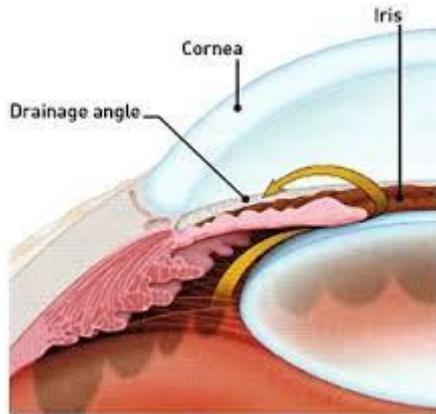


## Childhood Glaucoma – Information

Glaucoma is an eye disease where high pressure builds up inside the eye.

It is usually a problem that affects the elderly. However, rarely, it can occur in children.



**Aqueous Fluid** is internal eye fluid created and drained at a low, constant rate to maintain a healthy eye.

It is created behind the iris by the ciliary body and bathes the lens, passes through the pupil and drains out through the drainage angle. This angle is a 360-degree structure deep under the junction between the clear cornea and white scleral coat of the eye.

Aqueous fluid is separate to tear production and drainage.

The **common causes** of glaucoma in children are:

1. Isolated failure of the drainage angle also known as the **trabecular meshwork**
2. Structural change in the front of the eye
3. Baby or infant cataract surgery
4. High pressure in the aqueous-draining vein

Raised eye pressure affects the eye in several ways, most importantly:

1. In very young children (usually under 3 years of age) raised pressure causes the eye to **enlarge**. This stretches the cornea the front clear window of the eye) and the cornea can scar, or suddenly swell and appear cloudy. Any scar or swelling can make a child very light sensitive.
2. In all children the **optic nerve** is sensitive to pressure when it is excessively high for a long period of time. The higher the pressure, the more quickly damage occurs. When the nerve is damaged, vision is lost. There is currently no way of repairing an optic nerve once damage has occurred.

## Types of Childhood Glaucoma

**Primary Congenital Glaucoma (PCG)** is the term given to glaucoma that starts in the first couple of years of life. It can be detected at birth, but often takes many months or even years to become obvious. A prominent feature is enlargement or excessive growth of the eye.

### Primary Childhood Open Angle Glaucoma / Juvenile Open Onset Glaucoma

When childhood glaucoma commences after 3 years of age the high eye pressure will not usually enlarge the eye. Rather the affected eye may have unequal focus or be very short-sighted. These lead to a preference for the other eye and development of poor vision or amblyopia (lazy eye). If high pressures persist for months or years, before the problem is found, the optic nerve becomes damaged (“cupped”) and substantial vision is lost.

### Secondary Glaucoma

Eyes that have unusual features such as underdeveloped or incomplete iris, irregular pupils and wobbly lens are at risk of glaucoma. This is known as secondary glaucoma.

Another cause for childhood glaucoma is a history of **early cataract surgery**. Typically, this relates to cataract surgery in the first months of life.

Glaucoma may result from the same process that caused the cataract, or from the cataract surgery itself. The risk is greatest for a smaller than normal eye (microphthalmia) and those that need cataract surgery under 3 months of age.

Severe eye injuries or **eye trauma** can also cause glaucoma, sometimes many years later.

The drainage of the aqueous fluid can be impeded and pressure in the eye rises if there are unusual capillaries and veins as part of the blood vessels around the eye. This is classically seen in infants and children with **Port Wine Stain** birthmarks. If this is associated with changes in the blood vessels around the brain and possible seizures it is known as **Sturge Weber Syndrome**.

## How is an eye assessed for glaucoma?

At different childhood ages and developmental stages, the techniques to assess for glaucoma change.

Infants with uncertainty around the diagnosis may require an **examination under anaesthetic (EUA)**. If a child is too distressed to be examined, even when fed and well rested, an EUA may be required to find out what is going on, and initiate treatment.

For follow-up visits, patients are seen by an **orthoptist**, who is an experienced eye care professional who assesses children of all ages. The orthoptist will assess the vision (acuity) in each eye and ocular alignment. They may administer drops to help check refraction (focus).

An **ophthalmologist** will assess the eye pressure and measure a child's refraction. Refraction involves the process of assessing for glasses. In children the application of dilating eye drops assist in accurate measurement of refractive error.

