cardiac Imaging Guidelines

Evidence Discussion

"PHACE" (Posterior fossa malformations, Hemangiomas, Arterial anomalies, Coarctation of the aorta and Cardiac defects, and Eye abnormalities) syndrome or association (or "PHACE(S)" syndrome when also associated with sternal cleft and/or supraumbilical raphe) is frequently suspected when an infant has a large (5 cm in diameter or larger) infantile hemangioma of the face, scalp, or neck (risk of PHACE(S) Syndrome is then approximately 30%).

In rare cases, the face or scalp is not involved, with a large infantile hemangioma located on the torso and/or upper extremity instead. Cerebrovascular anomalies, present in more than 90% of individuals with PHACE(S) syndrome, are the most common extracutaneous feature of the syndrome, followed by cardiac anomalies (67%) and structural brain anomalies (about 50%).

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LUMBAR Syndrome (PEDPVD-5.4)

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Imaging Indications for LUMBAR Syndrome

- "LUMBAR syndrome" is reasonably suspected in a child with a large (5 or more cm in diameter) infantile hemangioma of any lumbosacral or perineal region or lower extremity. The following imaging is considered medically necessary:
  - Ultrasound spine (CPT® 76800) in infants up to 6 months of age, abdomen (CPT® 76700), and pelvis (CPT® 76856), with color Doppler.
  - MRI Lumbar Spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158) at 3 to 6 months of age, or earlier when either findings on an Ultrasound exam are inadequate or when requested by a hemangioma specialist or any provider in consultation with a hemangioma specialist.
  - MRI of other relevant spinal level (relevance based on proximity of observed infantile hemangiomas larger than 5 cm) without contrast or MRI of the relevant spinal level without and with contrast.
  - When ultrasound findings are inadequate and/or when recommended by a hemangioma specialist or any provider in consultation with a hemangioma specialist:
    - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72197) **and/or**
    - MRI Abdomen without contrast (CPT® 74181) or without and with contrast (CPT® 74183).
  - MRA Abdomen CPT® 74185 and/or Pelvis CPT® 72198, is considered medically necessary based on proximity of infantile hemangioma(s) at least 5 cm in diameter and/or other clinical evidence of vascular involvement, and/or when recommended by a hemangioma specialist or any provider in consultation with a hemangioma specialist.
  - Infantile hemangioma of the lower extremity that is at least 5 cm in diameter is an indication for MRI of the relevant portion of the lower extremity without contrast (CPT® 73718) or lower extremity without and with contrast (CPT® 73720) and/or lower extremity joint without contrast (CPT® 73721) or lower extremity joint without and with contrast (CPT® 73723).
  - When there is extensive lower extremity involvement with infantile hemangiomas the following are all considered medically necessary:
    - MRA (for both arterial and venous phase imaging) Abdomen
    - MRA Pelvis

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- MRA Lower extremities
- Note: this should be reported as CPT® 74185 and CPT® 73725; the CPT® code for MRA Pelvis (CPT® 72198) should not be included in this circumstance.

Evidence Discussion

The acronym "LUMBAR syndrome" refers to the association of lower body infantile hemangiomas of at least 5 cm in size (and other cutaneous defects), urogenital anomalies and ulceration, myelopathy (lipomyelocele/lipo-myelomeningocele and/or tethered spinal cord), bony deformities, anorectal malformations and arterial anomalies, and renal anomalies. Though not exclusively true, there is a general regional correlation between the location of the cutaneous large infantile hemangioma(s) with underlying structural anomalies.

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General Guidelines (PEDSP-1.0)

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Procedure Codes Associated with Spine Imaging (PEDSPINE)

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Procedure Codes Associated with Spine Imaging	
MRI	**CPT®**
MRI Cervical without contrast	72141
MRI Cervical with contrast	72142
MRI Cervical without and with contrast	72156
MRI Thoracic without contrast	72146
MRI Thoracic with contrast	72147
MRI Thoracic without and with contrast	72157
MRI Lumbar without contrast	72148
MRI Lumbar with contrast	72149
MRI Lumbar without and with contrast	72158
MRI Unlisted procedure (for radiation planning or surgical software)	76498
MRA	**CPT®**
MRA Spinal Canal	72159
CT	**CPT®**
CT Cervical without contrast	72125
CT Cervical with contrast	72126

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Procedure Codes Associated with Spine Imaging	
CT Cervical without and with contrast	72127
CT Thoracic without contrast	72128
CT Thoracic with contrast	72129
CT Thoracic without and with contrast	72130
CT Lumbar without contrast	72131
CT Lumbar with contrast	72132
CT Lumbar without and with contrast	72133
CT Pelvis without contrast	72192
CT Pelvis with contrast	72193
CT Pelvis without and with contrast	72194
CT Guidance for Placement of Radiation Therapy Fields	77014
CT Unlisted procedure (for radiation planning or surgical software)	76497
Nuclear Medicine	**CPT®**
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging: skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging: whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816

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Procedure Codes Associated with Spine Imaging	
Bone Marrow Imaging Limited Areas	78102
Bone Marrow Imaging Multiple Areas	78103
Bone Marrow Imaging Whole Body	78104
Nuclear Bone Scan Limited	78300
Nuclear Bone Scan Multiple Areas	78305
Nuclear Bone Scan Whole Body	78306
Bone Scan Three Phase	78315
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, single area (e.g., head, neck, chest, pelvis), single day imaging	78800
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, 2 or more areas (e.g., abdomen and pelvis, head and chest), 1 or more days imaging or single area imaging over 2 or more days	78801
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, single day imaging	78802
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), single area (e.g., head, neck, chest, pelvis), single day imaging	78803
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, requiring 2 or more days imaging	78804

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Procedure Codes Associated with Spine Imaging	
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (e.g., head, neck, chest, pelvis), single day imaging	78830
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), minimum 2 areas (e.g., pelvis and knees, abdomen and pelvis), single day imaging, or single area imaging over 2 or more days	78831
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, minimum 2 areas (e.g., pelvis and knees, abdomen and pelvis), single day imaging, or single area imaging over 2 or more days	78832
Ultrasound	**CPT®**
Ultrasound, spinal canal and contents	76800

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General Guidelines (PEDSP-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms, including a detailed history, physical examination with a thorough neurologic examination, appropriate laboratory studies and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) can serve as a pertinent clinical evaluation.
  - A thorough neurologic examination should include results of manual motor testing, specific dermatomal distribution of altered sensation, reflex examination, nerve root tension signs (e.g., straight leg raise test, slump test, femoral nerve tension test), and documentation of any specific radicular features.
- For those spinal conditions/disorders for which the Spine Imaging Guidelines require a plain x-ray of the spine prior to consideration of an advanced imaging study, the plain x-ray must be performed after the current episode of symptoms started or changed and results need to be available to the requesting provider of the advanced imaging study.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the spine is not supported. Advanced imaging of the spine should only be approved in individuals who have documented active clinical signs or symptoms of disease involving the spine.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the spine are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.

