



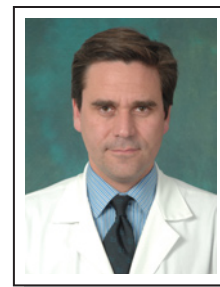
Grand Rounds

09-15-10

Introduction: Jose de la Cruz, MD (*Attending*)

This week's grand rounds presents several interesting cases encountered by the cornea service.

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Successful Boston Keratoprosthesis Type I for Bilateral Alkali Burn Injury: Renu Jivrajka, MD (*Resident*)



A 75 year old African American male presented to the UIC EEI general eye clinic in November of 2004 complaining of bilateral burning eye pain. At the time of his initial visit he had no complaints regarding his vision. His past ocular history was significant for an alkaline burn in both eyes in 1993 for which he required a 1 month hospital stay. He also underwent a penetrating keratoplasty of his right eye in 1994 that suffered a graft rejection in 1999. He had limbal stem cell transplant in 1995 in that same eye. He also had a questionable history of glaucoma secondary to topical prednisolone overuse. The patient had underlying dry eyes and was pseudophakic in both eyes.

On initial evaluation his vision was 20/400 in the right eye and 20/100 pin holing to 20/60+ in the left. His anterior segment exam revealed significant blepharitis and meibomian gland disease of both eyes with 3+ telangiectasis of his lids, eyelash tenting as well as overall eyelid skin thinning. The cornea of his right eye revealed central corneal thinning with neovascularization and an irregular surface. The left cornea similarly had an irregular surface with a centrally located anterior stromal scar. His posterior segment exam was limited due to the haziness of his corneas.

The patient was referred to the cornea clinic for further care. Initially, a conservative approach was taken through the use of aggressive lubrication and bandage contact lens. The patient however was unsatisfied and after discussing the risks and benefits of the procedure, it was determined that keratoprosthesis in the right eye would be the best option for him, since he already failed 2 corneal grafts and was limbal stem cell deficient.

Approximately 2 years after his first presentation the patient underwent Boston KPro type 1 transplantation in his right eye and just 3 months post-operatively his vision had improved from 20/400 to 20/25. He was monitored very closely post-operatively for any increase in IOP or retroprosthetic membranes which never developed in this case. The patient was also very compliant with his daily steroid and antibiotic drops.

Since the patient was so satisfied with his Kpro in the right eye even 2 years after the surgery, the discussion arose as to whether he would benefit from a Kpro in the fellow eye. At this point he had very low vision out of the left eye secondary to corneal scarring and was essentially monocular with the Kpro eye being his dominant eye. The patient was made well aware of the risk of diplopia postoperatively should there be a large discrepancy in the visual acuity in both eyes. After some deliberation, it was decided to proceed with the Kpro surgery in the fellow eye. Following surgery the patient was doing well with a vision of 20/40 in the 2nd eye just 1 week post-op. The patient was even able to regain some stereopsis which he never had after his binocular injury.

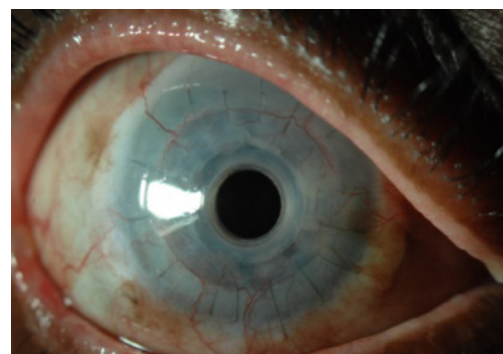


FIGURE 1
Keratoprosthesis of the right eye 2.5 years after surgery demonstrating a clear optical axis.

BACKGROUND In Keratoprosthesis type 1, the prosthesis consists of an optical front and back plates surrounding a corneal graft. The cylinder embedded in the prosthesis allows for an optically adjustable power to match refractive error of the patient. Some of the indications to perform Kpro surgeries include failed corneal grafts, visually impaired patients (unilateral or bilateral) with absent retinal or optic nerve disease and at least light perception vision. The documented complications resulting from Kpro surgery include glaucoma, retroprosthetic membranes, extrusion and endophthalmitis.

Cystoid Macular Edema after Boston Keratoprosthesis: Joshua Hou, MD (Resident)



A 45 year-old Caucasian female was referred to cornea clinic for evaluation for possible Keratoprosthesis (Boston K-pro type I) implantation in order to salvage vision in her right eye. Her past medical history was notable for Stevens-Johnson Syndrome, secondary to penicillin exposure as a child. Her past ocular history was notable for multiple surgeries in both eyes for chronic scarring and non-healing ulcers. The patient underwent four failed penetrating keratoplasty (PKP) surgeries in her right eye and was left hand-motion vision in that eye in 2004 (Figure 2). In her left eye,

she also underwent failed PKP, but subsequently underwent K-pro implantation in that eye. However, 2 years after the K-pro she developed *Candida albicans* fungal endophthalmitis in the left eye and became NLP in that eye in 2008 despite multiple vitrectomies and PKP surgeries.

