**Approach to a Case of Transient Visual Loss**

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Transient visual loss (TVL) is defined as an abrupt temporary monocular or binocular loss of vision. It may be due to vascular, neurologic or ophthalmic causes. It commonly results from impaired blood supply to the afferent visual system, due to primary arterial occlusion or stenosis, secondary arterial occlusion as a result of embolism from a distant site (eg. internal carotid artery (ICA), aortic arch, or heart), vasospasm (eg. migraine), or systemic hypoperfusion.

**Amaurosis Fugax:** The term amaurosis fugax is often used to describe transient ischemic attack of the retina. Because neural tissue has a high rate of metabolism, interruption of blood flow to retina for more than a few seconds results in transient monocular blindness, a term used interchangeably with amaurosis fugax. Amaurosis fugax commonly results from retinal embolus that transiently occludes a retinal arteriole. If the embolus breaks up or passes, flow is restored and vision returns quickly to normal without permanent damage. However, with prolonged interruption of blood flow the inner retina suffers infarction. Ophthalmoscopy reveals zones of whitened edematous retina corresponding to the distribution of branch retinal arterioles. Complete occlusion of the central retinal artery produces arrest of blood flow and milky retina with a cherry-red fovea. The most common source is an atherosclerotic plaque in the carotid artery or aorta. Emboli most commonly arise from an atheromatous plaque in the ICA, but can originate from the heart (as in patients with atrial fibrillation, valvular heart disease and dilated cardiomyopathy) and atheromatous plaque in the aortic arch. Less commonly, they arise from atheromatous plaque in the common carotid or ophthalmic arteries. Rarely, the emboli arise from an atrial myxoma or are paradoxical, traveling from the systemic venous system to the systemic arterial system via a cardiac septal defect or pulmonary arterio-venous fistula. Very rarely, septic, air, fat, silicon, or talc emboli can cause TVL.

Retinal emboli must be sought with a dilated funduscopic examination in patients presenting with transient monocular visual loss, although the absence of retinal emboli does not exclude them as
a cause. Three embolus subtypes may be identified: cholesterol, platelet-fibrin, and calcium.

Cardiac emboli can arise from the left atrial appendage in patients with atrial fibrillation, severe mitral stenosis with stasis of blood in the left atrium, thrombus in the dyskinetic left ventricular apex following anterior wall myocardial infarction, dilated cardiomyopathy with severe left ventricular dysfunction and calcific aortic valve in the elderly.

Rarely amaurosis fugax can occur from low central retinal artery perfusion pressure in patients with critical carotid stenosis with poor collateral flow from the circle of Willis. This can happen when there is transient hypotension. This may be associated with contralateral motor or sensory loss indicating concomitant hemispheric cerebral ischemia.

Transient ischemic attacks from vertebrobasilar insufficiency can result in acute homonymous visual symptoms. Many patients mistakenly describe symptoms in the right or left eye. Interruption of blood supply to the visual cortex causes a sudden fogging or graying of vision occasionally with flashing lights or other positive phenomena that may mimic migraine.

Ophthalmic Artery Stenosis and Occlusion: Stenosis of the ophthalmic artery due to atheroma can produce isolated episodes of transient monocular visual loss, as a consequence of retinal and optic nerve hypoperfusion or embolism to the retinal arteries.

Retinal arterial occlusion can occur rarely in association with retinal migraine, lupus erythematosis, temporal arteritis, hypercoagulable states like anticardiolipin antibodies, anticoagulant deficiency states (protein C, protein S and anti thrombin deficiency) and pregnancy.

Impending branch or central retinal vein occlusion can produce prolonged visual obscurations.

Sudden visual loss can occur in hypertensive crisis due to vasospasm of retinal arterioles and consequent retinal ischemia. Marked systemic hypertension causes sclerosis of retinal arterioles, splinter hemorrhages and focal infarcts of the nerve fiber layer (cotton–wool spots) and leakage of lipid and fluid (hard exudate) into the macula. Acute hypertension may produce visual loss from ischemic swelling of the optic disc.

