Reference number(s)
2991-A

SPECIALTY GUIDELINE MANAGEMENT

THIOLA (tiopronin)
THIOLA EC (tiopronin delayed-release tablets)
tiopronin (generic)
tiopronin delayed-release tablets (generic)

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 Indication
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, stone analysis, or urine microscopy supporting diagnosis; and
   2. Lab results documenting baseline urinary cystine levels.
B. Continuation of therapy requests: lab results or chart notes documenting a benefit from therapy.

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 cystinuria was established by one or more of the following:
   1. Biallelic mutations/variants in the SLC3A1 or the SLC7A9 gene confirmed by genetic testing.
   2. Stone analysis revealing 100 percent cystine calculi.
   3. Presence of pathognomonic hexagonal cystine crystals visualized on urine microscopy.
B. The requested medication is being used as an adjunct to high fluid intake, alkali, and diet modification.
C. The member has elevated urinary cystine levels at baseline.

IV. CONTINUATION OF THERAPY

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Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section III who are experiencing benefit from therapy (e.g., a decrease in urinary cystine levels compared to pretreatment baseline, reduction in stone formation/growth).

V. REFERENCES

1. Thiola [package insert]. San Antonio, TX: Mission Pharmacal Company; January 2021.
2. Thiola EC [package insert]. San Antonio, TX: Mission Pharmacal Company; March 2021.
3. Biyani CS, Cartledge JJ. Cystinuria- diagnosis and management. *Eur Urology*. 2006; 4:175-183.
4. Pearle MS, Goldfarb DS, Assimos DG, et al: Medical management of kidney stones: AUA Guideline. J Urol 2014; 192: 316.
5. Goldfarb DS, et al. Cystinuria and cystine stones. UpToDate, Lam, AQ (Ed), Waltham, MA, 2023. URL: www.uptodate.com. Accessed February 2, 2024.
6. Tiopronin [package insert]. Parsippany, NJ: Teva Pharmaceuticals USA, Inc.; February 2021.
7. Tiopronin delayed release [package insert]. Basking Ridge, NJ: Torrent Pharma Inc.; February 2024.

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