

Managing the Pediatric Glaucomas

What are the long-term implications?

BY DAVID S. WALTON, MD

Caring for children who have glaucoma is challenging and rewarding. The pediatric glaucomas are diverse, with the potential for many different primary and secondary causes in children who present from infancy through adolescence. In addition, the severity of defects in the filtration angle may be mild or advanced in both genetically determined glaucoma and secondary glaucoma.

The general difficulty of examining children is an inescapable obstacle to their successful assessment and the preparation for glaucoma surgery, which is more often than not the initial therapy for this disease. When managing pediatric glaucoma, physicians must recognize the uncertainty of the presently available therapies and realize that additional procedures may be necessary for young patients with a life expectancy of many decades.

CLINICAL EXAMINATION

Physicians have an obligation to every family to perform the examinations necessary to making appropriate

decisions on treatment. This work begins in the office, before the examination with general anesthesia. The goals are to confirm the presence of glaucoma and to determine its diagnostic type, the severity of the filtration angle anomaly, and the abnormalities secondary to the elevated IOP.

Successful tonometry, slit-lamp examination of the anterior chamber, and gonioscopy are necessary at this point as well as during postoperative follow-up. Tonometry is essential to establishing the presence of glaucoma and following patients who undergo medical and/or surgical treatment. By revealing the severity of the anomaly and/or secondary changes to the filtering mechanism, informative gonioscopy enables ophthalmologists to select and plan pediatric glaucoma surgery. Even very young patients will permit Koeppel gonioscopy in a clinical setting (Figure 1A), and this technique must also be successfully employed during the examination under anesthesia in preparation for surgery (Figure 1B). Pre-and postoperative B-scan ultrasonography is helpful as well, especially during inevitable periods of hypotony follow-

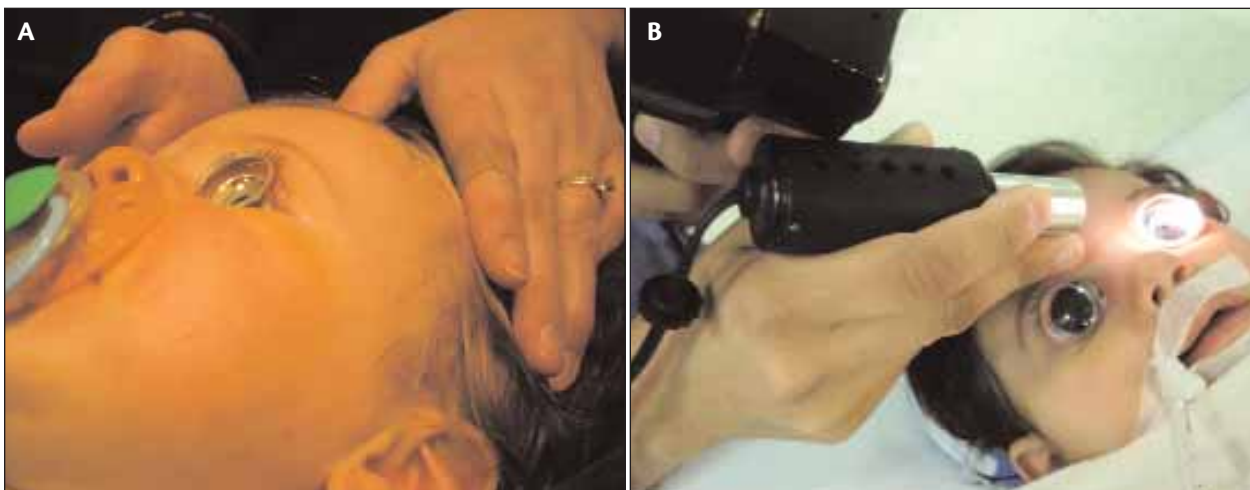


Figure 1. Koeppel gonioscopy is performed in an office setting as part of the assessment for primary congenital glaucoma (A). The physician performs Koeppel gonioscopy during an examination with anesthesia (B).

TABLE 1. THE INDICATIONS FOR GONIOTOMY FOR THE TREATMENT OF PEDIATRIC GLAUCOMA

	Favorable Prognosis	Unfavorable Prognosis
Primary Glaucomas	Infantile primary congenital glaucoma	Lowe syndrome
	Late-recognized primary congenital glaucoma	Neurofibromatosis (NF-1)
	Juvenile open-angle glaucoma	Sturge-Weber syndrome
	Hereditary iridotrabeculodysgenesis	Congenital iris ectropion syndrome
		Rieger syndrome
Secondary Glaucomas	Glaucoma with chronic anterior uveitis	Infantile aphakic glaucoma
	Congenital glaucoma with maternal rubella	Chronic anterior uveitis, aphakia, and anterior peripheral synechiae
	Steroid-induced glaucoma	

ing surgery.

