Amaurosis fugax (transient monocular or binocular vision loss)

Syndee Givre, MD, PhD Gregory P Van Stavern, MD *The next version of UpToDate (15.3) will be released in October 2007.*

INTRODUCTION AND DEFINITIONS — Amaurosis fugax (from the Greek "amaurosis," meaning **dark**, and the Latin "fugax," meaning **fleeting**) refers to a transient loss of vision in one or both eyes. Varied use of common terminology may cause some confusion when reading the literature. Some suggest that "amaurosis fugax" implies a vascular cause for the visual loss, but the term continues to be used when describing visual loss from any origin and involving one or both eyes. The term "transient monocular blindness" is also often used but is not ideal, since most patients do not experience complete loss of vision with the episode. "Transient monocular visual loss" (TMVL) and "transient binocular visual loss" (TBVL) are preferred to describe abrupt and temporary loss of vision in one or both eyes, since they carry no connotation regarding etiology.

Transient visual loss, either monocular or binocular, reflects a heterogeneous group of disorders, some relatively benign and others with grave neurologic or ophthalmologic implications. The task of the clinician is to use the history and examination to localize the problem to a region in the visual pathways, identify potential etiologies, and, when indicated, perform a focused battery of laboratory tests to confirm or exclude certain causes. Therapeutic interventions and prognostic implications are specific to the underlying cause.

This topic discusses transient visual loss. Other ocular and cerebral ischemic syndromes are discussed separately.

APPROACH TO TRANSIENT VISUAL LOSS — By definition, patients with transient visual loss almost always present after the episode has resolved; hence, the neurologic and ophthalmologic examination is usually normal. Reliance is placed on the patient's description of the nature of the visual symptoms and associated features (show table 1). Pertinent medical and family history may also provide valuable clues to the underlying diagnosis.

Few case series of patients with transient visual loss are reported. Details from the Framingham cohort provide some interesting insights into the challenges of evaluating this symptom. Between 1971 and 1989, participants were systematically questioned regarding specific symptoms of transient ischemic attack (TIA) and stroke; 186 of 2110 subjects reported onset of a sudden visual deficit (not necessarily transient) [4]. Follow-up evaluation determined the underlying cause to be stroke or TIA (24 percent), ocular disease (17 percent), transient monocular blindness (10 percent), and migraine (14 percent). The cause remained unknown in 22 percent, and a miscellany of etiologies comprised the remaining 12 percent.

Historical features — Important historical features include whether the visual loss affected one or both eyes, the duration of the episode, and a specific description of the symptoms.

Monocular versus binocular — It is important, but often difficult, to establish whether the visual loss was monocular or binocular. Transient monocular visual loss (**TMVL**) implies a disorder anterior to the optic chiasm (ie, the eye or the optic nerve); possibilities include ocular disease as well as ischemia due to ipsilateral carotid artery disease. Transient binocular visual loss (TBVL) suggests a more posterior process, involving the optic chiasm, tracts, or radiations, or the visual cortex.

Patients with homonymous visual field defects often report monocular visual loss and attribute it to the eye with the temporal field cut. The patient should be asked specifically whether each eye was alternately covered during the attack. Impairment of reading suggests that the visual loss was binocular rather than monocular [3]. In practice, many patients are not able to state definitively whether the episode affected one or both eyes. When the history is unclear, the clinician should assume that either a monocular or binocular etiology is possible.

Duration — Transient visual obscurations due to **papilledema** typically last **seconds**. **Thromboembolic** events from carotid disease or elsewhere generally last 1 to 15 **minutes** and only rarely an hour or more. **Migraine aura** typically lasts 10 to 30 minutes.

Description of visual symptoms — Transient visual loss (TVL) from any cause can be described as mild blurring or fogging to complete blackness, and may involve a part of or all of the visual field. As an example, patients with ocular ischemia may lose the upper or lower half of vision, the temporal or nasal aspect of vision, small areas of vision centrally or paracentrally, and the entire visual field. TMVL descending over the field of vision (like a curtain or shade) or, less commonly, ascending from below, is highly suggestive of retinal ischemia.