**Common Causes of Transient Visual loss**

**Monocular visual loss**

**Vascular**
- Internal carotid artery stenosis, occlusion, or dissection
- Common carotid artery stenosis, occlusion, or dissection
- Ophthalmic artery stenosis, occlusion, or dissection
- Aortic arch atheroma
- Cardioembolic source (atrial fibrillation, severe mitral stenosis, calcific aortic stenosis left ventricular thrombus in dilated/ischemic cardiomyopathy)
- Giant cell (temporal) arteritis
- Aortoarteritis
- Arterial vasospasm
- Hypercoagulable states
- Systemic hypoperfusion

**Neurologic**
- Retinal migraine

**Ophthalmic**
- Papilledema and optic disc edema
Optic disc drusen
Optic neuritis
Intermittent angle-closure glaucoma
Tear film dysfunction and dry eye

**Binocular visual loss**

**Vascular**
- Transient ischemic attacks
- Bilateral carotid artery stenosis or occlusion
- Systemic hypoperfusion

**Neurologic**
- Migraine aura
- Occipital seizures
- Posterior reversible encephalopathy syndrome
- Head trauma

**Ophthalmic**
- Papilledema and optic disc edema
- Optic disc drusen

**Anterior Ischemic Optic Neuropathy (AION):**
Is caused by insufficient blood flow through the posterior ciliary arteries supplying the optic disc. It produces sudden painless monocular visual loss. The optic disc appears swollen with splinter hemorrhages around it. AION is of two types: arteritic and non arteritic. Non arteritic form is the most common with no specific identifiable cause though diabetes and hypertension are frequent risk factors. 5% of patients of AION, especially those over 60 years develop the arteritic form in conjunction with giant cell arteritis.

**Posterior Ischemic Optic Neuropathy:** Is an uncommon cause of acute visual loss. It is induced by a combination of anemia and hypotension, causing infarction of the retrobulbar optic nerve. This may follow major blood loss during major surgery, exsanguinating trauma or gastrointestinal blood loss. Fundus usually appears normal.

**Optic Neuritis (Uhthoff phenomenon):** Is a common inflammatory disease of the optic nerve. In most patients demyelination is retrobulbar and hence fundus appears normal initially though disc pallor may develop later. They can develop episodic transient visual loss lasting minutes to hours with increase in body temperature (Uhthoff phenomenon). Episodes pose no threat to permanent vision. Vision returns to baseline when body temperature comes back to normal. This is due to transient conduction block within the optic nerve.

**Optic disc drusen:** Brief episodes of transient visual loss can occur in patients with optic disc drusen. Drusen may be visible on fundoscopy unless buried, when it may mimic papilledema. B scan ultrasound and CT scan can differentiate it from papilledema.

**Toxic optic neuropathy:** Can result in acute visual loss with bilateral optic disc swelling and central or centro-cecal scotomas. Cases have been reported to result from exposure to methyl alcohol, ethambutol, ethylene glycol or carbon monoxide.

**Papilledema:** Episodes of transient visual obscurations are characterized by complete or partial loss of vision lasting for seconds. Episodes are precipitated by postural changes and maneuvers that increase intracranial pressure such as coughing and straining. They are thought to be consequent to transient ischemia of the swollen optic nerve head.

**Classic migraine:** This usually occurs with a visual aura lasting about 20 minutes. Patients’ description of fortification spectra vary widely and can be confused with amaurosis fugax. Migraine pattern usually lasts longer and are perceived in both eyes where as amaurosis fugax is briefer and occurs in...
only one eye. Migraine phenomena remain visible in the dark or with the eye closed. After the visual symptoms recede headache develops in most patients. Cortical ischemic attacks are briefer in duration than migraine, occur in older patients and are not followed by headache. There may be associated signs of brainstem ischemia such as diplopia, vertigo, numbness, weakness or dysarthria.

**Occipital seizures:** Typically produce a sudden onset of binocular elementary positive visual phenomena with associated visual loss. In contrast with migraine, the positive visual phenomena consist of multiple, brightly colored, small circular spots, circles, or balls. They are usually located in the contralateral hemifield, but may be central. Vision is obscured in the area occupied by the hallucinations from the time of onset. It occurs in patients with posterior reversible encephalopathy syndrome (PRES), metabolic encephalopathies, malformations of cortical development, neoplasms, vascular lesions, prior head trauma, metabolic diseases (eg, mitochondrial disease), localized infections, or they may be idiopathic. They are diagnosed on the basis of the history, imaging findings, and electroencephalographic findings.

**Head Trauma:** Isolated transient cortical blindness may rarely result from minor blunt head trauma, usually direct occipital trauma, in children or adolescents. The visual loss commonly develops within minutes after the event.

**Orbital Masses and Foreign Bodies:** Orbital masses and foreign bodies may also give rise to episodic TVL. Episodes classically occur when the eye adopts a certain eccentric gaze position: this so-called gaze-evoked amaurosis rapidly remits once the eye moves out of the offending gaze position. There are usually obvious signs of orbital disease on examination or a history of trauma or penetrating injury. The episodes of TVL remit with treatment of the underlying cause.

