

## Syndromes and Anomalies (associated with Glaucoma)

### Sturge-Weber syndrome

The association between a port-wine stain on the face and scalp and raised eye pressure – the Sturge-Weber syndrome – is uncommon. It usually occurs at an early age, although it has occasionally been found to develop unknowingly in young adults, or even in later life.

People with a port-wine stain affecting the head and also involving the eye lid or conjunctiva (visible as prominent blood vessels on the white of the eye) are particularly prone to develop high pressure in the eye and this may result in glaucoma.

Any person who has a port-wine stain affecting the head, particularly if the upper eyelid is involved, should have their eyes examined regularly for all syndromes. There is considerable variation in the degree of glaucoma in these cases and only the eye specialist who has examined a patient can determine what treatment is relevant to an individual.

Mild cases can sometimes be controlled by medical treatment using pressure lowering eye drops but in most cases surgery is necessary to improve the drainage of fluid from the eye (**please ask for the IGA booklet 'Trabeculectomy'**).

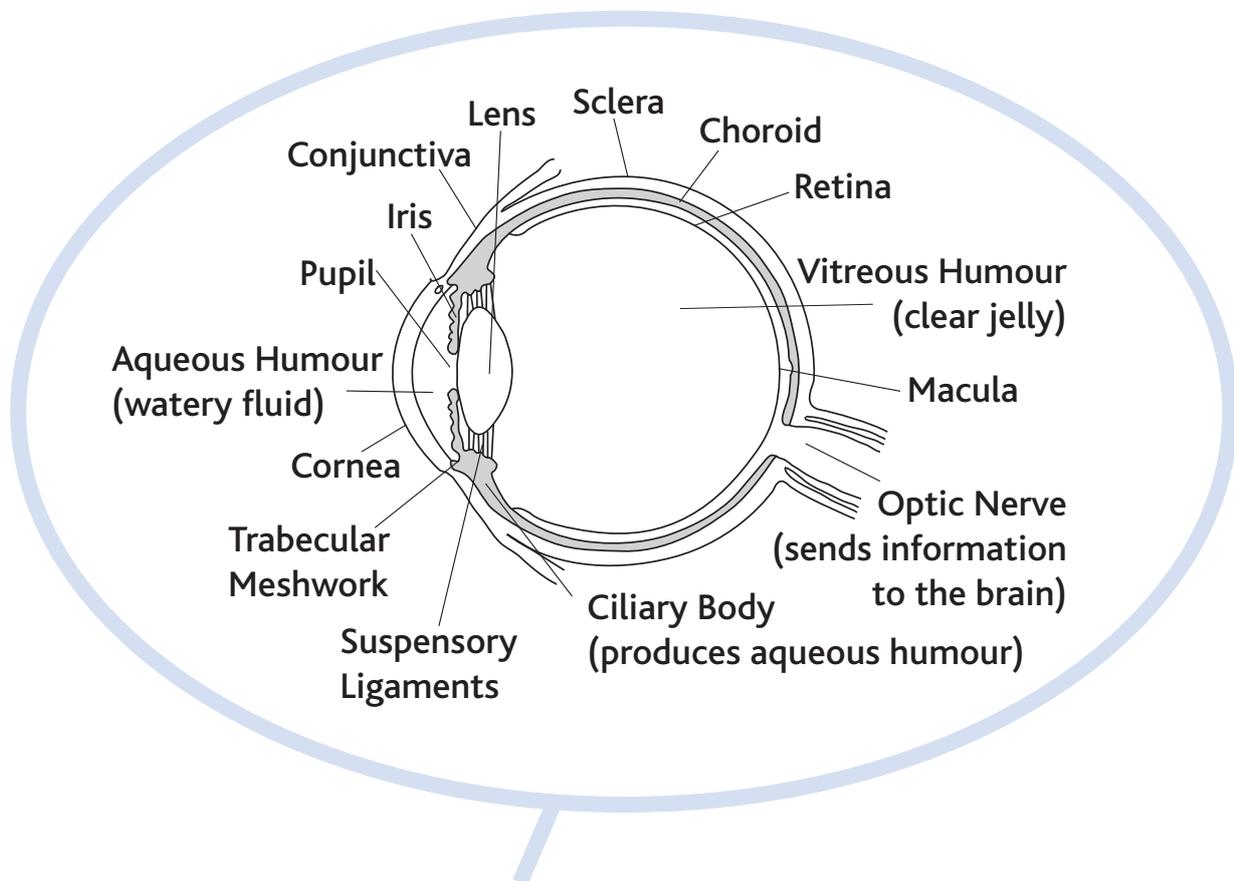
### Axenfeld and Reiger syndromes

In both of these inherited conditions, there is a tendency for the iris (coloured part of the eye) and the margin of the cornea (clear window of the eye) to develop abnormally.

As a result, the channels which drain aqueous (fluid produced in the eye) are less open and the pressure in the eye may rise causing a form of glaucoma. If that happens, treatment is similar to other types of chronic open angle glaucoma.

In general, treatment starts with pressure lowering eye drops. Should the pressure in the eye remain at a level which causes or is likely to cause damage to the field of vision, then surgery will be considered.

The most common operation is called a trabeculectomy and aims to reduce eye pressure by creating a new drainage channel within the eye but glaucoma drainage devices (tube) surgery is also often needed (**please ask for the IGA Booklet 'Trabeculectomy'**).



Cross section through the eye showing the major structures

## Peter's anomaly

Peter's anomaly is an abnormality of the eye which develops at the embryonic stage. It results in the central area of the cornea (the clear window in the front of the eye) being opaque.

The iris (coloured part of the eye) has holes and may be stuck to the back of the cornea and the lens. In some cases the lens is also opaque and this may be stuck to the back of the cornea.

