WHAT KIND OF PIGMENTARY **GLAUCOMA IS THIS?**

Surgeons discuss a diagnosis and the related treatment options.

BY WILLIAM H. BRAY, MD; JACOB W. BRUBAKER, MD; LEON W. HERNDON, MD; INDER PAUL SINGH, MD; AND STEVEN R. SARKISIAN JR, MD

CASE PRESENTATION

A 50-year-old white man with an IOP of 45 mm Hg on maximum tolerated medical therapy in his right eye was referred for unilateral glaucoma. The patient had undergone routine bilateral phacoemulsification cataract surgery and IOL implantation 3 months earlier. He reported that glaucoma had been diagnosed in his right eye but not his left eye just before cataract surgery. He stated that the IOP in the right eye had measured in the mid-20s mm Hg at that time and that the surgeon expected phacoemulsification would be adequate treatment. No notes or test results were available from the operating surgeon.

On examination, the patient had a BCVA of 20/60 OD and 20/20 OS. Humphrey visual field testing (Carl Zeiss Meditec) showed generalized depression in the right eye but was essentially normal in the left eye (Figures 1 and 2). The patient had cup-to-disc ratios of 0.85 OD and 0.4 OS. In each eye, gonioscopy showed a

deep, posteriorly bowed iris. Both eyes were symmetrically deep and had a visible ciliary body band. There was 4+ pigment of the trabecular meshwork in the right eye and 3+ in the left eye. The right eye exhibited no obvious transillumination defects, but 3+ pigmented cells were evident in the anterior chamber. Dense pigment was visible on the corneal endothelium of the right eye but not the left. Diffuse pigment was also present behind the IOL in the right eye. The pupil did not dilate enough to allow visualization of the haptics or the edge of the IOL optic in the right eye.

The patient expressed a desire to use as few medications as possible after further treatment. How would you proceed?

> -Case prepared by Steven R. Sarkisian Jr, MD

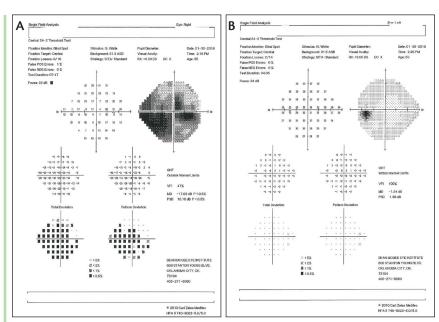


Figure 1. Visual field testing revealed generalized depression affecting fixation in the right eye (A), whereas results in the left eye were normal (B).

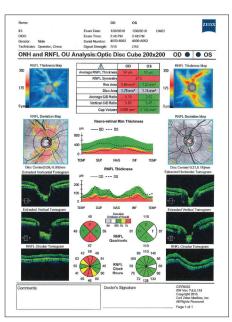


Figure 2. Corresponding profound thinning was evident on an OCT scan of the right eye.



WILLIAM H. BRAY, MD

Based on the case presentation, there is obvious pigment dispersion, with likely IOL-iris contact in the right eye that could be confirmed by ultrasound biomicroscopy (UBM). The question is why the pigment dispersion is so much worse after surgery than it was before.

One possibility is that there is reverse pupillary block on the right. This problem has been reported with sulcus-fixated IOLs, but it may also occur in the eyes of patients with a history of pigment dispersion. The second possibility is that the IOL is malpositioned, causing pigment dispersion as well as inflammation (uveitis-glaucoma-hyphema [UGH] syndrome). Because the visual acuity in the patient's right eye has decreased, an optical coherence tomography scan could help determine the cause. If cystoid macular edema is present, UGH syndrome is the likely culprit.

I would start by performing a laser peripheral iridotomy (LPI) to relieve any reverse pupillary block. If that procedure failed to alleviate the elevated IOP and pigment dispersion, then the IOL would have to be dealt with. In the OR, I would inspect the IOL to see if any element is out of the bag. If so, I would reposition the lens. If the IOL is in the bag, I would exchange it for a three-piece IOL with thinner haptics.

An IOP of 45 mm Hg must be addressed with surgery as well. I have no experience with the Xen45 (Allergan), so I would probably perform a filtering procedure instead. I doubt that microinvasive glaucoma surgery (MIGS) would be adequate to control the IOP.