On initial exam, the patient was HM OD and NLP OS with an opaque cornea OD and a phthisical OS.

The decision was made to attempt to salvage monocular vision in the patient with K-pro implantation to the right eye. Post-op she had improvement in visual acuity to 20/70 OD. Her post-op course, however, was complicated by elevated IOP poorly controlled on maximal medical therapy (approximately 3weeks post-op). She then underwent successful laser cyclophotocoagulation with normalization of pressures. She then developed increasing “floaters” by 4 weeks post-op and fundus exam revealed evidence of cystoid macular edema. At that time, she was referred to retina clinic.

OCT was obtained and patient was noted to have significant CME (Figure 3). She was treated conservatively with topical NSAIDs (Nevanac TID) and topical steroids. However, her CME continued to remain refractory to treatment after 10 months. Her visual acuity remained stable at 20/80 and after lengthy discussions with the patient regarding potential risks and benefits of more aggressive therapies such as intravitreal steroids, the patient opted to continue on conservative therapy.

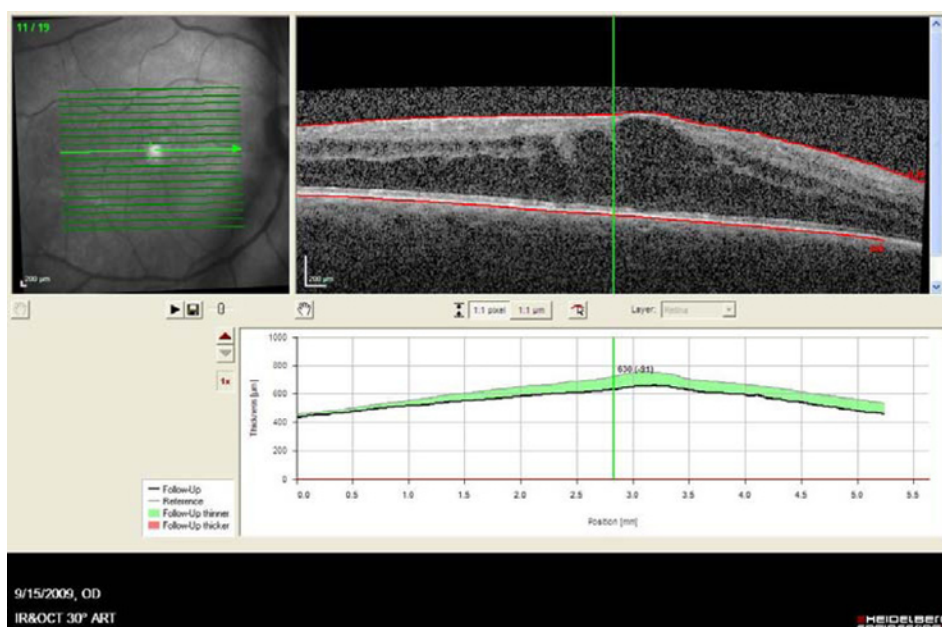


FIGURE 3
OCT OD showing persistent CME at 10weeks post-op despite therapy with topical NSAIDs and steroids.



FIGURE 2
Colored photo showing the right eye pre-op, after four failed penetrating keratoplasties.

BACKGROUND The Boston Keratoprosthesis type I is an increasingly viable surgical solution in patients with chronic, difficult to control ocular surface diseases such as Stevens-Johnson Syndrome or Ocular Cicatricial Pemphigoid, who have failed previous conventional PKP. The Boston K-pro I is not, however, without risks and numerous post-op complications have been documented.

In three major retrospective studies which included a combined total of 228 cases of Boston K-pro I implantation, the major post-op complications observed were retroprosthesis membranes (25-43%), elevated IOP (15-27%), sterile vitritis (3-10%), and infectious endophthalmitis (0-10%). Thus far there has been no published documentation of CME as a major post-op complication. Preliminary data from a retrospective review of 34 cases of Boston K-pro I implantation at the Illinois Eye & Ear Infirmary, however, found that 18% of cases developed significant CME by OCT. Of those cases, the majority (5/6) developed CME within the first 3 months post-op and there was a trend towards a positive association between increased numbers of concurrent procedures at the time of K-pro implantation and risk of post-op CME.

Post-op CME following K-pro implantation is most likely severely under diagnosed. Further research in the evaluation and treatment of post-op CME after K-pro is warranted to optimize K-pro outcomes.

Reference: Zerbe BL, Belin MW, Ciolino JB. Results from the multicenter Boston Type 1 Keratoprosthesis Study. *Ophthalmology*. Oct 2006;113(10):1779.

