SMALL EYE, BIG PRESSURE

Abnormal corneas render clinical decision-making more complex.

BY DEVESH K. VARMA, MD, FRCSC; LAUREN S. BLIEDEN, MD; GEORGES M. DURR, MD, FRCSC; AND H. GEORGE TANAKA, MD

CASE PRESENTATION

A 27-year-old man with microcornea and iris colobomas (Figure 1) was referred for elevated IOP, 34 mm Hg OD and 35 mm Hg OS without medication. Central corneal thickness was 634 µm in the right eye and 689 µm in the left eye. Testing with the Ocular Response Analyzer (Reichert) showed elevated corneal hysteresis bilaterally, 11.1 mm Hg in the right eye and 12.7 mm Hg in the left eye (Figure 2). Figures 3 to 6 show the results of diagnostic testing.

The horizontal white-to-white measurement for each eye was 8.5 mm. The average keratometry reading in each eye was 49.00 D. Anterior chamber depth measured with ultrasound was 2.28 mm in the right eye and 2.34 mm in the left eye. Axial length was 21.93 mm in the right eye and 21.75 mm in the left eye. The angle in each eye appeared to be anomalous but was open to the ciliary body, as was confirmed with ultrasound biomicroscopy (Figure 7). Trace rosette cataracts were evident, and BCVA was -2.25 +0.75 x 43° = 20/25 OD and -2.25 D = 20/30 OS.

How would you manage this patient? If you would treat him, what approach would you recommend? If not, what would be your threshold for treatment?

—Case prepared by Devesh K. Varma, MD, FRCSC

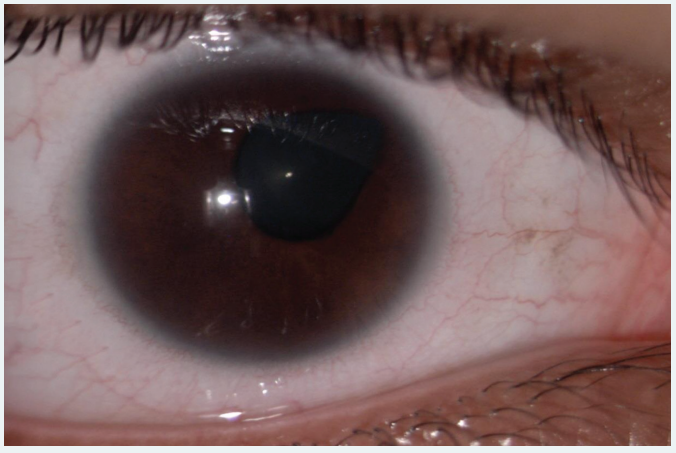

Figure 1. Microcornea and iris coloboma are evident at the slit lamp.

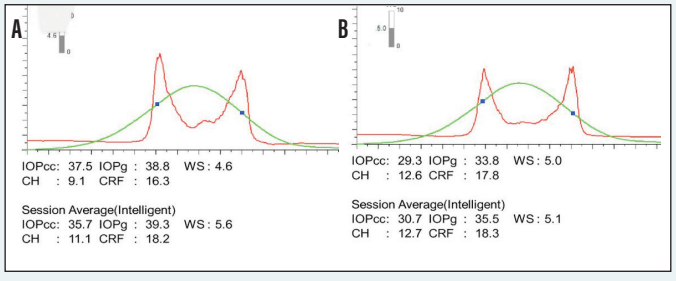

Figure 2. Corneal hysteresis measurements for the right (A) and left (B) eyes.

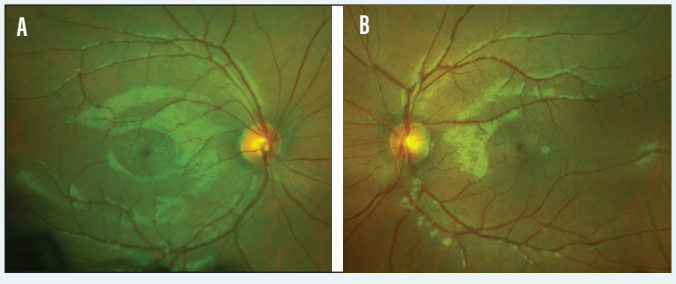

Figure 3. Optic nerves of the right (A) and left (B) eyes.

