

Aniridia and Severe Corneal Opacity

BY RICHARD J. MACKOOL, MD

CASE PRESENTATION

A 41-year-old white male with a history of congenital aniridia and developmental glaucoma in both eyes presented with a complaint of a gradually progressive loss of visual acuity in his left eye during the previous 3 months. He stated that the vision in his right eye was lost due to glaucoma (a Baerveldt valve [Pfizer Inc., New York, NY] had been implanted OD) and that until recently he had been able to function in his occupation as a computer programmer by relying entirely upon his left eye. His BCVA was limited to a recognition of hand motion OU. Nystagmus was present bilaterally. Corneal examination revealed a dense, vascularized corneal pannus in both eyes that was greater in his left eye (Figure 1). A poor view of the anterior segment was obtained due to corneal opacification, but there appeared to be a mature, or nearly mature, cataract present in his left eye. Both the corneal and scleral surfaces were dry despite the use of artificial tears bilaterally every 1 to 2 hours. Topical medications included Alphagan (Allergan Inc., Irvine, CA), Timoptic (Merck & Co., Inc., West Point, PA), and Xalatan (Pfizer Inc.) OS b.i.d. B-scan ultrasonography of both eyes was



Figure 1. Corneal examination revealed a dense, vascularized corneal pannus in both eyes that was greater in his left eye.

normal. Keratometry was not possible because of the diffusely irregular corneal surface. The axial length measured 28.5 mm in his left eye.

Because of the patient's history of adequate, functional vision with his left eye until recently, I presumed that progressive lens opacification was the cause of his recent loss of vision. Therefore, cataract-implant surgery was planned for his left eye.

HOW WOULD YOU PROCEED?

1. What method of cataract extraction would you select?
2. Would you address the corneal opacity? If so, would you do this before or during the cataract-implant procedure?
3. How would you determine the IOL power required?

SURGICAL COURSE

I planned to perform phacoemulsification in the patient's left eye. Before entering the eye, a limbal peritomy was performed, and the vascular pannus was dissected from the surface of the cornea (Figure 2). The pannus proved to be quite thick (approximately



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Figure 3. Adequate visualization for sculpting the nucleus was achieved with the use of a fiberoptic light.

0.3 mm). Beneath the pannus, the surface of the patient's cornea was irregular, with multiple depressions and surface abnormalities, and there was significant corneal stromal haze. The latter precluded adequate visualization of anterior segment details required to perform phacoemulsification. The placement of viscoelastic on the corneal surface provided somewhat improved visualization, but the view was still inadequate because of corneal stromal haze.

How would you address this problem?

The procedure was halted, and the patient was escorted from the OR to an examination area. Upon slit-lamp examination, the majority of the corneal opacity appeared to be within the anterior stromal layers, and the thickness of the cornea was adequate.

What methods might be attempted in order to improve corneal clarity?

The patient returned to the OR. Using a battery-driven, high-speed burr, the anterior layers of the cornea were removed until the cornea was relatively smooth and its clarity was maximal, although some deep stromal haze remained. I created the sideport and primary phacoemulsification incisions and stained the anterior capsule with trypan blue. I made a central incision in the anterior capsule with a bent-tipped needle, but visualization remained suboptimal and inadequate for the performance of a capsulorhexis.

What steps might now be taken to improve visualization of the anterior segment?

After enlarging the sideport incision to 1.2 mm, I inserted a fiberoptic light. The OR room and microscope lights were turned off, and the capsulorhexis was completed using intraocular illumination. The

fiberoptic light was removed and the microscope light was turned on. I performed hydrodissection and began phacoemulsification, but visualization for sculpting of the nucleus was inadequate. Therefore, the fiberoptic light was reintroduced, and the microscope and room lights were again turned off. Adequate visualization was achieved (Figure 3), and the lens was sculpted. The fiberoptic light was removed, the microscope light was turned on, and a phaco chopper was introduced. Visualization without the fiberoptic light was adequate for chopping, which I performed. The fiberoptic light was then reintroduced for both nucleus segment removal with the ultrasonic handpiece and cortex removal with the I/A instrument.

How would you determine the IOL power required for this patient?

