



Penetrating keratoplasty using collagen crosslinked donor tissue: A case report

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ABSTRACT

Purpose: In patients with corneal melt, pretreatment crosslinking (CXL) of donor tissue prior to placement of Boston keratoprosthesis (K-Pro I) decreases graft failure. We report a case of corneal sparing in a phthisical eye following penetrating keratoplasty (PKP) with pretreatment CXL of the donor cornea.

Observations: A 69-year-old female with a history of familial aniridia and bilateral K-Pro I placement. Her clinical course was complicated by recurrent corneal melt and hypotony in the left eye, resulting in extraction of the K-Pro I and successive PKP with pretreatment CXL of the donor cornea. She subsequently developed phthisis of the globe with notable retention of corneal structure. At 8 years, she maintains corneal contour without recurrence of keratolysis or extension of phthisis.

Conclusions and importance: This is the first reported case of corneal sparing in a phthisical eye with a history of PKP, suggesting a protective role of pretreatment CXL of donor tissue against keratolysis and phthisis.

1. Introduction

Phthisis bulbi is a rare condition that results in disorganized and shrunken ocular structures. The pathogenesis involves disorganization, calcification, and ossification of tissue secondary to prior insult.¹ The pathologic causes are broad and include trauma, hypotony, and inflammation.¹ Both anterior and posterior structures are frequently involved, including a thickened and disorganized corneal structure.¹

There are reports on the use of crosslinked carrier tissue in patients with severe keratolysis prior to the placement of Boston keratoprosthesis.^{2,3} To our knowledge, no study has reported the potential protective role of corneal crosslinking in maintaining corneal architecture in the context of phthisis. We describe a case of phthisis with corneal sparing in the setting of pretreatment collagen crosslinking of the donor tissue during a penetrating keratoplasty.

2. Case report

A 69-year-old female with familial aniridia complicated by corneal blindness initially underwent traditional bilateral Boston keratoprosthesis (K-Pro) placement, first in the right eye in June 2008, followed by a 9.5 mm K-Pro with a titanium backplate in the left eye in October 2009. The post-operative period in the left eye was complicated by

persistent post-operative sterile vitritis that cleared over the course of one year.

Two years later, the left eye developed Hand Motion vision. Exam was notable for a hypotonous eye with keratolysis under the front plate as well as Seidel positive leakage of the aqueous humor. The patient was brought to the operating room for replacement of the K-Pro with a new corneal donor carrier. During the procedure, the retroprosthetic membrane was noted to be adhered to the K-Pro backplate and the intraocular lens. Both were excised and removed from the eye. An anterior vitrectomy was performed and a new aphakic K-Pro was inserted.

Three months later, the patient presented following a gush of fluid from the left eye. Hypotony was again noted in the left eye, which had an irregular globe contour. SD-OCT showed choroidal thickening and retinal elevation.

Given that the patient had recurrent keratolysis in the left eye, the surgical team discussed collagen crosslinking the donor tissue prior to implantation to strengthen the tissue and help prevent keratolysis as part of an investigator-initiated study (NCT02863809). The next day the patient was brought to the OR for combined K-Pro replacement using the crosslinked corneal donor tissue and pars plana vitrectomy with membrane peel and silicone oil placement. Within the operating room prior to the surgery, the donor corneal tissue was placed on an anterior chamber maintainer and crosslinked using the Dresden Protocol.²

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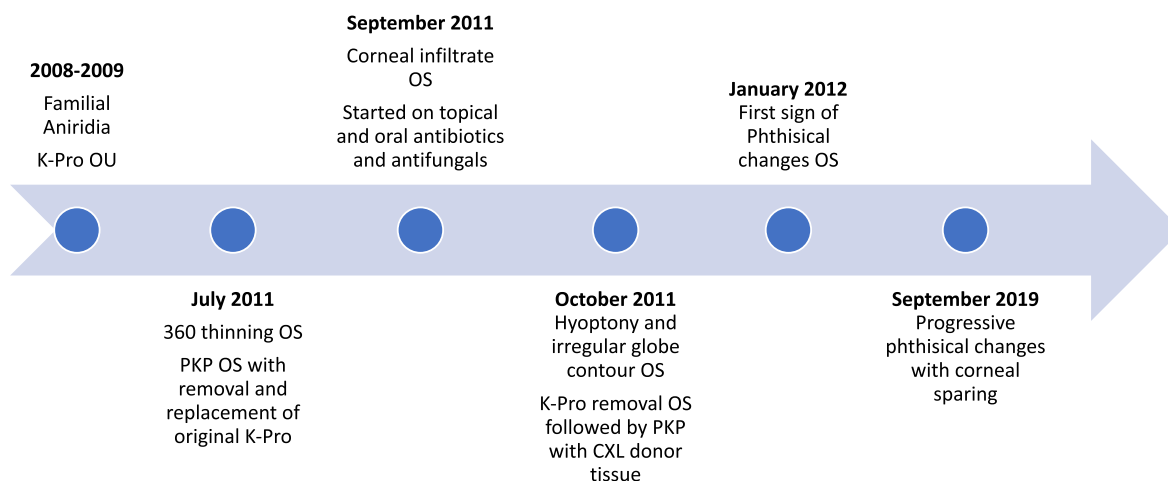


Fig. 1. Timeline of clinical events over 10 years of follow up.

