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D.A. Jacobs and S.L. Galetta

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REVIEW ARTICLE

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S.L. Galetta

SUMMARY: Combining an understanding of neuro-ophthalmologic anatomy with proper imaging techniques provides a powerful method to detect lesions involving the afferent and efferent visual pathways. Precise documentation of the extent of injury within the nervous system is becoming increasingly important to assess and monitor the effect of neurologic therapies. This review will focus on those common neuro-ophthalmologic problems that have exquisite localizing value on neuro-imaging.

The Retina and Optic Nerve

The pathway for afferent visual information begins in the retina. An important retinotopic organization is preserved throughout the course of the afferent visual pathways. The nasal retina receives information from the temporal field and the temporal retina information from the nasal field. The superior retina receives information from the inferior field and the inferior retina from the superior field. This upside-down and backward information is conveyed via the remainder of the afferent pathway.

The optic nerve head is formed from a coalescence of 1 million axons from the retinal ganglion cells. The optic nerve is divided into the intraocular, intraorbital, intracanicular, and intracranial segments. Patients with optic neuropathies may present with reduced acuity that is typically unilateral. Dyschromatopsia, visual field defects, and an afferent pupillary defect may be associated findings. A variety of visual field defects may be seen in optic neuropathies, including central, centrocecal, arcuate, altitudinal, and nasal step field defects.[1] An altitudinal visual field defect is suggestive of ischemic optic neuropathy but may also be seen in idiopathic optic neuritis.[2] Ischemic optic neuropathy is the result of posterior ciliary artery occlusion. Most cases of ischemic optic neuropathy are nonarteritic and related to a congenitally small optic cup and other vascular risk factors. Approximately 10% of patients with ischemic optic neuropathy may have underlying giant cell arteritis. In both nonarteritic and arteritic ischemic optic neuropathy, neuroimaging is usually normal. On rare occasion, enhancement of the optic nerve will be observed in giant cell arteritis.[3] The time course and clinical history may help to determine the etiology of the optic neuropathy. Ischemic optic neuropathy will present abruptly, typically in an older age group, and is usually painless. The prognosis for visual recovery is poor. On the other hand, demyelinating optic neuritis tends to occur in a younger patient population and is commonly associated with periorbital pain.[4] Although the vision loss is acute, it tends to progress over hours to days. In contrast to the poor visual recovery seen in ischemic optic neuropathy, the prognosis for recovery in optic neuritis is excellent and typically occurs within several weeks to months. Inflammatory optic neuropathy is the term used to broadly describe any inflammation of the optic nerve, resulting in vision loss. The differential diagnosis is extensive and includes demyelinating disease, sarcoidosis, orbital pseudotumor, infectious etiologies such as Lyme disease, and rheumatologic conditions, including lupus.

Orbital MR imaging may be helpful in demonstrating lesions in inflammatory optic neuropathies. In optic neuritis, axial T1-weighted postcontrast MR imaging may reveal enhancement along the periphery or diffusely within the nerve. However, peripheral enhancement may be seen in multiple diseases, including optic nerve meningioma, idiopathic inflammatory pseudotumor, sarcoidosis, leukemia, lymphoma, perioptic hemorrhage, metastatic disease, and may even be seen in normal patients.[5] The use of short-term inversion recovery (STIR) and fluid-attenuated inversion recovery (FLAIR) sequences may allow for detection of signal intensity abnormality within the inflamed optic nerve.[6-8] In fact, more than 80% of patients with optic neuritis will have signal intensity abnormality within the optic nerve in STIR sequences with fat-suppressed views of the orbit and orbital surface coil.[9] Clinically silent white matter lesions within the brain are often seen at the time of presentation with optic neuritis. In the Optic Neuritis Treatment Trial (ONTT), 59% of patients had cerebral white matter lesions.[10] Other studies have demonstrated that 48%–70% of patients with isolated optic neuritis will have brain lesions consistent with demyelination.[11-15] The presence or absence of these lesions is the critical point in determining the risk for the subsequent development of clinically definite multiple sclerosis (MS). In the ONTT, by 10 years, 56% of patients with 1 or more lesions developed MS versus 22% of patients with a normal results on MR imaging.[16] The characteristic MS lesions are periventricular in location and ovoid in shape, and they may or may not be associated with enhancement, which is often ring-enhancing. Patients with optic neuritis and white matter abnormalities are often administered a course of corticosteroids followed by the initiation of immunomodulatory therapy. Clinicians now use serial MR imaging scans (every 6 months to yearly) to establish the diagnosis of MS and to monitor the effects of therapy, based on the McDonald diagnostic criteria for MS.[17]