Positive visual phenomena, such as photopsias or scintillations that march across the visual field, suggest migraine as the most likely diagnosis. Seizures affecting visual cortex often involve positive visual symptoms, but are typically maximal at onset and lack the evolution or build-up characteristic of migraine. While the visual symptoms produced by retinal ischemia are usually "negative" (eg, shaded, black, blurred), positive symptoms are also known to occur in this setting. Similarly, migraine can produce visual loss without scintillations. Patients who experience intermittent hyphema (hemorrhage within the anterior chamber) may remark that the visual field appears red (erythropsia) during the episodes.

Precipitating factors — Severe carotid occlusive disease has been associated with dimming of vision in one eye after exposure to bright light (retinal claudication). Other retinal diseases (eg, photoreceptor dystrophy, macular degeneration) are also associated with transient worsening of vision in bright light. The mechanism is believed to be related to an ischemia-induced delay in regenerating visual pigments in the photoreceptors.

Episodes of TMVL due to carotid stenosis may be induced by changes in posture or neck position that temporarily reduce blood flow through the artery. Postprandial TMVL has been described in severe carotid narrowing, presumably as a vascular steal phenomenon.

TMVL occurring when the eye is moved into certain positions of gaze (gaze-evoked amaurosis) is suggestive of an orbital mass and is thought to be due to interruption of blood flow to the retina, choroid, or optic nerve.

Loss of vision after exercise or a hot shower (Uhthoff's symptom) is characteristic of demyelinating disease of the optic nerve. Exercise has also been reported to induce retinal vasospasm.

The transient visual obscurations of papilledema are sometimes brought on by postural change, exercise, or straining.

Recurrence — Patients with TVL from vasospasm, migraine, or ischemia may have multiple episodes separated by minutes or years. In one study, a high number of recurrent episodes (>10) of TMVL was more likely to be associated with complete rather than incomplete or absent occlusion of the ipsilateral internal carotid artery [5]. However, multiple, stereotypic episodes of TMVL recurring over many years in the absence of stroke or permanent visual loss would suggest a more benign etiology such as migraine or vasospasm.

Associated symptoms — TVL due to thromboembolic disease is painless in the vast majority of patients. The association of TVL with headache or periocular pain should alert the examiner to check for other causes such as migraine, angle closure glaucoma, and giant cell arteritis.

The association of TVL with transient contralateral hemiplegia is strongly suggestive of severe carotid occlusive disease. Vertebrobasilar symptoms (vertigo, dysarthria, diplopia) should suggest posterior circulation ischemia. Visual loss in the latter setting is binocular, but patients may misreport a transient hemianopia as monocular vision loss.

Medical history — Older age, a medical history of diabetes mellitus, hypertension, or hyperlipidemia all suggest atherosclerotic vascular disease. Known cardiac disease might indicate a cardioembolic event. A history of rash and previous miscarriage is a feature of the antiphospholipid antibody syndrome, which may uncommonly cause TVL. A family history of stroke at a young age or unexplained thrombosis might prompt evaluation for an underlying hereditary hypercoagulable disorder. A history of typical migraine headaches in the patient or family member suggests migraine as a cause.

Examination — The examination should include testing of visual acuity and visual fields, and examination of the optic fundus. The techniques employed are discussed in detail elsewhere.

Fundoscopic examination should include careful observation of the optic disc, retina, and blood vessels for clues as to the origin of the visual symptoms:

- Disc swelling in the setting of TMVL suggests an acute event involving the optic nerve, usually ischemia or inflammation. Because optic disc pallor does not develop until four to six weeks after an injury, its presence suggests a subacute or chronic process, rather than an acute event. Bilateral disc swelling with good visual acuity should prompt an evaluation for papilledema.

