Neurology

Fifth Year

Neuro ophthalmology

Functional Anatomy of the visual and optomotor systems

The system for peripheral vision begins in the periphery of the retina, where density of light-sensitive rods is greater than that of cones. The fibers of the optic nerve are the axons of the ganglion cells of the retina, of which the macula is the region of most acute vision. Ganglion cell axons exit the eye at the optic disc and travel through the optic nerve, optic chiasm and optic tract to reach the lateral geniculate body of the thalamus. There is a partial decussation of the fibers from each optic nerve at the optic chiasm, those from the nasal half of each retina crossing the midline to join those from the temporal half of the opposite retina.

The optic tracts give off collaterals as part of the **pupillary light reflex** (parasympathetic nucleus of the 3rd cranial nerve) and to the superior colliculus.

In the lateral geniculate body topographical overlap is a structural first step toward binocular single vision. Axons of neurons in the lateral geniculate body project to the visual cortex in the occipital lobe via optic radiation. The optic radiation passes through the posterior limb of the internal capsule. The cortical area of the occipital lobe comprises the visual cortex.

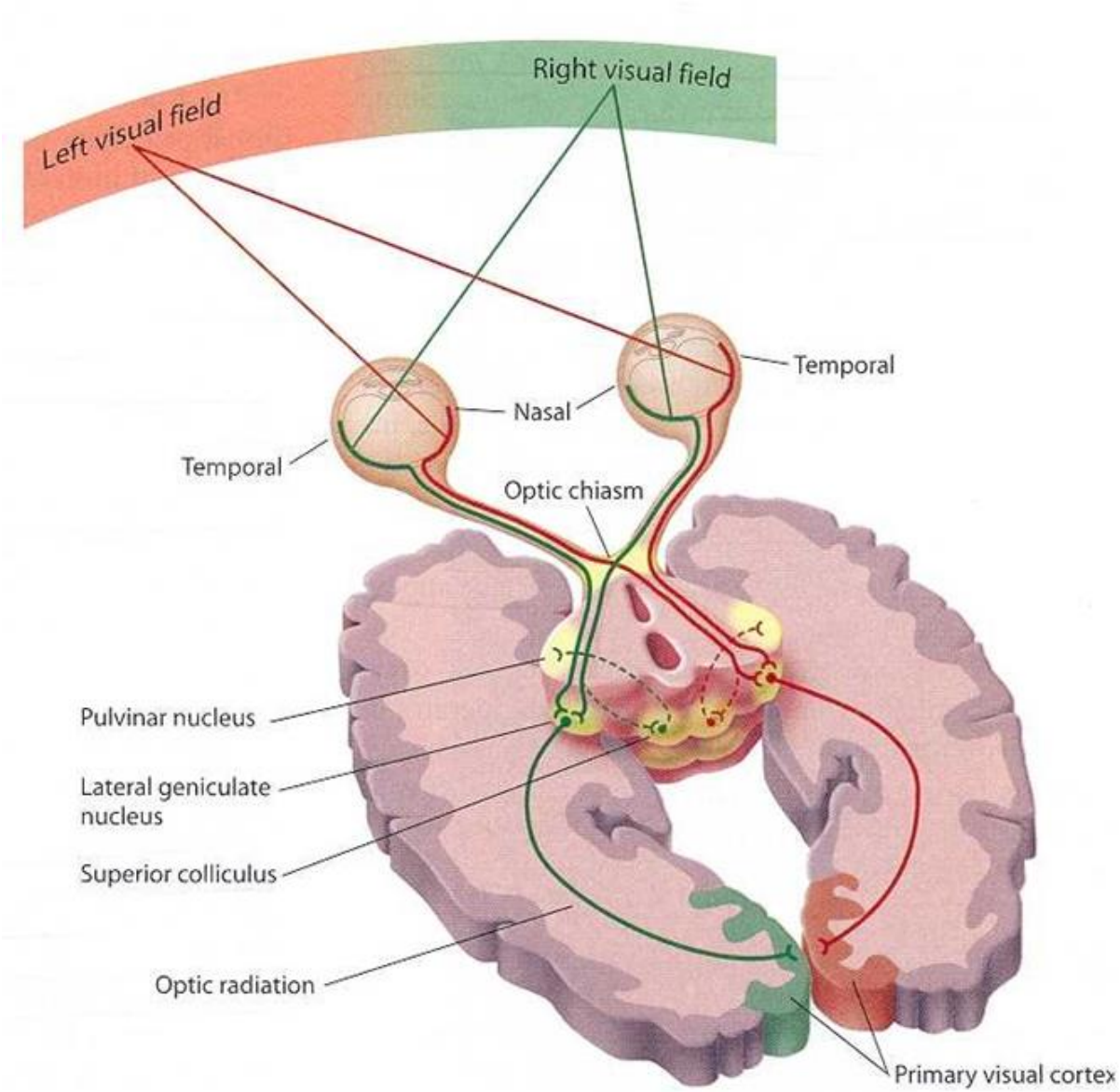
The visual pathway from the retina to the occipital cortex is topographically organized, so the pattern of visual field loss allows precise localization of the site of the lesion.

