PHARMACY POLICY STATEMENT

BILLING CODE	Must use valid NDC

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
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. 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 _____:

1. Chart notes must document continued stability/normalization of at least one of the following:
   NCC level
   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.

	New policy for Cuvrior created.

References:

1. Cuvrior [prescribing information]. Orphalan; 2022.
2. CHELATE STUDY: Trientine Tetrahydrochloride (TETA 4HCL) for the Treatment of Wilson Disease