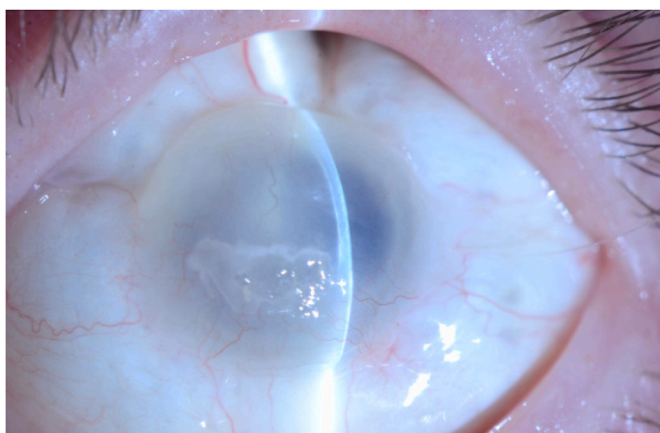


Fig. 2. Left eye 8 years after undergoing penetrating keratoplasty using crosslinked donor tissue. Normal corneal contour is surrounded by phthisical changes of the sclera with superior scleral buckling.

During surgery, the patient was found to have a large funnel retinal detachment that was not amenable to surgical repair. Given the limited visual potential for the eye, the decision was made to place the crosslinked corneal graft without the K-Pro.

The vision during the immediate post-operative period was Light Perception and the eye remained soft, with an intraocular pressure of 4 mmHg. The slit lamp examination was notable for a diffusely thickened penetrating keratoplasty graft with Descemet membrane folds. The graft was Seidel negative with silicone oil observed against the posterior surface of the graft. In January 2012, phthisical changes were first noted in the conjunctiva of the left eye. However, there were no phthisical changes in the cornea (Fig. 1).

The patient was followed regularly for the next eight years and the left eye became progressively more phthisical. Vision remained Light Perception only. There has been no recurrence of keratolysis. During her latest follow up in September 2019, the sclera and peripheral cornea appeared disfigured and shrunken while the central corneal graft retained its shape and contour (Fig. 2).

3. Discussion

We present a unique case of a patient with familial aniridia requiring bilateral Boston keratoprosthesis I (K-Pro I) placement that underwent penetrating keratoplasty with crosslinked corneal donor tissue due to recurrent keratolysis. The patient later developed phthisis with sparing

of the crosslinked corneal donor tissue. To our knowledge, this is the first reported case in the literature of a crosslinked corneal graft maintaining its structure in a phthisical eye.

Keratolysis is a common complication of K-Pro I, with a cited incidence of approximately 14% of eyes during the first 2 years.²⁻⁷ However, in certain high-risk populations, such as eyes with persistent inflammation, the incidence can be even higher.²⁻⁴ These eyes face many post-operative challenges, including the development of phthisis.

Phthisis bulbi involves pathologic changes to the entire eye. Early corneal involvement includes edema due to endothelial cell damage and degenerative pannus.¹⁻³ Later progression can lead to stromal scarring, vascularization, retro-corneal membranes, and dystrophic calcification.¹⁻³ The remaining cornea is thickened and opaque.¹⁻³

Corneal crosslinking uses riboflavin and UV-A light to arrest progression of corneal ectasias through the modification of collagen fibers by several proposed mechanisms.^{5,6} One such mechanism involves the induction of covalent bonds between collagen fibers.^{6,7} This results in corneal stiffening and strengthens the corneal stroma against keratolysis. Another proposed mechanism involves the observed increase in collagen fiber diameter following crosslinking.^{6,7} Together with the strengthened inter-fiber bonds, this increase in diameter serves to strengthen the cornea. However, the physiologic benefits of crosslinking go beyond the mechanical impact on corneal strength. Crosslinking increases resistance to enzymatic degradation, particularly collagenases, and improves resistance to heat degradation.^{5,6} While crosslinking has traditionally been used for patients with keratoconus, its unique properties serve a protective function against both keratolysis and disorganization, which occurs in phthisical eyes or eyes that have undergone chemical injuries.²⁻⁴

Our patient developed phthisis in the setting of recurrent hypotony and insult to the left eye. However, the crosslinked corneal donor tissue retained its normal structure. The literature supports such uses of collagen crosslinking in K-Pro patients in order to prevent keratolysis. In one trial there were no occurrences of keratolysis in the 11 patients treated with crosslinked tissue.³

Crosslinked corneal donor tissue retained normal architecture, thickness, and relative clarity despite the presence of phthisis. Beyond the advantages of improved tensile strength and improved resistance to degradation by collagenases, pretreated tissue may serve a protective role against the later development of phthisis. Our patient maintained corneal structure and contour in an otherwise phthisical eye, suggesting a possible protective role of crosslinking against the disorganized degeneration of phthisical eyes.

Patient consent

Consent to publish case details was obtained from the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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