

NEURO-OPHTHALMOLOGY

Objectives

As a primary care physician, you should be able to perform a **basic neuroophthalmic examination** and to **recognize and interpret the more common signs and symptoms of neuro-ophthalmic disorders.**

To achieve these objectives, you should learn:

- **To measure visual acuity**
- **To examine pupillary reactions**
- **To test the function of the extraocular muscles**
- **To evaluate the visual fields**
- **To inspect the optic nerve head**

Relevance

The visual pathways and the oculomotor system reflect much of the status of the nervous system as a whole. Approximately **35% of the sensory fibers** entering the brain are **in the two optic nerves**. It is estimated that **65% of intracranial diseases exhibit neuro-ophthalmic symptoms or signs**. Routine neurovisual examination allows the primary care physician to identify abnormalities indicating neurologic disorders. Important examples are **brain tumors, multiple sclerosis, cerebrovascular disease, and cerebral aneurysms**.

How to Examine

Visual Acuity Testing

The first step in any eye examination is to measure visual acuity in each eye. The chief complaint of most patients with ocular problems is some aberration of vision. Use of the conventional **Snellen eye chart** is currently the easiest and best method of measuring the function of the macular fibers, although any reading material may be useful for screening purposes. Both distance and near vision ideally should be measured and recorded using the patient's corrective lenses if available.

Visual Field Testing

Gross confrontation field-testing is an accepted screening procedure. Each eye must be tested separately. While fixing on the examiner's eye with the non-occluded eye, the patient is asked to count fingers in each of the four quadrants of the visual field.

Because most visual field defects affect central vision, attention should be paid to the area within the central 30' . A method of screening for central field defects is to ask the patient to look at a piece of graph paper or an Amsler grid, which is similar, but specific for this type of testing. (See Photo 3.3 in Chapter 3 for a depiction of the Amsler grid and further discussion on its use in central visual field testing.)

Pupillary Reactions

Pupils should be inspected while the patient is looking at distance to avoid the pupillary constriction that occurs with a near target. The pupils should be round and equal in diameter, although less than 1 millimeter of inequality may be a normal variation. Poor pupillary dilation in dim light may indicate dysfunction of the sympathetic nervous system, and poor pupillary constriction to bright light may indicate parasympathetic dysfunction.

The swinging-flashlight test is the most valuable clinical test for optic nerve dysfunction currently available to the general physician. The abnormality detected with this test is the afferent pupillary defect, also known as the Marcus Gunn pupil (see "Afferent Defect" in Illustration 7.2). To perform this test, dim room illumination and a bright light are helpful. The patient maintains fixation on an object 15 feet or more away, The bright light is held directly in front of one eye for 3 to 5 seconds, moved rapidly across the bridge of the nose to the front of the other eye for 3 to 5 seconds, and then shifted back to the first eye. This procedure should be repeated several times until the examiner is certain of the responses. The critical observation to be made is the behavior of the pupil when it is first illuminated. A normal response is initial pupillary constriction followed by variable amounts of redilation. An abnormal response is slow dilation without initial constriction. The relative afferent pupillary defect almost always indicates a lesion in the optic nerve on the affected side, although rarely a large retinal lesion may produce this defect.

Ocular Motility Testing

Eye movements should be tested, especially if the patient has a complaint of double vision or if any neurologic disease is suspected. First, alignment of the eyes should be assessed to identify a deviation that could be due to strabismus, extraocular muscle malfunction, or oculomotor nerve dysfunction. Ophthalmoplegia can be distinguished from nonparalytic strabismus by the presence of incomitancy. Rapid eye movements (saccades) are tested by asking the patient to direct the eyes quickly to a target that the examiner is holding right, left, up, and down. Tracking movements (pursuit) are tested by asking the patient to follow a slowly moving target first horizontally and then vertically.

