Update on Pediatric Glaucoma Surgery

This glaucoma subspecialist shares a 30-year perspective.

BY RONALD L. FELLMAN, MD

Health care professionals should be knowledgeable about the signs of congenital glaucoma. Unlike most adult forms, pediatric glaucoma is not a "silent" disease, because the elasticity of the infant eye allows for stretching of ocular tissues up to 3 years of age. This causes a variety of signs and symptoms, including epiphora (tearing), photophobia (light sensitivity), a cloudy or hazy cornea, and buphthalmos (ocular enlargement). Even something as subtle as a 4-month-old's not smiling back at his or her parent may be the first sign that something is wrong with the baby's visual system. An observation of this missed developmental milestone may prompt a parent to bring the child in for an evaluation, and glaucoma may be the culprit.

The management of pediatric glaucoma still relies heavily on angle surgery to cleave open a maldeveloped angle. Expert gonioscopic skills are essential to recognize and treat congenital glaucoma (Figure 1).[1] The big development in pediatric angle surgery over the past 3 decades is circumferential canal surgery, the ability to open the canal for 360º at one sitting in a highly reliable and accurate way with either a suture or a microcatheter (iTrack; iScience Interventional). The ophthalmologist's ability to visualize the position of the normally hidden canal probe through the tissues via the illuminated iTrack is a major advance in canal surgery and in teaching angle surgery. Another significant step forward is the use of drainage implants when angle surgery is unsuccessful or contraindicated. In years past, cyclocryosurgery was the mainstay when trabeculotomy and trabeculectomy failed. Today, glaucoma subspecialists such as myself increase aqueous outflow by implanting drainage devices instead of destroying the ciliary body through the application of cryosurgery. The days of cyclocryosurgery with phthisis bulbi are no more. If cyclodestruction becomes necessary, a kinder, gentler transscleral or endoscopic cyclodiode laser (Endo Optiks) works quite well.

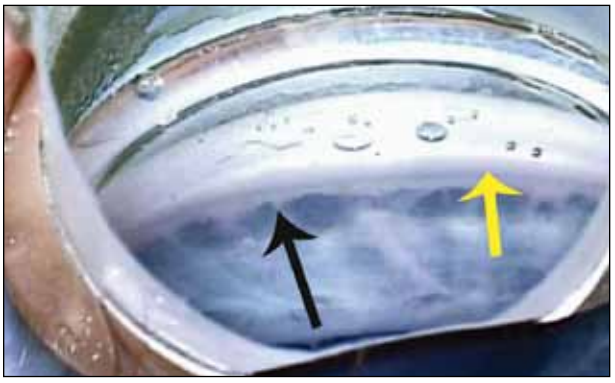

Figure 1. Iridotrabeculodysgenesis. The eye of an 8-month-old with congenital glaucoma demonstrates the classic features of a malformed angle. They include the scalloped margin of the iris (black arrow) that occurs from dysgenesis of the posterior layer (neuroepithelium) of the peripheral iris, a poorly defined scleral spur and ciliary body band (yellow arrow), high insertion of the iris into the ciliary body band area, and a thickened trabecular meshwork.

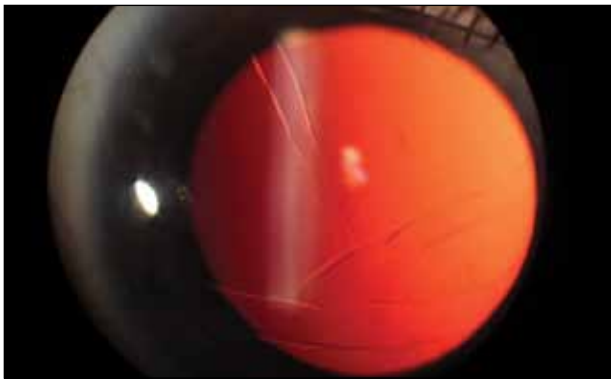

Figure 2. Haab striae. Elevated IOP during the first 3 years of life may stretch the cornea, rupturing Descemet membrane and causing corneal edema. When the IOP is normalized, the cornea repairs itself, the edema usually resolves, and a fine endothelial ridge of repaired tissue is best seen with retro illumination.

