Sudden loss of vision
a GP's guide to the many causes

Sudden loss of vision at any age is an alarming symptom that usually results in a rapid presentation for medical opinion. General practitioners need to have some understanding of the possible causes, an examination technique and a knowledge of conditions that require urgent referral for specialised care.

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History

In each case of sudden visual loss, the doctor of first contact needs to make an assessment that will determine whether any treatment should be administered immediately or if urgent referral to an ophthalmologist or neurologist is required. A sudden loss of vision can be:

- unilateral or bilateral
- partial or complete
- transient or permanent
- painful or painless
- of sudden onset or progressive.

Unilateral visual loss. Sudden visual loss in one eye is usually apparent at the time of onset. However, occasionally a patient may cover the normal eye and suddenly realise that the other eye is blind; in this situation there has probably been slowly progressive loss in the affected eye over months or years. Details of previous recordings of visual acuity are helpful (for example, driving licence tests and school medical or optometrical examinations).

Another cause of visual loss which may not have been noticed by the patient is amblyopia. Ocular defects in early childhood result in suppression of the image from the affected eye by the visual cortex, and lifelong poor vision in that eye. The condition is not progressive after 6 to 8 years of age, and there may be a history of a 'lazy eye' or squint in infancy.

IN SUMMARY

- **For a patient with a sudden visual loss, the GP needs to determine whether any treatment should be administered immediately or if urgent referral to an ophthalmologist or neurologist is required.**
- **An urgent ophthalmological examination is essential for any patient who presents with a sudden visual loss unless the cause is clearly neurological.**
- **Acute angle closure glaucoma produces rapid loss of vision in one eye. It is usually associated with severe pain (clearly localised to the affected eye), nausea, vomiting and haloes around lights. Urgent consultation with an ophthalmologist is required.**
- **Sudden painless loss of vision in one eye is the hallmark of central retinal artery occlusion. The occlusion occurs behind the optic disc, and causes such as emboli are not visible. If seen within minutes, the emergency treatment consists of massaging the globe, intravenous acetazolamide (Diamox) and paracentesis of the anterior chamber.**
- **Early diagnosis of giant cell arteritis is important because it is highly likely that the other eye will be affected within weeks if corticosteroids are not commenced. The prognosis for recovery is usually very poor.**

If sudden monocular visual loss is partial, the pattern of the scotoma can be of localising value. If the patient is aware of a shadow (that is, a positive scotoma), a retinal or vitreous cause is likely. On the other hand, scotomata are negative in optic nerve or visual pathway lesions and are not clearly apparent to the patient.

Bilateral visual loss. Sudden transient bilateral visual loss is usually immediately apparent to the patient, but sometimes there is difficulty distinguishing between transient loss in one eye and an homonymous hemianopia on the same side. The distinction is important because transient ischaemia in one eye is in the carotid territory, whereas loss in the half field of both eyes is in the vertebrobasilar territory.

Patients with bilateral occipital infarction may show denial of blindness (Anton's syndrome), but this is readily detected when visual acuity is tested in each eye. In addition, the fundi and pupillary reactions will be normal.

Examination

Unless the cause of sudden visual loss is clearly neurological, all patients should have an urgent ophthalmological examination.

Visual acuity. Some attempt should be made to record the best acuity in each eye – ideally with the Snellen chart or with typewritten text. The patient should use reading glasses if these are worn routinely. Small newsprint approximates N8. If the visual loss is severe and less than 6/60, record finger counting, hand movements, and light perception (or no perception).

Visual fields. Each quadrant is screened with finger movements with both eyes open. Visual fields are then checked separately by confrontation with a red pin, which will pick up a relative or complete homonymous hemianopia.

Fundus. Examination of the fundi with an ophthalmoscope is essential. The normality (or otherwise) of the optic discs, retinal arteries and veins and macula should be noted.

If no abnormality is seen in the fundus to account for unilateral visual loss, the lesion is probably behind the globe – that is, in the optic nerve. The pupil response to light is lost or reduced if visual loss is caused by lesions of the optic nerve, but the response is preserved in occipital lobe disease and amblyopia.