The operation was halted, and the patient was again taken to an examination area where an aphakic refraction was performed. I calculated the IOL power on the basis of this refraction, and the patient returned to the OR. I injected the AcrySof SA-60 single-piece acrylic IOL (Alcon Laboratories, Inc., Fort Worth, TX) into the eye after instilling viscoelastic to inflate the capsular sac. However, it was not possible to determine with certainty that the IOL had been delivered into the capsular sac.

How would you verify that the IOL was in the desired location?

A fiberoptic endoscope was inserted through the phacoemulsification incision. By viewing the monitor, I could determine the IOL location; the entire lens was within the capsular sac. A bandage contact lens was placed on the eye after irrigation of visco-

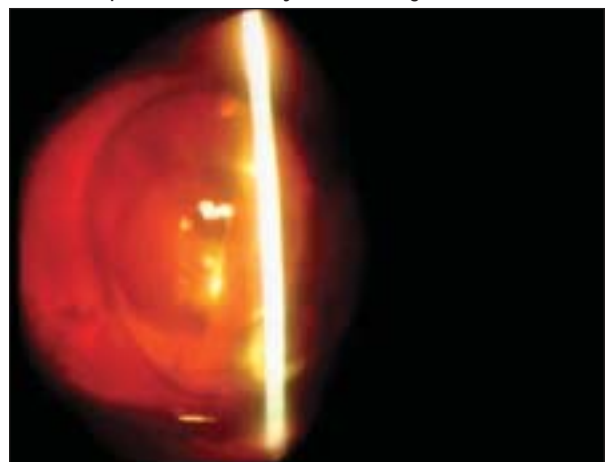


Figure 4. At 3 months postoperatively, the patient reported that his functional acuity was better than it had ever been in his adult life.

CATARACT SURGERY CHALLENGING CASES

elastic from the anterior chamber and verification that the incisions were watertight.

OUTCOME

The patient achieved a BCVA of approximately 20/300 within 2 weeks. In addition to the usual postoperative cataract medications, he used the topical lubricant, Systane (Alcon Laboratories, Inc.), every 15 minutes. At 3 months postoperatively (Figure 4), the patient reported that his functional acuity was better than it has ever been in his adult life (in fact, he stated that he was now able, for the first time, to see his wife's face). Careful observation for possible recurrent corneal pannus is ongoing, and the possibility of future limbal stem-cell transplantation has been discussed with the patient.

DISCUSSION

The successful outcome achieved in this case required the combination of a number of technologies and unusual surgical techniques. For phacoemulsification to be safely performed, adequate visualization is obviously necessary. In many eyes with mild-to-moderate corneal opacification, capsular staining dyes and the appropriate selection of microscope light intensity are sufficient. In cases of more severe corneal opacification, as in the present case, more extreme measures are required.

I first used fiberoptic endoillumination during phacoemulsification 15 years ago. This technique is common during the removal of lens material from the posterior segment. Because I had utilized the method many times during vitreoretinal surgery, I adapted it to the anterior segment.

In this case, the surgical procedure was interrupted twice, first to evaluate the cornea by slit-lamp examination and again to perform an aphakic refraction in order to determine the appropriate IOL power. Halting a surgical procedure in order to gather more data should be done whenever necessary. Other examples of this need include anterior chamber shallowing despite normal or elevated IOP (possible infusion misdirection syndrome or subchoroidal hemorrhage) and an uncooperative patient.

The aphakic refraction technique is extremely valuable. I have used it on hundreds of occasions during the past 10 years in order to determine the appropriate IOL power when keratometric or biometric problems are present (eg, eyes that have undergone LASIK/PRK or that have irregular corneas, posterior staphyloma, nystagmus, or vitreous opacities such as asteroid hyalosis). For more than 20 years, I have used a variant of this technique (intraoperative retinoscopy) in order to accurately determine the IOL power in children undergoing cataract extraction with general anesthesia.

Finally, the use of an endoscope to evaluate otherwise obscured intraocular anatomy can be extremely advantageous. Additional situations in which this technology/technique can be helpful include the evaluation of the position of the haptic of a subluxated IOL, the examination of an iris/ciliary body tumor, and the appropriate placement of a sulcus-fixation suture. ■

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