Keratoprosthesis in High-Risk Pediatric Corneal Transplantation: First 2 Cases

Infants with significant corneal opacities are at risk for development of profound deprivation amblyopia without surgical intervention. Early corneal transplantation is recommended. However, penetrating keratoplasty (PK) in young children is considered a high-risk procedure. Additionally, a poorer prognosis has been described in children with congenital corneal opacities when compared with acquired corneal opacities. Allograft rejection is the cause in the great majority of pediatric graft failures.

Implantation of a keratoprosthesis, or a synthetic cornea, is considered for patients who are at high risk for donor corneal transplantations. Although significant early and late postoperative complications can occur, favorable results have been achieved in patients with a relatively healthy ocular surface, such as in cases with multiple immunologic graft failures.

To date, keratoprosthesis implantation has been exclusively performed in adult patients. The success in children is unknown. Owing to the poor survival rate of conventional corneal transplantations in this group, we performed keratoprosthesis implantation in 2 infants with congenital corneal opacities and glaucoma to overcome deprivation amblyopia. In this article, we describe our short-term results.

Report of Cases. Case 1. A 21-month-old girl with bilateral Peters’ anomaly and associated glaucoma had undergone multiple surgical procedures in both eyes elsewhere. Initial examination under anesthesia revealed phthisis with hypotony of the right eye. The left eye had a small, opaque corneal graft with extensive neovascularization and a central epithelial defect. The intraocular pressure was 42 mm Hg. B-scan ultrasonography revealed aphakia, posterior vitreous detachment, and a thickened ocular wall. At age 22 months, the patient underwent a pars plana vitrectomy, epiretinal membrane peeling, Baerveldt tube shunt insertion, and implantaion of a permanent keratoprosthesis in the left eye (Figure).

Case 2. A 7-day-old boy was evaluated for congenital glaucoma. Examination under anesthesia revealed complete corneal opacification and an intraocular pressure of 40 mm Hg bilaterally. Ultrasonography suggested advanced cupping of the optic nerves. Trabeculotomy was performed in both eyes. At age 2 months, follow-up examination under general anesthesia revealed persistence of the elevated intraocular pressures, necessitating repeat bilateral trabeculotomy and trabeculectomy with mitomycin C. However, despite maximal topical treatment using latanoprost, dorzolamide hydrochloride–timolol maleate, and brimonidine tartrate, a tube shunt procedure had to be performed owing to the uncontrolled intraocular pressures. At age 4 months, the patient underwent a pars plana vitrectomy, lensectomy, Baerveldt tube shunt procedure, and permanent keratoprosthesis placement in the right eye. The left eye had a small area of relatively clear cornea in the inferotemporal quadrant and subsequently underwent a large optical sectoral iridectomy combined with a tube shunt procedure.

Surgical Technique and Postoperative Care. Adequate informed consent was obtained from the patients’ parents after extensive discussions regarding the risks and benefits of the procedure as compared with conventional corneal transplantation.

The devices were supplied by Dr Claes Dohlman, Massachusetts Eye and Ear Infirmary, Boston. Placement of the permanent keratoprosthesis was performed using the standard technique as described previously, using a donor corneal button oversized by 1 mm.

The postoperative regimen included 14 mg/mL of vancomycin hydrochloride 4 times daily, ofloxacin 4 times daily, 1% medroxyprogesterone acetate 4 times daily, 1% prednisolone acetate every 2 hours, combination dorzolamide-timolol twice daily, and erythromycin ointment every night. The patients were fitted with polycarbonate protective goggles. After the first 2 months, the eyedrops were each tapered to twice daily. The combination dorzolamide-timolol was discontinued after removal of the suture of the tube shunt at around 4 weeks.

An examination under general anesthesia was performed at approximately 1 and 4 weeks after the surgery and as necessary thereafter. Intraocular pressure was measured using a Tono-Pen XL tonometer (Medtronic Solan, Jacksonville, Fla) over the peripheral cornea. Retinoscopy and fitting of a large soft contact lens (Kontur Kontakt Lens Co, Inc, Richmond, Calif) was performed at 1 week postoperatively.

