Introduction

The cornea is the clear, dome-shaped tissue that covers the front of the eyeball. The cornea bends light rays to help focus vision. When the cornea becomes cloudy it’s called corneal opacity. This cloudiness can be from any number of eye problems such as inflammation, infection, ulcers on the eye, and many other conditions. When the cloudiness severely limits vision or leads to blindness, a corneal transplant may be done. Most corneal transplants use tissue from donors. If this surgery fails or a person isn’t a candidate for a corneal transplant using donor tissue, surgery using an artificial cornea may be an option. This policy describes when a surgery using an artificial cornea may be considered medically necessary.

Note: The Introduction section is for your general knowledge and is not to be taken as policy coverage criteria. The rest of the policy uses specific words and concepts familiar to medical professionals. It is intended for providers. A provider can be a person, such as a doctor, nurse, psychologist, or dentist. A provider also can be a place where medical care is given, like a hospital, clinic, or lab. This policy informs them about when a service may be covered.

Policy Coverage Criteria

Implantation of a keratoprosthesis is considered a high-risk procedure associated with numerous complications and probable need for additional surgery. Therefore, the likelihood of
regaining vision and the patient’s visual acuity in the opposite eye should be taken into account when considering the appropriateness of this procedure. Treatment should be restricted to centers experienced in treating this condition and staffed by surgeons adequately trained in techniques addressing implantation of this device.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Medical Necessity</th>
</tr>
</thead>
</table>
| The Boston (Dohlman-Doane) Keratoprosthesis (Boston KPro) | The Boston KPro may be considered medically necessary for the surgical treatment of severe corneal opacification under the following conditions:  
• The cornea is severely opaque and vascularized  
AND  
• Best-corrected vision is worse than 20/400 in the affected eye and worse than 20/40 in the opposite eye  
AND  
• No end-stage glaucoma or retinal detachment is present  
AND  
• The patient has at least one of the following indications:  
o History of 1 or more corneal transplant graft failures  
o Stevens-Johnson syndrome  
o Ocular cicatricial pemphigoid  
o Autoimmune conditions with rare ocular involvement  
o Ocular chemical burns  
o An ocular condition unlikely to respond favorably to primary corneal transplant surgery (eg, libel stem cell compromise or postherpetic anesthesia)  

**Note:** Patients should be able and expected to comply with postoperative care.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Investigational</th>
</tr>
</thead>
<tbody>
<tr>
<td>Permanent keratoprosthesis for all conditions not listed above</td>
<td>A permanent keratoprosthesis for all other conditions not listed in the Medical Necessity section above is considered investigational.</td>
</tr>
<tr>
<td>All other types of permanent keratoprosthesis</td>
<td>All other types of permanent keratoprosthesis not listed in the Medical Necessity section above are considered investigational.</td>
</tr>
</tbody>
</table>
Background

The cornea is a clear, dome-shaped membrane that covers the front of the eye and helps to bend light rays to give sharp vision. Diseases of the cornea can interfere with providing clear vision. Sometimes a corneal transplant, called a keratoplasty, is needed to treat corneal diseases. In certain conditions, such as Stevens-Johnson syndrome, cicatricial pemphigoid, chemical injury, or prior failed corneal transplant the survival of a transplanted cornea is poor. An artificial cornea, called akeratoprosthesis, has been developed to restore vision in patients for whom a corneal transplant is not an option.

There are different types of keratoprosthetic devices. The Dohlman-Doane Keratoprosthesis, most commonly referred to as the Boston Keratoprosthesis (Boston KPro), is manufactured under the auspices of the Harvard Medical School-affiliated Massachusetts Eye and Ear Infirmary. The AlphaCor, previously known as the Chirila keratoprosthesis (Chirila KPro), is another type of keratoprosthesis.
Summary of Evidence

The evidence for Boston KPro in individuals who have corneal blindness and failed (or are not candidates for) corneal transplantation includes several case series and systematic reviews. Relevant outcomes are observing a change in disease status, morbid events, quality of life, and treatment-related morbidity. Numerous case series have been published. Together, studies have assessed thousands of eyes. A 2015 systematic review of KPro efficacy included 22 series with a total of 2176 eyes. Systematic reviews and case series with longer follow-up (ie, at least 2 years) have shown improvement in visual outcomes in a substantial percentage of patients with Boston KPro. However, this procedure is high risk and is associated with numerous complications (eg, growth of retro prosthetic membranes) and a probable need for additional surgery. Thus careful patient selection is important. The evidence is sufficient to determine that the technology results in a significant improvement in health outcomes.

