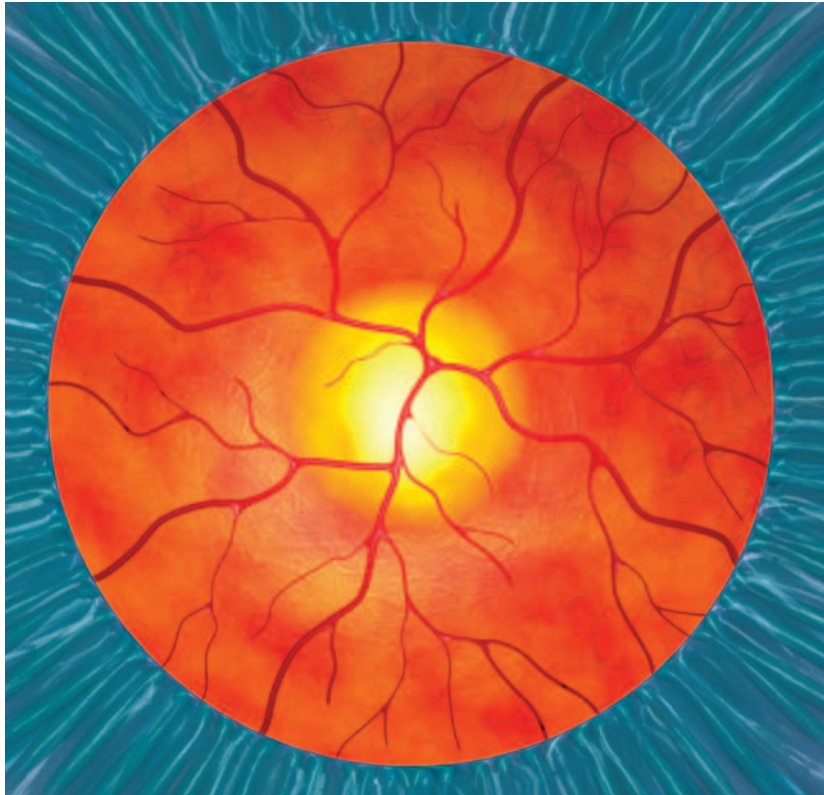


Fast Facts



# Fast Facts: Glaucoma

Paul R Healey and Ravi Thomas





# Fast Facts: Glaucoma



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**Declaration of Independence**

This book is as balanced and as practical as we can make it.  
Ideas for improvement are always welcome: [feedback@fastfacts.com](mailto:feedback@fastfacts.com)

Fast Facts: Glaucoma  
First published March 2010

Text © 2010 Paul R Healey, Ravi Thomas  
© 2010 in this edition Health Press Limited  
Health Press Limited, Elizabeth House, Queen Street, Abingdon,  
Oxford OX14 3LN, UK  
Tel: +44 (0)1235 523233  
Fax: +44 (0)1235 523238

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A CIP record for this title is available from the British Library.

ISBN 978-1-905832-40-8

Healey PR (Paul)  
Fast Facts: Glaucoma/  
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Cover: Glaucoma is an increased pressure in the eyeball due to an excessive amount of aqueous humour (the fluid that fills the eyeball). Seen here are the blood vessels (red) and the optic disc (yellow), which is raised with a central bulge or 'cup' (white).  
David Mack/Science Photo Library

Medical illustrations by Dee McLean, London, UK.  
Typesetting and page layout by Zed, Oxford, UK.  
Printed by Latimer Trend and Company Limited, Plymouth, UK.

Text printed on biodegradable and recyclable paper  
manufactured using elemental chlorine free (ECF)  
wood pulp from well-managed forests.



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## Glossary

**Amblyopia:** a decrease in vision for which no cause can be found on examination; in appropriate cases, amblyopia is correctable by therapeutic measures

**(Primary) angle closure:** obstruction of the trabecular meshwork by the iris in the absence of any detectable preceding causative disease

**Angle-closure glaucoma:** glaucomatous optic neuropathy due to raised IOP caused by obstruction of the iridocorneal angle by the iris

**CDR:** (optic) cup-to-disc ratio

**Cyclophotocoagulation:** procedure to destroy the ciliary processes, usually performed with a diode laser

**Cycloplegic agent:** topical drops used to paralyse the ciliary muscle; they also dilate the pupil

**Fundus:** the inner part of the eye visualized on ophthalmoscopy (fundoscopy): optic disc, retina, macula, blood vessels, etc.