Results. Shortly after the removal of the suture blocking the Baerveldt tube shunt, patient 1 was found to have significant hypotony with choroidal de-
tachments. The tube had to be occluded using a 6-0 Vicryl suture (Ethicon, Inc, Piscataway, NJ) 3 mm posterior to the limbus. Subsequent examination under general anesthesia revealed a dense retroprosthetic membrane with no view of the fundus. B-scan ultrasonography revealed a total choroidal and retinal detachment. The patient underwent revision of a pars plana vitrectomy with membrane peeling, scleral buckle, fluid-air exchange, endolaser photoacoagulation, and silicone oil tamponade. Two months following the retinal detachment repair, an avascular retroprosthetic membrane was noted. This thin membrane was successfully treated with Nd:YAG capsulotomy without complications. The keratoprosthesis remained in place 15 months after the surgery, allowing for a clear view of the fundus. The eye remained soft to palpation, with an intraocular pressure of less than 10 mm Hg as measured by the Tono-Pen. The patient currently wears a contact lens of +2 diopters (D) for refractive purposes. The patient’s vision is hand motions.

No postoperative complications occurred in patient 2. The most recent examination under general anesthesia 14 months following placement of the keratoprosthesis revealed a well-positioned keratoprosthesis and tube shunt in the right eye with a clear view of the fundus. The sectoral iridectomy temporally in the left eye allowed for a clear view of the fundus. The intraocular pressures remained in the high teens and low 20s in both eyes. The optic cups were large but improved to a cup-disc ratio of 0.7, and the rims were intact. The patient wears a contact lens of −4 D OD and is able to fixate and follow with each eye.

Comment. Although congenital corneal opacities in the pediatric population are infrequent, they pose a great challenge for cornea specialists owing to the difficulties in surgical management. Poor prognostic categories in the PK pediatric group include young age (particularly <1 year) at the time of transplantation, children with anterior segment dysgeneses such as Peters’ anomaly, associated comorbidity such as glaucoma, and refractoring.

Keratoprosthesis implantation has the goal of replacing the central cornea with a clear optical cylinder made of an immunologically inert material. Therefore, allograft rejection does not occur with the keratoprosthesis. Another significant advantage is that the keratoprosthesis maintains a spherical anterior shape. Furthermore, the Boston type I keratoprosthesis, a poly-methylmethacrylate device, can be custom made such that it can correct for refractive errors as well as aphakia. These advantages are particularly important in cases of pediatric PK. Immunologic graft rejection in children can be insidious and often eludes parents’ detection. Maintaining a clear visual axis while minimizing the induced astigmatism in the immediate postoperative period may minimize the development of amblyopia in this age group. The goal in cases of significant bilateral congenital corneal opacity is to minimize the amount of time that a child is not seeing clearly until he or she nears the end of the ambylopic age range. Theoretically, keratoprosthesis implantation may provide faster and superior visual results as compared with conventional donor corneal transplantation. The device can possibly be removed and replaced by a PK when the child is out of the ambylopic age range.

Corneal blindness remains the leading cause of pediatric blindness globally.5,6 In particular, infectious keratitis is the most common cause of corneal scarring in developing countries. Importantly, in the majority of these cases, the occurrence of corneal opacification is associated with poverty in areas devoid of eye-banking and transplantation-quality corneas. Regrettably, no significant developments in pediatric corneal transplantation have been achieved in recent years, and the results of pediatric PK remain poor even in developed countries. Although the postoperative complications may be severe enough to limit the use of currently available devices, we believe that the keratoprosthesis might have a role in the management of corneal blindness in carefully selected children with complex ocular diseases who are at high risk for graft failure.

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Intravascular B-Cell Lymphoma (Angiotropic Lymphoma) With Choroidal Involvement

Intravascular large B-cell lymphoma is a rare form of extranodal lymphoma characterized by the presence of large lymphoma cells in the lumen of small blood vessels, involving multiple organs. The diagnosis is usually made at the time of autopsy. We report a patient with intravascular large B-cell lymphoma who had vision loss from choroidal involvement, which improved with systemic therapy.

Report of a Case. A 44-year-old white man was initially seen with bilateral