CLASSIFICATION OF PRIMARY CHILDHOOD GLAUCOMAS

- Congenital glaucoma (iridotrabeculodysgenesis or goniodysgenesis)
  - newborn or neonatal (birth-1 month of age)
  - infantile (1 month-2 years of age)
  - late congenital (2-4 years of age)
- Juvenile glaucoma (trabeculodysgenesis or normal-appearing angle), 4 to 35 years of age
- Glaucoma associated with anterior segment abnormalities
  - Axenfeld-Reiger
  - Aniridia
  - Congenital microcornea
  - Megalocornea
  - Ectropion uveae
  - Others
- Glaucoma associated with systemic abnormalities
  - Phakomatoses
  - Chromosomal disorders
  - Connective tissue disorders
  - Others

TAKE THE CONGENITAL GLAUCOMA QUIZ

1. The incidence of congenital glaucoma is highest in the Western nations.
   - True
   - False

2. Most cases of primary congenital glaucoma are unilateral.
   - True
   - False

3. Which topical glaucoma medication is it essential to avoid in congenital glaucoma (in patients up to 3 years of age) and why?
   - Acetazolamide
   - Brimonidine
   - Latanoprost
   - Timolol

4. Barkan's membrane is real.
   - True
   - False

5. The treatment of congenital glaucoma largely involves angle surgery and is more successful when the disease is discovered while patients are 3 to 12 months of age.
   - True
   - False

6. Elevated IOP causes the cornea to stretch until 3 years of age.
   - True
   - False

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7. Newborn primary congenital glaucoma confers a worse prognosis than infantile glaucoma.
   - True
   - False

8. Approximately 50% to 75% of patients with aniridia develop glaucoma, and most cases are usually familial in origin.
   - True
   - False

9. In a patient with aniridia, Wilms' tumor is more likely with sporadic inheritance.
   - True
   - False

10. What are the white spots on Figure 3I, and what is the likely diagnosis?
    - Brushfield spots
    - Lisch nodules

IMPORTANCE OF AGE OF ONSET

The reader should be aware that the age of onset of congenital glaucoma is one of the most important factors that influence the surgical outcome and, therefore, the patient's visual potential. Although it is difficult to classify the childhood glaucomas (see *Classification of Primary Childhood Glaucomas*), the attempt helps surgeons to formulate a treatment plan and prognosis. This article excludes secondary pediatric glaucomas (eg, trauma, retinoblastoma, uveitis, medulloepithelioma), because they are far too complex for this discussion.

HISTOPATHOLOGIC SEVERITY OF OUTFLOW AND GONIODYSGENESIS

Alvarado and colleagues correlated the histopathologic appearance of the outflow system with goniodysgenesis.[2] Severely compromised aqueous outflow at birth causes a marked elevation in IOP and a cloudy cornea that may result in Haab striae (Figure 2). A diagnosis of glaucoma within the first month of life signifies a guarded prognosis. The disease results from a highly maldeveloped outflow system (severe goniodysgenesis, type 1; see Table). These patients have only a rudimen-