There are two types of visual streams that project away from primary visual, striate cortex to extrastriate cortical areas for further analysis:

- **The (What) system:** is concerned with object recognition and project inferiorly into the temporal lobe and limbic system.
- **The (Where) system:** is concerned with visuospatial perception and visuomotor performance, this functional stream project superiorly into parietal cortex.

The optomotor system consists of the frontal eye field (area 8), Brainstem gaze centers, the superior colliculus and the cranial nerves third, fourth, and sixth.

The superior colliculus is involved in coordinating eye and head movements to track, capture and maintain a visual image into central retina, especially the fovea, and for the quick jerk response of the eyes to appearance of new stimuli in peripheral vision. Visual information is coordinated with somatsensory and auditory information within the superior colliculus to allow proper tracking of environmental stimuli.



Eye movements

Normal eye movements share the goal of placing an object of visual interest on each fovea simultaneously to allow visualization of a single, stable object. Clear and stable vision is sustained by mechanisms that hold the object on the fovea.

Three major classes of eye movement are defined according to whether the movements are **conjugate** (version), **disconjugate** (vergence) and **reflex eye movement.**

The version system

The version system generates conjugate eye movements that can be further subdivided into two main types, rapid **saccades** and smooth (or slow) **pursuit** movements.

Fast eye movements or saccades are eye movements that rapidly shift the eyes from one object to another. Saccadic movements of the eye are modulated by a pathway arising in the frontal lobes. The supplementary motor area, substantia nigra, superior colliculus, cerebellum, reticular formation and vestibular system play roles in saccadic production and accuracy. You cannot move your eyes smoothly voluntarily. All volitional eye movements require saccades. But not all saccades are voluntary, rapid phase of jerky nystagmus is involuntary.

In contrast if the visual target moves, the **smooth pursuit system** holds the eyes on moving target. Pursuit movements are generated by a visual following reflex that detects the velocity of an image of an object that is moving across the fovea and then generates an eye movement to match that of the image of the target. The pursuit system functions independently of whether the head is still or moving. In the latter case, pursuit is used to cancel the vestibule-ocular reflex. Pursuit can also be used to suppress image movement generated by inappropriate movement of the eyes themselves.

The vergence system

The vergence system deals with problems of binocularly and generates disjunctive eye movements (converges or diverges eyes for near or distant targets) that bring the same image onto each fovea for best stereo-acuity. This system mediates the accommodation reflex. The eyes turn inward (off their parallel axes) and at the same time the pupils constrict and the ciliary muscles relax to thicken the lens. The systems that permit a moving target to remain sharply focused in the fovea (smooth pursuit and vergence systems) are largely involuntary, although the person may choose to glance or not to glance at a particular object.