The examiner should be looking for the following abnormalities:

1. Strabismus
2. Limitation of movement of one eye
3. Limitation of gaze (both eyes affected similarly)
4. Nystagmus (spontaneous jerking eye movements)

Ophthalmoscopy

Evaluation of a patient with neurologic symptoms is not complete without evaluation of the ocular fundus. Particular attention should be given to the appearance of the optic disc. In some neuro-ophthalmic conditions, the optic disc may be swollen and elevated; in others, it may be pale and atrophic.

How to Interpret the Findings

Pupillary Disorders

Dilated Pupil

A dilated pupil, especially one that does not react to light, usually indicates a lesion in the efferent limb of the pupillary reflex (see "Efferent Defect" in Illustration 7.2). A dilated pupil in a patient with a head injury or a cloudy sensorium may indicate compression of the third cranial nerve (oculomotor nerve) by herniation of the temporal lobe and may have grave importance. On the other hand, a dilated, fixed pupil in an otherwise asymptomatic, healthy patient is usually benign. In a healthy individual, a dilated pupil may reflect a benign lesion in the ciliary ganglion (an Adie' s tonic pupil) or may be secondary to instillation of a dilating eyedrops.

Tonic Pupil

The tonic pupil, known as *Adie's pupil*, is seen predominantly in young women and is usually unilateral. In ordinary light, the tonic pupil is usually larger than its counterpart; the reaction to light is either diminished or absent. Instillation of weak cholinergic agents (e.g., 1/8% Pilocarpine Hydrochloride) will cause constriction of a tonic pupil, indicating denervation hypersensitivity, whereas this constriction will not occur in a normal pupil. By itself, a tonic pupil is of no neurologic significance.

Unilateral Small Pupil

A small pupil in one eye that has normal reactivity to light and near stimuli is usually physiologic and is of no neurologic significance. However, when accompanied by ptosis of the upper eyelid, a small pupil may indicate Horner' s syndrome (Figure 57). Horner' s syndrome is caused by a congenital or an acquired lesion of the sympathetic pathways, either in the central or preganglionic portion from the hypothalamus to the superior cervical ganglion or in the postganglionic portion from the cervical ganglion to the eye. Detection can be accomplished through the topical instillation of 4% cocaine, which will dilate a normal pupil but not a desympathectomized pupil. Differentiation of preganglionic from postganglionic lesions is important and can usually be accomplished through the instillation of hydroxyamphptetamine drops if available.

Neuromotility Disorders

Some significant oculomotor disorders that the primary care physician should recognize are discussed below.

Third Cranial Nerve Paresis

Cranial nerve III (oculomotor nerve) supplies the levator palpebrae muscle of the upper eyelid, the superior rectus, medial rectus, inferior rectus, and inferior oblique muscles, as well as the parasympathetic fibers to the sphincter of the iris. Complete paralysis of the oculomotor nerve produces both horizontal and vertical diplopia, with ptosis of the upper eyelid and an inability to rotate the eye inward, upward, or downward (Figure 58). The pupil may be dilated and non-responsive. The most common causes of isolated third-nerve palsy include intracranial aneurysm (especially of the posterior communicating artery), vaso-occlusive disease within the nerve (usually associated with diabetes and hypertension), trauma, and brain tumor.

Fourth Cranial Nerve Paresis

Cranial nerve IV (trochlear nerve) innervates the superior oblique muscle so that complete paralysis causes vertical diplopia. The patient often notices more difficulty in downgaze and may tilt the head toward the opposite shoulder to minimize the diplopia. The most frequent cause of isolated fourth-nerve palsy is closed-head trauma, but the condition is also found in patients with small-vessel disease, especially associated with hypertension or diabetes.

Sixth Cranial Nerve Paresis

Cranial nerve VI (abducens nerve) supplies the lateral rectus muscle; therefore, complete paralysis produces loss of abduction and horizontal diplopia, with the greatest separation of images in gaze directed toward the affected side (Figure 59). Intracranial tumors account for approximately 30% of cases of isolated sixth-nerve paralysis. Head trauma, small vessel disease, viral infections, and increased intracranial pressure are also frequent causes of abducens paresis.

