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Sudden loss of vision

History and examination

Background
Sudden loss of vision requires careful history and examination to identify the underlying cause.

Objective
This article discusses the various causes of sudden loss of vision and provides the general practitioner with a guide to examination.

Discussion
Rapidity of onset, duration and associated symptoms provide vital clues to the nature of the disease process. Simple examination techniques such as visual acuity measurement, confrontational visual field testing, pupil assessment and fundoscopy are integral to the appropriate assessment, treatment and referral of patients presenting with sudden loss of vision.

The sudden loss of vision is alarming to both the patient and clinician alike. Sudden and transient visual loss or obscuration may simply be a symptom of dry eye, or it may herald the onset of irreversible visual loss or stroke. The key to determining diagnosis is a focused history and thorough examination.

History

Eliciting a relevant history is important in deciding upon the significance of transient visual obscuration. This will help to identify the anatomic site of the pathology and lead to a more focused examination. Considering the anatomical pathways involved in normal vision provides a sound framework for elucidating the cause of vision loss.

It is vital to distinguish between true sudden vision loss and sudden realisation of visual loss. Spontaneous loss of vision with eyes open is likely to be an acute event, while gradual monocular loss of vision may appear to be acute when the normally functioning eye is temporarily obscured revealing poor vision in the fellow eye. For appropriate diagnosis and treatment, a detailed and robust history of any previous vision loss allows comparison with the superimposed acute changes.

Retrobulbar causes of sudden vision loss

It is critical to determine whether the visual loss involves one eye or both (*Table 1*). The patient must be specifically questioned as some may, for example, confuse a left homonymous field loss for a loss of vision in the left eye. Bilateral visual disturbance rarely indicates bilateral ocular or optic nerve disease, but more commonly reflects disease involving or posterior to the optic chiasm, such as pathology of the optic tract or cerebral cortex, including impaired posterior cerebral circulation or migraine.

Transient loss of vision, particularly in the setting of significant cardiovascular risk, may be amaurosis fugax resulting from an arterial embolus of cholesterol or thrombus. The site of embolisation defines the extent and topography of visual field loss (eg. homonymous hemianopia from a retrochiasmal embolus versus inferotemporal scotoma from a branch retinal artery embolus).

Table 1. Causes of sudden vision loss

Onset	Duration	Differential diagnosis	Associations
Seconds	Seconds	Tear film abnormalities	Clears with blinking
		Papilloedema, optic disc drusen, vitreous debris (floaters)	Associated with eye movement
Seconds	Variable	Orbital tumour	Associated with other orbital cranial nerve palsies
Seconds	Less than 10 minutes	Carotid occlusive disease, vertebrobasilar disease	Cardiovascular risk factors
		Cardiac emboli	Cardiovascular risk factors
		Giant cell arteritis	Headache, jaw claudication, scalp tenderness, diplopia, shoulder girdle pain, weight loss, night sweats
		Postural hypotension	Dehydration, cardiovascular disease, presyncope
Minutes	Minutes to hours	Transient angle closure glaucoma	Pain, haloes, blurred vision, red eye
		Migraine	Zig-zag' lights, headache
Seconds	Minutes to indefinite	Retinal artery occlusion	Cardiovascular risk factors
		Retinal vein occlusion	Cardiovascular risk factors
		Retinal detachment	Flashing lights, floating spots, 'curtain' progressing from side of visual field
		Optic neuritis	Pain, exacerbated by exercise or elevated body temperature (Uhthoff phenomenon)

Inflammation of the optic nerve (optic neuritis) is an important cause of vision loss, and the index of suspicion should be particularly high in patients with multiple sclerosis (70% suffer optic neuritis).[1] Inflammation may occur at the optic nerve only (retrobulbar neuritis), the optic disc (papillitis), or with retinal involvement (neuroretinitis), with patterns of vision loss relative to the site of inflammation. Young women are predominantly affected, and vision loss is usually unilateral. Associated neurological symptoms should also be specifically assessed.

