

PEDIATRICS

The Challenge of Diagnosing Pediatric Glaucoma

BY ANNIE STUART, CONTRIBUTING WRITER

Glaucoma in children is not always a single disease that follows a predictable path. From anatomic defects and genetic syndromes to neoplastic and infectious processes, there are at least 100 pathologies that can include glaucoma in their presentation. Unfortunately, many of the tools used to diagnose glaucoma in adults may fail to detect the disease in children.

Why Early Diagnosis Is Critical

“If a glaucoma diagnosis is missed in an infant, the eye gets large and stretched out, the sclera becomes very thin, and the eye tends to collapse more easily with surgical treatment,” said Allen D. Beck, MD, professor of ophthalmology and director of the glaucoma section at Emory University in Atlanta.

“Finding landmarks during surgery becomes more difficult and the eye is prone to complications, even when surgery is otherwise successful,” he said, adding that it’s far preferable to diagnose the condition when it is most treatable and still possible to prevent sequelae like amblyopia.

Sharon F. Freedman, MD, agreed. To ensure early diagnosis, she and her staff follow two mottos:

1. Every child has glaucoma until proven otherwise.
2. The parent is always right.

Dr. Freedman is professor of ophthalmology and pediatrics and chief of the pediatric ophthalmology and strabismus service at Duke University

in Durham, N.C. Though a bit tongue in cheek, her first motto helps staff stay on guard to spot the signs of primary congenital glaucoma in infants: unusually large, hazy or sensitive eyes. The second motto arises from the regrettable fact that parents’ concerns about unusual eye features sometimes are dismissed, so their children are treated for an infection or blocked tear ducts instead of glaucoma.

It’s actually fortunate that glaucoma reveals signs in infants, said Dr. Beck, because screening at this age is difficult. Toddlers and school-age children, however, often lack clear external signs of glaucoma and are more often diagnosed through screening, especially if they were being screened because they have syndromes that are strongly associated with glaucoma, such as Sturge-Weber syndrome, neurofibromatosis or Axenfeld-Rieger syndrome.

Making a list and checking it twice. Dr. Beck advises looking for glaucoma in:

- Children whose sibling or parent had glaucoma in infancy or childhood. All siblings of a diagnosed child should be seen, since approximately 10 percent of cases of childhood glaucoma worldwide originate from autosomal recessive inheritance.
- Children with large myopic shifts during infancy.
- Children with a history of cataract.
- Any child with a port wine stain affecting the eyelid.
- Children with an ocular abnormality, such as aniridia.

Glaukos: Gleaming



Patient of Dr. Beck’s who presented with corneal hydrops caused by primary congenital glaucoma.

- Children with uveitis, trauma or intraocular neoplasm.
- Any child coming into the office for glasses. This is an opportunity to look at the size of the eye, check the clearness of the cornea, look at the optic nerve and check pressures.
- Any child exposed to high doses of steroids on the face.

Tools for Adults Are Tricky for Kids

Although new techniques have become available generally for the diagnosis of glaucoma, many are difficult to apply to children. “Those of us who take care of children look at what’s available in

the toolbox for adults and see which we can apply successfully to a child,” said Dr. Freedman.

Visual fields offer marginal help.

Something as simple as automated visual field testing to measure peripheral vision isn’t useful until a child reaches age 7 or 8, said Dr. Beck. Although the technology’s been around for a while and the algorithms are getting better, said Dr. Freedman, they’re difficult to apply to “trigger-happy” children. “Little kids look around, can’t sit still, can’t reach the buttons,” she said. “I usually tell the parents, ‘The first time we do this, we’ll just practice.’”

Pressures are elusive. Even measuring IOP in a pediatric patient is challenging, especially in the birth to 3-years-old range. “It’s hard enough to get a pressure when someone’s cooperative, much less when someone is screaming and squirming,” said Dr. Beck. “More likely, you have to do exams under anesthesia to assess the disease.”

