

# Community Eye Health Journal



Double vision is a distressing symptom that warrants investigation. **TANZANIA**

DANIEL ETYAALÉ  
(ORIGINAL PHOTOGRAPH)

## Neuro-ophthalmology

### Understanding vision and the brain



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Diseases which affect the visual pathway or the nerves to the eye muscles are often serious. This article summarises the anatomy and function of the 2nd, 3rd, 4th and 6th cranial nerves and the signs and symptoms which are important in making a correct diagnosis.

Neuro-ophthalmology includes the specialties of neurology and ophthalmology. Skills of both specialties are needed to assess patients. A detailed history and clinical examination are needed in order to make a differential diagnosis and create a management plan.

There are two broad groups of neurological conditions that can affect someone's vision:

- 1 Those which affect the visual pathways in the brain and
- 2 Those which affect eye movements.

If the condition affects the visual pathways, there can be loss of visual acuity, loss of visual field, or difficulties

in understanding the visual world, depending on where the lesion is in the visual pathway. If the condition affects the 3rd, 4th or 6th cranial nerves, the main symptom is double vision (diplopia) due to restricted eye movements.

**The visual pathways** comprise the optic nerve, optic chiasm, optic tract, optic radiation and the visual cortex in the occipital lobes.

Nerve impulses arising in the retina travel via the optic nerve to the optic chiasm. In the optic chiasm there is "crossing over" of some visual information, so that images which fall on the right visual field of each eye

*Continues overleaf* ➤



## About this issue

In this issue of the *Community Eye Health Journal* we briefly discuss the anatomy of vision, before explaining conditions presenting with double vision and/or sudden loss of vision which may be life and/or sight-threatening. There are articles on how to assess a patient with neuro-ophthalmological symptoms; the differential diagnosis of optic atrophy as well as how to counsel a patient with visual loss.

Large textbooks are written on neuro-ophthalmology and therefore it is not possible to discuss in detail the diagnosis and management of many neuro-ophthalmological diseases, but we trust this introduction will provide an insight to the important conditions and encourage you in further reading.

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go to left visual cortex via the optic tract and the optic radiation. Visual information from the left visual field in each eye goes to the right visual cortex.

## Neurological conditions which affect visual function

Conditions which affect the visual pathways can lead to loss of visual acuity (ranging from blurred vision to complete visual loss), impaired colour vision, or visual field loss (which may be central, nasal or temporal) or affect the right or the left visual field. The type of visual field loss depends on which part of the visual pathway is affected.

### Visual Loss

Loss of central visual acuity, with impaired colour vision and contrast sensitivity, suggest an optic nerve condition.

The two main clinical presentations are:

- **Optic neuritis**, with a decrease in vision over a few hours or days and sometimes pain on eye movement from inflammation of the optic nerve and surrounding sheath. Visual acuity is decreased, there is an afferent pupil defect (pupil on the affected side reacts poorly to light), and the optic disc may appear swollen (papillitis). It may be unilateral or bilateral. Causes include multiple sclerosis, some medications and toxins (e.g. cassava or methanol).
- **Optic atrophy**, in which the optic disc is pale. The visual acuity is reduced, colour vision is affected and there is an afferent pupil defect. There are many causes of optic atrophy (see page 69). Optic atrophy may develop slowly due to a progressive cause, or follow an episode of acute optic neuritis.

### Visual field defects

Lesions along the visual pathway, from the optic nerve to the occipital lobe, can present with localising visual field defects (**Figure 1**).

- **Optic nerve lesions** cause central, centrocaecal or altitudinal field defects. Central vision is primarily affected.
- **Optic chiasm lesions** caused by space-occupying lesions around the sella turcica (e.g. pituitary tumours) typically cause bitemporal hemianopia. Central vision is usually spared.

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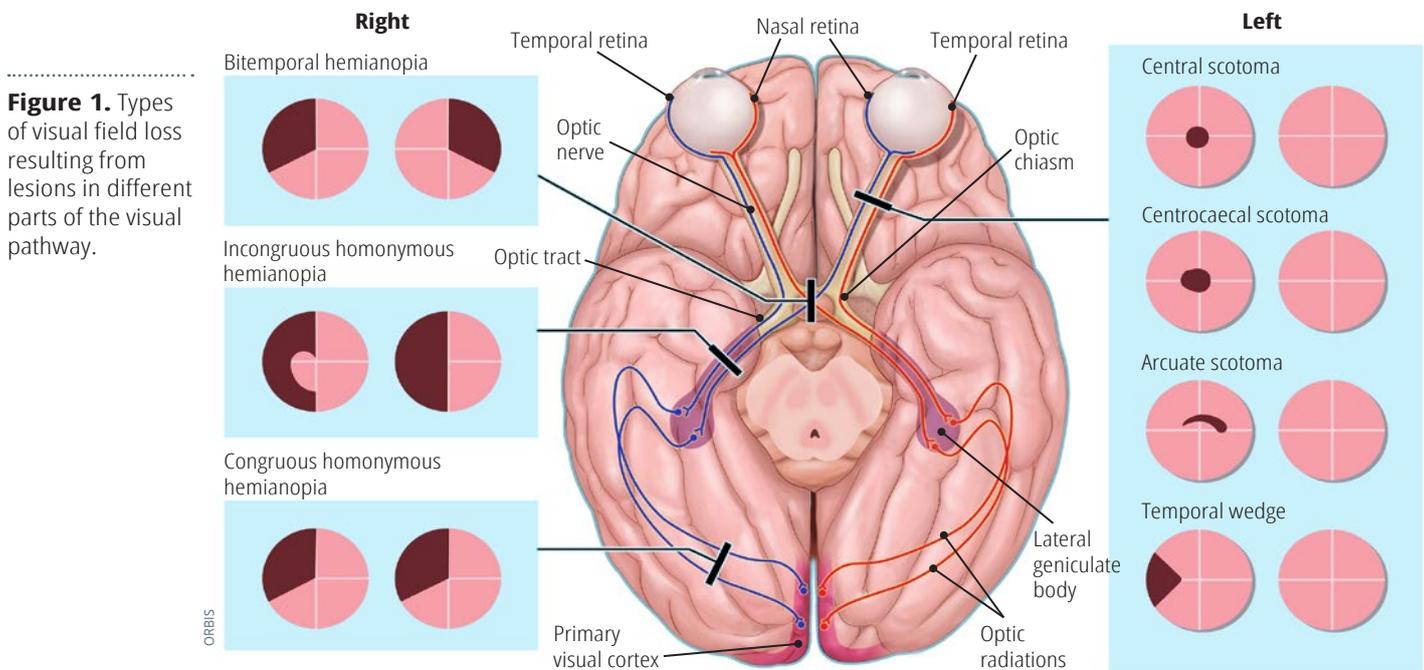
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**Figure 1.** Types of visual field loss resulting from lesions in different parts of the visual pathway.

- **Optic tract, optic radiation and occipital lobe** lesions result in loss of vision in the opposite hemifield which patients experience as loss of vision on the right or left.

#### Higher visual centres

Visual information goes from the visual cortex to the 'visual association areas' where what we see is interpreted. For example, visual information which goes from the occipital lobe to the temporal lobe enables faces and objects to be recognised, while visual information which goes from the occipital lobe to the parietal lobe allows objects to be localised in space.

#### Neurological conditions which affect eye movement

(See Figure 1 on page 64)

Diplopia (double vision) is the commonest symptom and results from misalignment of the visual axes in the eyes.

- Disorders causing diplopia may arise due to lesions affecting any part of the 3rd, 4th and 6th cranial nerves, or due to diseases affecting the extra-ocular muscles, e.g. myasthenia gravis or dysthyroid eye disease.
- Diplopia is experienced as either images lying side by side (horizontal diplopia) from involvement of the medial and lateral rectus muscles, or images

lying one above the other (vertical diplopia) from involvement of the eye elevators.

- Separation of images is greatest in the direction of the action of the weak muscles and the false image created by the affected eye is usually the outermost image.

#### Associated neurological symptoms

- Facial numbness (especially over the forehead), occurring in conjunction with diplopia from 3rd, 4th or 6th cranial nerve lesions, suggests disease at the cavernous sinus or orbit.
- Impaired vision on the same side of 3rd, 4th and 6th cranial nerve lesions suggests an orbital apex lesion.
- Weakness of the lower half of the face, arm and leg unilaterally (hemiparesis) occurring with 3rd or 6th nerve palsy (diplopia) suggest a lesion of the brainstem.
- Dysphagia (difficulty swallowing), dysarthria (slurred speech) with fluctuating double vision and eyelid drooping (ptosis) is suggestive of myasthenia gravis; a disorder of the neuromuscular junction.
- Patients with thyroid eye disease may have diplopia, proptosis, lid retraction (staring gaze) and an enlarged thyroid gland from Grave's disease.

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# Red flags in neuro-ophthalmology



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Some diseases of the brain presenting with visual symptoms are life threatening and need urgent management. This article discusses possible causes of double vision, vision loss with headache, and non-ocular vision loss.



