

Controlling IOP in Pigmentary Glaucoma

BY LOAN NGUYEN, MD, AND JOHN DANIAS, MD, PhD

CASE PRESENTATION

A 36-year-old white male presented to our clinic to inquire about alternative options for managing his glaucoma. He had been diagnosed with pigmentary glaucoma 1 year earlier and was placed on therapy with latanoprost ophthalmic solution 0.005% (Xalatan; Pfizer Inc., New York, NY) q.h.s. OS. Prior to initiation of therapy, the IOP in his left eye was in the low 30s.

His past medical history was noncontributory, a review of his systems was negative, and he had no family history of glaucoma. The patient denied sustaining any trauma to his left eye.

The patient's BCVA measured 20/20+1 OD and 20/20-1 OS with a correction of -1.00 D sphere OU. He had full motility, and there was no evidence of an afferent pupillary defect. Slit-lamp examination was significant for the presence of Krukenberg's spindles (pigment arranged in a vertical spindle pattern) bilaterally that were more prominent in his left eye. Examination revealed lightly pigmented irides, midperipheral transillumination defects, and a rupture of the iris sphincter inferiorly in his left eye.

On Goldmann applanation tonometry, the patient's IOPs measured 18 mm Hg OD and 11 mm Hg OS. His pachymetry readings were 533 μ m OD and 535 μ m OS. Gonioscopy with a four-mirror gonioprism revealed open angles (Shaffer grade 4) that appeared to be slightly deeper in his left versus his right eye. We could not identify any areas of recession (Figure 1). We noted significant pigmentary deposits in the trabecular meshwork bilaterally that were greater in his left eye.

An examination of the optic nerve heads with 78.00-D stereobiomicroscopy showed central cupping with cup-to-disc ratios of 0.5 OD and 0.6 OS. The left cup was more vertically elongated than the right, and the rims of both discs were intact without detectable nerve fiber layer loss. The rest of the fundus examination revealed normal blood vessels and maculae as well as no peripheral abnormalities.

A SITA-Standard 24-2 Humphrey visual field test (Carl

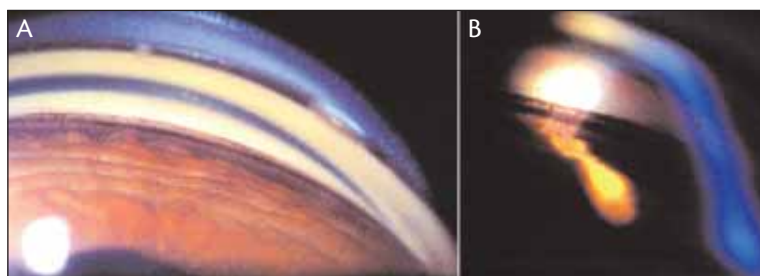


Figure 1. On gonioscopy, the angle of the patient's left eye was open to Shaffer grade 4 (A). Note the significant pigmentation of the trabecular meshwork (B).

Zeiss Meditec, Inc., Dublin, CA) showed full visual fields bilaterally (Figure 2). There was no evidence of thinning of the retinal nerve fiber layer in either eye with confocal scanning laser ophthalmoscopy (HRT II; Heidelberg Engineering GmbH, Heidelberg, Germany) or optical coherence tomography (Stratus OCT 3; Carl Zeiss Meditec, Inc.).

HOW WOULD YOU PROCEED?

1. Would you solicit additional information from the patient?
2. Would you perform additional diagnostic tests such as short wavelength automated perimetry (SWAP; Carl Zeiss Meditec Inc.), visual fields, ultrasound biomicroscopy (UBM), or anterior segment OCT?
3. What therapeutic options would you consider?

CLINICAL COURSE

Although the patient's high IOP and the presence of pigment in the anterior segments of both eyes were consistent with pigmentary glaucoma, the pupillary rupture in his left eye suggested a traumatic event. Despite repeated questioning, the patient could not recall any episodes that might have caused this damage. Furthermore, he reported that he did not exercise and described a mostly sedentary lifestyle.

The patient's normal visual fields, optic nerves, and retinal nerve fiber layer thickness suggested he had sustained minimal damage to his left eye due to high IOP.