Communication with parents must be active and informative to lay the foundation of trust necessary for a child's continued care.

MEDICAL THERAPY

An implicit goal of the care of children with glaucoma is minimizing, when possible, repetitive surgeries. Glaucoma medications can help achieve this aim. They also may allow the postponement of a glaucoma procedure until a child is healthier, more cooperative for examinations, older and thus a more suitable candidate for trabeculectomy surgery, or possessed of a clearer cornea in preparation for goniosurgery.

Topical beta-blockers can be well tolerated for long intervals without systemic complications. Both timolol 0.5% (Timoptic Ocudose; Merck & Co., Inc., Whitehouse Station, NJ) and the timolol 0.5%/dorzolamide 2% combined preparation (Cosopt; Merck & Co., Inc.) are available in preservative-free vials—highly desirable for children on prolonged topical glaucoma therapy.

Acetazolamide (Diamox; Wyeth Pharmaceuticals, Philadelphia, PA) may be the most effective IOP-lowering medication for use in children. The drug may be used cautiously in infants to lower pressure temporarily or for selected indications for chronic use among older children (eg, glaucoma related to Sturge-Weber syndrome). The agent's use in infants may become associated with anorexia, hyperpnea from renal acidosis, and dehydration. In older children, anorexia may occur associated with a subtle decrease in growth and chronic fatigue. Ten to 15 mg/kg orally per day in divided doses is appropriate. Most community pharmacists are willing to prepare a suspension for pediatric use with a concentration of 10 to 20 mg/mL for oral administration.

SURGICAL THERAPY

Goniosurgery

Until recently, the choice of goniosurgery was confined to the pediatric glaucomas. For children, it remains the first surgical choice for the treatment of the types of glaucoma for which it has proven effective (Table 1). Barkan developed the goniotomy technique and described its successful use for infants with glaucoma.^{1,2} This seminal accomplishment meant effective therapy for what had, until then, been considered a hopeless disease.

A preoperative gonioscopic evaluation is essential preparation for goniosurgery and may reveal encouraging or potentially discouraging findings. Inadequate pressure control after an initial goniotomy often indicates a need for repeat goniosurgery. The consideration



Figure 2. The cornea of the right eye is cloudy in this patient with newborn primary congenital glaucoma.

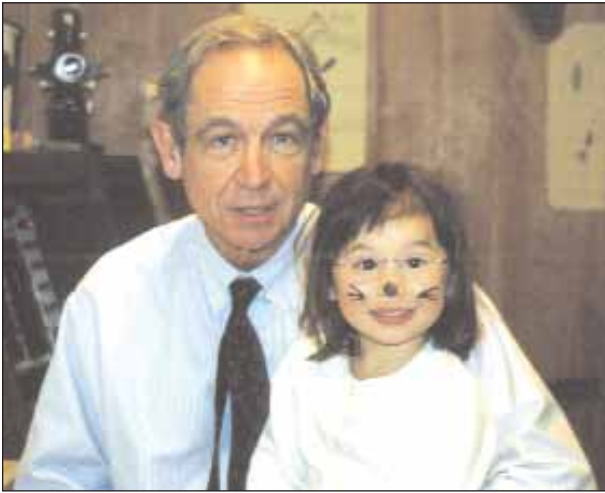


Figure 3. The author sits with a young patient who underwent goniosynechiolysis for angle-closure glaucoma.

of repeat goniotomy should be irrespective of the patient's age, even into adulthood.

Late-recognized primary congenital glaucoma can be difficult to distinguish from primary juvenile glaucoma, but, fortunately, goniotomy is indicated for both conditions. The expected outcome of goniosurgery for primary congenital glaucoma is very favorable overall, with a reported success rate of over 80%.³ The severe angle anomaly with extreme forward insertion of the iris and minimal development of the trabeculum typically seen in newborn primary glaucoma decreases the potential for a successful goniotomy.⁴ These patients recognized to have glaucoma between birth and 2 months of age have a lower success rate with goniotomy.⁵⁻⁷ A relapse following successful goniotomy is unusual⁷ and was reported to occur more frequently when the glaucoma was first recognized at birth (36%) versus before 6 months of age (16%).⁷ This observation holds true for successful procedures for both primary and secondary pediatric glaucomas.

