



United
Healthcare®
Community Plan

UNITEDHEALTHCARE® COMMUNITY PLAN:
RADIOLOGY IMAGING COVERAGE DETERMINATION GUIDELINE

Pediatric Cardiac Imaging Guidelines (For Ohio Only)

V2.0.2024

Guideline Number: CSRAD016OH.C

Effective Date: November 15, 2024

Application (for Ohio Only)

This Medical Policy only applies to the state of Ohio. Any requests for services that are stated as unproven or services for which there is a coverage or quantity limit will be evaluated for medical necessity using Ohio Administrative Code 5160-1-01.

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- Pediatric Peripheral Vascular Disease Imaging Guidelines
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Guideline Development (Preface-1)

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- The UnitedHealthcare's evidence-based, proprietary clinical guidelines evaluate a range of advanced imaging and procedures, including NM, US, CT, MRI, PET, Radiation Oncology, Sleep Studies, as well as Cardiac, musculoskeletal and Spine interventions.
- UnitedHealthcare reserves the right to change and update the guidelines. The guidelines undergo a formal review annually. UnitedHealthcare's 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.
- UnitedHealthcare supports the Choosing Wisely initiative (<https://www.choosingwisely.org/>) by the American Board of Internal Medicine (ABIM) Foundation and many national physician organizations, to reduce the overuse of diagnostic tests that are low value, no value, or whose risks are greater than the benefits.

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

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Investigational and Experimental Studies

- 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
 - if 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.

Clinical and Research Trials

- Similar to investigational and experimental studies, clinical trial imaging requests will be considered to determine whether they meet UnitedHealthcare's 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.

Legislative Mandate

- State and federal legislations may need to be considered in the review of advanced imaging requests.

References (Preface-2)

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1. Coverage of Clinical Trials under the Patient Protection and Affordable Care Act; 42 U.S.C.A. § 300gg-8.

Clinical Information (Preface-3)

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Clinical Documentation and Age Considerations

- UnitedHealthcare's 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. UnitedHealthcare's guidelines are framed by:
 - Clinical presentation of the individual, rather than the studies requested
 - Adequate clinical information that must be submitted to UnitedHealthcare in order to establish medical necessity for advanced imaging or other designated procedures includes, but is not limited to, the following:
 - Pertinent clinical evaluation should include a recent detailed history, physical examination²⁰ since the onset or change in symptoms, and/or laboratory and 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; x-ray imaging does not have to be within the past 60 days.
 - 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.
 - UnitedHealthcare's 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.

- Individuals who are 18 years old or younger¹⁹ 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.
- The terms “male” and “female” used in these guidelines refer to anatomic-specific diseases and disease predispositions associated with the individual's sex assigned at birth rather than their gender identity. It should be noted that gender identity and anatomic-specific diseases as well as disease predispositions are not always linked. As such, these guidelines should be applied to the individual's corresponding known or suspected anatomic-specific disease or disease predisposition. At UnitedHealthcare, we believe that it is important to understand how all individuals, including those who are gender-diverse, choose to identify themselves. To ensure that gender-diverse individuals are treated with respect and that decisions impacting their healthcare are made correctly and with sensitivity, UnitedHealthcare recognizes all individuals with the following gender marker options: Male, Female, Transgender Male, Transgender Female, “X”, and “Not Specified.”

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.¹

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 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:
 - 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.¹
- 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 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.²
- Both contrast CT and MRI may be 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 may be appropriate if clinical criteria for CT with contrast are met AND the individual has:
 - 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
 - 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.
- MRI utilizing Xenon Xe 129 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 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 may be considered to have the same risk profile with renal failure (GFR <30 mL/min).²
 - 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.^{3,4,5,6,7} 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.⁸

- A CT may be approved 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:
 - Caution should be taken in the use of gadolinium in individuals with renal failure.
 - The use of gadolinium contrast agents is 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 indicated 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.

Overutilization of Advanced Imaging

- A number of recent 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 indicated examination has been ordered for individuals, it may be possible to avoid exposing them to unnecessary risks.^{9,10} 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 indicated if the surgery/procedure is not indicated. Once the procedure has been approved or if the procedure does not require prior authorization, the appropriate pre-procedural imaging may be approved.

References (Preface-3)

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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)

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

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

Quantitative MR Analysis of Tissue Composition (Preface-4.8)

HCPCS Codes (Preface-4.9)

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3D Rendering (Preface-4.1)

PRF.CD.0004.1.UOH

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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 UnitedHealthcare 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, Mammogram, MRI Breast, US Breast, CT Colonography (virtual colonoscopy), Cardiac MRI, Cardiac CT, or Coronary CTA studies.

- 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.

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

PRF.CD.0004.2.A

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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

- 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.³
- 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.

- **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.

Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)

PRF.CD.0004.3.UOH

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

- 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) can be considered in the following clinical scenarios:
 - Studies done for navigation and planning for neurosurgical procedures (i.e., Stealth or Brain Lab Imaging)^{1,2}
 - 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.

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

PRF.CD.0004.5.UOH

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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 indicated 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.

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.

