

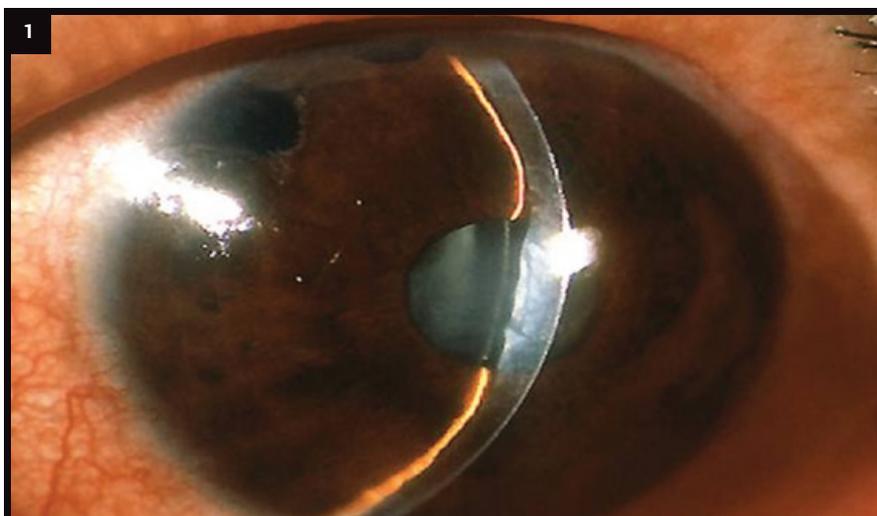
## Diagnosis and Management of Malignant Glaucoma

BY SYRIL DORAIRAJ, MD, IAN CHAN, MD, AND CHRISTOPHER C. TENG, MD  
EDITED BY INGRID U. SCOTT, MD, MPH, AND SHARON FEKRAT, MD

**M**alignant glaucoma, first described by von Graefe in 1869, is characterized by elevated IOP with a shallow or flat anterior chamber, and it usually occurs following ocular surgery. Over time, malignant glaucoma has taken on other names, including aqueous misdirection, ciliary block glaucoma and lens block angle closure. Malignant glaucoma is one of the most complex and difficult of all the glaucomas to treat, and it can progress to permanent blindness without prompt intervention.

### Mechanism

The mechanism of this disorder remains unclear. Shaffer proposed posterior segment pooling of aqueous due to a blockage of the normal anterior aqueous flow by an abnormal vitreociliary relationship. He coined the term ciliary block glaucoma. Another theory by Epstein and colleagues proposed anterior displacement of the vitreous due to posterior diversion of aqueous. They also proposed that there may be associated thickening of the anterior hyaloid, and they were able to demonstrate an impedance to flow across the intact anterior hyaloid in enucleated human and animal eyes with artificially elevated IOP. According to this theory, the accumulation of aqueous within the posterior segment forces the ciliary body and the anterior hyaloid face forward, shallowing the anterior chamber and causing secondary angle closure.<sup>1</sup>

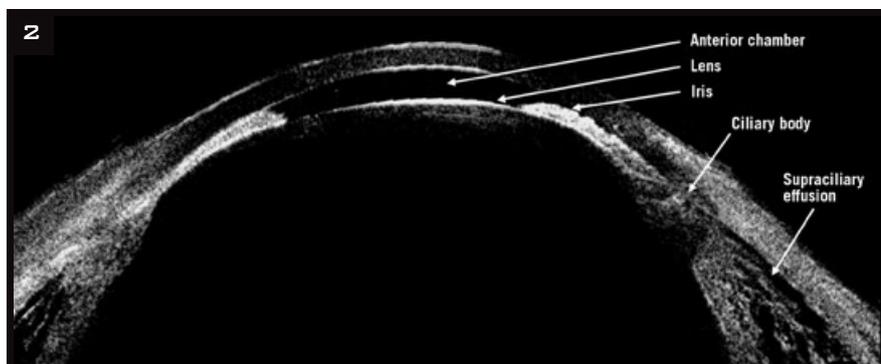


**DISEASE MANIFESTATION.** Slit-lamp photograph of the anterior segment in a phakic patient with malignant glaucoma, showing anterior displacement of the lens-iris diaphragm with shallowing of the central and peripheral anterior chamber in the presence of a patent peripheral iridotomy (1).

