

Infantile Glaucoma and Corneal Opacity

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CASE PRESENTATION

A 2-day-old Hispanic female was referred to us by her pediatrician for an evaluation of her hazy corneas and to rule out glaucoma. The patient was born by uncomplicated cesarean section at 37 weeks, at which time she weighed 7 pounds, 4 ounces. There were no exposures or complications during pregnancy, and the patient had no family history of childhood ocular problems.

Upon initial examination, the patient had mild blepharospasm and winced with both eyes when exposed to a bright light. The patient's lids and conjunctiva were normal, but her corneas showed diffuse haze (greater centrally than peripherally). Her irides and lenses were difficult to examine secondary to the corneal haze, but the anterior chambers seemed to be formed without any gross iris-to-cornea or lens-to-cornea adhesions. While she slept, we measured her IOP with a Tono-Pen XL (Medtronic Xomed Ophthalmics, Inc., Minneapolis, MN), at 42 mm Hg OD and 40 mm Hg OS. We prescribed acetazolamide 5 mg/kg t.i.d. and Trusopt (Merck & Co., Inc., West Point, PA) t.i.d. OU.

The patient was admitted to the OR when she was 5 days old. Her IOP measured 30 mm Hg OD and 34 mm Hg OS. Both corneas had a diameter of 12 mm and exhibited 4+ microcystic edema. Pachymetry measured 974 to 978 μ m OU. The anterior chambers were deep, the irides showed mild ectropion uvea, and the pupils were dilated to 8 mm,

despite preoperative pilocarpine. Axial length measured 20.5 mm OD and 20.3 mm OS. The B-scan ultrasound showed no retinal detachment or mass. Because we were unable to view the trabecular meshwork with gonioscopy, we performed bilateral temporal trabeculotomies using the metal trabeculotome (Storz Ophthalmics, St. Louis, MO) without complication (Figures 1 through 4).

The patient's corneas began to clear peripherally 2 weeks postoperatively. In the office, her IOP measured 15 mm Hg OU, and there appeared to be central Haab's striae in both eyes with overlying corneal edema. Timoptic 0.25% (Merck & Co., Inc.) was added to the patient's drug regimen. At 6 weeks postoperatively, her IOP increased to 38 mm Hg OD and 32 mm Hg OS. Lumigan (Allergan, Inc., Irvine, CA) lowered her IOP to 22 mm Hg OD and 16 mm Hg OS. The crystalline lens became visible in each eye, and there appeared to be mild lamellar opacities bilaterally. The optic nerves were not visible.

At 5 months old, the patient's IOP increased to 40 mm Hg OD and 32 mm Hg OS despite treatment with Cosopt (Merck & Co., Inc.), Lumigan, and Diamox (Wyeth Pharmaceuticals, Philadelphia, PA). We performed nasal trabeculotomies on both eyes. This intervention failed to decrease IOP, so we placed an Ahmed Glaucoma Valve (model S3; New World Medical, Inc., Rancho Cucamonga, CA) in her right eye when she was 6 months old and in her left eye

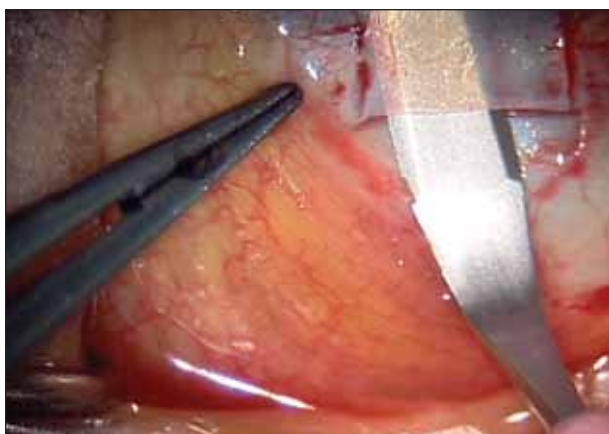


Figure 1. The surgeon dissected the scleral flap with a crescent blade.