**Papilloedema in a 23-year-old woman. Headache for two months. Worse on waking. Vision 6/6 in both eyes.**

The three most important 'red flag' symptoms that indicate that a patient may need neuro-ophthalmological assessment are:

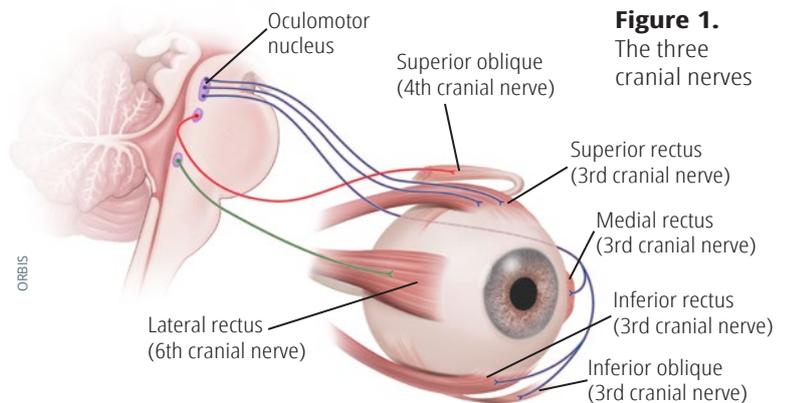
- 1 Sudden onset of double vision (diplopia)
- 2 Headache accompanied by vision loss (without an ocular cause)
- 3 Visual loss after ocular causes have been excluded

If a patient presents with any of the symptoms above, you must take a detailed history (Table 1).

## 1 Sudden onset of double vision

Each eye is moved by six muscles which are innervated by three "cranial" nerves (the 3rd, 4th and 6th nerve) (Figure 1). If the nerves are affected then the eye cannot move normally, which results in double vision. The 3rd nerve also innervates the upper eyelid (Table 2).

First exclude monocular diplopia by asking the patient to cover each eye in turn. If the double vision persists when looking with just one eye, then this is usually due to an ocular problem (e.g. cataract) and does not have a neurological cause.



**Figure 1.** The three cranial nerves

### Table 1 Taking a history

Questions to ask	Clinical interpretation
Is the double vision worse in any direction of gaze?	The direction of gaze in which the double vision is worst signifies the most likely eye muscle involved.
Are the images side by side; or is one image tilted and above the other?	In 6th nerve palsies the images are side by side; In 4th nerve palsies one image is tilted.
Has there been a recent head injury?	Trauma to the brain or orbit can affect the nerves which control eye muscle movements.
Does the double vision get worse as the day progresses or after exercise?	If the condition gets worse with use of the muscle then this is typical for myasthenia gravis; there may be eyelid drooping (ptosis) or diplopia as the day proceeds.
Is there any head or eye pain?	Pain is an important clue: it usually indicates infection or inflammation. Tumours are less likely to be painful.
Are there any systemic symptoms or diseases?	Hypertension and diabetes can both cause loss of vision and diplopia.

### Table 2 Examination

Questions to ask	Clinical interpretation
Is the upper eyelid drooping?	Ptosis may be due to myasthenia gravis or a third nerve palsy, or may be congenital.
How do the eyes move (each eye alone and both eyes together)?	Assess the position of the eyes looking straight ahead (check for squint), and the movements of each eye alone and together in all 9 positions of gaze (Figure 3 on page 67). Limitation of movement in a certain direction indicates disease of the affected muscle or the cranial nerve which innervates it. 6th nerve palsy: eye cannot look out (abduction). 3rd nerve palsy: ptosis and eye cannot look up and in.
Are the pupils of equal size?	If one pupil is larger than the other, this suggests 3rd nerve palsy relating to the eye with the larger pupil.
Does the pupil react normally to light?	A non-reactive pupil indicates a damaged optic nerve or prior use of dilating drops.

### What should I do?

Refer all patients with double vision for further investigation. Some may have life-threatening conditions.

## 2 Headache accompanied by vision loss (without an ocular cause)

The brain is encased by the skull and meninges and is bathed in cerebrospinal fluid. If the flow of fluid is blocked, by a tumour for example, this raises the pressure inside the head (intracranial pressure), causing headache, sometimes with nausea or vomiting. Raised intracranial pressure can lead to swelling of the optic nerve head (papilloedema), usually in both eyes. If the raised pressure persists, the optic nerves become atrophic; i.e. they become paler than normal.

Both headaches and visual loss are common. Before suspecting a neurological cause, examine the patient to exclude eye conditions which might be responsible for the visual loss (Table 4).

### Taking a history

Ask questions about any aches or pain using the mnemonic 'SOCRATES' (Table 3).

**Table 3** Structured history

Questions to ask	Clinical interpretation
<b>Site:</b> Where is the headache? The patient may describe or point to the location	Pain overlying the sinuses may suggest sinusitis, whereas periorbital pain suggests orbital pathology.
<b>Onset:</b> How long have they had the headache? Is it worse at any time of the day?	Migraine is a common cause of recurrent severe headache which may last hours or even days. Pain which is worse in the morning on waking up may be due to raised intracranial pressure. The headache may be associated with nausea and vomiting.
<b>Character:</b> Can they describe the quality or type of pain?	Dull, constant, unrelieved pain over days or weeks may suggest a space-occupying lesion. A sudden, throbbing pain is more typical of vascular problems like migraine or an aneurysm.
<b>Radiation:</b> does the pain start in one place and then extend/ spread to another?	Pain that starts in one place and seems to move or 'radiate' to another suggests that is generated by the irritation of a nerve.
<b>Associated vision loss</b>	Severe constant headache with gradual visual loss suggests either compression of the optic nerve, or longstanding raised intracranial pressure.
<b>Time course:</b> have the symptoms changed over time?	Symptoms which are constant and getting more severe may indicate a serious progressive condition e.g. a tumour. Intermittent headaches are more typical of vascular or inflammatory conditions.
<b>Exacerbating factors:</b> What makes the headache worse or better?	A headache which is worse when lying down or bending down may be due to raised intracranial pressure.
<b>Severity:</b> Ask the patient to rate the severity on a scale from 1 (mild) to 10 (very severe)	Any headache that interferes with the patient's daily activities should not be ignored.

**Table 4** Examination

Questions to ask	Clinical interpretation
Are the pupils of equal size and do they react normally to light?	See Table 2.
Is there optic disc swelling or atrophy?	Swelling of the optic disc can be due to raised intracranial pressure (papilloedema) or inflammation (papilitis). Optic atrophy may be due to longstanding compression of the optic nerve or vascular or toxic damage to the nerve.

### What should I do?

Refer all patients with headache and persistent visual loss for further investigation. The referral must be urgent if they have papilloedema. Some may have life-threatening conditions.

## 3 Visual loss after ocular causes have been excluded

Most causes of visual loss are due to diseases of the eye. Ocular conditions must be excluded by careful examination of the eye before considering a neurological cause of poor vision (Table 5).

Progressive vision loss with no ocular cause must be taken seriously.

**Table 5** Visual loss

Questions to ask	Clinical interpretation
Is the vision loss in one or both eyes?	Unilateral vision loss indicates a problem within the eyeball or optic nerve in the orbit.
Has there been any change in the vision since onset?	Vision loss that is progressively worsening may suggest a space-occupying lesion.
Are there any other symptoms?	Vomiting, seizures, and changes in mood or mental state may indicate increased intracranial pressure. Call for URGENT referral.
Is there a fever?	Fever indicates infection, check the sinuses, ears, orbit, and for neck stiffness.

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## More red flags

### Proptosis

Proptosis is anterior displacement of the globe. It may be due to space-occupying lesions in the orbit. Adults with acquired proptosis need to be evaluated for thyroid disorder. Pulsatile proptosis, painful proptosis and all cases of proptosis associated with vision loss should be referred for urgent evaluation.

### Ptosis

Drooping of the upper lid is called ptosis. All cases of acquired ptosis should be evaluated by an ophthalmologist. Marked unilateral ptosis with ocular deviation down and out are signs of a 3rd cranial nerve palsy. If associated with severe sudden onset of unilateral headache this can be due to an intracranial aneurysm (dilated artery). Patients must be referred for immediate neuro-ophthalmological review.

Bilateral ptosis which gets worse as the day progresses may be due to myasthenia gravis.

Partial ptosis with a smaller (constricted) pupil on the same side is due to damage to the sympathetic nerves which supply the muscles in the eyelid and iris – this is called Horner's syndrome and the cause needs to be investigated.

# Basic clinical examination of a patient with neuro-ophthalmology symptoms



**Anand Moodley**

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This article discusses how to clinically assess the visual pathway, examine the optic disc, check the pupil light reflexes and assess the extraocular movements in patients presenting with visual loss and/or diplopia.



Testing a patient's visual fields by confrontation. TANZANIA

HEIKO PHILIPPIN

If a patient presents with potential neuro-ophthalmology signs and symptoms (see article on page 64), a basic neuro-ophthalmology examination should be undertaken. If done systematically, as described in this article, it can be informative in making a differential diagnosis and deciding on management.

A basic neuro-ophthalmological examination can be done with a minimum of equipment (see box below). Subsequent examination depends on what has been found and may involve full ocular and neurological examinations as well as investigations.

## Equipment required for a neuro-ophthalmological assessment

- Snellen chart for far vision
- Pen torch for pupil reaction
- Occluder for cover testing
- Red probe or red mydriatic bottle top for colour desaturation testing
- Direct ophthalmoscope for funduscopy

Also useful:

- Ruler for lid function and pupil diameter
- Cotton wool and office pin for sensation testing
- Pseudo-isochromatic plates for colour vision

Both the **visual pathway** and **oculomotor functioning** should be assessed.

## Visual pathway

### Visual acuity

The presenting and best corrected (with pinhole) visual acuity is obtained using a Snellen or LogMar chart for distance vision.

### Colour vision

Colour vision is tested using the Ishihara pseudo-isochromatic plates. The number of correct responses as an index of the total number of plates used (**Figure 1**). Such quantification allows for comparison during follow-up evaluations.

### Funduscopy

Ophthalmoscopy is performed with particular attention to the optic disc, looking at possible swelling and the colour of the disc.