Toxic causes of vision loss such as methanol toxicity should be considered in patients with a history of drug ingestion, fire breathing,[2] or consumption of home distilled spirits. Vision loss typically occurs 18–48 hours following methanol ingestion and manifests in a dense centrocecal scotoma[3] (visual field loss extending between the point of fixation and the blind spot).

Trauma, although an important cause of acute visual loss, is considered in detail in a recent *Australian Family Physician* article so is not discussed here.[4]

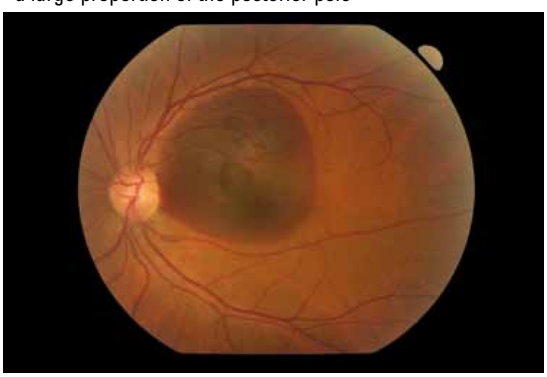
Visual loss in one eye is suggestive of a lesion anterior to the optic chiasm. This may be due to disease of the prechiasmal optic nerve or optic disc, carotid artery, retinal circulation or globe.

Intraocular causes of sudden vision loss

Discrete areas of monocular vision loss may represent intraocular lesions (eg. vitreous or retinal haemorrhage or retinal detachment), while monocular vision loss respecting the horizontal meridian may result from vascular lesions of the optic disc or retinal circulation.[5]

A fixed scotoma is likely to represent pathology in structures fixed relative to the globe, such as corneal, lenticular, retinal or optic nerve or neurological damage. A floating, mobile scotoma however, is consistent with vitreous pathology as the vitreous undulates relative to the retina with movement of the globe. Painless sudden vision loss in a diabetic or hypertensive patient may suggest vitreous haemorrhage, often described as 'a shower of black dots'. Subhyaloid, retinal or subretinal haemorrhages (*Figure 1*) are more discrete, less mobile, and may initially appear as a red, brown or black scotoma.

Figure 1. Subretinal haemorrhage involving the macular and a large proportion of the posterior pole



A painless, sudden onset fixed scotoma in a patient with significant cardiovascular risk may result from a retinal artery or vein occlusion. Artery occlusions are more rapid in their onset than vein occlusions (seconds compared with minutes), and the

site of vessel occlusion determines extent of the scotoma (ie. a central retinal vessel occlusion results in global monocular vision loss, while a branch retinal vessel occlusion causes a segmental scotoma). Sudden loss of vision associated with headaches, jaw claudication, scalp tenderness, unexplained weight loss, night sweats, diplopia or temporal artery tenderness is strongly suggestive of giant cell arteritis (GCA).[6] This diagnosis should be considered in any patient over the age of 50 years with sudden onset loss of vision or diplopia.

Loss of vision preceded by flashing lights, floating shadows or a 'curtain' progressing from the periphery of the visual field is highly indicative of retinal detachment. Particular attention should be given to such patients who also report a history of cataract surgery, high myopia, recent eye trauma, personal or family history of retinal detachment, or history of connective tissue disease.

Duration of vision loss

The duration of the visual loss (*Table 1*), the degree to which it recovers, and any associated symptoms are also important. Visual loss lasting for seconds to minutes may be due to embolic phenomena or optic nerve head drusen – deposits of extracellular material under the retina (*Figure 2*); whereas visual loss occurring over days to weeks may indicate either a compressive, infiltrative or inflammatory pathology. A visual obscuration that recovers with blinking is likely to be due to tear film abnormalities, whereas one that occurs in the setting of headache, photophobia and sonophobia, may be due to migraine.