Health Equity Considerations

Health equity is the highest level of health for all individuals; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which individuals are born, grow, live, work, and age. Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include the following: safe housing, transportation, and neighborhoods; racism, discrimination, and violence; education, job opportunities, and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

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Pediatric Spine Imaging Age Considerations (PEDSP-1.1)

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- Many conditions affecting the spine in the pediatric population have different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are ≤18 years old should be imaged according to the Pediatric Spine Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Spine Imaging Guidelines should be imaged according to the General Spine Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Spine Imaging Guidelines, except where directed otherwise by a specific guideline section.

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Pediatric Spine Imaging Appropriate Clinical Evaluation (PEDSP-1.2)

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- See: **General Guidelines (PEDSP-1.0)**

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Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)

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- MRI
  - MRI is the preferred modality for imaging the pediatric spine unless otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize individual movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years), as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
    - Recent evidence-based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
      - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
    - If multiple body areas are supported by these guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
- CT
  - CT is generally inferior to MRI for imaging the pediatric spine, but has specific indications in which it is the preferred modality listed in specific sections of these guidelines.
    - CT is the imaging study of choice in the setting of trauma

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        - CT should not be used to replace MRI in an attempt to avoid sedation unless it is listed as a recommended study in a specific guideline section.
    - Myelogram with post-myelogram CT imaging is rarely indicated in children except in certain limited indications (usually requested after specialist consultation), including:
        - Evaluation of spine in individuals with fixation hardware which limits utility of MRI.
        - Severe congenital scoliosis with inconclusive MRI.
        - Evaluation of nerve root avulsion in patients with a brachial plexus injury and inconclusive MRI.
        - Evaluation of paraspinal cyst to assess continuity with the subarachnoid space.
        - Coding note: CT of appropriate spinal level with or without contrast may be appropriate. If the radiologist performs the myelogram the exam should be coded with contrast. If a clinician performs the myelogram the exam should be coded without contrast.
- Ultrasound
    - Spinal canal ultrasound (CPT® 76800) describes the ultrasonic evaluation of the spinal cord (canal and contents) and should not be reported multiple times for imaging of different areas of the spinal canal.
    - Do not use CPT® 76800 for intraoperative spinal canal ultrasound as CPT® 76998 (intraoperative ultrasonic guidance) is the appropriate code in this circumstance.
    - Spinal canal ultrasound (CPT® 76800) is generally limited to infants up to 6 months of age because of the bone mass surrounding the spinal cord limits evaluation of the intraspinal contents in older infants.
        - **Exception:** the persisting acoustic window in children with posterior spinal defects of spinal dysraphism enables spinal canal ultrasound to be performed at any age (see: **Spinal Dysraphism (PEDSP-4)**).
        - In general, additional imaging studies of the spine are not indicated in asymptomatic individuals with normal spinal ultrasound findings.
- Nuclear Medicine
    - Nuclear medicine studies are rarely used in the evaluation of the spine, but are medically necessary in the following circumstances:
        - Evaluation of suspected loosening of orthopedic hardware when recent plain x-ray is nondiagnostic (see: **Nuclear Medicine (MS-28)**).
            - Bone scan (CPT® 78315) or
            - Distribution Of Radiopharmaceutical Agent SPECT (CPT® 78803, or 78831) or
            - SPECT/CT (CPT® 78830 or CPT® 78832)
        - For suspected spondylolysis, see **Spondylosis (PEDSP-2.4)**

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  - Evaluation of back pain when no cause is demonstrated on MRI, see **Back and Neck Pain in Children Age 5 and Under (PEDSP-2.2)** or **Back and Neck Pain in Children Age 6 and Older (PEDSP-2.3)**
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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Pediatric Back and Neck Pain and Trauma (PEDSP-2)

Guideline

Introduction (PEDSP-2.1)
Back and Neck Pain in Children Age 5 and Under (PEDSP-2.2)
Back and Neck Pain in Children Age 6 and Older (PEDSP-2.3)
Spondylolysis (PEDSP-2.4)
Spine Pain Due to Infectious Causes (PEDSP-2.5)
Spine Pain Related To Trauma and Painless Spine Trauma (PEDSP-2.6)
References (PEDSP-2)

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Introduction (PEDSP-2.1)

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- Currently, only about 20% of back pain in children over age 5 is from a discoverable cause. Scoliosis, spondylitic disorders, Scheuermann disease, tumor, and trauma are the most common causes.
- Back pain in children under age 5 is uncommon and often reflects underlying serious disease when present.
- Disc herniations are rare in children, but become more frequent as activity increases during adolescence.

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Back and Neck Pain in Children Age 5 and Under (PEDSP-2.2)

- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- Advanced imaging is medically necessary in all individuals in this age group except those with mild and transient back pain.
  - MRI of the symptomatic spinal region is medically necessary.
    - Individuals in this age group will require sedation to complete MRI imaging. See: **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)** for contrast and body area considerations.
  - CT without contrast of the symptomatic spinal region is medically necessary when:
    - plain x-rays suggest an isolated vertebral bone abnormality without any concern for spinal canal or cord abnormalities (which is rare in this age group)
    - a recent MRI does not provide sufficient detail of the bony anatomy to allow for acute patient care decision making
  - Bone scan is medically necessary for evaluation of suspected spinal fracture when x-ray is negative using any of the following CPT® code combinations:
    - CPT® 78300, CPT® 78305, or CPT® 78306 as a single study
    - CPT® 78315 or CPT® 78803 can be approved as a single study when stress fracture is suspected.
  - Bone scan is medically necessary for evaluation of suspected spondylolysis, or if recent spine MRI is inconclusive using any of the following CPT code combinations: SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period.
    - CPT® codes: CPT® 78300, CPT® 78305, CPT® 78306, CPT® 78315, or CPT® 78803 as a single study
    - CPT® 78305 and CPT® 78803 concurrently
    - CPT® 78306 and CPT® 78803 concurrently

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Back and Neck Pain in Children Age 6 and Older (PEDSP-2.3)

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Radicular back and neck pain is common in adult patients but is uncommon in adolescents and rare in children.

- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination including results of manual motor testing, the specific dermatomal distribution of altered sensation, reflex examination, and nerve root tension signs (e.g., straight leg raise test, slump test, femoral nerve tension test) and documentation of any specific radicular features, should be performed prior to considering advanced imaging.
- X-rays, while not required prior to conservative treatment, must be obtained before advanced imaging can be approved.
  - The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study.
- Advanced imaging is medically necessary following x-ray when one or more of the following pediatric “red flags” are present:
  - Accompanying systemic symptoms (fever, weight loss, etc.)
  - Functional disability (daily limitation in normal activities because of pain)
  - Pain which is extremely severe or worse at night
  - Constant or radicular pain lasting ≥4 weeks
  - Pain which worsens despite an attempt at symptomatic treatment
  - Neurological symptoms or abnormal neurological examination findings
  - An established diagnosis of cancer other than leukemia
  - Abnormal x-rays
  - Individuals having undergone spinal surgery
  - Associated bowel or bladder dysfunction
- In the absence of any “red flags”, a 4-week trial of provider-supervised conservative treatment (after the current set of symptoms or physical exam findings started or changed) should be attempted before advanced imaging is medically necessary.
  - It can be assumed that children who are being evaluated by a pediatric spine surgeon have failed a reasonable trial of conservative treatment under the care of the primary care provider, as this is by far the most common reason for such referrals.

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- X-rays of the involved regions should be obtained prior to advanced imaging in individuals with “red flag” findings, or who remain symptomatic after a 4-week trial of provider-supervised conservative treatment.
  - The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study.
- MRI without contrast of the symptomatic spinal region is medically necessary for the evaluation of pediatric spine pain, and should be approved unless one of the following conditions applies, in which case MRI without and with contrast is medically necessary:
  - Fever (≥100° F)
  - Clinical suspicion of infection (discitis, osteomyelitis, paraspinous or epidural abscess)
  - Physical examination or plain x-ray suggests a mass lesion
  - New or worsening pain in a patient with an established diagnosis of cancer
- CT without contrast of the symptomatic spinal region is medically necessary when:
  - the request is for re-evaluation of a known vertebral bony disorder
  - plain x-rays show spondylotic changes or suggest an isolated vertebral bone abnormality without any concern for spinal canal or cord abnormalities (which is rare in this age group)
  - a recent MRI does not provide sufficient detail of the bony anatomy to allow for acute individual care decision making
- Bone scan is medically necessary for evaluation of suspected spinal fracture when x-ray is negative, or if recent MRI is inconclusive using any of the following CPT® code combinations:
  - CPT® codes: CPT® 78300, CPT® 78305, or CPT® 78306 as a single study
  - CPT® 78315 or CPT® 78803 as a single study when stress fracture is suspected

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Spondylolysis (PEDSP-2.4)

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Most cases of childhood spondylolysis are believed to be caused by repeated microtrauma, resulting in stress fracture of the pars interarticularis. Heredity is also believed to be a factor in some cases. It is the most common cause of low back pain in children older than age 10.

- Activity modification, NSAID treatment, physical therapy, and/or immobilization with various braces are the initial treatments for symptomatic individuals.
- Surgical treatment is only recommended for individuals with disabling symptoms that have not responded to non-surgical care.
- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- Spondylolysis is screened with plain x-rays.
  - MRI without contrast of the symptomatic spinal level is medically necessary to evaluate for stress reaction in bone and visualizing nerve roots if symptoms have continued despite a provider-directed 4-week course of conservative care after the current set of symptoms or physical exam findings started or changed, or if there is a documented need for preoperative planning.
  - If additional imaging is needed because of radiological uncertainty or associated spondylolisthesis, SPECT Radiopharmaceutical Localization Imaging (CPT® 78803) or SPECT/CT (CPT® 78830) is medically necessary to identify stress reaction in spondylolysis cases which are radiographically occult. Bone scan has been demonstrated to be superior to MRI in detecting active spondylolysis.
  - SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period. CT without contrast of the symptomatic spinal level is medically necessary to provide detailed evaluation of bony anatomy, if there is a documented need for preoperative planning. CT scans have been considered the criterion standard for characterizing fractures and for detailing bone morphology and anatomy.

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Spine Pain Due to Infectious Causes (PEDSP-2.5)

- Entities including, but not limited to, discitis and vertebral osteomyelitis, typically present with sudden onset of back pain, fever, and elevated white blood cell count, occurring most commonly in the first decade of life.
- A detailed history and physical examination with thorough neurologic examination should be performed initially.

Initial Imaging Studies

- Plain x-rays should be performed initially.
  - The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study.
- MRI without and with contrast of the symptomatic spinal level is very sensitive at detecting early changes and is medically necessary when discitis or osteomyelitis is clinically suspected.
- Nuclear medicine imaging also can be positive as soon as 1 to 2 days after the onset of symptoms. Any of the following studies are medically necessary for initial evaluation of suspected osteomyelitis:
  - Bone scan (one of CPT® codes: CPT® 78300, 78305, 78306, or 78315)
  - Nuclear Bone Marrow imaging (one of CPT® codes: CPT® 78102, 78103, or 78104)
  - Radiopharmaceutical inflammatory imaging (one of CPT® codes: CPT® 78800, 78801, 78802, 78803, or 78804)
  - SPECT (CPT® 78831)
  - SPECT/CT (CPT® 78830, or CPT® 78832)

Follow-Up Imaging Studies

- Follow-up plain x-rays may show disc space narrowing and bony changes of osteomyelitis.
- MRI without and with contrast of the symptomatic spinal level or CT with contrast (including myelography) is medically necessary for follow-up evaluating bony changes of osteomyelitis or concern for epidural abscess.
- Any of the following studies are medically necessary for evaluation of response to treatment in established osteomyelitis:
  - Bone scan (one of CPT® codes: CPT® 78300, 78305, 78306, or 78315)