- Retinal whitening often reflects ischemia of the inner retina, and may be seen after a branch or central retinal artery occlusion (show picture 1). Mid-peripheral hemorrhages, tortuous veins, and neovascularization reflect reduced retinal arterial flow and venous stasis as the result of severe carotid stenosis.

- Examination of the arterioles should focus on caliber as well as the presence of embolic plaques. Investigators have attempted to identify the origin of retinal plaques by their appearance, but in practice the distinction is often difficult. A Hollenhorst plaque typically appears at vascular bifurcations, and has a highly refractile appearance (show picture 2). It strongly suggests ipsilateral carotid artery disease, but these cholesterol emboli may also arise from a more proximal source, such as the aortic arch. Platelet-fibrin and calcific emboli are less commonly seen.

A detailed eye examination usually requires an **ophthalmology consult**, which is indicated in many cases of TVL.

CAUSES OF TRANSIENT MONOCULAR VISUAL LOSS — Ischemia is the most common cause of transient monocular visual loss (TMVL); this in itself can have diverse etiologies. Papilledema, optic neuropathy, and ocular disease are other causes of TMVL.

Ischemia — The pathogenesis and pathophysiology for TMVL of ischemic origin are diverse, and include large artery occlusive disease (atherothrombosis, embolus, dissection), small artery occlusive disease (anterior ischemic optic neuropathy, vasculitis), venous disease, cardiac disease, hypercoagulable disorders, and systemic hypoperfusion. The final common pathway for these disorders is ischemia to the retina, the optic nerve, or both. TMVL secondary to retinal ischemia from carotid disease may be due to either embolism or hypoperfusion from fixed arterial stenosis, whereas optic nerve ischemia usually results from the latter mechanism.

Most patients with TMVL attributable to ischemia will report areas of blank or fuzzy vision ("negative visual phenomena"). Less common, but not unusual, are reports of positive visual phenomena. Symptoms generally begin abruptly and reach maximum severity within the first few minutes.

Funduscopic examination may be normal or may show evidence of vascular disease: cotton wool spots, retinal emboli, retinal hemorrhages, retinal whitening, or dilated retinal veins (show picture 1 and show picture 2).

Retinal embolism is frequently asymptomatic. In a population-based study of approximately 2000 older adults (≥49 years) whose retinas were examined at five and ten years, there was a baseline prevalence of retinal emboli of 1.4 percent. The cumulative 10-year incidence was 3 percent. Over 10 years follow-up, all-cause and stroke-related mortality was somewhat higher in patients with documented retinal emboli (HR = 1.3; CI 1.0 to 1.8).

Carotid artery disease — Ischemic visual loss related to occlusive disease in the cervical carotid artery may result from thromboembolism or hypoperfusion. The former mechanism is believed to be more common.

In contrast to transient ischemic attacks (TIAs) involving the cerebral hemispheres, retinal ischemia is more commonly associated with emboli originating from carotid stenosis rather than heart disease. This may represent a streaming effect of laminar blood flow, whereby particulates of uniform size are consistently deposited into one lamina, and swept into the same distal vascular bed. Small emboli from carotid disease may be more likely to drift to the edge of the bloodstream and enter the ophthalmic artery. Also, these small particles may be more prone to obstruct the smaller retinal arterioles.

Severe stenosis (90 to 100 percent) of the ipsilateral carotid artery may also cause visual loss due to retinal and/or choroidal hypoperfusion from low limiting stenosis. This ischemic mechanism may be more likely to include positive visual phenomena and recurrent episodes than thromboembolism, and it is often induced by activities that reduce ocular perfusion pressure (such as postural change, postprandial vascular steal) or

increase retinal oxygen demand (such as exposure to bright light). In contrast, episodes of TMVL due to carotid artery thromboembolism usually occur spontaneously, and are more often isolated events, although they can recur [7].

