Subject: Corneal Cross-Linking for Keratoconus and Ectasia

Medical Policy #: 28.0

Original Effective Date: 09-26-2018

Status: Reviewed

Last Annual Review Date: 03-26-2025

Disclaimer

Refer to the member's specific benefit plan and Schedule of Benefits to determine coverage. This may not be a benefit on all plans or the plan may have broader or more limited benefits than those listed in this Medical Policy.

Description

Keratoconus is a corneal disorder characterized by progressive corneal thinning, ectasia, and induced irregular astigmatism, which can lead to impaired vision. Available treatment options include rigid contact lenses, intracorneal ring segments, in advanced cases of keratoplasty. For progressive keratoconus cases, corneal cross-linking (CXL) is now used resulting enhancements in mechanical strength, provision of biochemical stability, and slowing or preventing progression.

DESCRIPTION OF TECHNOLOGY/THERAPY:
Conventional, epithelium-off, corneal collagen crosslinking (C-CXL) involves the use of riboflavin (vitamin B(2)) and ultraviolet-A (UVA) radiation. Only the use of U.S. Food and Drug Administration (FDA) approved drug/device system (e.g., Photrexa® Viscous or Photrexa® with the KXL® System) is considered medically necessary.

CLINICAL Features:
Patients present at puberty or early adulthood with blurry vision or a sudden decrease in visual acuity. The condition may progress throughout life, though progression often slows or halts after the fourth decade.

Clinical features which may more specifically suggest the diagnosis include the following:

1. **Asymmetric visual complaints** – Although keratoconus is usually a bilateral disease, patients may present with asymmetric symptoms as one eye may be much more severely affected than the other.
2. **Difficulty with visual correction** – As the disease progresses, patients experience difficulties with spectacle correction and contact lens fitting.
3. **Stereotypical findings on Corneal Topography/Tomography - such** as I/S asymmetry, skewing of axis, abnormal elevation of posterior and anterior surfaces, abnormal thinning and displacement of corneal apices
4. **Biomicroscopic stigmata** – such apical thinning, Vogt's Striae, and Fleischer Ring
5. **Munson's sign** – In advanced keratoconus, patients may have a v-shaped indentation of the lower eyelid on downgaze caused by a large protuberant cone.
6. **Corneal hydrops** – In advanced keratoconus, some patients can present with photophobia and a sudden painful drop in visual acuity that is due to corneal hydrops. The symptoms are caused by the sudden onset of severe corneal edema.

Coverage Determination

Prior Authorization is required. Logon to Pres Online to submit a request: https://ds.phs.org/preslogin/index.jsp

For Commercial, Medicare and Medicaid.

Conventional, epithelium-off, corneal collagen crosslinking (C-CXL) using an FDA approved drug/device system (e.g., Photrexa® Viscous or Photrexa® with the KXL® System).

PHP considers epithelium-off photochemical, collagen cross-linkage using riboflavin/ultraviolet-A, medically necessary for the treatment of Severe Keratoconus:

1. Progressive keratoconus or corneal ectasia must meet 1 or more of the following:
   a. An increase of 1 diopter (D) in the steepest keratometry value,
   b. An increase of 1 D in regular astigmatism evaluated by subjective manifest refraction,
   c. A myopic shift (decrease in the spherical equivalent) of 0.50 D on subjective manifest refraction,
   d. A decrease ≥0.1 mm in the back optical zone radius in rigid contact lens wearers where other information was not available.
2. When **ALL** of the following criteria are met:
   a. age 14–65 years

   AND

b. progression of the condition as defined above

AND

c. Absence of visual disturbance from a significant central corneal opacity or other eye disease (e.g., herpetic keratitis, neurotrophic keratopathy)

AND

d. Corneal thickness of at least 400 microns;

AND

e. Nonpregnant individuals

Exclusions

1. Corneal collagen cross-linking is considered experimental, investigational or unproven for any other indication including when combined with a second refractive procedure.
2. All other corneal collagen crosslinking procedures (e.g., epithelium-on/transepithelial) or non-FDA approve procedures are considered experimental, investigational or unproven.
3. The performance of photochemical collagen cross-linkage in combination with other procedures (CXL-plus) (e.g., intrastromal corneal ring segments, PRK or phakic intra-ocular lens implantation) experimental and investigational.

Coding

The coding listed in this medical policy is for reference only. Covered and non-covered codes are within this list.