Lower Eyelid Retraction After Boston Keratoprosthesis Type II (Treated with Medpor Implant): Elizabeth Grace, MD (Resident)



54yo Hispanic woman with a long history at UIC Ophthalmology Department, referred to cornea clinic for keratoprosthesis evaluation. Her ocular history was notable for biopsy proven mucous membrane pemphigoid (MMP) diagnosed in 2004. Her surgical history began with bilateral RK and LASIK, failed PKP OS initially performed for impending corneal perforation, amniotic membrane graft OD after corneal melt and impending perforation, and multiple eyelid surgeries. She was co-managed with heme-oncology who tried her on multiple systemic medications to control her condition, including: dapsone and Imuran, prednisone, Cytoxan, Methotrexate and Remicade, and lastly Rituxan/IVIg treatments. Her vision was 20/400 OD and hand motion with direction OS, and she had dense ankyloblepharon, and symblepharon formation OU, fornix foreshortening, deeply inflamed conj/cornea with failed PKP OS. It was decided to perform a Boston Keratoprosthesis Type II OS, due to the lack of fornice support and end-stage dry eye. After the surgery, her vision acuity improved to 20/30 with correction, however within a

few months she started to develop retraction of the lower eyelid around the telescopic portion of the anterior optic (Figure 4). Attempting to prevent recurrent retraction often seen in KPro Type II, her surgical repair included a novel combination of using a Medpor implant cut into a donut configuration and fashioned around the telescoping optic beneath the lid closure (Figure 5).

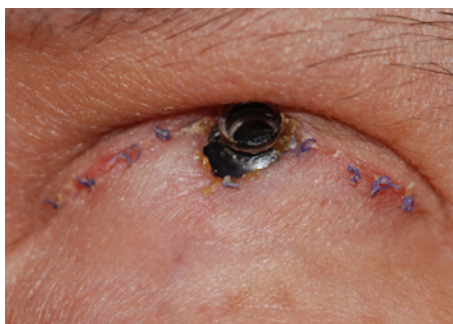


FIGURE 4

Color photo showing lower lid retraction around the telescopic portion of the anterior optic.



FIGURE 5

Intraoperative photo showing the medpor implant cut into a donut configuration and fashioned around the telescoping optic beneath the optic closure.

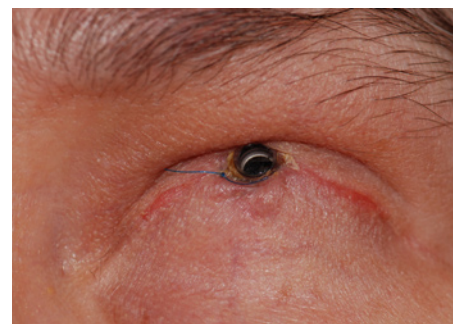


FIGURE 6

Post-operative photo. With vascularization of and incorporation of eyelid tissue into the Medpor implant over time, it may prevent the need for recurrent skin revision surgeries in the future.

BACKGROUND Patients with severe dry eye from MMP or Stevens-Johnson Syndrome often have severe ocular surface disease and lack of fornice support will not tolerate a contact lens needed for Boston KPro Type I. The Type II can be used in these patients. Complications such as retroprosthetic membrane, aqueous leak, glaucoma, can happen with either keratoprosthesis. Eyelid skin retraction around the telescoping optic is a unique complication to the KPro Type II. In one retrospective study, Sayegh RR et al. described lid skin retraction or necrosis in 40% (4/10) of eyes. This complication was noted 18.6 mo +/- 10 mo after surgery in their study. Their treatments consisted of multiple skin revisions, and one patient required replacement of Boston KPro Type II.

Patients with retraction around the telescoping optic often need repeat lid retraction repairs every few years to maintain the integrity of the keratoprosthesis. The Medpor implant was used in our patient's eyelid reconstruction with the hope that the implant will vascularize and stabilize the wound. If the implant incorporates the eyelid tissue it may prevent the need for recurrent skin revision surgeries in the future. To our knowledge this is the first use of the Medpor implant around the keratoprosthesis optic for eyelid wound stabilization.

Reference: Sayegh RR, Ang LPK, Foster S, Dohlman CH. The Boston Keratoprosthesis in Stevens-Johnson Syndrome. *Amer J of Ophthal*; 2008. 145(3): 438-444

Upcoming Grand Round

Illinois Eye and Ear Infirmary Ophthalmology Grand Rounds are held Wednesdays at 5:00 pm on the UIC campus at 909 S. Wolcott in the College of Medicine Research Building. For a complete schedule go to www.uic.edu/com/eye and click on Grand Rounds under the Education drop down menu. Or, call 312-996-6590.

UPCOMING CME COURSES

March 19-25, 2011

4th Annual Illinois Eye Review

April 22-23, 2011

UIC Cornea Symposium