Visual loss caused by either mechanism typically encompasses the entire visual field, but may affect only the upper, lower, nasal, or temporal portion. Small central or paracentral scotomas can also occur. The classic description of a "curtain" or "shade" descending over vision, is relatively uncommon; in one large study, only 23.8 percent of patients with TMVL experienced altitudinal visual loss. Nonetheless, when this is described, it is highly suggestive of retinal embolism. Patients may also experience gradual graying of vision that progresses from the periphery to the center. The visual loss may be associated with scintillations or the sensation of color. Episodes generally last from seconds to minutes, rarely more than 15 minutes.

When contralateral hemispheric symptoms (motor, sensory, aphasia) accompany visual loss, carotid disease is very likely; however, TMVL is more often an isolated symptom.

In a prospective study of 337 patients with TMVL, clinical features most predictive of high-grade (>70 percent) carotid stenosis included rapid onset of symptoms, altitudinal pattern of onset or resolution, and a duration of 1 to 10 minutes.

The diagnosis of carotid disease in the setting of TMVL is important because of the implications for stroke risk, approximately 2 to 3 percent per year. Most of these strokes are large-vessel infarcts within the territory of the ipsilateral carotid artery. Compared with cerebral ischemia, TMVL carries a lower risk for stroke in the setting of high-grade carotid disease; but still identifies a high-risk population for which carotid endarterectomy may be beneficial, particularly in high-risk subgroups.

Giant cell arteritis — TMVL, similar in character to that associated with carotid disease, can occur in patients with giant cell arteritis (GCA). Occlusion or near occlusion of the posterior ciliary arteries by granulomatous inflammation within the artery wall can cause an anterior ischemic optic neuropathy. Posterior ischemic optic neuropathy and central retinal artery occlusion are also manifestations of GCA.

GCA most often presents as acute sustained unilateral visual loss with a swollen optic disc [34]. In two large series (each >150 patients) of biopsy-confirmed GCA, TVL was the presenting symptom in just 10 to 15 percent. It was bilateral in 27 to 35 percent of these. Common accompanying symptoms include jaw claudication and headache (each in about half of patients). However, patients with occult giant cell arteritis may have no other symptoms.

While TVL is a relatively uncommon presentation for GCA, it represents an opportunity for intervention to prevent permanent visual loss.

Other ischemic etiologies

- Cardiogenic embolism — Emboli from the heart and great vessels may lodge in the ophthalmic circulation and cause TMVL, which is clinically similar to that associated with carotid disease. Cardiogenic embolism is less likely to underlie TMVL than a cerebral TIA.

  While an uncommon cause of TMVL, cardiogenic embolism is important to identify in this setting, because anticoagulation is often recommended to prevent more serious sequelae. A detailed review of cardiogenic sources of emboli is presented elsewhere.

- Hypotension — Hypoperfusion from cardiac failure and hypovolemia, reduced cardiac output from a dysrhythmia, and orthostatic hypotension may cause TVL. Anemia and intradialysis hypotension have also been implicated. In this setting, visual loss is usually binocular and associated with other suggestive features, such as postural change, lightheadedness, and presyncope. However, orthostatic or systemic hypotension can result in hypoperfusion to one eye, usually in association with carotid occlusive disease, which results in selective hypoperfusion to the ipsilateral eye. TMVL may also be precipitated by hypotension in the setting of underlying retinopathy, optic neuropathy, or glaucoma.

- Coagulopathy — Patients with coagulation disorders may have episodes of TMVL, with characteristics similar to those caused by atheromatous disease. Patients with coagulopathies usually have other symptoms in addition to TMVL (eg, rash associated with the antiphospholipid antibody syndrome) and/or

a medical history of other coagulation-related problems. However, there have been reports of patients with TMVL as the presenting symptom of a coagulation disorder.

- Others — Nonarteritic anterior ischemic optic neuropathy is more common than giant cell arteritis and is associated with small artery disease. However, it is not typically preceded by transient episodes of visual loss, and therefore is not usually considered in the differential diagnosis of TMVL.

