

The Charity for People with Glaucoma
Established 1974

Secondary Glaucomas



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Printed July 2014 Review date July 2017 This free booklet is brought to you by the International Glaucoma Association (IGA), the charity for people with glaucoma. We haven't charged for it because we want you to have easy access to information that will help you understand and manage glaucoma.

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Your feedback is also important to us. Please help us to improve our information by sending us your comments about the content and format of this publication at marketing@iga.org.uk or by writing to us at the address below.

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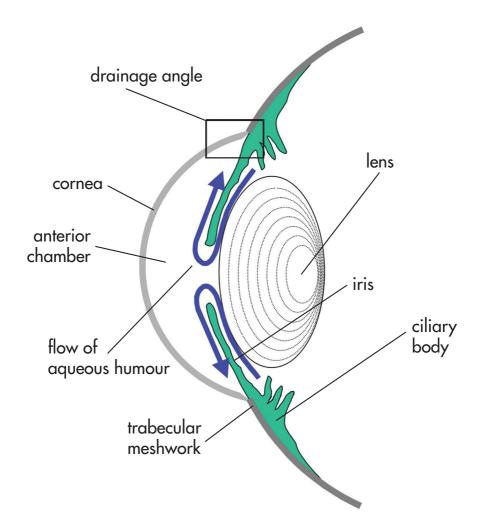
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1. Introduction - What is glaucoma?

Glaucoma is the name given to a number of conditions in which the optic nerve is damaged. The optic nerve is the nerve that transmits vision from the eye to the brain. Glaucoma damage in the optic nerve has a characteristic appearance and can affect the vision, causing progressive visual loss if untreated. Many eyes that develop glaucoma have raised intraocular pressure (high pressure in the eye) but there are other factors that may influence the development of glaucoma damage, such as poor blood supply to the optic nerve and weakness of the connective tissues supporting the optic nerve. A proportion of patients who develop glaucoma may therefore never have high pressure, although at present the treatment remains the same, which is to lower the eye pressure.

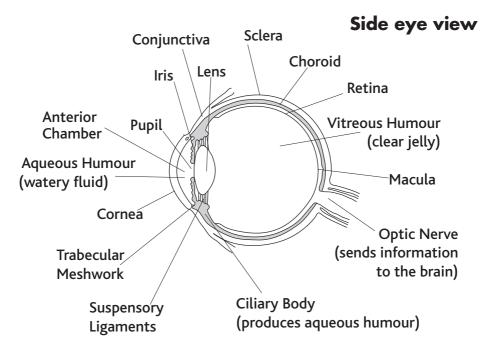
In most cases, glaucoma is of a 'primary' form, meaning that there is no identifiable cause for high pressure or the development of glaucoma. A 'secondary' glaucoma, on the other hand, occurs where there is an underlying ophthalmic or medical condition that predisposes to the development of high eye pressure.

The front inner part of the eye (anterior chamber) is bathed in a clear watery fluid called aqueous. This fluid keeps the eyeball firm and also nourishes the important tissues within the front part of the eye. The aqueous is produced by a structure called the ciliary body, located in the posterior chamber behind the pupil and lens. The aqueous passes through the pupil, which is the central hole in the coloured part of the eye (iris) and then circulates in the anterior



Normal flow of aqueous humour in the eye

chamber between the clear window at the front of the eye (cornea) and the iris. Aqueous production is continuous but drops down to a minimum at night. Obviously, if there was no mechanism for the aqueous to drain out of the eye, the pressure in the eye would become too high.



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There is therefore a drainage system called the trabecular meshwork located in the angle formed by the cornea and the iris. Once the aqueous drains through the trabecular meshwork it enters into the bloodstream. In secondary glaucomas there is some form of impairment to aqueous outflow, with an identifiable underlying cause.