**Other Ophthalmic Causes:** TVL may occur with several other ophthalmic diseases, although some of these produce visual blurring or visual distortion rather than actual loss of vision. Age-related macular degeneration can result in light-induced TVL due to increased photostress recovery time. The episodes are usually bilateral and affected patients have decreased acuity with obvious signs of macular degeneration on fundus examination. Intermittent angle-closure glaucoma can rarely present with isolated painless TVL although most patients have pain, vomiting, or other visual symptoms, such as halos, during episodes. TVL can rarely result from recurrent hyphaema, such as in the uveitis-glaucoma-hyphema syndrome that occasionally complicates cataract extraction with intraocular lens implantation. The patient reports a rapid reduction in vision over minutes, followed by gradual resolution over hours to days. There may be associated pain and erythropsia (red vision) and a microhyphema may be seen if the patient is examined during an attack. Corneal basement membrane dystrophy can also produce isolated TVL, but it is usually associated with pain. Signs of this disease can be detected on a careful anterior segment examination. Finally, dry eye and tear film dysfunction can result in TVL. The patient may report that their vision improves after blinking or after the application of artificial tears. Signs of dry eye may be present on examination.

**Idiopathic:** The cause of recurrent attacks of transient monocular visual loss frequently remains obscure in younger patients. As headache or orbital pain may be associated with the episodes of visual loss, it has been suggested that migraine or vasospasm is a possible cause. Alternatively, increased venous outflow resistance may play a role in the pathogenesis of the attacks. TVL can also occur in the setting of hypercoagulable state; the frequency of attacks may decrease with antiplatelet therapy or anticoagulation. If no cause is identified, the natural history of episodic transient monocular visual loss is frequently benign, especially in
adolescents and young adults. Calcium channel blockers may reduce the frequency of attacks in patients with presumed vasospasm.

**Fictitious visual loss:** This is claimed by hysterics or malingerers

**Clinical Evaluation of Patients with TVL**

**History**

A careful and detailed history is of paramount importance as in many cases, no abnormalities will be detected on physical examination or diagnostic investigations. It is especially important to establish what the degree of visual loss was, as the differential diagnosis, investigation, and management of transient blindness is quite different to that of transient visual blurring or distortion, which could be due to a relatively benign ophthalmic problem such as tear film dysfunction.

It is useful to focus on the following points to arrive at a diagnosis.

**Age:** Younger patients tend to have a more benign cause of TVL. (eg. Migraine), whereas older patients often have a more sinister cause.

**Monocular or binocular:** Monocular TVL suggests a pre-chiasmal lesion, whereas binocular TVL suggests a chiasmal, retrochiasmal or bilateral pre-chiasmal lesions. An important caveat is that homonymous visual field loss is frequently mistaken for monocular visual loss on the side with the temporal field defect; it is therefore useful to ask if visual loss was noted in the other eye when the affected eye was covered during the episode.

**Precipitating factors:** Although many episodes of TVL occur spontaneously, the presence of a precipitating factor may be helpful for narrowing the differential diagnosis. For instance, TVL precipitated by postural changes may occur with papilledema, giant cell arteritis, and hypotension. Gaze-evoked TVL may occur with intraorbital mass lesions or foreign bodies and heat-evoked TVL (Uhthoff’s phenomenon) may occur with demyelinating optic neuropathies.

**Pattern of onset and recovery:** Although not specific, a description of the pattern of visual loss may help in narrowing the differential diagnosis. For instance, an altitudinal onset of TVL (“like a curtain or shade descending”) may indicate embolic arterial occlusion, whereas a concentric onset of TVL, may indicate a vasospastic or neurologic cause of TVL.

**Duration of TVL:** TVL from papilledema and optic disc drusen typically lasts for seconds, TVL from retinal emboli or transient ischemic attacks (TIA’s) typically lasts for several minutes (usually less than 15 min), whereas TVL from migraine typically lasts for more than 15 minutes.

**Associated symptoms:** Many causes of TVL produce other symptoms during the attack such as headache, positive visual phenomena such as sparkles or flashes, and focal neurologic symptoms. The patient should be enquired about symptoms like headache, jaw claudication, scalp tenderness, or polymyalgia in patients with suspected giant cell arteritis.

**Comorbidities:** A past history of vascular risk factors (hypertension, diabetes mellitus, dyslipidemia, and tobacco smoking), cardiovascular disease (coronary artery disease, valvular heart disease, atrial fibrillation, aortoarteritis, stroke), and migraine should be specifically sought. Past history of polymyalgia rheumatica, connective tissue disease (eg systemic lupus erythematosus), hypercoagulable state or ophthalmic disease may be relevant.