Atherosclerosis involving the intracranial carotid or the ophthalmic artery may also produce TMVL [47]. TMVL also occurs in nonatheromatous disease of the carotid, such as fibromuscular dysplasia, aneurysm, dissection, and arteritis, such as Wegener's granulomatosis and Takayasu's arteritis [25,48]. Cocaine and other illicit drugs have been associated with TMVL through vasospasm or other mechanisms.

Retinal vein occlusion — TMVL may occur as a premonitory symptom of central retinal vein occlusion. Occlusion or thrombosis of the central retinal vein is associated with chronic glaucoma, atherosclerotic risk factors (age, diabetes, hypertension), hyperviscosity, and coagulopathy. The cause of retinal vein occlusion is often unknown.

In established retinal vein occlusion, the disc may be congested. Retinal hemorrhages extend beyond the posterior pole of the eye into the retinal periphery (show picture 3).

Episodes of visual loss are quite variable in length and may last several hours. Another potentially distinguishing feature is the description of "cloudy vision" rather than visual loss; however, episodes may be entirely similar to those occurring in arterial ischemia [53,55]. Treatment interventions for fixed visual loss associated with central retinal vein occlusion are often attempted, but are not of proven benefit.

Retinal vasospasm and retinal migraine — Idiopathic, reversible vasospasm affecting blood vessels in the retina and causing TMVL is a commonly described, but poorly understood and somewhat controversial phenomenon. Reported observations include:

- Studies of children, adolescents, and young adults with TMVL have shown that a majority have headaches either associated with or between episodes, a personal or family history of migraines, and a benign course

- Transient arterial narrowing has been observed in retinal arteries in symptomatic patients

- Some patients' symptoms respond to calcium channel blockers

Some of these observations have led to this entity being called retinal migraine. Retinal migraine is a diagnosis included in the International Headache Society Criteria [64], but is a debatable entity, and is always considered a diagnosis of exclusion. The formal criteria require reversible positive or negative visual phenomena accompanied or followed within 60 minutes by migraine headache. A personal or family history of migraine may suggest the diagnosis in the absence of headache. In general, migraine as a cause of TVL is best accepted as a manifestation of occipital cortex involvement, producing binocular, usually positive, visual symptoms, the "classic" or typical migraine aura. The etiology in this case is presumed to be a cortical spreading depression and not vasospasm.

Important caveats in the diagnosis of retinal vasospasm are that not all arterial narrowing is vasospasm; proximal occlusive disease and low blood flow may also give this appearance. Also, vasospasm may occur secondary to an underlying disease, such as vasculitis, migraine, hypercoagulability, or cocaine use.

Symptoms described in retinal vasospasm include characteristic positive phenomena of scintillations and photopsias as well as primarily negative symptoms—black, gray, white, or shaded areas, which are indistinguishable from ischemic symptoms. Patients may have several episodes in one day. A typical episode lasts longer than five minutes and less than 60 minutes. A first episode usually occurs in individuals less than 40 years old.

Optic neuropathy — Patients with a chronic optic neuropathy will occasionally complain of episodes of transient loss or blurring of vision, usually in association with elevation of body temperature (hot shower, exercise). This phenomenon is termed Uhthoff's symptom and reflects temporary conduction block along a previously demyelinated optic nerve [67,68].

The episodes typically last several minutes, but occasionally longer, until the body temperature returns to normal. Uhthoff's phenomenon is classically associated with multiple sclerosis, but has been reported in other optic neuropathies.

Papilledema — While rarely the presenting symptom, transient visual obscurations are common in patients with papilledema. These are described as a "whiting" or "graying out" of part or all of the visual field; they are often unilateral, and typically very brief (just seconds). They may occur spontaneously or with changes in position, and they are believed to represent transient fluctuations in nerve head perfusion. Other symptoms and signs (eg, headache, bilateral disc swelling) are usually present to suggest this diagnosis.