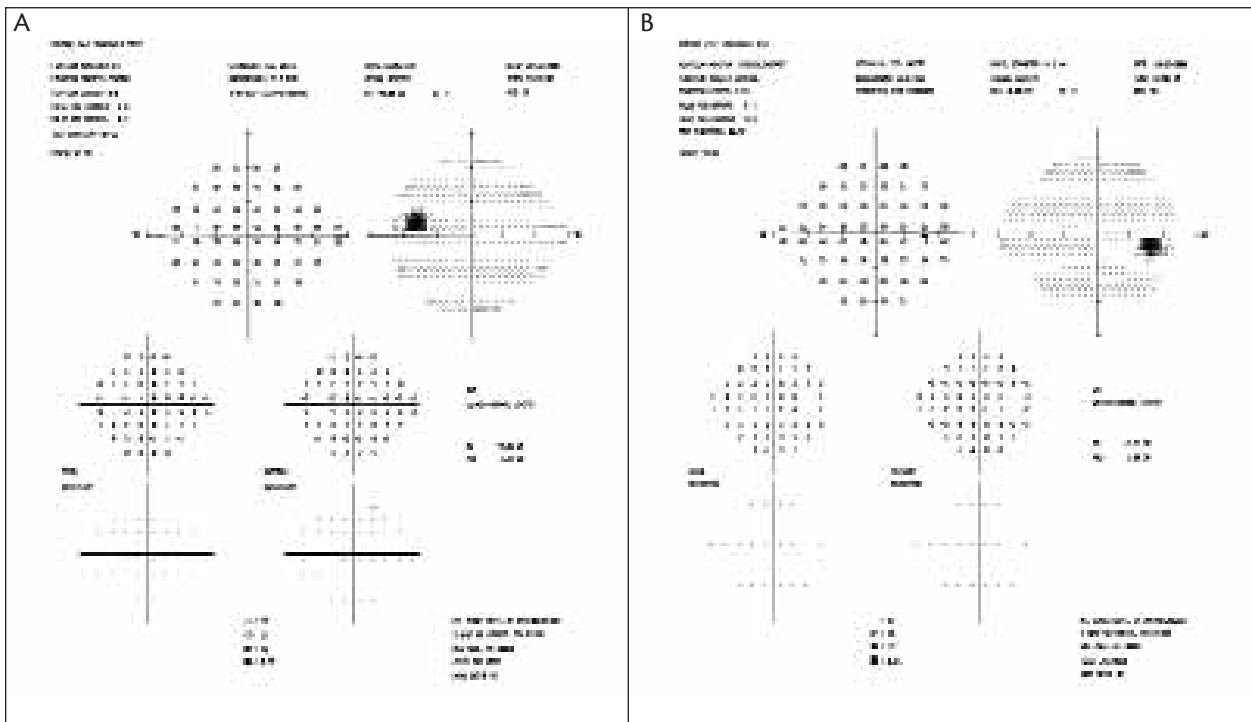


Figure 2. The patient had full visual fields in both eyes (A and B) on SITA-Standard 24-2 Humphrey visual field testing.

Although prostaglandin analogs may not be the ideal first-line therapy for a young patient with lightly colored irides who requires unocular therapy, he elected to continue using latanoprost ophthalmic solution 0.005% because of its IOP-lowering efficacy rather than changing his ocular regimen. We advised the patient to undergo regular visual field testing with short wavelength automated perimetry and to avoid vigorous exercise that could liberate pigment from his irides and cause his IOPs to spike.¹ We considered evaluating the patient with UBM but decided that the concavity of the iris in his left eye was not excessive enough to warrant this examination.

OUTCOME

To date, the patient's optic nerves have remained unchanged, and his visual fields are full. His IOP continues to be in the mid-to-low teens in his left eye, and he is tolerating the therapy well.

DISCUSSION

Pigmentary dispersion syndrome is characterized by increased pigmentation of the trabecular meshwork, Krukenberg's spindles on the corneal endothelium, transillumination defects of the midperipheral iris, and the deposition of pigment on the posterior lens capsule (Zentmeyer's line).²

Patients with pigmentary dispersion syndrome have a 25% to 50% risk of developing pigmentary glaucoma. This form of the disease—which typically affects myopes between 20 and 40 years of age^{3,4} and occurs more commonly in men than women (ratio, 3:1)—develops when iridal pigment becomes trapped in the trabecular meshwork and disrupts the physiological outflow of aqueous fluid from the anterior chamber. Slit lamp and gonioscopic examination of affected eyes usually shows posterior bowing of the midperipheral iris that creates iridozonular touch.