### Visual fields

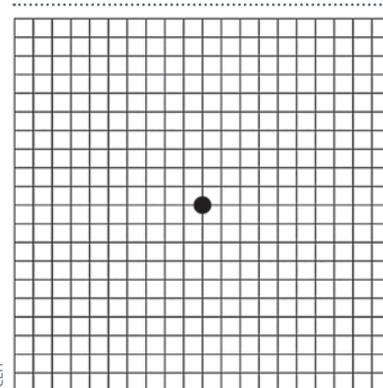
Visual field testing using a perimeter or, if not available, by confrontation, is done for each eye separately. Check the central vision with an Amsler grid (**Figure 2**) and then test the peripheral vision in each of the four quadrants.

**Figure 1.** Ishihara pseudo-isochromatic plates for colour vision testing



ICEH

ICEH



**Figure 2.** Amsler grid used for checking distortion of central vision

**Read more:**  
[www.cehjournal.org/article/visual-field-testing-for-glaucoma-a-practical-guide](http://www.cehjournal.org/article/visual-field-testing-for-glaucoma-a-practical-guide)

## Pupils

Examination of the pupil size, symmetry and reaction to a light stimulus provides information about both the afferent and efferent pathways.

Testing the pupil reflex in the dark is preferred as this makes it easier to appreciate constriction than in ambient room light.

A brisk, direct light reflex implies that the optic and 3rd nerve on that side are working well.

The swinging torch test is done by swinging the light source from one eye to the next while pausing for 3–5 seconds at each eye. It is used to detect a relative afferent pupillary defect (RAPD). This indicates that one optic nerve is not functioning as well as the other (the one that dilates as the torch light shines in it).

A dilated pupil that is non-reactive to a light source, together with ptosis, suggests a 3rd cranial nerve palsy; whereas a constricted, reactive pupil with partial ptosis suggests Horner syndrome from sympathetic impairment to the eye.

## Oculomotor functioning

### Ocular alignment

Check the alignment of the eyes. This is performed by comparing the light reflex from the cornea of both eyes. Hold a torch 1 metre in front of the eyes and look for the light reflex on the cornea (Hirschberg test). In the primary gaze (looking straight ahead at the torch light), the light reflexes should be in a symmetrical position on each cornea. If one eye is turning in, this is called esotropia, whereas if the eye is turning out it is called exotropia.

If you find that an eye is misaligned, use the cover test to confirm this. For example, say that you have observed the left eye turning in when both eyes look straight ahead. If you then cover the right eye (the normal eye), you should see the left eye (the deviated eye) turn out to take up fixation (i.e. look straight ahead).

If the left eye (in this example) does not realign when the other (normal) eye is covered, then the patient is either not cooperating or the eye is blind.

### Extraocular movements

Test the ocular movements of one eye at a time in the 9 positions of gaze. Check for any limitation of movement (Figure 3).

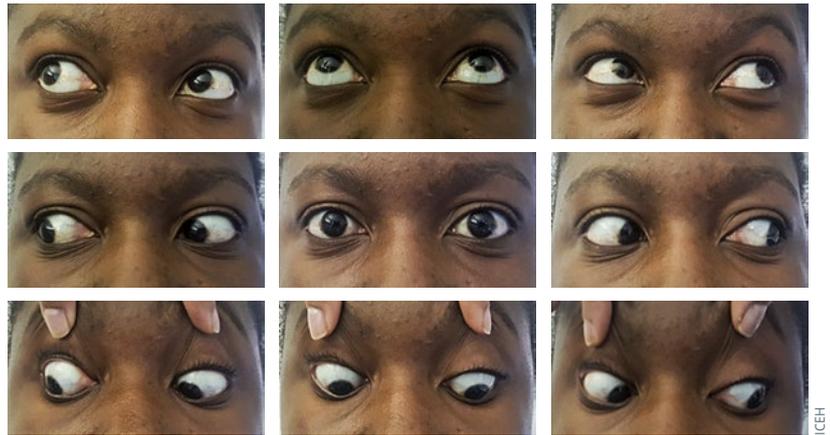
Test the ocular movements of both eyes together to see if double vision is elicited in any position of gaze.

### Examination of eyelid movement

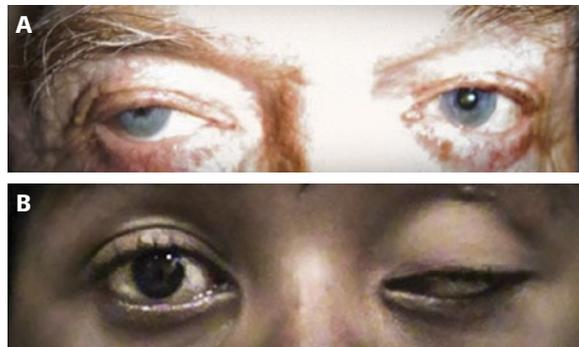
Two muscles are responsible for opening the upper eyelid: the levator palpebrae superioris (supplied by the 3rd cranial nerve) and Muller's muscle (supplied by the sympathetic pathway). Hence, in 3rd cranial nerve palsies and Horner syndrome, ptosis may result (Figure 4).

The eyelids are closed by the orbicularis oculi muscle, which is supplied by the facial (7th) cranial nerve. In 7th nerve palsy, there is lagophthalmos (an inability to close the eye).

**Figure 3.** Testing of the 9 cardinal positions of the eyes



**Figure 4.** Examination of the eyelids: image A shows right-sided ptosis from Horner syndrome and image B shows left-sided ptosis from 3rd nerve palsy



## Nystagmus

Nystagmus is rhythmical oscillations of the eyes. It may occur in the primary gaze or in horizontal or vertical gaze. Nystagmus has many causes, including ocular, vestibular and cerebellar conditions.

Jerk nystagmus is characterised by a slow 'drift' in one direction which is repeatedly corrected by a fast 'recovery' movement in the opposite direction.

Pendular nystagmus has equal speed in both directions and is commonly seen in congenital nystagmus.

### Further reading

Clarke C, Howard R, Rossor M, Shorvon S. *Neurology, Queen Square Textbook*. Institute of Neurology, University College London. 2009 Wiley-Blackwell

Kidd DP, Newman NJ, Biousse V. *Blue Books of Neurology, Neuro-ophthalmology*. 2008 Saunders

Rucker JC, Kennard C, Leigh RJ. *The neuro-ophthalmological Examination. Handbook of Clinical Neurology: Neuro-ophthalmology, Vol 102 (3rd series)*. Chapter 3, p71-94, 2011 Elsevier

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## Examination of the eyes in an unconscious patient

Examination of the eyes is important in the evaluation of the unconscious patient; it includes examination of the pupils and fundoscopy.

### Pupils

Pupillary testing involves assessment of the pupil size and reaction to light.

- In an unconscious patient, normal size and normally reactive pupils may suggest a metabolic encephalopathy from kidney failure, liver failure or electrolyte abnormalities.
- A unilateral or bilateral dilated pupil can suggest 3rd nerve palsy due to herniation of the brain through the tentorium cerebelli. This is a surgical emergency.
- Bilateral pinpoint pupils that react to light may indicate pathology in the pons of the brain such as haemorrhage; or opioid or organophosphate poisoning.

### Fundoscopy

- The presence of swollen optic discs suggest raised intracranial pressure.
- Preretinal haemorrhage may suggest ruptured intracranial blood vessels.

# How to test for a relative afferent pupillary defect (RAPD)



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This article explains how careful examination of the pupil light reflex can reveal valuable information about the afferent (optic nerve) and efferent (oculomotor nerve) light reflex pathway, and hence the functioning of these two cranial nerves.

The 'swinging light test' is used to detect a relative afferent pupil defect (RAPD): a means of detecting differences between the two eyes in how they respond to a light shone in one eye at a time. The test can be very useful for detecting unilateral or asymmetrical disease of the retina or optic nerve (but only optic nerve disease that occurs in front of the optic chiasm).

The physiological basis of the RAPD test is that, in healthy eyes, the reaction of the pupils in the right and left eyes are linked. In other words, a bright light shone into **one** eye leads to an equal constriction of **both** pupils. When the light source is taken away, the pupils of both eyes enlarge equally. This is called the **consensual light reflex**.

To understand how the pupils react to light, it is important to understand the **light reflex pathway** (Figure 1). This pathway has two parts.

- 1 The **afferent** part of the pathway (red) refers to the nerve impulse/message sent from the pupil to the brain along the optic nerve when a light is shone in that eye.
- 2 The **efferent** part of the pathway (blue) is the impulse/message that is sent from the mid-brain back to both pupils via the ciliary ganglion and the third cranial nerve (the oculomotor nerve), causing both pupils to constrict, even though only one eye is being stimulated by the light.

A **positive** RAPD means there are differences between the two eyes in the afferent pathway due to retinal or optic nerve disease. If the light used is sufficiently bright, even a dense cataract or corneal scar will not give a RAPD as long as the retina and optic nerve are healthy. Indeed, the test can be used to assess the health of the retina and optic nerve behind a dense cataract, for example.

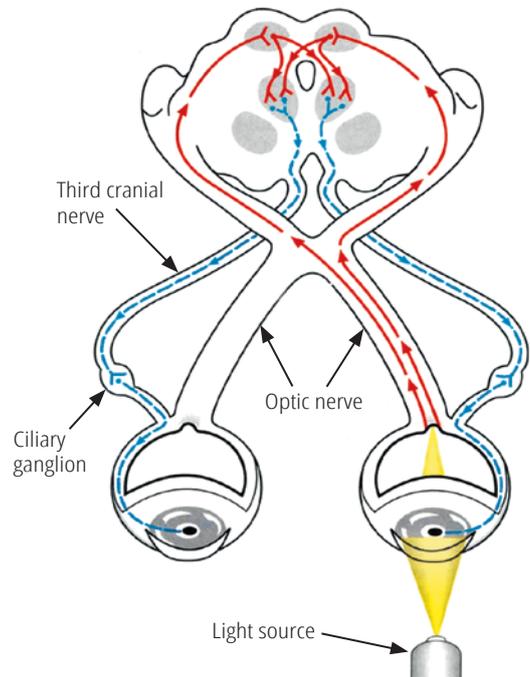
In glaucoma, if other tests of visual function (e.g. visual fields) are not possible, detecting a RAPD can be very useful as it indicates that there is more optic nerve damage in one eye than in the other, even if the visual acuity in both eyes is equal.

