Corneal Cross-Linking for Keratoconus and Ectasia
MPM 28.0

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 these criteria.

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., Photrex® Viscous or Photrex® 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](https://ds.phs.org/preslogin/index.jsp)

Conventional, epithelium-off, corneal collagen crosslinking (C-CXL) using a 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 less than 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
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Exclusion

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 intraocular 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.


<table>
<thead>
<tr>
<th>CPT Codes</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0402T</td>
<td>Collagen cross-linking of cornea (including removal of the corneal epithelium and intraoperative pachymetry when performed)</td>
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HCPCS Code

<table>
<thead>
<tr>
<th>HCPCS Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J2787</td>
<td>Riboflavin 5’ phosphate, ophthalmic solution, up to 3 ml</td>
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ICD-10 Diagnosis Codes

<table>
<thead>
<tr>
<th>ICD-10 Codes</th>
<th>Covered if selection criteria are met:</th>
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<tbody>
<tr>
<td>H18.601</td>
<td>Keratoconus, unspecified, right eye</td>
</tr>
<tr>
<td>H18.602</td>
<td>Keratoconus, unspecified, left eye</td>
</tr>
<tr>
<td>H18.603</td>
<td>Keratoconus, unspecified, bilateral</td>
</tr>
<tr>
<td>H18.609</td>
<td>Keratoconus, unspecified, unspecified eye</td>
</tr>
<tr>
<td>H18.611</td>
<td>Keratoconus, stable, right eye</td>
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<td>H18.612</td>
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<tr>
<td>H18.613</td>
<td>Keratoconus, stable, bilateral</td>
</tr>
<tr>
<td>H18.619</td>
<td>Keratoconus, stable, unspecified eye</td>
</tr>
</tbody>
</table>
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ICD-10 Codes | Covered if selection criteria are met:
-------------|-----------------------------------
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

Ernest Christman, MD - Ophthalmologist
Kenneth D. Himmel, MD – Eye Associates of New Mexico

References


3. Hayes, a Division of TractManager, Comparative Effectiveness of Corneal Cross-Linking for Treatment of Keratoconus, Annual Review Mar 15, 2019. [Cited 12/04/2019]

4. 3M- CPT Assistant, February 2016, Page: 12 Category Coding Brief: Corneal Collagen Cross-Linking (042T)


6. Hayes, a Division of TractManager, Evidence Analysis Research Brief, Concurrent Corneal Cross-Linking and Intacs Implantation for Treatment of Keratoconus, Apr 25, 2019. [Cited 12/04/2019]

7. Aetna, Corneal Remodeling, Number: 0023, Last review 05/22/2019, (see VII Collagen Cross-Linking for Keratoconus. [Cited 12-04-2019]

8. CMS, LCD: Services That Are Not Reasonable and Necessary (L35094), Revision date 09/12/2019, R41 with related Article (A56967), Rev date: 01/01/2020, R1. [Cited 01/24/2020]

Approval Signatures

Clinical Quality/Utilization Mgmt. Committee: Howard Epstein, MD
Medical Directory: Jim Romero, MD

Approval Date

January 22, 2020
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Publications 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

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