

Prior Authorization DRUG Guidelines

Cystagon (cysteamine)

Effective Date: 10/22/13

Date Developed: 9/3/13 by Albert Reeves MD

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Cystagon, a degradation product of the amino acid cysteine, lowers the cystine content of cells in patients with cystinosis (an inherited defect of cystine lysosomal transport).

Pre-Authorization Criteria: treatment for documented nephropathic or systemic cystinosis

Adult Dosing:

Immediate release: Adults: 2 g daily in 4 divided doses; maximum dose: 90 mg/kg/d

Delayed release: 1.3 g/m²/day divided every 12 hours; may increase in 10% increments to a maximum dose of 1.95 g/m²/day.

Note: Initiate therapy with $\frac{1}{6}$ to $\frac{1}{4}$ of maintenance dose; titrate slowly upward over 4-6 weeks.

Child and Adolescent Dosing:

Children and Adolescents weighing ≤ 50 kg: 1.3 g/m²/day or 60 mg/kg/day divided into four doses; maximum dose: 1.95 g/m²/day or 90 mg/kg/day; $c > 50$ kg, refer to adult dosing

PRECAUTIONS: osteopenia/pathologic fractures; pseudotumor cerebri; leukopenia; GI upset; uricaria

DRUG INTERACTIONS: none listed

REFERENCES

1. Dohil R, Fidler M, Gangoiti JA, et al, "Twice-Daily Cysteamine Bitartrate Therapy for Children With Cystinosis," *J Pediatr*, 2010, 156(1):71-75.e1-3. [PubMed [19775699](#)]
2. Gahl WA, Thoene JG, and Schneider JA, "Cystinosis," *N Engl J Med*, 2002, 347(2):111-21. [PubMed [12110740](#)]
3. Langman CB, Greenbaum LA, Sarwal M, et al, "A Randomized Controlled Crossover Trial With Delayed-Release Cysteamine Bitartrate in Nephropathic Cystinosis: Effectiveness on White Blood Cell Cystine Levels and Comparison of Safety," *Clin J Am Soc Nephrol*, 2012, 7(7):1112-20. [PubMed [22554716](#)]

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