

SPECIALTY GUIDELINE MANAGEMENT

THIOLA (tiopronin) and THIOLA EC (tiopronin delayed release tablets)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

Thiola and Thiola EC are indicated, in combination with high fluid intake, alkali, and diet modification, for the prevention of cystine stone formation in adult and pediatric patients 20 kg and greater with severe homozygous cystinuria who are not responsive to these measures alone.

All other indications are considered experimental/investigational and not medically necessary.

II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:

A. Initial requests:

1. Genetic testing results showing mutations in both alleles of the SLC3A1 or SLC7A9 genes; and
2. Lab results documenting baseline urinary cystine levels.

B. Continuation of therapy requests: lab results documenting a decrease in urinary cystine levels compared to baseline.

III. CRITERIA FOR INITIAL APPROVAL

Cystinuria

Authorization of 12 months may be granted for prevention of cystine stone formation in a member with severe homozygous cystinuria when all of the following criteria are met:

- A. Diagnosis of homozygous cystinuria was confirmed by genetic testing showing mutations in both alleles of the SLC3A1 or SLC7A9 genes.
- B. The requested medication is being used as an adjunct to high fluid intake, alkali, and diet modification.
- C. The member has elevated urinary cysteine levels at baseline.

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment of severe cystinuria in members requesting reauthorization who have experienced a decrease in urinary cystine levels compared to pretreatment baseline.

V. REFERENCES

1. Thiola [package insert]. San Antonio, TX: Mission Pharmaceutical Company; June 2019.

Reference number(s)
2991-A

2. Thiola EC [package insert]. San Antonio, TX: Mission Pharmaceutical Company; June 2019.
3. Biyani CS, Cartledge JJ. Cystinuria- diagnosis and management. *Eur Urology*. 2006; 4:175-183.
4. Pearle MS, Goldfarb DS, Assimos DG, Curhan G, Denu-Ciocca CJ, Matlaga BR, Monga M, Penniston KL, Preminger GM, Turk TM, White JR; American Urological Association. Medical management of kidney stones: AUA guideline. *J Urol*. 2014 Aug;192(2):316-24.