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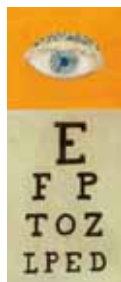
# Sudden loss of vision
## Investigation and management

### Background
Sudden vision loss usually requires urgent ophthalmic assessment. Diagnosis and management requires the judicious use of a wide range of serological and imaging investigations to guide appropriate treatment and referral.

### Objective
This article follows on from the previous discussion of the role of history and examination to discuss the appropriate investigation and management of common causes of sudden visual loss.

### Discussion
The key historical and examination findings have now been extracted and synthesised and these inform the next step. The general practitioner must now decide upon the most appropriate and timely investigation pathway or the need for, and urgency of, referral.

■ Investigation and management should be determined depending on the findings from the history and examination. Several of the more common and potentially sight or life threatening aetiologies will be discussed here, although this is not an exhaustive list. Many of the conditions discussed will cause temporary visual loss and several may become permanent, particularly if not promptly managed.

### Giant cell arteritis

Suspicion of giant cell arteritis (GCA) in the setting of sudden visual loss in a patient over 50 years of age should prompt immediate referral for ophthalmic review. Patients should have their erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and full blood count checked, followed by temporal artery biopsy where appropriate. In general terms, the upper limit of normal ESR can be considered to be half the age in males, and half the age plus 10 for females.[1] While ESR may be normal in 17% of patients with temporal arteritis, the sensitivity of ESR alone for the diagnosis of temporal arteritis is only 84.9%. The sensitivity of CRP alone is 97.5%, and the sensitivity of raised either ESR or CRP is 99.2%.[2] Platelet assay is also an important tool in the diagnosis of temporal arteritis, with temporal arteritis six times more probable in a patient with thrombcytosis than a patient with a normal platelet count.[2]

Temporal artery biopsy should be considered for all patients with a high clinical suspicion of GCA, as a positive histological diagnosis is definitive and is important in balancing the therapeutic benefits and adverse effects of prolonged oral corticosteroid therapy. Temporal artery biopsy results are not affected by corticosteroid administration for the first 4–7 days, and in suspected cases therapy should be commenced while urgent test results are pending. Therapy is aimed at limiting further vision loss, vision loss in the contralateral eye, and ischaemia of other organs such as bowel, myocardium and central nervous system.

### Transient embolic phenomenon

Where amaurosis fugax is suspected to be due to a transient embolic phenomenon, the patient requires carotid duplex and

an echocardiogram; those with critical stenosis warrant urgent surgical referral. Where these investigations prove unfruitful, but the suspicion is high (eg. an embolus can be seen in a retinal arteriole) the patient may require more sensitive investigation such as transoesophageal echocardiogram or computerised tomography (CT) angiography looking for more proximal carotid artery disease.

It is also imperative that the patient be screened for cardiovascular risk factors including hypertension, hypercholesterolaemia and diabetes mellitus. While not routinely performed, screening tests for thrombophilias and hyperviscosity syndromes should be considered in the setting of normal cardiovascular investigations and high clinical suspicion for embolic disease, and GCA should always be excluded.[3]

## Retinal artery occlusion

Retinal arterial occlusion results in sudden painless loss of vision, often to the extent of 'count fingers' visual acuity, with a fixed, nonresolving scotoma. It is not fleeting like amaurosis fugax. Emergency management with ocular massage and anterior chamber paracentesis within 90–120 minutes has anecdotally been reported to improve outcomes (theoretically by encouraging the passage of the embolus through the small arterioles and capillary bed into the venules), however these treatments are of no proven benefit. Retinal artery occlusion should be investigated in the same manner as embolic amaurosis fugax, but should also include a vasculitic screen.

Branch retinal artery occlusions create a more limited scotoma, while cilioretinal artery occlusions may only reduce the central vision. It is important to note that 20–30% of the population have a cilioretinal artery, and in these patients central retinal artery occlusion may not result in poor central vision. Retinal venous occlusion does not necessarily require carotid doppler or echocardiographic investigation, although investigation for hypertension, diabetes and hypercoagulable states is necessary.

