JOHNS HOPKINS MEDICINE JOHNS HOPKINS HEALTHCARE	Johns Hopkins HealthCare LLC **Medical Policy** **Medical Policy**	*Policy Number*	CMS02.16
		Effective Date	08/02/2021
		Review Date	05/18/2021
	Subject **Treatment of the Cornea**	*Revision Date*	05/18/2021
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This document applies to the following Participating Organizations:

EHP | Johns Hopkins Advantage MD | Priority Partners | US Family Health Plan

Keywords: Keratoprosthesis (Artifical Cornea), Collagen Cross-Linking for Keratoconus, Cornea, Endothelial Keratoplasty, Phototherapeutic Keratectomy, Refractive Surgery

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I. ACTION

	New Policy	
x	Revising Policy Number	CMS02.16
	Superseding Policy Number	
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II. POLICY DISCLAIMER

Johns Hopkins HealthCare LLC (JHHC) provides a full spectrum of health care products and services for Employer Health Programs, Priority Partners, Advantage MD and US Family Health Plan. Each line of business possesses its own unique contract and guidelines which, for benefit and payment purposes, should be consulted to know what benefits are available for reimbursement.

Specific contract benefits, guidelines or policies supersede the information outlined in this policy.

III. POLICY

For Advantage MD refer to: Medicare Coverage Database

- National Coverage Determination (NCD) 80.12 Intraocular Lenses (IOLs)
- National Coverage Determination (NCD) 80.7 Refractive Keratoplasty
- National Coverage Determination (NCD) 80.1 Hydrophilic Contact Lens for Corneal Bandage

- Local Coverage Determination (LCD) L33793 Refractive Lenses
- Local Coverage Determination (LCD) L33737 Eye Prostheses

For Employer Health Programs (EHP) refer to:

- Plan Specific Summary Plan Descriptions (SPD's)

For Priority Partners (PPMCO) refer to: Code of Maryland Regulations

- No specific information located in COMAR 10.67.01 – 10.67.13 (Accessed 03/31/2021)

For US Family Health Plan (USFHP) refer to: Tricare Policy Manuals

- TRICARE Policy Manual 6010.60-M, April 1, 2015, Chapter 4, Section 21.1 Eye and Ocular Adnexa
- TRICARE Policy Manual 6010.60-M, April 1, 2015, Chapter 7, Section 6.2 Lenses (Intraocular or Contact) and Eye Glasses

IV. POLICY CRITERIA

A. When benefits are provided under the member's contract, JHHC considers the following services and supplies medically necessary when the specific criteria are met:

1. Keratoplasty (e.g. penetrating, endothelial, anterior lameliar) when InterQual® criteria are met.
2. Keratoprosthesis (artificial cornea) using the Boston K-Pro device:
   a. The cornea is severely opaque and vascularized, AND;
   b. Documented history of previous corneal transplants failure and there is a poor prognosis for further grafting (except in infants who do not require previous transplant attempts).
3. Corneal relaxing incision or wedge resection:
   a. Correction of astigmatism resulting from a previous eligible surgery (e.g., cataract removal, corneal transplant), AND;
   b. Inability to tolerate glasses or contact lenses.
4. Phototherapeutic Keratectomy:
   a. Treatment of anterior corneal stromal pathologies (e.g. corneal scars and opacities, elevated or irregular corneal lesions, anterior corneal dystrophies, recurrent corneal erosions) when conservative methods are ineffective or contraindicated.
   b. Unless specific benefits are provided under the member's contract, JHHC considers phototherapeutic keratectomy not medically necessary for treatment of refractive errors (myopia or hyperopia with or without astigmatism).
5. Corneal Cross-Linking:
   a. The procedure is done using a conventional (epithelium-off) technique combining riboflavin (vitamin B2) and ultraviolet A (UVA) radiation for treatment of corneal ectasia.
6. Intrastromal Corneal Ring Segments (e.g. Intacs® Prescription Inserts, Addition Technology, Sunnyvale, CA):
   a. Age 21 years of age or older, AND;
   b. Diagnosis of keratoconus with progressive deterioration of vision, AND;
   c. Inadequate vision correction with eyeglasses or contacts, AND;
   d. Corneal transplantation is the only alternative to placement of intrastromal corneal ring segments for treatment of keratoconus.

