Nystagmus

- Nystagmus has traditionally been divided into two types on the basis of the clinical impression of the waveform. Thus, if the eyes appeared to oscillate with "equal speed" in either direction, it was called "pendular" nystagmus; if movement in one direction was faster than in the other, it was called "jerk" nystagmus.

- True pendular nystagmus is sinusoidal, whereas jerk nystagmus has a slow phase away from the object of regard, followed by a fast (saccadic) phase toward the target. The direction of the fast component, by convention, defines the nystagmus direction.
Nystagmus

Nystagmus should be described not only by its waveform and direction but also by its amplitude (A) and frequency (F), the product of which is intensity (I). The examiner should also note the positions of gaze in which the nystagmus occurs and whether the intensity changes with gaze direction. Jerk nystagmus is usually accentuated in amplitude upon gaze in the direction of the fast component, a characteristic referred to as Alexander's law.
The field of gaze in which nystagmus intensity is minimal is termed the "null zone".

The "neutral zone" is that eye position in which a reversal of direction of jerk nystagmus occurs and in which no nystagmus, any of several bidirectional waveforms, or pendular nystagmus is present. The null and neutral zones usually overlap; however, several cases have been recorded where they do not.
High gain instability

In some persons, because of abnormally high gain in the SEM subsystem, a runaway (increasing velocity) movement or a pendular oscillation is evoked. In this chapter, the term "high gain" can also imply excessive delay for the gain present (i.e., the control loop may have a normal gain but an increased delay). Control theory suggests how particular changes in gain can result in either a pendular or a jerk nystagmus.

- Pendular nystagmus can be congenital or acquired, whereas horizontal jerk nystagmus with slow phases of increasing velocity usually is congenital; however, the latter may result from an Arnold-Chiari malformation.
- Vertical nystagmus with an exponential slow phase of increasing velocity may be secondary to acquired cerebellar disease.
Vestibular tone imbalance.

- The nystagmus of vestibular tone imbalance results from the imposition of asymmetric vestibular input on an inherently normal horizontal gaze generator. This asymmetric input occurs if one vestibular apparatus (labyrinths, nerve, or brain stem nuclei) functions abnormally or if both sides are asymmetrically defective. The nystagmus recording always shows a linear (straight line) slow phase, reflecting a persistent tone to drive the eyes toward the side of the relatively damaged vestibular apparatus. The slow-phase amplitude is reduced by fixation and enhanced by darkness, Frenzel (high-plus) lenses, or closing the eyes. Fixation inhibition may be related to an opposing smooth-pursuit force and requires the integrity of the cerebellar flocculus.
Integrator leak.

- Nystagmus caused by a "leaky integrator" occurs only in an eccentric gaze position; thus, it is gaze-evoked. The eyes are unable to maintain the eccentric position and drift back to the primary position with a decreasing velocity, reflecting a passive movement resisted by the viscous forces of orbital soft tissues. The defect may reside in the brain stem "neural integrator" or its connections (such as in the cerebellum), which mediates eye deviation. This form of gaze-evoked nystagmus is called "gaze-paretic" nystagmus.
The localizing significance of nystagmus

- The localizing significance of nystagmus is often a mere indication of dysfunction somewhere in the posterior fossa (i.e., vestibular end-organ, brain stem, or cerebellum). However, certain nystagmus patterns are quite specific and permit reasonably accurate neuroanatomic diagnosis. When possible, the specific and nonspecific forms are separated on the basis of clinical appearance and associated signs and symptoms.
There are several types of benign nystagmus usually seen in infancy. Congenital nystagmus (CN) is the most common infantile nystagmus. Others are latent/manifest latent nystagmus (LMLN) and the pendular nystagmus of spasmus nutans.
NYSTAGMUS IN INFANCY

CN is the direct result of an ocular motor control instability that may develop with or without an accompanying sensory deficit. Thus, for those cases in which a sensory deficit exists, it can only be a subordinate factor in the development of CN, perhaps interfering with the normal calibration of a key ocular motor subsystem and thereby precipitating its instability. The common association of "pendular" CN with a sensory defect and the "jerk" form with a primary motor abnormality is both simplistic and erroneous.
**Congenital nystagmus (CN)**

