CHAPTER 7

Visual System Disorders

DIAGNOSTIC ASSESSMENT

The patient's complaints are usually caused by two types of abnormalities: defective visual perceptions and abnormal ocular movements. To analyze these visual disturbances, attention must be directed toward seven important points (Box 7-1). With these findings, it is usually possible to localize the lesion within the visual system.

SIGNS AND SYMPTOMS OF DISEASE IN THE AFFERENT VISUAL SYSTEM

Symptoms

Patients are frequently aware of bilateral visual acuity impairment, but if acuity loss is unilateral, it may not be detected early and only when patients close the nonaffected eye. Patients may complain of visual blurring or visual dimming. Ask the patient, "What did you initially notice was wrong with your vision?"

BOX 7-1

1. Precise delineation of visual symptoms including accompanying facial, orbital, and cephalic sensory (pain) symptoms or other neurological complaints
2. Measurement of best corrected visual acuity (use Snellen chart at distance of 20 ft. or Rosenbaum pocket vision screener held 14 in. from the patient)
3. Visual field examination
4. Analysis of pupillary size and reactivity
5. Examination of optic fundus
6. Eye movement analysis
7. Detection of associated neurologic findings

notice visual symptoms?" and "Has there been subsequent improvement or deterioration in your vision?" Determine exacerbating factors: one, exercise (Uhthoff's phenomenon in which visual dysfunction is worsened by exercise in patients with optic nerve demyelination) and elevated blood temperature, which also worsens vision in multiple sclerosis patients; and two, medication administration as it relates to visual symptoms (digitalis or ethambutol can cause visual disturbances).

Sudden unilateral visual loss is caused by retinal or optic nerve lesions. These include vascular conditions (retinal artery occlusion, temporal arteritis with optic nerve head infarction), demyelinating disorders (optic neuritis), or primary ophthalmologic disorders (detached retina). Amaurosis fugax is temporary unilateral loss of vision caused by a microembolism originating from the carotid artery. This can be accompanied by other signs of transient ischemia caused by ipsilateral cerebral hemisphere, for example, contralateral weakness, sensory disturbance, aphasia. Amaurosis fugax can be a harbinger of carotid artery or aortic arch disease (high-grade stenosis, occlusion) due to an embolus consisting of cholesterol (Hollenhorst plaque) material that occludes a retinal arteriole. If the embolus degenerates, flow is restored and vision normalizes. If there is prolonged retinal artery occlusion, retinal artery infarction with permanent visual loss may occur. Other causes of sudden unilateral visual loss include anterior ischemic optic neuropathy (AION), posterior ischemic optic neuropathy, and optic neuropathy. In CRAO, unilateral visual loss occurs rapidly. Findings include the pupil not reacting to light and funduscopy showing ischemic retina appearing pale (white) with normal red-colored choroid reflecting through fovea (cherry-red spot); optic atrophy develops later. In some cases, retinal embolus that is occluding the retinal artery can be visualized by funduscopic examination as it is observed moving from one vessel region to another, usually migrating to a more distal segment. In AION, sudden painless visual loss occurs; this is usually an altitudinal defect (described as if a curtain is pulled over on eye). Signs of AION include: unilateral altitudinal visual field defects or total blindness; impaired pupillary response to light; and funduscopy showing narrowed retinal arteries, hyperemic (swollen) pale disc, flame-shaped hemorrhages, and later optic atrophy. There is reduced perfusion through the posterior ciliary arteries which supply the optic disc. There is nonarteritic form (usually due to hypertension and diabetes) and arteritic form which occurs with temporal arteritis. Posterior ischemic optic neuropathy causes infarction of the retrobulbar optic nerve: therefore optic nerve may appear normal but the visual loss is accompanied by impaired pupillary response. This is caused by postural hypotension. Optic neuritis is a primary inflammatory disease of the optic nerve and may be bulbar (optic nerve appears swollen) or retrobulbar and optic fundus appears normal. Optic neuritis may be initial feature of multiple sclerosis.

Lesions responsible for sudden bilateral visual loss include pituitary apoplexy, cortical blindness caused by bilateral occipital lobe infarcts, and hysteria. Bilateral visual loss is due to bilateral retinal, optic nerves, optic chiasm, optic tract, optic radiation, or occipital lesions. Bilateral cortical (occipital) blindness is usually due to bilateral posterior cerebral or bilateral homonymous hemianopia; however, the patient may not be cognizant of blindness (Anton's syndrome).

Transient monocular visual loss that resolves rapidly within seconds or minutes indicates amaurosis fugax caused by carotid or aortic arch disease or papilledema caused by intracranial hypertension, although the later condition more commonly causes bilateral episodic visual obscurations (Box 7-2). Sudden monocular visual loss that stabilizes and improves during period of several days or weeks is consistent with demyelinating disease (optic neuritis). Visual loss can also develop insidiously. This indicates a compressive optic nerve lesion if unilateral.

Consider metabolic, toxic, or degenerative (Leber's hereditary optic neuropathy) optic nerve lesion if bilateral. These patients may report sensations that a film is developing over one eye or patient can have difficulty focusing when reading. Other visual symptoms of optic nerve dysfunction include dimming of color or light brightness under different sources of illumination (e.g., sunlight, twilight, watching color television).

