

Acute Visual Loss

Sudden onset of blindness is a disaster for most people and you should be able to evaluate such a patient and be able to recognize situations requiring urgent action.

History:

The pertinent history questions to be asked of a patient of sudden visual loss include:

1. Is the visual loss transient or persistent?
2. Is the visual loss monocular or binocular?
3. Did the visual loss occur suddenly or it developed over hours, days or weeks?
4. What is the patient's age and general medical condition?
5. Did the patient have normal vision in the past and when was vision last tested last, as quite a number of people will realize loss of vision from one eye when they cover the good eye.

In order to examine a patient with acute visual loss, you should be able to perform the **following tests:**

- Visual acuity testing
- Confrontation visual fields
- Pupillary reactions
- Ophthalmoscopy
- External examination of the eye with a pen light
- Tonometry to measure the intraocular pressure

Etiology of acute visual loss can be considered under the following headings:

1. Media opacities
2. Retinal disease
3. Optic nerve disease

4. Visual pathway disorders
5. Functional disorders
6. Acute discovery of chronic visual loss

1. **Media opacities** –

Any opacity of the clear refractive media of the eye, i.e. cornea, anterior chamber, lens, and vitreous will cause blurred vision or a reduction of visual acuity. These conditions do not cause a relative afferent pupillary defect, although pupillary reflexes may be altered.

Corneal edema can produce a sudden opacity of the cornea. The cornea appears like a ground glass rather than its normal clear appearance. The most common cause of corneal edema is increased intraocular pressure and occurs typically in angle closure glaucoma. Chronic damage to the corneal endothelium by dystrophies, or following cataract surgery can also produce corneal edema, but the visual loss will be gradual. Any acute infection of the cornea by a corneal ulcer may mimic corneal edema.

Hyphema – Hyphema is blood in the anterior chamber, significant hyphema reduces vision, and a complete hyphema may completely obliterate vision. The hyphema is a direct consequence of blunt trauma to a normal eye. However, it can occur with tumors, diabetes, intraocular surgery and chronic inflammation – which all cause neovascularization.

Cataract – most of the cataracts develop slowly, however, if a cataract progresses very rapidly, it can lead to sudden visual loss. The visual acuity in a diabetic can be altered, if the blood sugar rises very high and a patient may report this as a visual loss. The vision usually will come back to normal levels once the blood sugars are properly controlled.

Vitreous hemorrhage – Any bleeding into vitreous will also reduce the visual acuity. A large vitreous hemorrhages can occur after trauma, but have also been seen in diabetics or after a retinal vein occlusion and it may also accompany subarachnoid hemorrhage. If you cannot appreciate a red reflex with an ophthalmoscope and the lens appears clear, you should suspect a vitreous hemorrhage. The diagnosis is confirmed with slit lamp examination through a dilated pupil.

2. **Retinal diseases** -

The following retinal diseases may cause sudden visual loss.

Retinal detachment – an extensive retinal detachment involving the macular area would produce acute visual loss and this patient will complain of flashing lights followed by a large number of floaters and then a shade or blind covering the visual field. An afferent pupillary defect is usually present. The diagnosis is confirmed by ophthalmoscopy through a dilated pupil, and retina appears elevated with folds and the choroidal background is indistinct.

Macular disease – macular degeneration usually reduces visual acuity and there is no relative afferent pupillary defect. However, if there is a bleeding from a sub-retinal neovascular membrane, it can give rise to sudden loss of vision.

Retinal vascular occlusion - retinal vascular occlusion is a relatively common cause of sudden loss of vision. A transient episode of monocular visual loss is called amaurosis fugax. In a patient who reports transient visual loss in one eye lasting for several minutes followed by gradual recovery should be investigated by carotid Doppler's, as an atheroma in carotids may be the source of emboli to the retinal vessels. An urgent referral in such a situation is necessary to an Ophthalmologist, Neurologist or a vascular surgeon.