Other disorders of the optic nerve include papilledema, pseudotumor cerebri, and inflammatory and compressive optic neuropathies. Papilledema is bilateral optic disk swelling in the setting of increased intracranial pressure. The patient may have enlargement of the blind spot or transient visual obscurations (visual impairment lasting seconds) associated with postural changes. The etiology of the disk swelling is thought to be a disruption of axonal transport. Patients with true papilledema require urgent neurologic evaluation and brain MR imaging (or CT if the MR imaging is not immediately accessible). For those patients without a mass lesion or hydrocephalus, attention should be paid to the venous circulation to rule

From the Department of Neurology, University of Pennsylvania, Philadelphia, Penn.

Address correspondence to Dina A. Jacobs, University of Pennsylvania Department of Neurology, 3400 Spruce St, Three West Gates Building, Philadelphia, PA 19104; e-mail: daj37@mail.med.upenn.edu

Table 1: Localization of field defects and disorders of higher cortical visual function

Field Defect or Syndrome	Localization
Unilateral central scotoma	Optic nerve
Bitemporal hemianopsia	Chiasm
Junctional defect (ipsilateral central scotoma and a contralateral superior temporal field cut)	Anterior chiasm
Central temporal scotomas	Posterior chiasm
Incongruous homonymous hemianopsia, afferent pupillary defect, and bow-tie atrophy	Optic tract
Homonymous sectoranopia	Lateral geniculate nucleus
Incongruous homonymous hemianopsia	Lateral geniculate nucleus
Homonymous upper quadrant defect "pie in the sky"	Temporal lobe
Homonymous defect, denser inferiorly	Parietal lobe
Gerstmann syndrome and a homonymous defect, denser inferiorly	Parietal lobe
Complete homonymous hemianopsia	Not well-localized
Homonymous upper quadrantanopsia with macular sparing	Occipital lobe (lower bank)
Homonymous lower quadrantanopsia with macular sparing	Occipital lobe (upper bank)
Isolated homonymous defect (macular sparing) without other neurologic findings	Occipital lobe
Anton syndrome (cortical blindness)	Bilateral occipital lobe lesions
Balint syndrome	Bilateral occipitoparietal lesions
Alexia without agraphia	Left occipital lobe and angular gyrus
Central achromatopsia	Bilateral occipito-temporal lesions

out venous thrombosis as the cause of increased intracranial pressure. Pseudotumor cerebri (idiopathic intracranial hypertension) is an elevation of intracranial pressure in the absence of a mass lesion. It may be seen in the setting of venous thrombosis, systemic disorders such as anemia, certain medications (including steroids and tetracyclines), spinal cord tumors, and in obese young women. The most important concern for the neuroradiologist is to exclude sinus thrombosis. In addition, it may also be important to evaluate the spine for those patients who do not fit the typical profile of pseudotumor cerebri to exclude a compressive lesion that either blocks CSF flow or leads to increased CSF protein.