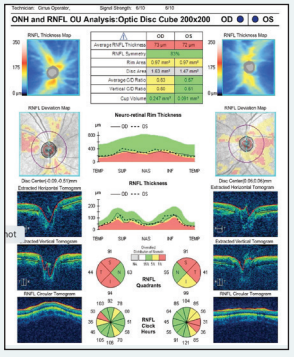

Figure 4. Analysis of the optic nerve head and retinal nerve fiber layer in each eye.

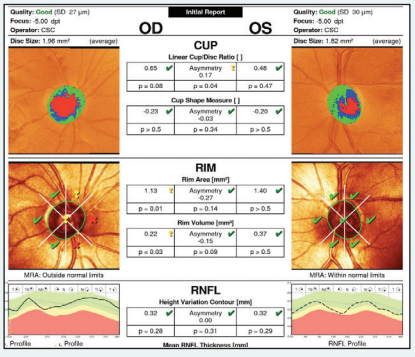

Figure 5. Confocal scanning laser ophthalmoscopy scan of right and left eyes.

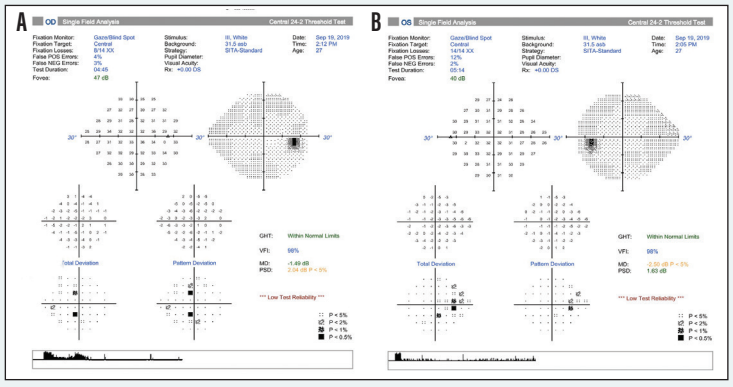

Figure 6. Visual field analysis of the right (A) and left (B) eyes.

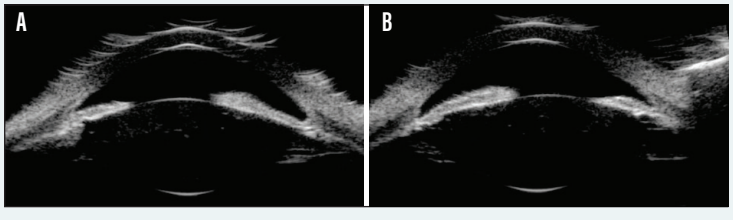

Figure 7. Ultrasound biomicroscopy of the right (A) and left (B) eyes.

LAUREN S. BLIEDEN, MD

The abnormal corneal properties make IOP interpretation difficult, although I suspect that the IOP is too high. Given the lower signal strength on OCT, the borderline results are more difficult to interpret. The fair-quality parameters on confocal scanning laser ophthalmoscopy ([CSLO] Heidelberg Retina Tomograph, Heidelberg) are reassuring. The results of visual field testing are unreliable here, but they also raise some concern about possible early arcuate defects superiorly and inferiorly in the right eye and a nasal step in the left eye. What to do?

This young patient has relatively healthy optic nerves but also high-risk features for developing glaucoma (ie, high IOP, anterior segment dysgenesis, and crowded anterior chamber anatomy). Regardless of whether treatment is initiated, this patient must be monitored closely. I would repeat visual field testing and OCT in hopes of obtaining better-quality results. Stability is the goal here. If test results at subsequent visits remain stable, then intervention is not required. If repeat testing shows glaucomatous changes, then, based on my experience with pediatric glaucoma patients, I would consider performing an angle-based procedure, either goniotomy or trabeculotomy. Given the patient's age and ability to cooperate, topical medications combined with close supervision and a very low threshold for surgery may be a more appealing approach.