**NOTE: If the glaucomatous damage is equal in the two eyes, there will be no RAPD, however severe the damage is.**

## The swinging light test

In a normal swinging light test (i.e. there is no RAPD) the pupils of both eyes constrict equally regardless of which eye is stimulated by the light (Figure 2). In an abnormal swinging-light test (i.e. there is a RAPD) there is less pupil constriction in the eye with the retinal or optic nerve disease (Figure 3).

**Figure 1.** The light reflex pathway showing the afferent path (red) and the efferent path (blue)



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## Steps

- Use a bright torch which can be focussed to give a narrow, even beam of light. Perform the test in a semi-darkened room. If the room is too dark it will be difficult to observe the pupil responses, particularly in heavily pigmented eyes.
- Ask the patient to look at a distant object, and to keep looking at it. Use a Snellen chart, or a picture. This is to prevent the near-pupil response (a constriction in pupil size when moving focus from a distant to a near object). While performing the test, take care not to get in the way of the fixation target.
- **Move** the whole torch deliberately from side to side so that the beam of light is directed directly into each eye. **Do not swing** the beam from side to side around a central axis (e.g. by holding it in front of the person's nose) as this can also stimulate the near response.
- Keep the light source at the same distance from each eye to ensure that the light stimulus is equally bright in both.
- Keep the beam of light steadily on the first eye for at least 3 seconds. This allows the pupil size to stabilise. Note whether the pupil of the eye being illuminated reacts briskly and constricts fully to the light. Also note what happens to the pupil of the other eye: does it also constrict briskly?
- Move the light quickly to shine in the other eye. Again, hold the light steady for 3 seconds. Note whether the pupil being illuminated stays the same size, or whether it gets bigger. Note also what happens to the other eye.
- As there is a lot to look at, repeat the test, observing what happens to the pupils of both eyes when one and then the other eye is illuminated.

When the test is performed on someone with unilateral or asymmetrical retinal or optic nerve disease, a RAPD should be present (Figure 3). The following happens:

- When the light is shone into the eye with the retinal or optic nerve disease, the pupils of both eyes will constrict, but not fully. This is because of a problem with the afferent pathway.
- When the light is shone into the other, normal (less abnormal) eye, both pupils will constrict further. This is because the afferent pathway of this eye is intact, or less damaged than that of the other eye.
- When the light is shone back into the abnormal eye, both pupils will get larger, even the pupil in the normal eye.
- It doesn't matter whether you start with the eye you think has the greater problem or the healthier eye: as long as the light is switched from one eye to the other and back again the signs should become apparent.

Sometimes the RAPD is obvious, as the pupil in the (most) affected eye very obviously gets larger when that eye is illuminated. But the signs can be more subtle (see **Table 1**).

## Specific situations

### Hippus

Normal pupils, particularly those of young people, sometimes show slight fluctuation in size (of less than 1 mm) even when the light shining into the eye is constant. This is called hippus and it can make eliciting a RAPD more difficult.

### Non-reactive pupils

A RAPD can still be detected even if one pupil cannot change size (i.e. it is fixed), because of trauma, posterior synechiae or because dilating or constricting eye drops have been used (**Figure 4**). Having established that the pupil of one eye does not change size, regardless of which eye has the light shone into it, concentrate on the eye where the pupil is reactive. Note what happens to the reacting pupil when the light is shone into each eye in turn. **Figure 4** shows what happens when the eye with the afferent pathway defect is also the eye with the fixed pupil. If the (more) normal eye is the one with the fixed pupil then, as the light moves from this eye to the other eye, the reacting pupil will dilate.

**Table 1** The grading of a RAPD in the swinging light test

<b>Amaurotic</b>	This is seen when one eye has no perception of light. The pupil of this eye only constricts when light is shone into the other eye. When the light is shone back into the eye with no perception of light the pupil rapidly enlarges against the light.
<b>3-4+</b>	The pupil enlarges as soon as the light is swung from the normal eye into the abnormal eye.
<b>1-2+</b>	The pupil enlarges, but only after a short delay, after the light is swung from the normal eye into the abnormal eye.
<b>Subtle/trace</b>	Sometimes the pupils of both eyes can enlarge in the short time interval between shining the light in the normal eye and the abnormal eye. If this happens, the pupil of the abnormal eye may constrict a little bit before enlarging.

### Asymmetric refractive errors and/or amblyopia

These occur when the vision is poor but the eye itself is normal, and are not associated with a RAPD.

### Maculopathy

Unless very severe, this is not usually associated with a RAPD and in eyes where the macular damage is sufficient to result in an RAPD, the grade is rarely more than 1-2+ (**Table 1**). Extensive retinal damage, major retinal vascular occlusion, or retinal detachment, by contrast, can lead to a high-grade RAPD.

## Causes of RAPDs

Common causes of unilateral optic nerve disorders that can be associated with a RAPD include ischaemic optic neuropathy, optic neuritis, optic nerve compression (orbital tumours or dysthyroid eye disease), trauma, and asymmetric glaucoma. Less common causes include infective, infiltrative, carcinomatous, or radiation optic neuropathy.

A RAPD is an extremely important localising clinical sign that can be detected by a simple, quick, non-invasive clinical test, provided that the test is performed carefully and correctly.

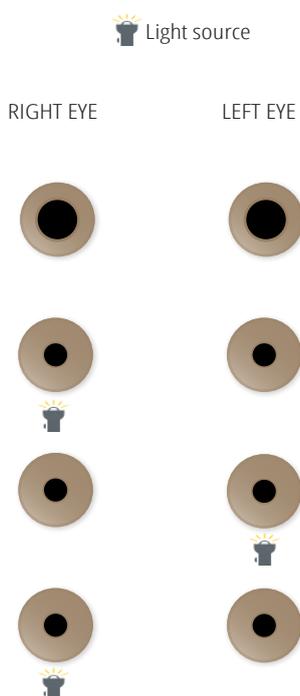
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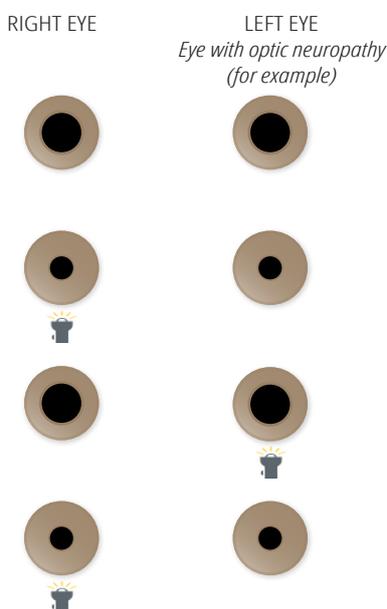
**Figure 2. Swinging-light test – normal (no RAPD)**

Illumination of either eye induces normal and equal pupil responses in both eyes (consensual responses).



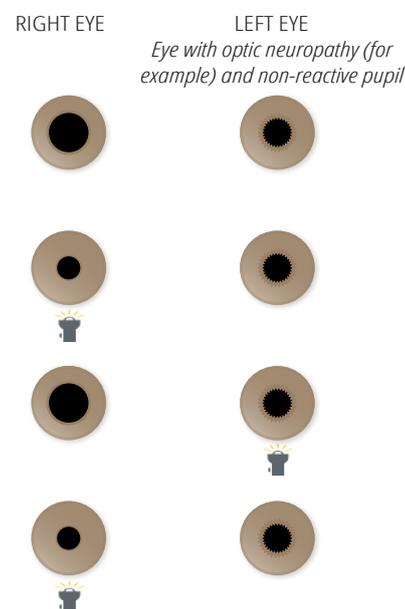
**Figure 3. Swinging-light test – left RAPD**

Illumination of the (more) normal right eye causes both pupils to constrict. When the light is moved to the (more) abnormal left eye (e.g. with optic neuropathy), both pupils dilate (constrict less), the left pupil dilating despite the light being shone directly at it. Returning the light to the (relatively) normal right eye results in constriction of both pupils again.



**Figure 4. Swinging-light test: left RAPD + non-reactive left pupil**

Illumination of the relatively normal right eye causes only right pupil constriction. When the light is moved to the abnormal left eye (e.g. fixed pupil and optic neuropathy), the right pupil dilates (constricts less). Returning the light to the right eye results in constriction of the right pupil again. In this situation it is only necessary to observe the eye with the reactive pupil in order to identify an RAPD.



# Counselling patients with sudden, irreversible sight loss



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Sudden loss of vision is devastating to the patient and close relatives. This article discusses how to talk with someone who has lost their vision and how to help them with their concerns and questions.

Sudden loss of sight is a life-changing event that changes the patient's view of the world and how they perceive themselves and others. In this article we will consider the emotional and psychological impact of those affected by sudden and irreversible sight loss.