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

PRF.CD.0004.7.UOH

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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.⁴
 - CPT[®] 76140 is the appropriate code to use for an exam which was completed elsewhere and a secondary interpretation of the images is requested.⁵

Quantitative MR Analysis of Tissue Composition (Preface-4.8)

PRF.CD.0004.8.A

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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. At present, these procedures are primarily being used in clinical trials and there is no widely recommended indications in clinical practice. As such, these procedures are considered to be investigational and experimental for coverage purposes.
 - CPT[®] 0648T (without diagnostic MRI) and CPT[®] 0649T (with diagnostic MRI) refer to data analysis with and without associate imaging of a single organ, with its most common use being LiverMultiScan (LMS).
 - See **Fatty Liver (AB-29.2)** in the Abdomen Imaging Guidelines.
 - CPT[®] 0697T (without diagnostic MRI) and CPT[®] 0698T (with diagnostic MRI) refer to data analysis with and without associate imaging of a multiple organs, with its most common use being CoverScan.

HCPCS Codes (Preface-4.9)

PRF.CD.0004.9.UOH

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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.⁶
 - 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.

References (Preface-4)

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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)

Whole-Body CT Imaging (Preface-5.1)

PRF.WB.0005.1.UOH

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- Whole-body CT or LifeScan (CT Brain, Chest, Abdomen, and Pelvis) for screening of asymptomatic individuals is not indicated. 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 CT is supported for oncologic staging in Multiple Myeloma. See **Multiple Myeloma and Plasmacytomas (ONC-25)** in the Oncology Imaging Guidelines.

Whole-Body MR Imaging (Preface-5.2)

PRF.WB.0005.2.A

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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)**, **Hereditary Paraganglioma-Pheochromocytoma (HPP) Syndromes (PEDONC-2.13)**, or **Constitutional Mismatch Repair Deficiency (CMMRD or Turcot Syndrome) (PEDONC-2.15)** in the Pediatric Oncology Imaging Guidelines.
 - Cancer staging and restaging:
 - While the feasibility of WBMRI has been established, data remain conflicting on whether WBMRI is of equivalent diagnostic accuracy compared with standard imaging modalities such as CT, scintigraphy, and PET imaging.
 - Evidence has not been published establishing WBMRI as a standard evaluation for any type of cancer.
 - Autoimmune disease:
 - WBMRI can be approved in some situations for individuals with chronic recurrent multifocal osteomyelitis.
 - For additional information, see **Chronic Recurrent Multifocal Osteomyelitis (PEDMS-10.2)** in the Pediatric Musculoskeletal Imaging Guidelines.

PET-MRI (Preface-5.3)

PRF.WB.0005.3.A

v2.0.2024

- 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 may be appropriate 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 is a pediatric patient or being treated under a pediatric guideline and treatment plan AND
 - 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.
- For more information, see **PET Imaging in Pediatric Oncology (PEDONC-1.4)** in the Pediatric Oncology Imaging Guidelines, and **PET Brain Imaging (PEDHD-2.3)** and **Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)** in the Pediatric Head Imaging Guidelines.

References (Preface-5)

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References (Preface-6)

Guideline

References (Preface-6.1)

References (Preface-6.1)

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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.
- The website addresses for certain references are included in the body of the guidelines but are not hyperlinked to the actual website.
- The website address for the American College of Radiology (ACR) Appropriateness Criteria[®] is <http://www.acr.org>.

Copyright Information (Preface-7)

Guideline

Copyright Information (Preface-7.1)

Copyright Information (Preface-7.1)

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Trademarks (Preface-8)

Guideline

Trademarks (Preface-8.1)

Trademarks (Preface-8.1)

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General Information

Guideline

General Guidelines (PEDCD-1.0)

General Guidelines (PEDCD-1.0)

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v1.0.2024

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, echo, 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 echo 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 echos in asymptomatic individuals
- Repeat imaging studies of the heart are **not** indicated unless one of the following applies:
 - Repeat imaging is indicated 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

- See **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** for indications for repeat echos 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** indicated, 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 indicated 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) – Other Indications (PEDCD-10)**
- 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 **Echocardiography - Other Indications (PEDCD-8)**
- Nuclear Medicine

- SPECT, PET stress may be indicated 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 can be approved 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), historically this method of imaging the myocardium, Myocardial Tc-99m Pyrophosphate Imaging, was used to identify recent infarction, hence, the term "infarct-avid scan." Although still available, the sensitivity and specificity for identifying infarcted myocardial tissue is variable 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 (CD-3.7)** and **Cardiac Amyloidosis (CD-3.8)**

MUGA (Multi Gated Acquisition) – Blood Pool Imaging	CPT [®]
Myocardial Imaging, infarct avid, planar, qualitative or quantitative	78466

MUGA (Multi Gated Acquisition) – Blood Pool Imaging	CPT®
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 **Adult Congenital Heart Disease (CD-11)** 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
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

MRI/MRA	CPT®
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
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; data preparation and transmission, analysis of fluid dynamics and simulated maximal coronary hyperemia, generation of estimated FFR model, with anatomical data review in comparison with estimated FFR model to reconcile discordant data, interpretation and report	0501T
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; data preparation and transmission	0502T

CT	CPT®
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; analysis of fluid dynamics and simulated maximal coronary hyperemia, and generation of estimated FFR model	0503T
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; anatomical data review in comparison with estimated FFR model to reconcile discordant data, interpretation and report	0504T