Box 7-2	Mechanisms of Acute Visual Loss
Transient	
Unilateral	Amaurosis fugax
	Retinal migraine
Bilateral	Systemic hypotension
	Systemic arterial hypertension
	Retinal dysfunction
	Intracranial hypertension
	Systemic hyperviscosity syndromes
Persistent	
Unilateral	Central retinal artery occlusion
Painless	Anterior or posterior ischemic optic neuropathy
	Macular or vitreous hemorrhage
	Retinal detachment
Painful	Glaucoma
	Temporal arteritis
	Optic neuritis
	Iridocyclitis
	Vitreous hemorrhage
Bilateral	
Painless	Bilateral occipital infarction
	Eclampsia
	Hysteria-conversion reaction
Painful	Pituitary apoplexy

Abnormal visual sensations can be initial migraine symptoms. These include flashing lights (scintillation), bright zigzag or picket fence lines (fortification spectrum), or flickering colored lights moving across the visual field. These symptoms can be accompanied by central areas of impaired vision (scotoma); this may be initially noted in center of visual field and extended peripherally within 10 to 30 minute interval. Perception of unformed flashing lights moving rapidly across the visual field within seconds or minutes is characteristic of occipital epilepsy.

In patients with sensory visual symptoms reporting cephalic or orbital pain, consider acute glaucoma, temporal arteritis, retrobulbar neuritis, migraine, or cluster headache; however, orbital pain can indicate a local ophthalmological disorder. In patients with diplopia and ocular motility disorders reporting cephalic or orbital pain, consider a ruptured carotid aneurysm or ischemic extraocular nerve lesion. Other causes of painful ophthalmoplegia include superior orbital fissure or cavernous sinus conditions of neoplastic, inflammatory, or vascular origin.

Examination

Determine patient's best corrected visual acuity at 20 feet; however, this is usually tested with a pocket-sized eye chart held 14 inches from the patient. It may be possible to improve acuity by having the patient view this card through a pinhole if reduced vision is due to refractive changes. Observe patient manipulate card; patients with hemianopic defects may ignore part of visual acuity chart extending into affected visual field. Visual acuity loss not correctable with lenses (refraction) or not attributed to retinal disease, strabismic amblyopia (lazy eye with loss of acuity caused by failure of visual potential to develop because of wandering or crossed eye), lens disease, corneal disease or other primary ophthalmological disease is presumed caused by optic nerve or chiasmal dysfunction. Test color vision using Ishihara color plates. Acquired color vision defects (color desaturation) is early sign of optic nerve dysfunction.

Examination of visual fields is best initially performed using confrontation techniques (Chapter 1). Visual field defects can be delineated by tangent screen examination, Goldmann kinetic perimetry, or automated static threshold perimetry, but these require specialized equipment. Confrontation technique is quite accurate if certain rules are considered. Box 7-3 lists these rules. Characteristic visual field defects are show in Figure 7-1.

Box 7-3

1. Lesions of the optic chiasm and postchiasmal region cause defects that change abruptly as object is moved across vertical meridian from nasal to temporal sectors.
2. Superior and inferior quadrants are separately tested because quadrantanopic defects have more localizing value than hemianopic defects.
3. Monocular visual field defects are due to retinal or optic nerve lesions.
4. Binocular visual field defects developing at the same time are caused by chiasmal and postchiasmal lesions. Bilateral scotomas caused by bilateral optic neuritis can develop at separate times, but it is unlikely that these will develop at exactly the same time.
5. The hallmark of postchiasmal visual lesions is homonymous hemianopsia; chiasmal lesions cause bitemporal hemianopsia.

Pupillary size, shape, and light reactivity must be assessed. Have patient fixate on distant object in dim illumination maximal pupillary size is then measured. Twenty to 30% of normal subjects have clinically observable differences in pupil size; however, light reactivity should be symmetrical in both eyes, and there should be no difference in the degree of anisocoria in different illumination states, such as sunlight versus a dark room. To test pupillary reactivity,

bright light is directed to one eye and rapidly moved to opposite eye. Rapidity and degree of change in pupillary size is determined in both the eye directly illuminated and the opposite eye (consensual) reaction. Response should be equal in both eyes because the Edinger-Westphal nucleus located in midbrain summates the total light received and sends identical pupil diameter message to both eyes.

If afferent (optic nerve) portion of light reflex is defective, response of defective eye is diminished when light is directly applied and normal when light is applied to opposite eye (consensual reaction). Unilateral optic nerve lesions can be detected by rapidly moving light from one eye to the other eye (swinging flashlight test). Abnormal response consists of less brisk pupillary constriction or dilation (pupillary escape) when light is rapidly moved from normal to defective pupil (Marcus Gunn phenomenon). This test is helpful in unilateral--but not bilateral--optic nerve lesions. Despite abnormal response to light stimulation in unilateral optic nerve lesions, the pupils are equal in size in resting state. If efferent portion of the light reflex (oculomotor nerve) is defective, pupils are unequal in size, with abnormal pupil dilated and poorly reactive to both direct and consensual stimulation.