Compressive or infiltrative optic neuropathies may present with unilateral or bilateral vision loss (depending on the extent of the lesion), reduced visual acuity, color vision loss, afferent pupillary defect, and optic nerve pallor. The compressive lesion may be a tumor or infiltrative process such as sarcoid or other inflammatory condition. Idiopathic orbital inflammatory syndrome (IOIS) is a rare disorder that causes localized inflammation within the orbit, resulting in pain, proptosis, and ophthalmoparesis. By definition, orbital pseudotumor is a diagnosis of exclusion, and it is important to exclude other entities, such as sarcoid, orbital cellulitis, thyroid ophthalmopathy, lymphoma, and rheumatologic conditions, such as Wegner granulomatosis. Contrast-enhanced MR imaging with multiple coronal views and fat saturation should be performed in those patients with suspected orbital pseudotumor or compressive optic neuropathies. In a study by Atlas et al,[18] the MR imaging was abnormal in all patients with a confirmed diagnosis of orbital pseudotumor, revealing mass lesions that were hypointense to orbital fat on T1-weighted images and isointense or minimally hyperintense to fat on T2-weighted images. Although this appears very different from the typical appearance of hemorrhage or malignancy, it can be quite similar in cases of meningioma, lymphoma, and sarcoidosis.[18,19] The radiologic appearance of IOIS consists of enlargement of multiple muscles, irregular borders with extension to the orbital fat, and enhancement around the globe. Thyroid eye disease will also have enlargement of multiple muscles, but the borders are more regular, and there is sparing of the tendons with more profound involvement of the belly of the muscle. The inferior and medial recti are typically the earliest and most severely affected muscles in thyroid eye disease; the lateral rectus muscle is rarely involved in this disease. In addition, the process does not extend to the orbital fat. Finally, cellulitis will be demonstrated by a decrease in signal intensity of the orbital fat; bony erosion, sinus disease, and venous thrombosis may also be seen.

Chiasmal and Retrochiasmal Pathways

The nature of the field defect may help with the localization of the abnormality and direct the radiologic search (Table 1). The fibers of the nasal retina (temporal field) of the optic nerves join at the chiasm. Thus, a bitemporal hemianopsia is the characteristic finding in a chiasmal lesion. The most common etiology is a pituitary tumor. Other etiologies include craniopharyngioma, meningioma, aneurysm, glioma, inflammation, MS, and ischemia, as can be seen with lupus or giant cell arteritis. There can be variations in chiasmal syndromes, depending on chiasmal anatomy (prefixed or postfixed pituitary) and the location of the lesion (anterior, medial, or posterior). The bitemporal hemianopsia is seen in lesions that affect the central portion of the chiasm. A lesion that is located more anteriorly will affect the ipsilateral optic nerve fibers and the contralateral fibers of the Wilbrand knee (the crossing of the temporal retinal fibers). This results in an ipsilateral central scotoma and a contralateral superior temporal field cut, and is also called a junctional chiasmal syndrome. A posterior chiasmal syndrome may result in centrally placed temporal scotomas. It is important to recognize whether the superior or inferior temporal field is most impaired. A lesion located near the third ventricle will result in a bitemporal defect that is denser inferiorly. On the other hand, a sellar lesion is more likely to cause involvement of the superior temporal fields.[20]

Proceeding retrochiasmally, the fibers of the optic tract synapse on the lateral geniculate body of the thalamus. The field defect associated with an optic tract lesion is typically an incongruous homonymous hemianopsia. An incongruous defect is one that asymmetrically involves the 2 eyes. There may be an afferent pupillary defect on the side of the greater field

loss (contralateral to the lesion) and optic atrophy. The combination of a homonymous hemianopsia and an afferent pupillary defect is the hallmark of an optic tract lesion. The optic atrophy will occur temporally in the eye ipsilateral to the tract lesion, and a bow-tie configuration in the contralateral eye. The optic tract fibers travel above and around the infundibulum and below the third ventricle. The blood supply of the optic tract is supplied by thalamic perforators of the posterior cerebral artery and branches of the anterior choroidal artery off the internal carotid artery. The etiology of an optic tract defect includes aneurysm, ischemia, pituitary tumor craniopharyngioma, sarcoid, and MS. The fibers of the optic tract then synapse in several different locations: the primary visual pathway synapses in the lateral geniculate body, the pupillomotor pathway synapses in the pretectum, and the subcortical visual pathway synapses in the superior colliculus. The lateral geniculate nucleus (LGN) has 6 layers. Layers 2, 3, and 5 receive visual input from the retinal ganglion cells from the ipsilateral temporal hemiretina or the nasal field. Layers 1, 4, and 6 receive information from the nasal retina or the temporal field. The vascular supply of the LGN is derived from the anterior and posterior choroidal arteries. Lesions of the LGN may result in congruous or incongruous field defects. If the posterior choroidal artery is affected, the result will be a congruous horizontal sectanopia. In contrast, lesions involving the anterior choroidal artery produce a quadruple quadrantanopsia (the horizontal sector is spared).