Trabeculotomy ab externo is an alternative to goniotomy for primary congenital glaucoma. Its successful use for infantile glaucoma was recognized early.^{8,9} Trabeculotomy and goniotomy for infantile glaucoma are equally effective and safe, but the former offers a significant advantage in eyes with a cloudy cornea that limits visualization of the angle¹⁰ (Figure 2). Investigators performed primary combined trabeculotomy/trabeculectomy on 144 eyes with primary congenital glaucoma, and they recommended the procedure for eyes with significant corneal edema based on a success rate of over 90% at 6 months.¹¹ A variation on this procedure is to thread a suture circumferentially through Schlemm's

canal and then to remove it into and out of the anterior chamber, which creates a communication between these structures that has been effective for primary congenital glaucoma.¹²

Research on the long-term effectiveness of trabeculotomy has confirmed its efficacy for primary congenital glaucoma. Investigators found a probability of success after one or more surgeries in 99 eyes to be 92% and 82% at 5 and 10 years, respectively.¹³ Trabeculotomy for 46 children with primary congenital glaucoma and a mean age of approximately 4 years (range, 13 to 88 months) was successful in 87% (68 eyes) with customized probes for 102 procedures.¹⁴

Trabeculectomy

The introduction of trabeculectomy lessened the risk of filtering surgery for young children but did not significantly improve control of their glaucoma. The addition of antimetabolites to trabeculectomy surgery for children improved its outcome.¹⁵

The indication for pediatric trabeculectomy surgery is the persistence of an unacceptable elevation in IOP following an appropriate goniosurgery. The condition of the superior conjunctiva related to the previous surgery, the patient's age, the potential for follow-up examinations, the postoperative use of contact lenses, and the risk of postoperative infection all must be considered. The adoption of the fornix-based approach has augmented the chance of long-term filtration and reduced the need for revision for infection due to conjunctival thinning.¹⁶

Trabeculectomy has a significantly lower rate of success among children under 1 year of age than older patients,¹⁷ and the use of an antimetabolite has not improved outcomes in the former group. The reason for the difference is unclear. The surgery is not more complicated. In patients under 1 year of age, the postoperative course of trabeculectomy is characterized by decreasing evidence of external filtration, even among patients who initially achieve improved IOP control and external evidence of filtration.

Glaucoma Drainage Devices

Glaucoma implant surgery is an important treatment alternative for pediatric glaucoma patients who are poor initial candidates for angle-incision therapy and trabeculectomy as well as for those whose disease has proven to be refractory to these procedures. Drainage devices have been widely adopted, and surgeons now better understand the implants' efficacy and complications.^{18,19}

Initially, devices were considered only after other procedures had failed, but their indications have expanded.

Most patients with primary congenital glaucoma will initially undergo goniosurgery. If that procedure is unsuccessful, trabeculectomy will be considered, followed by implant surgery. When preoperative gonioscopy reveals a severe angle anomaly in a case of newborn glaucoma, however, the expected outcome of both goniosurgery and trabeculectomy is poor. These infants are strong candidates for primary implant surgery. A study comparing the outcomes of 32 patients with a mean age of 7 months treated with glaucoma implants and 19 patients with a mean age of 5 months treated with trabeculectomy found a probability of success at 77 months of 53% and 19%, respectively.²⁰

Importantly, the potential success of implant surgery for refractory primary congenital glaucoma has greatly decreased the use of destructive cycloablative procedures. When placed in 60 pediatric glaucoma eyes, including 25 with primary congenital glaucoma, the Ahmed Glaucoma Valve (New World Medical, Inc., Rancho Cucamonga, CA) achieved IOP control in 73% (44 eyes) at last examination, with the continued use of medications in 77%. Complications, however, occurred in 50% (30 eyes).²¹ Early hypotony and tube-related problems are the most common complications.

Cyclodestruction

Ophthalmologists may use cyclodestructive procedures selectively in cases of pediatric glaucoma that is refractory to medical therapy and conventional surgical procedures. The required ciliary epithelial ablation can be clinically produced by cyclocryotherapy or by transscleral or endoscopic diode laser cyclophotocoagulation. The decision to perform these intentionally damaging procedures must take into account the long-term visual and anatomic prognoses and parents' expectations for the affected eye. The ocular indications include a blind painful eye, a blind eye with a high pressure and rapidly deteriorating cornea, an eye proven to be refractory to all more conservative treatment alternatives, an eye with anatomic defects that preclude other glaucoma procedures, and a patient who is not a candidate for either prolonged general anesthesia or intraocular surgery.

The complications of cyclodestruction include phthisis, retinal detachment, chronic hypotony, and cataracts. The chronic postcyclodestruction anterior segment syndrome seen frequently in children successfully controlled by this procedure is characterized by band keratopathy, corneal edema and a dropout of endothelial cells, chronic anterior chamber flare, posterior synechiae and microcoria, and slowly progressive cataracts.

CONCLUSION

The care of children with glaucoma has improved dramatically during the last decade. The use and development of goniosurgery continues, but filtering surgery in cases of glaucoma that is unresponsive to angle therapy is now more likely to produce a favorable outcome without the late complication of progressive conjunctival deterioration. Glaucoma implants are an appropriate and effective option for certain cases of pediatric glaucoma, and new devices that are smaller and more biocompatible should control IOP even more reliably.

It is important that academic centers, faculty, and the ophthalmic profession strive to increase residents' exposure to the challenge and rewards of caring for children with glaucoma (Figure 3). □

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