There are several reasons why a patient might be diagnosed with secondary glaucoma. There may be a disorder present locally in the eye, a general disease or the use of medication which is, or has been, the cause or contributory cause of raised eye pressure. Strictly speaking, the condition is called 'secondary ocular hypertension' if there is just raised intraocular pressure but with no detectable glaucomatous damage to the optic nerve. If however, the raised pressure damages the optic nerve, which therefore can lead to visual loss, this would then be called secondary glaucoma.

Secondary glaucoma is unlike primary open angle glaucoma in that when the underlying problem causing raised intraocular pressure is corrected or cured (if this is possible), the pressure may return to normal and further medication may be unnecessary. Unfortunately if optic nerve damage has already occurred and visual field damage already exists then this cannot be reversed but should stabilise once the pressure is normalised.

2. Causes of secondary glaucoma

1. Pigment dispersion syndrome and pseudo-exfoliation

These are two of the commonest causes of secondary open angle glaucoma. Because they are treated in an identical

fashion to primary open angle glaucoma, some doctors classify them as part of the same condition ('chronic open angle glaucoma'). In pigment dispersion syndrome, pigment granules are intermittently rubbed off the back of the iris by the lens and the granules get deposited in the trabecular meshwork. This causes a blockage of aqueous outflow leading to high intraocular pressure. When it causes optic nerve damage it is termed pigmentary glaucoma. The condition is commoner in young, myopic (short-sighted) men.

In pseudo-exfoliation, the trabecular meshwork gets clogged up with white flakes. These flakes are fibrillar deposits from the surface of the lens capsule that get shed by the continuous movement of the iris (when the pupil enlarges or gets smaller). If pseudo-exfoliation leads to damage to the optic nerve it is termed pseudo-exfoliative glaucoma. It tends to be more common with increasing age and is thought to be more prevalent in certain ethnic groups (such as Scandinavians and Southern Mediterraneans), although can be seen in any ethnicity.

2. latrogenic

latrogenic literally means 'caused by a doctor' and unfortunately this is perhaps the most common presentation of true secondary ocular hypertension seen in eye clinics. The main iatrogenic cause is following retinal

surgery. This is because sometimes the retinal surgeon needs to put substances such as gas or silicone oil in the eye in order to flatten the retina following a retinal detachment or after other retinal surgical treatments. These substances invariably cause the pressure to rise and sometimes the rise in pressure can be very high. With gas the effect is usually short-lived and some of the gas may be removed early to reduce the pressure. Silicone oil may need to stay inside the eye for many months, years or indefinitely and a high proportion of these patients may develop secondary glaucoma.

Another common 'iatrogenic' cause for high pressure is the use of steroids. Other medications can cause high pressure by a number of different mechanisms. Drug causes of secondary glaucoma will be discussed in a separate section below.

3. Inflammation in the eye (uveitic glaucoma)

Uveitis is inflammation of the layer of pigmented tissues inside the eye (uvea). There are a number of different ways in which uveitis can present – anterior uveitis or iritis will only affect the front of the eye, vitritis is inflammation in the vitreous and posterior uveitis implies inflammation in the back of the eye (retina, choroid layer and vitreous).

There are a number of different mechanisms by which

uveitis can cause high pressure.

- a) Uveitis may cause leakage of protein and white blood cells into the aqueous fluid and these can deposit in the trabecular meshwork, blocking aqueous outflow.
- b) The trabecular meshwork can become inflamed itself, a term called 'trabeculitis'. Doctors think that this may cause high pressure in situations where the eye has high pressure but only very minimal inflammation (such as the condition known as 'Posner-Schlossman syndrome' and also when shingles affects the eye).
- c) Uveitis may cause secondary angle closure. This is due to the fact that inflammation in the front part of the eye can cause the iris to stick to other structures. In the acute setting, the iris around the pupil can stick to the lens (posterior synechiae); if the pupil gets completely stuck down for 360 degrees then the aqueous cannot pass from the ciliary body to the anterior chamber. This means that aqueous builds up behind the iris, pushing it forwards and completely closing off drainage through the trabecular meshwork. This causes the pressure to go very high quickly and will require urgent intervention. The pressure can be relieved by making a hole (iridectomy) in the iris to allow the aqueous to pass. This is preferably achieved definitively using a

surgical procedure but laser can be used as a temporary measure in the emergency setting. When uveitis has been occurring over a long period of time, the iris can stick to the trabecular meshwork (peripheral anterior synechiae) and if these adhesions are extensive, there will be a significant reduction in aqueous outflow, leading to high pressure and glaucoma. Often the treatment for this will need to be surgical drainage.

d) One of the commonest causes of high pressure in uveitis is secondary to steroid treatment. This is discussed in more detail in a later section.

