

Hypotony After Trabeculectomy in a Patient With Juvenile Idiopathic Arthritis-Associated Uveitis

BY ANGELA TURALBA, MD, AND DOUGLAS J. RHEE, MD

CASE PRESENTATION

An 11-year-old girl presented to our clinic with glaucoma in her left eye secondary to juvenile idiopathic arthritis-associated uveitis. The patient's right eye was pseudophakic after undergoing a trabeculectomy 4 years earlier and cataract extraction with IOL implantation a few years later.

In 2006, an outside ophthalmologist performed a trabeculectomy with mitomycin C on the patient's left eye to resolve an elevated IOP of 36 mm Hg. Six weeks later, the patient presented with a flat anterior chamber that was subsequently reformed with intracameral Healon (Advanced Medical Optics, Inc., Santa Ana, CA). Three days after this intervention, however, the eye was again hypotonous and had a flat anterior chamber.

One week after the second episode of hypotony, the surgeon revised the trabeculectomy in the patient's left eye. Her postoperative regimen included topical prednisolone, atropine, and Vigamox (moxifloxacin; Alcon Laboratories, Inc., Fort Worth, TX) in her left eye only. The patient was also using systemic prednisone, infliximab, and methotrexate for her juvenile idiopathic arthritis.

Two months after the patient's initial surgery, she presented to our service with a BCVA of 20/40 and an IOP of 5 mm Hg OS. The examination was significant for a shallow anterior chamber, a negative Seidel test, and choroidal folds involving the foveal region (Figure 1). Ultrasound biomicroscopy (UBM) did not show choroidal effusions or abnormalities of the ciliary body (Figure 2).

An examination of the patient's pseudophakic right



Figure 1. Two months after the patient underwent a trabeculectomy in her left eye, a funduscopy examination revealed edema of the optic disc and macular folds consistent with hypotonic maculopathy. Her BCVA measured 20/40 OS.

eye was normal. The IOP measured 7 mm Hg, and a slit-lamp examination revealed a low bleb with minimal vascularity.

We decided to manage the patient conservatively and taper her oral and topical prednisone. This action was intended to stimulate conjunctival fibrosis and strengthen resistance to aqueous outflow. Over the next several weeks, the patient's visual acuity and maculopathy slowly improved, and the IOP in her left eye stabilized at 10 mm Hg. In September 2006, she developed herpes simplex virus



Figure 2. Postoperative UBM of the patient's hypotonous left eye did not show peripheral choroidal effusions or abnormalities of the ciliary body.

keratitis in her left eye while on topical prednisolone. The patient's BCVA dropped to 20/25, her IOP measured 4 mm Hg, and we observed worsening of the macular folds. Although the patient's keratitis and ocular inflammation resolved on a regimen of topical viroptic and oral acyclovir, her IOP remained very low, and her vision started to decline.

HOW WOULD YOU PROCEED?

1. Would you add more steroids to the patient's current medical regimen?
2. Consider a second revision of the trabeculectomy?
3. Perform scleral reinforcement if you decided to revise the trabeculectomy again?
4. Wait to perform another trabeculectomy or to implant a glaucoma drainage device? If so, for how long?

SURGICAL COURSE

In cases of uveitic glaucoma, hypotony can result from persistent inflammation, leaking blebs, or overfiltration. Our patient did not have intraocular inflammation, and her bleb was intact with no leaks. Because we presumed that her hypotony was caused by overfiltration, we did not think steroids were indicated at that time.

When the patient's vision started to decline in October 2006, we decided to revise the bleb with scleral reinforcement. During the procedure, we observed abundant drainage through a full-thickness hole that corresponded with the location of the bleb's internal osteum. This finding suggested that the scleral flap created during the previous trabeculectomy had melted. We reinforced the area by suturing a patch of pericardium over the scleral hole.

Postoperatively, the patient's IOP measured between 5 and 10 mm Hg, and her visual acuity stabilized at 20/20. Two months later, a follow-up examination by the patient's local ophthalmologist revealed an IOP of 1 mm Hg, with a Seidel-negative, cystic, avascular bleb in her left eye.

Over the following year, the patient developed persistently low IOPs that did not cause flattening of the anterior chamber. She also developed low-grade pain in her left eye that was thought to be the result of bleb dysesthesia and hypotony. Her vision fluctuated and gradually declined to 20/40. The patient continued to use systemic steroids for her juvenile idiopathic arthritis and was slowly tapering topical steroids to prevent intraocular inflammation.

Although the patient had already undergone two bleb revisions since her initial trabeculectomy, she continued to experience persistent hypotony and declining visual acuity. In April 2008, we decided to treat the hypotony by reinforcing the sclera at the site of the original sclerotomy. To provide long-term IOP control, we planned to implant a pediatric Ahmed Glaucoma Valve (New World Medical, Inc., Rancho Cucamonga, CA).