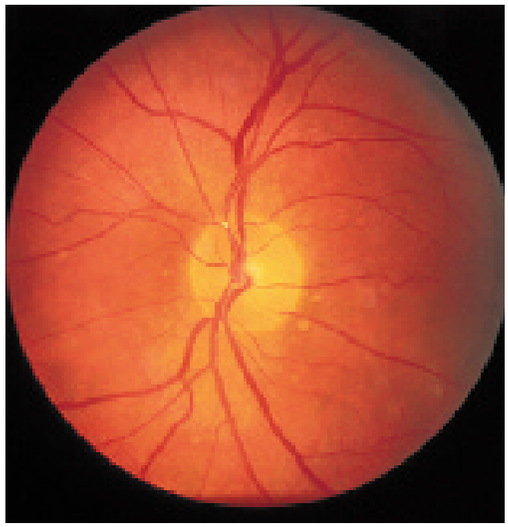

Figure 1. A highly refractile cholesterol embolus lodged in the bifurcation of the superior division of the central retinal artery.

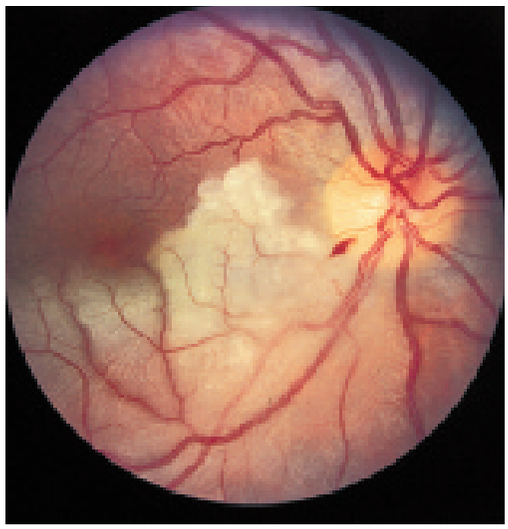

Figure 2. Calcific embolus from the aortic valve in a patient with rheumatic heart disease. An embolus in the central retinal artery is causing infarction of the inferior retina indicated by cloudy swelling and a hold up of axoplasmic transport at the junction of the normal and pale retina. There is a small haemorrhage in the nerve fibre layer of the optic disc.

Sudden transient unilateral visual loss

Causes of sudden transient unilateral visual loss are listed in Table 1.

Amaurosis fugax

Amaurosis fugax refers to painless uniocular visual loss of sudden onset lasting less than one hour. It is typically described by the patient as a 'shutter' coming down over the eye.

Amaurosis fugax is caused by the passage of an embolus from the carotid artery or the heart through the retinal arteries. Fundal examination at the time or soon after may reveal a highly refractile yellow cholesterol crystal within the retinal arterial branches near the optic disc

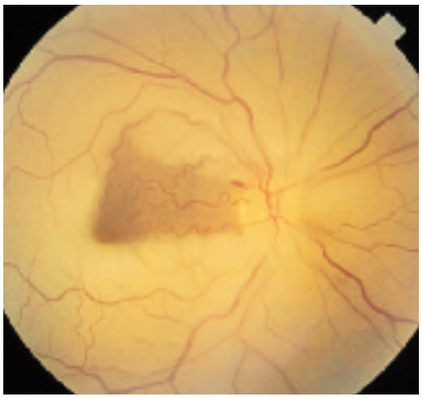

Figure 3. Central retinal artery occlusion with retinal infarction. An area of the macula supplied by a cilioretinal artery is not affected.