The evidence for the AlphaCor device in individuals who have corneal blindness and failed (or are not candidates for) corneal transplantation includes several case series and systematic reviews. Relevant outcomes are observing a change in disease status, morbid events, quality of life, and treatment-related morbidity. Only a few published case series have evaluated the AlphaCor device. There are insufficient data on improvement in vision outcomes using the AlphaCor device. Moreover, the device has been associated with complications, including thinning or melting of the anterior corneal surface and corneal necrosis. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have corneal blindness and have failed (or are not candidates for) corneal transplantation who receive an osteo-odonto-keratoprosthesis (OOKP), the evidence includes case series and a systematic review. Relevant outcomes are observing a change in disease status, morbid events, quality of life, and treatment-related morbidity. A 2012 systematic review of case series, all conducted outside of the United States, found high anatomic survival rates at 5 and 20 years, but vision outcomes were not well-described. OOKP is a complex surgical procedure and has been associated with a number of complications, including extrusion of the keratoprosthesis, retinal detachment, and vitreoretinal complications. The evidence is insufficient to determine the effects of the technology on health outcomes.

Ongoing and Unpublished Clinical Trials

Some currently unpublished trials that might influence this review are listed in Table 1.
Table 1. Summary of Key Trials

<table>
<thead>
<tr>
<th>NCT No.</th>
<th>Trial Name</th>
<th>Planned Enrollment</th>
<th>Completion Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ongoing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>NCT02084745</td>
<td>Timing of Glaucoma Drainage Device With Boston KPro Surgery (GDD-KPro)</td>
<td>60</td>
<td>Mar 2017</td>
</tr>
</tbody>
</table>

NCT: national clinical trial.

Clinical Input Received From Physician Specialty Societies and Academic Medical Centers

While the various physician specialty societies and academic medical centers may collaborate with and make recommendations during the review process, input received does not represent an endorsement or position statement by the physician specialty societies or academic medical centers, unless otherwise noted.

In response to requests, input was received from one specialty society and four academic medical centers while this policy was under review in 2009. Reviewers generally supported a limited role for the Boston KPro in selected patients. Some reviewers recommended use without first attempting a transplant in specific conditions that have a poor prognosis for corneal transplant; however, other input indicated that this is controversial. Some reviewers recommended use only in patients who have limited visual acuity in the contralateral eye. Overall, input indicated that the Boston KPro is reserved for cases in which no other alternative (ie, corneal transplantation) is available for treatment of corneal opacification.

Practice Guidelines and Position Statements

A 2013 Preferred Practice Parameter on ocular edema and opacification by the American Academy of Ophthalmology did not provide specific recommendations on the keratoprosthesis, but included this discussion of the technology\(^\text{20}\):

...Significant improvements in the design and postoperative management of the Boston type 1 keratoprosthesis has resulted in a steady rise in the number of these procedures performed both in the United States and abroad. Reduced
incidence of postoperative stromal necrosis and bacterial endophthalmitis due to the chronic use of protective soft contact lenses and topical antibiotics has resulted in improved retention and visual outcomes and has had a positive impact on surgeons’ perceptions of when to recommend keratoprosthesis. Once considered a procedure of last resort in patients with severe bilateral visual impairment, it is now being used for a variety of unilateral and bilateral indications, such as ocular trauma, herpetic keratitis, aniridia, Stevens-Johnson syndrome, and congenital corneal opacification. More recently, as corneal surgeons have gained a greater appreciation of the failure rate of repeat corneal transplantation, a role for a keratoprosthetic in cases of multiple graft failure has become clearer.

