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The Usage of a Conjunctival Flap to Improve Retention of Boston Type 1 Keratoprosthesis in Severe Ocular Surface Disease

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Abstract

Purpose: The Boston keratoprotheses type 1 devices (KPro) are utilized in cases unfavorable to penetrating keratoplasty. The prognosis remains guarded in cases of ocular surface disease due to risk of tissue necrosis. We describe a novel surgical approach using a conjunctival flap with a delayed opening to improve retention.

Methods: In three patients with advanced cicatrizing conjunctivitis, a Type 1 keratoprosthesis was stabilized using a full tarsal conjunctival flap. Three months postoperatively, an opening was created in the flap overlying the optical portion of the device.

Results: All patients had no device related complications over a mean follow-up period of 17.7 months (range 15–21 months) and vision remained excellent at better than 20/200 for all patients.

Conclusions: Utilization of a tarsal flap either primarily as part of a two stage modified technique or secondarily in cases of tissue necrosis and impending device extrusion might maximize retention of the type 1 KPro.

Keywords

Artificial cornea; complications; keratoprosthesis; surgical technique

Over the past decade the Boston type 1 keratoprosthesis (KPro) has emerged as a viable vision rehabilitation procedure for eyes with corneal blindness. Since the United States Food and Drug Administration (FDA) granted marketing clearance in 1992, the device has undergone multiple design revisions to maximize the outcomes.¹ As of August 2013, 8140 KPros have been implanted worldwide: 5406 domestically and 2734 abroad (L. Gelfand, Business Manager Boston Keratoprosthesis, Massachusetts Eye and Ear Infirmary, written communication, September 28, 2013).

While excellent outcomes have been reported in multiple previous studies, the retention rate is known to be poor with the type 1 device in patients with ocular surface diseases,

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DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

particularly from underlying autoimmune diseases. Indeed, the most recent multicenter study from the United States reported that the presence of autoimmune ocular surface disease is associated with the highest risk of device removal due to tissue necrosis and corneal melt (hazard ratio, 11.94; 95% confidence interval, 3.31–43.11).² Therefore, the type 2 KPro is the recommended procedure in these cases. An anterior extension of the type 2 device allows implantation through surgically closed lids, making it more suitable for patients with cicatrizing ocular surface disease, loss of conjunctival fornices, or keratinization from severe dry eye. However, implantation of this device is technically challenging requiring extensive dissection of the lids and a full tarsorrhaphy. Furthermore, intraocular pressure measurement becomes impossible after the procedure, increasing the likelihood of vision loss from glaucoma. When to use the type 2 KPro over the type 1 largely remains a clinical judgment. Nevertheless, due to its challenging surgical technique and unique complication profile, the type 2 device implantation is an uncommonly performed procedure aside from at a few select institutions.^{3,4} The osteo-odonto-keratoprosthesis (OOKP) and tibial keratoprotheses are other alternatives for patients with severe cicatrizing disease and are more commonly performed outside of the United States. They have better longer-term retention rates than the type 1 or 2 device, but require multiple lengthy and complex surgeries. Furthermore, visual field is limited with OOKP, the cosmetic appearance can often be unacceptable to patients, and more importantly measurement of intraocular pressure becomes impossible.⁵ For these reasons, this device has not been adopted in the United States.

The type 1 KPro remains the most commonly utilized device in the United States because of its surgical simplicity and well-studied side-effect profile. The device requires a donut-shaped donor corneal button that is sutured onto the host cornea as a carrier for the device. In patients with active immune disease, this donor button can potentially undergo a sterile melt that can subsequently result in extrusion of the device, infection, and loss of the eye. To date, there have been limited reports of novel surgical techniques for the implantation of this device in patients with cicatrizing disease who are prone to sterile keratolysis. One case report has mentioned the use of a conjunctival flap to cover a small area of sterile keratolysis.⁶ Another series has described the use of an oral buccal mucosal graft in addition to a corneal lamellar graft to cover areas of melt.⁷ Both of these described techniques were used as rescue procedures to cover an isolated region of device exposure. Prior reports have also described conjunctival flaps for keratoprotheses at high risk of melt, including the use of a delayed opening flap.⁸ Girard described a delayed-opening conjunctival flap technique for a keratoprosthesis, in which there was no corneal bed and conjunctiva was used for securing the device onto the eye.⁹ However, all of these techniques have largely employed tenon and bulbar conjunctiva in conjunction with mucous membrane grafts, and many of them have been described for other types of keratoprotheses. To date, no specific technique has described the usage of a tarsal-conjunctival flap in shortened fornices in the Boston type 1 KPro. Here, we report a series of three patients in which we performed a tarsal-conjunctival flap to cover the type 1 device, followed by a delayed window opening 3 months later, either as a two-step procedure in patients who were predicted to be at risk of poor retention or to salvage a device with significant tissue melting and impending or frank device extrusion.