In central retinal artery occlusions, it is critical to exclude GCA due to its propensity to involve the contralateral eye and other body systems. Fundus fluorescein angiography may be required to identify areas of vasculitis if this is a likely cause for retinal vascular pathology.

## Papilloedema

Papilloedema in any setting requires prompt imaging. While the term 'papilloedema' strictly refers to optic disc swelling secondary to raised intracranial pressure, optic disc swelling from any cause should be promptly referred for ophthalmic assessment. Nevertheless, the most common cause of papilloedema is idiopathic intracranial hypertension, which remains a diagnosis of exclusion. When idiopathic intracranial hypertension is suspected, a lumbar puncture will be required to assess opening pressure, and magnetic resonance imaging and venography (MRI and MRV) with gadolinium enhancement is required to investigate other causes of raised intracranial pressure such as cerebral venous sinus thrombosis.

In the obese patient, lumbar puncture may require CT guidance as the prevalence of this condition is approximately 20 times greater in obese patients.[4] While treatment for idiopathic intracranial hypertension ranges from weight loss to optic nerve sheath fenestration or neurosurgery, regular ophthalmic review is essential in monitoring the effect of the disease process on optic nerve and visual function.

Other causes of optic disc swelling should be considered, but should still be referred for specialist opinion. Optic disc drusen are chronic and nonprogressive, and can be demonstrated on ocular ultrasound or CT scan. The prevalence in caucasians is 0.5%.[4] Serial automated visual field testing may be helpful in assessing the stability of visual field loss. Disc swelling in the setting of systemic infection or neurological signs or symptoms should be treated as an emergency, and investigation for neurological infection should be commenced immediately.

## Optic neuritis

Optic neuritis occurs in 70% of patients with known multiple sclerosis,[4] and may be the first presentation of demyelinating disease. Patients with only classic signs and symptoms of optic neuritis may be diagnosed with typical optic neuritis. These include unilateral sudden vision loss in a patient aged 20–50 years, associated with retrobulbar pain, decreased colour perception and relative afferent papillary defect. Any atypical features should be investigated thoroughly to eliminate progressive optic neuropathies of other aetiologies using gadolinium enhanced MRI, chest X-ray, blood investigations and lumbar puncture.

Intravenous methylprednisolone (1 g/day for 3 days) followed by oral prednisolone for 11 days has been shown to decrease the time to recovery from a single episode, but not change long term outcome.[5] This treatment may therefore be of particular value to patients with significant pain, or pre-existing poor vision in the contralateral eye. Interferon-ß has been shown to delay the diagnosis of multiple sclerosis, and the accumulation of latent demyelinating plaques seen on MRI.[6]

Figure 1. Retinal detachment seen on B-scan ultrasound. A) The retina is tethered to the optic disc, creating the 'funnel' shape of a total retinal detachment; B) Duplex ultrasound showing flow in the retinal vessels within the funnel
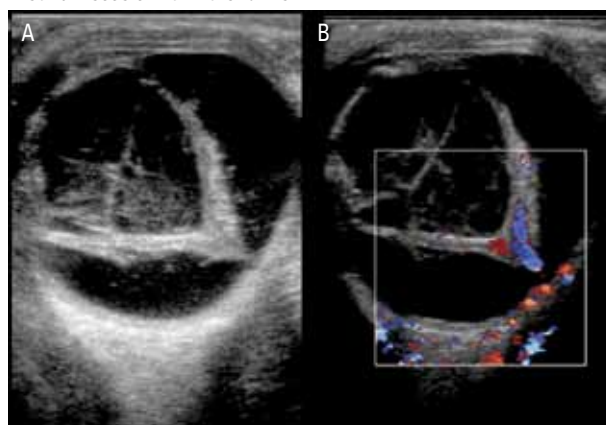
## Retinal detachment

Retinal detachment is a further cause of visual loss and requires immediate ophthalmic intervention. Patients symptomatic with flashes, floaters or decreased visual fields with one or more risk factors for retinal detachment should be treated with a high index of suspicion, and require an urgent dilated fundus examination by an ophthalmologist. (See the article 'Sudden loss of vision – history and examination' in this issue.) For all patients with media opacities obscuring the view of the retinal and posterior segment (including corneal pathology, dense cataract, vitritis and vitreous haemorrhage) ultrasound imaging is required (*Figure 1*).