The optic radiations are formed by the geniculocalcarine fibers leaving the LGN on their way to the calcarine cortex. The temporal radiations travel anteriorly around the lateral ventricle and form Meyer's loop. These fibers contain information from the superior visual fields. A temporal lobe lesion will result in a superior quadrantanopisa (a "pie-in-the-sky" defect). The parietal radiations carry the information from the inferior visual fields. Parietal lobe lesions will result in a homonymous defect that is denser inferiorly. Other associated symptoms may include neglect, constructional apraxia, and a Gerstmann syndrome (acalculia, agraphia, finger agnosia, and right-left confusion). A complete homonymous hemianopsia cannot be well-localized in the retrochiasmal pathways.

The optic radiations will terminate in the occipital lobe or the primary visual cortex. Lesions in the occipital lobes result in congruous defects. The occipital cortex is retinotopically organized based on superior-inferior anatomy. A lesion of the lower bank of the calcarine cortex will cause a contralateral congruous superior defect respecting both the horizontal and vertical meridians.[18] In addition, a homonymous hemianopsia with macular sparing and preservation of the temporal crescent is unique to the occipital lobe. Isolated homonymous defects without other neurologic findings, such as weakness, neglect, or aphasia, are most likely to be due to an occipital lobe lesion.

Several disorders of higher cortical visual functioning are worth mentioning, in that they can also aid in localization (Table 1). Bilateral occipital lobe lesions will result in cortical blindness. Patients with cortical blindness who are unaware of their visual loss have Anton syndrome. Occipital lobe lesions may also result in palinopsia, or a perseveration of the visual image once the stimulus has been removed. Bilateral occipitotemporal lesions have been associated with prosopagnosia (an inability to recognize familiar faces) and achromatopsia (an abnormality of color perception). Balint syndrome is a triad of simultanagnosia, optic apraxia, and ocular ataxia (visual misreaching). The lesion is located in bilateral parieto-occipital lobes or the visual association cortex. Alexia without agraphia is the ability to write but not read, and other language functions are intact. The syndrome is a result of a disconnection between the visual cortex from the language area or angular gyrus. The causative lesion is damage to the left occipital lobe and splenium of the corpus callosum. The visual information from the intact right occipital lobe is unable to reach the language areas. Complex visual hallucinations may be seen in the blind hemifield as a release phenomenon or as a seizure. The hallucinations are characteristically unformed when lesions are in the occipital cortex, and formed when derived from the temporal cortex, but there can be a lot of variability.

Table 2: Horner syndrome localization

Associated Symptoms	Consideration
Isolated, painful	Carotid dissection, cluster headache
Sensory level	Spinal cord
Arm numbness or weakness	Brachial plexus
Ipsilateral face and contralateral body numbness	Medulla
Sixth-nerve palsy	Cavernous sinus

Pupillary Abnormalities

The Marcus-Gunn pupil, or afferent pupillary defect, is seen in unilateral optic neuropathies but may also be observed in chiasmal and optic tract lesions. The affected pupil does not react to light as briskly as the unaffected pupil because of a decrease in afferent input reaching the midbrain (pretectal pathway responsible for the pupillary light response). Horner syndrome is a result of an interruption of the sympathetic input to the eye. The pathway begins in the hypothalamus and descends down the lateral brain stem to synapse in the intermediolateral cell column of the spinal cord at C8–T1 (Table 2). The second-order neuron then synapses in the superior cervical ganglion. The third-order neuron travels via the internal carotid artery into the cavernous sinus, following the nasociliary nerve into the orbit. Horner syndrome is characterized by miosis, with an increase in anisocoria in the dark because of the interrupted sympathetic pathway, ptosis of the upper and lower lids, variable anhydrosis, and slow pupillary redilation in the dark. If the lesion is congenital, there will be a heterochromic iris because of impairment of the pigmentary changes.