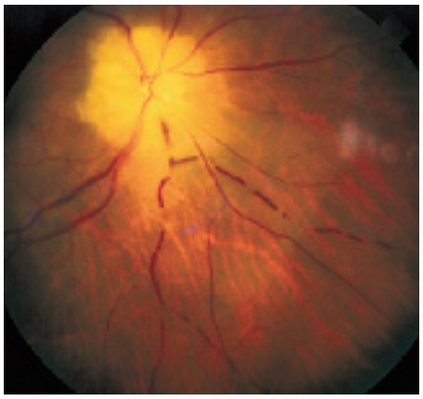

Figure 4. A case of ischaemic optic neuropathy showing pale swelling of the disc with 'cattle-trucking' of the inferior nasal branch vein caused by interrupted flow.

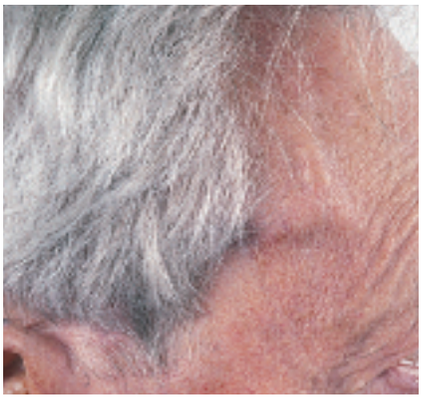

Figure 5. A patient with ischaemic optic neuropathy and a prominent, painful, occluded superficial temporal artery.

(Figure 1). The embolus will probably have arisen from the ipsilateral internal carotid artery, but it may have arisen (although it is much less likely) from the aortic arch or common carotid artery.

Calcific emboli from the aortic or mitral valves are larger, white, nonrefractile bodies that are usually seen lodged close to the disc and are often associated with permanent field defects (Figure 2). Fibrin platelet emboli are derived from ulcerated cholesterol plaques or the heart and, although they cause episodes of amaurosis, they are rarely seen in the fundus.

Ocular hypoperfusion

In patients with advanced extracranial vascular disease, transient unilateral visual loss may be provoked by activities that 'steal' blood from the retinal circulation, such as walking, facial heating, standing after bending, looking upwards or a postprandial state. The visual loss is often likened to a photographic negative (that is, colour is lost) or described as fragmented or constricted.

Venous stasis retinopathy (hypotensive retinopathy) may be found in patients suffering transient loss of vision caused by ocular hypoperfusion. The retinal veins are dilated, with branch occlusions, dot and blot haemorrhages in the midperiphery, and a central retinal artery pulsation that is readily abolished by minimal pressure on the globe.

Retinal migraine

Retinal migraine is a rare disorder of young adults. In classic migraine, the visual disturbance is bilateral and often associated with headache. However, in retinal migraine the visual loss is sudden, painless and uniocular, and typically lasts about five minutes. The mechanism is thought to be retinal arterial spasm.

Obscurations

Obscurations are episodes of unilateral or bilateral visual loss that last a few seconds in patients with critically raised intracranial pressure. Episodes are precipitated by standing, bending or straining. Papilloedema should be evident.

Uhthoff's phenomenon

Uhthoff's phenomenon is the name given to the transient loss of vision in one or both eyes that occurs at the time of a rise in body temperature, such as after a hot bath, exercise or hot drink. This distinctive symptom occurs in patients who have had demyelinating optic neuritis.

Glaucoma

Intermittent angle closure glaucoma may present with episodic unilateral loss of vision, and is likely to be triggered by pupillary dilatation that occurs in dim light. All patients with transient visual loss should have a complete ophthalmological examination that includes measurement of intraocular pressure and assessment of the anterior chamber angle.

Other causes

Giant cell arteritis may cause transient loss of vision resembling amaurosis fugax, and is presumably caused by inflammatory occlusion of the ophthalmic artery. Transient visual loss also occurs in hyperviscosity states such as:

- systemic lupus erythematosus (SLE)
- polycythaemia vera
- paraproteinaemia
- antiphospholipid antibody syndrome.

Sudden permanent unilateral visual loss

Causes of sudden permanent unilateral visual loss are listed in Table 2.