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- Nuclear Bone Marrow imaging (one of CPT® codes: CPT® 78102, 78103, or 78104)
- Radiopharmaceutical localization imaging (one of CPT® codes: CPT® 78800, 78801, 78803, 78830, 78831, or 78832)

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Spine Pain Related To Trauma and Painless Spine Trauma (PEDSP-2.6)

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- Imaging evaluation of traumatic spine injury in children is generally directed based on clinical examination. 60% to 80% of all spinal injuries in children involve the cervical spine as opposed to the thoracic spine and lumbar spine. Common causes are motor vehicle accidents, falls, and sports-related injuries.
- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, should be performed prior to considering advanced imaging.
- When advanced imaging is appropriate, MRI without contrast or CT without contrast of the involved level is medically necessary as discussed in **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)**
  - If the initial CT or MRI study is considered inconclusive, an exam of the other modality is medically necessary if needed to direct clinical management.

Cervical Spine

- The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study
- Advanced imaging of the cervical spine is medically necessary in children under 3 years of age following a relevant x-ray when one or more of the following “red flags” are present:
  - Glasgow Coma Scale <14
  - Individual does not open eyes regardless of stimulus
  - Motor vehicle collision
- Advanced imaging of the cervical spine is medically necessary in children ≥3 years of age following a relevant x-ray when one or more of the following “red flags” are present:
  - Altered mental status
  - Focal neurologic findings
  - Neck pain
  - Torticollis not present prior to trauma
  - Substantial torso injury
  - Diving or head-first injury
  - High speed motor vehicle collision
  - Predisposing conditions, e.g., Down syndrome

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- Advanced imaging of the cervical spine is not medically necessary in children older than 2 years of age if they meet ALL of the following criteria:
  - Absence of posterior midline cervical pain
  - Absence of focal neurologic deficit
  - Normal level of alertness
  - No evidence of intoxication
  - Absence of other clinically apparent pain which could distract patient from the pain of a cervical injury

Thoracolumbar Spine

- Advanced imaging of the thoracolumbar spine is medically necessary in children following a recent x-ray when x-rays are inconclusive, or there is an abnormal neurological examination.

Suspected Physical Child Abuse

- In children with suspected physical child abuse and documented findings suggesting abuse (e.g., fractures on skeletal survey or other clinical indicators), MRI Cervical (CPT® 72141), Thoracic (CPT® 72146), and Lumbar (CPT® 72148) Spine without contrast are medically necessary to search for associated abnormalities.
  - If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast is medically necessary. See: **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)**

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Kyphosis and Scoliosis (PEDSP-3)

Guideline

Juvenile Thoracic Kyphosis (Scheuermann Disease) (PEDSP-3.1)
Scoliosis (PEDSP-3.2)
References (PEDSP-3)

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Juvenile Thoracic Kyphosis (Scheuermann Disease) (PEDSP-3.1)

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- This condition is also known as Scheuermann Kyphosis, and these individuals generally present with chronic and recurrent back pain.
- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- X-rays will typically show anterior wedging in three or more adjacent vertebral bodies.
  - Lower thoracic kyphosis from developmental vertebral wedging with thoracic kyphosis varying between 20° and 45° should be identified by plain x-rays before considering advanced imaging.
  - MRI is not an effective diagnostic modality and is therefore not medically necessary for this condition since the incidence of false positive vertebral changes in normal individuals is high.
- MRI Thoracic Spine without contrast (CPT® 72146) is medically necessary preoperatively to rule out any associated spinal cord problems.
- MRI Lumbar Spine without contrast (CPT® 72148) is medically necessary preoperatively to rule out any associated spinal cord conditions when there is clinical or radiographic evidence of lumbar abnormalities.

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Scoliosis (PEDSP-3.2)

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- Scoliosis is an abnormal lateral curve of the thoracic or thoraco-lumbar spine in the frontal plane. A small lateral curve in a skeletally mature person is not uncommon and generally does not require further investigation.
- Using the Cobb technique for measuring these curves, a curve of under 10˚ is normal, a curve from 10˚ to 20˚ is mildly abnormal, a curve over 20˚ is significantly abnormal, and a curve over 40˚ is severely abnormal.
- Most individual with significant scoliosis have some element of kyphosis as well.
  - There are many ways of classifying scoliosis. These guidelines will classify scoliosis as congenital, idiopathic, and neuromuscular scoliosis.
- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, detailed examination of the spine in different body positions, and plain radiography should be performed prior to considering advanced imaging.
  - Standing posteroanterior (PA) and lateral x-rays of the spine are the initial imaging studies and are used for follow-up. If anteroposterior (AP) x-rays are to be performed, breast shields should be used to reduce breast radiation exposure.
  - Spine surgical specialists sometimes appropriately request both MRI and CT together for preoperative planning of scoliosis surgery.
    - In addition, MR and CT are useful to identify an underlying cause of scoliosis, such as congenital and developmental anomalies.
    - MR or CT Spine is medically necessary postoperatively when recent postoperative x-rays are inconclusive for managing individual treatment.
      - Individuals with severe scoliosis may have compromised lung development. CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) is medically necessary in the perioperative period as well as 2 and 5 years postoperatively to assess lung growth.

Congenital Scoliosis

Cases are recognized in infancy or early childhood. Most cases arise from anomalies of vertebral development, and many are associated with anomalies of the genitourinary system or of other organs.

- In infants under 6 months of age spinal ultrasound (CPT® 76800) is medically necessary after initial imaging with plain x-rays.
- MRI Cervical (CPT® 72141), Thoracic (CPT® 72146), and Lumbar (CPT® 72148) Spine without contrast are medically necessary to search for underlying anomalies.

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  - If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may, is medically necessary. See: **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)**
- MRI Brain without and with contrast is medically necessary if the clinical evaluation or preliminary imaging studies suggest an associated intracranial anomaly.
- Renal ultrasound (CPT® 76770 or CPT® 76775) is medically necessary since nearly one-third of individuals also have genitourinary anomalies.
  - CT, MRI, or nuclear medicine studies of the genitourinary tract are medically necessary if the ultrasound is abnormal.

