COMMON GROUND
HEALTHCARE COOPERATIVE

# PHARMACY POLICY STATEMENT
## Common Ground Healthcare Cooperative (CGHC)

| DRUG NAME | Cuvrior (trientine tetrahydrochloride) |
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| BENEFIT TYPE | Pharmacy |
| STATUS | Prior Authorization Required |

Cuvrior, approved by the FDA in 2022, is a copper chelator indicated for the treatment of adult patients with stable Wilson's disease who are de-coppered and tolerant to penicillamine. It eliminates absorbed copper from the body by forming a stable complex that is then eliminated through urinary excretion. It also chelates copper in the intestinal tract, reducing copper absorption.
Wilson's disease is a genetic disorder of copper metabolism leading to copper accumulation in the liver, brain, and corneas. It is caused by mutations in the intracellular copper-transporting ATP7B gene. Symptoms can include hepatic, neurologic, and/or psychiatric manifestations. The first phase of treatment is to remove existing copper that has accumulated using a chelator. The main chelator used is D-penicillamine, marketed as Depen or Cuprimine (both available generically). Another form of the chelator trientine, marketed as Syprine (also generic), is another option and has fewer side effects. Unlike Cuvrior, it is indicated in patients who do not tolerate penicillamine. The second phase of treatment focuses on preventing further copper accumulation.
Cuvrior was approved based on results of the phase 3 CHELATE trial in which it was found to be noninferior to penicillamine for the primary outcome that measured serum non-ceruloplasmin copper (NCC) at 6 months.

Cuvrior (trientine tetrahydrochloride) will be considered for coverage when the following criteria are met:

### Wilson's Disease

For **initial** authorization:
1. Member is at least 18 years of age; AND
2. Medication is prescribed by or in consultation with a hepatologist or gastroenterologist; AND
3. Member has a diagnosis of Wilson's disease confirmed by at least one of the following (a or b):
   a) Documentation of a Leipzig score of 4 or greater
   b) At least 2 of the following:
      i) Kayser-Fleischer rings identified on slit-lamp exam
      ii) Serum ceruloplasmin level < 20 mg/dL
      iii) 24-hour urinary copper excretion (UCE) > 40 mcg/24 hours
      iv) Liver biopsy (hepatic copper content 250 mcg/g or greater) OR genetic testing (*ATP7B* mutations) indicative of Wilson's disease; AND
4. Member tolerates penicillamine and has been adequately controlled with at least 3 months of treatment as evidenced by at least one of the following:
   a) NCC level of 25 to 150 mcg/L
   b) UCE 200 to 500 mcg/24 hours; AND

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5. Member does NOT have uncontrolled liver disease (e.g., decompensated cirrhosis, acute liver failure, etc.); AND
6. Cuvrior will not be used in combination with penicillamine or any other trientine product. (Current penicillamine use must be discontinued).
7. **Dosage allowed/Quantity limit:** 300 mg to 3,000 mg per day in divided doses (twice daily). (QL: 280 tablets per 28 days)

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:
1. Chart notes must document continued stability/normalization of at least one of the following:
   **a)** NCC level
   **b)** 24-hour UCE; AND
2. Member is clinically stable (e.g., stable hepatic, neurologic, psychiatric exam/labs).

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

| DATE | ACTION/DESCRIPTION |
|---|---|
| **08/16/2022** | New policy for Cuvrior created. |

References:


1. Cuvrior [prescribing information]. Orphalan; 2022.
2. CHELATE STUDY: Trientine Tetrahydrochloride (TETA 4HCL) for the Treatment of Wilson's Disease. ClinicalTrials.gov Identifier: NCT03539952. Accessed August 18, 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. European Association for Study of Liver. EASL Clinical Practice Guidelines: Wilson's disease. *J Hepatol*. 2012;56(3):671-685. doi:10.1016/j.jhep.2011.11.007
5. 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
6. Saroli Palumbo C, Schilsky ML. Clinical practice guidelines in Wilson disease. *Ann Transl Med*. 2019;7(Suppl 2):S65. doi:10.21037/atm.2018.12.53


Effective date: 01/01/2025
Revised date: 08/16/2022