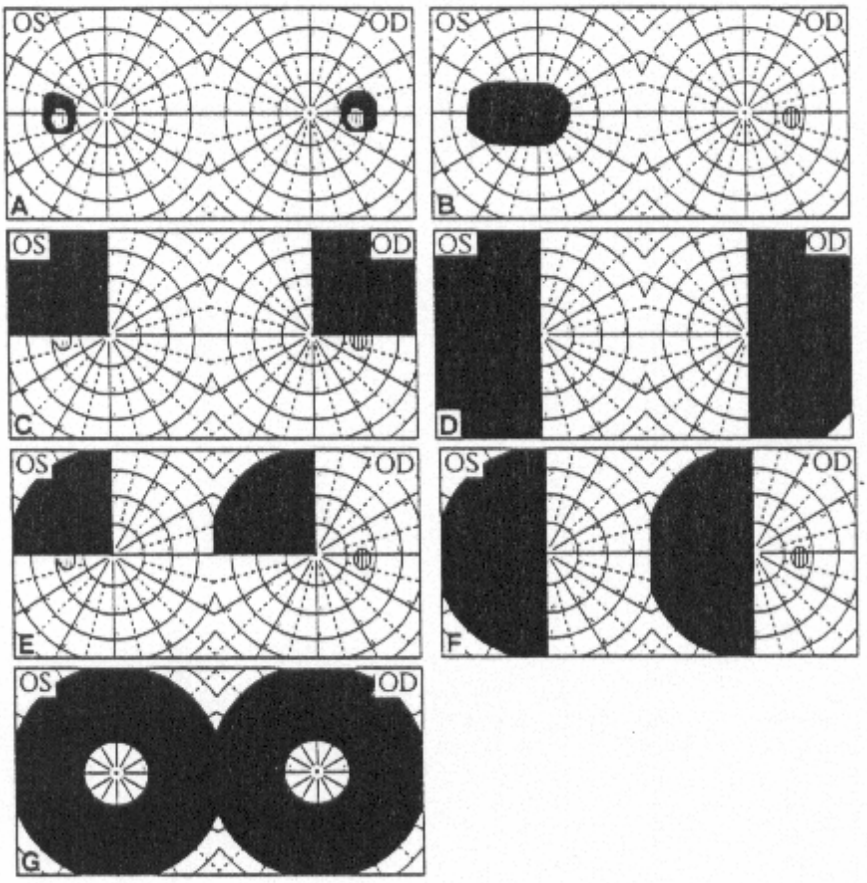

FIGURE 7-1. A, Bilateral enlarged blind spots caused by papilledema. B, Left centrocecal scotoma caused by dysfunction of the left optic nerve in a patient with unilateral optic neuritis. C, Bitemporal quadrantanopia caused by compression of the undersurface of the optic chiasm by a pituitary adenoma. D, Bitemporal hemianopia caused by optic chiasmal compression, which is due to pituitary tumor. E, Left superior homonymous quandrantanopia, which may be caused by a lesion involving the temporal lobe. F, Left homonymous hemianopia, which may be due to a right-sided cerebral lesion. G, Concentric constriction (tubular visual fields) is cylindrical and measures the same size irrespective of the distance of the patient from the visual chart or the size of the test object. This can be seen in hysteria.

Figure 7-1

In patients with optic nerve dysfunction, there is reduced visual acuity, central scotoma (blind or black spot within central field of vision), and afferent pupillary defect. Another feature of optic nerve dysfunction is defective color perception (dyschromatopsia); and decreased appreciation of color or light brightness. Impairment of color vision can be tested by having patient look at colored objects (such as the Ishihara color plates) with each eye independently. In assessing the presence of impaired color vision, remember 10% of men have congenital color blindness. The eye in which color is less rich and less bright is the affected eye. Patients rarely complain of this symptom, and this is demonstrated by visual testing only.

Examination of the retina and optic disc (papilla) is performed at bedside by the neurologist using hand-held ophthalmoscope. The ophthalmologist uses an indirect light-source ophthalmoscope; this has much brighter light source and pupil is fully dilated and prevented from constricting by locally instilled dilating agent. This allows better visualization of a larger portion of the retina (including periphery). Optic disc edema appears as optic nerve swelling. If this is due to increased intracranial pressure, it is referred to as papilledema. When caused by intracranial hypertension, papilledema is present bilaterally; papilledema can be asymptomatic or cause transient visual obscurations. Headaches are usually prominent in intracranial hypertension. If papilledema is unilateral, this indicates local vascular inflammatory or neoplastic orbital lesions; *unilateral* papilledema is rarely due to increased intracranial pressure. The diagnosis of papilledema is established by funduscopic findings (Figure 7-2). Box 7-4 outlines the diagnostic features of papilledema.

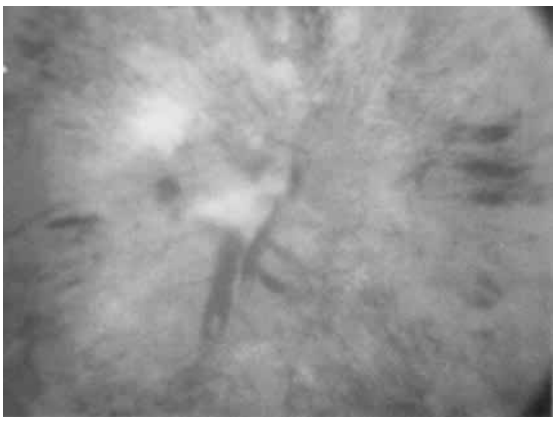

Figure 7-2. Optic disc blurring of disc margins with loss of central cup; consistent with papilledema.

