Penetrating Keratoplasty With a Valved Glaucoma Drainage Implant for Congenital Glaucoma and Corneal Scarring Secondary to Hydrops

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Abstract. The simultaneous management of glaucoma and corneal opacification is sometimes required in infants with severe congenital glaucoma if timely visual rehabilitation is to be achieved. A 1-month-old female infant presented with an enlarged, protuberant, opaque cornea in each eye and elevated intraocular pressure. An intrastromal, fluid-filled cleft was noted in both corneas. It resolved over 3 weeks as corneal scarring progressed. Peripheral corneal clearing allowed a view of an essentially normal anterior chamber. Penetrating keratoplasty and Ahmed (New World Medical Inc., Rancho Cucamonga, CA) valve implant surgery with mitomycin-C were performed simultaneously in the two eyes 1 month apart. At 15 months of age, the patient's grafts were clear and the intraocular pressure was well controlled in both eyes. One eye required multiple procedures for eventual glaucoma control. No postoperative overfiltration occurred. The authors conclude that the use of a valved implant should be considered in patients who require urgent simultaneous corneal and glaucoma surgery for severe congenital glaucoma. This combination may improve early postoperative control of aqueous outflow and positively affect long-term graft survival in these difficult cases. [Ophthalmic Surg Lasers 1998; 29:318–322.]

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Large elevations in intraocular pressure due to congenital glaucoma can cause enlargement of the globe with resultant hydrops and corneal scarring. When this occurs during the amblyogenic period, optically clear media needs to be provided in order to minimize the severity of amblyopia. If glaucoma has not been controlled, a procedure that will do so should be performed first in order to prevent permanent glaucomatous damage and optimize conditions for graft survival. However, during the first few months of an infant's life, the problems of amblyopia management and intraocular pressure control may require urgent and simultaneous surgery. In such situations, corneal transplantation requires the accompaniment of a procedure to control glaucoma. We present such a case here.

CASE REPORT

A 1-month-old Syrian female infant was brought to the King Khaled Eye Specialist Hospital by her parents, who reported that she was so severely proptotic at birth that she was unable to close her eyes. After the first few days of life, the proptosis subsided enough that she could close her eyes, but the parents noted that she had bilateral corneal opacification. Gestation reportedly was uncomplicated and led to a normal spontaneous vaginal delivery, unaided by forceps. The parents are first cousins.

On initial clinical examination, the patient was thought to have bilaterally hazy, thin, protuberant corneas of increased diameter. No other anterior segment details could be seen due to corneal opacification. The intraocular pressure was not significantly elevated. Ultrasonography revealed increased axial length in both eyes, but no optic nerve head cupping. A tentative diagnosis of keratoglobus was made, and the patient was scheduled for examination under anesthesia with possible placement of a limbus-to-limbus graft to reinforce the cornea in preparation for a cen-

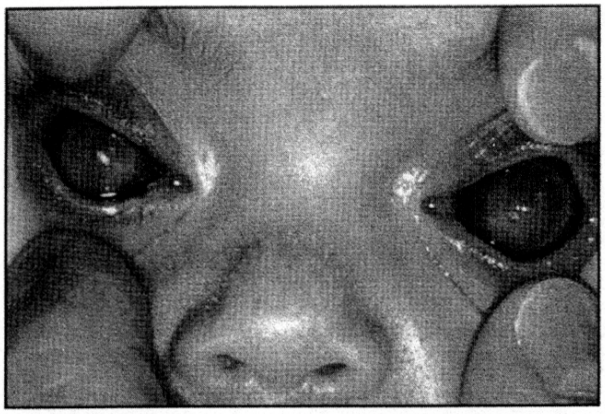

Figure 1. Preoperative photograph at 3 weeks of age.

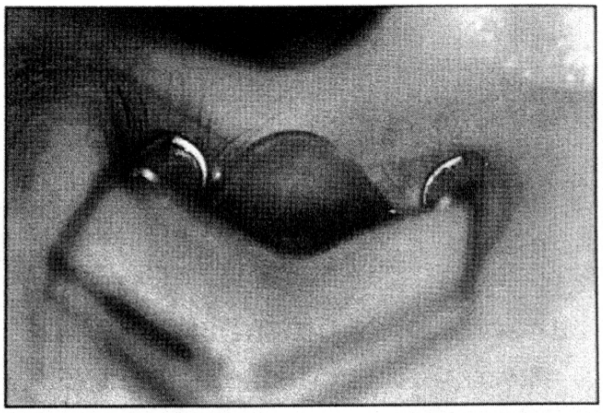

Figure 2. Temporal view of right eye at 3 weeks of age.

tral penetrating keratoplasty.