I would include regular gonioscopy as part of follow-up. Given the crowded configuration of the anterior segment, if he exhibits features of chronic angle closure, then I would discuss with him early lens extraction or temporizing iridotomy.

GEORGES M. DURR, MD, FRCSC

This case highlights several difficult considerations in managing a young patient who has possible glaucoma, microcornea, and open angles. Is treatment warranted? Although IOP is elevated, the corneas are thick. More important, the corneal hysteresis readings are reassuring. Nevertheless, OCT imaging shows possible blunting of the inferior peaks of the retinal nerve fiber layer (RNFL) to a greater degree in the left than right eye. Visual field testing may show an early superior nasal step in the left eye and shows nonspecific changes in the right eye.

The likelihood of glaucomatous damage, greater in the left than the right eye, suggests that this patient probably requires treatment or close monitoring. The next step is a thorough discussion with him of the findings and of the options for treatment, which are topical therapy, laser therapy, or surgery.

Topical therapy, either with a prostaglandin analogue or a fixed-combination drop, may be the first choice, but compliance is always an issue in glaucoma management, particularly with young patients.[1] Selective laser trabeculoplasty (SLT) is an alternative despite the anomalous angle structures. Microcornea may render SLT less effective, but, given the procedure's good safety profile, it is an option to consider.[2] If topical and/or laser therapy does not achieve the target IOP, surgery should be discussed with the patient earlier rather than later. Considering the anomalous angle, good visual acuity, and minimal cataracts, either a 180° or 360° standalone goniotomy procedure would be my first choice. Thorough informed consent would be required, including a discussion of the risks of hyphema and further cataract progression. Goniotomy can be effective for patients such as this one, and it can eliminate the need for a bleb-based procedure.[3]

H. GEORGE TANAKA, MD

There is no convincing structural or functional evidence of glaucoma: The optic nerves exhibit unremarkable cupping and good symmetry between the superior and inferior rims without focal thinning or RNFL defects. The OCT scan shows mild diffuse depression of the RNFL with suboptimal signal strength (6/10); it should be repeated, preferably after pupillary dilation. Visual field testing demonstrates nonspecific defects, and the results of further testing are likely to remain unremarkable with improved patient fixation. CSLO has largely been supplanted by OCT for glaucoma diagnosis. The inclusion of the CSLO scan is diagnostically irrelevant but also a reminder that technology progresses faster than glaucomatous damage to most optic nerves. With further advances in computerized optic nerve imaging, disc photographs will likely become the only valuable reference points for this patient in the years to come.

Clinical IOP measurements are indirect estimates that are affected by the biophysical properties of the human cornea. Short of performing manometry in this patient with markedly small, thick, and steep corneas, "true" IOP is an elusive value that should not dominate decision-making.

Considering the elevated corneal hysteresis, I would monitor this patient without prescribing IOP-lowering

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—H. GEORGE TANAKA, MD

treatment until reproducible ganglion cell loss occurs. I would stress to him the importance of regular checkups. Because the angles are open, initial SLT and/or topical monotherapy would be my first choice. When his cataracts become visually significant, cataract surgery combined with angle-based microinvasive glaucoma surgery could be considered.

WHAT I DID: DEVESH K. VARMA, MD, FRCSC

This patient has ocular hypertension and microcornea. Based on the corneal hysteresis and central corneal thickness, Goldmann tonometry might have overestimated the IOP. This consideration combined with the patient's age was reassuring. After a thorough discussion with the patient of risks and alternatives, I elected to monitor him closely. Should he develop glaucomatous changes to the visual field or optic nerve, I would recommend medical treatment.

The cataracts are mild. The patient is young. His ability to accommodate is intact. The small anterior segment could make cataract surgery challenging and increase his risk of developing postoperative malignant glaucoma. I will therefore try to delay surgery until the cataract becomes more symptomatic. ■

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2. Gupta V, Ghosh S, Sujeeth M, et al. Selective laser trabeculoplasty for primary open-angle glaucoma patients younger than 40 years. *Can J Ophthalmol*. 2018;53(1):81-85.
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