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) (eg, ejection fraction with probe technique) with or without pharmacologic intervention or exercise, single or multiple determinations	78414
Cardiac shunt detection	78428

Nuclear Medicine	CPT®
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
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 (eg, 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 (eg, 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

Nuclear Medicine	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
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

Nuclear Medicine	CPT®
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
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 (eg, 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 (eg, 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

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), single area (eg, 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
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
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

Ultrasound	CPT®
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
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
3D echocardiographic imaging and postprocessing during transesophageal echocardiography, or during transthoracic echocardiography for congenital cardiac anomalies, for the assessment of cardiac structure(s) (eg, 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, 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

Ultrasound	CPT[®]
Doppler echocardiography color flow velocity mapping (List separately in addition to codes for echocardiography)	+93325
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

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

Cardiac Cath 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

Cardiac Cath Procedure	CPT [®]
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
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

Cardiac Cath Procedure	CPT®
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.

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Congenital Heart Disease

Guideline

- Congenital Heart Disease General Information (PEDCD-2.1)
- Congenital Heart Disease Coding (PEDCD-2.2)
- Congenital Heart Disease Modality Considerations (PEDCD-2.3)
- Imaging and Surveillance per Congenital lesion (PEDCD-2.4)
- Atrial Defects-Secundum ASD, PFO, and Partial anomalous pulmonary venous return (PAPVR), Sinus Venosus defect (PEDCD-2.4.1)
- VSD (PEDCD-2.4.2)
- AVSD (Atrioventricular canal, endocardial cushion defect) (PEDCD-2.4.3)
- PDA (Patent ductus arteriosus) (PEDCD-2.4.4)
- TAPVR Total anomalous pulmonary venous return (PEDCD-2.4.5)
- Ebstein anomaly and TV dysplasia (PEDCD-2.4.6)
- Pulmonary Stenosis (PS) (PEDCD-2.4.7)
- Mitral valve disease (PEDCD-2.4.9)
- LVOT lesions (PEDCD-2.4.10)
- Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)
- Coronary Anomalies (PEDCD-2.4.12)
- Tetralogy of Fallot (TOF) (PEDCD-2.4.13)
- 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)

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 individuals over 18 years of age, see **Adult Congenital Heart Disease (CD-11)** in the 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 0-12 months
 - Subcategory of infant: Neonate or newborn 0-28 days
 - Child 1-18 years
 - Subcategory of child: Adolescent 11-18 years
 - "Children" refers to all pediatric individuals ages 0-18 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)
- 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, 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.
 - Because dose adjustments are done by weight, and infants are growing rapidly, they can have changing physiology, pulmonary vascular resistance, ductal size and weight changes, dose response and may require more than one echo during a medication adjustment.
- Heart surgery
 - TTE is indicated 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 echo is equivocal
- MRA/CTA Chest can be performed if prior echo 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 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 (PEDCD-11)**
- **Individuals with Pulmonary hypertension with CHD** should be reviewed for both their lesion and for PHT in section **Pediatric Pulmonary Hypertension (PEDCD-7)**

Congenital Heart Disease Coding (PEDCD-2.2)

CDP.CHD.0002.2.A

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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 indicated 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

- 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)

- 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)

Congenital Heart Disease Modality Considerations (PEDCD-2.3)

CDP.CHD.0002.3.A

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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 indicated unless otherwise indicated in a specific guideline section.
- Cardiac MRI either without contrast (CPT[®] 75557) or without and with contrast (CPT[®] 75561) is indicated, 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 indicated 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 indicated 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 indicated 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
 - Pulmonary artery (PA) and Pulmonary vein (PV) assessment
- CTA Chest (CPT[®] 71275) is indicated 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)
 - In individuals with congenital heart disease or suspected congenital heart diseases, who have clinical questions regarding relative pulmonary blood flow, can have perfusion imaging

Imaging and Surveillance per Congenital lesion (PEDCD-2.4)

CDP.CHD.0002.4.A

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- Echocardiography is often repeated frequently throughout a pediatric individual's 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 contact within 60 days.
- Adults 18 years and older who also have congenital heart disease should be imaged according to **Adult Congenital Heart Disease (CD-11)** and the general Cardiac Imaging Guidelines.

Atrial Defects-Secundum ASD, PFO, and Partial anomalous pulmonary venous return (PAPVR), Sinus Venosus defect (PEDCD-2.4.1)

CDP.CHD.0241.A

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See section on AVSD in **AVSD (Atrioventricular canal, Endocardial cushion defect) (PEDCD-2.4.3) for primum ASD**

PFO (Patent Foramen Ovale)

- Routine surveillance in an asymptomatic individual with PFO is not indicated
 - 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 echo and can be imaged according to the echocardiogram guidelines in **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** and **Frequency of echocardiography testing (CD-2.3)** in the general Cardiac Imaging Guidelines.
- Follow-up imaging with an echocardiogram is indicated when there is documentation of the following:
 - New cardiac symptoms
 - A concern that the last echo was equivocal for other cardiac issues
 - Question of a clot/embolism that has gone across the PFO
 - Prior echo did not differentiate the PFO from a secundum ASD
- TTE (CPT® 93306- non congenital echocardiogram) is indicated 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 but may be indicated 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
 - 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 indicated at any time post procedure when there is concern for **any** of the following:
 - Infection
 - Malposition
 - Embolization
 - Persistent shunt
- If persistent shunt TTE is indicated every 3 months until the finding is stable or there is a need for intervention

ASD and PAPVR asymptomatic isolated atrial septal defect (ASD)

This section reference secundum ASD, sinus Venosus, ASD and unobstructed partial anomalous pulmonary venous return

- Any surgical status
 - TTE is indicated 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
 - Echo (TTE) every 1 year
- 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.

