Effective Date 8/1/2023
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Coverage Policy Number IP0082

Cysteamine Ophthalmic Solution

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Related Coverage Resources

INSTRUCTIONS FOR USE
The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This policy supports medical necessity review for **Cystadrops®** (cysteamine ophthalmic solution) 0.37% and **Cystaran™** (cysteamine ophthalmic solution) 0.44%.

Coverage Policy

Cystadrops and Cystaran (cysteamine ophthalmic solution) are considered medically necessary when the following are met:

1. **Cystinosis, Corneal Cysteine Crystal Deposits.** Individual meets **BOTH** of the following (A and B):
   A. Individual has corneal cysteine crystal deposits confirmed by slit-lamp examination
   B. Prescribed by or in consultation with an ophthalmologist or a metabolic disease specialist (or specialist who focuses on the treatment of metabolic diseases)

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Note: Receipt of sample product does not satisfy any criteria requirements for coverage.

Reauthorization Criteria

Cystadrops and Cystaran (cysteamine ophthalmic solution) are considered medically necessary for continued use when initial criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval and reauthorization duration is 12 months.

Conditions Not Covered

Cystadrops and Cystaran (cysteamine ophthalmic solution) are considered experimental, investigational, or unproven for ANY other use.

Background

Overview
Cystamine ophthalmic solution is a cystine-depleting agent indicated for the treatment of corneal cystine crystal accumulation in patients with cystinosis.[1,2]

Disease Overview
Cystinosis is a rare autosomal recessive inborn error of metabolism in which cystine accumulates within lysosomes and forms crystals in many tissues, including the kidneys, liver, bone marrow, pancreas, muscle, rectal mucosa, brain, and eye.[3] Cystine deposits in the cornea cause photophobia. Patients may present only with corneal crystal deposition but no associated systemic manifestations; the kidney, retina, and other organs are free of cystine accumulation in these patients. In patients without systemic symptoms, diagnosis of ocular cystinosis is often in adulthood when corneal crystal deposits are noted on ocular examination.[4] Of note, with oral cysteamine the concentration obtained in corneal tissue is inadequate and does not affect corneal cystine crystals. Topical treatment is required to dissolve existing cystine crystals.

References

1. Cystadrops ophthalmic solution [prescribing information]. Lebanon, NJ: Recordati Rare Diseases; September 2020.
2. Cystaran ophthalmic solution [prescribing information]. Gaithersburg, MD: Leadiant Biosciences; March 2022.
3. Wilmer MJ, Schoeber JP, van den Heuvel LP, Levtchenko EN. Cystinosis: practical tools for diagnosis and treatment. Pediatr Nephrol. 2011; 26(2): 205–215.
4. Biswas S, Gaviria M, Malheiro L, et al. Latest clinical approaches in the ocular management of cystinosis: a review of current practice and opinion from the ophthalmology cystinosis forum. Ophthalmol Ther. 2018;7(2):307-322.