Myasthenia Gravis

Myasthenia gravis is a chronic autoimmune condition that interferes with neuromuscular transmission in skeletal muscles. The disease can affect any muscles, but ptosis and double vision are the presenting signs in about half the patients. Characterized by fatigability of muscle function on sustained effort, myasthenia gravis may mimic nearly any other oculomotor problem, including third-, fourth-, and sixth-nerve disease, gaze paresis, and internuclear Ophthalmoplegia. All patients with unexplained diplopia or ptosis should have an edrophonium chloride (Tensilon) test.

Internuclear Ophthalmoplegia

Lesions of the medial longitudinal fasciculus, which carries input from, the ipsilateral sixth-nerve nucleus to the contralateral third nerve medial rectus subnucleus, are usually of considerable diagnostic significance. The clinical manifestations of such a lesion include

straight eyes in primary gaze but weakness of the adducting eye and nystagmus of the abducting eye in lateral gaze. It may be unilateral or bilateral, and convergence is usually intact. In older individuals, unilateral internuclear Ophthalmoplegia suggests small-vessel disease within the distribution of the vertebral basilar arterial system. In young adults, bilateral internuclear Ophthalmoplegia is almost always due to demyelinating disease. In children, however, internuclear Ophthalmoplegia may be due to a pontine glioma.

Nystagmus

Spontaneous, rhythmic, back-and-forth movement of one or both eyes is referred to as *nystagmus*. The direction may be horizontal, vertical, rotary, or a combination. The three most common forms of nystagmus are benign and do not indicate central nervous system dysfunction. The first form of benign nystagmus occurs in end-gaze, when the patient is attempting to maintain the eyes in extremes of lateral gaze. In this position, it is not unusual for the eyes to drift back slightly from the extreme horizontal gaze position and then refixate with a small jerk movement. End-point nystagmus is usually not well sustained and disappears as the patient is permitted to move the gaze slightly away from the extreme position. The second form of benign nystagmus is induced by drugs: diphenylhydantoin, barbiturates and other sedatives. In this form, a jerk nystagmus may be present in all positions of gaze. The third form, a searching, pendular nystagmus, is commonly seen in individuals who are visually impaired from birth.

Nystagmus may indicate central nervous system dysfunction. Representative diseases that cause nystagmus are multiple sclerosis, brain tumor, and degeneration of the central nervous system.

Optic Nerve Disease

Optic Disc Elevation

Congenital Anomalous Disc Elevation- Occurring in slightly less than 1% of the population, congenital anomalous disc elevation is a benign, nonprogressive condition. The optic disc margins are blurred, the disc substance is elevated, and the optic cup is absent. However, there is no evidence of edema or hemorrhage. Congenital disc elevation may be associated with hyperopia and may be accompanied by the accumulation of calcified protein within the optic disc itself (optic disc drusen). Because of its deceptive appearance, congenital fullness of the optic nerve has been referred to as *pseudopapilledema* (Figure 60, left). The condition should not be confused with true, acquired papilledema (Figure 60, right). Such differentiation may be difficult, and the diagnosis of true papilledema sometimes cannot be made without evidence of progression obtained through serial examination or other diagnostic tests, including lumbar puncture.

Papilledema One of the most important ophthalmoscopic findings is papilledema; swelling of the optic disc secondary to increased intracranial pressure occurs in approximately 50% of patients with a brain tumor. Characteristics of fully developed papilledema include hyperemia of the disc, tortuosity of the veins and capillaries, blurring and elevation of the margins of the disc, and hemorrhages on and surrounding the nerve head. The signs of early

papilledema may not be distinct, with subtle elevation of the disc margins, loss of previously identified spontaneous venous pulsations, and mild hyperemia. In addition to intracranial mass lesions, papilledema may be seen in pseudotumor cerebri and in severe acute systemic hypertension.