VSD (PEDCD-2.4.2)

CDP.CHD.0242.A

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All

TTE is indicated for any of the following:

- with 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
 - Infancy every 6 months
 - Childhood 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.
 - Child –TTE every 3 months
 - Adolescent- TTE every 6 months

AVSD (Atrioventricular canal, endocardial cushion defect) (PEDCD-2.4.3)

CDP.CHD.0243.A

v1.0.2024

Any surgical status

TTE is indicated 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)
 - Newborn one addition study next 2 months.
 - TTE
 - Infancy every 3 months in infancy
 - 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

PDA (Patent ductus arteriosus) (PEDCD-2.4.4)

CDP.CHD.0244.A

v1.0.2024

Any surgical status

TTE is indicated 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

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
 - \geq Moderate PDA: TTE every month
- 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
 - Child
 - TTE annually
 - MRA/CTA Chest, or (lung perfusion for LPA stenosis) if questions remain unanswered after TTE
 - Adolescents
 - Every two years TTE and

- MRA/CTA Chest, or (lung perfusion for LPA stenosis) if questions remain unanswered after TTE

TAPVR Total anomalous pulmonary venous return (PEDCD-2.4.5)

CDP.CHD.0245.A

v1.0.2024

Any surgical status

- TTE, TEE, CMR/CT-CMRA/CTA, Lung perfusion scan are indicated 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
- Child: every 1 year
- Adolescence: TTE every 2 years

Ebstein anomaly and TV dysplasia (PEDCD-2.4.6)

CDP.CHD.0246.A

v1.0.2024

Any surgical status

- TTE, TEE, CMR/CT-CMRA/CTA are indicated 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
- Child: every year
- Adolescent: every 2 years
- Every 3 months if CHF or atrial arrhythmias

Pulmonary Stenosis (PS) (PEDCD-2.4.7)

CDP.CHD.0247.A

v1.0.2024

Any surgical status

- TTE is indicated 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
- 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
- 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

Mitral valve disease (PEDCD-2.4.9)

CDP.CHD.0249.A

v1.0.2024

Any surgical status

- TTE is indicated 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 \geq 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 \geq moderate MR
- Child
 - TTE every 2 years with mild MR, normal LV size and systolic function
 - TTE every 6 months with \geq 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 \geq 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

- TTE every 3 months
 - In a child with \geq moderate MS or MR, dilated LV, and no LV dysfunction
 - In a child with prosthetic mitral valve or ventricular dysfunction, and/or arrhythmias

LVOT lesions (PEDCD-2.4.10)

CDP.CHD.2410.A

v1.0.2024

Subvalvular Aortic stenosis

Any surgical status

- TTE, TEE, Cardiac MR/CT are indicated 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)

Any surgical status

- TTE, TEE, Cardiac MR/CT are indicated 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 \geq 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 \geq moderate AS or AR or LV dysfunction
 - Every 3 months \leq mild AS/AR and no LV dysfunction
- Child (TTE)
 - 6 months echo if \geq moderate AS or AR
 - 1-year echo if \leq 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 indicated for any of the following:
 - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
 - Preoperative
 - Williams syndrome
 - 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 \geq moderate AS

Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)

CDP.CHD.2411.A

v1.0.2024

All individuals

- TTE, MRA/CTA Chest are indicated 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 indicated 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

Unrepaired Aortic Coarctation

- Newborn, TTE weekly if assessing for ductal closure
- Infant with mild coarctation in absence of PDA
 - Echo every 3 months
- Child with mild coarctation
 - Echo 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 \geq moderate sequelae
 - MRA/CTA Chest every 3 years (include Cardiac MR/CT if issues not clarified on echo)

Coronary Anomalies (PEDCD-2.4.12)

CDP.CHD.2412.A

v1.0.2024

- CCTA or cardiac MRI is indicated 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, of 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 Valsavla 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
 - 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 indicated 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 or 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
 - Individuals with a history that includes one or more of the indications in **Indications for CCTA (CPT® 75574) (PEDCD-10.3)**

Tetralogy of Fallot (TOF) (PEDCD-2.4.13)

CDP.CHD.2413.A

v1.0.2024

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 planned 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
 - Child-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 $\leq 48\%$ or $\geq 6\%$ decrease in EF) or nearing imaging criteria for PVR.