Optic nerve compression — Gaze-evoked amaurosis has been described in association with compressive optic neuropathies from tumors, trauma, Graves' ophthalmopathy, and other causes. In cases due to slow growing masses, the examination will often reflect ipsilateral optic nerve dysfunction.

Ocular causes — Ocular causes of TMVL include:

- Increased intraocular pressure such as from intermittent angle closure glaucoma is typically associated with eye pain. A complaint of halos around lights is common in this condition.

- Spontaneous hyphema (hemorrhage within the anterior chamber) can occur after cataract surgery with lens implantation. The symptoms of the hyphema may be visual loss and erythropsia.

- Vitreous floaters occasionally obscure central vision.

- Congenital optic disc anomalies and optic disc drusen have been associated with TMVL.

Clues to an ocular cause might include:

- Eye redness
- Pain and tearing associated with visual loss (intermittent angle closure glaucoma)
- More prolonged episodes [3]
- Relief of symptoms with blinking or rubbing the eye (dry eye)
- Squinting (refractive error)

Idiopathic — Despite extensive workups, a cause of TMVL will not be found in some patients. These patients are at very low risk for either permanent visual loss or stroke and should be reassured.

CAUSES OF TRANSIENT BINOCULAR VISUAL LOSS — Transient binocular visual loss (TBVL) implies a process posterior to the optic chiasm, and may include migraine, seizure, and vertebrobasilar ischemia. Typical clinical features are summarized in the table (show table 1). In rare cases, TBVL may reflect bilateral, simultaneous involvement of the anterior visual pathways such as might occur with giant cell arteritis and systemic hypotension, as discussed above. Papilledema can produce unilateral or bilateral transient visual obscurations, also discussed above.

Migraine — Migraine is the most common cause of TBVL in young adults. Positive visual phenomena such as scintillations suggest migraine as the most likely diagnosis, especially if there is a history of headaches with migrainous features. This classic or typical migraine aura typically lasts 20 to 30 minutes, rarely as long as an hour, and has a characteristic build-up, or evolution, a feature lacking in other causes of TBVL (ischemia, seizure) [76]. The mechanism of visual loss in migraine is thought to be neuronal depression after a period of cortical excitation ("spreading depression of Leao").

Migraine visual aura may occur independently of headache (sometimes called migraine equivalent or acephalgic migraine) and may be confused with ischemia, especially in older patients [13]. In a Danish study, 38 percent of 163 patients reported having attacks of migraine aura both with and without headache; 4 percent had migraine aura that never occurred with headache [78]. The diagnosis of acephalgic migraine is supported by positive, binocular visual symptoms, a somewhat longer duration of episodes than is typical for ischemia, and a personal or strong family history of migraine [13].

Data from the Framingham cohort suggest that migrainous visual aura is not rare, even in older adults. In that study, 1.2 percent of individuals aged 50 to 82 years reported visual symptoms, subsequently attributed to migraine. In most (77 percent) of these 26 patients, the onset of these symptoms occurred after age 50 years; most episodes were never associated with headache (58 percent), and 42 percent had no history of recurrent headaches. Three strokes occurred subsequently in these 26 patients; this 11.5 percent incidence was lower than the 33 percent incidence among patients with transient ischemic attack (TIA).

Seizure — Visual seizure is an uncommon cause of TBVL. The visual loss may be ictal or postictal.

Among 20 patients in one series of epileptic visual aura, symptoms included elementary hallucinations (eg, flickering lights, stars, flashes of light) and visual loss, which was described as blurred vision, "white out," or scotomata. The visual loss and hallucinations could start in a specific part of the visual field, could move or be stationary, or could involve the entire visual field. These symptoms were not specifically localizing; underlying lesions (present in all) were found in either the occipital or temporal lobes. Ictal blindness may be an isolated epileptic phenomenon, but is more usually accompanied by other manifestations, such as tonic eye deviation, altered consciousness, or motor impairment. Prolonged ictal blindness ("status epilepticus amauroticus") is rarely reported.