JACOB W. BRUBAKER MD

Clearly, this patient urgently needs surgical treatment for his elevated IOP and progressive glaucoma. The presence of a deeply pigmented angle and freely liberated pigment suggests either primary or secondary pigmentary dispersion. The bilateral posterior bowing of the iris is typical in primary pigmentary dispersion prior to phacoemulsification. This configuration can exist after proper placement of an IOL in the bag because of reverse pupillary block.^{1,2} Treatment with an LPI should be the first step to address this potential cause. Alternatively, the pigmentary dispersion could be related to a secondary lens chafing syndrome. I would try to obtain records from the operating surgeon to determine if the IOL was placed in the sulcus or

Assuming the LPI proved unsuccessful, the next step would be to discuss with the patient all the diagnostic possibilities and potential procedures. He must be made aware of the challenges involved in his case. It will be necessary to evaluate the IOL through the placement of iris hooks and the use of endoscopy to determine its location. In the event that the IOL is poorly positioned, the surgeon will have to be prepared to perform optic capture with a three-piece lens, if it is in the sulcus, or to perform a complete lens exchange.

With regard to the elevated IOP, I have had great success with goniotomy, either with the Kahook Dual Blade (New World Medical) or via gonioscopy-assisted transluminal trabeculotomy. This procedure can often greatly reduce or even eliminate a patient's need for topical glaucoma therapy in cases of pigmentary

dispersion. It would be important to counsel this patient that, if goniotomy is unsuccessful, he may subsequently require a trabecular bypass procedure.



LEON W. HERNDON, MD

This patient was recently diagnosed with glaucoma in his right eye that was likely exacerbated by the release of pigment in the anterior chamber. Although this is not a classic presentation for pigment dispersion glaucoma, the patient does have some features (male sex, posteriorly bowed iris in the left eye, pigment accumulation in the trabecular meshwork of both eyes, diffuse pigment behind the IOL in the right eye) that are consistent with this diagnosis.

I would want to know more about the IOL. What type was used, and was it placed in the ciliary sulcus, creating an UGH-type of presentation due to iris chafing by the IOL? Although pigmented cells can be seen in the anterior chamber with pigmentary glaucoma, I would be more concerned about the contribution of the IOL to the exacerbation of this patient's glaucoma. Also, it would be necessary to rule out iris melanoma as the cause of his unilateral glaucoma. A UBM examination of the iris and ciliary body is indicated to assess the IOL's position and to rule out the presence of a ciliary body melanoma with anterior extension.

The loss of central visual acuity in the patient's right eye must be explored further as well. It would be unusual for the pigmented cells in the anterior chamber and the pigment behind the IOL to affect visual acuity to this degree, especially because there

is no mention of poor visualization to the fundus.

At this point, the patient's IOP is much too high for the health of the optic nerve, and surgical measures must be considered to manage his moderate to severe glaucoma. The fact that the disease was diagnosed prior to the cataract surgery leads me to believe that this is a case of pigmentary rather than IOL-related glaucoma. If further evaluation shows that the IOL is indeed well positioned in the capsular bag, I would offer the patient surgical options directed toward the management of pigmentary glaucoma. Because he has an active pigment storm, LPI could be considered to equalize the pressures in the anterior and posterior chambers and to pull the iris away from the zonules, but this approach might be less effective in a pseudophakic eye.

Angle-based surgery such as with the Trabectome (NeoMedix) or Kahook Dual Blade, goniotomy/trabeculotomy using the Trab360 (Sight Sciences), or ab interno canaloplasty using the iTrack 250 (Ellex) would all be options for this patient. Should one of these MIGS options fail to lower the IOP to a reasonable range (midteens), then trabeculectomy surgery would be considered.



INDER PAUL SINGH, MD

This case is a great example of the volatility of pigmentary glaucoma. The patient exhibits many of the characteristics of the condition: He is a young man with asymmetric damage, pigment on the corneal endothelium, heavy pigmentation of the trabecular meshwork, and a spike in IOP. Interestingly, patients such as this often exhibit decreased pigmentary dispersion as time passes.

Of concern here are the pigment in the anterior chamber and the sudden rise in IOP after cataract surgery. These findings suggest an active release of pigment, possibly from the continued concavity of the iris rubbing against the zonules or, more likely, because the haptics of the IOL are rubbing against the iris. Anterior segment optical coherence tomography, UBM, or swept-source biometry (IOLMaster 700; Carl Zeiss Meditec) would provide valuable information on the IOL's position in this eye with a small pupil.