Idiopathic Scoliosis

Idiopathic scoliosis is the most common form of pediatric scoliosis and is divided into infantile (0-3 years of age), juvenile (4-9 years of age), and adolescent (10-17 years of age). Idiopathic scoliosis is defined as having no underlying structural abnormality or accompanying syndrome.

- The following clinical features are associated with an increased risk of underlying vertebral or spinal cord abnormality:
  - Associated back pain
  - Age younger than 10 years
  - Neurological abnormalities on examination or neurological symptoms
  - Left sided curve (concave to right)
  - Absence of apical segment lordosis/kyphosis
  - Rapid curve progression (>1 degree per month)
  - Pes Cavus (see: **Occult Spinal Dysraphism (PEDSP-4.3)**
  - Double curves or high thoracic curves
  - Kyphosis
  - Spinal x-ray abnormalities other than the curve itself (widened spinal canal, dysplastic changes in spine or ribs, etc.)
  - Midline spinal cutaneous markers (esp. sacral) such as dermal tracts, tufts of hair, skin tags, etc.
  - Abnormal number or size of café au lait spots (neurofibromatosis)
- MRI Cervical (CPT® 72141), Thoracic (CPT® 72146), and Lumbar (CPT® 72148) Spine without contrast are the medically necessary studies for the evaluation of scoliosis when any of the above clinical features is present or if imaging is requested for individuals who are being actively evaluated for corrective surgery.

Neuromuscular Scoliosis

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Scoliosis can result from many disorders of the nervous system. In some conditions, including (but not limited to) cerebral palsy, muscular dystrophy, and spinal muscular atrophy, associated scoliosis may develop over time.

The appropriate spinal level, modality, and contrast level of advanced imaging will depend on the nature of the underlying disease.

- MRI without contrast or without and with contrast or CT without contrast of the cervical, thoracic, and/or lumbar spine is medically necessary in these individuals with painful neuromuscular scoliosis, or when they are actively being evaluated for spinal deformity corrective surgery.
- Bone scans (one of CPT® codes: CPT® 78300, CPT® 78305, CPT® 78306, or CPT® 78315) are medically necessary to evaluate cases of painful scoliosis and to identify tumors or infections. They are more sensitive than plain radiography.
  - Post-surgical considerations are similar to adult post-operative indications (see: **Post-Operative Spinal Disorders (SP-15)** in the General Spine Imaging Guidelines) except as follows:
    - Post-operative CT Chest without contrast (CPT® 71250) with 3D reconstruction is medically necessary for lung volume measurement in children with early onset scoliosis, (e.g., congenital/thoracogenic type), due to risk of restrictive lung disease and thoracic insufficiency syndrome which occur from failure of spine and chest to support normal lung growth.

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Spinal Dysraphism and Tethered Spinal Cord (PEDSP-4)

Guideline

Introduction (PEDSP-4.1)
Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)
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Introduction (PEDSP-4.1)

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Spinal Dysraphism

- Spinal dysraphism refers to a group of disorders characterized by incomplete or absent fusion of posterior midline structures. This includes a range of congenital and/or developmental anomalies of the spinal cord and associated spinal structures that can affect any level of the spine, but most commonly the lumbosacral region.
- Based on clinical classification, dysraphism is grouped into two categories:
  - Open dysraphism (spina bifida aperta), which are non-skin-covered, open neural tube defects (myelomeningocele).
  - Occult spinal dysraphism (also called closed spinal dysraphism), which includes skin-covered defects (either with or without an associated subcutaneous mass).

Normal position of spinal cord

- In newborns, the spinal cord should terminate (at the conus medullaris) at L2-3 or higher.
- By 3 months of age, the conus should lie at or above the L2 level.
- Afterwards, in normal infants and children, the conus medullaris should be positioned at L1-2.
- Of note, however, in premature infants, the conus medullaris may be located at the mid L3-level.
  - If such a finding on an initial spinal ultrasound results in uncertainty as to whether cord termination is low, repeat spinal ultrasound (CPT® 76800) is medically necessary in 4 to 6 weeks, since a normal cord will have “moved” higher within the spinal canal by this time.

Tethered cord

- Tethering is certain when the cord terminates at or below L4 and there is other supporting evidence of tethering such as limited spinal cord pulsatility, posterior positioning in the spinal canal, thick filum terminale, intraspinous mass, or lipoma.
- If the conus terminates at a normal position (at L2-3 under 3 months of age, at L2 by 3 months of age, at L1-2 in older infants and children), the cord may still be tethered by an abnormal structure. Such tethering of the spinal cord can be found in some (but not all) individuals with occult spinal dysraphism. Abnormalities can be found in both lumbosacral and thoracic regions and are often associated with spinal lipomas in either region.
- Open spinal dysraphism is frequently associated with tethering of the spinal cord; symptoms of or findings from that tethering may manifest initially or may increase

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after the newborn period and the initial imaging evaluation. See: **Open Dysraphism (PEDSP-4.4)**.

"Tethered cord Syndrome"

- "Tethered Cord Syndrome" refers to symptoms and abnormal physical findings (such as low back or leg pain, decreased or absent lower extremity reflexes, urinary urgency, urinary incontinence, bowel incontinence, and constipation) that arise when a pathologic attachment causes abnormal spinal tension (increased by axial growth), with ensuing pathophysiologic effects. Some of these patients do have an abnormally low conus medullaris; other patients have other spinal abnormalities (such as spinal dysraphism) that causes the spinal cord to be abnormally tethered. Other patients with spinal dysraphism who may present with symptoms or findings suggestive of "Tethered Cord Syndrome" may have those clinical manifestations caused by primary dysplasia of neural tissue, instead of being caused by abnormal tethering. See: **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**.
- Not all anatomically tethered spinal cords result in symptoms of "Tethered Cord Syndrome."

