Generic Name: Tiopronin

Therapeutic Class or Brand Name: Thiola®)

Applicable Drugs: N/A

Preferred: Tiopronin tablets (generic)

Non-preferred: Thiola, Thiola EC

Date of Origin: 5/7/2015

Date Last Reviewed / Revised: 6/17/2025

PRIOR AUTHORIZATION CRITERIA

(May be considered medically necessary when criteria I through VI are met)

I. Documentation that Thiola is used for the prevention of cystine (kidney) stone formation in patients with severe homozygous cystinuria.

II. Documentation that urinary cystine is greater than 500 mg/day.

III. Documented likely resistance or contraindication to a three-month trial with conservative measures of high fluid intake, alkali, and diet modification.

   A. Minimum age requirement: 9 years old.

   B. Minimum weight for pediatric patients is 20 kg.

IV. Treatment must be prescribed by or in consultation with a nephrologist or urologist.

V. Request is for a medication with the appropriate FDA labeling, or its use is supported by current clinical practice guidelines.

VI. Refer to the plan document for the list of preferred products. If the requested agent is not listed as a preferred product, must have documented treatment failure or contraindication to the preferred product(s).

EXCLUSION CRITERIA

- History of developing agranulocytosis, aplastic anemia, or thrombocytopenia while on Thiola.

OTHER CRITERIA

- N/A

QUANTITY / DAYS SUPPLY RESTRICTIONS

- Dosing should be based on the amount required to maintain urinary cystine concentration below its solubility limit (generally <250 mg/L).
  - Adult: usual starting dose of 800 mg/day
    - Average dosage in clinical studies was about 1,000 mg/day
  - Pediatric ≥20 kg: usual starting dose of 15 mg/kg/day

- Not to exceed 50 mg/kg/day

- The quantity is limited to a maximum of a 30-day supply per fill.

APPROVAL LENGTH

- **Authorization:** 6 months.
- **Re-Authorization:** 1 year. An updated letter of medical necessity or progress notes showing current medical necessity criteria are met and that the medication is effective. Must include documentation that the urinary cystine concentration is below its solubility limit (generally < 250 mg/liter) and that it is being measured at least every 6 months.

APPENDIX

N/A

REFERENCES

1. Pearle MS, Goldfarb DS, Assimos DG, et al. Medical management of kidney stones: AUA guideline. J Urol. 2014;192(2):316-324. doi:10.1016/j.juro.2014.05.006. Accessed June 17, 2025.

   https://pubmed.ncbi.nlm.nih.gov/24857648/

2. Qaseem A, Dallas P, Forciea MA, Starkey M, Denberg TD; Clinical Guidelines Committee of the American College of Physicians. Dietary and pharmacologic management to prevent recurrent nephrolithiasis in adults: a clinical practice guideline from the American College of Physicians. Ann Intern Med. 2014;161(9):659-667. doi:10.7326/M13-2908. Accessed June 17, 2025.

   https://pubmed.ncbi.nlm.nih.gov/25364887/

3. Thiola. Prescribing information. Mission Pharmacal Company; 2021. Accessed June 17, 2025.

   https://thiolaec.com/sites/default/files/pdfs/nda/Thiola_PI.pdf.

4. Tiselius HG. New horizons in the management of patients with cystinuria. Curr Opin Urol. 2010;20(2):169-173. doi:10.1097/MOU.0b013e328333b674. Accessed June 17, 2025.

   https://pubmed.ncbi.nlm.nih.gov/19887942/

5. Barbey F, Joly D, Rieu P, Méjean A, Daudon M, Jungers P. Medical treatment of cystinuria: critical reappraisal of long-term results. J Urol. 2000;163(5):1419-1423. doi:10.1016/s0022-5347(05)67633-1. Accessed June 17, 2025.

   https://pubmed.ncbi.nlm.nih.gov/10751848/