At surgery 6 days later, both eyes were observed to be buphthalmic with severely thickened, scarred, white corneas and narrow zones of peripheral corneal clearing that allowed a view of peripheral iris through formed anterior chambers (Figs. 1 and 2). The intraocular pressure exceeded 40 mm Hg, but varied according to the area of the cornea. An intrastromal cleft could now be recognized within each cornea, suggesting acute hydrops. Based on similar cases presenting to this institution, the diagnosis of congenital glaucoma with secondary corneal hydrops was made. Corneal surgery was deferred to allow resolution of the corneal hydrops. Topical timolol 0.25% was given to both eyes twice daily for control of intraocular pressure.

When the patient was 2 months old, an examination under anesthesia showed that both globes were markedly buphthalmic with horizontal diameters of 13.5 mm in the right eye and 14.0 mm in the left eye. The central 8.5 mm of each cornea was occupied by a thickened white scar. The intraocular pressure could not be reproducibly measured by pneumotonometry; however, both globes were firm by finger palpation. The patient underwent a penetrating keratoplasty in the left eye with insertion of an Ahmed glaucoma valve aqueous drainage implant following mitomycin-C application.

An inferotemporal quadrant peritomy was performed and a deep pocket was bluntly dissected beneath the conjunctiva and Tenon's flap. Mitomycin-C (0.4 mg/ml) was applied to the sclera for 5 minutes with filter paper pledgets. Once the pledgets were removed and the area was irrigated with balanced salt solution, the plate of a primed Ahmed implant was sutured to sclera such that the anterior edge of the plate was 5 mm posterior to the limbus. A Flieringa ring was sutured to the globe and trephination of an 8.5-mm button was performed. The anterior chamber was filled with viscoelastic and the button was excised with corneoscleral scissors. An 8.75-mm graft was attached to the host tissue with interrupted 10-0 nylon sutures. A large graft was used to encompass the area of elevated corneal scarring. It was believed that this large graft size would not pose a greater risk of rejection due to the presence of gross megalocornea. A 23-gauge needle was used to create a tunneled limbal tract through which the Ahmed tube was inserted into the anterior chamber. The tip was trimmed with an anterior bevel and extended 4 mm into the anterior chamber. The tube was anchored to sclera with 9-0 Prolene, and a scleral patch graft was sutured over the extraocular portion of the tube. After the conjunctiva closure, subconjunctival injections of gentamicin and dexamethasone were placed.

Postoperatively, the anterior chamber was deep and the tube was well positioned with a large inferotemporal filtering bleb. The retina was flat with no choroidal elevation. Topical steroids were tapered over 3 months to once daily. Corneal sutures were removed at 5 weeks. The graft has retained excellent clarity throughout the first 13 months of follow-up. The intraocular pressure has remained well controlled without medication, and the cup to disc ratio is 0.2.

The right eye underwent the same procedure 5 weeks after the left eye. The preoperative intraocular pressure was 33 mm Hg by pneumotonometry. The surgical procedure was similar to that for the left eye, but it was more difficult to push the plate of the Ahmed implant back into the orbit. The plate was sutured into place such that the anterior edge was

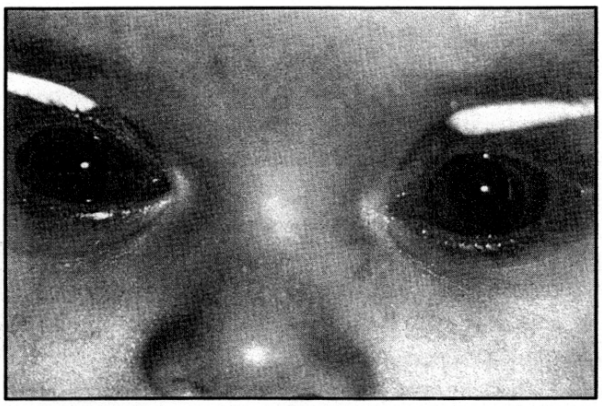

Figure 3. Postoperative photograph at 4 months of age.