## Grief and loss

It is recognised that people affected by sight loss will understandably experience very strong and at times over whelming feelings of grief and loss. In cases of sudden loss the reaction is often severe. Patients often experience panic and confusion, making it difficult to assimilate information and to make rational decisions. For the majority of patients with diagnosed sight-threatening conditions, the reality is feelings of helplessness, hopelessness and fear. These are often compounded by insomnia, loss of appetite, palpitations, visual hallucinations, aggression, anger, tension, frustration, disorganisation, irritability, restlessness, inability to concentrate, apathy, and depression.

## Case study 'Linda' a patient, affected by sudden irreversible sight loss

Linda is fifty years old, she is single and previously worked as a designer. She has had glaucoma for many years. Nine months ago her vision started to deteriorate. Following a period of further investigation and treatment Linda was assessed as having near complete sight loss.

Linda spent her first counselling sessions talking about her losses and frustrations since her sudden sight loss. Linda says that she is going out less often and admits to feeling lonely and isolated; she is feeling scared and fearful about the future. Her mood is low and at times she feels very depressed. Linda says that her loss of sight doesn't feel real and she keeps hoping that it will improve. Linda is angry about the impact her condition is having on her independence. Linda says she is particularly worried about becoming a burden to others.

It is common for patients with sudden sight loss to feel blame in relation to anger. Sometimes patients feel that more should have been done. In Linda's case at first she initially blamed herself, for not noticing her symptoms earlier.



Counselling a patient. UNITED KINGDOM

COMMUNICATIONS DEPT. MOORFIELDS EYE HOSPITAL

## Rehabilitation, stigma and shame

Linda was reluctant to use any aids to help with her sight loss including a white stick or cane. She was reluctant to work with the staff to begin her rehabilitation. She spoke of feelings of stigma and shame.

After some months Linda agreed to register as severely sight impaired. Registering with a certificate of vision impairment enables people in the UK to receive certain benefits from social services and tax concessions. But the acknowledgement of being registered as blind (severely sight impaired), often has a huge emotional impact, as it confirms that the sight loss is permanent. Eventually Linda decided that she was going to start rehabilitation training in the form of long cane training. She also attended a course entitled 'Learning to live with sight loss' at the National Association for the Blind.

Like all people affected by sudden sight loss, Linda is going through an on-going period of adjustment. She admitted that her certificate of vision impairment registration was significant in the ongoing process of her adjustment to her sight loss

## Summary

Adjusting to sight loss is hard and exhausting work, and is an active process. The psychological impact can be frightening and disturbing but it is the process by which a person can heal.

Linda said that she found the counselling sessions useful as they gave her space to think and to talk things through. In the sessions, she had been able to talk freely, express her anger, cry and vent her feelings in a way that she had not able to do with her friends or family.

As counsellors, we recognise that people who experience sudden sight loss have to be ready emotionally before they can begin rehabilitation. We talk of patients affected by sight loss going through an on-going period of adjustment involving re-conceptualisation of self, perhaps ultimately leading to acceptance.

**"You are just a visitor passing through my country"**, a statement made by a patient who had experienced sudden sight loss. It illustrates how patients can feel that at times, healthcare professionals do not understand the psychological impact of sudden sight loss.

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# Differential diagnoses of the pale / white / atrophic disc



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Optic atrophy, pallor of the optic nerve head, is a sign found in patients with visual loss due to pathology of the optic nerve or retinal ganglion cells. There are many causes. This article discusses the differential diagnosis of optic atrophy.



Examining the optic disc with a 90D lens at the slit lamp. NIGERIA

Optic atrophy is not a disease in itself but a clinical sign. It refers to pallor of the optic disc which results from irreversible damage to the retinal ganglion cells and axons. The axons of the retinal ganglion cells make up the optic nerve and continue onto the optic chiasm, optic tract and up to the lateral geniculate body before they synapse. Injury to the retinal ganglion cells and axons anywhere along their course from the retina to the lateral geniculate body may result in optic atrophy.

The causes of optic atrophy are numerous; they include:

- 1 Inflammation
- 2 Ischaemia
- 3 Compression, including raised intracranial pressure

## Clinical features in optic atrophy

- Diminution of vision (central acuity/colour vision/visual field defects)
- Afferent pupil defect
- Optic disc pallor
- Reduced number of small blood vessels on the disc surface (Kestenbaum sign)
- Attenuation (thinning) of blood vessels around the disc
- Thinning of the retinal nerve fiber layer.

## Causes of pseudo optic atrophy

### Non pathologic causes of a pale disc

- Disc is examined with very bright light
- Large physiologic cup in axial myopia
- Post-cataract extraction

### Other (non optic atrophy) causes of a pale disc

- Myelinated retinal nerve fibers
- Optic disc coloboma
- Optic disc hypoplasia

- 4 Nutritional deficiencies / effect of toxins, including epidemic
- 5 Trauma
- 6 Hereditary conditions and childhood optic atrophy

Clinically, optic atrophy is associated with a decrease in visual acuity and visual field defect (see upper box, left).

There are other causes of disc pallor which are not due to optic atrophy (see lower box, left) which should be excluded.

## Ophthalmoscopic classification

Optic atrophy can be classified into primary, secondary and consecutive optic atrophy. Each has characteristic features which may help to differentiate between them (**Table 1** on page 70).

In **primary optic atrophy** (**Figure 1** on page 71) there is no previous swelling of the optic disc. The disc is white, the margins are distinct and the retinal blood vessels at the optic nerve head appear normal. **Secondary optic atrophy** (**Figure 2** on page 71) is a consequence of long-standing swelling of the optic disc, which may be due to inflammation, ischaemia or raised intracranial pressure. The disc is greyish in colour and the margins are blurred. There is fibrosis (gliosis) of the optic nerve head and the blood vessels may appear indistinct or narrowed. **Consecutive optic atrophy** (**Figure 3** on page 72) results from chorioretinal disease such as retinitis pigmentosa or toxoplasmosis chorioretinitis. The cause of consecutive optic atrophy is usually obvious from the retinal appearance and so will not be discussed further.

Glaucoma is an important cause of optic atrophy. In glaucoma there is characteristic pathological cupping of the optic disc which together with the typical visual field loss distinguishes glaucoma from other causes of optic atrophy. Glaucoma will also not be considered further in this article. (For articles on glaucoma please see CEHJ Comm Eye Health Vol. 25 No. 79 & 80 2012).

Continues overleaf ➤

**Table 1** Ophthalmoscopic classification of optic atrophy

Sign	Primary	Secondary	Consecutive
Previous swelling of the optic disc	No	Preceded by long-standing swelling of the optic disc.	No
Disc colour	White: diffuse or sectoral pallor.	Grey	Waxy pale
Disc margins	Distinct	Blurred	Normal, attenuated arteries
Fibrosis (gliosis) of the disc	None	Gliosis of the optic nerve head	None
Cause	<ul style="list-style-type: none"> <li>• compression of the optic nerve or chiasm</li> <li>• hereditary optic neuropathy</li> <li>• nutritional optic atrophy</li> </ul>	<ul style="list-style-type: none"> <li>• chronic papilloedema</li> <li>• papillitis</li> <li>• anterior ischaemic optic neuropathy</li> </ul>	<ul style="list-style-type: none"> <li>• chorioretinal disease e.g. retinitis pigmentosa</li> <li>• central retinal artery occlusion</li> </ul>

### Differential diagnosis of optic atrophy

Careful evaluation of the optic disc pallor together with other clinical features and a careful history can give clues to the underlying cause. The cause of optic atrophy may be a threat to the patient's life or vision and identification of the cause may save the patient's life or vision.

### How to examine the optic disc

The optic disc can be examined using a direct ophthalmoscope or at the slit-lamp using a 70D or 90D lens. Special attention should be paid to the colour of the disc, and whether the whole disc is paler than usual, or only a segment. The edge of the disc should be examined to see whether it is distinct or indistinct, and lastly the retinal blood vessels should be examined as they course over the optic nerve head to see if they are distinct and of normal and regular thickness. Lastly, it is important to assess whether both optic discs are equally affected, or whether one disc is normal, less affected or is swollen.

### Other ocular assessment in patients with optic atrophy

As well as examining the optic disc the visual acuity should be measured and the visual fields assessed by perimetry. The pupils should be examined for response to light and colour vision should also be assessed.

Having established that optic atrophy is present, try and decide whether it is primary or secondary, as this will guide you on what questions to ask the patient as the causes can be quite different.

### History

For all patients with optic atrophy it is important to ask how long ago they noticed the loss of vision and whether it came on suddenly or gradually. More specific questions should then be asked for primary and for secondary optic atrophy (see across).

### Primary optic atrophy

Questions to ask the patient:

- Are they taking any medication for tuberculosis?
- Have they had a diagnosis of a sexually transmitted disease e.g. syphilis or HIV?
- Do they frequently take quinine for malaria?
- Do they smoke and / or drink alcohol, have they consumed methanol?
- Does their occupation entail working with chemicals?
- Do they eat cassava and if so how is it prepared?

### Secondary optic atrophy

Questions to ask the patient:

- Have they been suffering from headaches, and if so is the headache worse in the mornings?
- Is the headache accompanied by nausea or vomiting?

If the answer to these questions is yes, urgent referral is required to rule out raised intracranial pressure which may be due to a tumour.

- Have they noticed any pain behind the eye on eye movement?
- Have they had episodes of double vision, or weakness or tingling in their arms or legs?

If the answer is yes to these questions then the optic atrophy, which is often unilateral initially, may be due to multiple sclerosis and the patient should be referred to a neurologist.

- Have they had any pain in other areas of their face?

If yes, they should be referred to an ENT specialist to rule out sinus disease.

If the loss of vision was sudden, the following signs and symptoms are suggestive of giant cell arteritis:

- Scalp tenderness
- Pain in the jaws when chewing
- Muscle and joint pains
- Weight loss

The clinical diagnosis is confirmed by an urgent temporal artery biopsy and high dose systemic steroids are required. Once the diagnosis has been made these patients are best managed by a physician.