Vergence eye movements move the eyes in opposite directions so that images of a single object are placed on both foveas.

Reflex eye movement

In alert patients, three systems (reflexes) collaborate to hold the eyes on the target:

- **Ocular fixation (collicur) system:** Fixation reflexes tend to keep the eyes on target which is not moving, when the head moves. The fixation (collicur) system will lock the eyes on target until saccade or a reflex, moves them off.
- **Counter rolling system** (vestibular and neck proprioceptive system): The vestibulo-ocular reflex detects motion of the head and then generates an equal but oppositely directed eye movement in the orbit. In this way the position of the eye in space dose not changes during head rotation so that fixation of an object of interest can be steadily maintained.
- **The optokinetic system** which take over when the environment move.

So a fine corrective reflex movements, keeps the eye in the appropriate orbital position, despite ongoing head motion. Input for these corrective movements comes from the vestibular nuclei (vestibular system) or from the retina (optokinetic and smooth pursuit systems).

Cranial nerves supplied extraocular muscles

The third (oculomotor) cranial nerve:

Nerve nuclei of origin are in the midbrain. It supplies the superior rectus, inferior rectus, medial rectus, inferior oblique, and levator palpebrae superioris muscles. The parasympathetic fibers supply the sphincter, or constrictor, of the pupil, and the ciliary muscle and function in accommodation.

The fourth (trochlear) cranial nerve:

Its nuclei are situated in the lower mesencephalon (midbrain) immediately above the pons. It is the only cranial nerve whose fibers emerge from the posterior aspect of the brainstem. It supplies the superior oblique muscle on the side opposite to the nucleus of origin.

The sixth (abducens) cranial nerve:

Arises in the lower pons. The sixth nerve has the longest intracranial course of all the cranial nerves. It supplies the lateral rectus.

Supranuclear Control of Eye Movements by brainstem gaze centers which receive cortical input from the contralateral frontal eye field (area 8) of the frontal lobe.

Brainstem gaze centers divided into: Vertical and Horizontal gaze centers

Cortical input regulates eye movement and brainstem gaze centers control the position of both eyes.

- **Vertical gaze centers** are generated in structures lying in the area of the rostral mesencephalic (midbrain) reticular formation.
- **Horizontal gaze center** (paramedian pontine reticular formation). From here the output is to the ipsilateral sixth nerve nucleus and also to the contralateral third nerve unclear complex. The pathway joining these two structures is the medial longitudinal fasciculus.

Visual field

The normal monocular visual field subtends an angle of about 160 degrees in the horizontal plane and about 135 degrees in the vertical plane. Within the normal field of each eye is a blind spot, corresponding to the optic disk, which lacks receptor cells.

With **binocular vision,** the horizontal range of vision exceeds 180 degrees.

Vision can be impaired by damage to the visual system anywhere from the eyes to the occipital lobes. One can localize the site of the lesion with considerable accuracy by mapping the visual field deficit by finger confrontation and then correlating it with the topographic anatomy of the visual pathway.

Familiarity with the common types of visual field defects is important if testing is to be reasonably rapid and yield useful information.

Common visual field defects and their anatomical bases:

1. **Total blindness of the right eye** from a complete lesion of the right optic nerve.
2. **Bitemporal hemianopia** caused by pressure exerted on the optic chiasm.
3. **Right incongruous homonymous hemianopia** from a lesion of the left optic tract.
4. **Right congruous homonymous hemianopia** from a complete lesion of the left optic radiation.
5. Lesions of the primary visual cortex give rise to **dense, congruous hemianopic field defects.** Occlusion of the posterior cerebral artery supplying the occipital lobe is a frequent cause of total homonymous hemianopia. Some patients with hemianopia after

occipital stroke have **macular sparing**, because the macular representation at the tip of the occipital lobe is supplied by collaterals from the middle cerebral artery.

