

CHILDHOOD GLAUCOMA

Prepared by The Childhood Glaucoma Research Network (CGRN) (rev. 03/12)

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Introduction – What is Childhood Glaucoma?

Childhood glaucoma is a condition of childhood in which the optic nerve (nerve that goes from the eye to the brain) is damaged from high pressure in the eye. It can be present at birth or be diagnosed throughout childhood. Childhood glaucoma can cause permanent vision loss if left untreated.

Damage to the eye in children with glaucoma is almost always the result of raised intraocular pressure (IOP) in the eye. The inside of the eye is constantly making and draining fluid that fills the eye, providing nutrients and oxygen. Too little fluid being drained from the eye can cause the fluid to build up and thus the pressure in the eye to increase (think of it like pressure in a water balloon – the more water that is put inside the balloon, the higher the pressure rises inside the balloon). This rise in pressure is most often the result of a problem with the drainage channels in the eye. The most common type of childhood glaucoma is Primary congenital glaucoma in which the drainage channels do not develop properly. This condition is usually not inherited. Abnormal drainage can also be the result of other associated conditions described below.

When glaucoma occurs at a young age (usually <3 years old) it can cause the eye to stretch and grow. This can make the eye appear larger, the cornea (clear window of the eye) to become larger and more cloudy and the eye to become longer. Patients with childhood glaucoma may be light sensitive and have watering of the eyes. When childhood glaucoma occurs over the age of about 4 years, the patients may have fewer symptoms and the diagnosis must rely more on findings during an eye examination. The primary finding is evidence of optic nerve damage and increased pressure in the eyes; many children also become myopic (near-sighted) as their eyes stretch under elevated pressures.

Childhood glaucoma is an uncommon but serious disorder. The impact on a child's visual development can be profound. Early diagnosis with prompt and appropriate treatment by an ophthalmologist who specializes in treating childhood glaucoma, are keys to success in preserving the sight of children with glaucoma. Some children with glaucoma can have near normal vision. The most important thing is control of the eye pressure as early as possible followed by with the use of glasses for refractive errors (near sightedness / astigmatism) and patching if there is evidence of the eye/s being 'lazy' to ensure that the vision develops normally. As the child grows older the specialist will have a better idea of how good their long-term vision will be. Lifelong follow-up is a must.

How is it diagnosed?

The diagnosis of childhood glaucoma is confirmed by an ophthalmologist based on the eye examination. In young patients a thorough examination may require the child to have general anesthesia. Many children after the age of 5 years - and even some children as young as 2-3 years of age - can cooperate with examinations awake in the doctor's office.

Glaucoma is diagnosed based on certain eye examination findings:

- Increased eye pressure (there are various instruments that are used to measure this).
- Optic nerve damage diagnosed based on the appearance of the nerve. Photos can be taken to follow the appearance over time.
- Lines in the cornea (called Haab striae), which are stretch marks from the increased pressure.
- Increase in the length of the eye measured by ultrasound and glasses prescription (nearsightedness).
- Peripheral vision loss. This may be impossible to measure until a child is old enough (usually after 5 years of age) to cooperate with a formal side vision test (visual field).
- Abnormal appearance to the drainage system of the eye. This can be diagnosed only by using a special lens on the surface of the eye (gonioscopy).

Incidence

Primary congenital glaucoma (glaucoma without other eye or systemic abnormalities) has an incidence of around 1 in 10,000 – 1 in 20,000 live births in western developed countries. In certain ethnic groups and countries the rate is much higher. It occurs in both eyes in approximately 75% of cases. In approximately 50% of cases, glaucoma in children is associated with other eye or systemic disorders.

Associated conditions

Glaucoma can occur as an isolated disease or can be associated with other abnormalities of the eye or body. In patients with childhood glaucoma, a thorough eye exam and good history of the patient and family are important. Communication with the pediatrician can also be useful in identifying other body abnormalities.

Associated disorders include (partial list):

- Glaucoma following congenital cataract surgery— A cataract is clouding of the lens of the eye. Glaucoma can occur days, months or years after congenital cataract removal surgery, especially in younger children, those with small eyes and certain kinds of cataracts. The risk is life-long.
- Aniridia— No or very little development of the iris, which is the colored part of eye.
- Axenfeld-Rieger anomaly and syndrome and other types of abnormality in the development of the front portion of the eye.
- Sturge-Weber Syndrome — Blood vessel birthmark on the face, called a ‘port-wine’ birthmark is sometimes associated with brain abnormalities. The eye on the same side as the birthmark is at risk for glaucoma.
- Neurofibromatosis Type I — A disorder that causes tumors to grow in the nervous system. These patients have skin changes, eye changes, brown birthmarks (café-au-lait spots) and bone abnormalities.
- Uveitis and iritis— This is inflammation inside the eye from a variety of causes. The inflammation affects the drainage system of the eye causing glaucoma.
- Trauma— Injury to the eye can affect the drainage system of the eye resulting in glaucoma in some patients.