**TABLE 2. Characteristics of Congenital Nystagmus**

- Binocular
- Similar amplitude in both eyes
- Usually uniplanar (horizontal)
- Distinctive waveforms
- Diminished *(damped)* by convergence
- Increased by fixation attempt
- Superimposition of latent component
- "Inversion" of the optokinetic reflex
- Associated head oscillation
- No oscillopsia
- Abolished in sleep
The nystagmus blockage syndrome (NBS) is both a poorly understood and an overdiagnosed phenomenon related to CN. As the name suggests, the nystagmus of these patients diminishes or disappears with the act of willed esotropia while fixating a distant target.
Latent/Manifest Latent

- Latent/manifest latent nystagmus (LMLN) is a jerk nystagmus with either a linear or decreasing velocity exponential slow phase identical to that of gaze-paretic nystagmus. Occasionally, when both eyes are closed, a jerk nystagmus with a linear slow phase is present. Classically, "pure" or "true" latent nystagmus (LN) occurs only with uniocular fixation.

- There is no nystagmus with both eyes viewing, but when one eye is occluded, nystagmus develops in both eyes, with the fast phase toward the uncovered eye.

- LN is always congenital. However, several cases of manifest latent nystagmus (MLN) associated with retrolental fibroplasia have been recorded.
Acquired

- SECONDARY TO VISUAL LOSS. Nystagmus occurring in early childhood consequent to progressive bilateral visual loss should not be classified as CN unless CN waveforms are documented. The conceptual problems in the classification were discussed above. Usually, nystagmus secondary to visual loss cannot be distinguished from CN in a patient with coexisting primary visual abnormalities.
SPASMUS NUTANS

- Spasmus nutans is a rare constellation of ocular oscillation, head nodding, and torticollis that begins in infancy (usually between 4 and 18 months of age) and disappears in childhood (usually before 3 years of age). The nystagmus is generally bilateral (but it can differ in each eye and may even be strictly monocular), and it oscillates in horizontal, torsional, or vertical directions. An instance of spasmus nutans presenting with monocular nystagmus in monozygous twins has been reported.

- Spasmus nutans may sometimes be mimicked by tumors of the optic nerve, chiasm, or third ventricle.
Acquired pendular nystagmus may reflect brainstem or cerebellar dysfunction, or both. It occurs in patients with vascular or demyelinating disease. In the latter, it has been regarded as a sign of cerebellar nuclear lesions. The nystagmus is multivectorial (i.e., horizontal, vertical, diagonal, elliptic, or circular) and usually is associated with a head tremor. Marked dissociation between the two eyes often exists and may not correlate with differences in visual acuity from coexisting optic neuropathy. Despite the dissociation, the oscillations of the two eyes are phase locked, even when there is a difference in their frequencies.
Vestibular

We generally delimit vestibular nystagmus as being consequent to dysfunction of the vestibular end-organ, nerve, or nuclear complex within the brain stem. It is a horizontal-torsional or purely horizontal primary-position jerk nystagmus with a linear slow phase. The nystagmus intensity increases with gaze toward the fast phase (obeying Alexander's law); it decreases and, with central lesions, may reverse directions upon gaze toward the direction of the slow phase. The symptom of vertigo usually coexists. As might be expected, acute lesions of the cerebellar flocculus (the vestibulocerebellum) can produce a similar nystagmus. Cases of discrete cerebellar infarction are quite rare. Nystagmus may accompany episodic attacks of ataxia.
Gaze-Evoked (Gaze-Paretic) Nystagmus

- Gaze-evoked nystagmus is elicited by the attempt to maintain an eccentric eye position, and it is the most common form of nystagmus encountered in clinical practice. Patients recovering from a central gaze palsy show a phase in which lateral gaze movement is possible but cannot be maintained in the deviated position; that is, the eyes drift back slowly toward primary position.

