

Plateau Iris Syndrome Exacerbated by Lens/Angle Crowding in a Hyperopic Patient

BY GLORIA P. FLEMING, MD

CASE PRESENTATION

A 32-year-old white female presented to her community ER with an 8-week complaint of pain in her left eye, headache, and subsequent nausea. Her IOP measured 50 mm Hg OS, and the visual acuity in that eye was hand motions.

The patient was referred to an outside ophthalmologist, who diagnosed acute angle-closure glaucoma (ACG) and anterior uveitis. Her condition was managed with a topical steroid, a systemic steroid, a topical beta-blocker, systemic Diamox (acetazolamide; Duramed Pharmaceuticals, Inc, Pomona, NY), and Valtrex (GlaxoSmithKline, Research Triangle, NC), as it was initially thought that there was a viral etiology to the uveitis. Attempts to taper the medications caused a rebound IOP spike. The patient's past medical history was significant for non-insulin-dependent

diabetes mellitus of 1 year's duration, which was controlled by diet. She also had hypercholesterolemia and ill-defined psychosocial issues. Her systemic medication included alprazolam. The patient was referred to me for further evaluation and management.

She presented to me 2 days after her diagnosis with complaints of nausea, vomiting, diarrhea, and photophobia. Subjectively, she felt that her headache and vision had improved. Her BCVA was 20/30 OD and 20/150 OS. Her IOP measured 18 mm Hg OD and 9 mm Hg OS with Goldmann applanation tonometry. The patient's right pupil was round and reactive to light, whereas the left pupil was mid-dilated at 7 mm and minimally reactive. A slit-lamp examination of her left eye revealed ciliary flush with 1+ Descemet's folds. The anterior chamber was diffusely shallow with temporal iris atrophy and central



Figure 1. Disc photographs of the patient's right (A) and left eyes (B) revealed shallow physiologic cupping.

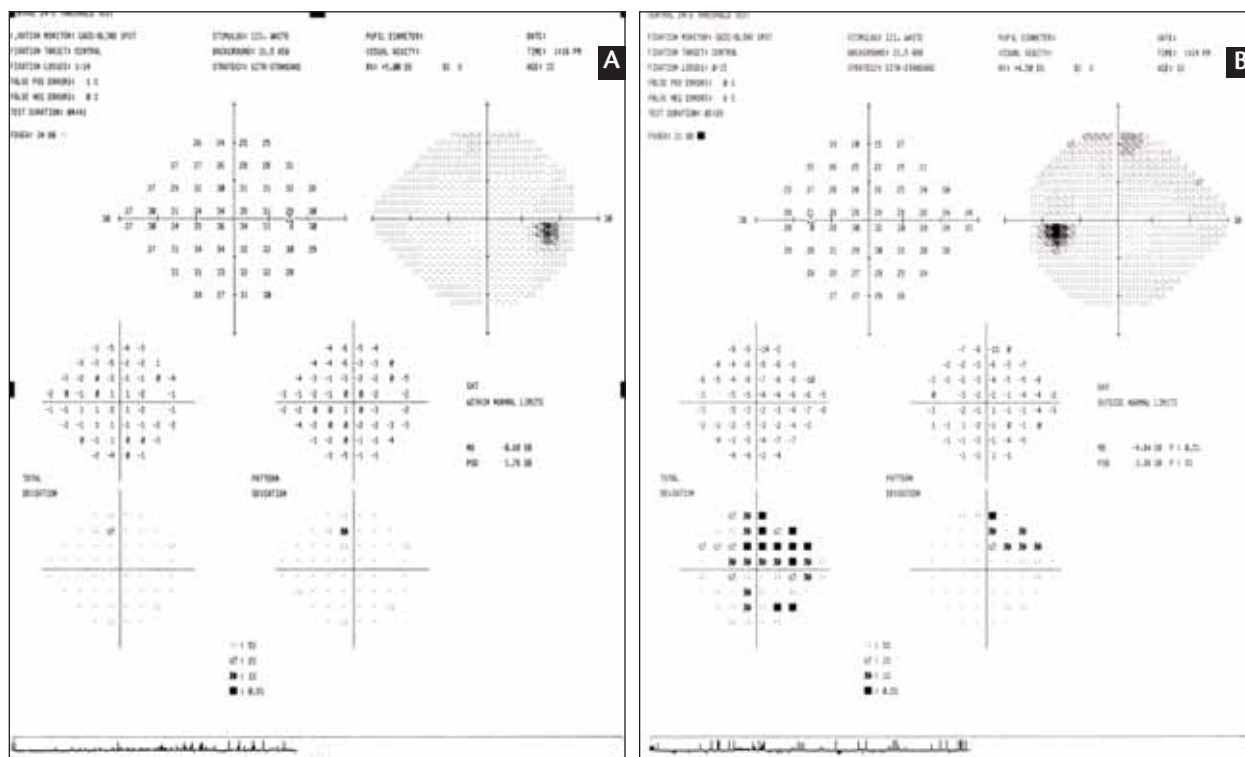


Figure 2. Humphrey visual field testing of the patient's right (A) and left eyes (B). First-time visual field testing shows superonasal depression in the left eye.