Patients with severe dry eye and autoimmune ocular surface diseases (particularly Stevens-Johnson syndrome and OMMP [ocular mucous membrane pemphigoid]) remain a difficult management group despite the other successes of the Boston type 1 keratoprosthetic. Epithelial defects, scleral necrosis, extrusion, and endophthalmitis are the principal concerns. This group of patients has had some success with a Boston type 2 keratoprosthetic designed to be used through the lid and the osteo-odonto-keratoprosthesis.

**Medicare National Coverage**

There is no Medicare national coverage policy. Medicare has established an Ambulatory Payment Classification 0293 for level V anterior segment eye procedures that includes CPT code 65770 (keratoprosthesis) and a HCPCS code for the prosthesis (C1818 - integrated keratoprosthesis OR L8609 - artificial cornea).²¹

**Regulatory Status**

A keratoprosthesis is a Class II FDA device intended to provide a transparent optical pathway through an opacified cornea in an eye that is not a reasonable candidate for a corneal transplant. Two permanent keratoprostheses have received 510(k) marketing clearance by FDA, the Boston KPro (Dohlman-Doane Keratoprosthesis) and the AlphaCor (Chirila keratoprosthesis). Both devices are indicated as permanent implantable keratoprostheses for eyes that are not corneal transplant candidates and are made of materials that have been proven to be biocompatible. According to the 510(k) summary, the AlphaCor keratoprosthesis was shown to
be substantially equivalent to the Dohlman-Doane Type I Keratoprosthesis. FDA product code: HQM.

References


<table>
<thead>
<tr>
<th>Date</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>01/97</td>
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</tr>
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</tr>
<tr>
<td>05/13/03</td>
<td>Replace Policy - Policy statement unchanged; expanded description and benefit application sections.</td>
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<td>Replace policy. Policy updated with literature review through December 2011; references 3 and 12-15 added; some references removed; policy statements</td>
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<td>Comments</td>
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<td>Update Coding Section – ICD-10 codes are now effective 10/01/2014.</td>
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<td>Replace policy. Policy updated with literature review through January 14, 2013; references 1, 7, and 15 added; policy statement unchanged.</td>
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<tr>
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<td>04/24/15</td>
<td>Annual Review. Policy updated with literature review through January 13, 2015. References 10-11 added; additional conditions that are likely to have poor outcomes from a corneal transplant were added to the medically necessary policy statement.</td>
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<tr>
<td>08/25/15</td>
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<td>06/01/16</td>
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</table>
Discrimination is Against the Law

LifeWise Health Plan of Washington complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability, or sex. LifeWise does not exclude people or treat them differently because of race, color, national origin, age, disability or sex.

LifeWise:

- Provides free aids and services to people with disabilities to communicate effectively with us, such as:
- Qualified sign language interpreters
- Written information in other formats (large print, audio, accessible electronic formats, other formats)
- Provides free language services to people whose primary language is not English, such as:
- Qualified interpreters
- Information written in other written languages

If you need these services, contact the Civil Rights Coordinator.

If you believe that LifeWise has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability or sex, you can file a grievance with:

Civil Rights Coordinator - Complaints and Appeals
PO Box 91102, Seattle, WA 98111
Toll free 855-332-6396, Fax 425-918-5592. TTY 800-842-5357
Email AppealsDepartmentInquiries@LifeWiseHealth.com

You can file a grievance in person or by mail, fax, or email. If you need help filing a grievance, the Civil Rights Coordinator is available to help you.

You can also file a civil rights complaint with the U.S. Department of Health and Human Services. Office for Civil Rights, electronically through the Office for Civil Rights Complaint Portal, available at https://ocrportal.hhs.gov/ocr/portal/lobby.jsf, or by mail or phone at:

U.S. Department of Health and Human Services
200 Independence Avenue SW. Room 509F, HHH Building
Washington, D.C. 20020, 1-800-368-1019, 800-537-7697 (TDD)

Getting Help in Other Languages

This Notice has Important Information. This notice may have important information about your application or coverage through LifeWise Health Plan of Washington. There may be key dates in this notice. You may need to take action by certain deadlines to keep your health coverage or help with costs. You have the right to get this information and help in your language at no cost. Call 800-592-6804 (TTY: 800-842-5357).