METHODS

This retrospective study was reviewed and approved by the Johns Hopkins University Institutional Review Board in accordance with the Declaration of Helsinki and was Health Insurance Portability and Accountability Act compliant. All patients provided informed, written consent to undergo surgery as described. For each patient we collected the following data: medical and ophthalmic history, Snellen visual acuity, clinical examination data, and clinical photographs.

RESULTS

Case 1

A 36-year-old man with a history of Stevens–Johnson syndrome presented for vision rehabilitation. His right eye had been enucleated due to complications following multiple corneal transplantations. The left eye had had three penetrating keratoplasties prior to presentation, the last of which was complicated by a hypopyon ulcer and melt a few days following the surgery. He was aphakic and had had a vitrectomy by history.

The patient's visual acuity was light perception. Clinical examination by slit-lamp revealed severely foreshortened fornices with symblepharon formation and the cornea had complete opacity and pannus formation. As a first step, buccal mucous membrane grafts were used to reconstruct upper and lower cul-de-sacs. The patient was placed on oral immunomodulation using mycophenolate 1 g TID. Six months later, an aphakic Boston keratoprosthesis was implanted in the left eye without complications. However, significant thinning of the donor as well as recipient corneas with overlying epithelial defect was noted at 1 week postoperatively. Despite aggressive use of topical lubricating agents and antibiotics, melting progressed over an additional week, with severe tissue necrosis and impending device extrusion. The patient was again taken to the operating room.

After debridement and cultures, the device was secured to the recipient scleral bed using several long 9–0 nylon sutures. A thick conjunctival flap with sub-Tenon's dissection was carried out inferiorly and superiorly. Dissection into the superior and inferior fornices allowed mobilization of the previously placed mucosal grafts and conjunctiva. These tissues were advanced over the device, such that the superior edge of the upper tarsus and inferior edge of the lower tarsus moved onto the bulbar surface. The conjunctival flap was closed with 6–0 Vicryl sutures.

Three months later, the patient was comfortable and vision was light perception. The conjunctival flap was stable with no signs of tissue necrosis (Figure 1). The patient was taken to the operating room. A 3 mm disposable dermal biopsy punch was used to trephine the conjunctiva overlying the optical portion of the device with no complications. The patient was placed on topical antibiotics, aqueous suppressing agents, and lubricants and kept on oral immunomodulation using mycophenolate. He successfully underwent diode laser treatment at 16 months to lower the intraocular pressure. At the last follow-up, 21 months postoperatively, the vision measured 20/50 with no device-related complications.

Case 2

An 80-year-old woman presented for evaluation of neurotrophic corneal ulcer in the right eye. She had a history of central retinal artery occlusion in the left eye with poor vision. She was pseudophakic in the right eye by history. On examination, the vision was counting fingers in the right eye. The cornea had a large epithelial defect, moderate thinning and almost complete stromal opacity; she also had severe dry eye with trichiatic eyelashes and lid margin scarring.

Initially, upper and lower lacrimal canaliculi were cauterized, cultures were taken, and the patient was placed on topical antibiotics and aggressive lubrication. No infectious etiology was identified. Despite maximum medical treatment, the corneal necrosis and thinning progressed and required first an amniotic membrane patch applied surgically along with a lateral tarsorrhaphy. Subsequently, due to gradual progression of stromal thinning and impending perforation, a 7 mm anterior lamellar keratoplasty had to be carried out along with a permanent medial and lateral tarsorrhaphies. Seven months after the lamellar graft, the tissue necrosis returned, despite aggressive topical treatment. The patient was placed on systemic immunosuppression using mycophenolate for clinical features of Sjögren syndrome. Although serology was positive for antinuclear antibodies, she did not consent to undergo a minor salivary gland biopsy for definitive diagnosis.

Four months into the systemic immunomodulating therapy, a decision was made to proceed with a pseudophakic Boston type 1 keratoprosthesis, in order to improve the vision. The surgery was combined with a glaucoma tube shunt insertion. Shortly after the uncomplicated surgery, the patient developed a rapidly progressing thick retroprosthetic membrane that could not be addressed with YAG laser. Her eye pressures, as measured by digital palpation, were also noted to be high and could not be controlled with maximum medical therapy. Therefore, she underwent further surgery with removal of the intraocular lens implant, repeat keratoprosthesis, and pars plana vitrectomy, followed by diode laser at 1 month postoperatively.