### What extra testing can be done, and why?

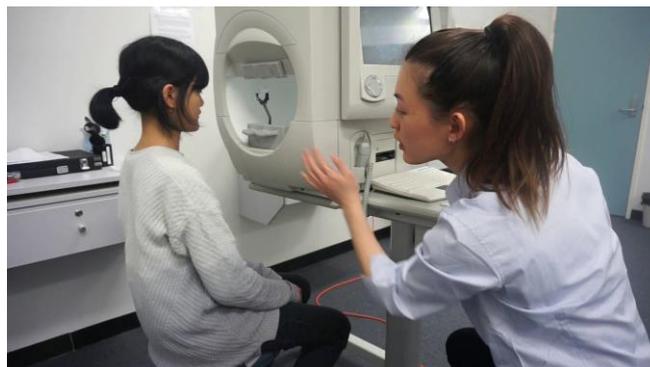
To show greater detail surrounding the health of the optic nerve and retina and to detect the presence of glaucoma worsening (progression), extra tests are performed in an age appropriate manner.

By the age of 3 or 4 many children can keep still for a brief **photo** of their optic nerves at the back of their eyes. Ideally this should be performed every 6 months.

Around age 5 or 6 the “photo” can be an **OCT scan** which shows, in greater detail, the health of the retina and optic nerve.

By age 10 it is common to start performing automated visual field tests (typically with the Humphrey field machine pictured) to further assess vision function.

Typically, these are yearly tests, but can be more common if the disease is unstable. Sometimes, the patient performing the test takes a couple of attempts to master it.



### How do you measure eye pressure in children?

Children can be distracted and without using any eye drops, an iCare tonometer (see picture) gently taps the eye to measure its pressure.

Those that do not have regular eye drops, are comfortable, fed and rested usually can be distracted while awake to allow measurement of their eye pressure. Some young babies need to be fed and allowed to sleep to attempt measurement.



## Is glaucoma treated with drops, medicine, or surgery?

**Eye drops** are a large part of glaucoma treatment in adults and particularly glaucoma in older children. There are many different types; some are given once a day, others twice a day. Some contain one agent whilst others contain a combination of two agents.

In children, **latanoprost** (XALATAN) is a common treatment drop, it is instilled once a day and usually before bedtime. It may cause some redness, darkening of the coloured iris as years pass, and longer eyelashes.

Another common drop is **timolol** (TIMOPTOL, TIMOPTOL XE, TENOPT). This can be instilled once or twice a day. This drop can worsen asthma or cause sleepiness. It should be stopped if a child has a prolonged cough or wheeze.

It is important to follow your doctor's instructions when using these drops.

If a drop is missed, please give the catch-up dose as soon as possible.

## Diamox (acetazolamide) Syrup or Tablets

This is a medicine that reduces eye pressure. It also causes a child to create more urine. They can become thirsty, and possibly grumpy and irritable. There may be more wet nappies or toilet training may worsen. In older children, as fluid is lost, potassium in the blood can decrease. It is wise to discuss how to replace this fluid and potassium by **drinking more and eating potassium rich food like bananas, oranges, apricots and raisins.**

**Diamox in syrup form** is generally only available from compounding pharmacies. Syrup can be mixed with food or given alone.

Wood's Pharmacy is a compounding pharmacy at the Royal Children's Hospital (you can call ahead to check they have it available on 93492315) The RCH outpatient Pharmacy can also make up Diamox syrup.



The syrup is typically 25mg per ml, and a typical dose may be 1-2mls three times a day in young infants. Ideally, large bottles with greater concentration (50mg per ml) are used for older children.

For older children, Diamox 250mg tablets can be easily halved, and they can be broken up and disguised in a sweet food.

Usually, Diamox is used only for a short time up to a few weeks or a few months around surgery. Its side effects on mood, increased risk of kidney stones and long-term taste distortion mean that the medicine is only rarely a long-term treatment option.

## Surgery for Childhood Glaucoma

There are several options for surgery in childhood glaucoma. They vary in duration and complexity. Your Ophthalmologist will discuss the approach best suited for your child.

### Requirements for all surgery:

1. FASTING – for any general anaesthetic short or long duration, your child will need to fast. You will be advised on timing when you are contacted by the pre-operative team.
2. GLAUCOMA MEDICATIONS – please give these as normal **the day before** the procedure. Do not take them on the day of surgery unless advised otherwise.
3. CHECK IN ON TIME – please arrive on time to meet the pre-admission clerk, nurses and the anaesthetist caring for your child

### Day stay or overnight stay?

Most children older than 6 months can be discharged home with parents/carer after surgery. It is very common to need a check-up the next day. For patients that live more than an hour from the hospital, it may be possible in some circumstances to arrange an overnight bed. Other alternatives are the care-by-parent ward of the Royal Children's Hospital, or local hotels.

Typically, children under 6 months of age need monitoring overnight in hospital following their anaesthetic. There may be exceptions, for shorter procedures, and larger healthy infants who are observed for a few hours in the theatre recovery suite before discharge.