Imaging Studies to Evaluate Suspected Occult Spinal Dysraphism and/or Tethered Cord

- Plain x-rays are not indicated for suspected occult spinal dysraphism and/or tethered cord.
- Spina bifida occulta, an incomplete fusion of the posterior lumbosacral bony elements (present in in about 25% of people), is often discovered as an incidental finding on x-rays and other imaging exams. In asymptomatic individuals it is of no consequence, and is not an indication for further imaging.
- A plain spine x-ray finding suggesting an absent or distorted pedicle (the "winking owl sign") can be indicative of occult spinal dysraphism, for which an initial MRI without contrast or MRI without and with contrast of the appropriate spinal level is medically necessary.
- When indicated (See: **Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)**, **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**, and **Open Dysraphism (PEDSP-4.4)** for indications), the following imaging is medically necessary:
  - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age, in premature infants whose "corrected age" (subtracting the number of weeks of prematurity from the infant's actual age) is less than or equal to 6 months, or in older individuals with open spinal dysraphism (see: **Open Dysraphism (PEDSP-4.4)**).
  - In a term infant, the diagnosis of tethered cord is likely if the conus terminates below the L2-L3 disc space. Of note, however, in premature infants, the conus medullaris may be located at the mid L3-level; if there is uncertainty as to whether

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cord termination is low in a premature infant, repeat spinal ultrasound (CPT® 76800) is medically necessary in 4 to 6 weeks, since a normal cord will have "moved" higher within the spinal canal by this time.

- MRI Cervical, Thoracic, and Lumbar spine without contrast (CPT® 72141, 72146, and 72148) or without and with contrast (CPT® 72156, 72157, and 72158) is medically necessary for initial evaluation in individuals older than 6 months of age.
  - MRI is medically necessary at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or when MRI imaging prior to 6 months of age has been ordered by (or in consultation with) an appropriate specialist for an indication from **Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)**, **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**, or **Open Dysraphism (PEDSP-4.4)**.
- The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually ordered by (or after consultation with) an appropriate specialist.
- Postoperative MRI is not done routinely but is medically necessary if there are recurrent symptoms or findings suggesting recurrent tethering or other deterioration. Contrast level per ordering specialist.
- A complete abdominal ultrasound (CPT® 76700) or retroperitoneal ultrasound (CPT® 76770) is medically necessary as an initial evaluation for patients with newly diagnosed neurogenic bladder, myelomeningocele (open spinal dysraphism), or occult spinal dysraphism.
  - A complete retroperitoneal ultrasound (CPT® 76770) is medically necessary every 6 to 12 months for follow-up/surveillance for any of the above conditions.
- CT of the effected spinal level is medically necessary for surgical planning when a complex bony deformity of the spine is present, or when the Guidelines support doing MRI of the spine in an individual for whom MRI is contraindicated.

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Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)

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- More than 80% of individuals with occult spinal dysraphism and/or tethered spinal cord will have a cutaneous lesion overlying the lower spine.
- Spine imaging is not medically necessary in the following situations:
  - Pilonidal cysts below the level of the intergluteal fold.
    - For discussion of imaging in pilonidal cysts, see: **Pilonidal Cyst (PV-21.4)** in the Pelvis Imaging Guidelines
  - Non-specific darkened areas of skin over the sacrum (such as dermal melanosis) unless there are other associated midline cutaneous abnormalities
  - Occult bony dysraphism incidentally noted on x-ray
- Screening with advanced imaging is medically necessary in the following clinical conditions which are associated with an increased risk of underlying spinal dysraphism:
  - Spinal dimples (midline soft tissue depression over the spine); or deviated or split (bifid) gluteal cleft
    - Spinal ultrasound (CPT® 76800) is medically necessary for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Follow-up of a normal screening spinal ultrasound with ultrasound is not medically necessary.
    - MRI of the involved spinal level without contrast or without and with contrast is medically necessary for initial evaluation in individuals older than 6 months of age. MRI is medically necessary at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or if ordered by (or in consultation with) an appropriate specialist.
    - A screening MRI is medically necessary after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not medically necessary.
  - Dermal sinuses overlying the lumbar, thoracic, or cervical spine, and sacral dermal sinuses, whether manifested by a dermal sinus tract (a small opening in the skin, which leads into a narrow duct; it may be associated with protruding hairs) or a dermal cyst. They may be associated with an overlying or nearby hairy patch or vascular nevus
    - Spinal ultrasound (CPT® 76800) is medically necessary for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up

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to 6 months of age). Follow-up of a normal screening spinal ultrasound is not medically necessary.
    - MRI of the involved spinal level without contrast or without and with contrast is medically necessary if an ultrasound shows abnormalities other than a cutaneous dermal cleft, if ordered after 6 months of age, or at a younger age if ordered by (or in consultation with) an appropriate specialist.
    - A screening MRI is medically necessary after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not medically necessary.
- Subcutaneous midline masses (including cysts and lipomas) at any level.
    - Plain x-rays are not required to approve other imaging for midline masses overlying the spine when occult spinal dysraphism and/or tethered cord is suspected.
    - Spinal ultrasound (CPT® 76800) is medically necessary for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age), but MRI of the involved spinal level without contrast or without and with contrast is the preferred medically necessary initial imaging for midline masses overlying the spine. Repeat ultrasound follow-up of a normal screening spinal ultrasound is not medically necessary.
    - MRI of the involved spinal level without contrast or without and with contrast is medically necessary for initial evaluation in patients older than 6 months of age. MRI is medically necessary at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or if ordered by (or in consultation with) an appropriate specialist.
    - A screening MRI is medically necessary after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not medically necessary.
- Caudal extensions (including tail-like appendages), midline skin tags, abnormal patches of hair over the spine at any level, infantile hemangiomas overlying any spinal level, and complex midline birthmarks above the upper sacral region.
    - Spinal ultrasound (CPT® 76800) is medically necessary for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Repeat ultrasound follow-up of a normal screening spinal ultrasound is not medically necessary.
    - MRI of the involved spinal level without contrast or without and with contrast is medically necessary for initial evaluation in individuals older than 6 months of age. MRI is medically necessary at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or if ordered by (or in consultation with) an appropriate specialist.

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        - A screening MRI is medically necessary after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not medically necessary.
    - Café au lait spots are a marker for type 1 neurofibromatosis
        - See imaging indications in **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** and/or **Neurofibromatosis (PEDPN-2)**.