Postictal blindness typically lasts minutes to hours, but may last days or weeks. As with Todd's paralysis (postictal weakness following a motor seizure), the mechanism is presumed to be prolonged enhancement of inhibitory stimuli in response to prolonged excitation (similar in some ways to migraine).

Adult onset visual seizures may be associated with cortical lesions in the posterior visual pathways, which produce a corresponding visual field defect. Therefore, visual field testing (preferably using static or kinetic perimetry) is important in all patients with TBVL.

Vertebrobasilar ischemia — Ischemia to the visual cortex may result in TBVL. If only one hemisphere is ischemic, the patient will experience homonymous visual field loss contralateral to the lesion. Visual loss may be an isolated symptom or may be accompanied by symptoms of brainstem ischemia (dysarthria, dysphagia, vertigo, diplopia) or cerebral ischemia (hemiparesis, hemisensory loss, aphasia). Isolated visual symptoms are usually the result of occipital lobe ischemia secondary to posterior cerebral artery occlusion. Difficulty seeing to one side is the most common symptom; lateralized flashing lights (photopsias) are also common.

As with anterior circulation ischemia, the mechanism of strokes in the posterior circulation may involve thrombosis, embolism, or hypoperfusion from fixed arterial stenosis. The most common identified mechanism underlying posterior cerebral artery territory infarcts is embolism, usually from cardiac disease. Artery to artery embolism, usually originating from proximal disease in the vertebral arteries, accounts for the second highest percentage. Atherostenosis in the posterior cerebral arteries has also been described.

DIAGNOSTIC EVALUATION OF TRANSIENT VISUAL LOSS — The diagnostic workup is tailored to the likely responsible conditions as suggested by the history and physical examination (show table 1). The overlap in clinical presentations and grave prognosis of some potential diagnoses require that some diagnostic testing is performed in most patients.

- Ophthalmologic evaluation — A detailed funduscopic evaluation is an important part of the evaluation of patients with transient visual loss. Ophthalmology referral is required for all patients with suspected giant cell arteritis, retinal vein disease, and ocular causes of visual loss, and will often reveal evidence of an otherwise unsuspected ischemic mechanism in patients with recurrent episodes of TMVL.

- Erythrocyte sedimentation rate and C-reactive protein - All older patients (>50 years) with transient monocular or binocular vision loss should have a sedimentation rate and C-reactive protein to exclude giant cell arteritis (GCA). If these are elevated, or if the history is very suggestive, patients should proceed to a confirmatory temporal artery biopsy. Treatment should not await pathology results.

- Carotid imaging — Carotid duplex ultrasound, magnetic resonance angiography, or computed tomographic angiography should be ordered in all older patients (>50 years) and in younger patients with vascular risk factors (diabetes, hypertension, hyperlipidemia) who have experienced TMVL. Magnetic resonance imaging (MRI) should be performed in a patient of any age with TMVL who has a history suggestive of carotid artery dissection.

Further evaluation and treatment of carotid disease in the setting of TMVL is discussed elsewhere.

- Cardiac evaluation — Once GCA and carotid disease have been excluded, an evaluation to seek a cardiogenic source of embolism should follow in all older patients (as well as in younger patients with risk factors) who have had TMVL. This is also indicated in patients with TBVL due to posterior circulation ischemia. Testing may include Holter monitoring and echocardiography. The details of this evaluation are discussed elsewhere.

A baseline **ECG** should also be included in the evaluation of these patients as cardiac morbidity and mortality in patients with TMVL and central retinal artery occlusion is significant.