Central Retinal artery occlusion – any prolonged interruption of the retinal arterial blood flow causes permanent damage to the ganglion cells and other tissue elements. A sudden, painless and often complete visual loss may indicate a central retinal artery occlusion. The fundus appearance of such a patient depends upon how soon the patient is examined. Very early on, the only finding may be a vascular stasis that is narrowing of the arterial blood column and interruption of the venous blood columns with the appearance of boxcarring. Several hours after a central retinal artery occlusion, the inner layer of the retina becomes opalescent. A cherry red spot is seen due to the pallor of the perifoveal retina in contrast to the normal color of the fovea. A chronic cherry red spot is also a feature of the storage diseases such as Tay-Sachs disease and Niemann-Pick disease.

The optic disk, which is supplied by other branches of the ophthalmic artery, does not swell unless the occlusion is in the ophthalmic or carotid artery, proximal to the origin of the central retinal artery. Sometimes the vision may be preserved, if the macular area is supplied by via a cilio-retinal artery. When a patient with central retinal artery occlusion is examined months later, the characteristic appearance is a pale disc and a blind eye, as the retinal edema slowly resolves and the death of the ganglion cells and their axons leads to optic atrophy.

Central retinal artery occlusion is a true ophthalmic emergency and restoration of blood flow may preserve vision if the occlusion is only a few hours old. In these cases, immediate treatment is required.

As an emergency measure, the primary care physician may compress the eye with the heel of the hand, pressing firmly for ten seconds and then releasing for ten seconds over a period of approximately five minutes. The sudden rise and fall in intraocular pressure could serve to dislodge small emboli in the central retinal artery and restore circulation before the retinal tissues sustain irreversible damage. An ophthalmologist might employ more invasive techniques including retrobulbar injection of anesthetic and paracentesis of the anterior chamber.

Branch Retinal Artery Occlusion – when only a branch of the central retinal artery is occluded, vision is only partially lost. This is more likely to be the result of an emboli and the source of the emboli should be sought. If the visual acuity is affected, attempts should be made to dislodge the emboli by ocular massage.

Central Retinal Vein Occlusion – a central retinal vein occlusion will show the ophthalmoscopic picture of disc swelling, venous engorgement, cotton wool spots and diffuse retinal hemorrhages like blood and thunder. Loss of vision may be severe. There is no generally accepted acute management. Central retinal vein occlusion is not a true ophthalmic emergency.

A central retinal vein occlusion is often encountered in old patients with hypertension and artero- sclerotic vascular disease. In rare cases, diseases that alter blood viscosity such as Polycythemia Vera, Sickle cell disease and Lymphoma. Leukemia may induce central retinal vein occlusion.

The acute hemorrhages and disc swelling resolve with time, however, they may be followed by development of shunt vessels from the retinal to the choroidal circulation and by ocular neovascularization. These patients need a general medical evaluation and follow up by an ophthalmologist and they do stand the possibility of developing neovascular glaucoma.

3. Optic Nerve Disease:

Any disease affecting the optic nerve can often result in acute visual loss. The pupillary reaction is abnormal with an afferent pupillary defect in unilateral disease, but the optic nerve head may appear quite normal initially.

Optic Neuritis: Optic Neuritis is inflammation of the optic nerve and is usually associated with multiple sclerosis in a significant number. The visual acuity is markedly reduced and an afferent pupillary defect is present. The optic disc initially appears hyperemic and swollen. The visual acuity usually recovers; however, repeated episodes of optic neuritis may lead to permanent loss of vision.

Sometimes high dose intravenous cortical steroids have been tried for optic neuritis. Optic neuritis should be referred to an ophthalmologist.

Retro-bulbar neuritis : Retro-bulbar neuritis typically occurs in a young adult with monocular loss of vision that has developed over hours to days and is often accompanied by pain on movement of the eye.

