

Case Presentation: Bilateral Angle-Closure Glaucoma

A thorough workup detects congenital changes in a patient's crystalline lenses.

BY TANIA PAUL, MD, AND NATHAN M. RADCLIFFE, MD

A 35-year-old woman presented to the ER with complaints of ocular pain that had persisted for 1 day. She reported a sensation of pressure and blurry vision in her left eye with no apparent exacerbating factors. In the ER, her UCVA measured 20/30 OD and 20/100 OS. A slit-lamp examination of her left eye revealed moderate conjunctival injection, corneal edema, and pigmentary deposits on the corneal endothelium. The anterior chambers of both eyes were shallow centrally and flat peripherally (Figure 1), but these findings were more pronounced in her left eye. The crystalline lenses were clear, and the IOP measured 46 mm Hg OD and 52 mm Hg OS. Both pupils were nonreactive and dilated midway, and they demonstrated posterior synechiae presumed to be from chronic iridolenticular contact.

Fundoscopy revealed pink optic nerves with sharp margins, bilateral vertical cup-to-disc ratios of 0.25, intact neuroretinal rims, and retinal nerve fiber layers without papillary atrophy. Darkroom gonioscopy revealed a convex

iris approach and intermittent peripheral anterior synechiae in both eyes for 360°. The patient was diagnosed with bilateral acute angle-closure glaucoma (ACG) and was started on topical ocular hypotensive treatment. No systemic medications or cyclopegic agents were prescribed.

When we evaluated the patient 2 days after her initial presentation to the ER, the eye drops had adequately reduced and stabilized her IOP, and the corneal edema in her left eye had resolved. The patient subsequently underwent bilateral peripheral iridotomies.

Postoperatively, the patient's UCVA returned to 20/15 OU, and her IOP was initially controlled with topical therapy in both eyes. Her anterior chambers remained shallow however, and she continued to demonstrate appositional closure on gonioscopy that was not relieved with compression. A subsequent fundoscopic and stereophotographic evaluation of the optic nerves showed cup-to-disc ratios of 0.4 OD and 0.7 OS. We also noted pathological changes in the retinal nerve fiber layer bilaterally (Figure 2) and an infe-

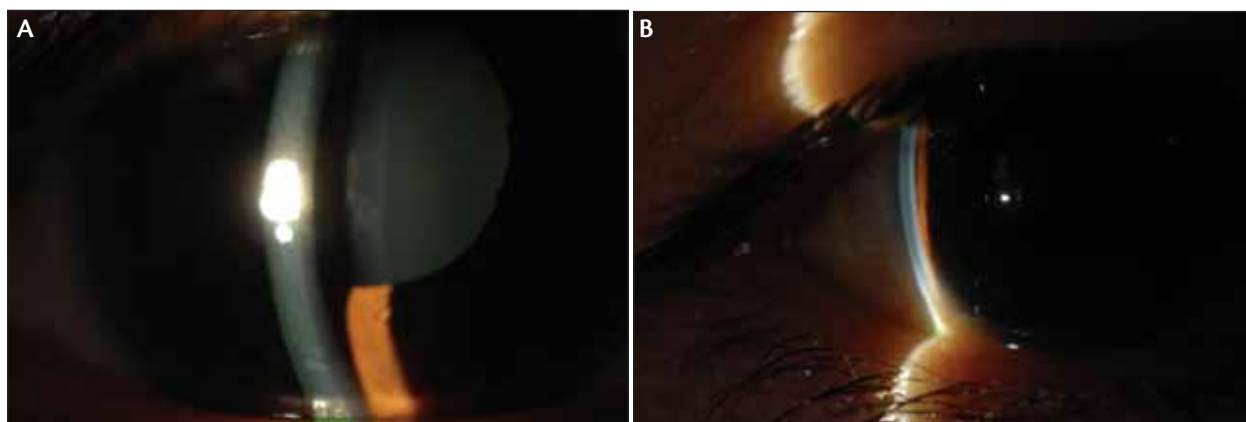


Figure 1. A slit-lamp photograph of the patient's right eye showed a fixed, middilated pupil (A) and shallowing of the central and peripheral anterior chamber (B).