6. Destruction of both occipital lobes produces **cortical blindness.**

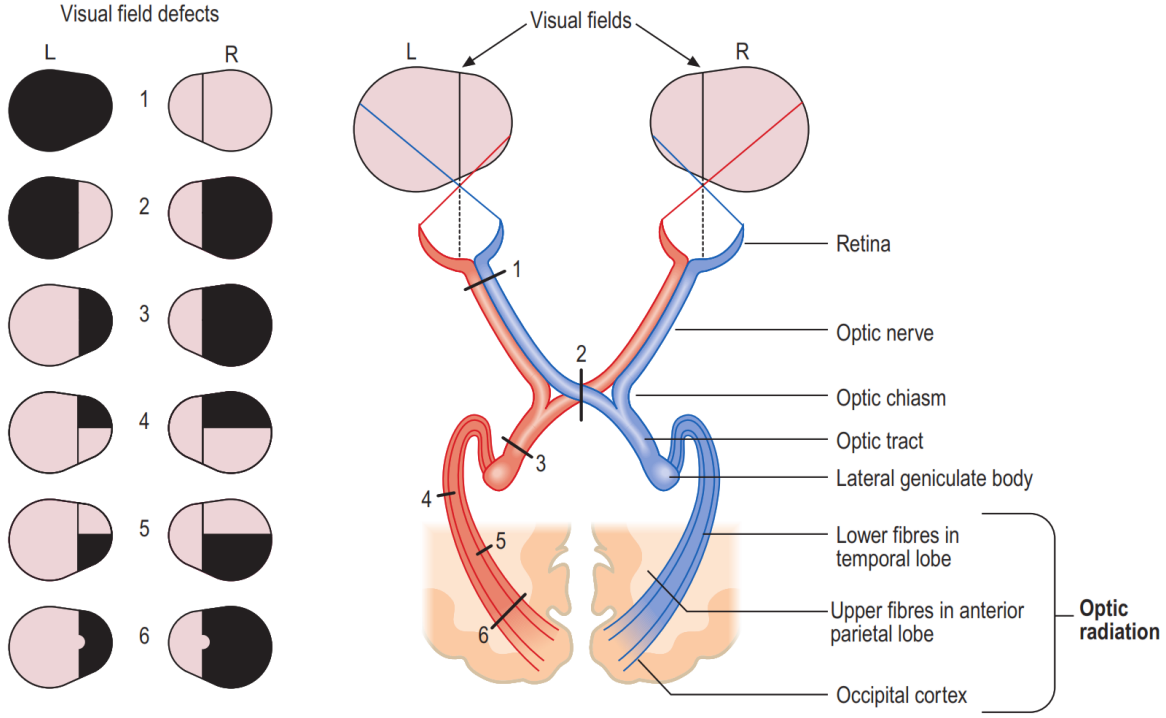


Fig. 12.3 Visual field defects. (1) Total loss of vision in one eye because of a lesion of the optic nerve. **(2)** Bitemporal hemianopia due to compression of the optic chiasm. **(3)** Right homonymous hemianopia from a lesion of the optic tract. **(4)** Upper right quadrantanopia from a lesion of the lower fibres of the optic radiation in the temporal lobe. **(5)** Lower quadrantanopia from a lesion of the upper fibres of the optic radiation in the anterior part of the parietal lobe. **(6)** Right homonymous hemianopia with sparing of the macula due to lesion of the optic radiation in the occipital lobe.

Characteristic clinical disturbance result from lesions of the second, third, fourth, and sixth cranial nerves:

Clinically the optic nerve behaves as though it were primarily carrying central visual information; therefore, lesions of the optic nerve may produce a combination of:

- Loss of visual acuity
- Diminished color perception
- Depression of the central field in a central scotoma
- Defective pupillary reaction to light.