There are a multitude of other disorders which can be associated with glaucoma but which are too numerous to fully discuss here.

Medical treatment

There are various eye drops and oral medications that can be used to manage glaucoma. In those born with glaucoma or those who develop it shortly after birth, glaucoma drops alone are usually not enough to treat the disease and surgery is needed.

Glaucoma drops and oral medications may also be needed after surgery. The goal of medications and surgery is to lower the eye pressure to a point where no further nerve damage is likely to occur. The eye drops work in two main ways - either by reducing the amount of fluid made in the eye (like slowing down the water coming out of a faucet) or by increasing the rate at which fluid drains out from the eye (like unclogging a plugged drain). The choice of eye drop medications depends on many factors including the age of the child, the presence of other eye abnormalities, and other health problems the child may have. Each drop has its own dosing schedule that may range from 1-4 times daily. Oral medications are usually a second choice to drops and if necessary may be used short term before surgery. In addition to glaucoma drops, other drop medications can be used (for example, to help to treat inflammation or dilate the pupil).

Surgical treatment

The decision to perform surgery and the type of surgery chosen is based on the specific findings in each patient's eye. There are several glaucoma surgeries that can be performed. Some eyes may require more than one surgery to help control the glaucoma.

- Goniotomy—A fine knife or needle is used to open the drainage channel (trabecular meshwork) that did not properly develop or became blocked by disease.
- Trabeculotomy— A fine probe is passed through the drainage system (Schlemm canal) from the outside of the eye and then pulled to create a connection between the drainage system and the front of the eye.
- Trabeculectomy—A small tunnel is created to make a new pathway (like a by-pass drain) for the fluid to leave the eye. The fluid from the front of the eye drains under conjunctiva (the fine clear layer covering the white of the eye). This new pocket of fluid is then absorbed. In some cases the new pocket can scar closed and thus a special medication (mitomycin) is used during surgery to prevent scarring.
- Drainage device— This procedure uses a special drainage tube that connects the fluid in the front of the eye to a pocket held open by the drainage tube under the conjunctiva. These tubes (sometimes called shunts or valves) have a variety of names such as Ahmed, Molteno, and Baerveldt.
- Laser—In some cases laser treatment is performed to damage the tissue in the eye making the fluid resulting in less fluid being produced. This laser can be done from the outside of the eye (contact laser) or with a special camera inside the eye (endoscopic laser). This treatment often needs to be repeated to help control the pressure.
- Cryotherapy - Very similar to laser but uses a freezing probe instead of laser to damage the fluid-making tissue (ciliary body) inside the eye.

There are many other variations on the operations listed here.

Follow-up

It's important that parents, caregivers, and health professionals work together in the care of a child with glaucoma. Frequent follow-up visits are needed after surgery. Children who have been diagnosed with childhood glaucoma must continue to receive follow-up care throughout their lives. Elevated eye pressure (IOP) can recur at any age in a child with glaucoma. At follow-up visits, the doctor will assess the child's vision, and check for amblyopia (lazy eye). They will also see if the eye has enlarged and whether there is any redness or clouding of the eye. The pressure of the eye (IOP) will also be measured.

Implications on development

The effect of glaucoma on vision is generally difficult to predict. In young children when the connection between the eye and brain is still developing, having a clear and sharp image becomes especially important. The effect of glaucoma on vision depends on the extent of damage to the optic nerve, the age at which it occurs, and how well treatment controls the eye pressure (IOP).

However, even when IOP is well controlled, approximately 50% of children with Primary congenital glaucoma do not achieve vision better than 20/50. Reduced vision can occur due to a number of reasons in addition to those mentioned above. Corneal damage from scarring, Haab striae and prolonged swelling with clouding can affect vision. Large refractive errors impair vision, but can be corrected or improved with glasses. Some patients with glaucoma may also have other eye abnormalities that could also affect vision e.g. cataract, eye misalignment (strabismus), shaking of the eyes (nystagmus), peripheral vision loss and lazy eye (amblyopia) to name a few.

Today, many children with glaucoma have good visual outcomes and develop normally. Yet, some children have more complex conditions that are associated with low vision. Those with low vision can obtain special vision services to help them best use the vision they have, including using enlarged print, extra lighting and special vision aids. Many resources are available to children and families of those with vision difficulties. It is important for families and teachers to foster the child's independence and self-confidence by normalizing their environments and routine. For more information, go to www.gl-foundation.org and find additional information and links to support organizations.