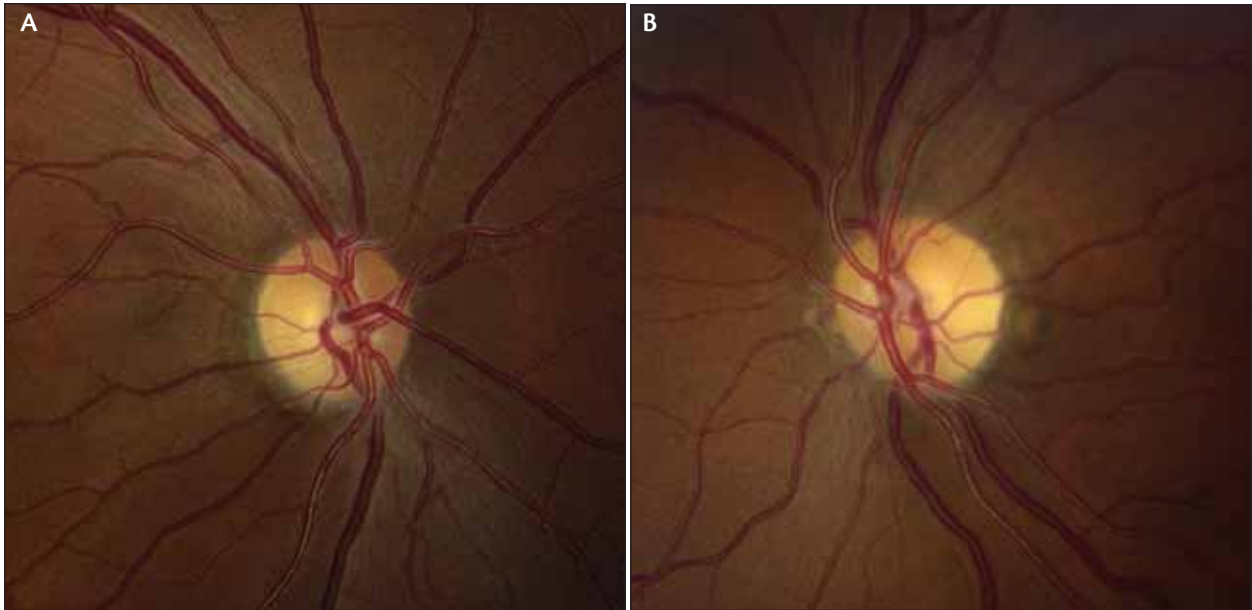


Figure 2. Fundus photographs of the patient's right (A) and left (B) eyes showed pathological changes in the retinal nerve fiber layer secondary to acute ACG.

rior notch in the left optic nerve that correlated with superior visual field defects (Figure 3) (mean deviation, -10.99 dB OD and -17.75 dB OS) on Humphrey visual field testing (Carl Zeiss Meditec, Inc., Dublin, CA).

Two months after the patient initially presented to the ER, she was re-evaluated in our office, at which time her IOPs were elevated despite self-reported adherence to a regimen of three topical ocular hypotensive medications. She did not report experiencing ocular pain, headache, blurred vision, or erythema despite IOPs of 48 mm Hg OS and 50 mm Hg OD. The examination did not reveal any corneal edema or intraocular inflammation in either eye.

HOW WOULD YOU PROCEED?

1. What further testing would you order to identify the underlying cause of this patient's ACG? Is her presentation characteristic of a syndrome?
2. If the patient requires surgery, would you perform cataract extraction, a trabeculectomy, or a combined procedure?
3. Would you perform goniosynechiolysis? What are the indications for this procedure in this setting?
4. If you performed a lensectomy, what refractive endpoint would you target for this 35-year-old emmetrope who has never experienced asthenopia?

SURGICAL COURSE

Ultrasound biometry and biomicroscopy showed axial lengths of 21.6 mm OD and 21.7 mm OS and bilateral lens thicknesses of 4.75 mm. An evaluation with an Artemis 2

very high-frequency ultrasound (Arcscan, Inc., Golden, CO; not available in the United States) confirmed persistent iridotrabecular apposition with lens-induced angle closure (Figure 4). We noted that the ciliary body was somewhat hypoplastic and anteriorly rotated in both eyes. We did not observe any uveal effusions.

These pathological changes were suggestive of spherophakia. A review of systems showed that the patient was of short stature but did not have brachydactyly or abnormal dentition. Because she had been adopted, she could not provide a family history.

When we discussed her options for treatment, the patient, who was an executive in the fashion industry, stated that she preferred to avoid spectacle correction for near and distance vision postoperatively. Specifically, she desired the ability to read and use her handheld personal digital assistant without eyeglasses.