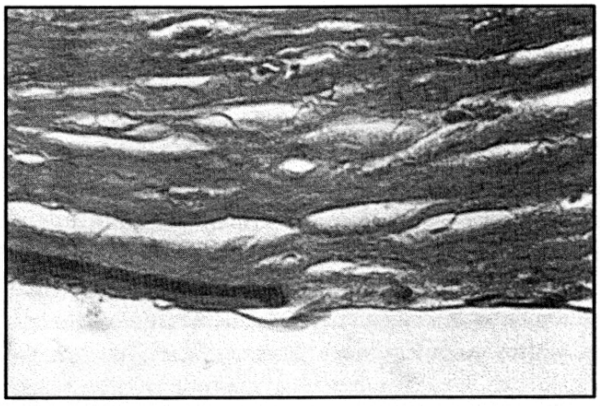

Figure 4. Histopathology of the cornea of the right eye.

diagonally oriented between 3 and 5 mm posterior to the limbus.

Three weeks postoperatively, the intraocular pressure in the right eye was 10 mm Hg by pneumotonometry. The graft was clear and there was good tube position. The retina was flat and the cup to disc ratio was 0.3. The graft sutures were removed at the sixth postoperative week. Dehiscence of the conjunctival wound at the limbus occurred and was repaired with 10-0 Dexon sutures. The conjunctival dehiscence recurred, was repaired again, and recurred a third time.

Three months after the initial surgery in the right eye, the pediatric-size Ahmed implant became available at this hospital. Because of recurrent retraction of the conjunctiva, the adult-size implant was removed from the eye and replaced with the pediatric Ahmed implant in the superonasal quadrant. Postoperatively, the graft was clear, with a deep anterior chamber and good tube position (Fig. 3). Despite a bleb over the implant, the intraocular pressure was 38 mm Hg.

During the following 8 months, both corneal grafts remained clear and the intraocular pressure was well controlled in the left eye. However, the intraocular pressure in the right eye remained around 35 mm Hg despite maximal medical therapy, trabeculectomy using mitomycin-C (0.4 mg/ml for 5 minutes), and two cycloablations using the contact YAG laser. Eventually, the patient required a third Ahmed implant. The postoperative course was uneventful and the intraocular pressure was 16 mm Hg 1 month later.

During the entire postoperative period, visual development was encouraged with early correction of the refractive error. The patient was placed in her full cycloplegic refraction 2 weeks after the second corneal graft at 3½ months of age. Her most recent refraction was -9.00 -4.00 × 180 in the right eye, and -11.00 sphere in the left eye. The patient has rapid small amplitude nystagmus in both eyes, and relative amblyopia in the right eye. She is undergoing occlusion therapy, shows good visual interest in her environment, and will fix and follow objects in both eyes.

A histopathologic examination of both excised corneal buttons showed interruption of Bowman's membrane by fibroblastic activity, thick, nonlamellar stromal collagen strands, and thin, heavily convoluted, incomplete Descemet's membrane with loss of endothelium (Fig. 4). The patient had round pupils, and no iridocorneal or lenticulocorneal adhesions were seen at surgery. This did not appear to be a case of Peter's anomaly, but rather a primary congenital glaucoma with acute hydrops and corneal scarring.

DISCUSSION

Uncontrolled glaucoma and opaque cornea require urgent attention in the infant. The media must be cleared to minimize amblyopia, and the pressure must be controlled to both allow for graft survival and prevent irreversible glaucomatous damage.

Pediatric penetrating keratoplasty has become increasingly successful. However, lower success rates have been associated with younger age.[1-4] In a study by Cowden, nearly half of the grafts remained clear in patients 1 to 4 years of age as compared with fewer than one fifth remaining clear among patients younger than 1 year of age.[1] Several factors make this procedure less successful in infants. The procedure is technically more demanding in infant eyes, which have low scleral rigidity with resulting intraoperative anterior displacement of the lens–iris diaphragm. Postoperative care is more difficult. Because infants

heal much more rapidly, there is early loosening of sutures and the need for prompt suture removal.[5] The process of graft rejection is accelerated in infants compared with in adults, and the diagnosis may be delayed because an infant is less likely to complain reliably about symptoms.[3] In cases with long-term graft survival, visual acuity may be limited by amblyopia or preexisting glaucomatous damage.

It is difficult to evaluate penetrating keratoplasty in children with congenital glaucoma because of the relatively low number of reported cases. There has been better success in eyes undergoing keratoplasty after glaucoma has been controlled. Erlich et al. reported discouraging results of 13 penetrating keratoplasty procedures performed on 8 patients with congenital glaucoma.[6] None of the grafts remained clear during the follow-up period (range, 3 months to 4 years), and 85% failed within 6 months. Four eyes became phthisical. Waring and Laibson found that penetrating keratoplasty was more successful in eyes with acquired corneal opacities (87%) than in eyes with congenital opacities (1 of 9 eyes [11%] retained a clear graft and 4 became phthisical).[7] Among the latter group were 3 eyes with either elevated preoperative intraocular pressure or previous glaucoma surgery. All 3 of these grafts failed, and 2 of the eyes became phthisical. Cowden found encouraging results in 7 congenitally glaucomatous eyes in which intraocular pressure was controlled medically or surgically before penetrating keratoplasty.[1] Although Cowden does not report the ages of his patients with congenital glaucoma, all 7 eyes retained clear grafts 1 year postoperatively.

