UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2024 P 2325-1
Program	Prior Authorization/Medical Necessity
Medication	Cuvrior™ (trientine tetrahydrochloride)*
P&T Approval Date	2/2024
Effective Date	5/1/2024

1. Background:

Cuvrior (trientine tetrahydrochloride) is a copper chelator indicated for the treatment of adult patients with Wilson's disease who are de-coppered and tolerant to penicillamine.

Wilson's disease is an autosomal genetic disorder of copper metabolism that results in an inability to maintain copper balance. Excess copper accumulates and is deposited in several organs and tissues, and eventually produces pathological effects primarily in the liver, where damage progresses to post necrotic cirrhosis, and in the brain, where degeneration is widespread. Copper is also deposited as characteristic, asymptomatic, golden-brown Kayser-Fleischer rings in the corneas of all patients with cerebral symptomatology and some patients who are either asymptomatic or manifest only hepatic symptomatology. Chelating agents (e.g., pencillamine, trientine hydrochloride) are well-established as the standard treatment of Wilson's disease. Cuvrior (trientine tetrahydrochloride) has only been studied for maintenance treatment in patients with Wilson's disease who are pencillamine tolerant.

2. Coverage Criteria[a]:

A. <u>Initial Authorization</u>

1. **Cuvrior*** will be approved based upon **<u>all</u>** of the following criteria:

   a. Diagnosis of Wilson's disease

   -AND-

   b. Patient is de-coppered [i.e., serum non-ceruloplasmin copper (NCC) level ≥ 25 and ≤ 150 mcg/L]

   -AND-

   c. Patient is tolerant to penicillamine

   -AND-

   d. Patient will discontinue penicillamine before starting therapy with Cuvrior

   -AND-

   e. History of intolerance, failure or contraindication to trientine hydrochloride

-AND-

f. Prescribed by a hepatologist.

Authorization will be issued for 12 months.

B. <u>Reauthorization</u>

1. **Cuvrior** will be approved based on **both** the following criteria:

   a. Documentation of positive clinical response to Cuvrior therapy (e.g., increased 24-hour urinary copper excretion from baseline, normalization of serum free copper, prevention of or improvement in symptoms)

-AND-

   b. Prescribed by a hepatologist.

Authorization will be issued for 12 months.

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

*Cuvrior is typically excluded from coverage. Tried/Failed criteria may be in place. Please refer to plan specifics to determine exclusion status.

3. Additional Clinical Rules:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

4. Reference:

1. Cuvrior [package insert]. Chicago, IL: Orphalan; April 2022.
2. Trientine hydrochloride [package insert]. Parsippany, NJ: Teva Pharmaceuticals; January 2022.
3. Roberts EA, Schilsky ML; American Association for Study of Liver Diseases (AASLD). Diagnosis and treatment of Wilson disease: an update. Hepatology. 2008;47(6):2089-2111. doi:10.1002/hep.22261
4. Socha P, Janczyk W, Dhawan A, et al. Wilson's Disease in Children: A Position Paper by the Hepatology Committee of the European Society for Paediatric Gastroenterology, Hepatology and Nutrition. J Pediatr Gastroenterol Nutr. 2018;66(2):334-344. doi:10.1097/MPG.0000000000001787

Program	Prior Authorization/Medical Necessity - Cuvrior (trientine tetrahydrochloride)
Change Control	
2/2024	New program.