

Juvenile Idiopathic Arthritis and Glaucoma

The challenge is balancing the control of IOP and of inflammation.

BY BRENDA L. BOHNSACK, MD, PhD, AND SHARON F. FREEDMAN, MD

Uveitis is commonly associated with the systemic disease juvenile idiopathic arthritis (JIA) and affects 10% to 18% of children diagnosed with JIA.¹⁻³ Risk factors for intraocular involvement include positive antinuclear antibody titers, oligoarthritis, and early age at JIA diagnosis. Poor prognostic factors for visual outcome include a visual acuity of less than 20/60 and high levels of intraocular inflammation at initial presentation, uveitis onset either prior to or within 6 months of arthritis onset, and the presence of cataracts, macular edema, or glaucoma at the time of diagnosis.⁴⁻⁸ The reported rates of visual impairment due to cataracts, macular edema, band keratopathy, synechiae formation, amblyopia, and glaucoma have been as high as 30%, indicating the importance of adequate screening and treatment of chronic inflammation.^{9,10}

Glaucoma affects between 14% and 48% of children with JIA-associated uveitis and is a common cause of irreversible visual impairment.¹¹⁻¹³ Elevated IOP in the setting of chronic inflammation is often multifactorial.¹⁴⁻¹⁷ Trabecular meshwork dysfunction and the formation of synechiae increase resistance through outflow pathways, a process that corticosteroids can exacerbate. Managing the complex balance between the control of IOP and of inflammation can therefore be challenging.

TREATMENT OF PEDIATRIC UVEITIC GLAUCOMA: OVERVIEW

Achieving and maintaining control of intraocular inflammation are key to the successful management of JIA-associated uveitic glaucoma. Although topical corticosteroids remain the mainstay of treatment, the agents may worsen cataracts and increase IOP. Systemic oral corticosteroids and steroid-sparing therapies such as methotrexate, etanercept, infliximab, and adalimumab may be required to control inflammation, especially in cases of refractory JIA-associated uveitis.¹⁸⁻²⁰ Alternatively, local treatments such as sub-Tenon or intravitreal corticosteroid injections may improve inflammation, but cataracts and glaucoma may be



Figure. Gonioscopic view of a goniotomy cleft (arrows) in an eye with uveitic glaucoma. The angle also has a few scattered peripheral anterior synechiae, but in this case, IOPs have been well controlled after goniotomy.

exacerbated, particularly by long-acting steroids.^{21,22}

The medical treatment of childhood uveitic glaucoma includes topical β -blockers and topical and systemic carbonic anhydrase inhibitors. α -agonists (especially brimonidine and, to a much lesser extent, apraclonidine) should be administered with considerable caution, especially in children younger than 5 years, due to a risk of central nervous system suppression (eg, sleepiness and cardiorespiratory depression).^{23,24} Prostaglandin analogues in the setting of uveitis are typically avoided due to concerns over inducing or reactivating inflammation.²⁵⁻²⁷ Given the limitations of medical treatment, cases of childhood uveitic glaucoma may require surgical intervention.

When planning a procedure for a child with glaucoma, the surgeon needs to consider a strategy that maximizes the control of IOP and preserves vision over decades. The surgical management of uveitic glaucoma is particularly challenging due to underlying inflammation. Often, an intense postoperative inflammatory response complicates the control of IOP and uveitis. Although high-dose oral steroids (approximately 1 mg/kg/day) administered just before and during

the immediate postoperative period can help suppress this inflammation, glaucoma surgery is most successful when the uveitis is well controlled preoperatively. Furthermore, with control of intraocular inflammation, glaucoma surgery can achieve long-term control of IOP and preservation of vision.²⁸ Reports regarding pediatric uveitis and specifically JIA-associated glaucoma are limited, and there is no consensus on the strategy for treating these difficult cases.

ANGLE SURGERY

Angle surgery—specifically goniotomy—may be an effective IOP-reducing procedure in childhood uveitic glaucoma. Goniotomy involves incising the trabecular meshwork, creating a cleft, and presumably removing inflammatory debris blocking the outflow pathway (Figure). This procedure requires excellent preoperative control of the uveitis to help avoid inflammation-mediated synechiae formation and cleft closure. Goniotomy requires a relatively clear cornea, a gonioscopic view of the angle, and surgical expertise, but this relatively noninvasive procedure preserves the conjunctiva and can be repeated for additional IOP-lowering effect.

