Glaucoma for Children

What is glaucoma?

Glaucoma is a group of diseases characterized by damage to the optic nerve that often occurs when the eye pressure is too high. This optic nerve damage can eventually result in severe vision loss. Most often the pressure is too high because the eye is able to make the fluid it needs but is unable to sufficiently drain the fluid out. Glaucoma is more common in the elderly but can develop at any age. Infants and children with glaucoma typically have different signs and symptoms than adults.

What are the different types and causes of pediatric glaucoma?

One way to classify glaucoma is based on the age of onset. Congenital glaucoma is present at birth. Infantile glaucoma develops between the ages of 1-24 months. Glaucoma with onset after age 3 years is called juvenile glaucoma. Another way to classify glaucoma is to describe the structural abnormality or systemic condition which underlies the glaucoma.

Many cases of pediatric glaucoma have no specific identifiable cause and are considered primary glaucoma. When glaucoma is caused by, or associated with a specific condition or disease, it is called secondary glaucoma. Examples of conditions which can be associated with childhood glaucoma include Axenfeld-Reiger Syndome, aniridia, Sturge-Weber Syndrome, neurofibromatosis, chronic steroid use, trauma, or previous eye surgery such as childhood cataract removal. Not all patients with these conditions will develop glaucoma, but their incidence of glaucoma is much higher than average, and they should be monitored regularly.

How common is pediatric glaucoma?

Childhood glaucoma is relatively rare. Primary congenital/ primary infantile glaucoma occurs in the general population at a rate of approximately 1 in 10,000 births. However, if a child has cataract surgery or one of the other conditions listed above, the incidence of glaucoma will be much higher. For example, 50% of patients with aniridia will develop glaucoma during their lifetime.

Is pediatric glaucoma hereditary?

Some types of pediatric glaucoma are hereditary. About 10% of primary congenital/infantile glaucoma cases are inherited. Recent research has identified some specific gene mutations linked to this disease; for which genetic testing and counseling for affected families is may be available.

Other conditions that cause secondary glaucoma can be inherited. For example, neurofibromatosis and aniridia are dominantly inherited and are passed on to the children of

affected individuals approximately 50% of the time. The incidence of glaucoma that occurs in association with these conditions, however, is less predictable.

What are the symptoms of congenital/infantile glaucoma?

The most common symptoms of congenital/infantile glaucoma are excessive tearing, light sensitivity and a large, cloudy cornea (the normally clear front surface of the eye) which can cause the iris (colored part of the eye) to appear dull. Excessive tearing accompanied by mattering/discharge in a child is usually not caused by glaucoma but instead is the result of congenital nasolacrimal duct obstruction (blocked tear duct).

Enlarged, cloudy corneas in a child with congenital glaucoma

What are the symptoms of juvenile glaucoma?

Juvenile glaucoma tends to develop without any obvious symptoms, similar to adult glaucoma. Patients with juvenile glaucoma often have a positive family history. On exam the eye pressure will typically be elevated and there may be signs of optic nerve cupping (enlargement of the center “cup” portion of the optic nerve).

Fig. 1: Glaucoma examination.

How is pediatric glaucoma evaluated and diagnosed?

There are several components to the examination that help the ophthalmologist determine the nature and extent of glaucoma. Depending on the age and cooperation of the child, some or all of these components may be better evaluated while the child is under anesthesia. Specifically, the doctor will evaluate the intraocular pressure corneal diameter (for increased size), corneal clarity (for cloudiness and/or Haabs striae, breaks in the back surface of the cornea), corneal thickness (which can potentially change the accuracy of pressure measurements), the drainage angle of the eye (using a technique called gonioscopy), axial length (for elongation of the eye caused by increased pressure), refractive error (for near-sightedness, also caused by elongation and the optic nerve (for abnormal cupping indicative of optic nerve damage Older children may also be able to participate in a diagnostic visual field test to evaluate peripheral vision. This can help

determine if there has been any significant optic nerve damage. Additional imaging tests, such as Optical Coherence Tomography (OCT), a test that looks for thinning of the actual nerve fibers, can be useful for detecting more subtle changes in the progression of the disease.

How is pediatric glaucoma treated?

Pediatric glaucoma is treated by lowering the intraocular pressure (IOP) via medical and/or surgical means. Most cases of primary pediatric glaucoma are treated with surgery. Trabeculotomy and goniotomy, which open the drainage canals, are the most common surgical interventions. Other procedures create a bypass route for the aqueous (fluid made by the eye) to drain out of the eye.

A trabeculectomy creates a guarded opening from the front of the eye to a space underneath the conjunctiva- which is the clear membrane that covers the sclera (the white part of the eye). A tube shunt is a device which is inserted into the front of the eye or into the vitreous cavity (back part of the eye). Fluid from the eye then drains to a reservoir that is located underneath the conjunctiva. Laser procedures can also be beneficial in some cases. Control of the glaucoma often requires multiple procedures and examinations under anesthesia. Eye drops, and oral medications are the primary treatments for secondary and juvenile glaucoma and are often used as adjuvant therapy after surgery in primary pediatric glaucoma.

One or more medications may be necessary to control the intraocular pressure (IOP), even after surgery. Achieving the best vision in patients with pediatric glaucoma is not solely dependent on successful reduction of IOP. Many children with pediatric glaucoma develop myopia (nearsightedness) and require glasses. Also, amblyopia (“lazy” vision) and strabismus (misalignment of the eyes) occur more frequently and may require treatment with patching or surgery.

Despite timely and aggressive treatment, pediatric glaucoma can still cause significant and permanent vision loss. Early diagnosis and treatment, as well as close monitoring are crucial for obtaining a long-term successful outcome.

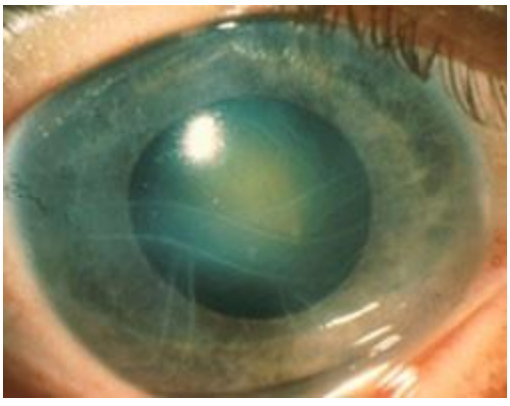

Fig. 2: Multiple back surface corneal breaks (Haab striae) in a child with congenital glaucoma.

Associated information site: Pediatric Glaucoma and Cataract Family Association (PGCFA)

More technical information may be found on the EyeWiki Site.

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