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Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)

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- Imperforate anus
- VACTERL (vertebral malformations, anal atresia, cardiac anomalies, tracheo-esophageal fistula, renal abnormalities, and limb defects) syndrome
- Currarino triad (sacral dysgenesis, presacral mass, anorectal malformation), OEIS (omphalocele, exstrophy, imperforate anus, spinal defects) syndrome
- Caudal regression syndrome
- Sacral agenesis (when 2 or more of the sacral vertebral bodies are absent; about 20% of children with sacral agenesis are not detected prior to age of 3 years).
- For all of the above conditions, the following imaging is medically necessary:
  - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Repeat ultrasound follow-up of a normal screening spinal ultrasound is not medically necessary.
  - MRI Lumbar Spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158); and/or MRI Pelvis without contrast (CPT® 72195) or MRI Pelvis without and with contrast (CPT® 72197).
  - Appropriate MRI (or other modality) imaging (including contrast level) of any other spinal level will depend on the nature of the underlying disease, usually ordered by (or in consultation with) an appropriate specialist.
  - Follow-up of a normal screening MRI imaging study is not medically necessary, but an initial MRI can be approved if the first screening study was an ultrasound.
  - Postoperative MRI is not done routinely but is medically necessary if there are recurrent symptoms or findings suggesting recurrent tethering. Contrast level per ordering specialist.
- For Rubinstein-Taybi syndrome (gait abnormalities, short stature, short limbs, characteristic facies, developmental delay, tethered spinal cord), the following imaging is medically necessary:
  - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Repeat ultrasound follow-up of a normal screening spinal ultrasound is not medically necessary.
  - MRI Lumbar spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158)

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- Appropriate MRI (or other modality) imaging (including contrast level) of any other spinal level will depend on the nature of the underlying disease, usually ordered by (or in consultation with) an appropriate specialist.
- Follow-up of a normal screening MRI imaging study is not medically necessary, but an initial MRI can be approved if the first screening study was an ultrasound.

- For individuals with known DiGeorge Syndrome (22q11.2 deletion syndrome), the following imaging is medically necessary when tethered cord syndrome or occult spinal dysraphism is suspected:
  - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age
  - MRI Lumbar Spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158)
  - Appropriate MRI (or other modality) imaging (including contrast level) of any other spinal level will depend on the nature of the underlying disease, usually ordered by (or in consultation with) an appropriate specialist.
  - Follow-up of a normal screening MRI imaging study is not medically necessary, but an initial MRI can be approved if the first screening study was an ultrasound.
- MRI of the involved spinal level without contrast or without and with contrast is medically necessary for neurologic related symptoms and physical exam findings suggestive of occult spinal dysraphism, tethered cord syndrome, and/or low lying conus medullaris when any of the following are present:
  - Asymmetry of the feet, with one smaller foot, a high arch, and/or clawing of the toes. This is sometimes called the “neuroorthopedic syndrome”, and is associated with lack of an ipsilateral ankle jerk deep tendon reflex and calf atrophy.
  - Cavus foot (also called pes cavus or pes cavovarus)
  - Toe walking, when associated with upper motor neuron signs including hyperreflexia, spasticity, and positive Babinski sign
  - Ataxia (see: **Ataxia (PEDHD-20)**)
  - Absent perineal sensation
  - Lower urinary tract dysfunction, including urinary urgency or urinary incontinence. Though not a requirement for advanced imaging, some of these patients will have had abnormal urodynamic studies (such as cystometrography and/or sphincter electromyography).
  - Constipation, especially if there are abnormal physical exam findings related to the spine (such as lower extremity weakness, decreased lower extremity tone, abnormal lower extremity reflexes, a tuft of hair over the spine or covering a pilonidal dimple, a sacral dimple, gluteal cleft deviation, or absent anal or cremasteric reflex), failure of maximal laxative therapy (see: **Constipation, Diarrhea, and Irritable Bowel Syndrome (PEDAB-12)**) and/or bowel incontinence, when tethered cord syndrome or occult spinal dysraphism is suspected as the cause

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  - Back or leg pain when tethered cord syndrome or occult spinal dysraphism is suspected as the cause. In this setting, neither a plain x-ray of the spine nor a recent period of provider directed conservative treatment is required to approve an MRI spine).
- See: **Myelopathy (SP-7.1)**, **Myelopathy (PEDSP-6)**, and **Developmental Motor Delay (PEDHD-19.3)** for spinal cord involvement suspected in individuals with developmental motor delay.

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Spinal Dysraphism (PEDSP-4.4)

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- Clinically significant dysraphism includes findings ranging from complex vertebral anomalies to myelomeningocele.
- Dysraphism is categorized into 2 major groups:
  - Open Dysraphism - lack of skin covering with exposed neural elements
  - Closed Dysraphism - skin covered
- It is rare to perform MRI in neonates with open dysraphism as the diagnosis is usually made with obstetric ultrasound and confirmed with visual inspection
  - MRI of the entire spine is medically necessary for preoperative planning if ordered by a specialist.
- MRI Brain without contrast (CPT® 70551) or with and without contrast (CPT® 70553) is medically necessary in all cases of open dysraphism as Chiari II malformation will be present
- Closed Dysraphism
  - MRI of the entire spine without contrast or without and with contrast is medically necessary at the time of initial diagnosis.
    - MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) or CT without contrast of the brain (CPT® 70450) is medically necessary in cases with associated hydrocephalus, signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
    - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72196) is medically necessary once if there are clinical signs of pelvic malformation or anorectal anomaly.
  - MRI Cervical, Thoracic, and Lumbar spine without contrast (CPT® 72141, 72146, 72148) or without and with contrast (CPT® 72156, 72157, 72158) is medically necessary when ordered for preoperative planning.
  - Spinal canal ultrasound (CPT® 76800) is medically necessary as an alternative to MRI, if requested, in individuals with open dysraphism as the posterior bony defect provides an acoustic window for ultrasound.
  - MRI of the appropriate spinal level without contrast or without and with contrast is medically necessary when there are new and/or worsened neurologic symptoms and/or physical exam findings suggestive of new or worsened tethering of the spinal cord, such as any of the following:
    - New or worsened cavus foot
    - New or worsened toe walking and/or upper motor neuron signs (including hyperreflexia, spasticity, and positive Babinski sign)
    - New or worsened leg weakness or numbness or difficulty in ambulation

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- New or worsened loss of perineal sensation
- New or worsened lower urinary tract dysfunction (including urinary urgency or urinary incontinence, or new or worse changes on diagnostic urodynamic studies)
- New or worsened constipation
- New or worsened pain in the back or legs suspected to have been caused by tethering of the spinal cord
  - MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) or CT without contrast of the brain (CPT® 70450) is medically necessary in cases with associated hydrocephalus, signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
  - MRI Pelvis without contrast (CPT® 72195) or without and with contrast (CPT® 72196) is medically necessary once if there are clinical signs of pelvic malformation or anorectal anomaly.