7. Corneal and Scleral Contact Lenses (e.g. soft, bandage, rigid, hybrid, toric, scleral shell (e.g.BostonSight®):
   a. Therapeutic contact lenses are utilized specifically for treatment of the following conditions:
      i. Keratoconus or other corneal disorders associated with irregular astigmatism (e.g., keratoglobus, corneal degeneration, ectasia due to refractive surgery, corneal scarring), OR;
      ii. Severe ocular surface diseases (e.g. corneal stem cell deficiency, neurotrophic (anesthetic) cornea, severe dry eyes, epidermal ocular disorders, corneal exposure, trauma), OR;
      iii. To support orbital tissue (covering a shrunken, sightless or damaged eye), OR;
      iv. Aphakia.
   b. Unless specific benefits are provided under the member's contract, JHHC considers corrective contact lenses provided solely for the correction of a refractive error (i.e. myopia, hyperopia, presbyopia, astigmatism) not medically necessary.
8. Intraocular lenses:
   a. Standard non-accommodating monofocal intraocular lenses are medically necessary when used to carry out the function of the human lens, lost as the result of intraocular surgery or ocular injury or congenital absence.
   b. Unless specific benefits are provided under the member's contract, JHHC considers placement of multifocal, accommodating, toric, or phakic intraocular lenses for correction of refractive errors (myopia, hyperopia, presbyopia, astigmatism) not medically necessary because these lenses is considered to be predominately for comfort and convenience, to eliminate the need for glasses or contact lenses after the surgery.
9. Amniotic Membrane Graft and Conjunctival Flap:
   a. Treatment of severe ocular surface disease resulting in persistent epithelial defects (e.g. bullous keratopathy; chemical or thermal burns or trauma; pterygium; recurrent corneal abrasions, erosions, ulcers, wounds; recurrent severe keratitis, limbal stem cell deficiency), AND;
   b. Documented failure of, intolerance to or contraindication to treatment using conventional medical management (lubricating and anti-inflammatory eye drops, gels, ointments; prescription medications, contact lenses, treatment of underlying cause, environmental and lifestyle modifications).

B. Limitations:
1. Unless specific benefits are provided under the member's contract, JHHC considers laser assisted or conventional refractive surgery (e.g. Radial Keratotomy (RK), Laser-in-situ Keratomileusis (LASIK), Epi-LASIK/LASEK, Photoastigmatic Keratectomy (PARK), Photorefractive Keratectomy (PRK), refractive lens exchange, phakic intraocular lenses) performed solely to correct refractive errors (myopia, hyperopia, presbyopia, astigmatism) not medically necessary.
2. Unless specific benefits are provided under the member's contract, JHHC considers procedures for treatment of cornea other than specified above experimental and investigational because they do not meet Technology Evaluation Criteria (TEC). Examples of experimental and investigational procedures:
   a. Orthokeratology
   b. Injection of micronized or particulated human amniotic membrane
   c. Electrothermal heat (e.i. TearCare system)
   d. Intranasal neurostimulation (i.e. TrueTear)
   e. Hexagonal keratotomy
   f. Scleral expansion surgery
   g. AlphaCor keratoprosthesis

V. DEFINITIONS

Amniotic Membrane Graft: Amniotic membrane harvested from the innermost layer of the placenta. The membrane is placed on the eye and secured to the cornea or episclera with sutures or fibrin glue or via a self-retaining mechanism (ring or contact lens). The application of an amniotic membrane to eyes with partial stem cell deficiency can improve ocular surface health and, in some cases, even restore a near-normal corneal epithelium (Nikpoor&Perez, 2019).

Aphakia: Aphakia is defined as an absence of crystalline lens; usually secondary to surgery, trauma (total dislocation of crystaline lens), or very rarely congenital (Friedman et al., 2021).

Astigmatism: Astigmatism is a condition in which the light rays are unequally focused, producing two or more focal points rather than a single point because the curvature of the cornea or, less commonly, the curvature of the lens varies in different meridians, or because the surface of the cornea is irregular (Friedman et al., 2021).