Vitreous haemorrhage

In patients with vitreous haemorrhage the fundus cannot be seen, and a dark

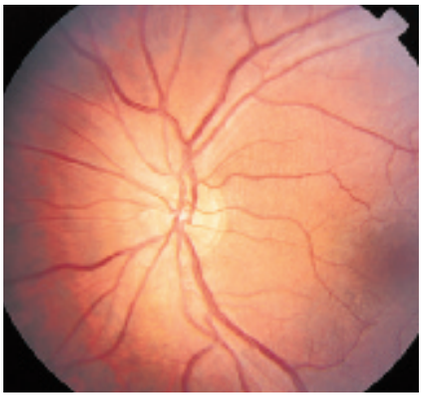

Figure 6a. Methanol poisoning. A left fundus is shown 48 hours after ingestion of methanol showing early nerve fibre swelling.

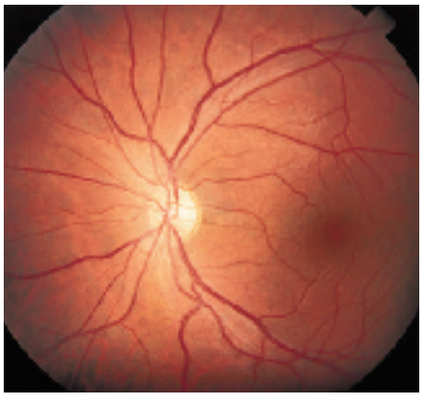

Figure 6b. The same patient. Ten weeks after methanol ingestion, the fundus shows severe optic atrophy. The changes were bilateral.

or grey reflex is present. It occurs with bleeding from new vessels (such as in diabetes), in subarachnoid haemorrhage and after trauma.

Retinal detachment

In retinal detachment, a patient is aware of a dark shadow in the peripheral field of vision. This is followed by loss of acuity as the macula becomes involved.

Central serous retinopathy

Central serous retinopathy typically occurs in young men who complain of impaired central vision with distortion of objects, metamorphopsia and micropsia. The fundus may or may not show a macular lesion, but the Amsler grid will reveal a central scotoma and the photostress test will be positive. (The photostress test involves shining a bright light in the affected eye for 10 seconds and measuring the time required for the acuity to return to baseline, compared with the other eye.)

Central retinal vein occlusion

Central retinal vein occlusion is a common condition that is diagnosed by the fundal appearances of dilated tortuous veins, flame-shaped haemorrhages and soft exudates. The visual loss occurs when the macula is involved by haemorrhage or oedema.

Acute glaucoma

Acute angle closure glaucoma produces rapid loss of vision in one eye, usually associated with severe pain clearly localised to the affected eye, and nausea, vomiting and haloes around lights. The globe feels hard and is very tender. Urgent consultation with an ophthalmologist is required.

Central retinal artery occlusion

Sudden painless loss of vision in one eye is the hallmark of central retinal artery occlusion. Examination reveals vision of light perception and an unreactive pupil; in the earliest stages, the fundus may be near-normal. The retinal arteries may appear attenuated and a 'cherry red spot' may be apparent at the macula.

In the presence of a cilioretinal artery, some macular vision will be preserved (Figure 3). Later, the retina shows pale cloudy swelling caused by infarction. The occlusion occurs behind the optic disc, and causes such as emboli are not visible. If seen within minutes, emergency treatment consists of massaging the globe, intravenous acetazolamide (Diamox) and paracentesis of the anterior chamber.

Table 1. Sudden transient unilateral visual loss

- Amaurosis fugax
- Ocular hypoperfusion
- Retinal migraine
- Obscurations
- Uhthoff's phenomenon
- Subacute glaucoma

Table 2. Sudden permanent unilateral visual loss

- Vitreous haemorrhage
- Retinal detachment
- Central serous retinopathy
- Central retinal vein occlusion
- Acute glaucoma
- Central retinal artery occlusion
- Optic neuritis
- Ischaemic optic neuropathy
- Optic nerve compression
- Carcinomatous or lymphomatous optic neuropathy

Optic neuritis

The onset of visual loss in optic neuritis usually occurs over several days and may be preceded by pain on eye movement. It is typically described as a lace curtain in front of the eye, but can vary from minimal loss of acuity to no light perception. Some degree of recovery should occur between two and six weeks after onset.

