

Specialty Guideline Management

tiopronin products

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Thiola	tiopronin
Thiola EC	tiopronin delayed-release tablets
Venxxiva	tiopronin delayed-release tablets

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^{1,2,3,7,8}

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.

Documentation

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

Initial requests

- Genetic testing results, stone analysis, or urine microscopy supporting diagnosis; and
- Lab results documenting baseline urinary cystine levels.

Continuation of therapy requests

Lab results or chart notes documenting a benefit from therapy.

Coverage Criteria

Cystinuria¹⁻⁸

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:

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

Continuation of Therapy

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

References

1. Thiola [package insert]. San Antonio, TX: Mission Pharmacal Company; January 2024.
2. Thiola EC [package insert]. San Antonio, TX: Mission Pharmacal Company; March 2021.
3. Venxxiva [package insert]. Cambridge, UK: Cycle Pharmaceuticals LTD; December 2024.

Reference number(s)
2991-A

4. Biyani CS, Cartledge JJ. Cystinuria- diagnosis and management. *Eur Urology*. 2006; 4:175-183.
5. Pearle MS, Goldfarb DS, Assimos DG, et al: Medical management of kidney stones: AUA Guideline. *J Urol* 2014; 192: 316.
6. Goldfarb DS, et al. Cystinuria and cystine stones. UpToDate, Lam, AQ (Ed), Waltham, MA, 2023. URL: www.uptodate.com. Accessed February 11, 2025.
7. Tiopronin [package insert]. Parsippany, NJ: Teva Pharmaceuticals USA, Inc.; February 2021.
8. Tiopronin delayed release [package insert]. Parsippany, NJ : Teva Pharmaceuticals USA, Inc.; June 2024.