An Adies-Tonic pupil is typically idiopathic and benign and is a result of the interruption of the parasympathetic pathways. The clinical features are a dilated pupil with poor or absent response to light but preserved near response. It is more common in women and may be seen in association with absent reflexes as part of the Holmes Adies syndrome. It is most likely viral in origin. Imaging is usually normal. Argyll-Robertson pupils may be seen in diabetes or syphilis. Pupils are small and irregular with impaired light response and intact near response. The lesion is likely in the dorsal midbrain with interruption of the light reflex pathways and preservation of the near response. Pupillary abnormalities seen with third nerve palsies will be covered later.

Efferent Abnormalities

The third cranial nerve (oculomotor) complex is located in the dorsal midbrain, anterior to the cerebral aqueduct. It innervates the superior rectus, inferior rectus, inferior oblique, medial rectus, pupillary sphincter, and levator palpebrae. There are subnuclei that supply the individual muscles. The central caudal nucleus actually innervates both levator muscles. The superior rectus receives information from the contralateral superior rectus subnuclei, but the rest of the innervation of the third nerve is ipsilateral. The nuclei converge to give rise to the fascicle that travels through the ventral midbrain and exits near the medial aspect of the cerebral peduncle. It enters the subarachnoid space coursing medial to the posterior communicating artery. It is susceptible here to compression in the setting of uncal herniation secondary to cerebral edema or a mass lesion. It then courses through the lateral wall of the cavernous sinus superior to the fourth cranial nerve. As the nerve enters the orbit, it separates into the superior and inferior divisions. The superior division innervates the levator palpebrae and superior rectus, whereas the inferior division innervates the papillary sphincter, medial and inferior recti, and the inferior oblique muscle. However, divisional third nerve palsies may occur all the way back to the midbrain.

A lesion of the third nerve nucleus is exceedingly rare but will result in an ipsilateral third nerve palsy with contralateral superior rectus weakness and bilateral partial ptosis, because the central caudal nucleus innervates both levator muscles. The best way to localize the oculomotor palsy is by the associated features. The association of a third nerve palsy with a hemiparesis would localize to the anterior midbrain involving the cerebral peduncle and third nerve fascicle (Weber syndrome). Benedikt syndrome is a lesion of the third nerve fascicle and the red nucleus and results in a third nerve palsy and contralateral tremor. If multiple cranial nerves are involved, then a diffuse process involving the subarachnoid space would be implicated, such as meningitis, or an inflammatory condition such as sarcoid. If there are concomitant fourth and sixth nerve palsies, then the pathology would arise from the cavernous sinus. All of the above with an optic neuropathy would implicate the orbital apex. Isolated pupil-involving third-nerve palsy is most likely due to an aneurysm of the posterior communicating artery as it arises from the internal carotid artery. On the other hand, an isolated pupil-sparing third nerve palsy would be most commonly due to microvascular ischemia (Table 3).

The fourth cranial nerve (trochlear) nucleus is also located in the midbrain, and it supplies the contralateral superior oblique muscle. The nerve crosses near the roof of the aqueduct and exits the brain stem dorsally through the anterior medullary velum. It is here that it is very susceptible to damage from trauma, which is the most common cause of a fourth nerve palsy. In addition, it has the longest course of any of the oculomotor nerves. The fourth nerve then travels through the cavernous sinus en route to the orbit.