After 10 months, the patient's trichiasis and lower lid entropion had to be repaired as she frequently developed epithelial defects. Lid margin rotation and buccal mucous membrane grafts were applied to correct lower eyelid cicatricial entropion. The trichiatic eyelashes were addressed with electrolysis at the same session. One month following the lid surgeries, severe corneal stromal thinning with epithelial defect was appreciated, again adjacent to the optic of the device.

The patient underwent complete tarsal-conjunctival flap to rescue the KPro. A crescent knife was used to dissect the superior bulbar conjunctiva and forniceal tissue as well as palpebral conjunctiva to the superior edge of the tarsus. This was then advanced over the cornea and attached to the sclera inferiorly (Figure 2). The flap remained stable for 3 months, at which point a window opening was produced using a 3 mm trephine, followed by medial and lateral tarsorrhaphies to stabilize the ocular surface. The tarsorrhaphies were opened 2 months later, and after 5 months, she underwent successful right upper eyelid ptosis repair. At 20 months after flap construction, vision measured 20/70 and there were no further complications.

Case 3

The patient, a 65-year-old Middle-Eastern male, presented with poor vision in the setting of long-standing, bilateral cicatrizing conjunctivitis and history of herpetic keratouveitis of both eyes. He was diagnosed with atopic keratoconjunctivitis. By history, he was pseudophakic in both eyes and had undergone pars plana vitrectomy for proliferative diabetic vitreoretinopathy.

On examination, visual acuity was hand motions in the right eye and light perception in the left eye. Slit-lamp examination demonstrated severe burnt out cicatrizing conjunctivitis with fornix foreshortening, subepithelial fibrosis, fully scarred and neovascularized cornea without an overlying epithelial defect in the right eye. Complete ankyloblepharon was present in the left eye. The eye pressure in the right eye was 3 mmHg with applanation and the eye was soft to palpation. Left eye pressure was normal by digital palpation. A B-scan sonogram revealed an attached retina in both eyes.

Right upper eyelid entropion was repaired with buccal mucous membrane grafting and a diagnostic conjunctival biopsy was performed in the right eye. The biopsy showed nonspecific, chronic changes with no significant active inflammation. The patient was placed on chronic suppressive oral antiviral. A combined surgery was performed with explantation of a subluxed intraocular lens, placement of a Boston type 1 KPro, pars plana vitrectomy, and silicone oil injection in the right eye. An amniotic membrane graft was placed to cover the device at the completion of the surgery. The amniotic membrane patch had to be repeated at 3 months and a tarsorrhaphy was added to stabilize the ocular surface as the patient continued getting epithelial defects. He could not keep a bandage contact lens due to forniceal conjunctival scarring.

At 10 months postoperatively, the patient was diagnosed with stromal necrosis adjacent to the device by his local referring physician and underwent another amniotic membrane patch that dissolved shortly after the grafting (Figure 3). The patient was asked to return to our institute for further management. Severe tissue necrosis with impending device extrusion and thick retroprosthetic membrane were noted at the slit-lamp, demonstrating rapid progression of the melt. The patient was taken to the operating room and a repeat type 1 KPro was performed with further silicone oil injection after removal of thick retroprosthetic and ciliary body membranes. At the completion of the case, a dissection was carried superiorly posterior to Tenon's capsule for 360° to mobilize the conjunctiva. The superior forniceal flap was advanced to obtain complete device coverage and sutured to sclera inferiorly with 8-0 Vicryl sutures, which were also utilized to close remaining conjunctiva. In this case, the scarring and shortening of the superior cul-de-sac necessitated the eversion of the superior aspect of the upper lid tarsus to allow enough advancement of tissue to complete the conjunctival flap.

Three months later, a 3 mm punch was used to trephine in the conjunctiva directly overlying the center of the optical part of the device. Once a partial thickness incision had been made using a disposable trephine, a full thickness incision was made using a 15° surgical blade and the conjunctiva was removed using small scissors.

At 3 months postoperatively, the KPro was well-positioned and visual acuity corrected to 20/200. The patient returned to the clinic 14 months after conjunctival flap placement, with the optical window maintained. Uncorrected distance visual acuity measured 20/250 and mild retroprosthetic membrane was noted at the slit-lamp. He underwent superior cul-de-sac reconstruction with buccal mucous membrane graft at 15 months to allow elevation of the upper lid. The conjunctival opening was expanded to approximately 4 mm diameter at this time, and the patient successfully underwent YAG retroprosthetic membrane removal after 1 week.