Associated symptoms

It is also important to elicit associated symptoms such as double vision, sore eyes, red eyes, haloes and flashing lights. These symptoms may be associated with other cranial nerve lesions, ocular surface disorders, angle closure glaucoma and migraine. Past history of migraine, childhood motion sickness, family history of migraine, transient ischaemic attack or stroke, and atherosclerotic risk factors including smoking or recent trauma (eg. neck manipulation, motor vehicle accident, roller coaster ride) may also provide clues to the aetiology. Acute angle closure glaucoma is typified by vision loss associated with intense pain, haloes, conjunctival injection, nausea, vomiting and photophobia.[7]

Examination

A simple but thorough ophthalmic examination in the general practice setting will greatly assist in the diagnosis and management of acute vision loss. Assessment of visual acuity, visual fields to confrontation, pupil reaction, quality of the red reflex and assessment of cranial nerves may all be performed in the general practice surgery with minimal specialised equipment and is highly informative.

Inspection

Inspection of the face and eye should be directed toward looking for ptosis, proptosis, injection or chemosis (oedema of the conjunctiva).

Figure 2. Optic nerve head drusen may mimic optic disc swelling

Figure 3. Testing visual fields to confrontation

General inspection of the patient's level of functioning and navigational abilities may augment information obtained from formal visual acuity testing. Visual acuity testing should be performed to assess the presenting vision (ie. wearing glasses if the patient usually wears them), and using a pinhole to correct for refractive error (even if the patient wears glasses). This should be performed before other steps in the examination to avoid confounding from bright lights.

Visual fields

Visual fields to confrontation are of particular importance in cases where history points to a retinal or neurological cause for vision loss. The physician should sit directly opposite the patient, have the patient cover one eye, and the physician should occlude his/her opposite eye like a mirror image. While fixating on the physician's pupil, the patient should indicate when a moving target is first seen when slowly brought in from the periphery by the physician (*Figure 3*). The patient's visual field should be compared with the physician's visual field during testing.

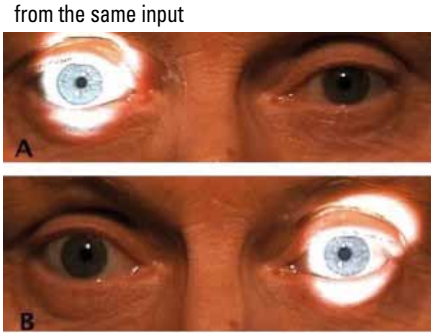
Pupil assessment

Arguably one of the most important parts of the ophthalmic examination is the assessment of the pupils. Normal pupil reactions require normal functioning of both oculomotor and optic nerves. For this reason, the direct and consensual pupillary responses should be assessed in each eye. Subtle optic nerve deficits may be detected by

Table 2. Examination steps for a relative afferent pupillary defect

- Darken the room
- Have the patient fix on a distant target (eg. the top letter on a Snellen chart)
- Alternate a bright light rapidly (<1 second) between the two eyes, spending 2 seconds on each eye
- Compare the initial constriction and the initial dilatation of the pupils
- Shining the light on the normal eye will result in bilateral pupillary constriction
- Swinging across to the abnormal eye will result in bilateral pupillary dilatation (Figure 4a, b)

Figure 4a, b. This man had acute onset of painless visual loss in the left eye (6/60) following admission for herpes simplex encephalopathy. a) When a bright light was shone on the right pupil there was equal constriction of both pupils due to the direct and consensual light reflex; b) After the light was quickly swung across to the left pupil, both pupils dilate due to a relative reduction in the afferent impulse from the left optic nerve. Both pupils in Figure 4a are smaller than both pupils in Figure 4b, despite stimulus from the same input



testing for a relative afferent pupil defect (*Table 2, Figure 4*). Pupils should be specifically checked for anisocoria (unequal pupils) as this may be suggestive of trauma, third nerve palsy or Horner syndrome. Teardrop shaped pupils are suggestive of penetrating eye injury (as the iris is drawn to the laceration site), and irregular pupils in a painful red eye with no history of trauma may indicate acute anterior uveitis.