Conjunctival Flap: In a conjunctival flap procedure, a flap of conjunctiva is created to cover an unstable or painful corneal surface. Conjunctival flaps, partial or total, is an effective procedure for the treatment of challenging ocular surface disorders in patients with poor visual prognosis (Chang et al., 2019).

Cornea: The cornea represents the transparent anterior wall of the globe. A healthy cornea, together with the overlying tear film, is necessary to provide a proper anterior refractive surface and to protect the eye against infection and structural damage to the deeper components of the eye (Farjo et al., 2019).

Corneal Cross-Linking: The purpose of corneal cross-linking (CXL), also known as corneal collagen cross-linking, is to increase the biomechanical strength of collagen fibrils of the cornea in order to avoid the progression of corneal ectasia. CXL uses a combination of riboflavin (vitamin B2) eye drops, absorbed throughout the cornea stroma, with ultraviolet A (UVA) radiation to trigger a photochemical reaction that changes the cross-links between and within collagen fibers in the corneal stroma. This is believed to strengthen and increase the biomechanical stiffness of the corneal stroma. The procedure has variable effects on changing the shape of the cornea, but generally produces a relatively small amount of flattening. Conventional CXL (C-CXL) involves removing the epithelium, after which riboflavin drops are applied to the cornea and the UVA irradiation is performed (Hayes, 2020).

Corneal ectasia: Corneal ectasia is a group of disorders affecting the shape of the cornea including keratoconus, pellucid marginal corneal degeneration, post–refractive surgery corneal steepening, and keratoglobus (Sugar&Garcia-Zalisnak, 2019).

Corneal Wedge Resection: Corneal wedge resection is an effective treatment for high astigmatism after penetrating keratoplasty. The length of the incision is centered at the axis of the flatter meridian of the cornea and generally extended over a range of 60-90 degrees (Ezra, 2007).

Corneal Relaxing Incision: Corneal relaxing is done by partial thickness incisions made at the corneal periphery for the treatment of astigmatism. These incisions make the misshapen cornea more spherical, which improves visual clarity (Kozak, 2018).

Intraocular Lenses (IOLs): An intraocular lens implant is a synthetic, artificial lens placed inside the eye that replaces the focusing power of a natural lens that is surgically removed or absent. Most commonly used types of IOLs:

- Monofocal IOLs: Most often used IOLs. Fixed focus lenses, typically providing distance vision; glasses are still required for near activities. Monovision is a good option for many patients, with one eye set for distance and the other eye set for midrange or near vision. The eye set for distance is typically the dominant eye.

- Accommodative IOLs: Allow for correction of presbyopia. Designed to change position or shape in the eye with accommodative effort. It can allow for good distance and improved near vision without eyeglasses or contact lenses.
- Multifocal IOLs: Provide for near to distance focal lengths within the optic zones of the lens. It allows an improved ability to see well over a continuous range of distance without glasses.
- Piggyback IOLs: A second lens implant can be inserted in the ciliary sulcus to correct for low levels of a postoperative refractive error after cataract or refractive lens exchange surgery or to treat negative dysphotopsia (unwanted shadow) after otherwise routine cataract/IOL surgery.
- Phakic IOLs: IOLs implanted when the natural lens is present. These lenses are implanted in the anterior or posterior chamber of the eye to correct refractive errors. They preserve the natural crystalline lens to maintain accommodation in patients who are not presbyopic (Friedman et al., 2021; Taneri et al., 2019).

Intrastromal Corneal Ring Segments (ICRSs): ICRSs are thin, clear, semicircular, plastic implants placed beneath the cornea that are designed to reduce or eliminate myopia and astigmatism by stretching and flattening the cornea in patients with keratoconus. The surgery is minimally invasive. The implanted inserts do not directly alter the passage of light through the pupil. Instead, the inserts correct vision indirectly by stretching and flattening the cornea. Intacs® (Addition Technology INC., Sunnyvale, CA) is one type of the ICRSs (Hayes, 2020).

Irregular Astigmatism: Irregular astigmatism occurs when the cornea or lens are so irregular that they do not form well-defined meridians (Diniz et al., 2019).

Keratoconus: Keratoconus is a disorder characterized by progressive corneal steepening, most typically inferior to the center of the cornea, with eventual corneal thinning, induced myopia, and both regular and irregular astigmatism (Sugar&Garcia-Zalisnak, 2019).