Two studies, each with more than 30 patients who had childhood uveitis, demonstrated that goniotomy (1-3 total procedures) has a 10-year success (by Kaplan-Meier life table analysis) of 69% to 71% for controlling IOP.²⁸⁻³⁰ These studies included many forms of childhood uveitis, but the majority of patients (56%-75%) had the diagnosis of JIA-associated uveitis.^{28,29} Goniotomy is less successful in the setting of significant peripheral anterior synechiae and a predominantly closed angle. Although angle surgery can be performed in eyes that are phakic, pseudophakic, or aphakic, goniotomy may be less successful in aphakic eyes.²⁹ Reduced goniotomy success after cataract removal may be due to structural changes in the angle after the lens' removal or overall worse uveitic control indicated by the occurrence of visually significant cataracts.

Given the low risk associated with goniosurgery and the relatively high success of IOP control, goniotomy should be considered as a first-line surgery for childhood uveitic glaucoma secondary to JIA, especially in phakic eyes with minimal synechiae and an open angle.

GLAUCOMA DRAINAGE DEVICES

Glaucoma drainage devices (GDDs) lower IOP and are a standard treatment for adult uveitic glaucoma.³¹⁻³⁵ Reports on the use of GDDs in pediatric uveitic glaucoma are limited both in number of eyes and length of follow-up. In a few small studies, both the Ahmed Glaucoma Valve (New World Medical, Inc.) and Baerveldt glaucoma implant (Abbott Medical Optics Inc.) were shown to decrease IOP

and the number of glaucoma medications in children with uveitic glaucoma.^{28,36-38}

Due to the nature of GDDs, additional surgeries to address hardware-related problems such as exposure and migration of the tube and failure of the valve mechanism are common. GDDs in the pediatric population can also cause strabismus, increased inflammation, cataract formation, pupillary abnormalities, and corneal decompensation.^{36,37,39} Furthermore, a recent study demonstrated that Ahmed valves in children with uveitic glaucoma are correlated with decreased corneal endothelial cell densities that worsen over time.⁴⁰ Thus, care must be taken to implant GDDs as far posteriorly as possible to prevent irreversible damage to the cornea. Despite problems related to hardware in and around the eye, GDDs can be useful in cases of JIA-associated uveitic glaucoma, especially in eyes in which goniotomy has failed or in eyes that are not amenable to angle surgery.²⁸

TRABECULECTOMY

Trabeculectomy surgery in children with uveitic glaucoma typically does not control IOP.^{41,42} After intraocular surgery, early hypotony usually flattens the bleb and causes early bleb failure. Antifibrotics used in conjunction with trabeculectomy help prevent immediate scarring of the scleral flap but result in thin-walled blebs that are susceptible to infection.⁴³⁻⁴⁹ Early failure and a lifelong risk of infection combined with the difficulty of monitoring blebs in young children make trabeculectomy less than ideal for treating cases of pediatric uveitic glaucoma.

CYCLOABLATION

Cycloablation is commonly reserved for eyes that have late or end-stage glaucoma. Whether from an external or internal approach, these procedures can have an unpredictable effect on IOP.⁵⁰⁻⁵⁴ In a small series of patients with JIA, one study demonstrated that transscleral laser cyclophotocoagulation had less than a one-third success rate in decreasing IOP after three treatments.⁵² On the other hand, overtreatment can result in hypotony and subsequent phthisis bulbi. Cycloablation also causes a significant intraocular inflammatory response that can exacerbate the uveitis. Cycloablation should therefore be used sparingly, if at all, in children with JIA-associated uveitis.

SUMMARY

The management of childhood glaucoma associated with uveitis and specifically JIA is challenging, with no consensus on optimal management. Treatment of this form of glaucoma must address both inflammation and IOP. The use of topical and systemic steroids to control intraocular inflammation may increase IOP, often requiring systemic steroid-



sparing therapy. The options for glaucoma medication are limited due to underlying inflammation and the young age of many children affected by JIA-associated uveitis, and a large number of them require surgical intervention for adequate IOP control.

Glaucoma surgery is most successful when inflammation is controlled preoperatively; goniotomy and GDDs offer long-term IOP control for many cases. Goniotomy effectively decreases IOP and can be used as a first-line surgery, especially in phakic eyes that have open angle configurations. GDDs also lower IOP and should be considered in eyes that have previously undergone goniotomy or in eyes in which angle surgery is not advisable.

The vision of children with JIA-associated uveitic glaucoma can be preserved if inflammation and IOP are well controlled through medical and surgical treatments. ■

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