Français (French):


Deutsche (German):


Italiano (Italian):


中文 (Chinese):

本通知有重要的訊息。本通知可能有關於您透過 LifeWise Health Plan of Washington 提交的申請或保險的重要訊息。本通知內可能會有重要日期。您可能需要在截止日期前採取行動。以保留您的健康保險或費用補貼。您有權利免費以您的母語得到本訊息和幫助。請撥電話 800-592-6804 (TTY: 800-842-5357).

Oromo (Cushite):


Ilokano (Ilocano):

Daytoy a Pakdaar ket naglaon iti Napateg nga Impormasion. Daytoy a pakdaar mabalin nga adda ket naglaon iti napateg nga impormasion maipanggep ti aplikasyonyo wengo coverage baben Iti LifeWise Health Plan of Washington. Daytoy ket mabalin dagiti importante a petsa iti daytoy a pakdaar. Mabalin nga adda rumbeng nga aramidenyo nga addang sakbay dagiti partikular a naituding nga adday tapo mapagaldaanidyo ti coverage ti salun-ayto weno tulong kadagiti gastos. Adda karbenganyo a mangala iti daytoy nga impormasion ken tulong ti pukodo y a pagasanga nga awan ti bayadanyo. Tumawag iti numero nga 800-592-6804 (TTY: 800-842-5357).

Iloko (Ilocano):

Daytoy a Pakdaar ket naglaon iti Napateg nga Impormasion. Daytoy a pakdaar mabalin nga adda ket naglaon iti napateg nga impormasion maipanggep ti aplikasyonyo wengo coverage baben Iti LifeWise Health Plan of Washington. Daytoy ket mabalin dagiti importante a petsa iti daytoy a pakdaar. Mabalin nga adda rumbeng nga aramidenyo nga addang sakbay dagiti partikular a naituding nga adday tapo mapagaldaanidyo ti coverage ti salun-ayto weno tulong kadagiti gastos. Adda karbenganyo a mangala iti daytoy nga impormasion ken tulong ti pukodo y a pagasanga nga awan ti bayadanyo. Tumawag iti numero nga 800-592-6804 (TTY: 800-842-5357).
Este aviso contém informações importantes. Talvez seja necessário que você tome providências dentro de determinados prazos para manter sua cobertura de saúde ou ajuda de custos. Você tem o direito de obter esta informação e ajuda em seu idioma e sem custos. Ligue para 800-592-6804 (TTY: 800-842-5357) para obter ajuda.

Français (French):

Votre couverture d'assurance santé ou de aide de couverture peut être mise en danger s'il n'est pas maintenue ou si une certaine date est manquée. Vous avez le droit d'obtenir cette information et de l'aider dans votre langue et sans frais. Appelez 800-592-6804 (TTY: 800-842-5357) pour obtenir de l'aide.

Polskie (Polish):

Oświadczenie może zawierać ważne informacje. W spectrum informacji można również dostać się na język polski i bezpłatnie. Wybierz numer 800-592-6804 (TTY: 800-842-5357) i zatrzymaj jak pożądane.

Португез (Portuguese):


Română (Romanian):


Русский (Russian):

Настоящее уведомление содержит важную информацию. Это уведомление может содержать важную информацию о вашем заявлении или страховом покрытии через LifeWise Health Plan of Washington. В настоящем уведомлении могут быть указаны ключевые даты. Вам, возможно, потребуется принять меры к определенным предельным срокам для сохранения страхового покрытия или помощи с расходами. Вы имеете право на бесплатное получение этой информации и помощь на вашем языке. Звоните по телефону 800-592-6804 (TTY: 800-842-5357).

Español (Spanish):

Este aviso contiene información importante. Es posible que este aviso contenga información importante acerca de su solicitud o cobertura a través de LifeWise Health Plan of Washington. Es posible que haya fechas clave en este aviso. Es posible que deba tomar alguna medida antes de determinadas fechas para mantener su cobertura médica o ayuda con los costos. Usted tiene derecho a recibir esta información en su idioma sin costo alguno. Llamé al 800-592-6804 (TTY: 800-842-5357).

Tagalog (Tagalog):


ไทย (Thai):