Keratoplasty: Keratoplasty is a procedure for replacement of corneal tissues with a healthy donor graft. Most common techniques include the following procedures:

- Penetrating Keratoplasty (PK): PK involves full-thickness replacement of corneal tissue with a healthy donor graft. PK may be used to provide tectonic support (as in corneal thinning or perforation) and to improve visual outcomes (as in the replacement of an opaque or irregular cornea). The rate of success of PK is excellent, but the long-term risk of graft rejection increases significantly with active or recurrent infection, inflammation, corneal neovascularization, previous graft rejection, and each subsequent penetrating graft.
- Endothelial Keratoplasty (EK): EK, also referred as posterior lamellar keratoplasty, is a procedure in which a partial-thickness graft of the donor tissue is used to replace the inner layer of the cornea (diseased endothelium). EK is indicated for patients with any type of endothelial dysfunction such as endothelial dystrophies, bullous keratopathy, endothelial failure post PK, and trauma.
- Anterior Lamellar Keratoplasty (ALK): In ALK, the outer layer of the cornea is replaced with transplanted tissue. The graft does not include corneal endothelium, thus avoiding endothelial rejection or failure. Indications for ALK mainly include anterior corneal pathology in which the posterior cornea is unaffected, such as keratoconus, anterior corneal scars, and corneal dystrophies limited to the stroma (Gibbons et al., 2019).

Keratoprosthesis: Keratoprosthesis is a surgical procedure where a diseased cornea is replaced with an artificial cornea. The Boston Keratoprosthesis (K-Pro) (formerly known as the Dohlman–Doane Kpro) is the most commonly used keratoprosthesis in the world. The keratoprosthesis is made of clear polymethyl methacrylate (PMMA) plastic and has two pieces that take the

shape of a collar button. The device is inserted into a corneal graft, which is then transplanted into the recipient's cloudy cornea (Gibbons et al., 2019).

Phototherapeutic Keratectomy: Phototherapeutic keratectomy is an excimer laser-based surgical procedure widely performed by corneal surgeons for treating anterior corneal epithelial and stromal pathologies. Phototherapeutic keratectomy helps by ablating the corneal stroma, thereby improving corneal clarity and smoothening the surface or by better enabling corneal epithelium to adhere to the underlying tissue. It can be considered as a bridge between medical and surgical management of different corneal diseases. This procedure can also be used to correct refractive errors (Nagpal et al., 2020).

Presbyopia: Presbyopia is the loss of accommodation due to hardening of the crystalline lens with age (Diniz et al., 2019).

Refractive Error: Refractive errors, or ametropias, are anomalies of the optical state of the eye that cause imperfect focus on the retina, leading to a poor quality of the final image. Refractive errors include myopia (nearsightedness), hyperopia (farsightedness), and astigmatism (imperfection in the curvature of the cornea or lens (Diniz et al., 2019).

Refractive surgery: Refractive surgery is the surgical correction of refractive errors of the eye such as myopia, hyperopia, astigmatism, and presbyopia (Taneri et al., 2019).

Therapeutic Contact Lenses: Therapeutic contact lenses are special contact lenses used for treatment of a corneal or anterior eye disease. Although therapeutic contact lenses can be used to correct refractive error, and in some cases enhance drug delivery to the cornea, they are used primarily for treatment purposes, mechanical eye protection, and as an aid to healing. Based on covered surface area, they can be referred to as corneal, cornea-scleral, or scleral. The following are the most frequently used therapeutic lenses:

- Special use soft lenses:
  - Bandage lenses for healing after ocular surgery or treatment of ocular injury;
  - Specially designed lenses for keratoconus or irregular astigmatism, including toric lenses;
- Rigid gas-permeable lenses used for patients with keratoconus and irregular corneal topography;
- Hybrid lenses have a rigid central portion fused to a peripheral soft skirt. These lenses can be used for treatment of keratoconus, post-surgical eyes, and other irregular astigmatism cases;
- Scleral lenses are larger diameter (up to 24 mm) lenses prescribed to completely vault over the cornea. These lenses are designed to extend beyond the cornea to rest on the conjunctiva overlying the sclera and are comfortable to wear. Because they completely clear the corneal surface, they retain a reservoir of tears between the lens and the cornea. Scleral lenses are helpful for correcting vision in cases of irregular or distorted corneas (e.g. keratoconus, post-corneal transplant, post-refractive surgery or corneal scarring, and ocular surface disease (Lim&Lim, 2020).