Treatment of a retinal tear may be performed in the ophthalmology clinic through the use of barrier laser or cryopexy, whereas treatment of retinal detachment requires emergency operative management, extended postoperative care and prolonged visual rehabilitation. The urgency of the situation is heightened when the macular is uninvolved (ie. the central vision is preserved), as visual prognosis for 'macular off' retinal detachments is limited.

## Migraine

Partial or complete vision loss is a rare association of classic migraine. More typically, the visual aura of migraine presents as a positive scotoma (zigzag lights, shimmering or flickering colours) preceding the headache. The aura characteristically begins paracentrally and progresses temporally, with the leading edge a positive scotoma, and the trailing edge a negative scotoma.[4] Other visual associations include blurred or tunnel vision.

In keeping with the general investigation and management pattern of migraine, atypical cases such as those occurring in patients over 55 years of age, those with occipito basal headaches, or those with unresolving neurological signs and symptoms should be referred to a neurologist.[4]

## Indications for brain CT

Indications for CT of the brain and orbits include:
• suspicion of raised intracranial pressure
• proptosis
• chemosis
• homonymous field defect, or
• eye movement disorders.

If an orbital lesion is suspected, orbital CT should be specifically requested, as head CT in isolation does not completely image the orbit, sinuses and cavernous sinuses. Contrast should be requested in patients with adequate renal function when vascular, inflammatory or infective aetiologies are suspected. Magnetic resonance imaging is particularly useful in the diagnosis of orbital or intracranial masses (*Figure 2*), cavernous sinus lesions, neurogenic tumours and optic neuritis. Cortical blindness is a rare cause of sudden loss of vision due to occipital lobe infarction. This pathology preserves functions controlled by more primitive nuclei such as pupillary reflexes and reflex eye movements, as well as functions controlled by higher order

Figure 2. MRI revealed a large retro-orbital mass in a young man with vision loss, orbital pain and proptosis. When this man looked straight ahead his vision was 6/9 in the left eye, but in left gaze the optic nerve was stretched and his vision fell to 6/60. Intraorbital masses may also present with gradual loss of vision if they are not intricately related to the optic nerve in this manner
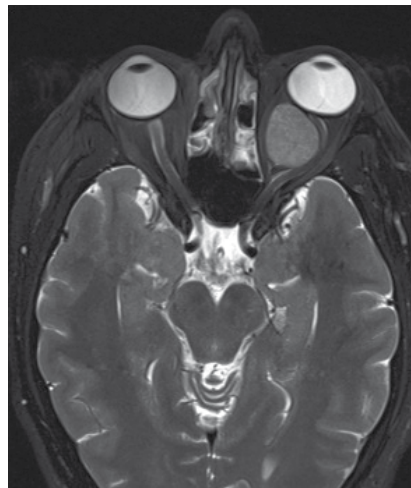


cortical areas such as motion perception, however dense bilateral loss of vision may result from bilateral occipital lobe infarction and requires urgent imaging and referral to a neurologist.

## Conclusion

Patients may present with transient changes in vision for which no cause can be found. It may be attributed to tear film abnormalities and less commonly migraine. However, it is important to remember that more sinister causes for transient visual obscuration exist, including GCA, carotid or cardiac emboli, retinal detachment, vitreous haemorrhage (especially in diabetic patients), orbital tumours and cerebral vascular events. A careful history, complete examination, astute investigation and appropriate referral will aid in early diagnosis and management and prevent further loss of vision or life.

Conflict of interest: none declared.

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