glaukomflecken of the lens. Gonioscopy revealed an appositionally closed angle of 270° in her right eye and no visible angle structures in her left eye despite gonio-compression. Funduscopy revealed physiological cupping of 0.25 bilaterally (Figure 1).

My working diagnosis was acute ACG. I discontinued the Valtrex, and the patient was scheduled for a prophylactic laser peripheral iridotomy (LPI) in her right eye and a trabeculectomy with mitomycin C in her left eye. Both procedures were uncomplicated. The angle in the right eye deepened to the level of midtrabecular meshwork following the LPI. After the trabeculectomy, her visual acuity improved to 20/40 OS, the IOP by applanation tonometry was 8 mm Hg, and the posterior bleb was diffuse. Once stable, she continued her follow-up care with her original ophthalmologist.

The patient was referred back to me 14 months later for progressive narrowing of the angle in her right eye, without an increase in IOP. Her BCVA ($+4.75$ -1.00 X 180) was 20/30 OD and ($+4.25$ -0.75 X 180) 20/50 OS. IOP by Goldmann applanation tonometry measured 18 mm Hg OD and 9 mm Hg OS. The iridotomy was patent by slit-lamp examination, but gonioscopy revealed appositional closure inferonasally, with 1 clock hour of peripheral anterior synechiae (PAS) nasally to anterior midtrabecular meshwork. Corneal horizontal diameters measured 10 mm OU.

The lens was clear and not intumescent in the patient's right eye. Corneal pachymetry measured 569 μ m OD and 567 μ m OS. A SITA-standard 24-2 Humphrey visual field test (Carl Zeiss Meditec, Inc., Dublin, CA) revealed a full visual field in her right eye and superonasal depression in her left eye (Figure 2).

HOW WOULD YOU PROCEED IN THE PATIENT'S RIGHT EYE?

1. Would you begin miotic therapy?
2. Would you perform a goniotomy?
3. Would you extract the lens and implant an IOL?

SURGICAL COURSE

I started the patient on low-dose pilocarpine 0.5% b.i.d. while arrangements were made for an iridoplasty. The brief course of pilocarpine therapy significantly relieved the iridotrabecular apposition as seen by gonioscopy, and subsequent ultrasound biomicroscopy (UBM) revealed an open angle with evidence of plateau iris configuration (Figure 3A). The thickness of the posterior sclera could not be assessed. Given the crowded anterior segment, I performed optical biometry using the IOLMaster (Carl Zeiss Meditec, Inc.), which measured an axial length of 19.79 mm OD and 19.61 mm OS and an anterior chamber depth of 2.05 mm OD and 2.16 mm OS (Table 1).

I performed an iridoplasty 360° and lysis of the small area of PAS. Postoperatively, she was started on prednisolone acetate 1% q.i.d.

OUTCOME

Three weeks postoperatively, the anterior chamber remained shallow, although the angle in the patient’s right eye remained open without pilocarpine. Repeat UBM imaging demonstrated a continued narrow but open angle (Figure 3B). Her IOP measured 12 mm Hg OD by applanation, and the anterior segment was quiet on biomicroscopy.

Because she was able to maintain her angle depth off of pilocarpine, I deferred lens extraction in favor of close observation.

DISCUSSION

As people age, the depth and volume of the anterior chamber decreases as the crystalline lens thickens and assumes a more anterior position.¹ The incidence of primary angle closure (PAC) therefore increases with age, reaching a peak between 55 and 70 years.² PAC rarely occurs in adults as young as this patient.

The major mechanism of PAC in older adults is pupillary block³ versus plateau iris in younger adults.² Other conditions such as iridociliary cysts, trauma, uveal effusion, and nanophthalmos should also be considered in younger patients.

