VENTURA COUNTY
HEALTH CARE PLAN

Prior Authorization DRUG Guidelines

Cystagon (cysteamine)

Effective Date: 10/22/13

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

Last Approval Date: 1/26/16, 1/24/17, 1/23/18, 1/22/19, 2/18/20; 8/3/21, 2/1/22, 1/31/23, 2/13/24

Cystagon, 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: 2 g daily in 4 divided doses; maximum dose: 90 mg/kg/d

Delayed release: 1.3 g/m^2/day divided every 12 hours; may increase in 10%

increments to a maximum dose of 1.95 g/m^2/day.

Note: Initiate therapy with $^1/_6$ to $^1/_4$ of maintenance dose; titrate slowly upward over 4-6 weeks.

Child and Adolescent Dosing:

Children and Adolescents weighing ≤50 kg: 1.3 g/m^2/day or 60 mg/kg/day divided into four doses; maximum dose: 1.95 g/m^2/day or 90 mg/kg/day; c>50kg, refer to adult dosing

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

DRUG INTERACTIONS: none listed

DOSAGE FORMS:

Capsule, Oral: Cystagon: 50 mg, 150 mg

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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