

Pediatric Glaucoma

Advice on diagnosing and managing a rare but potentially devastating group of diseases.

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Childhood glaucomas are infrequently encountered by many eye care providers and can therefore be challenging to diagnose. Because this heterogeneous group of diseases can cause a rapid loss of vision or even blindness, however, the timely recognition and optimal treatment of pediatric glaucoma is critical. Fortunately, ophthalmologists often have at their disposal the tools needed to diagnose and begin to manage these children. Usually, the ideal care of patients with pediatric glaucoma involves the efforts of more than one ophthalmologist, unless a pediatric glaucoma specialist is located near the child's home.

BACKGROUND

The most common form of pediatric glaucoma, primary congenital glaucoma, occurs in approximately one in 10,000 individuals.¹ As with adult forms of the disease, pediatric glaucoma may be primary or secondary. The former involves intrinsic disease of the aqueous outflow system and often has a genetic basis, such as with primary congenital glaucoma. Secondary pediatric glaucoma is caused by an injury or another ocular condition (eg, glaucoma after the removal of congenital cataracts). Pediatric glaucoma is sometimes associated with systemic disease in children. In contrast, primary open-angle glaucoma often occurs in an otherwise healthy adult.

PRESENTATION

The signs and symptoms of pediatric glaucoma vary depending on the age of the child and on the acuteness and magnitude of the elevation in IOP. Although glaucoma is often silent in adults, its presence is frequently noticeable to the families of affected children, particularly those younger than 1 year. Infants with glaucoma usually present because the high IOP has caused one or both of their eyes to expand. The cornea may have enlarged and clouded (due to edema), and there may be asymme-

try between the corneas of the infant's eyes. Rapid corneal stretching may lead to breaks in Descemet's membrane called *Haab's striae*, which will leave permanent scars (Figure 1). Oftentimes, the affected infant is photophobic (Figure 2) and may have tearing that the clinician must distinguish from nasolacrimal duct obstruction.^{1,2}

Newborns with healthy eyes have a corneal diameter of approximately 9.5 to 10.0 mm. A corneal diameter of greater than 12.0 mm in any infant younger than 1 year is therefore suspicious for glaucoma.³ The IOP usually ranges from 10 to 15 mm Hg in newborns, whereas the IOP in children of school age resembles that of adults. Most children with glaucoma will have an IOP that is higher than 22 mm Hg.

Although infants and children under the age of 2 often present with signs and symptoms related to rapid ocular expansion under high IOP, older children without an acutely elevated IOP typically present to an eye care provider with myopia due to glaucoma-related elongation of the eye. Occasionally and tragically, visual loss may already be permanent if the high IOP is not detected for a prolonged period and severe damage to the optic nerve has occurred. It is important to note that glaucomatous stretching of the eye is usually irreversible in older infants and children.

The optic nerve can undergo similar morphologic changes in pediatric glaucoma as in adult forms of the disease. A major difference is that, after a decrease in IOP, some cupping is reversible in infants and very young chil-



Figure 1. Primary congenital glaucoma was recognized late in this 8-month-old infant. Note the significant corneal scarring from Haab's striae in both eyes despite IOP control with goniotomy surgery.

dren, presumably due to the flexible lamina cribrosa in children compared with the more rigid tissues of the optic nerve in adults. Newborns usually have a cup-to-disc ratio of less than 0.4, and there is good symmetry between the optic nerves of both eyes.⁴ Whereas older children often have a vertical optic nerve cupping pattern similar to that in adults, the cupping in younger children with elevated IOP tends to be more generalized and central, although cupping at the poles can also occur.

EVALUATION

Clinical Examination

In addition to examining the child and measuring the visual acuity of each eye, ophthalmologists should inspect the eye, estimate its horizontal corneal diameter, assess the corneal clarity, and observe the patient's sensitivity to light. They should also note associated conditions such as a facial port wine stain involving the upper lid and face and suggesting Sturge-Weber syndrome, an absence of normal iris tissue indicative of aniridia, and aphakia. A handheld slit lamp is valuable for evaluating the anterior segment in infants and young children, because the majority of them cannot sit properly at a freestanding slit lamp.

When evaluating the optic nerve in children, many ophthalmologists favor a 28.00 D lens with the indirect ophthalmoscope, but this technique will often underestimate true cupping in children. A 14.00 D lens is preferable for infants and young children, although a direct ophthalmoscope used through a gonioscopy lens in the OR (if an examination under anesthesia is necessary) also affords an excellent view of the optic nerve, even through a relatively small pupil. A 90.00 D lens or the equivalent can be used for a binocular view of the optic nerve at the slit lamp and is appropriate for viewing the optic nerve of an older child in the office.

