

Secondary Glaucoma in a Child With Microcornea and Aphakia

BY NALINI K. AGGARWAL, MD, AND JESS T. WHITSON MD, FACS

CASE PRESENTATION

In February 2004, a 10-year-old black male presented for an initial consultation regarding elevated IOP. The patient had a history of congenital cataracts, the removal of which when he was 4 months old had left him bilaterally aphakic. He also had bilateral microcorneas with corneal diameters of 8 mm. At the ages of 2 and 3 years, he had undergone unsuccessful attempts at penetrating keratoplasty in his left eye that resulted in a failed graft.

Upon initial evaluation, the patient's BCVA was 20/40 in his right eye and count fingers at 1 foot in his left eye. His right eye's refractive error measured $+7.00 +2.00 \times 75$. His right pupil was reactive with a relative afferent pupillary defect on the left side. The child demonstrated complete suppression of his left eye on Worth 4-dot testing, and there was no stereopsis. His IOP measured

34 mm Hg OD and 17 mm Hg OS by pneumotonometry. The referring ophthalmologist had noted that the patient's IOP had fluctuated between 20 and 35 mm Hg OD on various topical glaucoma medications during the past 2 to 3 years.

An external examination was unremarkable. The patient's extraocular movements were full with a small-angle esotropia in his left eye and an intermittent horizontal nystagmus in both eyes. The anterior segment examination was remarkable for scarred conjunctiva superiorly in both eyes. The right cornea was clear, whereas an opaque corneal graft was present in the left eye. The right anterior chamber was quiet. On gonioscopy, the anterior chamber angle in the patient's right eye was open to the scleral spur for 360°. There was no view of the angle in his left eye. A fundus examination of his right eye revealed a moderate cup-to-disc ratio of approximately 0.8 (Figure 1). Pachymetry yielded a central corneal thickness of 685 μm OD. A visual field test of his right eye showed a mean deviation of -17.16 dB with a dense inferonasal step and a generalized depression superiorly (Figure 2).

The referring ophthalmologist had followed the patient, who was taking Cosopt (Merck & Co., Inc., West Point, PA), Alphagan (Allergan, Inc., Irvine, CA), and pilocarpine gel on his initial visit to our clinic.

HOW WOULD YOU PROCEED?

1. What would be your next step in managing this functionally monocular patient?
2. When assessing glaucoma, what are the implications of an increased corneal thickness in a patient with microcornea?
3. If you chose to intervene surgically to manage the glaucoma in the patient's better seeing eye, would you perform a trabeculectomy, or would you implant a

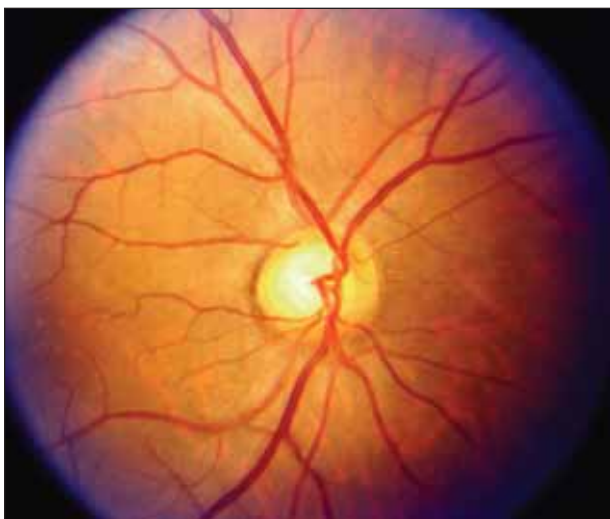


Figure 1. This fundus photograph shows optic nerve cupping with an approximate cup-to-disc ratio of 0.8 in the patient's right eye.

glaucoma drainage device?

4. If you chose to place a tube shunt, which type and position would you select?

CLINICAL AND SURGICAL COURSE

Despite the addition of a prostaglandin analog to the patient's existing medical regimen, his IOP remained elevated at between 30 and 35 mm Hg OD. Considering the moderate cupping of the optic disc, visual field changes, and elevated IOP on maximal medical therapy, we elected to place a glaucoma drainage implant. With the patient under general anesthesia, we performed an anterior vitrectomy. We then implanted an Ahmed Glaucoma Valve Flexible Plate (FP-7; New World Medical Inc., Rancho Cucamonga, CA) in the superotemporal quadrant of his right eye, with a pars plana insertion of the tube behind the iris plane.

OUTCOME

On the first postoperative day, the patient's visual acuity was 20/100, and his IOP measured 14 mm Hg with pneumotonometry. The bleb over the plate was diffuse, and the anterior chamber was well formed. A fundus examination revealed a flat retina. We pre-

scribed a topical antibiotic and steroid q.i.d. for his right eye.

The patient's IOP remained between 13 and 14 mm Hg OD until approximately 3 weeks postoperatively, when it measured 32 mm Hg. We felt that this elevation was due to a hypertensive phase sometimes encountered in the early postoperative period following the implantation of a glaucoma tube shunt. We had him restart a b.i.d. regimen of Cosopt.