◦ The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

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Tethered Cord (PEDSP-5)

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- See: **Spinal Dysraphism and Tethered Spinal Cord (PEDSP-4)**

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Myelopathy (PEDSP-6)

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Myelopathy (PEDSP-6)

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Myelopathy imaging indications in pediatric individuals are similar to those for adult individuals. See: **Myelopathy (SP-7)** in the Spine Imaging Guidelines and/or **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**

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Other Congenital and Pediatric Spine Disorders (PEDSP-7)

Guideline

General Guidelines - Other Congenital and Pediatric Spine Disorders (PEDSP-7.0)
Achondroplasia (PEDSP-7.1)
Inflammatory Spondylitis (PEDSP-7.2)
Atlantoaxial Instability in Trisomy 21 (Down Syndrome) (PEDSP-7.3)
Basilar Impression (PEDSP-7.4)
Chiari Malformation (PEDSP-7.5)
Klippel-Feil Anomaly (Congenital Fusion of Cervical Vertebrae) (PEDSP-7.6)
Marfan Syndrome (PEDSP-7.7)
Neurofibromatosis (PEDSP-7.8)
Von Hippel-Lindau Syndrome (VHL) (PEDSP-7.9)
References (PEDSP-7)

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General Guidelines - Other Congenital and Pediatric Spine Disorders (PEDSP-7.0)

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- Many congenital spine disorders also affect adults as survival continues to improve for these individuals. Adults with disorders covered in this section may follow these guidelines except where contraindicated by specific statements in the general imaging guidelines.

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Achondroplasia (PEDSP-7.1)

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- The diagnosis of achondroplasia is made clinically. Individuals with achondroplasia are at risk for hydrocephalus as well as myelopathy from spinal stenosis with increasing age.
- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- MRI without contrast or without and with of the symptomatic spinal region is medically necessary when new or worsening clinical symptoms suggest achondroplasia-related spinal stenosis.
- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is medically necessary when new or worsening clinical symptoms suggest hydrocephalus.

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Inflammatory Spondylitis (PEDSP-7.2)

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- Except as listed below, imaging considerations in pediatric and adult individuals are identical for this condition and these individuals should be imaged according to **Inflammatory Spondylitis (SP-10.2)**.

For pediatric individuals with juvenile idiopathic arthritis, the following imaging is medically necessary:

- MRI without and with contrast or without contrast of the involved levels
- An initial x-ray is not necessary prior to MRI in these individuals.
- For evaluation of facet arthropathy in patients with ankylosing spondylitis, osteoarthritis, or rheumatoid arthritis:
  - Whole body radiopharmaceutical localization imaging (CPT® 78802) and SPECT (CPT® 78803) OR
  - SPECT/CT (CPT® 78830)

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Atlantoaxial Instability in Trisomy 21 (Down Syndrome) (PEDSP-7.3)

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- The diagnosis of atlantoaxial instability is a recognized complication of trisomy 21, and individuals are routinely screened with lateral x-rays of the cervical spine.
- MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) is medically necessary in individuals where the lateral cervical spine x-ray demonstrates an atlantodental (pre-dens) interval of ≥4.5 mm, and/or a neural canal width of ≤14 mm.
- MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) is medically necessary when new or worsening clinical symptoms suggest myelopathy in a trisomy 21 individual.

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Basilar Impression (PEDSP-7.4)

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See: **Basilar Impression/Basilar Invagination (PEDHD-9.4)** in the Pediatric Head Imaging Guidelines

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Chiari Malformation (PEDSP-7.5)

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See: **Chiari and Skull Base Malformations (PEDHD-9)** in the Pediatric Head Imaging Guidelines

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Klippel-Feil Anomaly (Congenital Fusion of Cervical Vertebrae) (PEDSP-7.6)

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This is generally an incidental finding. A detailed history and physical examination with thorough neurologic examination, and plain x-rays should be performed initially. Klippel-Feil can occur in conjunction with platybasia and/or Chiari malformation.

- Plain x-rays of the cervical spine are sufficient to establish the diagnosis. Advanced imaging is medically necessary if there are acute or worsening neurologic symptoms (including pain), or if multiple levels are involved.
  - MRI Cervical Spine without contrast (CPT® 72141) or CT Cervical Spine without contrast (CPT® 72125) for these indications.

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Marfan Syndrome (PEDSP-7.7)

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Individuals with Marfan syndrome are at risk for scoliosis (see **Scoliosis (PEDSP-3.2)**) and dural ectasias. Dural ectasias are usually asymptomatic but can be associated with other spinal lesions.

- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- MRI without contrast of the symptomatic spinal region is medically necessary for the following:
  - New or worsening clinical symptoms suggest a complicated dural ectasia.
  - The individual is under active consideration for surgery.

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Neurofibromatosis (PEDSP-7.8)

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- See: **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines for screening recommendations in neurofibromatosis.
- See: **Neurofibromatosis (PEDPN-2)** in the Pediatric Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines for imaging considerations in neurofibromatosis individuals with known plexiform neurofibromas.
- See: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (PEDONC-8.3)** in the Pediatric and Special Populations Oncology Imaging Guidelines for imaging in individuals with neurofibromatosis and malignant peripheral nerve sheath tumors.

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Von Hippel-Lindau Syndrome (VHL) (PEDSP-7.9)

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- See: **Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)** in the Pediatric and Special Populations Oncology Imaging Guidelines for screening recommendations in VHL patients.
- MRI without and with contrast of the affected spinal level is medically necessary for individuals with known spinal hemangioblastomas in the following conditions:
  - Annually for asymptomatic individuals with unresected spinal hemangioblastoma(s).
  - Preoperative planning for resection of a hemangioblastoma.
  - New or worsening symptoms suggesting progression of a known hemangioblastoma.

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