The optic nerve on ophthalmoscopic examination appears completely normal. The visual acuity is markedly reduced and an afferent pupillary defect is present. A compressive optic neuropathy should be considered in the differential diagnosis. MRI and CT will identify most compressive lesions which are potentially treatable with surgery.

Papillitis and Papilledema: Papillitis is a sub-type of optic neuritis and is an inflammation of the optic nerve head. Papilledema on the other hand refers to swelling of the optic disc from increased intra-cranial pressure.

In optic neuritis, (retro-bulbar / optic neuritis) vision is usually decreased and an afferent pupillary defect is usually present. In papilledema, the visual acuity and pupillary reflexes are usually normal.

Some patients with acute papilledema may complain of momentary blurring or transient obscurations of vision. However, the visual acuity may be lost to some extent with chronic papilledema.

Ischemic optic neuropathy: Ischemic optic neuropathy is a vascular disorder that presents with a pale swollen disc often accompanied by splinter hemorrhages and loss of visual acuity and visual field. The visual field loss is usually superior or inferiorly field defect. Ischemic optic neuropathy usually occurs in older individuals and represents a vascular event.

Giant Cell Arteritis / Temporal arteritis: In ischemic optic neuropathy in a patient over age 60 raises the possibility of giant cell or temporal arteritis. This is often associated with complaints of malaise, headache, fever, weight loss, pain and tenderness of muscles and joints, scalp tenderness or discomfort when combing the hair and a virtually pathognomonic pain in the jaws on chewing termed as jaw claudication.

Ocular complaints may include sudden visual loss and diplopia.

A sedimentation rate should be obtained immediately and many elderly patients with temporal arteritis have very elevated sedimentation rates greater than 60 mm per hour. C-reactive protein levels will also be elevated. Temporal arteritis is treated with high dose systemic cortico-steroids and this may preserve the vision in the remaining eye and prevent vascular occlusion, otherwise it could cause a stroke or myocardial infarction. Immediate referral to an ophthalmologist is indicated, if temporal arteritis is a strong possibility.

Temporal artery biopsy usually confirms the diagnosis.

If no systemic arteritis is demonstrated, there is no clear evidence that systemic cortico-steroids benefit patients with ischemic optic neuropathy. There is approx. 40% chance that the other eye will become involved with non-arteritic ischemic optic neuropathy with or without treatment.

Trauma: a concussion or head trauma may shear the vascular supply to the optic nerve thus producing blindness. These cases are termed as traumatic optic neuropathy and surgical decompression of the optic canal may be undertaken in selected cases.

4. Visual Pathway Disorders :-

Homonymous hemianopia - is loss of vision on one side of both visual fields and may result from occlusion of one of the posterior cerebral arteries with infarction of the occipital lobe. Other vascular abnormalities occurring in the middle cerebral artery distribution may produce a hemianopia, but usually other neurological signs are prominent. Any patient with a hemianopia needs a CT or MRI to localize and identify the cause.

Cortical Blindness: A rare extensive bilateral damage to the cerebral visual pathways results in complete loss of vision. This condition is referred to as cortical, central or cerebral blindness. As the pathways serving the pupillary light reflex separate from those carrying visual information at the level of the optic tracts, a patient who is cortically blind has normal pupillary reactions. Thus a patient with normal fundus examination along with normal pupillary reactions, most likely has cortical blindness. Most patients with cortical blindness either improve or will die due to severe neurological damage. Transient cortical blindness has been observed in children after sub-concussive head trauma.

5. Functional Disorders

A functional disorder is used in preference to hysterical or malingering to describe visual loss without organic basis. A patient may report complete blindness in one eye and normal vision in the other eye, but has normal stereopsis and no relative afferent pupillary defect.

6. Acute Discovery of chronic visual loss

A large number of cases of chronic visual loss turn up as acute visual loss discovery. A person may claim acute visual loss in one eye but has advanced optic atrophy, which has been unrecognized so far.