Box 7-4

1. Absence of spontaneous venous pulsations
2. Presence of hemorrhages and exudates located contiguous to optic disc region
3. Inability to focus on vessels at center and periphery of disc without changing diopter settings; this is due to central disc elevation
4. Blurring of nasal disc margins and later extending to temporal margins
5. Loss of central physiologic optic cup; this is usually a late finding

6. Blurring of the retinal nerve fiber layer as seen with green light on the ophthalmoscope
7. Engorged veins and increased disc redness (disc hyperemia)

Spontaneous venous pulsations are present in 80% of normal people. Their visualization usually excludes diagnosis of increased intracranial pressure; however, absence of spontaneous venous pulsations is not helpful as 20% of normal people have no evidence of pulsations despite normal cerebrospinal fluid (CSF) pressure. It may not be possible to differentiate ophthalmoscopic signs of optic disc edema from pseudopapilledema such as optic disc drusen. Fluorescein angiography, three-dimensional fundus photographs, and serial ophthalmoscopic examinations can improve diagnostic accuracy. The presence of intracranial hypertension can only be *definitively* established by finding of elevated opening pressure when lumbar puncture is performed with the patient in lateral recumbent position. Optic disc edema can occur in the absence of elevated intracranial pressure without any neurological signs, and in this case ophthalmologic evaluation for possible vascular or inflammatory causes is warranted.

Funduscopic findings of optic atrophy reflect the loss of nerve fibers with decrease in number of small blood vessels crossing optic disc with accompanying gliotic reaction (Figure 7-3). Funduscopic criteria for optic atrophy include the following: white coloration of optic disc representing glial changes, decrease in number (usually 8 to 12) of arterioles seen extending across disc margin, and thinning of retinal nerve fiber layer, best appreciated by looking at very small caliber branches of retinal arterioles. In ischemic optic atrophy, such as temporal arteritis, retinal arteries can be markedly narrowed; in glaucomatous atrophy disc is enlarged and deeply cupped. In patients with optic atrophy there is decreased visual acuity in the involved eye with afferent pupillary defect (minimal pupillary constriction with direct light stimulation but brisk response of involved eye when light is applied to normal eye).

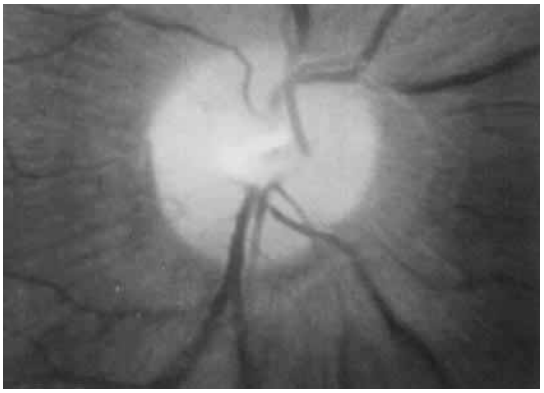

Figure 7-3. Pale gliotic-appearing optic disc with loss of nerve fibers consistent with optic atrophy.

Patients with retrochiasmal lesions have homonymous visual field defects. Certain patients can be unaware of this deficit (visual inattention), whereas others initially complain of difficulty reading or problems bumping into objects in affected visual field. In patients with retrochiasmal hemispheric (temporal, parietal, occipital) lesions and during certain drug intoxications, unusual visual disturbances can be present, including these:

- Prosopagnosia--inability to recognize faces

- Palinopsia--persistent visualization of an image after stimulus has been removed
- Metamorphopsia--distortion of shape or form of objects
- Micropsia or macropsia--abnormal perception of size--or teleopsia--distance of an object from the patient
- Phosphenes--flashing of bright lights or colors
- Chromatopsia--perception that the environment is tinted in single color
- Visual hallucinations

With occipital lesion such as neoplasm or hemorrhage, perceived visual sensations are unformed and consist of flashing lights or spots, whereas parietal-temporal lesions cause complex hallucinations and illusions, including entire pictures or scenes. It is important not to falsely identify the cause of these bizarre and unusual visual phenomena as hysteric in origin or caused by psychotic disorders without careful assessment of visual system.

SIGNS AND SYMPTOMS OF DISEASE IN THE EFFERENT VISUAL SYSTEM

Symptoms

Patients with ocular motility abnormalities complain of double vision (diplopia) or movement of objects in the environment (oscillopsia). In patients with congenital strabismus (misalignment or lack of parallelism of visual axes of eyes), there may be no diplopia because the visual image from the deviated eye is suppressed. If ocular misalignment is observed, it is called tropia (turning), for example, exo- or esotropia, hypo- or hypertropia; however there can be phoria, which is misalignment brought out by cover-uncover testing of each eye. Observe whether the eyes appear in parallel alignment and whether they move conjugately. Specific characteristics of diplopia must be determined. Box 7-5 lists four questions specific to the charactistics of diplopia.