Post-surgical or catheter based pulmonary valve replacement

- TTE

- 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

Double Outlet Right Ventricle (DORV) (PEDCD-2.4.14)

CDP.CHD.2414.A

v1.0.2024

Any surgical status

- TTE, CMR/CT-CMRA/CTA are indicated 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

D-Loop Transposition of the Great Arteries (D-Loop TGA) (PEDCD-2.4.15)

CDP.CHD.2415.A

v1.0.2024

Any surgical status

- **TTE, CMR/CT-CMRA/CTA, stress imaging** are indicated 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 \geq 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

- 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 \geq moderate systemic AV, valve regurgitation, systemic RV dysfunction, LVOT obstruction, or arrhythmias, or CHF.
 - Routine **CMR/CT-CMRA/CTA** every 3 years

Congenitally Corrected Transposition of the Great Arteries (ccTGA, LTGA) (PEDCD-2.4.16)

CDP.CHD.2416.A
v1.0.2024

Any surgical status

- TTE, TEE, **CMR/CT-CMRA/CTA** are indicated 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 if cardiac symptoms and moderate findings
 - Child
 - <2 years every 3 months
 - >2 years every 1 year
 - Every 6 months if \geq moderate AV regurg
 - 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
 - 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 \geq 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 \geq moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction, or with CHF symptoms

Truncus Arteriosus (PEDCD-2.4.17)

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Any surgical status

- TTE, CMR/CT-CMRA/CTA are indicated 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
 - \geq 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 \geq moderate Truncal stenosis or regurgitation

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 indicated 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 cath procedure
- Routine weekly TTE

Stage 2 palliation (TTE)

- Often referred to as Glenn procedure
- Within 30 days after surgical or cath 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 indicated for all Single Ventricles once every 10 years

Eisenmenger and PHT (with CHD)- (PEDCD-2.4.19)

CDP.CHD.2419.A

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PHT without CHD is covered in section **Pediatric Pulmonary Hypertension (PEDCD-7)**

These are in addition to studies supported by lesion

Any surgical status

- TTE, CMR/CT-CMRA/CTA are indicated 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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Condition Specific Imaging

Guideline

Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)
Heart Murmur (PEDCD-3.1)
Chest Pain General (PEDCD-4.1)
Syncope (PEDCD-5.1)
Pediatric Pulmonary Hypertension General (PEDCD-7.1)
Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade (PEDCD-9.5)
Kawasaki Disease (PEDCD-6)

Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)

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MIS-C General Information (PEDCD-12.1)

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 [PEDHD-12.7](#) for the outpatient management of head imaging.

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 \(PEDCD-6.2\)](#)) echo (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 echo (TTE) can be approved at the time of presentation and followed by serial echos (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 echos 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 echo (TTE):
 - Within 1 week of discharge
 - 4 weeks post-discharge
 - At 6 months post-discharge
 - One year post-discharge
- 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 echo (TTE) may have a Cardiac MRI based on **Cardiac MRI and MRA Chest – Indications (excluding Stress MRI) (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 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		

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

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

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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
CPT [®] 93303, CPT [®] 93306, CPT [®] 93320, and CPT [®] 93325
CPT [®] 93303, 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)

- Any one of the above echocardiography code combinations is indicated for evaluation of either:
 - Any pathologic murmur
 - Any innocent murmur with associated cardiac signs or symptoms
- Repeat echocardiography is not indicated if the initial echocardiogram was normal and the murmur has not changed in quality.

Background and supporting information

- 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 indicated, and is performed as part of the office visit.

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Chest Pain General (PEDCD-4.1)

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Initial pediatric TTE combinations

Cardiologist only submits charges for the procedure actually performed
CPT [®] 93303, CPT [®] 93306, CPT [®] 93320, and CPT [®] 93325
CPT [®] 93303, 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)

- Any one of the TTE combinations above is indicated 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 indicated 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 indicated 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 indicated if other imaging suggests coronary artery abnormality, or ETT suggests ischemia. EKG is uninterpretable. Any indication in section **Stress Testing with Imaging – Indications (CD 1.4)** in the Cardiac Imaging Guidelines. This can include Stress SPECT, echo or MR

Background and supporting information

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 (PEDCD-5.1)

CDP.CS.0005.1.A

v1.0.2024

Initial pediatric TTE combinations

Cardiologist only submits charges for the procedure actually performed

CPT[®] 93303, CPT[®] 93306, CPT[®] 93320, and CPT[®] 93325

CPT[®] 93303, 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)

- Echocardiography is not indicated for most individuals with isolated syncope.
- Echocardiography is indicated 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 indicated 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

- Cardiac MR or Cardiac CT is indicated 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 indicated (any);
 - If other imaging suggests coronary artery abnormality
 - ETT suggests ischemia
 - EKG is uninterpretable
 - Any indication in section **Stress Testing with Imaging – Indications (CD 1.4)** in the Cardiac Imaging Guidelines

Background and supporting information

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 (PEDCD-7.1)

CDP.CS.0007.1.A

v1.0.2024

- 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 indicated 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 PAH-targeted therapy, and for individuals with concomitant congenital heart disease
- CT Chest (CPT[®] 71250) may be indicated 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 indicated for evaluation of inconclusive echocardiogram findings, or for monitoring right ventricular function during follow-up.
- Stress echocardiograms may be indicated (as in the general cardiac imaging guidelines) see **Stress Echocardiography – Indications other than ruling out CAD (CD-2.7)** in the Cardiac Imaging Guidelines.