The sixth cranial nerve (abducens) nucleus lies dorsally in the pons near the genu of the seventh cranial nerve or the region of the facial colliculus. The nucleus innervates the ipsilateral lateral rectus and sends interneurons to the medial longitudinal fasciculus (MLF) that will go on to innervate the contralateral medial rectus to coordinate horizontal gaze. The fascicle travels ventrally and exits the pontomedullary junction anterolaterally. It also travels through the cavernous sinus on the way to the orbit. However, it floats freely in the cavernous sinus lateral to the internal carotid artery, as opposed to the third and fourth nerves, which are in the lateral wall of the sinus. Because of its proximity to the cavernous carotid artery, it may be the first sign of a carotid cavernous fistula. A lesion of the sixth nerve nucleus causes a conjugate gaze palsy because it affects the interneurons that travel to the MLF. The facial colliculus syndrome is a peripheral facial palsy and conjugate gaze palsy. Bilateral sixth nerve palsies may be seen in the setting of increased intracranial pressure as a false localizing sign, in the setting of meningitis, or in a skull-based tumor. A "pseudosixth" nerve palsy may be seen in thyroid eye disease or in myasthenia gravis. The etiology of a sixth nerve palsy is age-dependent; in younger patients, the likely culprits are tumor, demyelination, and the effects of viral infection; in older patients, it is more likely to be vascular in cause.

Table 3: Localization of motility abnormalities

Motility Disturbance	Localization/Etiology
Weber syndrome (Third-nerve palsy and hemiparesis)	Anterior midbrain
Benedikt syndrome (third-nerve palsy and contralateral tremor)	Red nucleus and third-nerve fascicle
Isolated pupil-involving third-nerve palsy	Posterior communicating artery aneurysm
Pupil-sparing third-nerve palsy	Microvascular ischemia of the third nerve
Isolated fourth-nerve palsy	Doral midbrain/anterior medullary velum
	Microvascular ischemia
Isolated sixth nerve palsy	Pons or sixth-nerve fascicle
	Demyelination/microvascular ischemia
Gaze palsy and facial weakness	Dorsal pons/facial colliculus
Bilateral sixth-nerve palsies	Elevated intracranial pressure
Third-, fourth-, and sixth-nerve palsies	Cavernous sinus
Third-, fourth-, sixth-nerve palsies, and optic neuropathy	Orbital apex
Multiple cranial neuropathies	Subarachnoid space
Internuclear ophthalmoplegia	Medial longitudinal fasciculus
Gaze palsy	Dorsal pons
Parinaud syndrome (upgaze palsy, eyelid retraction)	Dorsal midbrain

Supranuclear and Internuclear Motility Problems

A lesion of the medial longitudinal fasciculus results in an ipsilateral adduction deficit and a contralateral abducting nystagmus, referred to as an internuclear ophthalmoplegia (INO). Vertical nystagmus may also be seen, particularly when there is bilateral MLF involvement, as can be seen in demyelinating disease. The paramedian pontine reticular formation (PPRF) is also responsible for ipsilateral horizontal conjugate gaze, in particular quick eye movements such as horizontal saccades. An isolated lesion of the PPRF can be distinguished from a sixth-nerve nuclear lesion in that the gaze palsy will be overcome by the oculocephalic maneuver. A lesion that involves the sixth-nerve nucleus near the facial colliculus produces an ipsilateral gaze palsy. The interneurons of the MLF are intermixed with the abducens neurons in the sixth-nerve

nucleus. A one-and-a-half syndrome is a caused by a lesion to either the sixth-nerve nucleus or the PPRF (or both) and the MLF, resulting in an ipsilateral conjugate gaze paresis and an ipsilateral INO.

A skew deviation is a vertical misalignment resulting in vertical diplopia and a hypertropia. It is the result of a lesion in the pathways between the medullary vestibular nuclei and the third and fourth cranial nerve nuclei in the midbrain. Medullary lesions will result in a hypertropia (higher eye) contralateral to the lesion and midbrain, or pontine lesions will result in hypertropia ipsilateral to the lesion. The ocular tilt reaction combines a skew deviation with a conjugate ocular torsion and a head tilt. It may be seen in association with lesions of the midbrain, pons, or medullary vestibular regions.