**Examination**

A thorough examination is essential to document the current state of the afferent visual system and to detect signs that suggest a cause for the TVL. An
ophthalmic examination, including an assessment of the orbits, anterior segment, and intraocular pressures, and funduscopy, is essential to find out the etiology (eg, asymptomatic retinal emboli or signs of the ocular ischemic syndrome due to atheromatous carotid stenosis). Assessment of the pulse (for presence of atrial fibrillation) and blood pressure are also important. Cardiovascular exam may reveal features of valvular heart disease (like mitral stenosis and aortic stenosis which is most often calcific in the elderly), dilated and restrictive cardiomyopathy. Carotid bruit is audible when there is significant internal carotid artery stenosis unless there is total occlusion. Asymmetric pulses, bilateral subclavian and often carotid bruit in a young person may suggest a diagnosis of aortoarteritis.

**Investigations and Management**

The investigation of patients with TVL due to possible retinal embolism is primarily directed toward identifying an embolic source. Doppler ultrasound, computed tomography (CT) angio and magnetic resonance angiography(MRA) are useful for identifying ICA stenosis, but catheter angiography remains the gold-standard technique for quantifying the degree of stenosis. Trans-thoracic echocardiography is required to identify structural cardiac abnormalities associated with intramural thrombus formation and systemic or paradoxical embolism, and may also be used to assess the aortic arch for atheromatous plaque. Often when the clinical suspicion of a cardiac source of embolism is high transesophagel echocardiography is very useful especially for evaluation of the left atrial appendage. Prolonged ambulatory electrocardiographic(holter) monitoring may be needed to diagnose paroxysmal arrhythmias like intermittent AF. Investigations should be directed towards modifiable vascular risk factors, such as hypertension, diabetes, and dyslipidemia, or other unusual causes of retinal emboli.

Management is directed toward the underlying cause. In patients with a cardiac source of embolism, anticoagulation with warfarin and treatment of the underlying cardiac disease is indicated, whereas those with atherothrombotic disease should be commenced on antiplatelet therapy. Modifiable vascular risk factors should be addressed.

In those with ICA stenosis, antiplatelet therapy with aspirin and clopidogrel should be initiated and modifiable vascular risk factors addressed(statins). Management of high-grade ICA (70% to 99%) stenosis in patients with isolated TVL remains controversial. Carotid stenting with distal protection device during the procedure might produce an equivalent long-term outcome as carotid endarterectomy and may be a safer option for poor surgical candidates.

As TVL may be a warning symptom for impending anterior ischemic optic neuropathy in patients with giant cell arteritis and the patient may be otherwise asymptomatic, an erythrocyte sedimentation rate, C-reactive protein, platelet count, and fibrinogen level should be obtained urgently in all older patients with TVL. The diagnosis is confirmed by finding the characteristic histopathologic changes on temporal artery biopsy. The patient should be admitted for bed rest, as this may decrease the risk of infarction due to posturally-induced hypoperfusion, and commenced on high-dose intravenous steroids as soon as the diagnosis is suspected. One should not wait for the biopsy report to start steroids.

**Common causes for systemic Hypoperfusion**

include vasovagal attacks, cardiac arrhythmias, valvular heart disease (eg, aortic stenosis), and orthostatic hypotension. Investigations and subsequent management are directed toward the underlying cause.

For patients with migraine an evaluation for alternative diagnoses, such as vertebrobasilar ischemia, is indicated, especially in older patients
or those with vascular risk factors. The treatment approach for migraine involves avoidance of precipitating factors and the use of abortive treatments, such as the triptans, at the onset of attacks. Patients with frequent or disabling attacks may respond to prophylactic therapies, such as propranolol, amitriptyline, or topiramate. Attacks of transient monocular visual loss due to vasospasm may remit with calcium channel blockers.

Occipital seizures can usually be adequately controlled with anticonvulsants.

In patients with papilledema neuroimaging should be obtained urgently to exclude a structural cause, such as a mass lesion, obstructive hydrocephalus, or venous sinus thrombosis. Patients with normal imaging should undergo lumbar puncture for measurement of opening pressure and cerebrospinal fluid analysis. Treatment is directed towards the underlying cause.

Optic disc drusen may be visible on funduscopy. B-scan ultrasonography or CT may be required to demonstrate their presence. Rarely, episodes of TVL can be harbinger for central retinal artery occlusion in these patients. No treatment is available.

Uhthoff’s phenomenon causing TVL is self limiting with good visual recovery. No specific treatment is required.

**Summary**

TVL is a sudden onset, temporary, monocular or binocular loss of vision that is often caused by a temporary disruption of blood supply to the eye or visual pathways. It can also result from a wide variety of other causes, including neurologic conditions, such as migraine, and ophthalmic conditions, such as optic disc edema. Diagnostic investigations should be tailored toward the suspected cause. As some causes of TVL, such as TIAs and GCA are true medical emergencies, the clinician should have a low threshold for urgent evaluation of patients who may have these conditions.

**References**