### 1 Inflammatory optic neuropathy

In inflammatory disease of the optic nerve the initial appearance may be one of a swollen disc due to papillitis which over the course of a few months becomes atrophic. The visual loss may be sudden or gradual, unilateral or bilateral and may be associated with pain on eye movements.

Inflammatory optic neuropathy due to multiple sclerosis may present as a history of unilateral blurring of vision associated with pain on eye movements in a person aged 20-40 years, associated with other neurologic features such as diplopia, paraesthesia, loss of muscle power and loss of sphincter control. Neuromyelitis optica (Devic's disease) is severe and rare and presents acutely as bilateral optic neuritis with paralysis due to a transverse myelitis.

Other causes of inflammatory optic neuropathy include tuberculosis or syphilis; and it can also occur in the presence of systemic lupus erythematosus, or sarcoidosis; occasionally it is associated with orbital or sinus infection.

### 2 Ischaemic (vascular) optic neuropathy

Optic neuropathy due to a compromised blood supply of the optic nerve usually presents as a sudden loss of vision. The optic disc pallor may be diffuse or segmental (sectoral). Segmental pallor occurs if part of the blood supply to the optic nerve is occluded, and it will be associated with an appropriate altitudinal field defect.

An important treatable cause of ischaemic optic neuropathy is giant cell arteritis (GCA). It may present with sudden acute visual loss associated with scalp tenderness, jaw pain on chewing, muscle aches and weight loss, usually in people aged over 60 years. Diagnosis is suggested by tenderness over an easily palpable temporal artery and confirmed histologically by a temporal artery biopsy. Often

the ESR is highly raised. If GCA is the suspected diagnosis, treatment with high dose systemic steroids should be started urgently in order to reduce the risk of vision loss.

Anterior ischaemic optic neuropathy which is not due to arteritis may also occur in people (often elderly) with vascular risk factors such as diabetes mellitus, hypertension, hypercholesterolaemia and/or smoking. It presents as one or more episodes of acute visual loss due to vessel occlusion at the optic nerve head. Treatment is that of the cause, however one should be careful about lowering the blood pressure when the optic nerve already has evidence of poor perfusion.

Sometimes acute visual loss may follow surgery (cardiac, neck or spine surgery), or massive blood loss due to ischaemia of the blood supply posterior to the optic nerve head.

### 3 Compressive optic neuropathy

Compressive lesions commonly present with unilateral or bilateral gradual progressive vision loss. Lesions around the orbital apex, superior orbital fissure or cavernous sinus may also present with limitation of extraocular motility from involvement of the cranial nerves 3rd, 4th and 6th.

If the optic nerve is affected there may be a central or caecocentral field defect with an afferent pupil defect while optic chiasmal lesions which affect the decussating nasal fibres cause a bitemporal hemianopia (loss of vision in the temporal visual fields of the two eyes).

Compression of the optic nerve may be due to a meningioma, orbital tumour, thyroid eye disease or intracranial tumours such as pituitary adenoma or carotid aneurysm.

#### Raised Intracranial pressure – papilloedema and secondary atrophy

Papilloedema refers to optic disc swelling as a result of raised intracranial pressure and it is usually bilateral. There may be diminution in vision and other features of raised intracranial pressure such

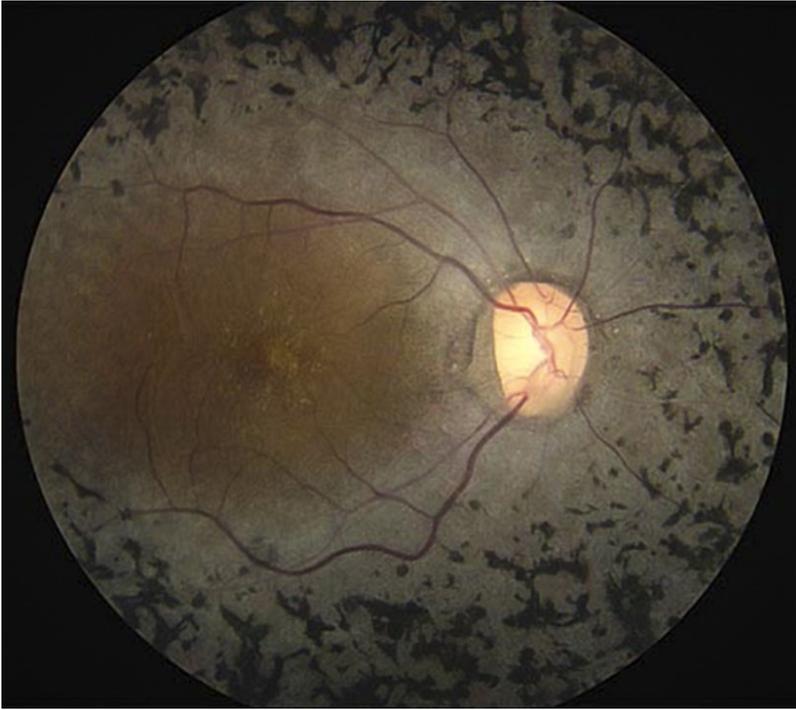
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**Figure 1.** Primary optic atrophy



**Figure 2.** Secondary optic atrophy



**Figure 3.** Consecutive optic atrophy (with retinitis pigmentosa)

HTTP://CLINICALGATE.COM/VIDEO-LIBRARY-OF-NEURO-OPHTHALMOLOGY/

as headache, seizures, nausea and vomiting. Initially there is a swollen disc (papilloedema) followed later by secondary optic atrophy.

Intracranial pressure rise may result from many causes including intracranial space occupying lesions, idiopathic intracranial hypertension and meningitis. Long-standing papilloedema results in optic atrophy.

#### 4 Toxic/nutritional optic neuropathy

Toxic/nutritional optic neuropathies present with bilateral, painless, symmetric vision loss which is gradual and progressive. The neuropathy typically affects the papillomacular bundle causing temporal pallor of the optic disc (the papillomacular bundle inserts into the temporal optic disc). The visual fields show central or centrocaecal field defects.

It is important to take a history to rule out medications that are toxic to the anterior visual pathway such as ethambutol, isoniazid or sildenafil. The patient's occupation may also expose him or her to toxic substances such as lead in paints, antifreeze agents or cyanide from improper processing of cassava. A monotonous diet of cassava products with little protein has been associated with optic atrophy in the "tropical amblyopia syndrome". Deficiency of vitamin B12, B1 (thiamine) and folate can result in optic atrophy and may be seen in persons who are heavy cigarette smokers, consume alcohol and have a poor diet. Accidental ingestion of methanol (methyl alcohol) may be fatal and may cause rapid and severe visual loss.

#### Epidemics of Optic Atrophy

There have been some reports of optic neuropathy affecting large numbers of people over a few months most notably in Cuba, Tanzania and Sierra Leone. Those affected are often adolescents who develop

optic atrophy, sometimes with other neurological signs. The optic atrophy can be mild and limited to the temporal aspect of the disc. The cause is not fully understood, but is thought to be due to nutritional deficiencies and/or toxic effects. Those affected should be treated with multi-vitamins, including the B group, and given advice about a healthy diet.

#### 5 Ocular trauma to the optic nerve

Trauma to the optic nerve may result from indirect injury (blunt trauma) to the head or from direct injury from bony fragments or bullets, or from compression by haematoma within the orbit or optic nerve sheath. A previous history of head trauma with visual loss may be obtained.

#### 6 Hereditary causes and optic atrophy in children

Children with unexplained visual loss and/or unilateral or bilateral optic atrophy should always be referred for further investigation, as an underlying cause should always be sought. Optic atrophy in children may be due to genetic factors (e.g. Leber's hereditary optic atrophy), but it might also signify that something is pressing on the nerve, or there is raised intracranial pressure from a brain tumour which may be benign or malignant.

#### Investigations

Appropriate investigations may include: blood pressure; blood glucose; erythrocyte sedimentation rate; blood test for vitamin B12, and red blood cell folate for those suspected of having nutritional optic atrophy. Chest X-ray is indicated if there are associated respiratory symptoms.

Neuroimaging of the orbit and the brain with attention to the course of the optic nerve, optic chiasm and optic tract may reveal the cause of the optic atrophy in compressive lesions.

#### Management

There is no specific treatment for optic atrophy itself. The underlying cause whether inflammatory, ischaemic, compressive or metabolic should be treated if known. If there is a causative medication or toxin it should be avoided while vitamin deficiencies should be replaced. Patients with low vision may benefit from low vision devices.

#### Summary

Optic atrophy is not a disease but a clinical sign. It refers to pallor of the optic disc which results from irreversible damage to fibers of the anterior visual pathway. The causes of optic atrophy are numerous, some of which may be life or sight threatening. A detailed clinical evaluation is helpful in the differential diagnosis and management of optic atrophy.

#### Further reading

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Kline LB, Bhatti MT, Chung SM, Eggenberger E, Foroosan R, Golnik KC et al. Basic and Clinical Science Course. Neuro-Ophthalmology. San Francisco: American Academy of Ophthalmology; 2011.p.116-169.

# Visual effects and rehabilitation after stroke



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[www.liverpool.ac.uk/psychology-health-and-society/research/vision/about](http://www.liverpool.ac.uk/psychology-health-and-society/research/vision/about)

Strokes, or cerebrovascular accidents (CVA) are common, particularly in older people. The problems of motor function and speech are well known. This article explains the common visual problems which can occur with a stroke and gives information about diagnosis and management.