The dorsal midbrain syndrome, or Parinaud syndrome, is the result of a lesion in the posterior commissure. The clinical features are lid retraction, convergence-retraction nystagmus, pupillary light-near dissociation (the pupils respond to near stimuli but not light), and an upgaze paresis that may be overcome by the oculocephalic maneuver. The patient may present with diplopia, blurred vision at near distances, and difficulty looking up. Other associated features include skew deviation, INO, and convergence insufficiency.

Nystagmus

Nystagmus is a rhythmic oscillating movement of the eyes. Two types of nystagmus may be seen. The first is a jerk nystagmus in which there is a slow phase and a fast "jerk" phase in the opposite direction. Pendular nystagmus consists of only one phase of movements in a sinusoidal pattern. Nystagmus may either congenital or acquired. Congenital nystagmus will present shortly after birth. It is horizontal, even in upgaze and downgaze. Patients will exhibit a head turn to move their eyes to the "null position," which will dampen the amplitude of the nystagmus. In addition, it may be dampened by convergence. Spasmus nutans is a congenital nystagmus that typically presents between 4 and 14 months of age and disappears by age 5. It is associated with a clinical triad of head nodding, head tilt, and monocular nystagmus. Although spasmus nutans is typically a benign condition, it is important to exclude a chiasmal glioma, which is typically seen in association with vision loss.

There are several types of acquired nystagmus, which can be well-localized within the central nervous system (Table 4). Seesaw nystagmus may be seen in lesions involving the midbrain and parasellar region (pituitary tumor or craniopharyngioma). It appears clinically as a conjugate pendular elevation and intorsion of one eye with depression and extorsion of the opposite eye. In the setting of parasellar lesions, a bitemporal hemianopsia may also be seen. Downbeat nystagmus is seen in disorders affecting the cervicomedullary junction. Conditions causing downbeat nystagmus include Arnold-Chiari malformations, skull base tumors, spinocerebellar degenerations, and toxic metabolic conditions such as medication toxicity (specifically phenytoin, carbamazepine, and lithium). Periodic alternating nystagmus is another condition localizing to the cervicomedullary junction. It is characterized by a horizontal jerk nystagmus in one direction for 90 seconds, no nystagmus for 10 seconds, and then recurrent horizontal jerk nystagmus in the opposite direction for 90 seconds. It may be seen in association with a downbeat nystagmus. Dissociated nystagmus may be seen in lesions involving the medial longitudinal fasciculus, as is most commonly seen in internuclear ophthalmoplegias. It appears as a nystagmus of the abducting eye, which appears to be a corrective phase to "catch up" with the contralateral adducting eye. Convergence retraction nystagmus has been described in the setting of Parinaud syndrome and is associated with dorsal midbrain lesions.

Table 4: Localization of nystagmus

Nystagmus	Location
Spasmus nutans	Congenital or chiasm
Seesaw	Midbrain/parasellar region
Downbeat	Cervicomedullary junction
Periodic alternating	Cervicomedullary junction
Dissociated	Medial longitudinal fasciculus
Rebound	Cerebellum
Convergence retraction nystagmus	Dorsal midbrain
Oculopalatal myoclonus	Mollaret triangle (central tegmental tract)
Brun nystagmus	Cerebellopontine angle
Gaze evoked	Vestibular, cerebellum
Upbeat	Pontomesencephalic or pontomedullary junction and cerebellum
Torsional (pure)	Central vestibular or cerebellum

Oculopalatal myoclonus is a pendular horizontal nystagmus seen in association with rhythmic elevation of the palate at the same frequency of 1–3 Hz. Damage to the pathways connecting the inferior olive, dentate nucleus of the cerebellum, and red nucleus, or Mollaret triangle, results in oculopalatal myoclonus typically months after the initial injury, once olivary hypertrophy occurs. Etiologies include stroke, MS, head trauma, and hemorrhage. Gaze-evoked nystagmus that is pathologic may be seen ipsilateral to a lesion of the brain stem or cerebellum and contralateral to a peripheral vestibular pathway lesion. Brun nystagmus is an ipsilateral gaze paretic and contralateral high frequency, low amplitude nystagmus that can be seen in association with cerebellopontine angle tumors.

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