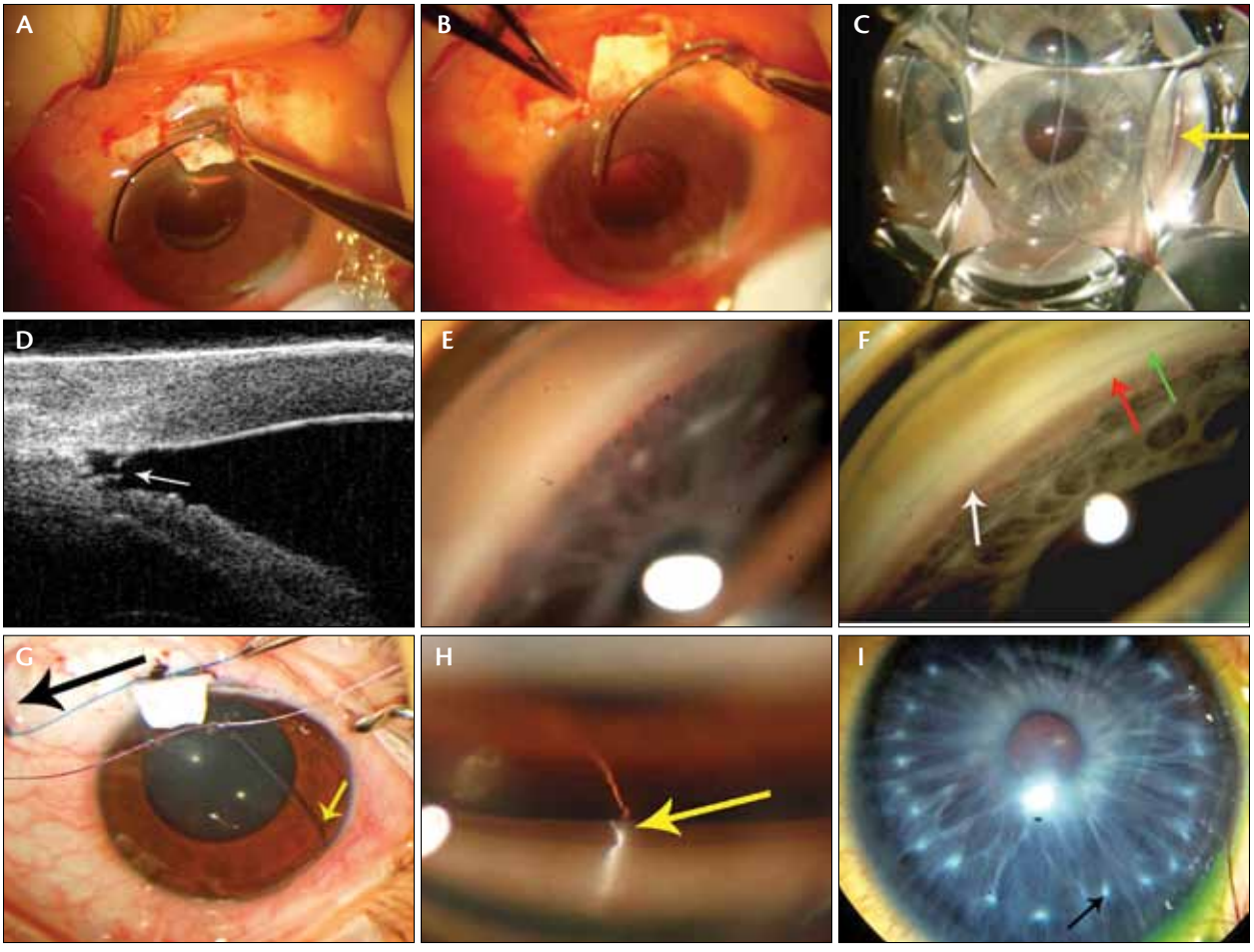

Figure 3. Evolution of trabeculotomy (segmental to circumferential). The surgeon positions a metal trabeculotome into a segment of Schlemm canal (A). Rotation of the trabeculotome opens 2 to 3 clock hours of the canal (B). Goniophotography shows the trabeculotome cleaving the angle and entering the anterior chamber. Note the blood (C). High-frequency ultrasound reveals the separation of the anterior and posterior trabecular leaflets after trabeculotomy with a metal trabeculotome (D). No landmarks are clearly visible in the trabeculodysgenic angle of a 6-month-old (E). Six months after 360° trabeculotomy with a Prolene suture in the eye shown in Figure 3E, a wide ciliary body band is present (white arrow). A red arrow indicates the scleral spur, and a green arrow indicates the trabecular meshwork (F). Circumferential suture trabeculotomy. As it is pulled in the direction of the black arrow, the Prolene suture cleaves open the angle in the direction of the yellow arrow for 360° (G). The eye shown in Figure 3G after a successful circumferential trabeculotomy. Note the shelf of white tissue (yellow arrow), which is the cleaved inner wall of the canal as it prolapses into the anterior chamber (H). The arrow points to one of many spots in the eye shown in Figure 3E and 3F. What are these spots, and what disease comes to mind (I)? (See *Take the Congenital Glaucoma Quiz*.)