A complete third nerve lesion causes ptosis and an inability to rotate the eye upward, downward or inward. When the lid is passively elevated, the eye is found to be deviated outward and slightly downward because of the unopposed intact actions of the lateral rectus and superior oblique muscles. Since the preganglionic parasympathetic fibers lie near the surface compressive lesions of the nerve dilate the pupil (surgical 3rd nerve palsy) but infarction of the central portion of the oculomotor nerve may spare the pupil (medical 3rd nerve palsy) as occur in diabetic opthalmoplegia.

Fourth nerve lesions result in extorsion and weakness of downward and inward movement of the affected eye, most marked when the eye is turned inward, so that the patient commonly complains of special difficulty in reading or going downstairs.
Head tilting to the opposite shoulder is especially characteristic of fourth lesion, this maneuver causes a compensatory intorsion of the unaffected eye, and ameliorate the double vision.
Lesions of the sixth nerve result in a paralysis of lateral or outward movement. With incomplete sixth nerve palsies turning the head toward the side of the paretic muscle may overcome diplopia.

Vision Loss

The visual pathways represent one third of the supratentorial brain mass and are frequently affected by structural lesions and a wide range of neurological disorders. The most important part of the history when evaluating a patient with visual loss is to establish whether the visual loss is:

- Monocular or binocular
- Painful or painless
- The temporal profile:
  - ✓ Onset (sudden, acute, subacute or chronic)
  - ✓ The course (transient or permanent).

Monocular visual loss results from lesions anterior to the chiasm (prechiasmal: the eye itself or the optic nerve), whereas binocular visual loss results from either bilateral prechiasmal lesions or, more likely, from a chiasmal or retrochiasmal lesion.

Diplopia

The majority of normal eye movements are conjugate, i.e. the two eyes move an equal distance in the same direction. Most cases in which an abnormality of eye movement is the primary neurological problem involve an acquired weakness of one of the extraocular muscle supplied by the oculomotor nerves 3rd, 4th and 6th cranial nerves that results in diplopia.

The primary function of the extraocular muscle and the optomotor system in general is to position the globes so that the object of regard is seen as a single image in the visual space regardless of the fact that it is viewed simultaneously by two eyes. Every point in the visual space has a corresponding retinal location in either eye. Squint and diplopia are features of nuclear and infranuclear lesion.

Clinical approach to a patient with diplopia

The most important part of the history when the evaluating a patient with diplopia is to establish whether the diplopia is:

1. Binocular or monocular
2. Horizontal or vertical or oblique
3. The temporal profile: onset and the course (fluctuating or constant).

Examination

1. The resulting diplopia can be analyzed by **measuring the direction of maximal image separation and the distance between the two images**, which is a direct measure of the angle at which the two visual axes diverge. Deviation of the eyes relative to one another is greatest when the patient attempts to look in the direction in which the weak muscle exerts its primary or maximal action.

2. **The cover-uncover test** is useful to localized the affected eye by disappear – reappear of one of the two image. The false image is the peripheral (outer) image. False image is from the affected (paralyzed) eye.

Nystagmus

Nystagmus: a repetitive, to and fro movement of the eye. It may consist of smooth oscillation of approximately equal velocity and amplitude **(pendular nystagmus)** or it may consist of alternating slow drifts (slow phase) in one direction and corrective, resetting saccades (quick phase) in the other **(jerk nystagmus)**. Points should be determined for the description of nystagmus:

- The symmetry of movement speed (jerk or pendular).
- The direction of fast phase (horizantal,vertical,rotatary).
- The position of the eye which exacerbates or provokes nystagmus (right or left – upward or downward).
- The amplitude (fine or coarse).

Pendular nystagmus (ocular nystagmus): Ocular nystagmus due to a defective vision represents a searching movement of the eyes, seemingly made in a useless effort to find a fixation point.

Pathological jerk nystagmus arises most commonly from lesions most commonly located in:

1. Vestibular end organ or nerve
2. The nystagmogenic zone of the neuraxis, essentially the brainstem tegmentum and cerebellum.