**Hypermetropia:** condition of the eye where, with the accommodation at rest, parallel rays of light focus at a point behind the retina, usually because the axial length of the eyeball is smaller than normal

**Hypotony:** a syndrome of reduced vision and/or retinal swelling or folds caused by a very low eye pressure

**IOP:** intraocular pressure

**Iridocorneal angle:** the angle created by the junction of the cornea and iris; the outflow channels for aqueous drainage (trabecular meshwork) are located here

**Lamina cribrosa:** a sieve-like structure in the optic nerve head through which nerve fibers from the retina pass; blood vessels enter and leave the eye through this structure

**Myopia:** condition of the eye where, with the accommodation at rest, parallel rays of light focus at a point in front of the retina, usually because the axial length of the eyeball is larger than normal

**Neuroretinal rim:** the area of the optic disc occupied by the axons of the optic nerve (the 'left over' space is the cup)

**Neuroretinal rim notch:** a focal area of loss of the neuroretinal rim; notches are often seen in glaucoma

**Open-angle glaucoma:** glaucoma in the presence of anatomically normal iridocorneal angle structures, as seen on gonioscopy

**Optic cup:** the space left over in the optic disc after accommodating the retinal axons that form the optic nerve

**Optic disc:** the region of the fundus where the axons aggregate to form the optic nerve and exit the eye

**PAS:** peripheral anterior synechiae; an adhesion of the iris to the iridocorneal angle

**PGA:** prostaglandin analog

**Primary angle-closure suspect:** a patient whose angles are at risk of closure but who has no structural or functional signs of the disease

**Primary glaucoma:** glaucoma with no detectable preceding causative disease

**Relative afferent pupillary defect (RAPD):** an abnormal response in which one pupil dilates, rather than constricts, when a light is shone alternately on it and the other eye

**Scotoma:** a visual-field defect

**Secondary exotropia:** an outward deviation of the eye due to loss of vision

**Secondary glaucoma:** glaucoma with an identifiable cause of angle damage

**Tonometry:** measurement of the pressure within the eye

**Trabecular meshwork:** mesh-like structure at the iridoscleral angle, which allows aqueous humor to flow from the eye

**Vascular dysregulation:** a condition in which blood flow is not properly distributed to meet the demands of different tissues (includes Raynaud's phenomenon)

## Introduction

Glaucoma is a chronic neurodegenerative disease of the optic nerve (the second cranial nerve). It is the most common neurodegenerative disease, affecting about 70 million people worldwide. Glaucoma is the second most common disease causing blindness after cataract, although the speed and degree of vision loss vary. Glaucoma blindness is irreversible but preventable. However, most people with glaucoma are not diagnosed or treated.

As yet, we have no tests that show the pathogenic mechanism at work in glaucoma. So, clinically, the disease is diagnosed and treated as a syndrome. Unfortunately, symptoms do not usually become noticeable until the late stages of glaucoma, because of neural compensation mechanisms. Nevertheless, quality of life can still be impaired relatively early in the course of the disease.

Clinical diagnosis consists of identifying the signs of structural damage in the eye (spatially localized loss of ganglion cells on the retina and at the optic disc) with matching loss of function (reduction in differential light sensitivity or amplitude of visual evoked potentials in the corresponding part of the visual field).

A number of risk factors for the onset and progression of glaucoma have been identified, of which raised intraocular pressure (IOP) is the most important. Corticosteroid use and contact between the iris and trabecular meshwork (angle closure) are modifiable risk factors for glaucoma, which act via raised IOP. Cardiovascular disease (including high and low blood pressure) is also a risk factor, acting via both raised IOP and possibly reduced perfusion of the optic nerve.

Treatment of glaucoma is based on reducing risk factors (almost always IOP) and improving quality of life. Lowering the IOP is a generic strategy for protecting the optic nerve, even when the initial IOP is not particularly high. The aim is to keep the IOP at a level at which disease progression is anticipated to be at an acceptably low rate. Medicines that may protect the visual pathways at a cellular level are being researched and developed, but no treatment can regenerate the optic nerve.

The management of glaucoma requires life-long monitoring for risk factors and checking optic nerve structure and function to determine whether the risk or disease state has changed. Results can be fed back into a management plan and the desired (target) IOP revised where necessary.