When high pressure occurs during an acute episode of anterior uveitis, it is usually termed 'hypertensive uveitis'. A rare variant of this, when the pressure is very high but the inflammation is minimal, is called 'Posner-Schlossman' syndrome. In most cases, the episodes are short-lived and respond well to topical steroids to treat the inflammation and glaucoma medications (eye drops and occasionally Diamox tablets) to lower the eye pressure. The medications can usually be stopped once the episode has resolved. In some patients, the episodes can recur but can eventually wane out completely. However, in some patients who get frequent sustained episodes of high pressure, the optic nerve may become progressively damaged leading to

glaucoma. These patients usually need long term treatment and in some cases surgery.

Fuch's Heterochromic Cyclitis is a specific form of uveitis associated with loss of pigment from the iris of the eye, which becomes blue if previously brown. Symptoms are often absent in the early stages but raised pressure and cataract development may occur later and affect vision.

4. Lens related problems

Cataracts may cause a secondary rise in pressure through an angle closure mechanism. When a cataract is very advanced it can become swollen and block the flow of aqueous from the ciliary body through the pupil. This causes a build up of aqueous behind the iris, pushing the iris forward, closing off drainage through the trabecular meshwork. This type of mechanism is termed 'phacomorphic' and is relieved by removing the cataract. A similar 'pupil block' mechanism may occur if the lens within the eye is unstable due to weakness or loss of the zonular ligaments that support the lens. This can happen following trauma and in patients with pseudo-exfoliation (where the zonules are weaker than normal).

Very rarely, when a patient has advanced cataract, proteins from within the lens can leak into the aqueous and this can cause high pressure in the same way as with uveitic glaucoma. This mechanism is termed phacolytic and will respond to removal of the cataract. Much more rarely, if there is a break in the capsule surrounding the lens (for example following trauma), leakage of lens material may cause a severe allergic reaction in the eye causing a lot of inflammation and secondary raised pressure. This is termed phacoanaphylaxis and once again requires removal of the cataract.

Occasionally secondary ocular hypertension can occur due to the position of the artificial lens inserted into the eye at the time of cataract surgery. Occasionally it is necessary to insert a lens in the anterior chamber, rather than in the posterior chamber. This mostly happens when there isn't enough support for an artificial lens within the remaining lens capsule (where lenses are usually inserted) following complicated cataract surgery. Some lenses used for refractive surgery are also inserted into the anterior chamber. It is possible for these lenses to block aqueous passage through the pupil causing a secondary angle closure. It is therefore essential that a peripheral iridectomy (a hole in the peripheral iris) is made at the time of inserting the lens. Occasionally the pressure can become very high in these eyes if the peripheral iridectomy has not been made or closes off. Sometimes the artificial lens is inserted behind the iris but in front of the capsule (in the 'sulcus'). Once again this may be undertaken in the

context of complicated cataract surgery. Sometimes the artificial lens in this position can rub off pigment from the back of the iris and cause secondary pigment dispersion. This pigment can lead to blockage of the trabecular meshwork with resultant high intraocular pressure. If this problem is identified early, it can be reversed with removal or re-siting of the lens. If the problem has been present for a long time the pigment will have caused permanent damage to the trabecular meshwork so removal/resiting of the lens alone may not lower the pressure. This may therefore require glaucoma drainage surgery.

Occasionally patients have cataracts removed and do not have an artificial lens inserted. This is called aphakia and it can cause a secondary glaucoma, although the precise mechanism is unknown. This is by far the commonest cause of secondary glaucoma in children.