Quigley and colleagues implicated the poor conductivity of fluid through the vitreous and stated that past theories proposing misdirected aqueous violate the laws of physics. In his Jackson Memorial Lecture of 2009, Dr. Quigley postulated that if aqueous humor can migrate from the ciliary body through the vitreous gel to the fluid compartment behind it to cause a pressure differential, it would be able to circulate back in the opposite direction just as easily. A functional ball-valve mechanism would be needed to cause the one-way movement of aqueous humor posteriorly, yet none has been documented thus far.<sup>2,3</sup>

Dr. Quigley believes that the inciting event for malignant glaucoma is

more likely to be the inability to generate sufficient fluid flow across the vitreous to compensate for aqueous outflow anteriorly under the higher pressure conditions generated by choroidal expansion. With such choroidal expansion, the pressure within the corneoscleral shell increases and anterior outflow increases. There is a finite ability to transmit fluid through the vitreous cavity. When transvitreous flow is insufficient to equalize the pressure differential, the vitreous compresses more, further decreasing its fluid conductivity and establishing a self-perpetuating cycle. The vitreous gel moves forward, carrying the lens and iris with it, which causes direct angle closure by physically pushing the



**IMAGING.** Composite picture of ultrasound biomicroscopy, showing swelling or anterior rotation of the ciliary body with forward rotation of the lens-iris diaphragm and direct angle closure by physical pushing of the iris against the trabecular meshwork (2). Shallow supraciliary effusion with detachment, which may be responsible for anterior rotation of the ciliary body, is also evident.

iris against the trabecular meshwork with a shallow or flat anterior chamber.<sup>2</sup>

Whatever the underlying pathophysiology may be, it is clear that the iris, ciliary processes, lens and vitreous are intimately involved in the development of malignant glaucoma.

### Risk Factors

Malignant glaucoma occurs in 2 to 4 percent of eyes undergoing surgery for angle-closure glaucoma, and though it may occur at any time during the postoperative period, most cases occur shortly after incisional surgery. It may occur within hours to days or years after iatrogenic causes such as trabeculectomy, cataract extraction with or without IOL implantation, glaucoma drainage implantation, laser iridotomy, capsulotomy, laser suture lysis or argon laser photocoagulation, miotic therapy, needling of filtering blebs, viscoelastic use or intravitreal injection. Infectious and endogenous pathologies include *Aspergillus flavus* intraocular infection and acute hydrops in Down syndrome patients with keratoconus.

It is imperative to be aware of those eyes at particular risk of developing malignant glaucoma, to follow them in the immediate postoperative period and to avoid factors that may aggravate or precipitate malignant glaucoma. This evaluative process should begin preoperatively, with particular attention to risk factors such as hyperopia, chronic angle closure with plateau iris

configuration, nanophthalmos or a history of malignant glaucoma in the fellow eye. Women are three times more likely than men to develop malignant glaucoma, possibly because they have a smaller mean axial length than men, their ocular volume is 10 percent less and they have shallower anterior chambers.<sup>1</sup>

The postoperative development of malignant glaucoma is more frequent in eyes with preexisting angle closure. The anatomical configuration of the ciliary body and iris in these individuals seems to be associated with the future onset of malignant glaucoma. Plateau iris configuration in patients with an anatomically narrow angle should be considered as a possible predictor of postoperative malignant glaucoma development.

Preoperative IOP is not a good indicator of the likelihood of developing malignant glaucoma postsurgically. There have also been cases in which postoperative IOP is normal due to the low rate of aqueous humor formation that occurs as a result of a spontaneous reaction to a previous acute attack of angle closure or secondary to medication use. Unlike in pupillary block angle closure, miotics can exacerbate malignant glaucoma.