The Perkins handheld applanation tonometer or the Tono-Pen have typically been used to obtain pressures in children. **But one newer device, the rebound tonometer by Icare, is especially helpful, said Dr. Freedman. Working on a different principle than other tonometers, it’s easier to use on a child and lowers the frequency of exams under general anesthesia. Anesthesia is costly, exposes the child to well-known risks and can sometimes falsely alter**

the awake pressure readings.

“The tiny tip moves on and off the surface of the eye so quickly that you may not feel it, or may perceive it as simply a minimal ‘tickle,’ causing an extra blink,” said Dr. Freedman.

The good news is there’s no need to use a stinging anesthetic beforehand or to hold the eye open. The pressure can be measured between blinks while a child is watching television. Although not yet marketed for this use, the Icare tonometer may be used in special home situations to monitor suspected volatile pressures throughout the day. “Being able to check pressure painlessly and faithfully at home in selected cases might allow easier acquisition of pressure data,” said Dr. Freedman, referring to cases where pressure measured in the office appears fine, yet the optic nerve is inexplicably getting injured.

This challenge in getting readings in children is a reason pressure is just one aspect of the glaucoma assessment, said Dr. Beck. “We also frequently measure corneal diameters and the axial length of the eye, as well as look at the optic nerve, and then use all of these to assess stability.”

Pachymetry needs a grain of salt.

Dr. Freedman agreed, but cautioned against relying too heavily on the connection between central corneal thickness and measured pressure. If a cornea is thinner due to genetics, you may underestimate pressure, she said. “I would strongly caution my colleagues against feeling reassured because a child with known glaucoma has a thick cornea,” she said, adding that other factors may also influence pressure, including edema and the elasticity of the cornea. In adults, a thicker cornea may correlate with a thicker support network of the optic nerve at the back of the eye, a possible explanation for the correlation between thicker corneas and a lower risk for glaucoma damage shown in some adult studies.

OCT useful? Maybe. Together with the gold standard—photographing the nerve head—newer technologies such as optical coherence tomography can look at nerve fibers on the surface of

the retina, just around the optic nerve or even in the macula. This may be very useful in adults, said Dr. Freedman, but is often difficult to apply in children who can’t sit still for the test and may also have nystagmus.

“You’ve really got to depend on your clinical skills to figure out what’s happening with the child,” said Dr. Beck. Dr. Freedman said that OCT gives one more set of objective measurements that can be very good for kids age 5 and up, and it’s much easier than taking a visual field test. “Some children are fine with a pressure of 20 mmHg. Others may need a pressure of 15 to resist further optic nerve damage. Sorting this out is very difficult.” Spectral domain OCT may be more helpful, said Dr. Freedman. This high-definition technology takes less time to obtain data, producing clear pictures quickly in children with nystagmus or who are unable to sit still for a standard OCT.

After Diagnosis: Management

Treatment protocols applied to this very heterogeneous group are, at best, very complex flow diagrams, said Dr. Freedman. “Does the child weigh more than 50 pounds—yes/no? Will the child put in eyedrops at school—yes/no? Does the child have asthma—yes/no? How low do I need the pressure? And so forth.”

Hey Doc, I’m a kid. As with all things pediatric, it’s essential to remember that kids are not little bitty adults, said Dr. Beck. They might have systemic responses to eyedrops, for instance, or not respond well to treatment due to a congenital anomaly.

Many ophthalmologists avoid pediatric glaucoma because it clearly takes a lot of time and is difficult, said Dr. Beck. “But to me, it’s always worth the effort. It may not affect a huge number of patients, but it will make a huge difference in whether this child goes on to be an adult who can see and function in society.”

Dr. Beck has received research support from Alcon and Pfizer. Dr. Freedman reports no related financial interests.

Squirmy Patient?

Dr. Freedman offered these tips for a successful pediatric exam:

- Have the parent bring the child in hungry and provide a bottle or snack during the exam.
- Help the parent relax and be calm.
- Put the parent and child in a comfortable position.
- Coach the person who is doing the testing to develop a level, unhurried mind-set. Although 10 patients are waiting, the clock stops for kids.
- Offer the child a post-exam treat.