DISCUSSION

In this retrospective small case series, we report the use of a conjunctival flap to cover and protect the entirety of a Boston type 1 KPro device, followed by a delayed window opening for visual improvement to allow better tissue healing with vascularization to prevent tissue necrosis. Our duration of follow-up for each of these patients is >1 year with no further episodes of sterile keratolysis, making this a promising technique in this challenging population. A recent review of eyelid rehabilitation in Boston type 1 KPros was published, and described a “bucket handle technique,” in which the superior or inferior forniceal and palpebral conjunctiva is used to cover up an area of keratolysis. Mucosal membrane was then used to cover the conjunctival defect. However, they reported a 50% failure rate in their series.¹⁰ Our technique utilizes the forniceal conjunctiva and upper palpebral conjunctiva as a flap for advancement over the cornea. The flap is then secured with a 6–0 Vicryl suture to the inferior sclera, and the inferior conjunctiva is then closed with 7–0 Vicryl sutures. Corneal epithelial debridement is performed routinely in these cases in order to secure the conjunctival tissues over the cornea. Although the patient still has some residual lid function because the tarsus remains intact, because the forniceal tissues have been advanced over the cornea, the eyelid anatomy changes. Patients may require a ptosis repair later to increase the interpalpebral fissure. However, the long-term sustainability of this technique outweighs the potential derangement of structure. This technique can be utilized primarily as a two-stage procedure to allow delayed and cautious exposure of the device in patients with severe ocular surface disease and or dry eye in whom a corneal melt is anticipated. A delayed opening is particularly useful because it allows for stability of the advanced tissue, allowing for safer opening free from immediate keratolysis as published previously.¹¹ It can also be deployed as a rescue procedure in cases where there is impending or frank device extrusion. When the bulbar conjunctiva is too fragile or inadequate due to advanced scarring, which can often happen in these patients, the superior palpebral conjunctiva offers coverage for an additional surface area. In severe cases of forniceal shortening, the superior tarsal margin can be advanced onto the globe and, if need be, the upper tarsus can be everted as the tissues are advanced. Although a buccal graft is also an alternative rescue procedure, harvesting appropriate oral mucosa may be challenging to most anterior segment surgeons. The relative accessibility of this technique may be more practical to a greater number of practitioners.

Although the OOKP and tibial keratoprosthesis have the highest likelihood of retention secondary to biointegration for cicatrizing disease, their use remains limited in the United States. Furthermore, most practitioners outside of a few select institutions have not adopted the use of the type 2 device, and practically, the type 1 device remains the most commonly

utilized. In contrast to the type 2 KPro implantation, our patients did not require additional hardware or full tarsorrhaphy. The type 1 has several advantages, including greater visual field, more acceptable cosmesis, and simpler surgical technique. If further interventions are needed, such as glaucoma diode laser application and re-opening of the tarsorrhaphy, an extensive surgery would not be required. In addition, we have found that the process of rearranging the tarsal conjunctiva induces a decrease in the interpalpebral fissure that serves to further protect the drying of the ocular surface.

It is important to note that our patients were watched closely for the duration of their follow-up and needed several interventions in order to optimize results, including a two-step procedure and the use of immunomodulatory or antiviral therapy. Indeed, the success of these cases may be attributable to both the surgical techniques involved, as well as their medical management. The importance of a multidisciplinary approach and close follow-up for these challenging cases deserves highlighting.

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FIGURE 1.

Case 1, a 36-year-old man with a history of Stevens–Johnson Syndrome. The photograph shows the external appearance of the left eye 3 months following the conjunctival flap procedure to rescue an extruding Boston type 1 keratoprosthesis, which occurred immediately after the implantation.

FIGURE 2.

A shortened superior fornix with the red line demonstrating conjunctiva to be utilized for the flap (left). After surgery, a conjunctival flap has been sutured onto the inferior scleral bed with additional shortening of the superior fornix (right). Note the tarsus has changed position to slightly more horizontal than before.

FIGURE 3.

Case 3, a 65-year-old Middle-Eastern man with bilateral chronic cicatrizing conjunctivitis. (A) Slit-lamp appearance of the right eye demonstrates a partially intact amniotic membrane (stained yellow with fluorescein), Vicryl sutures superotemporally, significant stromal thinning and epithelial defect inferonasally. (B) Progression of tissue necrosis measured about 180° nasally and partial device exposure despite aggressive lubrication. (C) Six months after application of a conjunctival flap and 3 months after creation of a small window opening directly overlying the device, vision was 20/200. (D) External appearance of the patient at his last visit, 15 months after the second stage of the modified Boston type 1 keratoprosthesis implantation. The conjunctival opening remained patent. Additional surgery for elevation of the upper lid and expansion of the conjunctival opening to approximately 4 mm diameter were subsequently performed.