Colour vision

Full examination of optic nerve function also requires assessment of red desaturation, brightness perception (commonly performed by asking the patient to compare the brightness of a torch shone in each eye), and Ishihara colour vision testing. The assessment of red desaturation can be performed using a red hat pin or the red lid of a dilating eye drop bottle. The patient is asked whether the intensity of red is the same in each eye. An eye affected by some forms of optic neuropathy will perceive a dull red colour or no red at all.

Fundoscopy

Fundoscopy should be performed following dilation if the pupil examination is normal and trauma or acute angle closure glaucoma is not suspected. Phenylephrine 2.5% and tropicamide 1% are often used with a duration of action of 2–6 hours. Caution should be exercised in elderly or hypertensive patients when considering phenylephrine use. Before examining retinal and optic nerve features, the red reflex should be assessed in each eye. This may be obscured by media opacities such as cataract or vitreous haemorrhage, or intraocular pathologies such as retinal detachment (*Figure 5*). As a general rule of thumb, the quality of the red reflex often gives an indication of the limitation of visual impairment due to media opacities.

The optic disc may be located by following blood vessels and should be inspected for pallor, swelling (*Figure 6*), haemorrhage (*Figure 7*) or gross cupping (*Figure 8*). Retinal blood vessels may reveal signs of hypertensive retinopathy, emboli (*Figure 9*), or venous congestion. The macular, found temporal to the optic disc, should be examined last. Particularly in the undilated eye, this minimises patient discomfort and reduces the effect of pupillary constriction on the remainder of the fundus examination. A cherry-red spot (*Figure 10*) is indicative of retinal ischaemia (as the macular supplied by the choroidal vascular bed beneath becomes more prominent relative to the surrounding ischaemic retina supplied by the retinal vessels).

Figure 5. Colour fundus photograph of a retinal detachment. The pale inferior retina has detached from the retinal pigment epithelium, resulting in loss of the normal choroidal hue beneath. The retinal vessels, particularly as they emerge from the inferior aspect of the optic disc, can be seen billowing forward with the retina

Figure 6. Colour fundus photograph showing pallid disc swelling in giant cell arteritis anterior ischaemic optic neuropathy

Figure 7. Colour fundus photograph of a central retinal vein occlusion. (Note tortuous dilated vessels, optic disc swelling and widespread retinal nerve fibre layer haemorrhages)

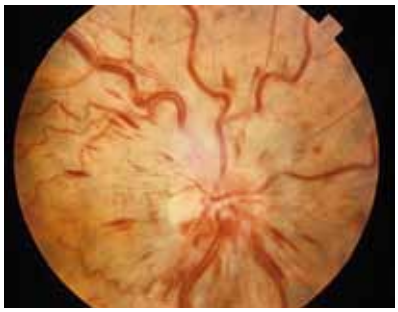


Figure 8. Gross optic disc cupping consistent with chronic glaucoma

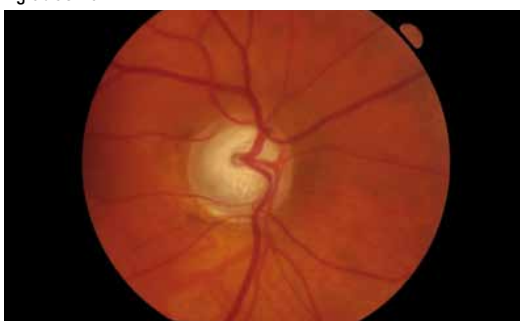
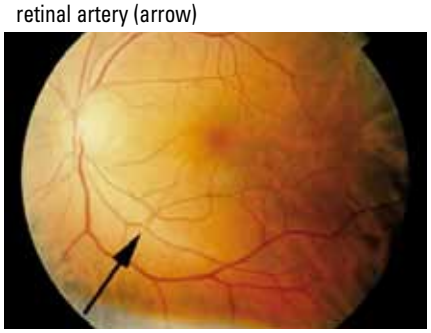