5. Trauma

- a) A blow to the eye or a penetrating injury may cause high pressure through uveitis, haemorrhage (with blood cells clogging up the trabecular meshwork) or damage to the lens (such as dislocation/instability of the lens or capsule rupture as discussed above).
- b) A chemical injury (such as an alkali burn) can lead to severe inflammation of the cornea and uveitis,

leading to secondary raised pressure.

c) Blunt trauma to the eye may cause the drainage angle to become pushed backwards permanently (angle 'recession'). This can lead to the development of high pressure up to a number of years after the injury. Most patients found to have significant angle recession following an injury will be monitored for a number of years to ensure they do not develop glaucoma.

6. Drug related

a) The commonest drug-related cause of secondary raised intraocular pressure is called 'steroid response' and this is due to the use of steroids. Steroids may be inhaled, taken orally, injected, used topically on the skin or taken locally in the eye. Steroid response pressure elevation may be caused by each of these routes but it is most common when the steroid is applied locally to the eye, in other words via a steroid eye drop, injected into or around the eye. In most cases of steroid response, the intraocular pressure may reduce back to baseline levels if the steroid is withdrawn. It is important not to use topical eye drop steroids for a long-time without having the intraocular pressure checked intermittently.

Sometimes steroids may be necessary for long periods

for the treatment of uveitis or other ocular inflammatory conditions. It is becoming increasingly common to inject steroid into the eye or to insert a long acting steroid implant. The steroid-related elevation may be very prolonged in these cases and may require glaucoma drainage surgery.

b) In a susceptible person, any eye drop which dilates the pupil may result in raised pressure by closing an already narrow drainage angle.

7. Neovascular (newly formed blood vessels)

In certain retinal conditions, such as proliferative diabetic retinopathy and central retinal vein occlusions, the poor blood supply within the eye may cause new blood vessels to grow onto the surface of the eye and into the drainage angle. The presence of the new vessels in the angle can cause the pressure to rise. If the new vessels progress and are present for a long time they can scar up and completely zip up the drainage angle (synechial closure). If detected early, treatment of the underlying retinal condition (with laser and perhaps an injection of an anti-VEGF drug) may reverse the new vessels and lower the pressure. If the angle has become zipped completely, the high pressure often will not respond adequately to drop treatment so glaucoma drainage surgery or cyclodiode laser is usually required. Often patients will already have poor vision in

the affected eye due to the underlying retinal problem.

However, if the pressure is not adequately treated in a timely fashion (and the underlying retinal condition treated), the secondary glaucoma due to the new vessels (also called 'rubeotic glaucoma') can lead to severe optic nerve damage with a high risk of blindness.

8. Following corneal disease and corneal surgery

Secondary ocular hypertension can occur in patients who attend the ophthalmologist because of disorders of the cornea (the clear window at the front of the eye). Corneal infection (whether bacterial or viral) and corneal burns may cause inflammation in the eye (similar to uveitis) leading to elevated intraocular pressure.

Many corneal conditions will require the use of long-term topical eye drop steroids and this can lead to sustained steroid response pressure elevation. This is commonly seen in patients who have undergone corneal grafts, where topical eye drop steroids are needed for many months to prevent the corneal graft from failing. If this cannot be treated with eye drops and the optic disc is showing glaucoma damage then glaucoma drainage surgery is usually recommended.

Some patients with complicated corneal problems may

develop scarring adhesions between the iris and the trabecular meshwork (peripheral anterior synechiae); this also occurs in patients following multiple graft surgery. If the synechiae are extensive, the outflow of aqueous can be severely compromised leading to high intraocular pressure. In many cases, glaucoma drainage surgery may be required. Very rarely, patients with multiple failed corneal grafts may need to have an artificial keratoprosthesis (artificial corneas) inserted to allow them to see. If the patient can produce sufficient tears to keep the surface of the eye wet, a 'K-Pro' can be sutured to the eye, attached to a ring of donor cornea. In patients who cannot produce sufficient tears and therefore have a dry eye, the keratoprosthesis can be incorporated into a piece of tissue hewn from one of the patient's teeth. The tissue and prosthesis is then implanted under the patient's cheek to enable it to develop its own blood supply over a few months. The implant is then removed and sutured to the patient's eye. Both forms of keratoprostheses have a high risk of developing secondary glaucoma, with the additional problem that one cannot measure the pressure in eyes with artificial corneas. Such patients will require glaucoma drainage surgery (invariably tube implantation).

