

Prior Authorization DRUG Guidelines

Cystagon (cysteamine)

Effective Date: 10/22/13

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

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Cysteamine is a degradation product of the amino acid cysteine. It lowers the cystine content of cells in patients with cystinosis (an inherited defect of cystine lysosomal transport) by conversion to cysteine, which can then exit the lysosome.

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

Adult Dosing:

Immediate release: Adults: **Initial:** $\frac{1}{6}$ to $\frac{1}{4}$ of target maintenance dose administered in 4 divided doses; gradually increase dose over 4 to 6 weeks to a target maintenance dose of 500 mg 4 times daily (maximum: 1.95 g/m²/day).

Delayed release: 0.2 to 0.3 g/m²/day in 2 divided doses every 12 hours; increase dose gradually over 4 to 6 weeks to target maintenance dose of 1.3 g/m²/day in 2 divided doses every 12 hours. May further increase in 10% increments if needed up to a maximum daily dose.

Child and Adolescent Dosing:

1.3 g/m²/day divided into 4 doses up to 2,000 mg/day (500 mg 4 times daily); maximum daily dose: 1.95 g/m²/day

NOTE: To avoid intolerance, initiate therapy with $\frac{1}{6}$ to $\frac{1}{4}$ of maintenance dose (see below for maintenance doses); titrate slowly upward over 4 to 6 weeks. Dosage adjustments should be made based on target WBC cystine levels.

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

DRUG INTERACTIONS: none listed

DOSAGE FORMS:

Capsule, Oral: Cystagon: 50 mg, 150 mg; Delayed Release: 25 mg, 75 mg

REFERENCES

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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.
- 3 Cystagon (cysteamine) [prescribing information]. Morgantown, WV: Mylan Pharmaceuticals Inc; August 2021.
- 4 Belldina EB, Huang MY, Schneider JA, et al. Steady-state pharmacokinetics and pharmacodynamics of cysteamine bitartrate in paediatric nephropathic cystinosis patients. *Br J Clin Pharmacol*. 2003;56(5):520-525
- 5 Bouazza N, Tréluyer JM, Ottolenghi C, et al. Population pharmacokinetics and pharmacodynamics of cysteamine in nephropathic cystinosis patients. *Orphanet J Rare Dis*. 2011;6:86. doi: 10.1186/1750-1172-6-86

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