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**Restoring Vision after Multiple Corneal Graft Failures with the Boston Keratoprosthesis**

**Abstract:** Severe corneal complications often require treatment with penetrating keratoplasty. This report discusses a patient who was treated with the Boston Keratoprosthesis due to a history of multiple corneal graft failures.

**I. Case History**

- **Patient demographics:** 90 y/o Hispanic Female
- **Chief complaint:** Doctor ordered 6 week follow up for Boston Keratoprosthesis OS
- **HPI:** (-) Discomfort (-) Discharge (+) Compliance with ocular medications
- **Past Ocular history**
  - Pseudophakic OU
  - Moderate Primary Open Angle Glaucoma OU, diagnosed prior to BKPro transplantation, date unknown to patient
  - Phthisis bulbi OD 2011
  - History of multiple corneal graft failures OU
- **Past Medical History**
  - High cholesterol
  - Osteoporosis
- **Ocular Surgical History:**
  - Cataract extraction OU 2000
  - S/P PKP OU 2001
  - Boston Keratoprosthesis with tube shunt inserted OS 2011
  - S/P yag to remove mild retroprosthetic membrane OS 2012
  - Tube repair OS 7/2013 (Scleral patch to fix tube erosion)
- **Social History:** No tobacco or alcohol use, living with daughter who cares for her
- **Systemic Medications:**
  - alendronate sodium
  - simvastatin
  - acetazolamide 500 mg extended release 12 hour tab PO BID
- **Ocular Medications:**
  - timolol 1 gt OS QD
  - latanoprost 0.005% 1 gt OS qHS
- fluorometholone 0.1% susp 1 gt OS BID
- tacrolimus PF 0.03% 1 gt OS BID
- vancomycin fortified PF 25 mg/ml solution 1 gt OS QD
- moxifloxacin 0.5% 1 gt OU QD

**Other salient information:** Initially, the patient had a BKPro consultation due to multiple failed corneal transplants in 2011 in both eyes. Her visual acuity was counting fingers at 1 meter in both eyes, however her right eye was beginning to become phthisical. A Boston Keratoprosthesis was recommended to salvage remaining vision in the left eye and the surgery was performed in 2011. For the past five years, the OS visual acuity has fluctuated from 20/60 (2014) to 20/400 and typically measures in the 20/200 to 20/400 range. The right eye became hand motion in January 2012. The patient is currently prescribed moxifloxacin in both eyes prophylactically to prevent secondary infection.

**II. Pertinent Findings**

- **Clinical**
  - **Unaided Distance Visual Acuities:** OD: Hand Motion and OS: 20/200
  - **Extraocular muscles:** SAFE OU
  - **Confrontation Visual Fields:** OD not tested due to phthisis bulbi; OS restricted in all quadrants
  - **Pupils:**
    - OD: Poor view due to phthisis bulbi; unable to assess
    - OS: Miotic with minimal reaction to light; unable to assess for RAPD due to phthisis bulbi of OD
  - **Slit Lamp Exam:**
    - OD: phthisis bulbi; corneal opacification due to edema, corneal neovascularization
    - OS: tube shunt covered with scleral patch graft secondary to tube erosion, BKPro with Kontur bandage contact with mild deposits
  - **Intraocular Pressure:** OD: soft with palpation; OS: Unable to take IOP measurement secondary to BKPro
  - **Fundus exam**
    - OD: No view secondary to phthisis bulbi
    - OS: Stable from previous exams; C/D ratio: 0.60/0.60

- Fundus photos: shows cupping of OS ONH

**III. Differential Diagnosis**

- Corneal graft rejection
- Corneal graft injection
- Corneal graft failure

**IV. Diagnosis and Discussion**
• **Primary Diagnosis:** S/p corneal transplant with Boston Keratoprosthesis OS, stable. No signs of infection, rejection or failure
  o Continue close monitoring every 4-6 weeks with bandage contact lens replacement. Replaced bandage contact lens in office: Kontur Precision Sphere (Plano power, DIA 18.0, BC 9.8)
  o Continue: preservative free fortified vancomycin 25 mg/ml solution 1 gt OS QD; Preservative free tacrolimus 0.03% 1 gt OS BID; fluorometholone suspension 0.1% 1 gt OS BID; moxifloxacin 0.5% 1 gt OU

• **Secondary Diagnosis:** Moderate POAG, OU. Stable
  o Due to the artificial cornea, unable to test IOP in OS. Monitor OS with regular fundus exams to watch for changes in the optic nerve head due to history of poor reliability with visual fields and OCT.
  o s/p tube shunt with scleral graft OS; tube causing small conjunctival erosion. Increased bandage contact lens diameter to 18 mm
  o OD: Non-painful eye currently phthisical state; IOP stable with palpation (soft).
  o Continue: timolol 1 gt OS QD, latanoprost 0.005% 1 gt OS qHS, acetazolamide 500 mg extended release 12 hour tab PO BID

**Discussion**

• **Pathophysiology of Corneal Graft Failure**
  o Corneal graft failure occurs when the graft is no longer viable. This most commonly occurs due to endothelial decompensation, but can also be caused by rejection, infection or primary failure of the transplanted graft. Endothelial decompensation refers to poor to non-functioning endothelial cells, which lead to corneal edema and loss of clarity. Graft rejection is caused by an immune reaction by the host tissue against the donor tissue. Primary failure of the graft refers to when the donor tissue itself is not viable prior to transplant and eventually fails after transplantation.
  o Risk factors for graft failure include: previous graft failure, corneal neovascularization, glaucoma, past history of herpes simplex and increased size of transplanted graft.