During the patient's last examination, 3 months postoperatively, his IOP measured 14 mm Hg OD on Cosopt b.i.d. A slit-lamp examination revealed a quiet right eye with a clear cornea and a well-healed incision in the superotemporal quadrant (Figure 3). The pressure in his left eye has remained stable at 15 to 16 mm Hg on the drug regimen described earlier in the case presentation.

DISCUSSION

Congenital microcornea may result in secondary open-angle glaucoma or secondary angle-closure glaucoma. The former is thought to be similar to developmental glaucoma with early angle maldevelopment.¹ Angle-closure glaucoma may appear at a younger age than in patients with simple pupillary block, and it is most likely related to more complex mechanisms.¹ Additionally, increased corneal thickness can complicate the accurate assessment of IOP, particularly with Goldmann applanation tonometry. Pneumotonometry may be less affected by differences in corneal thickness than Goldmann applanation tonometry.²

Glaucoma has also been recognized as an infrequent, but serious, complication following cataract surgery in children; its prevalence has ranged from 6.1% to 24% in various series.³ Most of the children who developed glaucoma after cataract surgery had an open-angle mechanism, but little is known about the risk factors.

In a series of 240 eyes on which physicians performed cataract surgery, investigators found an increased relative risk for developing glaucoma in patients aged less than 1 year and in patients with microcornea.³ In another series, researchers identified 48 eyes with aphakic glaucoma and found that 45 of them (94%) had microcornea when compared with the normal corneal diameter for the subjects' age.⁴ Notably, children's eyes with aphakia or pseudophakia have recently been reported to have a significantly increased corneal thickness when compared to their phakic fellow eyes.⁵

Microcornea can present a significant obstacle to the success of glaucoma implant surgery due to the crowded anterior chamber angle structures. The decision to insert the tube posterior to the iris is typically made preoperatively, and an anterior vitrectomy is usually required to

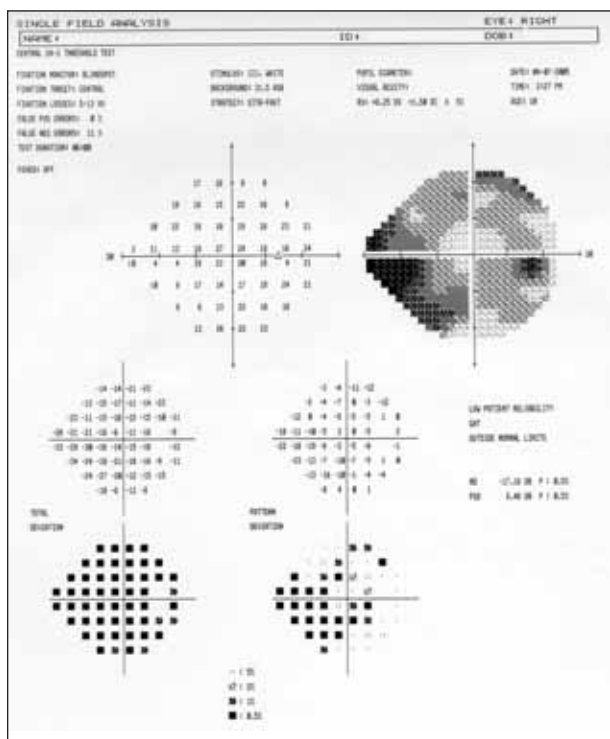


Figure 2. A Humphrey Visual Field 24-2 (Carl Zeiss Meditec Inc., Dublin CA), stimulus III, shows a dense inferonasal step along with generalized depression in the superior field of the patient's right eye.

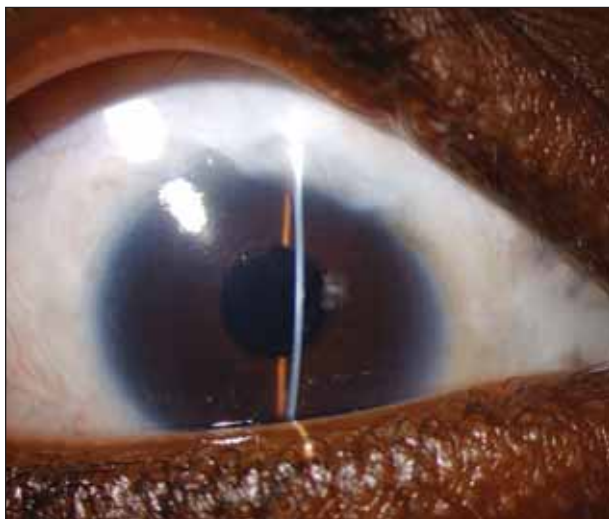


Figure 3. This slit-lamp photograph shows a quiet right eye at 3 months postoperatively with a well-formed anterior chamber. The tube shunt (not seen here) was observed at the slit lamp to be just behind the iris in the superotemporal quadrant.