In this case, gonioscopy and UBM ruled out relative pupillary block, trauma, iridociliary cysts, and uveal effusions. Nanophthalmos is characterized by a small eye, hyperopia, small corneal diameters, shallow anterior chambers,⁴ high lens/eye volume, and thickened sclera.⁵ The condition is quite rare and usually bilateral.⁴ Although the patient in this case possessed several of the aforementioned features

TABLE 1. SUMMARY OF OCULAR MEASUREMENTS		
	OD	OS
Central corneal thickness, μm	569	567
Axial length, mm	19.79	19.61
Anterior chamber depth, mm	2.05	2.16
Corneal diameter, horizontal, mm	10.00	10.00

of nanophthalmos, they are also typical risk factors seen in patients with PAC.³ Although the characteristic double-hump sign—as seen on gonioscopy—was absent, UBM confirmed anteriorly rotated iris processes. I made the diagnosis of plateau iris syndrome *incomplete*² based on continued appositional closure, despite the iridotomy’s confirmed patency without elevated IOP. A crowded anterior segment only exacerbates the shallow anterior chamber, which I suspect contributed to the initial presentation of acute angle closure in the patient’s left eye.

The options for surgical management include LPI, argon laser peripheral iridoplasty, trabeculectomy, and lens extraction. Psychosocial issues may have delayed this patient’s initial presentation to the ER. By the time of my evaluation, the angle of her left eye revealed synechial closure, without evidence of pupillary block. I felt a primary trabeculectomy with antimetabolites would afford her the best long-term IOP control. Although others have advocated argon laser peripheral iridoplasty in chronic angle closure, it has been reported that more than 36% of study patients required trabeculectomy by 3 months after the procedure.⁶ An alternative approach could have been initial argon laser peripheral iridoplasty followed by cataract extraction and

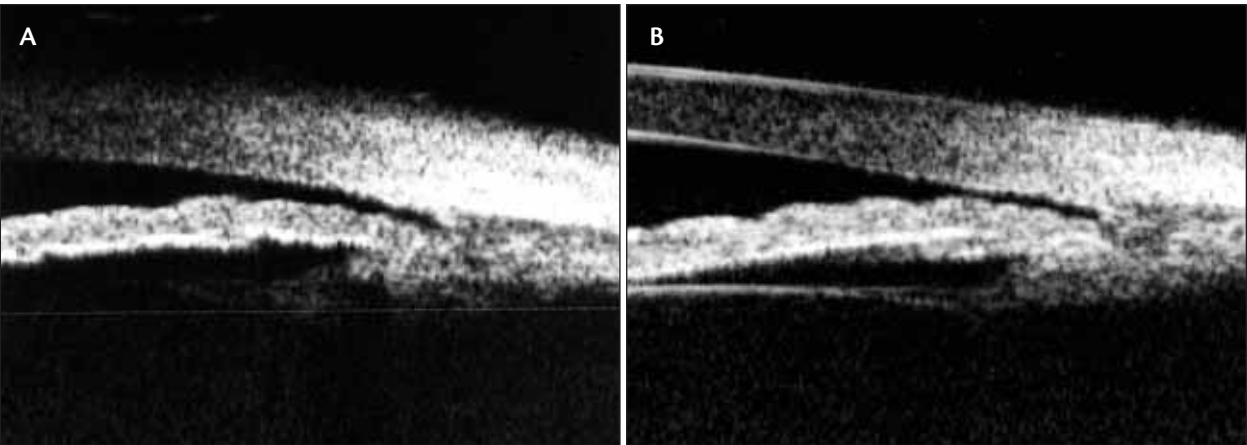


Figure 3. UBM of the right eye following pilocarpine use demonstrates deepening of the iridotrabeular angle, which was maintained after argon laser peripheral iridoplasty (A). Note the anteriorly rotated iris processes and loss of iridociliary sulcus (B).

ANGLE-CLOSURE GLAUCOMA IN YOUNG PATIENTS

By Steven D. Vold, MD

Angle-closure glaucoma (ACG) in young patients presents unique diagnostic and managerial challenges. Controversy continues to exist in regard to how best to approach these patients. Dr. Fleming presents an instructive case summary illustrating potential diagnostic difficulties and the rather wide range of managerial options available to us.

NEW DIAGNOSTIC CONCEPTS

In recent years, our understanding of ACG has been challenged. Iris thickness, choroidal expansion, and vitreous conductivity have been implicated as potential etiologic factors of ACG.¹ Classically, the term *plateau iris syndrome* inferred a specific iris configuration, increased postiridotomy IOP after dilation, and myopic refractions. Recent imaging advances have made the identification of anterior rotation of the ciliary processes much more commonplace and ultimately have affected our management of ACG patients.