During the procedure, we carefully dissected fibrotic subconjunctival tissue from the area of the previous revision. We again observed an area of leakage and a full-thickness scleral hole in the 12-o'clock position of the limbus. Instead of sizing the donated pericardium to cover the hole, we prepared a graft that was large enough to fill the focal scleral defect. We secured the patch by passing a partial-thickness Vicryl suture (Ethicon, Inc., Somerville, NJ) through one of the defect's walls before passing it transversely across the graft and through the opposite wall. We then placed a pediatric Ahmed Glaucoma Valve (model FP8) in the superotemporal region of the left eye and positioned the tube away from the area of reinforced sclera.

OUTCOME

Six months postoperatively, the patient's IOP was stable (between 7 and 10 mm Hg OS). Although she had an early cataract, her visual acuity had returned to 20/20. The patient continues to use methotrexate and infliximab for juvenile idiopathic arthritis as well as a dose of acyclovir to prevent a recurrence of herpes virus keratopathy. Despite the discontinuation of all topical and systemic steroids, she has not experienced any active intraocular inflammation.

DISCUSSION

Between 15% and 42% of patients with juvenile idiopathic arthritis-associated uveitis develop ocular hypertension or secondary glaucoma in response to steroid

treatment or due to mixed mechanisms.¹⁻³ Secondary glaucoma in this setting is associated with a poor visual prognosis, because as many as 18% of patients end up with a BCVA of 20/50 or worse.³ In one case series, the IOPs in 37% (15 of 41) of affected eyes were controlled with medications alone. Two-thirds of the eyes in the same series, however, required surgery to achieve lower controlled IOPs.¹

The abundant inflammation and uveitis associated with juvenile idiopathic arthritis in young patients could contribute to the failure of trabeculectomy in this population.⁴ This high risk of failure could explain why surgeons are implanting drainage devices with increasing frequency in young patients who have glaucoma secondary to juvenile idiopathic arthritis-associated uveitis.⁴⁻⁶

Some devices may provide better long-term control of IOP compared with others. Whereas the Molteno implant (Molteno Ophthalmic Limited, Dunedin, New Zealand) provided good IOP control in uveitic patients after 10 years, the Ahmed Glaucoma Valve was only moderately successful at controlling IOP at 4 years of follow-up.^{5,6} We chose to use the Ahmed device in this case because its valved system reduces the patient's risk of developing postoperative hypotony.

Overall, glaucoma drainage devices appear to be a safe and well-tolerated treatment for uveitic glaucoma. The most common postoperative complication of the Ahmed Glaucoma Valve in patients with uveitic glaucoma is corneal decompensation (3.2% per person/years). The chance of developing severe hypotony with an Ahmed Glaucoma Valve is low but possible (1.3% per person/years).⁶ Given the poor visual prognosis associated with glaucoma secondary to juvenile idiopathic arthritis-associated uveitis, however, the potential benefits of surgical treatment must be weighed against the risk of postoperative complications.

Hypotony is a relatively uncommon complication of juvenile idiopathic arthritis-associated uveitis. Abnormally low IOPs, which are thought to be the result of persistent inflammation of the ciliary body and a decrease in the production of aqueous, can occur in up to 9% of patients suffering from the underlying systemic disease. Affected patients are at risk of developing delayed vision loss secondary to hypotonic maculopathy.³

Of patients experiencing chronic hypotony, those with uveitis had the longest duration of abnormally low IOPs. UBM images obtained from uveitic patients showed atrophy and inflammation of the ciliary body, a finding that confirmed the mechanisms thought to underlie persistent hypotony.⁷ Treating underlying inflammation is thought to improve low IOP in patients

with uveitis-related hypotony. Because our patient was already taking steroids for her juvenile idiopathic arthritis and we did not see active intraocular inflammation on UBM, we did not pursue this treatment option.

Other significant causes of hypotony in uveitic patients who have undergone trabeculectomy include bleb leaks and overfiltration. In children, intraoperative mitomycin C and postoperative steroids reportedly help minimize the abundant inflammatory response that can lead to a trabeculectomy's failure.⁴ The anti-inflammatory effects of steroids and antimetabolites could also impair postoperative healing and thus lead to bleb leaks and overfiltration. Glaucoma drainage devices are a reasonable option for stabilizing IOP in patients with uveitic glaucoma, and surgeons can consider filling scleral defects in eyes that are leaking aqueous from an area of focal thinning.

The poor visual prognosis associated with abnormally high or low IOP in patients with juvenile idiopathic arthritis-associated uveitis supports the use of aggressive anti-inflammatory treatment and careful monitoring of IOP. Glaucoma specialists may improve patients' prognoses by working with uveitis specialists to manage steroidal and immunomodulatory therapy. □

Douglas J. Rhee, MD, is an assistant professor of ophthalmology at Harvard Medical School at the Massachusetts Eye and Ear Infirmary and Children's Hospital Boston. He acknowledged no financial interest in any products or companies mentioned herein. Dr. Rhee may be reached at (617) 573-3305; dougrhee@aol.com.



Angela Turalba, MD, is the chief resident and director of the Eye Trauma Service at the Massachusetts Eye and Ear Infirmary in Boston. She acknowledged no financial interest in any products or companies mentioned herein. Dr. Turalba may be reached at (617) 573-3529; angela_turalba@meei.harvard.edu.



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