The diagnosis and treatment of glaucoma is made difficult by a number of factors.

- Visual disability often does not become apparent until the patient is almost blind.
- The 'normal' appearance of the optic disc varies enormously, making early diagnosis of structural damage difficult.
- Accurate measurement of vision loss from glaucoma requires expensive visual field analyzers and training for the clinician and their staff. False positive visual field test results are common in inexperienced patients.
- Risk factors for glaucoma in the absence of glaucomatous optic neuropathy are usually not sufficient to warrant prophylactic treatment (with the exception of a very high IOP or angle closure).
- The degree of IOP lowering required to stabilize glaucoma is different for each patient.
- Worsening of glaucoma usually occurs over years, making change difficult to recognize.
- Lowering the eye pressure surgically is challenging.
- We do not fully understand how glaucoma occurs, and have no treatments to directly prevent or cure it.

The aim of *Fast Facts: Glaucoma* is to provide a clear understanding of glaucoma: what it is, how to detect it and how to treat it. We hope this book will serve as a ready reference for all medical and eyecare practitioners, an aid for students and scientists involved in the study of eye disease and a sound overview for anyone interested in this challenging disease.

Several well-conducted population-based studies have investigated the prevalence and incidence of glaucoma and the risk factors associated with the disease. Additional information about risk factors and the natural history of glaucoma has been gleaned from well-conducted randomized controlled trials of treatment.

### **Definitions**

Because it is diagnosed as a syndrome, the frequency (prevalence) of glaucoma varies depending on how it is defined. As the condition develops slowly, most definitions come from cross-sectional studies. Loose definitions are based on either the appearance of the optic nerve head or a visual field defect that is typical in glaucoma. The perceived influence of intraocular pressure (IOP) has been so strong that some (mostly older) studies defined glaucoma as any eye with a pressure above the normal range (8–21 mmHg for most populations), even in the absence of any detectable nerve damage.

Stricter (more modern) definitions of glaucoma require correlation between structural damage at the optic disc and functional abnormalities of the visual field typical of glaucoma, or an amount of nerve tissue in the optic nerve head that is less than the extreme end of the normal population distribution (i.e. 97.5th or 99th percentile). This is based on the assumption that the smaller the area of nerve tissue in the optic nerve head, the more likely it is to have been destroyed by glaucoma. In longitudinal studies glaucoma is defined as either the loss of tissue from the neuroretinal rim of the optic nerve head or the new development of a reproducible visual field defect that is typical of glaucoma. The ‘typical’ findings of glaucoma are summarized in Table 1.1.

### **Subtypes of glaucoma**

Because IOP is such an important modifiable risk factor for glaucoma, subtypes of glaucoma are classified according to the cause or mechanism

TABLE 1.1

### **Findings 'typical' of glaucoma**

No one sign is diagnostic. There are no standardized criteria for diagnosis, but a combination of signs provides the strongest indicator, particularly when structural and functional signs match.

#### **Structural signs – loss of optic nerve tissue within the eye**

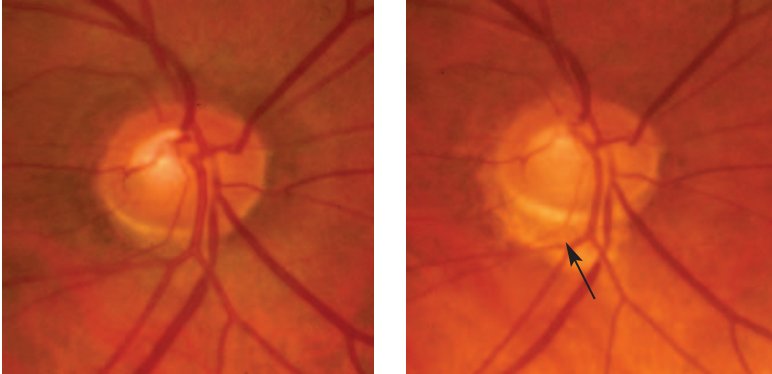
- Neuroretinal rim notch – focal (usually lower or upper) thinning or a complete absence of the rim of the optic nerve
- Large cup-to-disc ratio – generalized loss of neuroretinal rim tissue manifesting as a large optic cup compared with the size of the optic disc
- Focal or generalized defects (loss) of the retinal nerve fiber layer