OPTIONS FOR MANAGEMENT

Medication

In patients with ACG, topical, oral, or intravenous glaucoma medications and topical steroids may be useful in immediately lowering IOP and quieting inflamed eyes. These methods are often considered temporizing measures prior to more definitive surgical treatment. In eyes with spherophakia-induced angle closure, cycloplegic agents may be helpful.

Laser Iridotomy and Iridoplasty

Laser peripheral iridotomy (LPI) remains the most common first-line treatment for ACG and plateau iris configuration. If LPI does not successfully open the anterior chamber angle or the IOP rises significantly after post-LPI dilation, argon laser peripheral iridoplasty may be considered.

Lens-Based Surgery

Cataract surgery is an extremely effective method of managing many cases of ACG. In patients with pupillary block, this surgery may be completely curative. In patients with a mild plateau iris configuration as well, lens removal alone may be enough to eliminate angle closure. In patients with anterior synechial formation of short duration, combining cataract surgery with goniosynechialysis may also be beneficial. In patients with prominent anterior ciliary processes, endoscopic cyclophotocoagulation may

be effective in retracting the ciliary processes posteriorly and opening the angle. In younger patients, loss of accommodation and placement of a presbyopia-correcting IOL should be considered. In nanophthalmic eyes, a limited pars plana vitrectomy may be necessary to successfully complete lens removal and placement of a posterior chamber IOL.

Filtration Surgery

Trabeculectomy with an antifibrotic agent is certainly reasonable in patients with substantial synechial angle closure. Surgeons must be wary of the increased risk of ciliary block postoperatively. To prevent this potentially serious complication, tight scleral flap sutures are generally advocated. Combining trabeculectomy with cataract surgery is often beneficial to these patients as well. Combined surgery may reduce the incidence of ciliary block or at the very least make treatment of this condition with a YAG laser possible. In nanophthalmic eyes, some surgeons advocate the placement of scleral windows to prevent postoperative choroidal effusions. Although devices such as tube shunts may be considered, caution is encouraged, as they may potentially be associated with corneal decompensation in eyes with more shallow anterior chambers.

OTHER CONSIDERATIONS

In young patients with angle closure, uncontrolled diabetes mellitus must be considered as an etiology of lens swelling-induced pupillary block. Oral medications such as Topamax (Ortho-McNeil-Janssen Pharmaceuticals, Inc., Titusville, NJ) and other sulfa-based drugs may induce uveal effusions that may lead to ACG. In these cases, discontinuation of the drug and short-term hypotensive glaucoma medication may be all that is required for successful treatment. Iridociliary cysts, retinopathy of prematurity, uveitis, Weill-Marchesani syndrome, Marfan's syndrome, lens subluxation, persistent fetal vasculature, and nanophthalmos with uveal effusion syndrome must also be considered.

Steven D. Vold, MD, is the chief executive officer and a glaucoma and cataract surgery consultant at Boozman-Hof Regional Eye Clinic, PA, in Rogers, Arkansas. He acknowledged no financial interest in the product or company mentioned herein. Dr. Vold may be reached at (479) 246-1700; svold@cox.net.

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goniosynechialysis for residual PAS, as described by Ritch et al.⁷

Kobayashi et al demonstrated by UBM that pilocarpine widens the iridotrabecular angle in patients with narrow angles⁸ and may have a temporary role in managing these patients. In this case, pilocarpine was used until argon laser peripheral iridoplasty could be performed. During this time, UBM confirmed deepening of the angle.

The role of the crystalline lens in acute ACG has been described. Tarongoy et al explained that, despite a patent LPI, lens/angle crowding as seen in plateau iris syndrome can result in acute or chronic angle closure, which necessitates cataract extraction for definitive management.¹ Certainly, over time, this procedure may become necessary in this case. Despite cataract extraction and IOL implantation, however, iridociliary apposition is not eliminated in these eyes, as described by Tran et al.⁹

Regardless of one's surgical approach, the ultimate goal is to prevent trabecular dysfunction from chronic angle closure and the development of glaucomatous optic neuropathy. A well-planned, systematic approach can be achieved with close observation of the patient, including compression gonioscopy, which is essential in monitoring the development of any new PAS. □

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Gloria P. Fleming, MD, is an assistant professor of ophthalmology at The Ohio State University in Columbus, Ohio. She acknowledged no financial interest in the products or companies mentioned herein. Dr. Fleming may be reached at (614) 293-8119; gloria.fleming@osumc.edu.



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