Although a central scotoma is regarded as the field defect of optic neuritis, almost any pattern of field loss can be seen. The optic disc may be near-normal or swollen with haemorrhages and exudates (papillitis) in the first few weeks, but goes on to become atrophic. At presentation, the ability to read the Ishihara plates accurately is lost and there is a relative afferent pupil defect (that is, the pupil of the affected eye does not respond as briskly to light as the normal pupil).

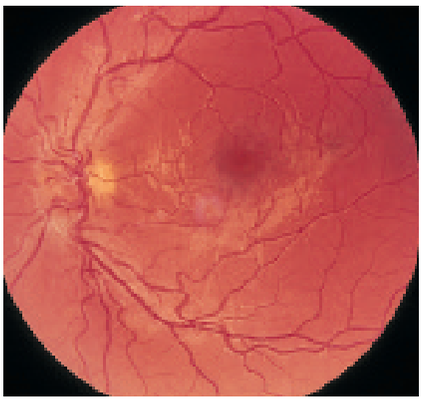

Figure 7. An acute case of Leber's hereditary optic atrophy. The nerve fibre layer is oedematous and the circumpapillary vessels show increased tortuosity.

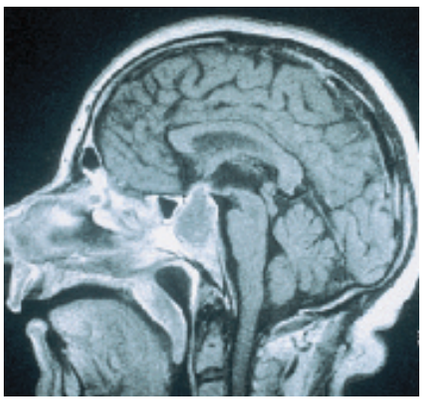

Figure 8. A sagittal MRI of the brain in a case of pituitary apoplexy with sudden visual loss caused by suprasellar extension compressing the optic chiasm from below.

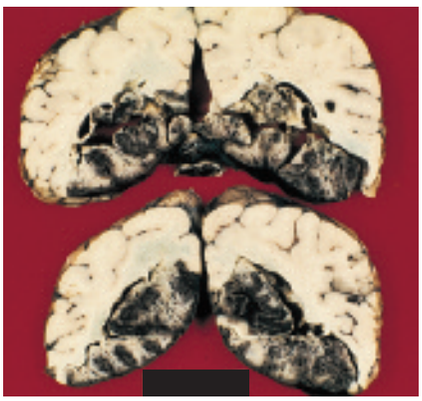

Figure 9. A postmortem specimen showing bilateral posterior cerebral artery territory infarction. The patient died after presenting with sudden cortical blindness.

Ischaemic optic neuropathy

Ischaemic optic neuropathy causes sudden painless loss of vision in elderly patients. The condition is associated with a pale swollen optic disc, with or without peripapillary haemorrhages (Figure 4). The superficial temporal arteries may be tender, prominent and nonpulsatile (Figure 5). The visual field defect is a combination of arcuate scotomata and altitudinal hemianopia.

The pathology is infarction of the head of the optic nerve: 15% of cases are caused by giant cell arteritis; the remainder are associated with diabetes, hypertension, atherosclerosis and severe hypotension. Early diagnosis of giant cell arteritis is important because it is likely that the other eye will be affected within weeks if corticosteroids are not commenced.

The prognosis for recovery after ischaemic optic neuropathy is usually very poor.

Optic nerve compression

Visual loss caused by compression of the optic nerve is not usually of sudden onset, except in cases of pituitary apoplexy and expanding anterior communicating artery aneurysm. The discovery of poor vision in one eye may be thought to be of sudden onset because slowly progressive unilateral blindness may go unnoticed for some time. The findings are:

- decreased acuity
- poor colour vision
- a central scotoma
- a pale optic disc
- a relative afferent pupillary defect.