A patient receiving information on rehabilitation. INDIA

## What is a stroke?

A stroke occurs when there is an interruption to blood flow to the brain either because of a blood clot blocking the blood vessel or a haemorrhage in the brain<sup>1</sup>. Strokes can cause signs which are obvious, such as loss of speech, drooping of one side of their face, or weakness or paralysis of the arm and/or leg on one side of the body<sup>1</sup>. The vision is affected in about two thirds of people who have had a stroke, but this is often not obvious to the patient or their carers. For example, someone who has weakness down one side may bump into things or not eat all the food on their plate, not realising that this may also be because they have visual field loss<sup>2</sup>.

## What causes a stroke?

A stroke or cerebrovascular accident, (CVA) is the result of a blocked blood vessel in the brain (thrombosis or embolus), or haemorrhage into the brain<sup>1</sup>. Strokes are more likely in the elderly, and those who have high blood pressure, diabetes or cardiovascular disease.

## Types of visual loss in people who have had a stroke

There are four ways in which vision can be affected following a stroke:

- 1 Loss of central vision
- 2 Visual field loss
- 3 Visual perceptual abnormalities
- 4 Eye movement abnormalities

These may occur in isolation but more frequently occur in combination<sup>3</sup>.

Problems with **central vision** are quite common after a stroke. The symptoms include blurred or altered vision. In many the vision improves, but the visual loss can be permanent.

**Visual field loss** occurs in up to half of people with a stroke, with the commonest defect being homonymous hemianopia in which vision is lost in the right or the left visual fields<sup>4</sup> (**Figure 1**). Patients may not be aware of this, and bump into doorframes or trip over things on the affected side. Reading can also be difficult (**Figure 2** on page 74).

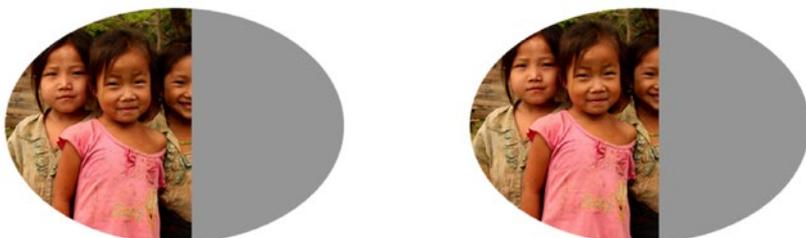
**Visual perceptual deficits** are many and varied affecting about a third of people with a stroke. Problems that may develop include neglect one side of their body; difficulty recognising faces or objects, or difficulties with colour vision, depth perception and motion<sup>5</sup>.

**Eye movement abnormalities** can also be varied, including strabismus (misaligned eyes), difficulty in converging the eyes to look at near objects, or double vision due to the cranial nerves which control eye movement being affected<sup>6</sup>. Typical symptoms include double vision, or jumbled, blurred and/or juddery vision (**Figure 2** on page 74).

## Impact

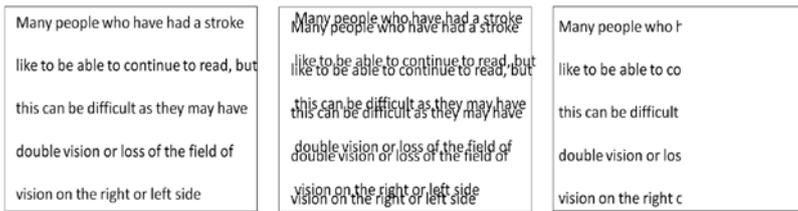
Blurred vision, double vision and loss of visual field are significant symptoms that impair daily functioning<sup>7</sup>.

**Figure 1:** Right homonymous hemianopia: the right-hand field of view is lost in both eyes



*Continues overleaf* ➤

**Figure 2:** Impact of vertical double vision (central image) and right hemianopia (right image) on reading



The patient or their close relatives may report that they frequently bump into objects such as door frames; have difficulty finding things on surfaces; are unsure of their footing while walking and stumble; may leave food uneaten on one side of the plate and have difficulty with reading. Other impacts on the quality of life include loss of confidence, fear of falling, fear of going out alone, social isolation and loss of independence<sup>8</sup>.

### How to assess visual function in someone who has had a stroke

Examination for visual loss<sup>9</sup> is essential for stroke survivors. There are various assessment tools which can be used to examine visual function after a stroke:

- UK National Clinical Guidelines for Stroke: [www.rcplondon.ac.uk/guidelines-policy/stroke-guidelines](http://www.rcplondon.ac.uk/guidelines-policy/stroke-guidelines)
- UK stroke/vision resources and factsheets: [www.parallelvisionproblems.org.uk/index.php/eye-conditions/](http://www.parallelvisionproblems.org.uk/index.php/eye-conditions/)
- UK Stroke Association stroke/vision factsheet: [www.stroke.org.uk/what-stroke/common-problems-after-stroke/vision-problems](http://www.stroke.org.uk/what-stroke/common-problems-after-stroke/vision-problems)
- UK Royal National Institute for the Blind stroke/vision factsheet: [www.rnib.org.uk/eye-health-eye-conditions-z-eye-conditions/stroke-related-eye-conditions/](http://www.rnib.org.uk/eye-health-eye-conditions-z-eye-conditions/stroke-related-eye-conditions/)

### Management

Treatment options aim to restore visual function to as normal as possible<sup>10</sup>. For eye movement abnormalities, prisms and patching one eye can be effective in reducing double vision<sup>6</sup>. For visual field loss a Cochrane systematic review<sup>11</sup> reports favourable evidence of visual scanning training which aims to compensate for the visual field loss. It is available as a paper training option ([www.strokevision.org.uk](http://www.strokevision.org.uk)) or through computer training ([www.eyesearch.ucl.ac.uk](http://www.eyesearch.ucl.ac.uk); [www.readright.ucl.ac.uk](http://www.readright.ucl.ac.uk)).

Stroke survivors with persistent impairment of central vision may be helped by low vision services which can include magnifiers, reading aids, computerised adaptations and improved lighting<sup>12</sup>. Furthermore, simple adaptations can be made by stroke survivors such as using large print, ensuring good lighting at home, putting labels or coloured stickers on cooking equipment, decluttering areas and having a companion when going out, particularly in busy, crowded places<sup>10</sup>.

### Conclusion

Post-stroke difficulties in visual function are an under-recognised problem that cause significant impact to the quality of life of stroke survivors. Carers and health workers need to be aware that problems with vision are a common consequence of stroke that is not outwardly obvious.

Assessment including visual functioning is best provided as part of a multi-disciplinary team on acute stroke units, or in neuro-rehabilitation units. A careful history about visual problems from the patient and carers followed by examination of visual acuity, eye movements and visual field are important in understanding the difficulties in visual functioning.

Management should be tailored to each individual, their visual difficulties and visual needs. With about one quarter of stroke survivors being of working age, rehabilitation in the context of adaptation of the work place environment is vital if younger people are to return to work after stroke. Rehabilitation requires patience and perseverance on the side of the client, relatives and the health provider.

Despite improvement in stroke prevention and acute stroke management, the increasing ageing population will result in more stroke survivors requiring rehabilitation. Policy makers need to understand the importance of providing post-stroke rehabilitation services including visual functioning.

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# New ways of working to support sustainable disease elimination



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 .....

How can we ensure that Neglected Tropical Diseases (NTDs) are not just eliminated, but eliminated once and for all?

This article explores the key role that water, sanitation and hygiene (WASH) interventions can play and what partnerships, programs and policies can be adopted to help see the end of certain diseases for good.

When talking about the end goal for a number of NTDs, we use the term *elimination as a public health problem* instead of control. This requires disease prevalence to be reduced to below specific threshold levels so that transmission levels are sufficiently low for fixed health facilities to treat cases so that specific community outreach programs are not required.

This carries a risk of resurgence to public health problem levels if the conditions for transmission have not changed. For diseases in which access to water, and poor sanitation and hygiene (WASH) plays a fundamental role, undertaking efforts to improve these conditions will reduce the risk of resurgence and, ultimately, enhance the sustainability of elimination efforts.

The transmission of schistosomiasis, lymphatic filariasis and trachoma is closely linked to poor WASH conditions, yet programs often focus on medical interventions, particularly mass drug administration. In order to prevent the spread of these diseases, a greater focus on WASH services is needed to reach elimination goals faster, reduce competition for resources and increase the value of programs in the eyes of the public and politicians by offering other health and non health related benefits.

This approach is being championed at the highest level. In August 2015, the World Health Organization (WHO) unveiled a global strategy and action plan to better integrate WASH services with public health interventions to accelerate progress in eliminating and eradicating NTDs by 2020. The emphasis is further stated in the standard operating procedures developed by WHO for validation or elimination of trachoma as a public health problem. The procedures require that gains against disease are sustained in the absence of antibiotic pressure and that evidence that environmental and behavioural conditions for transmission have been addressed. This provides added incentive for trachoma partners to work with WASH, education and other stakeholders.

While efforts have been made over the years by those working on NTDs to engage with agencies that deliver WASH services, NTD and WASH programs have tended to work separately. This has led to concerns over both the sustainability of achievements made through mass drug administration and over the lack of targeting of WASH services to endemic communities, which are almost always the communities most in need of those services.

This challenge has been picked up within trachoma elimination efforts, predominantly through the two



BILLY WEEKS FOR THE TASK FORCE FOR GLOBAL HEALTH

**Good hygiene and access to water is essential for ensuring clean faces and reducing transmission, an essential strand of the strategy to eliminate trachoma. MALAWI**

large-scale programs funded by the UK Department for International Development (DFID) and The Queen Elizabeth Diamond Jubilee Trust (The Trust). Planning workshops in seven countries aimed specifically at trachoma brought entirely new groups together, including different sectors of government (WASH, Education, Health) as well as NGOs and academics. This allowed for real-time development and testing of innovative planning tools. To enhance this collaboration and ensure lessons were captured and shared, in 2015 ICTC released *All you need for F&E – a toolkit for planning and partnering*, a resource aiming to strengthen coordination and maximise impacts in the field by supporting program managers working on trachoma to engage stakeholders from other/dependent sectors.

