

# Severe Scleritis, Weak Zonules, and a Rock-Hard Cataract in a One-Eyed Patient

BY DAVID F. CHANG, MD

I always approach surgery on a complex eye by mentally rehearsing a backup plan in the event of a complication. My most challenging case ever was also my most terrifying, because I could not envision what my backup plan would be.

## THE PATIENT'S HISTORY

I first saw this 61-year-old woman in 1997. She had a long history of uveitis and scleritis since the age of 12, which is when she was first diagnosed and managed by Phillips Thygeson, MD. This was complicated by severe secondary uveitic glaucoma. Her left eye underwent cataract surgery in 1984 by one of the leading phaco surgeons in the country. Unfortunately, complications had led to corneal decompensation and severe secondary glaucoma that progressed to optic atrophy and blindness despite all treatment. Her functioning right eye had undergone a successful trabeculectomy in 1977 by H. Dunbar Hoskins, Jr, MD, who, according to the patient, called hers the worst case of scleritis he had ever seen. Her IOP was well controlled in the single digits. As her right cataract became increasingly brunescient and mature, she consulted one of the country's leading phaco surgeons in 1995. His consultation letter described the "formidable anatomic obstacles" and concluded, "There is nothing to be lost by waiting, because the cataract is nearly as hard and fully developed as it will be."

The patient was referred to me by her general ophthalmologist in 1997, because, as a resident of Northern California, she wanted to have her cataract surgery performed close to home. I learned that she was a nationally syndicated writer and an accomplished author of many books who was well known in her field. Although hoping to delay surgery for as long as possible, she was struggling to read and work with 20/400 vision in her only sighted eye.

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## CLINICAL FINDINGS

Examining this eye with extensive scleromalacia at the slit lamp made my heart stop. The entire superior one-fourth of the cornea had thinned seemingly to the thickness of Descemet's membrane. The corneal and scleral thickness looked to be no more than 100  $\mu$ m all along the superior 8 clock hours of her limbus. There was a large, thin-walled bleb inferiorly that was rather bullous and encroached onto the peripheral cornea. There was only approximately 3 mm of normal corneal thickness at the inferior-temporal limbus adjacent to the bleb. The pupil did not dilate to more than 3 mm in diameter, and it remained eccentric due to broad inferior or posterior synechiae. An ultrabrunescient lens obscured any view of her fundus.

Of the two of us, I am not sure who was more scared, but it might have been me. Because of the limited limbal space, which could barely accommodate a phaco incision, I had no backup plan if there was damaged or inadequate capsular support. Converting to a large incision would be impossible if I encountered complications. The tiny 3-mm wide island of nearly normal peripheral corneal thickness would not allow me to insert an ACIOL. Furthermore, there was insufficient tissue to create a scleral flap anywhere else to suture fixate a PCIOL. Suturing both haptics of a PCIOL to the iris had not yet been described, and it would have been difficult



Figure 1. Surgeon's view of the patient's right eye with an inferior bleb, two inferior iris retractors, and the phaco tip placed through an inferotemporal clear corneal incision. There is no red reflex, making visualization of the capsulorhexis extremely difficult. Normal corneal thickness disappears at the whitish areas that involve most of the peripheral cornea, beyond which the cornea looks to be no more than 100- $\mu$ m thick.



Figure 2. A capsular tension ring (CTR) is manually inserted without an injector. The inability to see the margin of the capsulorhexis makes it difficult to tell whether the CTR is entering the capsular bag. Note the position of the sculpted area showing that the insertion of the CTR is decentering the entire loose lens complex superiorly.

because of her eccentric pupil. Wearing an aphakic contact lens was not an option because of the bullous, thin-walled inferior bleb.

There were other potential nightmarish scenarios that I could envision. I knew that a penetrating keratoplasty would be impossible if her corneal endothelium decompensated, and I doubted that a pars plana vitrectomy/lensectomy could be performed in the event that any lens fragments dropped posteriorly. I wondered how I would close the eye if there were any thermal damage to the phaco incision. I was not even sure I could close a sideport incision because of the severe peripheral scleral/corneal thinning.

After we discussed the many risks, I was somewhat relieved when she elected to postpone surgery until she could no longer read with low vision aids. She returned 12 months later and again 6 months after that before she finally decided to proceed with cataract surgery. I had this case on my mind for the ensuing 4 weeks leading up to the date of her surgery in October 1998. Other than a prayer, my only backup plan was to give her aphakic spectacles if the posterior capsule ruptured, and I explained this to her in advance.

## SURGERY

Her referring ophthalmologist came to observe her surgery, and she appreciated the moral support. Assuming the posterior sclera to be equally thin as the

anterior sclera, I used topical anesthesia to avoid the risks of a retro- or peribulbar injection. I made the 2.6-mm clear corneal incision in the only place possible before encountering the first major problem. Despite lysing the posterior synechiae, inserting two iris retractors inferiorly, and maximizing the microscope's illumination and zoom, I simply could not get enough of a red reflex to see the anterior capsule (Figure 1). Capsular staining with trypan blue dye had not yet been described. The difficulty in puncturing her anterior capsule and the excessive mobility of her lens as I maneuvered the capsular flap indicated that her zonules were extremely loose. I had not expected this. After struggling along at a snail's pace, I thought that I had successfully completed a capsulorhexis, but I could not be absolutely sure.