### Diagnosis

Malignant glaucoma presents difficult diagnostic and treatment challenges. Slit-lamp examination reveals anterior displacement of the lens-iris dia-

phragm in phakic and pseudophakic patients and of the anterior hyaloid face in aphakic patients, shallowing of both the central and peripheral anterior chamber and elevated IOP (Fig. 1). Most cases occur in the presence of a patent peripheral iridotomy, though cases have been reported without an iridotomy. Physical findings include unequal anterior chamber depths, increasing myopia and progressive shallowing of the anterior chamber.

If there is no iridotomy or if patency is in doubt, an iridotomy must be created to rule out pupillary block. If there is a patent iridotomy and the IOP remains high, choroidal detachment and suprachoroidal hemorrhage must be excluded clinically or by ultrasound biomicroscopy (UBM).

### Imaging

Anterior segment imaging devices have been used to improve our understanding of the pathophysiology, diagnosis and treatment of malignant glaucoma and can complement the clinical examination. UBM has documented swelling or anterior rotation of the ciliary body with forward rotation of the lens-iris diaphragm and relaxation of the zonular apparatus, causing anterior lens displacement and direct angle closure by physically pushing the iris against the trabecular meshwork (Fig. 2). In some eyes, a shallow supraciliary detachment, which is not evident on routine B-scan examination, is responsible for anterior rotation of the ciliary body. Consequently, there is diffuse shallowing of the anterior chamber. In ciliary block, a pressure differential is created between the vitreous and aqueous compartments, a situation analogous to pupillary block, in which the angle becomes occluded by the iris because of a pressure differential between the posterior and anterior chambers.

Anterior segment optical coherence tomography (AS-OCT) is a new imaging method that allows for objective and quantitative imaging of the anterior segment. AS-OCT is similar to UBM in its ability to determine angle parameters quantitatively. Although the resolution is lower and details

behind the iris are not reliably seen, AS-OCT has the benefit of being easier to use and does not require a coupling agent.

### Management

Initial medical therapy is directed at lowering IOP with aqueous suppressants, shrinking the vitreous with hyperosmotic agents and attempting posterior displacement of the lens-iris diaphragm with a strong cycloplegic such as atropine. A laser iridotomy must be performed if one is not present or if patency of a previous iridotomy cannot be established. The effect of medical therapy is often not immediate, but approximately 50 percent of cases will be relieved within five days.

If medical management is unsuccessful, or if lens-cornea touch occurs, Nd:YAG laser therapy may be attempted to disrupt the posterior capsule and anterior hyaloid face. Argon laser through a peripheral iridectomy may also be used in an attempt to shrink the ciliary processes and relieve ciliolenticular blockage of the anterior flow of aqueous. When laser therapy is not possible or is unsuccessful, posterior vitrectomy should be performed with disruption of the anterior hyaloid face.

Once the anterior chamber deepens and the IOP has been normalized, medical treatment can be withdrawn gradually. The patient may require indefinite treatment with cycloplegics to prevent recurrence if vitrectomy was not performed.

1 Ruben, S. et al. *Br J Ophthalmol* 1997;81: 163–167.

2 Quigley, H. A. et al. *J Glaucoma* 2003;12: 167–180.

3 Quigley, H. A. *Am J Ophthalmol* 2009; 148(5):657–669.

*Drs. Dorairaj and Chan are second-year ophthalmology residents at the New York Eye and Ear Infirmary, and Dr. Teng is a clinical assistant professor at New York Medical College and the New York Eye and Ear Infirmary.*

Learn how to write a Pearls article at [www.eyenetmagazine.org](http://www.eyenetmagazine.org).

# 2010 – 2011 Member Directory

*The most comprehensive  
“Who’s Who” in ophthalmology.*

Browse detailed, individual listings by city/state or country for more than 32,000 members of the American Academy of Ophthalmology and the American Academy of Ophthalmic Executives (AAOE).



The Member Directory is a handy desk reference for:

- Providing patient referrals
- Finding colleagues around the world
- Locating state and subspecialty societies
- Contacting Academy departments

**To order your copy, visit [www.aao.org/store](http://www.aao.org/store).**

 **AMERICAN ACADEMY  
OF OPHTHALMOLOGY**  
*The Eye M.D. Association*

 **AMERICAN ACADEMY  
OF OPHTHALMIC EXECUTIVES\***  
Solutions for Practice Management