Unfortunately, the patient was not a good candidate for pseudoaccommodating or accommodating IOLs, because her pupils were irregular and she had advanced visual field loss. We were also concerned that she could have zonular instability secondary to spherophakia and that her short axial length would make it difficult to predict her refractive outcome. We instead implanted a monofocal IOL and targeted a mildly myopic refraction (-1.50 D) in the patient's left eye only. This refraction was chosen to help the patient obtain spectacle independence, with or without monovision, depending on the targeted refraction in her second eye. We decided to extract the lens from her left eye, because the angle closure had a phacomorphic component

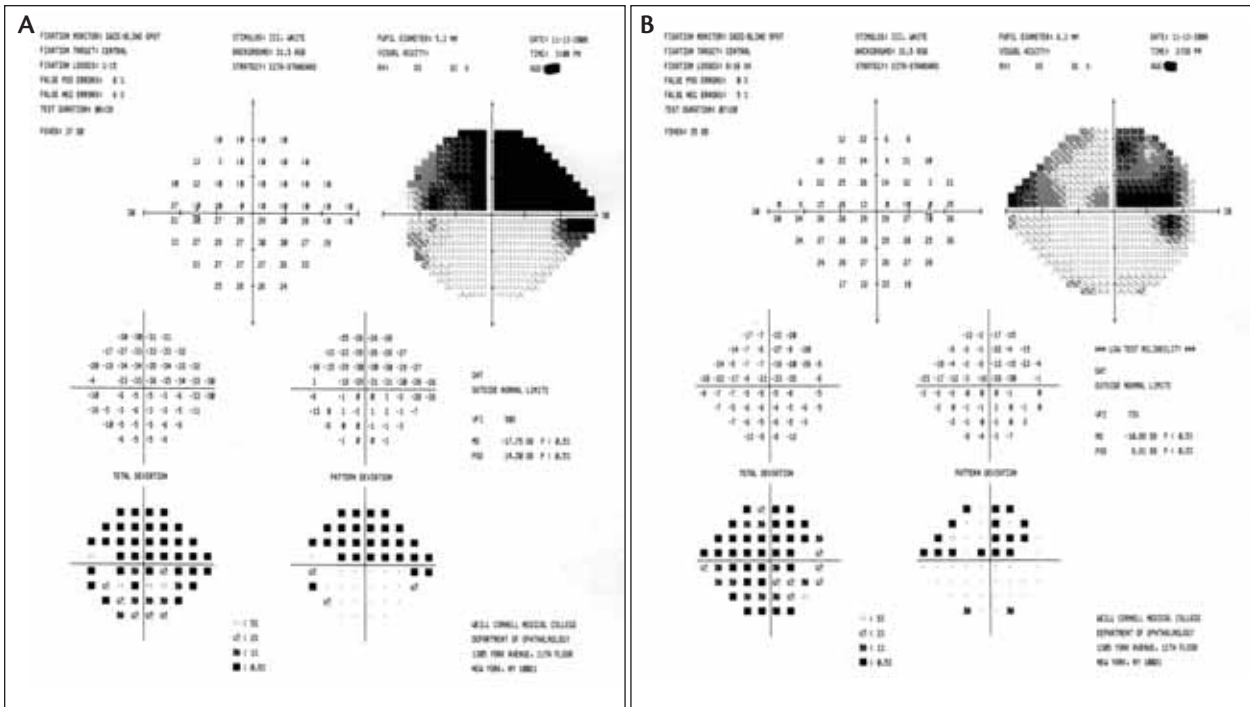


Figure 3. Perimetry showed superior visual field defects in the patient's right (A) and left (B) eyes. The defect in the left eye correlated with a notch in the left optic nerve.