Following hydrodissection, I was unable to rotate the bulky nucleus within the loose capsular bag. As I began to sculpt a central trough through the brunescient nucleus, I was confronted with a surprising degree of phacodonesis. Despite proceeding very slowly, I got increasingly nervous as the lens jiggled with each phaco stroke. I decided to stop to regroup and ponder my options. After becoming a subinvestigator in Morcher's expanded investigational device exemption study, I had earlier purchased a single CTR (Stuttgart, Germany) from the company to be used in an emergency. Reasoning that desperate times required desperate

measures, I decided that now was the time to implant my first CTR. Doing so proved to be extremely difficult, because the pupil was small, I did not have an injector, and I really could not visualize the margins of the capsulorhexis (Figure 2). It was extremely hard to tell if the ring was even entering the bag, and I nervously released the trailing eyelet with mixed feelings of hope and trepidation.

I had heard others speak about how helpful CTRs were for eyes with abnormal zonules. To my dismay, however, there was no reduction in phacodonesis when I resumed sculpting. The idea of using capsule or iris retractors to support the bag had not yet been described, and I did not understand at the time that a CTR can only redistribute instruments' forces to areas of healthy zonules—of which this patient had none. Nevertheless, the naïve notion that the CTR was somehow going to fortify the capsular bag gave me just enough confidence to continue.

After what seemed like an eternity, I eventually succeeded in chopping and removing her nucleus. I accomplished cortical cleanup in the presence of the CTR with great difficulty. I was drenched in perspiration but elated. I filled the loose capsular bag with an ophthalmic viscosurgical device and prepared to inject a foldable IOL. To my dismay, the zonules were so lax that the entire bag bobbed posteriorly away from the approaching IOL. I doubted that I could get the IOL into such a mobile capsular bag and elected to leave it in the ciliary sulcus instead.

### OUTCOME

Postoperatively, the eye displayed a large choroidal detachment that persisted for many years, as the IOP remained in single digits. The IOL was slightly decentered within the sulcus but was optically well tolerated thanks to the patient's small pupil. Her BCVA improved to 20/70, and she could happily read J3 with a simple magnifier!

### LESSONS LEARNED

#### No. 1.

As a surgeon, it is much better to be lucky than good. For example, a CTR does not really stabilize the capsular bag intraoperatively if there are 360° of zonular deficiency. Although it probably did nothing to enhance intraoperative safety in this case as I had intended, the CTR fortuitously prevented postoperative capsular contraction with the IOL positioned in the sulcus.

#### No. 2

We should endeavor to master new technologies. Were I to do this case now (10 years later), I would use

VisionBlue dye (DORC International BV, Zuidland, the Netherlands) and a Malyugin pupil expansion ring (MicroSurgical Technologies, Redmond, WA) to avoid the need for multiple paracenteses. Microcoaxial phaco instrumentation would have been ideal for this case, and I would certainly use hyperpulse power modulation with OZil torsional ultrasound (Alcon Laboratories, Inc., Fort Worth, TX) or Ellips Transversal Ultrasound (Advanced Medical Optics, Inc., Santa Ana, CA) to minimize heat and endothelial trauma from particulate turbulence.

Finally, I would use polypropylene capsule retractors (eg, Mackool Capsule Support System [FCI Ophthalmics, Inc., Marshfield Hills, MA]) once severe capsulodonesis became apparent. Then, I would implant a CTR (using an injector) in the bag after cortical cleanup to prevent postoperative capsular contraction. I prefer a 13.5-mm long STAAR AQ2010V foldable three-piece IOL (STAAR Surgical Company, Monrovia, CA) for placement in the sulcus. I would consider anchoring one haptic to the iris with a 10-0 Prolene (Ethicon Inc., Somerville, NJ) McCannel suture or suturing both haptics to the iris if the posterior capsule were ruptured. In the event of a dropped nucleus, 25-gauge vitrectomy microinstrumentation would most likely be used.

### POSTSCRIPT

I saw this patient several times during her first postoperative year but only twice thereafter. During that second visit in the summer of 2007, we congratulated each other on her 9 years of good visual function and reminisced about how scared we had both been while approaching her cataract operation. Ironically and tragically, she apparently tripped and hit her head on a curb 1 month later, causing a ruptured globe with a spontaneous expulsion of the IOL and much of her iris and vitreous. Her globe was repaired, but she remained limited to counting fingers vision and became very depressed. While preparing this article, I was heartbroken to learn that she had passed away in September 2008 of a possible suicide. Her story is a tragic and sobering reminder of the grave risks we and our patients face whenever we operate on someone's only seeing eye. ■

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