



Aniridia and Glaucoma

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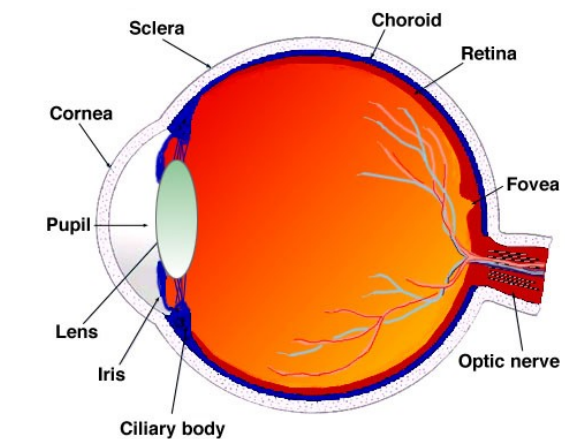
Glaucoma occurs in up to 50% or more of people with Aniridia. It may occur at any time in life, from birth through adulthood, but is probably more common in adults.

Below you will find information, diagrams, and photographs describing this common and very challenging problem.

What Is Glaucoma?

In the normal eye, there is a small amount of fluid between the iris and the cornea. This fluid is called the **Aqueous Humor**. By regulating the amount of fluid that is produced and the amount that is drained, the structures of the eye maintain the aqueous humor at a constant pressure (usually below 22mm Hg)

In people with Aniridia, these structures may be under-developed or poorly formed, or at some point may lose



their ability to function. If the fluid is unable to drain properly, the pressure within the eye will begin to increase. This is called "Increased Intraocular Pressure." Glaucoma occurs when this pressure is high enough to cause damage to the eye. If left untreated, Glaucoma can cause blindness. While there are effective treatments for Glaucoma, the unique anatomy of the aniridic eye may make it very difficult to treat. If a person with Aniridia develops glaucoma,

referral to an experienced specialist is very important.

Tonometry

The tonometry test measures the inner pressure of the eye (called the **IOP**, or **IntraOcular P** ressure) Usually, drops are used to numb the surface of the eye. Then the doctor places a special device directly on the cornea. This gives a pressure reading. Two devices commonly used for this are the TonoPen, and the Perkins Tonometer. These devices give a more accurate measurement than the "puff of air" machines, which are used for routine screening.



Special Challenges In Children

Measuring eye pressure in young children is often very difficult, because they cannot cooperate with the examination. If a child is

How Is Glaucoma Diagnosed?

Several basic tests are used to examine the eye, to determine the status of the optic nerve or optic discs, and to measure the pressure within the eye. These tests are:

- 1) Tonometry**--to measure the pressure within the eye
- 2) Ophthalmoscopy**--to examine the structures at the back of the eye
- 3) Gonioscopy**--to examine the structures near the front of the eye.



crying when his or her eye pressure is measured, the reading will appear to be higher than what it really is. Some physicians suggest using a mild sedative (such as

Chloral Hydrate, which is given by mouth, or Ketamine, which is given by injection) These drugs can be used in the physician's office, but may not work well for some children, depending on their age or general health status. Another option for getting a pressure reading in children is the

" Exam Under Anesthesia" or EUA .

This is an examination done under general anesthesia. Unlike a surgical procedure, only a small amount of anesthesia is needed for an EUA, and only for a short period of time. Another benefit of the

Ophthalmoscopy



Ophthalmoscopy is used to examine the inside of the eye, especially the optic nerve and optic discs. In a darkened room, the doctor will look at the eye with an ophthalmoscope (an instrument with a small light on the end). This helps the doctor see the shape and color of the optic nerve and the other structures at the back of the eye.

EUA is that it allows the physician to conduct a very thorough, complete examination of the whole eye. It should be noted, though, that some drugs used for anesthesia can cause the eye pressure to appear *lower* than what it really is.

If crying elevates eye pressure, and general anesthesia can lower it, how does a physician tell what a child's eye pressure really is?

This is usually done by monitoring the pressure readings over several visits, and by monitoring changes in the appearance of the structures within the eye.

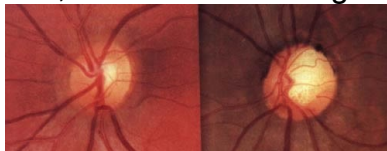


What the doctor sees with the ophthalmoscope

This picture above is what a normal **Retina** looks like through the ophthalmoscope. The doughnut-shaped, bright circle on the left side is the **Optic Disc** (the head of the optic nerve)

The darker spot to the right of the optic disc is the **Macula** (the part of the retina responsible for clearest vision)

Normal Optic Disc/Glaucoma. On the left is a normal optic disc (the doughnut-shaped ring in the center) with a thick outer ring of nerve tissue. The optic cup is small. On the right is an optic nerve showing the changes characteristic of glaucoma. The outer ring is thin, and the hole is larger.