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Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade (PEDCD-9.5)

CDP.CS.0009.5.A

v1.0.2024

- 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 (PEDCD-6)

CDP.CS.0006.A

v1.0.2024

Kawasaki Disease Initial Imaging (PEDCD-6.1)

- Echocardiography (CPT[®] 93306) is indicated 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 indicated for evaluation of inconclusive echocardiogram findings, or significant coronary artery abnormalities.
 - Screening of other body areas for aneurysms is not routinely indicated 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 should be performed when the diagnosis of KD is considered
 - Uncomplicated individuals, echocardiography can be repeated after treatment **both:**
 - Within 1 to 2 weeks
 - Within 4 to 6 weeks
 - For individuals with important and evolving coronary artery abnormalities (Z score >2.5) detected during the acute illness, more frequent echocardiography (at least twice per week) should be performed until luminal dimensions have stopped progressing to determine the risk for and presence of thrombosis.

- Expanding large or giant aneurysms:
 - 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
- It is reasonable to obtain advanced imaging studies such as computed tomographic angiography (CTA), cardiac magnetic resonance imaging (CMRI), or invasive angiography on individuals' severe proximal coronary artery abnormalities in the acute phase when results will impact management decisions.
- Transesophageal echocardiography, invasive angiography, CMRI, and CTA can be of value in the assessment of selected individuals but are not routinely indicated for diagnosis and management of the acute illness.
 - Invasive angiography is rarely performed during the acute illness.
 - Transesophageal echocardiography, CTA, and CMRI can be useful for the evaluation of older children and adolescents 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 indicated 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.

Background and supporting information

- 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 abnormalities¹
 - 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 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.

- 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
 - 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
- 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

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
		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
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

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
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
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)
- Fetal Echo (PEDCD-8.5)
- References (PEDCD-8)

Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)

CDP.000.8.1.A

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- CPT[®] codes for echocardiography are listed in **General Guidelines (PEDCD-1)**

Echocardiogram coding notes	CPT [®]
<ul style="list-style-type: none"> • The most commonly performed study is a complete transthoracic echocardiogram with spectral and color flow Doppler (CPT[®] 93306). <ul style="list-style-type: none"> ◦ CPT[®] 93306 includes CPT[®] 93320 and CPT[®] 93325, so those codes should not be approved along with CPT[®] 93306. 	93306
<ul style="list-style-type: none"> • 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
<ul style="list-style-type: none"> • For a 2D transthoracic echocardiogram without Doppler, report CPT[®] 93307. 	93307
<ul style="list-style-type: none"> • A limited transthoracic echocardiogram is reported with CPT[®] 93308. <ul style="list-style-type: none"> ◦ 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
<ul style="list-style-type: none"> • 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** echo on a pediatric individual will not know what procedure codes they will be reporting until the initial study is completed.
 - If congenital heart disease is found on the initial echo, a complete echo 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 indicated, 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 indicated, 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

Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)

CDP.000.8.1.A

v1.0.2024

- In addition to indications listed in previous guideline sections, initial TTE evaluation is indicated 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
 - Sick 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)

- 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:

- 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

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 indicated 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 echo every 12 months
 - Repeat echo 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** indicated 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 echo is allowed to assess stability of findings
 - Individual with a phenotype positive cardiomyopathy (with or without a positive gene) can be imaged as follows:
 - Infants (under one year)

- TTE is indicated at frequency requested by pediatric cardiology or provider in consultation with pediatric cardiology
- Children (over one year) yearly testing is indicated 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 echo 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 echo 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 echo 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 Sβ⁰, severe thalassemia, etc.)
 - History of acute chest syndrome or intrinsic lung disease
 - History of stroke
 - 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 echo 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)

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) – Indications (CD-2.5)** in the Cardiac Imaging Guidelines.

Fetal Echo (PEDCD-8.5)

CDP.EC.0008.5.A
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Fetal Echocardiography - coding

- Supported fetal echocardiography (echo) codes include:
 - Initial Fetal Echo, CPT[®] 76825 and Doppler Echo CPT[®] 76827 are performed only once per fetus/per facility (i.e. Maternal Fetal Medicine versus Pediatric Cardiology request)
 - Follow-up-Fetal echo and/or Follow-up Doppler echo (CPT[®] 76826/CPT[®] 76828)
 - CPT[®] 93325 for Doppler color flow velocity mapping
- An initial fetal echo is usually not performed prior to 16 weeks.
- Doppler echo procedure codes (CPT[®] 76827 or CPT[®] 76828) include the evaluation of veins, arteries, and valves. Guidelines do not support the billing of additional codes (CPT[®] 76820 and/or CPT[®] 76821)

Background and Supporting Information

- The minimal use of color Doppler (CPT[®] 93325) alone, when performed for anatomical structure identification during a standard ultrasound procedure, is not separately reimbursable

Fetal Echocardiography - Indications for Fetal Conditions

Initial Fetal echocardiography (CPT[®] 76825) and/or Doppler echocardiography (CPT[®] 76827) with or without Doppler color flow velocity mapping (CPT[®] 93325) can be performed if ≥16 weeks, for the indications listed below (See **Fetal Echocardiography – Coding (OB-12.1)**):