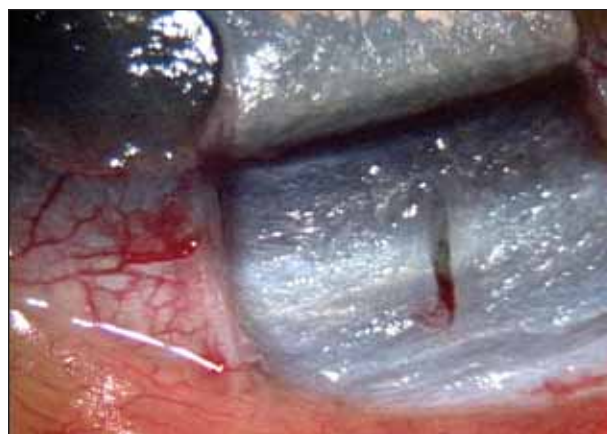


Figure 2. A radial incision unroofed the external wall of Schlemm's canal.

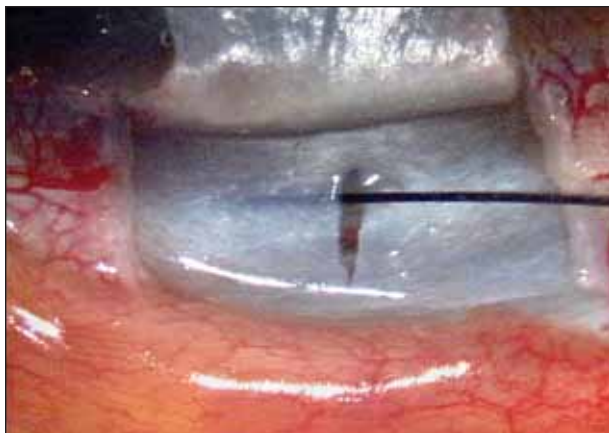


Figure 3. The surgeon placed a 6-0 nylon suture into Schlemm's canal.

when she reached 7 months of age.

The patient's corneas cleared peripherally but remained scarred centrally (Figure 5). She developed nystagmus but appeared to have sufficient vision to reach for toys. Her IOP remained in the midteens until she was 10 months old but then measured in the high 20s under anesthesia while she was on Lumigan OU bilaterally. The tubes appeared to be well positioned, and the blebs seemed to have encapsulated the Ahmed Glaucoma Valves. Axial length measured 24.1 mm in both eyes.

HOW WOULD YOU PROCEED?

1. How would you assess glaucoma control when the optic nerves are not visible and the patient cannot perform visual field testing?
2. What would be your next step in lowering this patient's IOP?
3. What should be done about the patient's central corneal scarring?

SURGICAL COURSE

Despite the patient's thickened corneas, we felt that her increasing axial length meant her IOP was too high.¹ The needling of both blebs resulted in their diffusion and decreased the IOP to the low teens bilaterally. No antime-tabolite was used. In addition, we performed transscleral diode cyclophotocoagulation inferotemporally in both eyes, six spots in her right eye and eight spots in her left at 2,000 mW.

OUTCOME

The patient's IOP has remained in the teens on Lumigan and Cosopt, and she can locate a 15-mm long candy. No corneal replacement has been performed to date. We think her large, tonic pupils have limited her deprivation ambly-

opia. Her parents have elected not to pursue corneal transplant or keratoprosthesis unless the corneal opacity becomes larger or if her vision decreases dramatically. The patient has also undergone genetic evaluation for aniridia because of her tonic mydriasis. DNA analysis and serial abdominal ultrasounds have been negative.

DISCUSSION

Infantile glaucoma may be caused by a primary defect in the development of the aqueous drainage pathways, or it may be associated with a number of ocular and systemic syndromes, including aniridia, Axenfeld-Rieger syndrome, Peter's anomaly, persistent fetal vasculature, congenital rubella, Lowe's syndrome, and Sturge-Weber syndrome.² The initial surgical management often involves drainage-angle surgery, either goniotomy or trabeculotomy. The former is a quicker procedure and does not require a conjunctival incision, but it requires a gonioscopic view of the trabecular meshwork and can be difficult in cases with corneal opacities or Haab's striae. Trabeculotomy can be performed in cases where the cornea is too cloudy to allow goniotomy, but the procedure requires a conjunctival incision and the surgeon's familiarity with locating Schlemm's canal. No large, prospective, randomized trial has compared goniotomy and trabeculotomy.