Box 7-5

1. Are images horizontal side by side (this indicates dysfunction of lateral or medial recti), or located vertically one above the other (this indicates superior or inferior recti or oblique muscles)?
2. Is diplopia more prominent on near gaze as a result of superior oblique or medial recti paresis or on far gaze as a result of lateral rectus paresis?
3. With closure of one eye, does diplopia disappear or persist (monocular diplopia is usually caused by ocular disease, e.g., lens opacities, dislocated lens, psychogenic origin, or occipital-parietal lesions causing metamorphopsia)?
4. Has double vision subsequently improved (vascular), become more severe and persistent (tumor), or does it fluctuate during the day (myasthenia gravis)?

Examination

Evaluation of eye movements includes observation of eyelids, orbits, and facial movements. The presence of proptosis (protrusion of globe) is evaluated by standing behind the seated patient and looking directly down at the orbits. To measure proptosis objectively, a small and inexpensive exophthalmometer is necessary. The upper limit of normal forward eye protuberance is 22 mm; there should be no more than 3 mm intereye difference. When observing eyelids, note any degree of eyelid asymmetry. Ptosis caused by levator palpebrae weakness is initially detected on upward gaze. The finding of ptosis does not necessarily imply neurologic abnormality. Compare patient's present appearance with old photographs to differentiate congenital from recently developing ptosis. Ptosis can be caused by myopathic conditions such as myasthenia gravis, by neurogenic conditions such as third nerve or sympathetic disorders, or by loss of tension (laxity) in levator tendons (dehiscence); the latter is of nonneurological origin. Ptosis can cause visual impairment if the pupil is covered. The diagnosis is based on these general facts:

- Ptosis of myopathic origin is usually bilateral, and the pupil is spared.
- Ptosis caused by third nerve disorders is unilateral and produces paresis of medial and superior gaze; the pupil is dilated and unreactive.
- Ptosis caused by oculosympathetic paresis (Horner's syndrome) is unilateral, the pupil is miotic, reduced sweating occurs on the involved side of face, and no motility abnormality occurs.

If third nerve is compressed by extrinsic mass lesion (aneurysm or neoplasm), pupil is dilated and unreactive to light stimulation, whereas pupil is usually normal in diabetic ophthalmoplegia caused by vascular ischemia. This results because pupillary fibers are located peripherally and are initially affected by compressive lesions, yet are spared in ischemic lesions as blood vessels initially traverse external nerve surface and are less vulnerable to vascular ischemia.

Pupils that respond poorly to light should be tested for reactivity (constriction) or near fixation. Miotic pupils have normal near reflex (pupilloconstriction, convergence, lens accommodation) but reacting poorly to light are indicative of neurosyphilis (Argyll Robertson pupil). Tonic (Adie's) pupil shows absent constriction to light with slowed and tonic constriction to near target (accommodation). The pupil can show slowed writhing movements when viewed with a slit lamp. This condition can be unilateral or bilateral, is most common in young women, and can be associated with decreased or absent deep tendon reflexes. Tonic pupil is a benign condition; however, certain patients report visual blurring especially when shifting from far to near objects. The diagnosis of tonic pupil is confirmed by pharmacologic testing. There is rapid and complete pupillary constriction using 0.125% pilocarpine; this indicates denervation cholinergic sensitivity. Tonic response to accommodation and pharmacologic testing differentiates Adie's pupil from Argyll Robertson pupil (Box 7-6).

Box 7-6 Features of Pupillary Syndromes
Argyll-Robertson Syndrome
Visual function intact
Intact accommodation response
Abnormal pupillary light response
Irregular shaped miotic and asymmetric pupils
Iris atrophy
Usually caused by neurosyphilis
Adie's Tonic Pupil Syndrome
Dilated pupils
Absent reaction to light stimulation
Slow pupillary reaction to near response
Slow (tonic) dilation after near response
Iris sphincter sector paresis
Impaired accommodation
Pupils constricts to 0.125% pilocarpine
Associated diminished deep tendon reflexes

The effect of drugs on pupillary size and reactivity must be considered; pupillary dilation can be secondary to anticholinergic drugs; constriction can result from heroin abuse. In comatose patients, pupillary size or reactivity is quite important, and four general rules should be remembered (Box 7-7).

Box 7-7
1. Metabolic disturbances do not alter pupillary size or reactivity.
2. Unilateral cerebral hemispheric lesions cause a unilateral dilated and poorly reactive pupil if uncal (transtentorial) herniation has occurred.
3. Midbrain lesions cause bilateral dilated and fixed pupils.
4. Pontine lesions cause bilateral miotic but reactive pupils.

If the patient has fixed dilated pupils without ptosis or extraocular muscle paresis, this may have resulted from local application of sympathomimetic (dilating) drug. Failure of dilated pupil to constrict following local application of 1% pilocarpine indicates pharmacologic blockade because pupillary constriction occurs with neurologic disorders. If patient has miotic pupil, there can be an oculosympathetic chain lesion (Horner's syndrome), or this can represent normal variant. In Horner's syndrome, the miotic pupil does not respond (dilate) as well as a normal pupil to ocular instillation of cocaine solution, whereas if miosis represents normal asymmetry, the response in both eyes should be equal. Sympathetic fibers originate in the hypothalamus and traverse the brain stem to the spinal cord (C7-T2), then travel to superior cervical ganglion. Postganglionic sympathetic fibers traverse carotid artery sheath, cavernous sinus, and orbit. Lesions at these locations cause Horner's syndrome (Box 7-8).