- A corrective saccade repositions the eyes eccentrically, and repetition of this pattern produces nystagmus, aptly designated "gaze-paretic."
SPECIAL NYSTAGMUS TYPES

- Physiologic (End-Point)
- There are three basic types of nystagmus that are regarded as normal (physiologic) phenomena
Fatigue nystagmus

- begins during extended maintenance of an extreme gaze position and has been found in up to 60% of normals when horizontal gaze is maximally deviated for a time exceeding 30 seconds. It may become increasingly torsional with prolonged deviation effort and may be greater in the adducting eye. Fatigue nystagmus is not a clinically important phenomenon, because routine examinations do not include the maintenance of far eccentric gaze.
Unsustained end-point nystagmus

- is certainly the most frequently encountered physiologic nystagmus. Its incidence and characteristics have never been studied quantitatively. All experienced clinicians recognize that a few beats of nystagmus are within perfectly normal limits at gaze deviations of 30° or more.
Sustained end-point nystagmus

- Sustained end-point nystagmus begins immediately upon, or within several seconds of, reaching an eccentric lateral-gaze position. It has been found in more than 60% of normal subjects with horizontal-gaze maintenance greater than 40°.
Torsional

Torsional nystagmus describes a torsional movement of the globe about its anteroposterior axis; the term "rotary nystagmus" is used interchangeably. Most nystagmus consequent to vestibular end-organ dysfunction has a torsional component admixed with a major horizontal or vertical nystagmus. A purely torsional nystagmus never occurs with vestibular end-organ disease. When of small amplitude, torsional nystagmus may reflect a medullary lesion. Larger amplitude torsional nystagmus may be congenital, but when it is acquired it often indicates diencephalic (thalamic) involvement, in which case it is the underlying pattern in see-saw nystagmus.
See-saw nystagmus

See-saw nystagmus is characterized by a conjugate, pendular, torsional oscillation with a superimposed disjunctive vertical vector. The intorting eye rises and the opposite, extorting eye falls. Repetition of this sequence in the alternate direction provides the see-saw effect. The torsional movements predominate in all fields of gaze, but the see-saw feature may be restricted to the primary position or, more commonly, to downward or lateral gaze. See-saw nystagmus can be of the jerk type (with one phase being slow and the other fast) with unilateral, mesodiencephalic lesions.

Most patients with acquired see-saw nystagmus have bitemporal hemianopias consequent to large parasellar tumors expanding within the third ventricle. It is occasionally evoked transiently after blinks or saccades.
Convergence-retraction "nystagmus"

- Convergence-retraction "nystagmus" as a manifestation of the dorsal midbrain syndrome here, the initiating convergence movements are saccadic and thus not a true nystagmus. Fast divergent movements followed by a slow convergence, associated with epileptic electroencephalographic activity, occurred in a neonate with an intraventricular hemorrhage.
Periodic alternating nystagmus (PAN)

Periodic alternating nystagmus (PAN) is an extraordinary ocular motor phenomenon in which a persisting horizontal jerk nystagmus periodically changes directions. PAN may be congenital or acquired. The congenital variety, which may be associated with albinism, has the slow-phase waveform of an increasing-velocity exponential and usually lacks the well-defined stereotyped periodicity seen in acquired PAN (i.e., it is aperiodic alternating nystagmus). The periodicity of the congenital PAN is markedly influenced by changes in gaze position, supporting the hypothesis that the PAN is a result of a temporal shift in the null zone.
Downbeat