## Examination under anaesthetic (EUA)

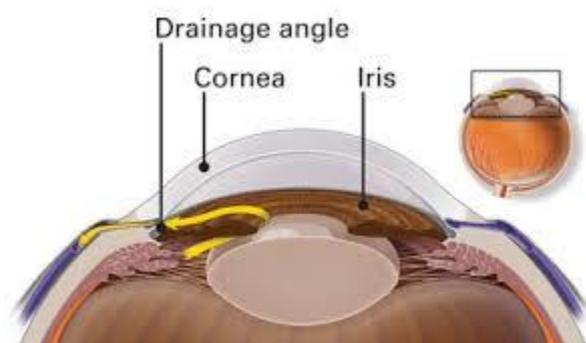
Examination under anaesthetic can last between 15-30 minutes. For children older than a few months of age, a parent accompanies them to the anaesthetic room to provide reassurance as an expert paediatric anaesthetist explains carefully each step of the procedure. The anaesthetic helps children to forget this experience.

Under anaesthetic, the eye pressure and eye features can be carefully examined. If the diagnosis is clear and further surgery has been discussed, this is then performed.

Examination under anaesthesia to remove sutures, check pressures, check eye focusing and give anti-scarring injections is often a part of assessing the progress of bigger glaucoma surgical operations.

## Angle surgery for glaucoma

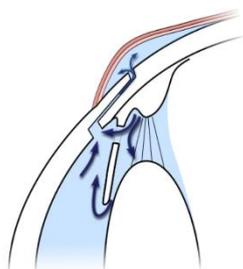
The angle of the eye is the junction between the coloured iris and the cornea. Angle surgery can be either a goniotomy or trabeculotomy. In general, goniotomy is faster but treats a smaller amount of the angle. This surgery can be repeated.



**Goniotomy:** a blade is passed across the front of the inside of the eye to incise an arc of the tissue that is blocking access of aqueous fluid. It is the least invasive of glaucoma surgeries.

**Trabeculotomy:** a tiny window is made through the outside of the eye to expose the drainage channel passage known as Schlemm's canal. A tiny microcannula with tiny fiberoptic light source (iTrack) or fine trabectome probe is used to explore and open the canal into the front of the eye.

## TRABECULECTOMY SURGERY



When angle procedures fail, or glaucoma appears severe, a **trabeculectomy** operation is offered. At this procedure a window of scleral tissue is lifted to allow controlled aqueous fluid flow from the eye. The rate of flow is controlled by **releasable sutures** in the flap. Fluid collects under the subconjunctival space and is then absorbed into surrounding tissues.

**Anti-scar medicine** such as Mitomycin C (MMC) is applied in tiny topical amounts for a few minutes during surgery. The body responds to surgery by healing and scar formation, so it is not unusual for eye pressures to rise again as healing takes place after trabeculectomy. In adults, releasable sutures and top-up anti-scar 5FU injections can be given awake with topical anaesthetic. After a trabeculectomy in young children, a repeat, brief general anaesthetic needs to be arranged (see above - Examination under anaesthetic). These typically occur at 2 and 4 weeks after initial surgery.

## TUBE SURGERY / GLAUCOMA DRAINAGE DEVICE SURGERY

When angle surgery fails, or is not possible, and when trabeculectomy is likely to fail due to rapid scar tissue formation, glaucoma tube surgery is a great alternative.

Glaucoma tubes consist of a silicone plate and its extension of a small tube. The plate is placed on the outer surface of the eyeball, back in the space between the eye muscles. This receives aqueous fluid from the front chamber of the eye via a thin, almost invisible tube.



Over 8-12 weeks following glaucoma surgery, a healing fibrosis develops around the plate to restrict excessive flow through the tube. Until this occurs, **flow needs to be controlled** by either a self-dissolving ligating suture around the tube or a **suture placed inside the tube** and sometimes both. The most common suture material is called Supramid Extra. It is white so not easily seen. During this period of flow-control it is common to continue pressure lowering drops.

### What happens after surgery? What about that night and the next day of recovery?

Once the surgery finishes, while children are still sleepy from anaesthetic, they are moved to the recovery area. Here, they wake up under the watchful eye and monitoring of a recovery nurse. Parents and care-givers will rejoin their child in the recovery area.

Typically, the operated eye remains patched overnight. Regular children's paracetamol or ibuprofen, or both, can be used for any discomfort. The recovery nurses will advise when drops are due.

Monitoring continues until your child is awake and alert, drinking and eating small amounts and is comfortable with distraction and cuddles.

### Post-Operative Drops and Pain Relief

Local anaesthetic, antibiotic and local steroid injections are given at the end of these procedures, then the eye is gently covered by an **eye pad** and a clear plastic **shield**. Ideally these will remain in place until review the next morning. No glaucoma or post-operative drops are needed before review.

The day after surgery the pad and shield are removed. If possible, pressure is measured, though the appearance of the eye and comfort will guide the surgeon in assessing your child.

**Drops** now commence, usually 4 times a day for antibiotics, and every 2 hours for steroid drops. Chlorsig (Chloramphenicol) is the most common antibiotic used. Maxidex (Dexamethasone 0.1%) is the most common steroid drop.