Another initiative will enhance joint WASH and NTD monitoring processes. An extensive consultation with NTD and WASH experts has resulted in an agreed set of core indicators to be applied at program level. By sharing goals and indicators, the common ground between partners is made explicit, making collaboration easier.

At the country level, Zambia and Kenya are providing us with some concrete examples of how data sharing can support joined-up working. In Zambia, a DFID supported trachoma elimination program has built upon an already successful monitoring platform to track the WASH-related elements of trachoma interventions in rural communities and monitor facial cleanliness and environmental improvement. In Kenya, a Trust supported trachoma elimination program in 12 counties has sourced demographic data from community health unit chalkboards, area and sub-county records (verified by education and health departments) to inform tailored planning for WASH components of SAFE, the trachoma intervention strategy, that would not have been possible using analysis of national-level data.

Ensuring the sustainability of elimination efforts is possible. There are practical examples to support the sustainable elimination of some of the world's most disabling and neglected diseases. We look forward to seeing these efforts scaled up and adopted by more partners working on trachoma and other NTDs.

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# Test your knowledge and understanding

This page is designed to help you to test your own understanding of the concepts covered in this issue, and to reflect on what you have learnt.



MURRAY MCGAVIN

Is this a sudden onset of a 6th nerve palsy or a longstanding esotropia? This boy may have a longstanding right esotropia (if he is looking straight) or a left 6th nerve palsy (if he is attempting to look left).

We hope that you will also discuss the questions with your colleagues and other members of the eye care team, perhaps in a journal club. To complete the activities online – and get instant feedback – please visit [www.cehjournal.org](http://www.cehjournal.org)

## Tick ALL that are TRUE

### Question 1 Anisocoria:

- a. May be due to an oculomotor (3rd nerve) palsy
- b. May be due to bilateral optic atrophy
- c. May occur in association with partial ptosis
- d. May be due to mydriatics
- e. May be associated with heterochromia of the iris

### Question 2 Homonymous hemianopia:

- a. Means loss of field of vision in one eye with normal visual field in the other eye
- b. May be due to a stroke (CVA) affecting the occipital cortex
- c. Can cause difficulty in reading, eating and driving
- d. May be due to an orbital lesion causing proptosis
- e. May occur during a migraine attack

### Question 3 Diplopia:

- a. May follow a head injury with reduced abduction on eye movements
- b. Can be due to myasthenia gravis
- c. If it persists when one eye is closed, then it is due to a lesion in the brain
- d. May be associated with proptosis
- e. The separation of images is greatest in the direction of action of the paralysed muscle

## ANSWERS

1. a, c, d and e. Anisocoria is a difference in size between the two pupils. Third nerve palsy may paralyse the constrictor muscle of the iris and cause a dilated pupil on the affected side and may cause a constricted pupil on the affected side together with partial ptosis and sometimes depression of the iris (heterochromia). In optic atrophy (unilateral or bilateral) the pupils are the same size, although they will have a poor reflex reaction to light.

2. b, c and e. Homonymous hemianopia means half the field of vision is lost in both eyes – the nasal field in one eye and the temporal field in the other. The result is loss of half the bilateral field of vision. It may occur with any lesion affecting the optic tract, radiation or cortex. As half the bilateral visual field is lost, only half the page, half the plate and half the driving field of vision is seen. Occasionally it can occur with a migraine attack, usually recovering but not always.

3. a, b, d and e are correct. A head injury may cause a palsy of the 6th cranial nerve which supplies the lateral rectus. Myasthenia gravis causes weakness of muscles on use and can lead to diplopia and ptosis. Monocular diplopia is due to ocular conditions e.g. media opacities. Proptosis, particularly associated with dysthyroid eye disease can cause diplopia. The diplopia is greatest in the direction of action of the paralysed muscle.

## Reflective learning

Please visit [www.cehjournal.org](http://www.cehjournal.org) to complete the online 'Time to reflect' section.

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## Picture quiz



This 30-year-old woman presents with headache, nausea and confusion developing over the last 4 weeks. The appearance in both fundi is similar. The visual acuity is 6/6 and 6/9. Pupils respond equally to light. Extraocular movements are full.

### Tick ALL that are TRUE

#### Question 1

The following signs are visible:

- a. Hard exudates
- b. Intra-retinal haemorrhage
- c. Swelling of the disc
- d. Cotton wool spots
- e. Optic pallor / atrophy

#### Question 2

The following are likely diagnoses:

- a. Papillitis due to multiple sclerosis
- b. Hypertensive retinopathy
- c. Diabetic retinopathy
- d. Raised intracranial pressure
- e. Migraine

#### Question 3

The following is indicated:

- a. Lumbar puncture
- b. X-ray of the orbit
- c. Referral for brain imaging (MRI / CT scan)
- d. Careful history taking of medications
- e. Tensilon test.

### ANSWERS

1. Answer b and c. There is a swollen disc (disc edge not clearly visible) with a flame shaped haemorrhage above the disc superiorly.  
 2. Answer d. In papillitis the visual acuity is decreased. Hypertensive and diabetic retinopathy often have hard and soft exudates. Migraine would not last for so long. The symptoms of headache, nausea and confusion are typical of raised intracranial pressure (ICP) which is confirmed by the presence of swollen discs (papilloedema).  
 3. Answer c and d. Lumbar puncture should not be performed in the presence of papilloedema as it may cause 'coning' and death. There is no evidence of orbital disease (no proptosis and normal ocular movements). An MRI is indicated to look for an intra-cranial space occupying lesion. Some medications cause raised ICP so a careful history of medicine use is important. The Tensilon test is used to investigate myasthenia gravis which is not indicated here.

## IAPB Vision Atlas



The IAPB Vision Atlas website is a compilation of the latest data and evidence relevant to all those who

believe that in the 21st Century no one should have to live with avoidable blindness or sight loss – from eye conditions many of which can be easily treated or prevented and for which cost-effective solutions are readily available.

The IAPB Vision Atlas is designed around two main sets of data: the estimates of the burden of blindness and visual impairment made by the Vision Loss Expert Group (VLEG) and national level performance against the key indicators laid out in the World Health Assembly resolution 66.4 'Universal Eye Health: a Global Action Plan 2014 – 2019' (the GAP).

Do visit it here:

<http://atlas.iapb.org>

## Courses

### MSc Public Health for Eye Care, London School of Hygiene & Tropical Medicine

10 fully funded scholarships available for Commonwealth Country Nationals. Course aims to provide eye health professionals with the public health knowledge and skills required to reduce blindness and visual disability in their setting. For more information visit:

[www.lshtm.ac.uk/study/masters/mscpec.html](http://www.lshtm.ac.uk/study/masters/mscpec.html) or email [Romulo.Fabunan@Lshtm.ac.uk](mailto:Romulo.Fabunan@Lshtm.ac.uk)

**Eye Banking Course:** New international qualification course for eye bankers. Suitable to all service and experience levels. The *Specialist Certificate* course starts in September 2017 (with option to work towards *Graduate Certificate* afterwards). Expressions of Interest via: <http://commercial.unimelb.edu.au/custom-education/courses/eyebanking> or please contact Heather Machin, Subject Coordinator, via: [heather.machin@unimelb.edu.au](mailto:heather.machin@unimelb.edu.au)

### University of Cape Town Community Eye Health Institute

[www.health.uct.ac.za](http://www.health.uct.ac.za)

or email

[chevron.vanderross@uct.ac.za](mailto:chevron.vanderross@uct.ac.za)

### Kilimanjaro Centre for Community Ophthalmology International

[www.kcco.net](http://www.kcco.net) or contact Genes Mng'anga at [genes@kcco.net](mailto:genes@kcco.net)

### Lions Medical Training Centre

Write to the Training Coordinator, Lions Medical Training Centre, Lions SightFirst Eye Hospital, PO Box 66576-00800, Nairobi, Kenya.

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## Next issue



CLAIRE WALKER

The next issue of the *Community Eye Health Journal* will be on the theme: : **Continuing Professional Development**

# Key community eye health messages

## Double vision is an important symptom which may be due to serious even life-threatening diseases



DANIEL TYAALE (ORIGINAL PHOTOGRAPH)

- It can be caused by diseases affecting the 3rd, 4th or 6th cranial nerves or by diseases of the extraocular muscles e.g. myasthenia or thyroid eye disease.
- All patients with double vision need a full examination of their eyes including extra-ocular movements in order to make a diagnosis.

## Sudden loss of vision in one (or both) eyes with an afferent pupil defect and normal globes may be due to vascular occlusion of retinal vessels or inflammation of the optic nerves



NICK ASTBURY

- In elderly people check for temporal (giant cell) arteritis and in younger patients check for symptoms of multiple sclerosis.

## A swollen optic disc may be unilateral or bilateral



ICEH

- If associated with severe headache and relatively normal vision this probably indicates papilloedema due to raised intracranial pressure which requires urgent investigation and diagnosis.
- If the vision is significantly reduced and there is no headache it is probably due to inflammation of the optic nerve (optic neuritis / papillitis) which is likely to require treatment.

## Sudden loss of vision is devastating to the patient and close family members



ICEH

- A person trained in counselling patients with vision loss should be available in eye departments to spend time listening to and advising people who have just lost their vision.