- Downbeat nystagmus is defined as nystagmus gaze position with the fast phase beating in a downward direction. Patients with brain stem disease or drug intoxications usually lack gaze-evoked downward nystagmus despite nystagmus in all other fields of gaze. Thus, nystagmus beating downward in the primary position is a striking phenomenon and is highly suggestive of a disorder of the craniocervical junction, such as Arnold-Chiari malformations.
The other major cause of downbeat nystagmus is spinocerebellar degeneration. Indeed, it is difficult to ascertain from the literature whether an Arnold-Chiari malformation or spinocerebellar degeneration is the most common cause. However, because the latter is correctable, a defect of the craniocervical junction must be carefully considered in all patients with downbeat nystagmus. Downbeat nystagmus may coexist with PAN, another type of nystagmus suggestive of an abnormality of the craniocervical junction. A variety of miscellaneous conditions have also been reported to produce downbeat nystagmus. These include anticonvulsant, alcohol, and lithium intoxication; magnesium deficiency; B12 deficiency; brain stem encephalitis; alcoholic cerebellar degeneration; dolichoectasia of the ventral artery; and vertebral artery occlusion.
Rebound

- Rebound nystagmus is either the diminution and direction change of gaze-evoked horizontal nystagmus during sustained ocular deviation or a horizontal gaze-evoked nystagmus that, on refixation to primary position, transiently beats in the opposite direction. The sign is often present in patients with cerebellar disease.
A paretic eye muscle, from whatever cause, can fatigue quickly during contraction, and muscle-paretic nystagmus can be observed. This is often evident as gaze-evoked nystagmus in myasthenia gravis, in which there is usually asymmetry between the two eyes. Another form of oscillation in myasthenia is "nystagmus" of the abducting eye (may be saccadic pulse trains, discussed below) coexisting with paresis of adduction; this mimics an internuclear ophthalmoplegia. Here the oscillation is not due to lateral rectus paresis but rather to excessive innervation by increased central gain, the result of paresis of the contralateral yoke medial rectus. Cessation of both muscle-paretic and contralateral yoke nystagmus in myasthenia usually follows administration of anticholinesterase medication.
Acoustic Neuroma

- Schwannomas of the eighth nerve grow so slowly that adaptive mechanisms often obscure clinical vestibular manifestations. Vestibular nystagmus beating contralateral to the lesion may be present, particularly if fixation is eliminated. As the tumor expands to compress the brain stem, a slow, gaze-evoked ipsilateral nystagmus is often added. The combination of a small-amplitude, rapid primary-position jerk nystagmus beating contralateral to the lesion and a slower, larger-amplitude, gaze-evoked (Bruns') nystagmus ipsilateral to the lesion also occurs with other extra-axial masses, including cerebellar tumors, compressing the brain stem. Rarely, Bruns' nystagmus is inverted.
Albinism and Achiasma

- Ocular albinism is associated with anomalous visual projections that result in a variety of eye movement disturbances, with considerable intersubject variability. These persons may have pendular or jerk nystagmus, absent OKN, "inverted" pursuit, or "defective" pursuit (see section on CN) when targets are projected onto the temporal half-retina. Periodic alternating nystagmus may also occur. Albinism can also exist in the absence of any nystagmus.

- Achiasma is a very rare condition, first documented in dogs, then in humans, that has associated with it the combination of horizontal CN and see-saw nystagmus. In this condition, all retinal fibers remain ipsilateral, passing to the ipsilateral lateral geniculates and visual cortices.
Lateral Medullary Syndrome

- The lateral medullary syndrome (Wallenberg) is a distinctive constellation of signs. The nystagmus in this syndrome tends to be stereotyped. With the eyes open there is horizontal-torsional jerk nystagmus beating contralateral to the lesion; when recorded with the eyes closed, the nystagmus beats ipsilateral to the lesion. Other rare manifestations, confined to single cases, are gaze-evoked eyelid and ocular nystagmus inhibited by the near reflex90 and horizontal gaze-evoked monocular downbeat nystagmus.
Upbeat

Primary-position nystagmus with the fast phase beating upward rarely reflects drug intoxication. Most often, the nystagmus is acquired and indicates structural disease, usually of the brain stem. The location of the lesions in patients with upbeat nystagmus after meningitis, Wernicke's encephalopathy, or organophosphate poisoning is uncertain. With convergence, upbeat may enhance or convert to downbeat. The slow-phase waveform is usually linear but may be an increasing-velocity exponential.