The goal of treatment for pigmentary glaucoma is the same as for any other variation of the disease: to reduce the IOP and prevent damage to the optic nerve. Although ocular hypotensive agents such as beta blockers, carbonic anhydrase inhibitors, alpha adrenergic agonists, and prostaglandin analogs all effectively reduce IOP in pigmentary glaucoma, it has been proposed that the long-term use of prostaglandin analogs may exacerbate this condition by increasing iridal pigmentation.⁵ Miotic drugs such as pilocarpine can stretch the iris, thus decreasing its posterior bowing and reducing the amount of contact between the iris and the zonules (Figure 3A and B). This effect can often be dramatic and reduce the liberation of pigment, but the use of this class of drugs is limited, because these agents can worsen myopia and increase the risk of retinal detachment in the young

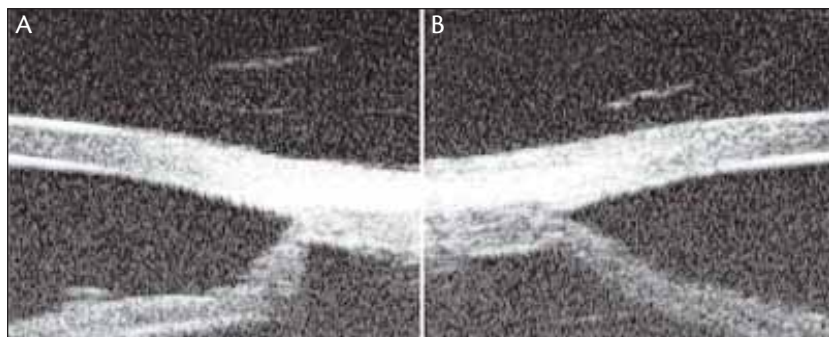


Figure 3. UBM of an angle before (A) and 30 minutes after (B) the instillation of pilocarpine showed a significant decrease in the posterior bowing of the iris after the drug's application.

myopic population.² Young patients are also more symptomatic of the brow ache caused by pilocarpine. Long-term pilocarpine use may additionally induce the formation of posterior synechiae.

Potential laser-based treatments for pigmentary glaucoma include argon laser trabeculoplasty and laser peripheral iridotomy (LPI). One study showed that argon laser trabeculoplasty lowered IOP similarly in patients with pigmentary or primary open-angle glaucoma and that the change in IOP was inversely related to the age of the patient.⁶ Selective laser trabeculoplasty (SLT) on the other hand may be relatively contraindicated in eyes with pigmentary glaucoma, because the trabecular meshwork is heavily pigmented, increasing the risk of significant post-SLT elevations in IOP.⁷

LPI reportedly reduces IOP in pigmentary glaucoma by eliminating reverse pupillary block in the presence of a posteriorly bowed iris.⁸ It is presumed that a defect in the iris causes its midperipheral portion to bow posteriorly and hug the lens' anterior surface, bringing it in apposition to the anterior lens zonules. As the eye blinks, it forces aqueous humor into the anterior chamber, thereby causing a reverse pupillary block that further presses the iris onto the lens' surface and zonules. Contact between the iris and the zonules creates friction that liberates pigment from the iris.

The irides of eyes with pigmentary dispersion syndrome appear to be flatter and less bowed on ultrasound biomicroscopy after Nd:YAG iridotomy.⁹ Studies have not shown, however, that LPI improves the outcomes in pigmentary glaucoma,¹⁰ because the procedure does not affect pigment that is already deposited in the trabecular meshwork. Nonetheless, LPI may make it easier to control the acute elevations in IOP that are often observed with pigmentary glaucoma.¹¹

Incisional glaucoma surgery is another alternative for lowering IOP in patients with pigmentary glaucoma, but

it is usually more successful for treating primary open-angle glaucoma. This is because patients with pigmentary glaucoma are usually younger and thus tend to have a more intense fibrovascular response to penetrating procedures than those with primary open-angle glaucoma. In addition, postoperative hypotony is more common with pigmentary glaucoma, and high myopes have a greater chance of maculopathy with hypotony compared to a similar patient without myopia.¹²

Given the complications that patients with pigmentary glaucoma could develop after trabeculectomy, it would be prudent to treat them with medical or laser therapy before progressing to more aggressive surgical procedures. □

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