VI. BACKGROUND

The cornea is a complex structure which, as well as having a protective role, is responsible for about three-quarters of the optical power of the eye. The normal cornea is free of blood vessels. Nutrients are supplied and metabolic products removed via the aqueous humor posteriorly and the tears anteriorly. The cornea is the most densely innervated tissue in the body and conditions such as abrasion and bullous keratopathy are associated with marked pain, photophobia and reflex lacrimation. Management options for corneal pathologies range from excimer laser–based ablation procedures to partial- and full-thickness grafts, depending on the depth of corneal involvement (Salmon, 2019).

Corneal ectasia (thinning) disorders represent a range of disorders involving either primary disease conditions, such as keratoconus and pellucid marginal corneal degeneration, or secondary iatrogenic conditions, such as corneal thinning occurring

after laser in situ keratomileusis (LASIK) refractive surgery. Corneal thinning is a disease that occurs when the normally round dome-shaped cornea progressively thins, causing a cone-like bulge or forward protrusion in response to the normal pressure of the eye. The thinning occurs primarily in the stroma layers and is believed to be a breakdown in the collagen structure and organization. This bulging can lead to irregular astigmatism or the shape of the cornea. Because the anterior part of the cornea is responsible for most of the focusing of the light on the retina, this can then result in loss of visual acuity (Pron et al., 2011).

Corneal ectasias cause progressive and often unpredictable decreases in best-corrected vision. Early in the course of the disease, this effect on vision may be adequately corrected with eyeglasses. As the disease progresses, therapeutic contact lenses (to address the abnormal curvature of the cornea) are often required or corneal cross-linking can be used to slow or halt the progression of keratoconus. When these methods are no longer effective, surgical procedures such as intracorneal ring segment implantation or full or partial thickness corneal transplantation are required to maintain visual function (Reynolds et al., 2020).

Corneal transplantation has changed dramatically since Eduard Zirm performed the first one on a human just over a century ago. As an avascular tissue, corneal transplantation is associated with one of the lowest rejection rates of all human organs. In recent years, the evolution of techniques and technologies has substantially improved outcomes and enabled a shift toward replacement of only the diseased layers. The indications for transplantation are numerous and include optical, tectonic and reconstructive, therapeutic and, in rare cases, cosmetic needs. Patients who have a significantly reduced best-corrected visual acuity (BCVA) from a corneal pathology usually benefit from a graft. Occasionally, transplantation is required to save an altered corneal structure from perforation or thinning. Pain management for bullous keratopathy and non-healing ulcers can also be a primary therapeutic indication (Steele, 2019).

Ocular surface diseases (OSDs) is an umbrella term encompassing a wide variety of conditions affecting the ocular surface. These conditions include dry eye disease, blepharitis, meibomian gland dysfunction, and immunologic disorders. Current conservative management options for OSD include artificial tears, topical antihistamines, steroids and immunologics and therapeutic contact lens (TCL) usage. Indications of TCL include pain relief, enhancing corneal healing, corneal sealing, corneal protection, and drug delivery. For painful corneal diseases such as bullous keratopathy, epidermolysis bullosa, and epithelial abrasions/erosions, bandage contact lenses (BCLs) provide symptomatic relief. BCLs are used post-keratoplasty, post-trabeculectomy, and post–amniotic membrane transplantation to enhance healing. BCLs, with or without glue adhesives, are used to seal small corneal perforations and sometimes also used as bridging treatment before penetrating keratoplasty in corneal perforations. In patients with eyelid conditions such as trichiasis, ptosis, and tarsal scarring, BCLs are also effective in forming a mechanical barrier to protect the cornea (Lim&Lim, 2020).

VII. CODING DISCLAIMER

Note: The following CPT/HCPCS codes are included below for informational purposes and may not be all inclusive. Inclusion or exclusion of a CPT/HCPCS code(s) below does not signify or imply that the service described by the code is a covered or non-covered health service. Benefit coverage for health services is determined by the member's specific benefit plan document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee of payment. Other policies and coverage determination guidelines may apply.