Fetal Echocardiography - Indications for Fetal Conditions

- Known or suspected abnormal fetal cardiac evaluation on fetal anatomic scan.
 - Known or suspected abnormality must be documented as hard copy or acknowledged verbally by provider of known or suspected fetal cardiac evaluation
 - Suboptimal cardiac evaluation alone is not an indication for fetal echogram. If the 4-chamber view is adequate and there is no other suspicion of a cardiac abnormality, a fetal echocardiogram is not considered medically necessary. A follow up ultrasound (CPT[®] 76815 or CPT[®] 76816) is indicated for suboptimal visualization. If the follow-up ultrasound fails to show a 4-chamber view or there is suspicion of a cardiac abnormality, fetal echocardiogram is indicated.
- Fetal cardiac arrhythmia; persistent fetal tachycardia or bradycardia

Fetal Echocardiography - Indications for Fetal Conditions

- Major fetal extra-cardiac anomaly
- Fetal Echo is NOT indicated for an isolated soft marker found on routine imaging including:
 - Choroid plexus cyst, or
 - Echogenic intra-cardiac foci, or
 - Thickened nuchal fold (≥ 6 mm at 15 to 20 weeks), or
 - Absent or hypoplastic nasal bone, or
 - Echogenic bowel, or
 - Shortened long bones, or
 - Pyelectasis
- Congenital heart disease (CHD) in a 1st degree relative of the fetus (i.e. CHD in the mother, father, or sibling of the fetus) or a half-sibling of the fetus
- Known fetal chromosomal abnormalities (fetal aneuploidy) or ultrasound findings of a suspected chromosomal abnormality (excluding soft markers as only ultrasound findings)
 - Early onset FGR (<32 weeks) may be a sign of fetal aneuploidy^{11,12}
- Single umbilical artery
- Chorioangioma or Umbilical cord varix (if suspicion of fetal hydrops)
- Fetal intra-abdominal venous anomaly (persistent right umbilical vein)
- Fetal effusion (pericardial, pleural effusion, ascites, etc.)
- Fetal hydrops, See **Alloimmunization/Rh Isoimmunization/Other Causes of Fetal Anemia/Parvo/Hydrops (OB-16)**
- Monochorionic twins/TTTS
- Abnormal Fetal Nuchal Translucency scan (NT ≥ 3.0 mm or above the 95th percentile for the CRL) during current pregnancy.
- Abnormal ductus venosus waveform⁵
- Fetal echocardiography may be indicated with severe or unexplained polyhydramnios, or if there are also other suspicious findings on an anatomy scan

Fetal Echocardiography - Indications for Maternal Conditions

Initial Fetal echocardiography (CPT[®] 76825) and/or Doppler echocardiography (CPT[®] 76827) with or without Doppler color flow velocity mapping (CPT[®] 93325) can be performed if ≥ 16 weeks, for the indications listed below (See **Fetal Echocardiography – Coding (OB-12.1)**):

Maternal Conditions:

- Maternal pre-gestational DM or early diagnosed GDM (1st or early 2nd trimester)
- Maternal gestational diabetes mellitus, if HbA1C >6% [in the third trimester (≥32 weeks)]
- Maternal connective tissue disease (SLE, RA, Sjogrens) with Anti-Ro/SSA or anti-La/SSB antibodies present
 - Weekly follow-up Fetal echocardiography (CPT[®] 76826) and/or Doppler fetal echocardiography (CPT[®] 76828) or CPT[®] 76815 from the 18th through the 26th week of pregnancy and then every other week until 30 weeks
- Phenylketonuria
- Infections associated with cardiac anomalies (such as parvovirus, Rubella, Coxsackie virus)
- Genetic conditions associated with CHD in a first degree relative of the fetus (e.g. Marfan syndrome, 22q11.2 deletion syndrome (DiGeorge Syndrome) or Noonan syndrome)
- Prior child with CHD born to mother and/or father of the fetus⁵
- Pregnancy conceived by assisted reproductive technology:¹
 - In Vitro Fertilization (IVF)
 - Intracytoplasmic sperm injection (ICSI)¹

Background and supporting information

If diabetes is diagnosed prior to pregnancy or in the first or early second trimester (typically before 20 weeks gestation) with standard diagnostic criteria of: HbA1C ≥6.5%, fasting plasma glucose ≥126 mg/dL, or 2-hour glucose ≥200 mg/dL on a 75-g oral glucose tolerance test, then image as above

For those with GDM on medication, if HbA1c levels are >6%, fetal echocardiogram in the third trimester to assess for ventricular hypertrophy can be performed.

In cases of extreme obesity (BMI≥40-50) where the 4-chamber view is inadequately documented after 2 separate ultrasound visits with MFM, fetal echo can be performed.

With positive SSA/SSB antibodies, the most vulnerable period for the fetus is during the period from 18 to 24 weeks gestation. Normal sinus rhythm can progress to complete block in seven days during this high-risk period. New onset of heart block is less likely during the 26th through the 30th week, and it rarely develops after 30 weeks of pregnancy.