avoid blockage of the tube by vitreous postoperatively. The pars plana insertion also has the advantage of preventing cornea-tube touch, which may lead to corneal edema or decompensation. In this case, we decided to proceed with this approach, and we fashioned the tube with the bevel down to prevent its occlusion by iris tissue. A mean postoperative IOP of 14 mm Hg with an average of 0.6 glaucoma medications has been reported following pars plana tube insertion.⁶

Aphakia itself can also raise the risk of complicated glaucoma surgery by multiple mechanisms, including distortion of the anterior chamber angle, inflammation secondary to retained lens fragments, vitreous in the anterior chamber, pupillary block by the anterior vitreous face, and peripheral anterior synechiae.¹ Preoperative considerations to reduce the volume of vitreous such as digital pressure or the use of acetazolamide may prevent immediate postoperative increases in IOP. The judicious use of viscoelastics and the intraoperative injection of miotics have been found to significantly affect IOP following cataract and glaucoma surgery, especially in eyes with preexisting aphakia.⁷⁻⁹

Our decision to implant an Ahmed Glaucoma Valve in this case was primarily based on the need for expedient IOP control. Overall success rates of 55% to 95% have been reported in the pediatric population, with no clear advantage between Molteno (Molteno Ophthalmic Limited, Dunedin, New Zealand), Baerveldt (Advanced Medical Optics, Inc., Santa Ana, CA), or

Ahmed implants.¹⁰⁻¹² Given the multiple barriers to success in our patient, it is fortunate that the surgical outcome has been successful thus far. Our plans are to maintain close follow-up with a possible discontinuation of the topical glaucoma medication if his IOP remains adequately controlled. □

Nalini K. Aggarwal, MD, is Clinical Assistant Instructor and currently a Clinical Glaucoma fellow, Department of Ophthalmology, University of Texas, Southwestern Medical Center, Dallas. She acknowledged no financial interest in the products or companies mentioned herein. Dr. Aggarwal may be reached at (214) 648-2020; guptana@yahoo.com.



Jess T. Whitson, MD, FACS, is Associate Professor, Department of Ophthalmology, University of Texas, Southwestern Medical Center, Dallas. He acknowledged no financial interest in the products or companies mentioned herein. Dr. Whitson may be reached at (214) 648-4733; jess.whitson@utsouthwestern.edu.



1. Shields MB, Allingham RR, Damji K, et al. *Shields's Textbook of Glaucoma*. 5th ed. Philadelphia, PA: Lippincott, Williams, and Wilkins; 2005.
2. Bayraktar S, Bayraktar Z. Central corneal thickness and intraocular pressure relationship in eyes with and without previous LASIK: comparison of Goldmann applanation tonometer with pneumotonometer. *Eur J Ophthalmol*. 2005;15:355-360.
3. Mills MD, Robb RM. Glaucoma following childhood cataract surgery. *J Pediatr Ophthalmol Strabismus*. 1994;6:355-360.
4. Wallace DK, Plager DA. Corneal diameter in childhood aphakic glaucoma. *J Pediatr Ophthalmol Strabismus*. 1996;5:230-234.
5. Simon JW, O'Malley MR, Gandham SB, et al. Central corneal thickness and glaucoma in aphakic and pseudophakic children. *J AAPOS*. 2005;4:326-329.
6. Luttrull JK, Avery RL, Baerveldt G, et al. Initial experience with pneumatically stented Baerveldt implant modified for pars plana insertion for complicated glaucoma. *Ophthalmology*. 2000;107:143-149.
7. MacRae SM, Edelhauser HF, Hyndiuk RA, et al. The effects of sodium hyaluronate, chondroitin sulfate, and methylcellulose on the corneal endothelium and intraocular pressure. *Am J Ophthalmol*. 1983;95:332-341.
8. Bomer TG, Lagreze W-DA, Funk J. Intraocular pressure rise after phacoemulsification with posterior chamber lens implantation: effect of prophylactic medication, wound closure and surgeon's experience. *Br J Ophthalmol*. 1995;79:809-813.
9. Wood TO. Effect of carbachol on postoperative intraocular pressure. *J Cataract Refract Surg*. 1988;14:654-656.
10. Netland PA, Walton DS. Glaucoma drainage implants in pediatric patients. *Ophthalmic Surg*. 1993;24:723-729.
11. Eid TE, Katz LJ, Spaeth GL, et al. Long term effects of tube-shunt procedures on management of refractory childhood glaucoma. *Ophthalmology*. 1997;104:1011-1016.
12. Djodeyre MR, Peralta CJ, Abelairas GJ. Clinical evaluation and risk factors of time to failure of Ahmed Glaucoma Valve implant in pediatric patients. *Ophthalmology*. 2001;108:614-620.

CONTACT US

Glaucoma Today accepts submissions of challenging cases. Articles must be approximately 1,200 words and arranged in the format shown (case presentation, questions, surgical or clinical course, outcome, discussion). Please include one or two figures. E-mail your case to gmcdermott@bmctoday.com.