#### **Functional symptoms – decreased optic nerve function in visual field locations that correspond to structural damage**

- Focal visual field defects (scotomas) that appear to arise from the blind spot (the position of the optic nerve head in the visual field) but do not cross the horizontal midline
- Scotomas can occur in isolation or in combination in the nasal, paracentral or peripheral parts of the upper or lower hemifields
- Coalescence of scotomas with loss of central vision but retention of temporal visual field (in advanced glaucoma)

of the increased IOP, which usually means the mechanism of damage to the aqueous outflow pathways of the eye. Figure 1.1 shows the two pathways through which aqueous fluid leaves the eye: the trabecular and uveoscleral outflow pathways. Aqueous production from the cells lining the ciliary body does not increase in eyes with high IOP. The uveoscleral pathway usually drains only a small proportion of aqueous. The main determinant of IOP is the resistance to flow through the trabecular meshwork; diseases that damage the trabecular outflow pathway frequently raise IOP.

It is important to remember that the subtypes, as shown in Figure 1.2, refer to the different causes of raised IOP, not to different types of optic neuropathy. Glaucomatous optic neuropathy – which



**Figure 3.20** Parapapillary atrophy (lighter area around black arrowhead) developing with open-angle glaucoma over 9 years. The visual field of this patient is shown in Figure 2.5 (page 26).

**Acquired pit of the optic nerve.** An acquired pit is a discrete oval-shaped depression in the lamina cribrosa (Figure 3.21). It is usually associated with localized excavation of the neuroretinal rim. The presence of an acquired pit of the optic nerve (APON) is strongly associated with glaucoma. It is usually located in the inferotemporal or superotemporal sectors of the disc. A congenital pit of the optic nerve is rarer and is not associated with glaucoma.

**Nerve fiber layer (NFL) defect.** The NFL should be examined on well-focused and illuminated optic disc photographs or using a green filter on the slit-lamp microscope or ophthalmoscope. The green light is reflected by the NFL and is absorbed by the pigment in the retinal

**Figure 3.21** Acquired pit of the optic nerve (arrowed). Reproduced with permission from Healey PR, Mitchell P. The prevalence of optic disc pits and their relationship to glaucoma. *J Glaucoma* 2008;17:11–14.



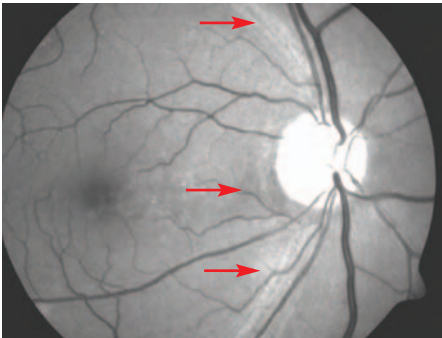
epithelium and choroid layers to create a dark background. The normal arcuate NFL appears as fine bright striations. Viewed from the superior arcuate NFL to the inferior NFL, a bright–dark–bright pattern is visible, the dark area being the region between the disc and the macula (Figure 3.22). The inferior arcuate NFL is a larger area and is easier to see than the superior arcuate NFL.

A localized NFL defect appears as a dark wedge that follows the pattern of the NFL (Figure 3.23). Characteristically a defect:

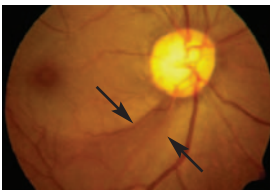
- is wider than an arteriole
- touches the edge of the disc
- increases in width towards the periphery.

Such defects have a strong predictive value for future functional changes. The specificity is very high ('SpPIN'; see Box 3.1, page 31) but the sensitivity is poor. The defects are a definite sign of pathology, but can also occur in diseases other than glaucoma. Localized defects are sometimes easily seen on indirect ophthalmoscopy.

Diffuse NFL defects are more difficult to detect because the normal bright–dark–bright pattern is lost. The pattern looks more like dark–dark–dark (Figure 3.24). Better visibility of the superior NFL defect relative to the inferior NFL is also suspect.



**Figure 3.22** Normal retinal nerve fiber layer, showing distinct bright–dark–bright striations (arrowed).



**Figure 3.23** Localized (wedge-shaped) retinal nerve fiber layer defect, increasing in width towards the periphery.