- Brain **MRI** — Older patients with binocular visual symptoms (TBVL) that accompany symptoms suggestive of vertebrobasilar ischemia, should have a brain MRI. Diffusion-weighted imaging (DWI) increases the yield of abnormalities in patients with TIA, and intracranial and cervical magnetic resonance angiography (MRA) may turn up occlusive disease of the posterior circulation.

Patients whose history or examination suggests optic neuropathy should have an MRI to look for other evidence of demyelinating disease. A history suggestive of compressive optic neuropathy (gaze evoked amaurosis) or seizure should also prompt a brain MRI with contrast enhancement.

- **EEG** — Electroencephalography (EEG) is not a routine test for TVL, but should be undertaken in a patient with TBVL whose symptoms suggest possible seizure. EEG monitoring may increase the diagnostic yield, especially in patients with frequently recurring symptoms.

- **Hypercoagulable testing** — When brain or ocular ischemia is the suspected cause of TVL, hypercoagulable testing should be performed in individuals who have suggestive histories (prior thrombosis, miscarriage, or family history), as well as in individuals with probable ischemia and otherwise negative workup. A complete blood count should also be obtained to screen for conditions such as polycythemia vera and essential thrombocythemia.

Young patients without risk factors — Patients in whom there is a strong presumptive diagnosis of migraine may have a somewhat limited workup or perhaps no testing at all. An example might be a young person with typical binocular visual symptoms that include positive phenomena and are followed by migraine headache. Other factors that support (but obviously do not confirm) a diagnosis of migraine include a family history of migraine and a past history of migraine headaches. The more the symptoms deviate from classic migraine, the more extensive the workup should be.

In the majority of young patients (<50 years) with TMVL who are otherwise healthy, the diagnostic yield of extensive testing is very low. The most reasonable presumed cause is vasospasm or migraine, although this remains speculative. Their future risk of stroke is low. However, TMVL due to carotid disease and cardiac disease does occur in young people. The extent to which these patients should be subjected to diagnostic investigation is debated, and should be decided individually. A conservative approach recommends performing noninvasive studies that identify high-risk pathology. This might include MRI, carotid imaging, ECG, echocardiography, and selective clotting studies.

SUMMARY AND RECOMMENDATIONS — Underlying causes of transient visual loss range from benign conditions to those with serious neurologic and ophthalmologic sequelae.

- With transient symptoms, the diagnosis is guided by historical features. The most important of these is the distinction between monocular and binocular involvement; however, this is often unclear. Other helpful features include symptom duration, description of symptom onset, the presence of positive visual phenomena, precipitating factors, and associated symptoms (show table 1).

- Ischemia originating from carotid artery disease is a common cause of transient monocular visual loss (TMVL), and has serious implications for future stroke risk that can be reduced by carotid endarterectomy. Suggestive features include rapid onset in an altitudinal pattern (visual loss descending over the field of vision like a curtain or shade) and brief duration (<10 minutes).

- Giant cell arteritis is not a common cause of TMVL but is a treatable cause of otherwise permanent visual loss. It is clinically indistinguisable from TMVL of carotid disease except for frequent accompanying headache.

- Other causes of TMVL include ischemia from cardioembolic disease, some ocular conditions, and others.

- TMVL in young patients may be due to retinal vasospasm or may be idiopathic, but other etiologies must be excluded.

- Migraine typically produces binocular symptoms with positive phenomenology and a characteristic build-up, lasting 10 to 30 minutes, and associated with or followed by migraine headache. (

- Binocular visual loss, usually isolated to one visual field, can also occur during vertebrobasilar ischemia or seizure.

- Unless symptoms are very typical for migraine, some diagnostic testing is required. Erythrocyte sedimentation rate and C-reactive protein should be performed in all individuals over age 50 years with transient monocular or binocular visual loss. We recommend ophthalmologic examination and carotid duplex ultrasound for all patients with TMVL. Other tests are ordered according to symptoms and clinical setting and may include cardiac workup, hypercoagulable testing, magnetic resonance imaging (MRI), and electroencephalogram (EEG) (show table 2).