Papillitis Inflammatory edema of the disc, known as *Papillitis or anterior optic neuritis*, may be indistinguishable from papilledema by its ophthalmoscopic appearance. Whereas papillitis is more commonly unilateral, papilledema is usually bilateral.

Optic Atrophy

Optic atrophy (Figure 61), or pallor of the optic disc, results from damage to the nerve fiber layer of the retina, the optic nerve itself, the optic chiasm, or the optic tracts. With progressive loss of axons and alteration in glial tissue, the optic disc becomes pale. After extensive damage, the disc may become white.

There is a wide range of normal coloration of the disc. A diagnosis of optic atrophy should not be made unless there is decreased visual acuity or visual field loss accompanying the paleness of the optic disc or, if unilateral, a relative afferent pupillary defect. Common causes of optic atrophy include previous optic neuritis or long-standing papilledema, compression of the optic nerve by a mass lesion, such as a meningioma, or pituitary adenoma, ischemic damage to the optic nerve due to small-vessel disease or giant cell arteritis and glaucoma.

Visual Field Defects

The following terms are commonly used to discuss visual field loss:

Scotoma An area of reduced or absent vision within an otherwise intact visual field.

Hemianopia- Loss of half the visual field. Usually, this involves loss of either the right or the left half of the visual field in either eye; however, the term *altitudinal hemianopia* may be used to distinguish loss of the superior or inferior half of the visual field.

Homonymous hemianopia- Loss of either the right or the left half of the visual field in both eyes.

Bitemporal hemianopia- Loss of the right half of the visual field in the right eye and loss of the left half of the visual field in the left eye.

Lesions anywhere in the visual system, from the retina to the occipital lobes, will produce visual field defects (Illustration 7.3). Although detection and analysis of these visual field defects have led to entire texts written on the science of perimetry, most physicians need be concerned with only a few types of visual field loss. Neurologically significant field defects are most often central scotomas (due to optic nerve lesions), bitemporal field defects (due to chiasmal disease), or homonymous visual field defects (due to retrochiasmal damage to the optic tracts, the radiations, or the occipital cortex). In almost all locations, the effects produced are most profound within the central 30' of the visual field.

Isolated lesions anterior to the chiasm within the optic nerve produce visual field defects in one eye only. Optic nerve dysfunction typically produces a central scotoma, with

accompanying reduction in visual acuity. Common causes of unilateral optic neuropathy include optic neuritis, optic nerve glioma, and meningioma.

Lesions in the optic chiasm produce visual field defects that affect both eyes, but in a dissimilar fashion. The most common example is a bitemporal hemianopia caused by pituitary adenoma. Another common visual field defect due to disease near the chiasm is loss of central field in one eye and a temporal visual field defect in the other eye.

Lesions affecting the visual pathways behind the chiasm produce homonymous hemianopia or homonymous defects that are less than a complete hemianopia. Because the fibers serving the corresponding portions of the two retinas lie increasingly closer together as the fibers pass back toward the occipital cortex, there is greater correspondence of the field defects in the two eyes as the lesions occur more posteriorly. Central visual acuity is not affected in homonymous hemianopia unless both hemispheres are involved. Stroke is the most common cause of a homonymous hemianopia. Middle cerebral artery occlusion tends to cause a complete hemianopia with other neurologic signs, whereas posterior cerebral artery occlusion causes isolated, congruous (identical) hemianopic scotomas.

Points to Remember

- 1.** Testing of both visual acuity and visual fields is critical in the evaluation of the abnormal optic disc.
- 2.** A patient with a unilateral optic nerve lesion should have equal pupils in ambient light, but a positive swinging-flashlight test.
- 3.** A blurred disc margin is not diagnostic for papilledema. Other signs and symptoms must be considered.
- 4.** Chiasmal disease is most likely to cause a bitemporal field defect.