CPT® code	Description
0402T	Collagen cross-linking of cornea (including removal of the corneal epithelium and intraoperative pachymetry when performed) (Report medication separately) ******Do not report 0402T in conjunction with 65435, 69990, 76514

HCPCS Code	Description
J2787	Riboflavin 5' phosphate, ophthalmic solution, up to 3 ml

ICD-10 Codes	Covered if selection criteria are met:
H18.601	Keratoconus, unspecified, right eye
H18.602	Keratoconus, unspecified, left eye
H18.603	Keratoconus, unspecified, bilateral
H18.609	Keratoconus, unspecified, unspecified eye
H18.611	Keratoconus, stable, right eye
H18.612	Keratoconus, stable, left eye
H18.613	Keratoconus, stable, bilateral
H18.619	Keratoconus, stable, unspecified eye
H18.621	Keratoconus, unstable, right eye
H18.622	Keratoconus, unstable, left eye
H18.623	Keratoconus, unstable, bilateral
H18.629	Keratoconus, unstable, unspecified eye
H18.711	Corneal ectasia, right eye
H18.712	Corneal ectasia, left eye
H18.713	Corneal ectasia, bilateral
H18.719	Corneal ectasia, unspecified eye

Reviewed by / Approval Signatures

Population Health & Clinical Quality Committee: Clinton White MD
Senior Medical Director: Jim Romero MD
Date Approved: 03-26-2025
Reviewed by: Kenneth D. Himmel, MD – Eye Associates of New Mexico

References

1. CMS, NCD - 80.7, Refractive Keratoplasty, V1, Effective Date: 05/01/1997. [Cited 02/10/2025]

Not every Presbyterian health plan contains the same benefits. Please refer to the member's specific benefit plan and Schedule of Benefits to determine coverage [MPMPPC051001].

2. MCG, Corneal Cross-Linking, 28th Edition, ACG: A-1040 (AC), Last Update: 3/14/2024 [Cited 02/10/2025]
3. UpToDate Inc., Keratoconus, Author: Laura L Wayman, MD, Literature review current through: Jan 2025, Updated May 31, 2024. [Cited 02/10/2025]
4. Hayes, Health Technology Assessment, Comparative Effectiveness of Corneal Cross-Linking for Treatment of Keratoconus, Annual Review Jan 13, 2022. [Cited 02/10/2025]
5. Hayes, Health Technology Assessment, Conventional Corneal Collagen Cross-Linking for Treatment of LASIK-Related Ectasia, Mar 24, 2023 [Cited 02/10/2025]
6. Aetna, Corneal Remodeling, Number: 0023, Next review 01/09/2025. [Cited 02/10/2025]
7. Humana, Keratoconus Surgical Treatments – Effective Date: 07/25/2024, Review Date: 07/25/2024 Policy Number: HUM-0314-019, Line of Business: Commercial. [Cited 02/10/2025]
8. Glaukos, Photrexa Viscous and Photrexa ISA (for Riboflavin), Revised: 11/2018. [Cited 02/10/2025]

Publication History

09-26-18	Policy created September 26, 2018
01-22-20	Annual review. No change to Hayes. Added additional references and updated HCPCS code (J2787) for riboflavin. Update: On 04-02-2020, CPT 0402T is added to PA grid.
03-24-21	Annual review. Reviewed by Medical Directors on 02/02/2021. No change. Will continue to use the current criteria and to leave CPT 0402T and J2787 on the PA grid.
03-23-22	Annual review. Reviewed by PHP Medical Policy Committee on 03-02-2022. No change. Continue to use the homegrown criteria for Medicare, Medicaid and commercial. Aetna, Humana and Cigna cover CXL for keratoconus are comparable. There is no NCD or LCD identified for CXL for treatment of keratoconus. Continue PA requirement for 0402T and riboflavin (managed by Pharmacy), HCPCS code (J2787).
03-22-23	Annual review. Reviewed by PHP Medical Policy Committee on 02-03-2023. The age requirement has changed, from “age less than 65 years” to “age 14–65 years”. Code 0402T will continue to require PA.
03-20-24	Annual review. Reviewed by PHP Medical Policy Committee on 01/05/2024. Minor formatting adjustments were done in section two under coverage indication. There has been no change in coverage with this revision. Continue coverage for Medicare, Medicaid and commercial. Continue PA requirement for 0402T. The listed ICD-10 codes are in good standing.
03-26-25	Annual review. Reviewed by PHP Medical Policy Committee on 02/14/2025. No change. Continue with the criteria stated and continue to require PA for 0402T to monitor misuse.

This Medical Policy is intended to represent clinical guidelines describing medical appropriateness and is developed to assist Presbyterian Health Plan and Presbyterian Insurance Company, Inc. (Presbyterian) Health Services staff and Presbyterian medical directors in determination of coverage. The Medical Policy is not a treatment guide and should not be used as such.

For those instances where a member does not meet the criteria described in these guidelines, additional information supporting medical necessity is welcome and may be utilized by the medical director in reviewing the case. Please note that all Presbyterian Medical Policies are available online at: ***Click here for Medical Policies***

Web links:

At any time during your visit to this policy and find the source material web links has been updated, retired or superseded, PHP is not responsible for the continued viability of websites listed in this policy.

When PHP follows a particular guideline such as LCDs, NCDs, MCG, NCCN etc., for the purposes of determining coverage; it is expected providers maintain or have access to appropriate documentation when requested to support coverage. See the References section to view the source materials used to develop this resource document.