Carcinomatous or lymphomatous optic neuropathy

Rapid loss of vision can occur in one or both eyes with infiltration of the optic nerve by neoplastic cells. The diagnosis should be suspected in patients with known carcinoma, lymphoma or leukaemia, and is confirmed by finding neoplastic cells in the cerebrospinal fluid.

Sudden bilateral visual loss

Causes of sudden bilateral visual loss are listed in Table 3.

Toxic optic neuropathy

Patients with toxic optic neuropathies usually present with visual loss that is painless, bilaterally symmetrical and progressive. The field defect is typically bilateral central or centrocaecal scotomata with loss of colour vision and late optic atrophy. The diagnosis is relatively easy if the patient is taking drugs known to cause toxic optic neuropathy (such as ethambutol) or abuses alcohol and tobacco.

Quinine poisoning can occur with a deliberate overdose or in susceptible individuals. The visual loss is sudden, bilateral and severe, but substantial recovery usually occurs. The visual fields are constricted and the arterioles are attenuated in the acute phase.

Poisoning by methanol (methyl alcohol) or wood alcohol is now relatively rare. It produces sudden bilateral visual loss with central and centrocaecal scotomata, optic atrophy and widely dilated unreactive pupils (Figures 6a and b). The prognosis for recovery of vision is poor.

Leber's hereditary optic neuropathy

Leber's hereditary optic neuropathy typically presents in young men, with rapid visual loss in one eye and then in the other eye within days to weeks. It is caused by one of several mutations in mitochondrial DNA.

Patients may have a family history of blindness in young males, but the retinal appearances are characteristic in the acute stages: a peculiar tortuosity of

arterioles around the optic disc and swelling of the fibre layer of the retinal nerve (Figure 7). After some weeks the optic discs become pale and the patient is left with dense central scotomata.

Despite advances in diagnosis, there is no known treatment for Leber's hereditary optic neuropathy.

Pituitary apoplexy

Sudden haemorrhage or infarction in a pituitary adenoma causes rapid expansion of the mass with onset of headache, chiasmal compression, ophthalmoplegia and disturbed consciousness. The diagnosis is difficult because the pupils are dilated and unreactive as a result of third nerve involvement, but ophthalmoplegia serves to exclude bilateral occipital lesions as the cause of the patient's visual loss.

CT scanning may not show the characterisistic appearance of haemorrhage, but the sella is enlarged. MRI will reveal the suprasellar mass (see Figure 8).

Other causes of chiasmal compression

Most suprasellar lesions cause slowly progressive visual loss. However, sudden enlargement of suprasellar cysts associated with craniopharyngioma, dermoid or expanding aneurysms of the internal carotid or anterior cerebral arteries may produce acute chiasmal compression.

Infarction of the occipital cortex

Vertebrobasilar thromboembolism to the posterior cerebral arteries results in bilateral calcarine infarction that presents as bilateral homonymous hemianopia with variable macular sparing (Figure 9). Initially, patients are cortically blind and often unaware of the visual loss until formally examined. The pupils remain fully reactive. There may be other manifestations of brainstem ischaemia, such as dysarthria, gaze palsies, limb weakness and ataxia.

Table 3. Sudden bilateral visual loss

Retina and optic nerve
- Toxic optic neuropathy (including quinine and methanol)
- Leber's hereditary optic neuropathy

Optic chiasm
- Pituitary apoplexy
- Craniopharyngioma

Occipital cortex
- Infarction of occipital cortex

Concluding remarks

Patients with sudden visual loss will frequently need rapid referral to the appropriate specialist, but they are likely to present initially to their general practitioner. A prompt assessment and consideration of the causes listed will ensure patients receive optimal treatment.

Finally, in some patients sudden visual loss may have a psychogenic basis. The most common field defect is concentric contraction; however, the diagnosis is often extremely difficult and requires repeated examination by an ophthalmologist or neurologist. MT