



PRIOR AUTHORIZATION POLICY

- POLICY:** Metabolic Disorders – Cysteamine Ophthalmic Solution Prior Authorization Policy
- Cystadrops® (cysteamine 0.37% ophthalmic solution – Recordati Rare Diseases)
 - Cystaran® (cysteamine 0.44% ophthalmic solution – Leadiant Biosciences)

REVIEW DATE: 03/12/2025

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 AND HAVE DISCRETION IN MAKING INDIVIDUAL COVERAGE DETERMINATIONS. 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.

CIGNA NATIONAL FORMULARY COVERAGE:

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.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of cysteamine ophthalmic solution. All approvals are provided for the duration noted below. 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 or in consultation with a physician who specializes in the condition being treated.

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

is(are) covered as medically necessary when the following criteria is(are) met for FDA-approved indication(s) or other uses with supportive evidence (if applicable):

FDA-Approved Indication

1. Cystinosis, Corneal Cysteine Crystal Deposits. Approve for 1 year if the patient meets BOTH of 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 in the treatment of metabolic diseases).

CONDITIONS NOT COVERED

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

is(are) considered experimental, investigational or unproven for ANY other use(s) including the following; 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.

HISTORY

Type of Revision	Summary of Changes	Review Date
Annual Revision	No criteria changes.	03/29/2023
Annual Revision	No criteria changes.	03/20/2024
Annual Revision	No criteria changes.	03/12/2025

"Cigna Companies" refers to operating subsidiaries of The Cigna Group. All products and services are provided exclusively by or through such operating subsidiaries, including Cigna Health and Life Insurance Company, Connecticut General Life Insurance Company, Evernorth Behavioral Health, Inc., Cigna Health Management, Inc., and HMO or service company subsidiaries of The Cigna Group. © 2025 The Cigna Group.