



Drug Coverage Policy

Effective Date6/15/2025

Coverage Policy Number.....IP0082

Policy Title..... Cysteamine Ophthalmic Solution

Metabolic Disorders – Cysteamine Ophthalmic Solution

- Cystadrops® (cysteamine 0.37% ophthalmic solution – Recordati Rare Diseases)
- Cystaran® (cysteamine 0.44% ophthalmic solution – Leadiant Biosciences)

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. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted for services that are not accompanied by covered code(s) under the applicable Coverage Policy will be denied as not covered. 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

Cysteamine ophthalmic solution, a cystine-depleting agent, is 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.

Coverage Policy

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Cysteamine ophthalmic solution. All approvals are provided for the duration noted below. In cases where the approval is authorized in months, 1 month is equal to 30 days. Because of the specialized skills required for evaluation and diagnosis of patients treated with Cysteamine ophthalmic solution as well as the monitoring required for adverse events and long-term efficacy, approval requires Cysteamine ophthalmic solution to be prescribed by a physician who has consulted with or who specializes in the condition.

Cysteamine ophthalmic solution is considered medically necessary when the following criteria are met:

FDA-Approved Indication

- 1. Cystinosis, Corneal Cysteine Crystal Deposits.** Approve for 1 year if the patient meets the following (A and B):
 - A)** Patient has corneal cysteine crystal deposits confirmed by slit-lamp examination; **AND**
 - B)** The medication is 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.

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

Conditions Not Covered

Any other use is considered experimental, investigational, or unproven (criteria will be updated as new published data are available).

References

1. Cystadrops® ophthalmic solution [prescribing information]. Lebanon, NJ: Recordati Rare Diseases; August 2020.

2. Cystaran® ophthalmic solution [prescribing information]. Gaithersburg, MD: Leadiant Biosciences; May 2023.

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.

Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	No criteria change.	7/15/2024
Annual Revision	No criteria change.	6/15/2025

The policy effective date is in force until updated or retired.

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