Gabapentin and **memantine** considered as treatment for acquired forms of nystagmus.

Optic nerve disorders (optic neuropathy)

Types of optic neuropathy:

1. Inflammatory
2. Vascular : ischemic optic neuropathy
3. Compressive/infiltrative
4. Toxic/nutritional
5. Hereditary
6. Traumatic
7. Elevated intracranial pressure
8. Elevated intraocular pressure

Classic Features

1. Decreased visual acuity
2. Visual field defect
3. Relative afferent pupillary defect
4. Can see the optic disc in the fundus by the funduscopic (ophthalmoscopic) examination
5. Swollen or shrinkage optic nerve (change the shape of optic disc: color, margin and vessels)

Optic Neuritis

This is a common inflammatory disorder of the optic nerve. Females more than males. Ocular pain (especially with eye movements) very common.

Optic Neuritis is the general term used to describe involvement of the optic nerve. When there is swelling of the optic disc the terms **anterior optic neuritis or papillitis** have been used. When the clinical history and examination suggest optic neuritis but the optic disc appears completely normal, the term **retrobulbar neuritis** is used.

Optic neuritis is usually idiopathic but is occasionally caused by contiguous inflammation of the meninges, orbital, or paranasal sinuses and may be also be caused by tuberculosis, syphilis and fungus. Optic neuritis is a common manifestation of multiple sclerosis but in a patient who has no past history of neurological dysfunction the diagnosis of multiple sclerosis is not justified.

The majority of cases occurs in patients 20 to 40 years old and is unilateral. In patients less than 20 years of age optic neuritis is commonly bilateral.

Visual loss may develop very rapidly but usually progresses for several days to weeks and an aching orbital pain exacerbated by eye movements is common.

Treatment with high dose intravenous methylprednisolone makes the recovery of visual function occurs more rapidly. Vision usually begins to improve by the end of 2 weeks and most patients experience good visual recovery.

Virtually all patients experience a gradual recovery of vision after a single episode of optic neuritis, even without treatment. This rule is so reliable that failure of vision to improve after a first attack of optic neuritis casts doubt upon the original diagnosis.

Papilledema

This connotes bilateral optic disc swelling from raised intracranial pressure. Headache is a frequent, but not invariable accompaniment.

Transient visual obscurations are a classic symptom of papilledema. They can occur in only one eye or simultaneously in both eyes. They usually last seconds but can persist longer if the papilledema is fulminant. Obscurations follow abrupt shifts in posture or happen spontaneously. When obscurations are prolonged or spontaneous, the papilledema is more threatening.

Visual acuity is not affected by papilledema unless the papilledema is severe, long-standing, or accompanied by macular edema and hemorrhage.

Vision is not affected initially, but the blind spot is enlarged. If the intracranial pressure is not reduced, secondary optic atrophy and loss of vision eventually occur.

Optic atrophy

Optic atrophy is not a disease. It is a morphologic sequel of any disease that causes damage to ganglion cells and axons (first order neuron). Optic atrophy developed 4-6 weeks after the onset in visual loss.

The term optic atrophy is therefore a pathologic generalization applied to optic nerve shrinkage from any process that produces degeneration of axon in the anterior visual system (the retrogeniculate pathway), including ischemia, inflammation, compression, infiltration and demyelination.

The common features of all varieties of optic atrophy, is pallor of the optic disc and loss of visual acuity. Visual loss is roughly proportional to the degree of nerve atrophy. Total blindness with a pupil that is unreactive to light could be seen.

Optic atrophy is a sign of chronic optic nerve disease and not a diagnosis in itself, it demands search for an etiology.

Optic atrophy can be divided into primary and secondary. **Primary optic atrophy** follow acute or chronic lesion of the optic nerve or retina. The disc is chalk- white with cookie-cutter sharp borders. **Secondary optic atrophy** follows papilledema. The disc is gray with shaggy borders from connective tissue proliferation.