If angle surgery fails to control the IOP, the surgeon may perform trabeculectomy, shunt surgery, or cyclodestruction. Trabeculectomy has a higher incidence of failure and blebitis in children than in adults.³ Shunt surgery may be limited by the size of the infant's eye and orbit. In this case, we used the pediatric size of the Ahmed Glaucoma Valve. Alternatively, one could trim the Baerveldt plate (Advanced Medical Optics, Santa Ana, CA) if it were too large. Surgeons should be careful when performing cyclodestruction (especially in the presence of a functioning bleb or tube) to prevent hypotension or phthisis.

Aniridia is a rare ocular condition affecting approximately

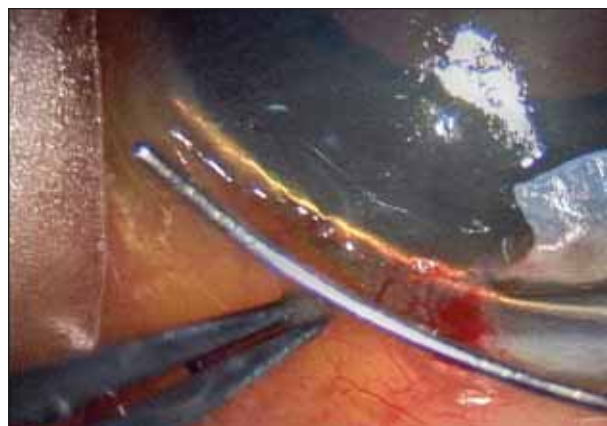


Figure 4. The metal trabeculotome is within Schlemm's canal.



Figure 5. The patient had central corneal scarring in both eyes. Notice the visible pupillary edge beyond the corneal opacity.

1 in 61,000 newborns in the US. Reduced visual acuity often results from foveal and optic nerve hypoplasia, cataracts, glaucoma, and corneal opacification. In 85% to 92% of patients with aniridia, sensory nystagmus develops. Although the pathogenesis of aniridia is unknown, a mutation in the PAX6 gene on chromosome 11 is implicated.⁴ Aniridia presents as autosomal dominant with complete penetration but variable expressivity in approximately 85% of cases.⁵ Another 15% of cases are sporadic with the same deletion or mutation in the short arm of chromosome 11 as that found in patients with Wilms tumor. Approximately 30% of patients with sporadic aniridia have Miller syndrome and develop Wilms tumor, other genitourinary abnormalities, craniofacial dysmorphism, hemihypertrophy, and severe mental retardation.⁶ Gillespie syndrome occurs in approximately 2% of aniridia patients and causes cerebellar ataxia and mental retardation.⁶

These syndromes make it imperative for a pediatrician to examine carefully all infants with aniridia. Further workup should include imaging of the abdomen and brain, chromosome analysis of the patient and family for the PAX6 gene mutation, and genetic counseling.

Corneal opacification in children often necessitates the tissue's replacement to prevent amblyopia. Corneal transplantation is challenging in infants for several reasons. First, the pliability of an immature cornea makes the surgical technique of penetrating keratoplasty difficult in comparison to that of an adult's cornea. The success of the graft is also affected by several factors, including extensive corneal vascularization and infantile glaucoma, both of which are common among patients with aniridia. Furthermore, close follow-up and a dedicated family are

essential not only for the graft's viability but also for maximum visual rehabilitation.

The infrequency of penetrating keratoplasty in infants makes determining graft survival and visual outcomes after keratoplasty difficult as well. Several retrospective studies, however, show that approximately 60% of grafts remain clear at 1 year and approximately 50% of eyes achieve a visual acuity of 20/400 or better.^{7,8} Because of the relatively high rate of graft rejection and the amblyogenic effect of irregular astigmatism, we have been performing keratoprosthesis in infants with corneal opacities. Currently, we use two distinct keratoprosthesis designs, both of which offer the potential of establishing a clear visual pathway in a short time frame. We have implanted five devices to date, but the time frame is too short for us to offer an assessment. It seems that, when we implant the device very early in life, the infant is less likely to disrupt the integrity of the device before the eye has healed. Protecting the eye is critical and becomes more difficult with time. □

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