Gonioscopy

Gonioscopy is an eye exam to look at the anterior chamber of the eye, which is the front part of the eye between the cornea and the iris (or, in people with Aniridia, the area where the iris should be) Gonioscopy is used to determine whether this area is damaged, blocked, or clogged.



What Is A "Normal" Pressure Reading?

"Normal" eye pressure is usually quoted as being somewhere between 8 and 22 (mmHg)

But every individual's optic nerves are different. So some people may begin to show signs of damage from an IOP (intraocular pressure) of 28, while others may not show damage until the pressure reading is much higher.

The decision about whether to diagnose and/or treat for glaucoma does not rest solely on the pressure reading. A combination of factors, such as the IOP, the appearance of the optic nerve, or changes in the appearance of the optic nerve over time, are all taken into account by the physician.

High intraocular pressure is abnormal, though, and should be followed closely to detect early optic nerve damage. Some patients with high pressures may need treatment to reduce the risk, even though their optic nerves remain normal.

Once Glaucoma Is Diagnosed, How Is It Treated?

Glaucoma is treated with medication (usually eye drops) or surgery, or in some cases, both. Both eye drops and surgery work by helping the fluid to drain from the eye, or by decreasing the amount of fluid produced by the eye.

Glaucoma medications come in different strengths and combinations. Doctors try to use the smallest amount of medication that offers the best results with the fewest

side effects.

Medications must be taken on a daily, regular basis to control the pressure in the eye. Most medications have some side effects, but very often these side effects lessen after a few weeks.

Medications. The four main families of medications are: beta blockers (**Timoptic, Betoptic**), carbonic anhydrase inhibitors (**Trusopt, Azopt**), alpha agonists (**Alphagan, lopicidine**) and prostaglandin (**Xalatan**)
The first three types of medications decrease

production of fluid in the eye, while prostaglandin helps fluid leave the eye through a different pathway located at the back of the eye.

Laser procedures. An argon laser may be used to perform a procedure called a trabeculoplasty. The laser is focused into the trabecular meshwork (see the "anatomy of the eye" diagram, above) where it alters the cells there to let aqueous fluid leave the eye more efficiently.

Implantation of a "Glaucoma Drainage Device." This type of procedure involves placing a drainage tube into the eye. The tube exits at the junction of the cornea and sclera (the white portion of the eye) This may be the most effective procedure for people with aniridic glaucoma. There are different types of glaucoma implants or glaucoma drainage devices (see photo below) The most common of these are the **Molteno** implant, the **Baerveldt** implant, and the **Ahmed** implant.

With these devices, a plastic tube is inserted into the front of the eye

behind the cornea. The tube is like an artificial drain, and fluid passes through it to the outer layers of the eye.



Glaucoma implants come in many shapes and sizes. A clear thin tube (at the base) is placed into the front part of the eye, and is connected to a reservoir (white) outside the eye. After the implant procedure, the reservoir will not be visible except to a physician.

Can Glaucoma Be Prevented in People With Aniridia?

There is a procedure called a "Preventative Goniotomy." In this procedure, the drainage part of the eye is surgically incised (opened) This procedure may be very effective for preventing glaucoma in people with Aniridia, but whether a patient is a good candidate for this surgery depends on many factors.

The physician who pioneered this approach will discuss it with

interested patients,
parents, and physicians.

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An abstract of the 1999
article describing
Preventative Goniotomy
can be found here:

[http://archophth.ama-
assn.org/issues/v117n9/
abs/ecs80019.html](http://archophth.ama-assn.org/issues/v117n9/abs/ecs80019.html)>

**Thank you to Dr. Peter
Netland
for his assistance with
this article.**

**Need More Information
and Support?**

These organizations
can help:

**The International
Glaucoma Association**
<http://www.iga.org.uk/ig>

...."Medical Information presented in this newsletter is
for informational purposes only. Please consult your
physician if you have questions or concerns regarding
your child's health."

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The National Eye
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ealth/glaucoma/glaucom
a_facts.htm\]\(http://www.nei.nih.gov/health/glaucoma/glaucoma_facts.htm\)](http://www.nei.nih.gov/health/glaucoma/glaucoma_facts.htm)

**The Glaucoma
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**Just For Kids
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coma.com/index.html](http://www.childrensglaucoma.com/index.html)
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and Cataract Family
Association.**
www.pgcf.org