I. Medication Description

Penicillamine is a chelating agent recommended for the removal of excess copper in patients with Wilson's disease. From *in vitro* studies which indicate that one atom of copper combines with two molecules of penicillamine, it would appear that one gram of penicillamine should be followed by the excretion of about 200 milligrams of copper; however, the actual amount excreted is about one percent of this. Penicillamine also reduces excess cystine excretion in cystinuria. This is done, at least in part, by disulfide interchange between penicillamine and cystine, resulting in formation of penicillamine-cysteine disulfide, a substance that is much more soluble than cystine and is excreted readily. Penicillamine interferes with the formation of cross-links between tropocollagen molecules and cleaves them when newly formed. The mechanism of action of penicillamine in rheumatoid arthritis is unknown although it appears to suppress disease activity. Unlike cytotoxic immunosuppressants, penicillamine markedly lowers IgM rheumatoid factor but produces no significant depression in absolute levels of serum immunoglobulins. Also unlike cytotoxic immunosuppressants which act on both, penicillamine *in vitro* depresses T-cell activity but not B-cell activity.

II. Position Statement

Coverage is determined through a prior authorization process with supporting clinical documentation for every request.

III. Policy

Coverage of oral penicillamine products can be provided when the following conditions are confirmed:

- Cystinuria
- Lead toxicity
- Rheumatoid arthritis
  - Prescribed by a rheumatologist **AND**
  - Used for severe, active disease that has not responded to conventional therapy
- Systemic sclerosis:
  - Prescribed by a rheumatologist or other pertinent specialist **AND**
  - Used for rapidly progressive disease **AND**
  - Disease is unable to be managed with other pharmacologic therapies
- Wilson’s disease

IV. Quantity Limitations

240 capsules/tablets per each 30 day period are covered. Additional quantities may be considered for coverage on a case-by-case basis.
V. Coverage Duration

Coverage is provided for the following durations and may be renewed in cases where medically necessary:

- Cystinuria: up to 12 months
- Lead toxicity: up to 12 weeks
- Rheumatoid arthritis: up to 4 months for initial coverage; may be renewed in 6 month intervals
- Systemic sclerosis: up to 6 months
- Wilson’s disease: up to 12 months

VI. Coverage Renewal Criteria

Coverage can be renewed based upon the following criteria:

- Stabilization of disease or in absence of disease progression AND
- Absence of unacceptable toxicity from the drug

VII. Billing/Coding Information

- Cuprimine: 250mg oral capsules
- Depen: 250mg oral tablets

VIII. Summary of Policy Changes

6/15/16: new policy

IX. References


The Plan fully expects that only appropriate and medically necessary services will be rendered. The Plan reserves the right to conduct pre-payment and post-payment reviews to assess the medical appropriateness of the above-referenced therapies.

Drug therapy initiated with samples will not be considered as meeting medical necessity for coverage for non-preferred or prior authorized medications.

The preceding policy applies only to members for whom the above named pharmacy benefit medications are included on their covered formulary. Members with closed formulary benefits are subject to trying all appropriate formulary alternatives before a coverage exception for a non-formulary agent will be considered.

The preceding policy is a guideline to allow for coverage of the pertinent medication/product, and is not meant to serve as a clinical practice guideline.