Box 7-8 Etiologies of Horner's Syndrome
First Neuron
Hypothalamic lesion
Pituitary-chiasmal lesion
Basal meningitis--arachnoiditis
Brain stem lesion
Cervical cord lesion
Second Neuron
Cervical rib
Apical lung lesion (Pancoast tumor)
Aortic aneurysm
Cervical adenopathy
Brachial plexus lesion
Third Neuron
Carotid Artery lesion
Cavernous sinus lesion
Paratrigeminal syndrome
Oribital lesion
Cluster headache

Determining the location of the lesion causing Horner's syndrome is simplified by the presence of certain accompanying findings, for example, radiographic evidence of mediastinal mass or neurologic signs of brain stem involvement. Pharmacologic agents can be used to differentiate central (hypothalamus, brain stem, and spinal cord) and preganglionic (mediastinum) causes from postganglionic (carotid artery sheath, cavernous sinus, and orbit) lesions causing Horner's syndrome. If postganglionic system is involved, there is denervation hypersensitivity; pupil dilates in response to local application of 1:1000 epinephrine solution. If the postganglionic system is intact (neurons from superior cervical ganglion in the neck to pupillodilator fibers in orbit), hydroxyamphetamine (Paredrine), which releases norepinephrine from postganglionic vesicles, causes pupillary dilation. Failure to obtain this dilatation with Paredrine indicates postganglionic lesions as the cause of Horner's syndrome.

Conjugate gaze can be tested for command (ask the patient to look to the right, left, upward, downward) and pursuit or slow tracking eye movements (ask patient to follow the pointer). The pathway for supranuclear control of gaze for saccades (rapid eye movements) begins in frontal lobe (area 8) and descends through internal capsule and diencephalon to reach midbrain, where it crosses in paramedian pontine reticular formation (lateral gaze center). Cortical lesions can be irritative (neoplasm or abscess) or destructive (infarction or hemorrhage). These lesions cause differential effects on lateral conjugate gaze; certain lesions, for example, glioma and meningioma, can act as both destructive and irritative lesions. A destructive lesion above decussation causes paresis of horizontal conjugate eye movements to contralateral side; lesions below decussation cause ipsilateral paresis. During simple partial seizures caused by irritative lesions originating in left frontal region, frontal eye fields are stimulated with eyes deviating to right side. Following left-hemispheric destructive lesion, horizontal eye movement to right is impaired and eyes deviate to left (in direction opposite hemiparesis) such that eyes look toward the lesion. When lesions located below oculomotor decussation, there is paralysis of conjugate

gaze to the side of the lesion such that eyes deviate away from brain stem lesion.

Another type of disorder involving horizontal (lateral) conjugate gaze is internuclear ophthalmoplegia caused by medial longitudinal fasciculus lesion located in the brain stem. This disorder causes impaired medial (adduction) movement of eye ipsilateral to the lesion with horizontal nystagmus in the abducting opposite eye. In resting state eyes are conjugate and convergence movements are normal; this indicates that medial rectus muscle functions normally. Bilateral internuclear ophthalmoplegia occurs most frequently in patients with multiple sclerosis. Other causes include brain stem tumors and ischemic vascular disease; these usually cause unilateral internuclear ophthalmoplegia.

Using red lens placed over one eye (by convention, right eye) helps clarify defective extraocular muscle(s) and/or cranial nerves in patients with diplopia. Box 7-9 lists some of the causes of extraocular nerve lesions. Two rules are inviolate: one, diplopia is maximal in the greatest separation of images in field gaze of involved muscle; and two, an image belonging to paretic muscle projects peripherally. For example, in isolated right lateral rectus lesion with abductor muscle paresis, diplopia is horizontal and maximal on far gaze. With red lens placed over right eye, two images are seen on gaze to right side with red image (caused by the right lateral rectus) projecting peripherally to right (uncrossed diplopia). In right third-nerve lesions with adductor muscle paresis, there is maximal separation of two horizontal images on gaze directed to left with red image (with red glass held over right eye) projecting peripherally (crossed diplopia). Muscles functioning as torters (rotation of globe) are tested by noting the degree of separation of objects when head is tilted. For example, with right superior oblique palsy, images are closer together when head is tilted to the left and further apart when head is tilted to the right. If patient reports diplopia, ocular deviation can be due to muscle paresis or muscle restriction (as in thyroid ophthalmopathy). In restrictive disorders (such as thyroid eye disease), forced duction test is performed with forceps to move the eye, and cannot be moved because of eye muscle restriction by the pathologic process. Determination of paretic muscles in patients with diplopia can be very complex if multiple extraocular muscles are paretic.