Figure 9. Colour fundus photograph demonstrating an embolus within a branch retinal artery (arrow)

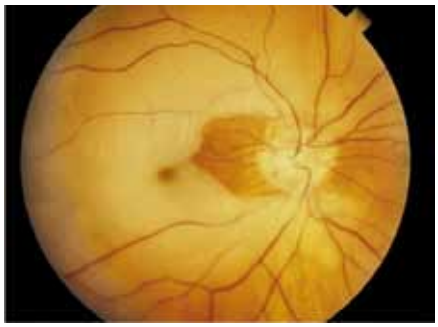


Other examinations

Careful examination of the cranial nerves may determine the cause of vision loss through reconciliation of symptoms with anatomical structures. Vision loss in concurrence with lesions of the intraorbital cranial nerves (optic, oculomotor, trochlear, the ophthalmic branch of the trigeminal and abducens) may indicate an orbital mass, while central retinal vein occlusion and involvement of the ophthalmic and maxillary branch of the trigeminal nerve may suggest cavernous sinus thrombosis.

Historical features and examination findings may also lead the physician to check blood pressure or blood sugar levels and to auscultate the carotid arteries for bruits or the heart for murmurs. It may also be necessary to check the function of more inferior cranial nerves, or to conduct a more thorough neurological examination, especially in the setting of a homonymous field defect. Where GCA is suspected it is also imperative to palpate the temporal arteries looking for tenderness or reduced pulsatility.

Figure 10. Colour fundus photograph demonstrating central retinal artery occlusion with sparing of the cilioretinal artery. The cilioretinal artery is present in 20–30% of the population, and derives its blood supply from the choroidal rather than the retinal circulation. Superotemporal (upper right) vessels exhibit cattle trucking (segmentation of the blood column in the arterioles) and narrowed retinal arterioles



Conclusion

Patients often present with transient changes in vision for which no obvious cause can be determined. In these cases the cause of the visual obscuration may innocuous. But one must not be tempted into premature closure of diagnostic hypotheses. It is important to remember that more sinister causes for visual obscuration exist, including GCA, carotid or cardiac emboli, orbital tumours, cerebral vascular events and retinal detachment. A considered history and thorough examination will result in more appropriate investigation and a better informed management plan.

Further reading

Donders RC. Dutch TMB Study Group. Clinical features of transient monocular blindness and the likelihood of atherosclerotic lesions of the internal carotid artery. J Neurol Neurosurg Psychiatry 2001;71:247–9.

Conflict of interest: none declared.

References

1. Denniston AKO, Murray PI. Oxford handbook of ophthalmology. Oxford: New York: Oxford University Press, 2006.
2. Cursiefen C, Bergua A. Acute bilateral blindness caused by accidental methanol intoxication during fire 'eating'. Br J Ophthalmol 2002;86:1064–5.
3. McKellar MJ, Hidajat RR, Elder MJ. Acute ocular methanol toxicity: clinical and electrophysiological features. Aust N Z J Ophthalmol 1997;25:225–30.
4. Hodge, C, Lawless, M. Ocular emergencies. Aust Fam Physician 2008;27:506–9.
5. Kanski JJ. Clinical ophthalmology: A systematic approach. 6th edn. Edinburgh: New York: Butterworth-Heinemann/Elsevier, 2007.
6. Danesh-Meyer HV, Savino PJ. Giant cell arteritis. Curr Opin Ophthalmol 2007;18:443–9.
7. Ehlers JP, Shah CP, Wills Eye Hospital (Philadelphia Pa.). The Wills eye manual: Office and emergency room diagnosis and treatment of eye disease. 5th edn. Philadelphia, PA: Lippincott Williams & Wilkins, 2008.

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