Iridocornealendothelial (ICE) syndrome is a rare disorder that can cause glaucoma in adults aged 30-50 years, more commonly in women. It is unilateral (occurring in only one eye) and is related to an abnormality of one of the cell layers of the cornea. ICE syndrome is associated with swelling of the cornea and acquired defects of the iris. High pressure is caused by the formation of adhesions of the iris to the trabecular meshwork (peripheral anterior synechiae) reducing the outflow of aqueous. It can be difficult to control the pressure in such eyes using drops, so treatment will often require the implantation of a glaucoma drainage device (tube surgery).

9. Bleeding inside the eye

As mentioned in the section on trauma, blood cells in the anterior chamber can clog up the trabecular meshwork and lead to high pressure. Blood in the vitreous cavity (vitreous haemorrhage – often occurring in proliferative diabetic retinopathy) can cause secondary raised pressure by a similar mechanism.

10. Raised episceral venous pressure

This refers to conditions where the pressure in the veins into which the aqueous drains is elevated. This high pressure causes resistance to outflow of aqueous and therefore causes high eye pressure. This can happen when there is an increase in pressure in the tissues of the orbit (e.g. in the presence of an orbital tumour, thyroid eye disease), or abnormalities of the veins, or an abnormal

communication between the arteries and veins (a fistula).

11. Tumours

Occasionally tumours may develop in the eye. Tumours in the iris may involve the drainage angle and therefore reduce drainage outflow causing high eye pressure. Likewise a tumour in the ciliary body may push the peripheral iris forward, closing off the drainage angle. Large tumours in the back of the eye may push the lens forward, closing off aqueous passage through the pupil leading to a secondary pupillary block angle closure. Tumours may also cause new vessel formation, with the same effect as described in section seven.

3. Treatment

The good thing about secondary glaucomas is the fact that reversal or treatment of the underlying cause will often normalise the pressure and may indeed be curative if permanent. Most secondary glaucomas will require treatment of the underlying cause, drops to lower the pressure (and possibly tablets in the short-term) and anti-inflammatory drops if there is an inflammatory component.

There are certain circumstances where surgical or laser treatments are required. Specific interventions, such as cataract surgery or lens removal/resiting have been discussed for lens-related causes. The creation of a peripheral iridectomy for secondary angle closure in uveitis has also been discussed in the section pertaining to uveitic glaucoma.

Surgical or laser intervention needs to be considered if the high pressure is refractory (i.e. inadequately responsive) to medical treatment and measures to reverse the underlying cause. This is because there is a significant risk of visual loss unless the high pressure is definitively addressed. The commonest surgical procedure undertaken for glaucoma (other than cataract surgery) is called trabeculectomy and this is usually augmented with the use of antiscarring drugs such as 5-fluorouracil or mitomycin C. Trabeculectomy is usually quite successful in the management of uveitic glaucoma. However there is a high risk of trabeculectomy failure in other forms of secondary glaucoma such as neovascular (rubeotic) glaucoma, aphakic glaucoma and glaucoma related to retinal surgery (particularly if silicone oil has been used in the eye). In these cases, it is usually preferable to insert a tube shunt. This tube diverts aqueous from the anterior chamber to a plate secured to the orbital part of the eyeball. From here, the aqueous can drain into the bloodstream. This differs from trabeculectomy where the aqueous collects in a 'bleb' nearer the front of the surface of the eye just below the upper eyelid. The tube procedure is less prone to scarring and therefore failure for certain secondary glaucomas as

compared to trabeculectomy.