BOX 7-9 Etiologies of Extraocular Nerve Lesion	
Isolated Oculomotor	
Lesion	Carotid aneurysm
Painful	Parasellar lesion
	Ophthalmoplegic migraine
	Ischemic (diabetic disease
	Head injury
Painless	Basal meningitis
	Parasellar lesion
	Nasopharyngeal lesion
	Multiple sclerosis
	Myasthenia gravis
	Thyroid ophthalmology
	Wernicke syndrome
Isolated Trochlear	Head injury
Paresis	Multiple sclerosis
	Myasthenia gravis
	Thyroid ophthalmopathy
	Wernicke syndrome
	Ischemic diabetic syndrome
	Neoplasm
	Orbital lesion
Isolated abducens	Intracranial hypertension
paresis	Lumbar puncture
Nonlocalizing	Head trauma
	Basal meningitis--arachnoiditis
	Myasthenia gravis
	Wernicke syndrome
	Brain stem (pontine)
	Clivus
Localizing	Temporal bone
	Parasellar region
	Cavernous sinus
	Superior orbital fissure
	Orbital lesion

If there is abduction defect (lateral rectus is innervated by sixth nerve), the underlying pathologic process can be myopathic (thyroid myopathy, myasthenia gravis) or neuropathic (such as caused by cranial nerve dysfunction). If neuropathic, sixth-nerve palsy can be caused by the effects of increased intracranial pressure or meningitis process (neoplastic, infectious-inflammatory) at skull base. The sixth nerve may be damaged anywhere along its intracranial (e.g., mid pons, cerebellopontine angle, clival or skull base, parasellar or cavernous sinus) or extracranial (superior orbital fissure, orbit) course. If there is hypertropia in adduction, superior oblique muscle is usually involved. This nerve is susceptible to traumatic injuries; other causes (vascular, neoplastic) are less common. A droopy eyelid with only preserved lateral (abduction) movement is characteristic of oculomotor nerve paresis. This nerve may be damaged along its intracranial (midbrain, parasellar or cavernous sinus, tenoorial notch) or extracranial (orbital) course. If pupil is involved, compressive lesion should be considered, whereas if pupil is spared, there is usually ischemic process.

CLINICAL SYNDROMES

The following clinical syndromes are best understood when the symptoms, acuity, visual field, funduscopy, pupils, motility, and etiology are all analyzed and taken into consideration.

Optic Neuritis

Symptoms. Loss of vision may worsen over several days; orbital or ocular pain especially moving eyes. Visual loss is usually unilateral but can be bilateral; however, this rarely develops simultaneously.
Acuity Usually substantial impairment***.***
Visual Field. Central scotoma(s)
Funduscopy. Blurring of disc margins, presence of hemorrhages and exudates; if involvement is retrobulbar, disc can initially appear normal and later show optic atrophy.
Pupils. Equal in size; involved pupil responds more rapidly and briskly to consensual than direct light stimulation.
Motility. Normal.
Etiology. Demyelinating disease; less commonly, syphilis, systemic lupus, infectious, including sinus disease. "Chronic" or slowly progressive optic neuritis can be incorrectly diagnosed when visual loss is actually due to juxtasellar mass lesion and there is usually unilateral visual impairment.
Associated Findings. Can be none or other signs of multiple sclerosis (cerebellar, brain stem, corticospinal) or juxtasellar involvement.
Diagnostic Studies. Visual evoked potentials, cerebrospinal fluid analysis with multiple sclerosis profile, orbital computed tomography or, magnetic resonance imaging, serology (VDRL and fluorescent treponemal antibody), and studies for systemic lupus (LE prep and antinuclear antibodies).

Optic Neuropathy

Symptoms. Loss of visual acuity is characteristic. This can be sudden in ischemic lesions and slow in compressive (mass) lesions or hereditary, toxic, or metabolic deficiency conditions. Visual loss is usually unilateral with compressive and vascular lesions and bilateral with hereditary, toxic, and metabolic deficiency conditions.
Acuity. Can vary from minimal reduction to severe impairment. Color vision is usually severely impaired.
Visual Fields. Central or ceocentral scotomas in hereditary, toxic, or metabolic deficiency. In compressive optic neuropathy, it is imperative to check carefully visual fields in the eye with normal acuity for possible temporal defect that suggests chiasmal extension. In ischemic optic neuropathy there can be an altitudinal visual defect.
Funduscopy. In ischemic and toxic lesions there is acute disc swelling. In hereditary optic neuropathy there is optic atrophy. In compressive lesions the disc can appear normal or atrophic.
Pupils. Equal in size. In bilateral conditions, pupillary response can be symmetric. In unilateral (ischemic, compressive) lesions there can be an afferent pupillary defect.
Motility. Normal
Etiology. Ischemic (collagen vascular disorders, temporal arteritis, syphilis, diabetes mellitus); toxic (ethambutol, isoniazide, tobacco-alcohol, disulfiram); metabolic (vitamin B_{12} deficiency); compressive (optic glioma, meningioma); hereditary (Leber's optic atrophy).
Associated Findings. Can be none.
Diagnostic Studies. Same as for optic neuritis. To include investigation for metabolic, infectious immune-mediated disease.

Papilledema

Symptoms. Headache, nausea, vomiting, horizontal diplopia caused by lateral rectus paresis, and transient visual obscurations caused by intermittent optic nerve dysfunction.
Acuity. Usually normal until late in course; however, more sensitive vision parameters, including tests of contrast sensitivity, can show abnormalities.
Visual Field. Enlargement of blind spots, constriction of visual fields, and inferior nasal quadrant defect.
Funduscopy. Loss of spontaneous venous pulsations; loss of physiologic cup, hemorrhages, and exudates; blurring of disc margins; venous engorgement with disc hyperemia; and blurring of retinal nerve fiber layer.
Pupils. Normal.
Motility. Can demonstrate lateral rectus paresis
Etiology. Intracranial hypertension caused by mass lesion or idiopathic intracranial hypertension.
Associated Findings. Related to underlying cause of intracranial hypertension.
Diagnostic Studies. Skull roentgenogram, computed tomography/magnetic resonance imaging (CT/MRI), angiogram to search for venous sinus thrombosis, and CSF analysis if CT/MRI normal.