Note: All inpatient admissions require preauthorization.

Compliance with the provision in this policy may be monitored and addressed through post payment data analysis and/or medical review audits			
Employer Health Programs (EHP) refer to specific Summary Plan Description (SPD). If there is no criteria in the SPD, apply the Medical Policy criteria.	Priority Partners (PPMCO) refer to COMAR guidelines and then apply the Medical Policy criteria.	US Family Health Plan (USFHP), TRICARE Medical Policy supersedes JHHC Medical Policy. If there is no Policy in TRICARE, apply the Medical Policy Criteria.	Advantage MD, LCD and NCD Medical Policy supersedes JHHC Medical Policy. If there is no LCD or NCD, apply the Medical Policy Criteria.

VIII. CODING INFORMATION

CPT® CODES ARE FOR INFORMATIONAL PURPOSES

CPT® CODES	DESCRIPTION
0290T	Corneal incisions in the recipient cornea created using a laser, in preparation for penetrating or lamellar keratoplasty (List separately in addition to code for primary procedure)
0402T	Collagen cross-linking of cornea (including removal of the corneal epithelium and intraoperative pachymetry when performed)
65710	Keratoplasty (corneal transplant); anterior lamellar
65730	Keratoplasty (corneal transplant); penetrating (except in aphakia or pseudoaphakia)
65750	Keratoplasty (corneal transplant); penetrating (in aphakia)
65755	Keratoplasty (corneal transplant); penetrating (in pseudophakia)
65756	Keratoplasty (Corneal Transplant); endothelial
65757	Backbench preparation of corneal endothelial allograft prior to transplantation
65760	Keratomileusis
65765	Keratophakia
65767	Epikeratoplasty
65770	Keratoprosthesis
65771	Radial keratotomy
65772	Corneal relaxing incision for correction of surgically induced astigmatism
65775	Corneal wedge resection for correction of surgically induced astigmatism

65778	Placement of amniotic membrane on the ocular surface; without sutures
65779	Placement of amniotic membrane on the ocular surface; single layer, sutured
65780	Ocular surface reconstruction; amniotic membrane transplantation, multiple layers
65782	Ocular surface reconstruction; limbal conjunctival autograft (includes obtaining graft)
65785	Implantation of intrastromal corneal ring segments
66840	Removal of lens material;aspiration technique,1 or more
66940	Removal of lens material; extracapsular (other than 66840, 66850, 66852)
66985	Insertion of intraocular lens prosthesis (secondary implant), not associated with concurrent cataract removal
68360	Conjunctival flap; bridge or partial (separate procedure)
68362	Conjunctival flap; total (such as Gunderson thin flap or purse string flap)

HCPCS CODES ARE FOR INFORMATIONAL PURPOSES	
HCPCS CODES	**DESCRIPTION**
C1780	Lens, Intraocular (new technology)
C1818	Integrated keratoprosthesis
L8609	Artificial cornea
Q1004	New technology, intraocular lens, category 4 as defined in Federal Register notice
Q1005	New technology, intraocular lens, category 5 as defined in Federal Register notice
S0596	Phakic intraocular lens for correction of refractive error
S0800	Laser in situ keratomileusis (LASIK)
S0810	Photorefractive keratectomy (PRK)
S0812	Phototherapeutic keratectomy (PTK)
V2630	Anterior chamber intraocular lens
V2631	Iris supported intraocular lens
V2632	Posterior Chamber intraocular lens
V2785	Processing, preserving, and transporting corneal tissue
V2787	Astigmatism correcting function of introcular lens
V2788	Presbyopia correcting function of intraocular lens

IX. REFERENCE STATEMENT

Analyses of the scientific and clinical references cited below were conducted and utilized by the Johns Hopkins HealthCare LLC (JHHC) Medical Policy Team during the development and implementation of this medical policy. Per NCQA standards, the Medical Policy Team will continue to monitor and review any newly published clinical evidence and adjust the references below accordingly if deemed necessary.

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XI. APPROVALS

Historical Effective Dates: 03/03/2017; 02/19/2019; 08/02/2021