If the problem relates to the IOL haptics, the lens should be surgically repositioned or replaced. In an eye with a posteriorly bowed iris, one could perform an LPI or iridoplasty to attempt to flatten the iris and help decrease apposition of this tissue and the haptics. Unfortunately, these procedures will not be effective if the IOL is the main culprit, and the IOP will likely remain elevated because a significant amount of pigment is already blocking outflow. Laser trabeculoplasty probably would not lower the IOP sufficiently in the patient's right eye, but the procedure would be a good option for his left eye, although it would be important to monitor him thereafter for IOP spikes. Because the patient is pseudophakic, the use of a trabecular bypass stent or supraciliary stent would be off-label.

If the patient understands that he will likely continue to require topical glaucoma therapy and may require additional surgery in the future, one could address the IOL in combination with a MIGS procedure. Options include viscodilation; a gonioscopyassisted transluminal trabeculotomy using the Visco360 (Sight Sciences) or Trab360 or ab interno canaloplasty using the iTrack 250; and/or removal of the trabecular meshwork tissue with the Kahook Dual Blade or Trabectome. These procedures probably would not achieve the target IOP without topical

glaucoma therapy, but they might decrease the patient's medication burden. A subconjunctival filtering procedure, such as with the Xen45, might get the patient off most medications, and the device would offer a reasonable balance of safety and efficacy if the surgeon chose to bypass internal mechanisms. A traditional trabeculectomy and tube shunt surgery would also be options, but, considering the many MIGS procedures available, I would offer traditional filtration surgery as a last resort in this case.



WHAT I DID: STEVEN R. SARKISIAN JR, MD

This case was interesting because the patient had both primary and secondary pigmentary glaucoma in his right eye. His left eye had pigment dispersion syndrome. The active release of pigment and acute glaucoma were caused by the haptics of a one-piece acrylic IOL that were protruding through the capsular bag. The IOL was completely in the bag, but it was a large plate-haptic lens that was obviously touching the peripheral iris. The status of the IOL was unclear until the patient was in the OR.

An IOL exchange was my top priority. In addition, however, I felt that removing the dense pigment occluding the trabecular meshwork would help prevent further glaucomatous damage after the IOL's removal because the patient had been diagnosed with the disease prior to cataract surgery and his outflow system was already compromised.

I exchanged the IOL for a threepiece acrylic lens placed in the capsular bag without complication. I

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combined the procedure with a 360° ab interno goniotomy/trabeculotomy using the Trab360 device. One month after surgery, the unmedicated IOP measured between 7 and 9 mm Hg, and his visual acuity was 20/60. ■

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STEVEN R. SARKISIAN JR, MD | SECTION EDITOR

- Clinical Professor and Glaucoma Fellowship Director, Dean McGee Eye Institute, University of Oklahoma, Oklahoma City
- Member, Glaucoma Today Editorial Board
- steven-sarkisian@dmei.org
- Financial disclosure: Consultant (Alcon, Allergan, BVI, Katena, Glaukos, New World Medical, Santen, Sight Sciences); Equity owner (Sight Sciences); Grant support (Alcon, Glaukos, Sight Sciences)

WILLIAM H. BRAY, MD

- Assistant Clinical Professor of Ophthalmology, University of Kansas, Kansas City, Kansas
- Kansas City VA Medical Center, Missouri
- drwhbray@gmail.com
- Financial disclosure: None

JACOB W. BRUBAKER MD

- Glaucoma and anterior segment surgery, Sacramento Eye Consultants, Sacramento, California
- jbrubaker@saceye.com
- Financial disclosure: Consultant (Allergan, New World Medical); Research funding (Allergan, Glaukos, Ivantis); Speakers' bureau (Alcon, Allergan, New World Medical)

LEON W. HERNDON, MD

- Professor of Ophthalmology, Duke University Eye Center, Durham, North Carolina
- leon.herndon@duke.edu
- Financial disclosure: Advisory board (Glaukos, Sight Sciences); Research support (Glaukos)

INDER PAUL SINGH, MD

- The Eye Centers of Racine and Kenosha, Wisconsin
- Member, Glaucoma Today Editorial Board
- ipsingh@amazingeye.com
- Financial disclosure: Consultant (Alcon, Allergan, Ellex, Glaukos); Research funding (Alcon, Allergan, Glaukos, Ellex, Ivantis, New World Medical); Speakers' bureau (Alcon, Allergan, Ellex, Glaukos)