TABLE. CORRELATION BETWEEN THE EXTENT OF GONIODYSGENESIS AND HISTOLOGICAL PHENOTYPE OF CONGENITAL GLAUCOMA

Phenotype	Extent of Goniodysgenesis	Histology
Type 1	Severe	Absence of the canal of Schlemm and deep scleral venous channels
Type 2	Moderate	Dense collagenous tissue bands in the corneoscleral or juxtacanalicular tissue
Type 3	Mild	Deposition of loose connective tissue (glycosaminoglycans) in juxtacanalicular meshwork

tary venous channel system and therefore require an artificial drainage system such as a drainage implant or filter. Infants with mildly compromised outflow channels will demonstrate signs and symptoms of congenital glaucoma several months after birth. These patients have a much better surgical prognosis, because their canal system has developed enough to allow for surgical cleavage. In contrast, in phenotype 1, the outflow channels have not formed enough for surgical correction.

If possible, especially in infants, I avoid trabeculectomy because it will require a lifetime of vigilance for blebitis, bleb dysesthesia, hypotony, and endophthalmitis.

SURGERY FOR PEDIATRIC GLAUCOMAS

The majority of pediatric glaucomas are due to abnormal development of the chamber angle with incomplete cleavage of the outflow structures associated with iridotrabeculodysgenesis. Types 2 and 3 dysgenesis may respond quite nicely to a trabeculotomy, a procedure designed to interiorize a maldeveloped canal system. Trabeculotomy has evolved from cleaving Schlemm canal with a metal trabeculotome to 360° trabeculotomy with a Prolene suture (Ethicon, Inc.)[3] to circumferential cleavage with an illuminated microcatheter (Figure 3 and see *Take the Congenital Glaucoma Quiz*).[4,5] If the microcatheter comes out of the canal and courses over the suprachoroidal space, it readily becomes visible beneath the sclera. It is impossible to perform trabeculotomy procedures successfully in infants with type 1 goniodysgenesis, because they do not have a canal system. These patients benefit from drainage device surgery.

Goniotomy and metal trabeculotome trabeculotomy produce similar results, and the former remains a staple for many surgeons. I prefer 360° trabeculotomy,[6] because the entire angle is opened at one sitting, circumferential return of the probe confirms the correct location of the canal, a cloudy cornea is not an impediment, and recent studies point to lower IOP with circumferential treatment.[5]

Even with excellent IOP control after trabeculotomy, the eye care professional must be wary of strabismus, amblyopia, cataract, and corneal disease. Thus, a multifaceted approach is necessary to achieve the triumph of good vision in these cases.

When angle surgery is unsuccessful, most glaucoma subspecialists turn to drainage implants, including valved and nonvalved devices. The major problem with tubes in children revolves around their revision. Strabismus may be a problem after the implantation of a drainage device. If an implant fails to control IOP, a second drainage device may be necessary, or improved types of cyclodestruction such as transscleral diode or endocyclophotocoagulation can be useful.

CONCLUSION

Knowledge gained through managing angle surgery in childhood glaucoma has spilled over into the treatment of adult glaucomas.[7] Several types of adult glaucomas respond well to circumferential trabeculotomy, which allows good IOP control without a bleb.[8,9] Increasing aqueous outflow through resident collector channels in all age groups remains an exciting avenue of glaucoma management, and it represents a significant advance in eye care. ■

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