Prechiasmal Lesions (Optic Nerve--Optic Chiasm Junction)

Symptoms. Progressive dimming of vision in only one eye is usually initial disorder.
Acuity. Ipsilateral impairment of visual acuity.
Visual Field. Contralateral monocular temporal hemianopia, and ipsilateral central scotoma.
Funduscopy. Optic atrophy is late sign.
Pupils. Delayed and incomplete constriction in ipsilateral eye in response to direct light stimulation with brisk consensual response (afferent pupillary defect).
Motility. Normal.
Etiology. Chromophobe adenoma, craniopharyngioma, parasellar meningioma, aneurysm.
Associated Findings. Can be none or signs of pituitary dysfunction.
Diagnostic Studies. CT/MRI scan and carotid angiogram to detect tumor and exclude aneurysm.

Chiasmal Lesions

Symptoms. Gradual onset of visual blurring that progressively worsens; impaired peripheral vision and abnormal depth perception.
Acuity. Unilateral or bilateral visual loss, or can be normal.
Visual Field. Bitemporal defect: if optic chiasm is compressed from undersurface, superior quadrantanopsia results; if the chiasm is compressed from above, inferior quadrantanopsia results.
Funduscopy. Optic atrophy or can be normal.
Pupils. Usually normal, but can see unilateral afferent defect.
Motility. Diplopia caused by extraocular muscle involvement as the lesion extends laterally into cavernous sinus.
Etiology. Chromophobe adenoma, craniopharyngioma, tuberculum sella meningioma, aneurysm.
Associated Findings. Hypopituitarism with amenorrhea, infertility, hypothyoridism, hypoadrenalism; hyperpituitarism as expressed by amenorrhea-galactorrhea, acromegaly, or Cushing's syndrome.
Diagnostic Studies. CT/MRI scan and angiography, Hormonal studies to detect hypothalamid-pituitary dysfunction.

Postchiasmal Lesions

Symptoms. Occasional difficulty seeing in involved visual field; more commonly no awareness of visual field abnormality occurs.
Acuity. Normal, but may ignore involved hemifield of eye chart.
Visual Field. Homonymous defects: Temporal lobe: Superior homonymous quadrantanopsia. Parietal lobe: Inferior homonymous quadrantanopsia. Occipital lobe: Homonymous hemianopsia, usually with macular sparing
Funduscopy. Normal for postgeniculate lesions. Can see optic atrophy with pregeniculate lesions (optic tract and lateral geniculate body).
Pupils. Normal with postgeniculate lesions, or can have hemianopic pupillary afferent defect with optic tract lesions (e.g., when point source of light is directed into hemiretina corresponding

to hemianopic field, diminished light response is present).
Motility. Normal.
Etiology. Vascular, neoplasm, trauma, and inflammatory.

Associated Findings.
Temporal lobe: Partial complex seizures, aphasia, hemiparesis, hemianesthesia.
Parietal lobe: Aphasia, extinction of sensory stimuli, agraphia, acalculia, right-left confusion, and finger agnosia.
Occipital lobe: Usually none.

Diagnostic Studies.
EEG, CT/MRI scan, carotid angiogram.

Hysterical Blindness

Symptoms. Decreased vision or blindness; however patient has no difficulty walking and does not bump into objects.
Acuity. Impaired, but severity of visual loss is constant and does not change at varying distances.
Visual Field. Tubular with no change in size of the field with increasing distance from target (tunnel vision).
Funduscopy. Normal.
Pupils. Normal.
Associated Findings. If patient with unilateral visual loss is asked to read red letters with red lens over normal eye and clear lens over "blind eye," this can only be accomplished with the blind eye. This is because it is not possible to see red letters with a red lens over the eye; therefore, vision must originate from the eye alleged to be blind.

SUMMARY

Diminished vision is a sign that the afferent (sensory) visual system is impaired, whereas diplopia (double vision) caused by abnormal ocular motility is a manifestation that the ocular motor (efferent) system is impaired. In patients who report visual loss, it is important to assess the primary visual apparatus (ophthalmological system). If this is not the cause of the visual impairment, it is necessary to assess the optic nerve, optic chiasm, and visual pathways traversing the parietal and temporal lobes and their termination in the occipital lobe, which is the final common pathway for the visual system as the suspected cause of the visual impairment. Adequate visualization of the optic nerve head (papilla) is mandatory as the funduscopic findings can reflect the presence of normal or increased intracranial pressure as well as showing inflammation or demyelination of the optic nerve. The visual field examination is crucial to determine if the abnormality involves the retina, optic nerve, or pre- or postchiasmal region. Patients with ocular motility disturbances usually report double vision or the feeling of objects moving in the environment (oscillopsia); however, it is possible for patients with ocular motility disturbances to have no symptoms. Evaluation of the patient's eye movements as well as the eyelids, orbital region, and facial movements is necessary to determine whether the pathological process involves the extraocular muscles or cranial nerves supplying these muscles or is due to CNS dysfunction.

Suggested Readings

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