IOP Measurement

An elevated IOP is necessary to confirm the diagnosis of glaucoma in a young child. The most convenient devices in infants and young children are a Perkins tonometer (Veatch Ophthalmic Instruments, Tempe, AZ) and the Tono-Pen XL (Medtronic Xomed Ophthalmics, Inc., Minneapolis, MN).

Attempting to measure IOP is pointless in a screaming child. If the patient is relatively calm, the physician may try a number of strategies for measuring the IOP. Infants who are comfortable, warm, and being fed will often submit to testing. Toddlers may be sufficiently distracted by a lollipop, a spinning, lighted toy, or a television at close range. With all children, relaxation on the part of the examiner and parents can be helpful. When recording the IOP in the medical record, it is worthwhile to note the technique used for measuring the IOP, the child's



Figure 2. This 6-month-old infant has severe photophobia from corneal edema and Haab's striae in the setting of primary congenital glaucoma and uncontrolled IOP in both eyes.

level of cooperation, and the time of day.

When the diagnosis of glaucoma is uncertain, as in a school-aged child with a suspicious-appearing optic nerve and a borderline IOP, performing a modified diurnal curve of the IOP can be helpful. The IOP of children with healthy eyes will vary by only 1 or 2 mm Hg between time points and between eyes. Patients with abnormal aqueous outflow can exhibit a variation of 10 mm Hg or more over the course of a day, and the difference in IOP between their eyes will often be 3 mm Hg or higher.

Pachymetry

Central corneal thickness varies among children, with pachymetry readings averaging 550 μm in normal eyes and thinner corneas in black versus white or Hispanic children.^{5,6} Moreover, children with aniridia and those who are aphakic after the removal of a congenital cataract seem to have unusually thick corneas. Children with enlarged corneas from congenital glaucoma often have thinner-than-average corneas. Despite the report of a positive association between increasing central corneal thickness and recorded IOP among normal pediatric eyes,⁷ there are no well-established nomograms for altering the IOP measure-

ments in children with especially thick or thin corneas, especially if they are in any other way abnormal. It seems reasonable to take central corneal thickness as one factor in the selection of a target IOP for older children with glaucoma, but this measurement plays a minor role in the management of most young children with the disease.

Ancillary Testing

Visual field testing rarely assists the initial diagnosis in infants or young children. On the other hand, a cooperative child without nystagmus and with reasonably good central vision can usually begin automated visual field testing by the age of 8 years. Visual fields are useful for following glaucoma over time in older children. Stereoscopic photography of the optic nerve head is recommended as a baseline for older children with glaucoma, although the presence of nystagmus will limit the quality of the images. The utility of ocular coherence tomography in children is the subject of current research, with the values for the normal eyes of children recently reported.⁸ One study⁹ showed that the peripapillary nerve fiber layer and macular thickness volume was higher in healthy versus glaucomatous pediatric eyes. At present, it seems prudent to use ocular coherence tomography as a baseline that may prove useful for following patients over time.

OBSERVATION VERSUS TREATMENT

Ophthalmologists must determine the ideal treatment regimen on a case-by-case basis. At times, it may be appropriate to follow a child carefully without treatment if the diagnosis of glaucoma is uncertain. The most important issue in such a case is to ensure that the family understands the importance of regular follow-up so that treatment may begin if necessary. Most often, a child with glaucoma will require intervention—medical, surgical, or both.

MEDICAL THERAPY

Background

Although the first-line treatment of primary congenital glaucoma is surgical, medical therapy is important with many other open-angle forms of pediatric glaucoma. Children with aphakic, uveitic, and juvenile open-angle glaucoma usually should first receive medication and may avoid surgery for a long time (or even indefinitely) if therapy achieves the target IOP. Medication also plays a vital role in cases in which surgery did not lower IOP adequately by avoiding the need for more aggressive (and often riskier) surgery.

Children with glaucoma often adhere to their prescribed therapy quite well, particularly with strong familial support. Complicated dosing regimens can profoundly affect the child's and the family's quality of life, however,

as well as their ability to comply with prescribed therapy. Although the clinician may add a medication if the current regimen does not achieve the desired IOP, it is useful first to re-evaluate the effect of each prescribed drug. Whenever possible, a monocular trial and changing one drug at a time are advisable.