In *angle surgery*, pilocarpine can be used to constrict the pupil and prevent scar in the area of surgery. After *tube surgery* because the tubes are blocked, glaucoma drops may continue. Most commonly after *trabeculectomy* **Diamox and glaucoma drops will cease.**

For ongoing **pain relief**, it is sensible to give regular doses of paracetamol syrup every 4-6 hours, particularly before sleep times. Your surgeon may not wish you to use ibuprofen (Nurofen) for the first 48 hours as this can interfere with clotting.

### Do I need to keep the eye covered?

After the soft eye pad is removed your child's normal glasses can be worn. Generally it is not necessary to keep the eye covered. Older children may like to wear sunglasses. Overnight, a clear plastic shield can be worn for a few days to prevent inadvertent rubbing or pressure during sleep.

### Follow-up appointments

After the first check on the day of surgery or morning after, the next appointment is the following Monday or Wednesday. After that, reviews will occur weekly, and then fortnightly with a gradual decrease in frequency.

### What about other activities after surgery?

Encourage **no rubbing** of the eye. It is fine to **wash normally**, in the bath or shower. Pouring of water directly onto the eye is discouraged.

Returning to childcare, pre-school activities and school itself can occur once the eye is comfortable, and drop regime settles. This generally takes a 5-10 days. A gradual return to more vigorous activities is suggested: participation in 2 weeks, then competitive in 4 weeks is a rule-of-thumb after eye surgery.

There are **no restrictions on flying**.

Please report any sudden swelling, or sudden increases in pain that don't settle with simple pain medications. If your child is old enough to report a sudden change or decline in their vision is another reason to seek urgent review.

### How do we know when the operation is successful?

It takes several weeks of decreasing post-operative drops and pressure monitoring to declare a glaucoma operation success. In younger anxious children, procedures such as examination under anaesthetic and suture removal may be required to confirm success.

Successful glaucoma surgery stops any progression of the disease, sometimes for years, sometimes for decades. Success is usually defined as normal eye pressure, sometimes whilst continuing drops and sometimes off all pressure-reducing drops.

Some types of surgery in mild glaucoma have success rates as high as 90% at five years following surgery. For more severe secondary glaucoma, or repeat surgeries, success rates of 60% at 5 years are more common.

Failure of surgery does not mean inevitable blindness. If intraocular pressures rise or the disease appears to be progressing, glaucoma interventions such as recommencing drops, using laser or newer surgical devices are all possible options.

## The pressure is better, what next?

**Monitoring of the pressure** should continue. Usually every 3 months at first, then six-monthly. If you live a long way from Melbourne, we are happy to liaise with a well-qualified and available local ophthalmologist or optometrist to reduce the number of visits, or for peace of mind regarding eye pressures in between visits.

Glasses may be needed in many cases, to account for the changed size, shape and focus of eyes with glaucoma.

Amblyopia (lazy eye) can develop through the glaucoma and the journey of surgery. It requires glasses and daily part time patching of the good eye.

## The future?

Caring for a child with glaucoma is a journey. At all points, information and resources are available to help you navigate, please do not hesitate to ask. Along the journey, there may be steady periods with regular appointments and drops to administer.

Children should have the issue explained in age-appropriate language and gradually, as adolescents, begin to take control of the condition themselves.

Members of the treating team include your ophthalmic surgeon, orthoptists, nurses, and reception team. Your GP and optometrists are key members who will be kept informed by your surgeon. Although it is a rare condition, this large team have seen many people face the challenge and thrive.

## Support and References

Congenital Glaucoma Research Network <https://www.gl-foundation.org/>

Glaucoma Australia <https://www.glaucoma.org.au/>

Vision Australia <https://www.visionaustralia.org/services/children>

Statewide Vision Resource Centre <https://svrc.vic.edu.au/>

## Glossary

**Ophthalmologist:** A medical doctor who has undertaken further specialist training to diagnose eye disease and initiate medical and surgical treatments. Some ophthalmologists can have extra training in caring for glaucoma, or paediatric patients or both!

**Orthoptist:** A university-trained specialist in assessment of children's vision and eye alignment. All orthoptists work closely with ophthalmologists, have expertise in measuring vision (regardless of age) and ocular alignment, and in performing clinical testing for glaucoma.

**Optometrist:** a university-trained community provider for providing refraction (glasses) and detecting eye disease. Some may monitor treatment in glaucoma with Ophthalmologists.

**Geneticist:** A doctor that explains the investigation of possible genetic diseases to establish the risks of glaucoma recurrence and guide estimation of glaucoma severity.

**Anaesthetist:** A medical doctor specializing in preparing, initiating, and maintaining anaesthesia (keeping a patient pain-free and having no-awareness of a procedure).