Fetal Echocardiography - Indications for Medication or Drug Exposure

Initial Fetal echocardiography (CPT[®] 76825) and/or Doppler echocardiography (CPT[®] 76827) with or without Doppler color flow velocity mapping (CPT[®] 93325) can be performed if ≥ 16 weeks, for the indications listed below (See **Fetal Echocardiography – Coding (OB-12.1)**):

- Ace inhibitors
- Alcohol (excessive quantities)
- Anti-seizure medication, e.g. carbamazepine, hydantoin, valproate
- Folate antagonists (methotrexate)
- Lithium
- NSAIDS (Ibuprofen, Indomethacin) 2nd and 3rd trimester
- Paroxetine (Paxil)
- Retinoids (e.g. Isotretinoin, Retinoic acid, Vitamin A -over 10,000 IU per day, etc.)
- Thalidomide
- Venlafaxine (Effexor)
- This may not be an all-inclusive list, however, exposure to other potential teratogens associated with cardiac anomalies in the fetus are typically adequately assessed with a detailed fetal anatomy ultrasound. (CPT[®] 76811) (See **Potentially Teratogenic Medications/Substances (OB 10.1)**)

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Computed Tomography

Guideline

CT Heart and Coronary Computed Tomography Angiography (CCTA) General Considerations (PEDCD-10.1)

Indications for CCTA (CPT[®] 75574) (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)

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) states “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) states, “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 (CD-1)** in the general Cardiac Imaging Guidelines

Indications for CCTA (CPT[®] 75574) (PEDCD-10.3)

CDP.CT.0010.3.A

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- In addition to indications listed in previous guideline sections, CCTA is indicated 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 arteriosus, aortic root surgery)
 - Evaluation of the arterial supply and venous drainage in children with bronchopulmonary sequestration
- See **Coronary Anomalies (PEDCD-2.4.12)**

Indications for Cardiac CT (CPT[®] 75572) (PEDCD-10.4)

CDP.CT.0010.4.A

v1.0.2024

- In addition to indications listed in previous guideline sections, CCT is indicated 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 may also be indicated during a Cardiac CT if there are issues regarding the chest vessels that are inconclusive after echocardiogram or Cardiac MRI

Indications for Chest CTA with Cardiac CT or CTA (PEDCD-10.5)

CDP.CT.0010.5.A

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- A Chest CTA may be indicated in individuals who require Cardiac CT or Cardiac CTA when:
 - A TTE or MRI is inconclusive for issues regarding chest vasculature
 - Routine imaging is indicated based on **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)**

References (PEDCD-10)

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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)

Cardiac MRI Coding (PEDCD-9.2)

CDP.MR.0009.2.A
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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.

Indications for Cardiac MRI (PEDCD-9.3)

CDP.MR.0009.3.A

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- In addition to indications listed in previous guideline sections, Cardiac MRI (CPT[®] 75557 or 75561) evaluation is indicated 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 arteriosus, 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's 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.

- 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 indicated 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 indicated 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 indicated unless there was an independent indication for either of those codes.

Indications for Chest MRA for Congenital heart disease (PEDCD-9.4)

CDP.MR.0009.4.A

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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 indicated 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 indicated following an inconclusive TTE:
 - MRI Chest (CPT[®] 71552)
 - MRA Chest (CPT[®] 71555)
- MRA Chest (CPT[®] 71555) is indicated 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).

References (PEDCD-9)

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Diagnostic Heart Catheterization

Guideline

Cardiac Catheterization General Information (PEDCD-11.1)

Cardiac Catheterization Indications (PEDCD-11.2)

References (PEDCD-11)

Cardiac Catheterization General Information (PEDCD-11.1)

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Cardiac Catheterization Procedure Codes

Cardiac Cath 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

Cardiac Cath 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 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 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 catheterizations are done for many purposes, including diagnosis and intervention of congenital and acquired heart disease.
- 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 cath may be considered on a case-by-case basis if there are unanswered issues via noninvasive imaging.

- A right heart cath can be approved for pulmonary artery interventions (e.g., stents, coils).

Cardiac Catheterization Indications (PEDCD-11.2)

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- Diagnostic catheterization is indicated:
 - When other advanced imaging has failed to resolve a clinical issue and results will impact the individual's management
 - For example, a cath 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 indicated 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

References (PEDCD-11)

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Policy History and Instructions for Use

Guideline

Policy History and Instructions for Use

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Instructions for Use

This Medical Policy provides assistance in interpreting United HealthCare Services, Inc. standard benefit plans. When deciding coverage, the federal, state (Ohio Administrative Code [OAC]) or contractual requirements for benefit plan coverage must be referenced as the terms of the federal, state (OAC) or contractual requirements for benefit plan coverage may differ from the standard benefit plan. In the event of a conflict, the federal, state (OAC) or contractual requirements for benefit plan coverage govern.

Before using this policy, please check the federal, state (OAC) or contractual requirements for benefit plan coverage. United HealthCare Services, Inc reserves the right to modify its Policies and Guidelines as necessary. This Medical Policy is provided for informational purposes. It does not constitute medical advice.

United HealthCare Services, Inc. uses InterQual[®] for the primary medical/surgical criteria, and the American Society of Addiction Medicine (ASAM) for substance use, in administering health benefits. If InterQual[®] does not have applicable criteria, United HealthCare Services, Inc. may also use United HealthCare Services, Inc. Medical Policies, Coverage Determination Guidelines, and/or Utilization Review Guidelines that have been approved by the Ohio Department for Medicaid Services. The United HealthCare Services, Inc. Medical Policies, Coverage Determination Guidelines, and Utilization Review Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

Policy History/Revision Information

Date	Summary of Changes
02/01/2024	Annual evidence-based updates
07/01/2024	Interim evidence-based updates