It is important to note that most of the glaucoma drugs on the market today were approved without any data on their safety in children. The FDA is currently encouraging pharmaceutical companies to test their newer drugs in this population, and some have complied.¹⁰

Carbonic Anhydrase Inhibitors

Topical carbonic anhydrase inhibitors (CAIs) such as dorzolamide and brinzolamide are quite useful in pediatric glaucoma and are frequently the first choice for childhood glaucoma, especially when beta-blockers are contraindicated. CAIs are well tolerated systemically and may be used b.i.d. or t.i.d. with significant efficacy (often a reduction in IOP of 20% to 25% in the author's experience). They are also additive to beta-blockers. CAIs are relatively contraindicated for pediatric eyes with clear corneal transplants, however.¹¹

When a decrease in IOP is urgent, oral CAIs are an option but should be used with caution in infants, who may develop respiratory alkalosis or tachypnea. For young or school-aged children, oral acetazolamide is the most potent IOP-lowering drug available. It may be administered as a pill or as a suspension, dosed at 10 to 20 mg/kg per day based on the child's weight and given b.i.d. or t.i.d. with food. Common side effects include a general decrease in appetite, some tingling of the fingers and toes and around the mouth, and fatigue.¹¹

Beta-Blockers

Beta-blockers can be an excellent choice for lowering IOP in older children because the drugs may be dosed q.d. or b.i.d., depending upon their formulation, and usually reduce IOP by approximately 25%. In infants and young children, practitioners should start with the lowest available dose (eg, timolol 0.25% gel-forming solution) to minimize the systemic blood levels and beta blockade that may result.

These drugs are contraindicated in premature babies and children with known cardiac or pulmonary disease, and they should be used with care in young infants. Beta-blockers should be used with caution in any child who has asthma or reactive airways, for whom it may be wise to begin with betaxolol, a beta-2 selective agent.¹

Miotics

Miotics are generally useful in children only for constricting the pupil in order to protect the crystalline lens

during surgery and to keep the cleft open after angle surgery in infants and young children. Pilocarpine or echothiophate iodide may be used in select cases of aphakic glaucoma.

Alpha-2 Agonists

Brimonidine is absolutely contraindicated in infants and young children due to its depression of the central nervous system. The drug also causes sedation and fatigue in school-aged children, so it must be used with caution in this population, if at all.^{12,13} In contrast, even fairly young children usually tolerate apraclonidine 0.5% well, and the drug can help achieve a short-term reduction in IOP and can decrease bleeding at the time of angle surgery.

Prostaglandin Analogs

Latanoprost seems to be quite useful for the treatment of juvenile open-angle glaucoma and select cases of primary congenital and aphakic glaucoma.¹⁴ Anecdotally, the author has found that latanoprost, travoprost, and bimatoprost all cause children to grow impressively long and thick eyelashes. For patients receiving monocular treatment, the difference in the appearance of the lashes between the treated and untreated eyes is readily apparent. Moreover, the eyelashes can grow so long as to cause smearing on the inside of spectacle lenses.

Data are limited on prostaglandin analogs in children. Response rates have been relatively low in published reports compared with those seen in adults. For that reason and because of cosmetic concerns, they are generally not first-line therapy for pediatric glaucoma.¹⁴

Fixed-Combination Agent

A fixed combination of timolol and dorzolamide can be an excellent choice in older children who have responded well to a beta-blocker and a topical CAI. Clinicians should be on the alert for an afternoon rise in IOP, however. In such cases, a single, additional drop of dorzolamide administered in the afternoon can be helpful.

SURGERY

This article does not provide a detailed analysis of surgical intervention for pediatric glaucoma but will touch on a few relevant points. Although medications can serve a temporizing role by clearing the cornea and slowing the rate of damage, two types of childhood glaucoma almost uniformly require surgery. They are primary congenital glaucoma and all forms of angle-closure glaucoma (which occur in the setting of advanced retinopathy of prematurity and in some cases of uveitis and aphakia).

As in adults, uveitic glaucoma in children can be difficult to treat. When medications fail and physicians are

considering surgical intervention, they should remember that goniosurgery can be an effective option in children who have glaucoma related to chronic anterior uveitis.^{15,16} This procedure is less invasive than trabeculectomy or the placement of a tube shunt.

CONCLUSION

Fortunately, pediatric glaucoma is rare. Treatment modalities have decades longer to produce side effects and fail in a pediatric versus an elderly patient, however. In addition, achieving a low IOP in children will not spare them from visual loss due to amblyopia, corneal scarring, or significant ametropia. Simply reducing the IOP to a safe level for the optic nerve is thus only one part of caring for a child with glaucoma.

The successful management of pediatric glaucoma can be highly rewarding, and it is well within the purview of most ophthalmologists to evaluate and treat this group of patients, either alone or as part of a team. Thanks to the variety of medical and surgical options available today, most children with glaucoma can become visually independent adults. □

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