• **Symptoms**
  o Blurry vision
  o Redness
  o FB sensation
  o Watery eyes
  o Pain

• **Signs**
  o Decrease in visual acuity
Endothelial decompensation presents with: decrease in clarity of graft due to edema, increase in corneal thickness due to edema, and loss or pleomorphism of endothelial cells visible on specular microscopy.

The signs of graft rejection vary based on the specific location of rejection. Epithelial rejection presents with a rejection line that is an area of donor tissue destruction. Stromal rejection presents with haziness of the stroma. Endothelial rejection typically presents with a Khoudadoust line (keratic precipitates), stromal edema, ciliary flush and possible mild anterior chamber reaction.

Graft infection typically presents with corneal infiltrate and possible anterior chamber reaction.

- Diagnosis
  - The diagnosis of corneal graft failure is based on clinical findings.

V. Treatment and Management
- Corneal graft failure is typically treated by a repeat penetrating keratoplasty. Unfortunately, with each repeat procedure, the chances for failure are increased. An artificial cornea transplant is an option for patients with a history of multiple failures and poor vision in both eyes, as well as monocular patients.
- The Boston Keratoprosthesis (BKPro) is the most common artificial cornea that is being transplanted currently. It is indicated when multiple corneal transplants have failed or if the patient is a poor candidate for repeat penetrating keratoplasty. BKPro was FDA approved in 1992. The most recent model consists of a polymethyl methacrylate (PMMA) front-plate and a titanium back plate that holds the donor cornea in between.
  - Klufas and Colby found that patients with the BKPro had improved visual acuities, reduced incidence of infectious keratitis and better graft retention compared to repeat penetrating keratoplasty.
- Prior to surgery, the donor graft is inserted between the front and back plates, which are then snapped together. The optics of the front plate can be changed according to the patient’s refractive error. The back plate contains 16 holes, which allows the cornea to have access to aqueous humor. This is important because the aqueous humor provides essential nutrients to the cornea. In earlier models without the holes, stromal necrosis and corneal melt were common. Two forms of the Boston Keratoprosthesis exist currently; which are called “Type 1” and “Type 2.” Type 1 is the most common; type 2 is the same except for a two millimeter nub on the front plate. Patients who receive “Type 2” have a desiccated cornea secondary to end stage ocular surface disease and have been treated with a complete tarsorrhaphy. The nub protrudes through the patient’s superior eyelid and allows for vision in that eye.
- After the prosthesis is put together, the device is then transplanted surgically, similar to penetrating keratoplasty, with interrupted sutures. Typically a cataract extraction is
performed at the same time to prevent future complications. If the patient has glaucoma, a tube shunt is also placed to aid in maintaining decreased intraocular pressure.

- Post-operative care includes: one day, one week, and two week follow up visits. The patient is then seen every four to six weeks for close monitoring and replacement of bandage contact lenses.
- A bandage contact lens, typically Kontur precision sphere, is worn full time after BKPro transplantation. This contact lens is made of methafilcon A material and is preferred due to durability, adequate thickness and large diameter. It allows for “even hydration” of the cornea and prevents corneal melt.
- Common complications associated with the BKPro are retroprosthetic membrane, infection, corneal melt and glaucoma.
  - Retroprosthetic membranes can be treated with a yag laser.
  - Infection is controlled by using a low dose prophylactic antibiotic ophthalmic drop daily. A broad spectrum antibiotic, like a fourth-generation fluoroquinolone, is most commonly used. In addition, monocular patients are prescribed long-term fortified vancomycin.
  - Corneal melt has been decreased by long-term use of a bandage contact lens.
  - A majority of patients with the BKPro also have glaucomatous changes before or after the transplant. As stated before, a glaucoma tube shunt is commonly implanted along with the BKPro in patients with a prior diagnosis of glaucoma. Unfortunately, accurate measurements of the intraocular pressure cannot be taken after BKPro transplantation. These patients should be monitored closely for glaucomatous changes on fundus examination and ancillary testing (OCT and HVF), if possible.

VI. Conclusion (Clinical Pearls)

- Knowing alternative treatment options can help restore lost vision in a patient with multiple corneal graft failures.
- Patients with the BKPro must be monitored routinely for glaucoma with close assessment of the optic nerve and ancillary testing (OCT/HVF).

VI. Bibliography, literature review encouraged