Our patient had early corneal decompensation that caused irreversible opacification as evidenced by the histopathology. Penetrating keratoplasty was indicated to reduce the severity of amblyopia. We considered other surgical procedures to control intraocular pressure. We chose to do simultaneous penetrating keratoplasty and glaucoma surgery because performing a glaucoma procedure first would have delayed restoration of an optically clear cornea.

Other options for glaucoma management in this patient include trabeculotomy, trabeculectomy, cyclodestructive surgery, and other setons. Schanzlin et al. described 15 eyes of 11 children with congenitally opaque corneas in whom glaucoma was evident either before or after penetrating keratoplasty.[8] Several of the cases they presented were eyes that underwent trabeculotomy, trabeculectomy, thermal sclerectomy, and/or cyclocryotherapy before or after penetrating keratoplasty. In each case, the patient underwent multiple procedures in various combinations before intraocular pressure became controlled with or without medication. Cyclocryotherapy prior to or in combination with penetrating keratoplasty has met with mixed results. It culminated in phthisis in 1 of 9 cases,[7] and required repetition and/or medication to control intraocular pressure in 3 other cases.[9] In general, we prefer to leave cyclodestructive procedures as a last choice for treatment of glaucoma in these cases because of the high incidences of hypotony and phthisis that have been reported with this modality.[10,11]

Other authors have reported the use of Molteno and Baerveldt implants in combination with penetrating keratoplasty. Ariyasu et al. presented eight eyes with congenital glaucoma, all of which had undergone previous surgeries that included goniotomies, trabeculotomies, and seton operations.[12] Despite these previous procedures, five eyes had poor control of intraocular pressure at the time of penetrating keratoplasty and underwent simultaneous implantation of a Molteno or a Baerveldt implant. In this study, 67% of the grafts remained clear 30 months postoperatively, and phthisis developed in one eye. Astle et al. described a newborn with Peter's anomaly who underwent bilateral penetrating keratoplasty, with simultaneous placement of a double-plate Molteno implant in one eye.[13] Both grafts were clear 19 months after surgery. However, prolonged hypotony and medial rectus muscle palsy were reported in the eye with the Molteno implant.

We chose the Ahmed glaucoma valve implant for our case because it is a valved seton, unlike the Molteno and Baerveldt implants. A significant incidence of prolonged hypotony and flat anterior chamber has been reported with use of the single-plate Molteno implant in advanced infantile glaucoma.[14] In a recent study, Coleman et al. reported that although long-term success rates with the Ahmed implant were similar to those of other setons in adult patients, the advantage of this valved implant was the lower frequency of complications resulting from overfiltration in the early postoperative period.[15] This advantage is especially important for very young patients, in whom complications are more difficult to evaluate and manage.

Because our patient was at high risk for failure of a glaucoma filtering procedure, we used mitomycin-C with the hope of improving the long-term success of intraocular pressure control with the implant. Whether mitomycin-C provides any long-term advantage when used in such a situation with a glaucoma drainage

implant is unknown. It is possible that this antimetabolite contributed to the success of our procedure in the patient's left eye, while it may have contributed to the occurrence of conjunctival dehiscence in the patient's right eye. The long-term risks of use of mitomycin-C on the eye of a newborn are also unknown. This issue deserves further study because of the potential for this antimetabolite to enhance success rates in eyes with congenital glaucoma at high risk for failure of glaucoma filtering procedures.

We were successful in achieving a clear graft while controlling intraocular pressure with a single procedure in one eye of our patient. Although the other eye required multiple procedures, including cyclodestruction and replacement of the drainage implant, we were able to avoid postoperative loss of the anterior chamber and hypotony, which may have resulted with other nonvalved setons or trabeculectomy. Possible explanations for a more complicated course in the patient's right eye may include the occurrence of multiple surgical interventions to repair the conjunctival wound dehiscence prior to placement of the second implant, or older age at the time of the second implant. Avoidance of flat anterior chamber and hypotony as well as the potential for simultaneous management of both corneal opacification and glaucoma in a newborn are advantages of our approach. For these reasons, we believe that this is a worthwhile approach to consider in the initial management of corneal opacification with congenital glaucoma in infants.

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