There is a form of laser treatment called 'cyclodiode' or ciliary body ablation which can be very useful for treating some secondary glaucomas. The laser treatment works by reducing the production of aqueous by the ciliary body. The effect is usually temporary. It is often used in eyes that are painful and blind due to secondary glaucoma. It can, however, be very useful as a temporising measure in eyes with good visual potential. It should be avoided in people with uveitis as it can increase the degree of inflammation and can make the vision worse through accumulation of fluid at the macula in these eyes.

A popular laser treatment for primary open angle glaucoma known as selective laser trabeculoplasty (SLT) is unlikely to work in most forms of secondary glaucoma. It is relatively contraindicated in pigment dispersion syndrome and pseudo-exfoliation as it can cause high pressure spikes post-laser; however experienced laser surgeons can safely and successfully use this treatment in patients with these conditions by using a lower amount of laser energy.

There are a number of newer surgical glaucoma treatments such as trabectome, iStent and canaloplasty. Whilst these may be showing some promise in the management of ocular hypertension and primary open angle glaucoma, at the time of writing there is very little published evidence demonstrating their long-term effectiveness in secondary glaucomas.

4. Remember

- 1. If you have bilateral glaucoma, you **MUST** inform the DVLA.
- Prescribed drops should be used as recommended by your specialist, on a daily basis unless otherwise informed.
- 3. If you develop any untoward side effects you should inform your specialist as soon as possible.

5. Disclaimer

Whilst every step have been taken to compile accurate information and to keep it up-to-date, we cannot guarantee its correctness and completeness. The information provided in this information sheet is designed as an adjunct to, and not a substitute for, professional healthcare advice by a qualified doctor or other healthcare professional, which will be tailored to a patient's individual circumstances. The International Glaucoma Association cannot take responsibility if patients rely soley on the information in the information sheet.

Further help and information

Telephone us

The IGA operates its own free telephone advice line, called Sightline.

If you would like to find out more about any of the information contained in this booklet, or you would like to discuss any concerns you may have about glaucoma, you can call the IGA's Sightline, Monday to Friday 9.30am to 5.00pm. Out of office hours there is an answer phone service where you can leave a message and you will be called back.

Sightline: 01233 64 81 70Monday-Friday 9.30am-5.00pm

Visit our website: **www.glaucoma-association.com** for a wide range of information, to order other booklets or leaflets, or to participate in our on-line discussion forum.

Email us at: info@iga.org.uk

Other IGA booklets and leaflets

The IGA produces a range of informational booklets and leaflets. These are constantly being reprinted, reviewed and updated so call Sightline, or visit the website, to find out what is currently available.

Secondary Glaucomas

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A full list of references and information sources used in the compilation of this booklet is available on request by phone: 01233 64 81 70 (Sightline) or by email: info@iga.org.uk

Don't Forget!

- Use your eye drops as prescribed by your consultant to avoid further sight loss in most cases.
- Tell your close relatives that you have glaucoma. They
 are at higher risk than average so should be tested
 regularly, and first degree relatives over the age of 40
 are entitled to free eye tests.
- Contact the IGA Sightline if you have any questions.
 We are here to help.
- Join us! A membership form is enclosed in the middle of this booklet. If you are a member already, please pass it to a relative or friend. You may save someone's sight:
 - Support leaflets for other patients!
 - Support research into the causes and treatment of glaucoma
 - Receive the quarterly IGA News

The information contained in this booklet was correct at the time of printing.



The Charity for People with Glaucoma Established 1974

Our Mission and Vision

The International Glaucoma Association is the charity for people with glaucoma. Our mission is to raise awareness of glaucoma, promote research related to early diagnosis and treatment and to provide support to patients and all those who care for them.

Our vision is to ensure that all people with or at risk of glaucoma have the knowledge and access to care that will enable them to maintain a good quality of life.

To donate:

Call 01233 64 81 64 or visit www.glaucoma-association.com

For help, advice and information Sightline: 01233 64 81 70 Monday - Friday 9.30am - 5.00pm

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