products to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

I. Coverage of Cuprimine and penicillamine capsules is recommended in those who meet one of the following criteria:

FDA-Approved Indications

1. Cystinuria. Approve for 1 year if the patient meets the following criteria (A and B):
- **A)** According to the prescriber, patient has tried increased fluid intake; restriction of sodium and protein; and urinary alkalinization; AND
- **B)** Patient meets ONE of the following (i or ii):
  - **i.** Generic penicillamine capsules are requested; OR
  - **ii.** If brand Cuprimine is prescribed, patient has tried generic penicillamine capsules AND cannot take generic penicillamine capsules due to a formulation difference in the inactive ingredient(s) [e.g., difference in dyes, fillers, preservatives] between the Brand and the bioequivalent generic product which, per the prescriber, would result in a significant allergy or serious adverse reaction.

2. Wilson's Disease. Approve for 1 year if the patient meets the following criteria (A, B, C, and D):
- **A)** Diagnosis of Wilson's disease is confirmed by ONE of the following (i or ii):
  - **i.** Genetic testing results confirming biallelic pathogenic *ATP7B* mutations (in either symptomatic or asymptomatic individuals); OR
  - **ii.** Confirmation of at least TWO of the following (TWO of a, b, c, and d):
    - **a)** Presence of Kayser-Fleischer rings;
    - **b)** Serum ceruloplasmin level < 20 mg/dL;
    - **c)** Liver biopsy findings consistent with Wilson's disease;
    - **d)** 24-hour urinary copper > 40 mcg/24 hours; AND
- **B)** Patient meets ONE of the following criteria (i, ii, iii, or iv):
  - **i.** Patient has tried Galzin (zinc acetate capsules); OR
  - **ii.** Patient has tried another zinc product (e.g., zinc sulfate, zinc gluconate, zinc acetate); OR
  - **iii.** According to the prescriber, patient has symptoms of Wilson's disease and zinc would not be an appropriate therapy; OR
  - **iv.** Patient has been started on therapy with a penicillamine product; AND
- **C)** Patient meets ONE of the following (i or ii):
  - **i.** Generic penicillamine capsules are requested; OR
  - **ii.** If brand Cuprimine is prescribed, patient has tried generic penicillamine capsules AND cannot take generic penicillamine capsules due to a formulation difference in the inactive ingredient(s) [e.g., difference in dyes, fillers, preservatives] between the Brand and the bioequivalent generic product which, per the prescriber, would result in a significant allergy or serious adverse reaction; AND
- **D)** The medication is prescribed by or in consultation with a gastroenterologist, hepatologist, or liver transplant physician.

II. Coverage of Depen and penicillamine tablets is recommended in those who meet one of the following criteria:

FDA-Approved Indications

1. **Cystinuria.** Approve for 1 year if the patient meets the following criteria (A and B):
   - **A)** According to the prescriber, patient has tried increased fluid intake; restriction of sodium and protein; and urinary alkalinization; AND
   - **B)** Patient meets ONE of the following (i or ii):
     - **i.** Generic penicillamine tablets are requested; OR
     - **ii.** If brand Depen is prescribed, patient has tried generic penicillamine tablets AND cannot take generic penicillamine tablets due to a formulation difference in the inactive ingredient(s) [e.g., difference in dyes, fillers, preservatives] between the Brand and the bioequivalent generic product which, per the prescriber, would result in a significant allergy or serious adverse reaction.

2. **Wilson's Disease.** Approve for 1 year if the patient meets the following criteria (A, B, C, and D):
   - **A)** Diagnosis of Wilson's disease is confirmed by ONE of the following (i or ii):
     - **i.** Genetic testing results confirming biallelic pathogenic *ATP7B* mutations (in either symptomatic or asymptomatic individuals); OR
     - **ii.** Confirmation of at least two of the following (a, b, c, d):
       - **a)** Presence of Kayser-Fleischer rings;
       - **b)** Serum ceruloplasmin level < 20 mg/dL;
       - **c)** Liver biopsy findings consistent with Wilson's disease;
       - **d)** 24-hour urinary copper > 40 mcg/24 hours; AND
   - **B)** Patient meets ONE of the following criteria (i, ii, iii, or iv):
     - **i.** Patient has tried Galzin (zinc acetate capsules); OR
     - **ii.** Patient has tried another zinc product (e.g., zinc sulfate, zinc gluconate, zinc acetate); OR
     - **iii.** According to the prescriber, patient has symptoms of Wilson's disease and zinc would not be an appropriate therapy; OR
     - **iv.** Patient has been started on therapy with a penicillamine product; AND
   - **C)** Patient meets ONE of the following (i or ii):
     - **i.** Generic penicillamine tablets are requested; OR
     - **ii.** If brand Depen is prescribed, patient has tried generic penicillamine tablets AND cannot take generic penicillamine tablets due to a formulation difference in the inactive ingredient(s) [e.g., difference in dyes, fillers, preservatives] between the Brand and the bioequivalent generic product which, per the prescriber, would result in a significant allergy or serious adverse reaction; AND
   - **D)** The medication is prescribed by or in consultation with a gastroenterologist, hepatologist, or liver transplant physician.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of penicillamine products is not recommended in the following situations:

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria.

Criteria will be updated as new published data are available.

REFERENCES

1. Cuprimine® capsules [prescribing information]. Bridgewater, NJ. Valeant; November 2019.

2. Depen® tablets [prescribing information]. Somerset, NJ. Meda; January 2019.
3. Roberts EA, Schilsky MI. AASLD Practice Guidelines: Diagnosis and treatment of Wilson disease: an update. *Hepatology*. 2008;47(6):2089-2111.
4. European Association for Study of the Liver (EASL) clinical practice guidelines: Wilson's disease. *J Hepatol*. 2012;56(3):671-85.
5. Fraenkel L, Bathon JM, England BR, et al. 2021 American College of Rheumatology Guideline for the treatment of rheumatoid arthritis. *Arthritis Care Res (Hoboken)*. 2021 Jul;73(7):924-939.