In addition there may be malformations of the drainage mechanism of the eye and parts of the iris may block this drainage mechanism leading to a rise in the level of intraocular pressure (the pressure within the eye). If this raised pressure is high enough and prolonged enough it may cause damage to the optic nerve (glaucoma).

Most cases of Peter's anomaly occur without there being any family history of this condition, although it can run in families. In the majority of cases both eyes are affected, and in about half of these cases the intraocular pressure is, or may become raised. Treatment of Peter's anomaly may include performing a surgical iridotomy (making a hole in the iris) to improve vision, or sometimes performing an early corneal transplant.

Raised pressure, if present, is usually difficult to treat using eye drops and surgery is the most common option with a trabeculectomy or glaucoma drainage devices being the most usual choice for the surgeon.

If drainage surgery is not fully successful then the use of laser treatment of the ciliary body (part of the eye which produces fluid) may be advised to reduce the pressure of the eye and control the glaucoma.

This may be described as '**cyclodiode laser**' or '**cyclo-photocoagulation**'.

Author: **Cecilia Fenerty** MD PGCertME FRCOphth

Medical editor: **Gus Gazzard** MBBChir MA MD FRCOphth



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## For more information

Please call: **01233 64 81 70** or email: [info@iga.org.uk](mailto:info@iga.org.uk) to receive free copies of:

- Glaucoma - A Guide
- Dry Eye Syndrome - A Guide
- Ocular Hypertension - A Guide
- Eye Drops and Dispensing Aids

**International Glaucoma Association**  
**Woodcote House**  
**15 Highpoint Business Village**  
**Henwood, Ashford**  
**Kent TN24 8DH**

Administration: **01233 64 81 64**  
Email: [info@iga.org.uk](mailto:info@iga.org.uk)  
Website: [www.glaucoma-association.com](http://www.glaucoma-association.com)

A full list of references is available on request.

Formed in 1974, the IGA has the mission 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. Funded entirely by its members and donors (no government or statutory funding) the Association provides its services free of charge to anyone in need of assistance.

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International Glaucoma Association,  
Woodcote House, 15 Highpoint Business Village, Henwood, Ashford, Kent TN24 8DH  
Administration: 01233 64 81 64 • Email: [info@iga.org.uk](mailto:info@iga.org.uk)  
Charity registered in England & Wales No. 274681 and in Scotland No. SC041550