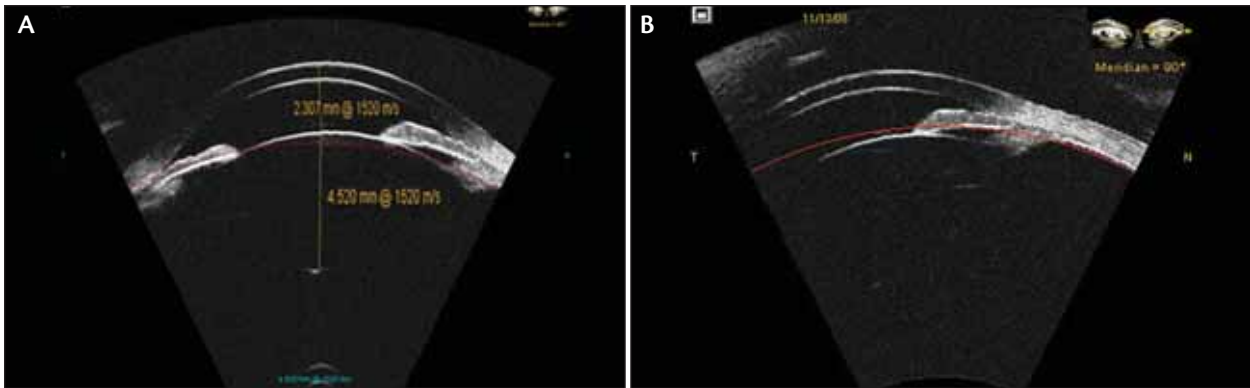


Figure 4. Imaging with very high-frequency ultrasound demonstrated a large crystalline lens and a shallow anterior chamber (A) with persistent iridotrabecular apposition (B).

and the patient had more visual field defects in this eye than in her right eye. In addition, we performed goniosynechiolysis to resolve the extensive peripheral anterior synechiae (presumed to be of less than 1 year's duration) and to re-establish the left eye's anatomic angle.

OUTCOME

Postoperatively, the patient achieved an acuity of 20/20 OD with mild myopic correction. Unlike her right eye, which still required three topical medications to maintain a stable IOP, her left eye had excellent IOPs, ranging between 7 and 10 mm Hg without hypotensive drugs. We also noted

significant postoperative deepening of the anterior chamber (Figure 5), and gonioscopy revealed an essentially open angle with some residual peripheral anterior synechiae nasally. Despite these positive results, the patient is bothered by asthenopia in her left eye and has deferred surgical intervention in her right eye. We continue to monitor her closely.

DISCUSSION

Bilateral ACG is an unusual and uncommon occurrence. The differential diagnoses for bilateral ACG are limited and include uveal effusion related to medication, general anesthesia, snake venom, and lenticular/zonular



Figure 5. Surgical intervention significantly deepened the anterior chamber of the patient's left eye.

abnormalities.¹⁻⁵ In this case, we ruled out uveal effusions. Based on the size and shape of the patient's crystalline lenses, we instead considered a diagnosis of spherophakia.

Spherophakia is a rare congenital abnormality in which the crystalline lens' unusually large anteroposterior diameter causes it to take on a spherical formation. Clues to the diagnosis of spherophakia include a shallow anterior chamber, ACG, and high lenticular myopia (diameters of 4.5 to 4.9 mm).⁶ Increased curvature of the lens is associated with weak, elongated zonules that can lead to ACG with pupillary block and the formation of peripheral anterior synechiae, subluxation of the lens into the anterior chamber, inflammation of the ciliary body, or progressive narrowing of the angle by the lens' anterior movement.^{7,8}

Microspherophakia (the presence of a spherical lens with a reduced equatorial diameter) is associated with systemic diseases such as Weill-Marchesani syndrome, Marfan syndrome, homocystinuria, Klinefelter syndrome, Alport syndrome, and Meyer-Schwickerath-Weyers syndrome.⁸⁻¹²

Investigators have hypothesized that spherophakia occurs when an incompletely developed ciliary body and its loose elongated zonules do not exert sufficient pressure to flatten the developing lens. The lenses of patients with spherophakia therefore retain a fetal spherical conformation.¹³

Pupillary block in spherophakia is exacerbated by treatment with miotic drugs, because the relaxation of the ciliary body allows the lens to move forward and obstruct the pupillary aperture. On the other hand, mydriatic agents can sometimes relieve pupillary block by increasing tension on the zonules and pulling the lens complex posteriorly.¹⁴

Surgeons can attempt to break an attack of ACG with laser peripheral iridotomy or lensectomy. The latter option may not be effective for eyes that have peripheral anterior synechiae or a damaged trabecular meshwork. Surgeons have advocated that patients who have periph-

eral anterior synechiae over more than 270° of their angle undergo goniosynechiolysis to help open the angle.¹⁵⁻¹⁷

If damage to the trabecular meshwork is advanced, the previously described surgical interventions may not prevent glaucomatous progression. In these cases, patients may require filtering surgery or the implantation of a tube to lower their IOP. These options present their own problems, however, because filtering surgery tends to flatten the chambers of eyes with ACG and may contribute to the development of malignant glaucoma.¹⁵ □

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