A Practical Approach to Nystagmus and Saccadic Oscillations

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Cover images. Left: Patient with a right face turn, left gaze preference due to a right-beating jerk nystagmus. Right: Selected videos featured in this module; see them online. (Left image reproduced, with permission, from Raab EL. Pediatric Ophthalmology and Strabismus. Basic and Clinical Science Course, Section 6, 2013–2014.)
LEARNING OBJECTIVES

Upon completion of this module, the reader should be able to:

1. Summarize the differences between nystagmus, saccadic intrusions, and oscillations
2. Identify the different types of involuntary eye oscillations by their clinical features
3. Improve clinical management of involuntary eye oscillations

Introduction

Nystagmus is an involuntary oscillation that is initiated by a slow eye movement driving the eyes off target (Figure 1). A second movement brings the eyes back to target. If the second phase is also slow, it is called **pendular nystagmus**. If the second phase is quick (a saccade), it is called **jerk nystagmus**. By convention, the direction of nystagmus is named in the direction of the quick phase. Jerk nystagmus usually increases during gaze in the direction of the quick phase (eg, left-beating nystagmus will increase in left gaze), a phenomenon known as Alexander’s law. Nystagmus arises from imbalances in the vestibulo-ocular or gaze-holding system; it may also arise in the smooth pursuit, optokinetic, or rarely the vergence system.

Saccadic intrusions and oscillations, on the other hand, are fast eye movements that drive the eyes off target. Intermittent inappropriate saccades are called saccadic intrusions (eg, square wave jerks, macro square wave jerks), whereas those that are continuous are called saccadic oscillations (eg, square wave oscillations, macrosaccadic oscillations, opsoclonus, ocular flutter, voluntary flutter). Saccadic intrusions and oscillations occur with lesions of the cerebellum or brainstem.

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This module reviews 6 categories of involuntary eye movements: infantile-onset/congenital nystagmus; nystagmus due to defective gaze-holding; acquired pendular nystagmus; vestibular nystagmus; saccadic intrusions and oscillations; and other involuntary eye movements. See the sidebar, “Six Key Questions,” for guidance on...
Six Key Questions

WHEN SEEING A PATIENT with involuntary eye oscillations, the ophthalmologist should ask the following important questions:

1. Does the patient experience oscillopsia, an illusion of motion in the stationary environment? Patients with infantile nystagmus (infantile idiopathic motor nystagmus) usually do not complain of oscillopsia, whereas patients with acquired nystagmus generally do.

2. Is there a slow phase? Nystagmus is initiated by slow smooth movements whereas saccadic intrusions and oscillations are initiated by fast movements.

3. Is there nystagmus in primary position? Gaze-evoked nystagmus is present only on eccentric gaze.

4. Do the eyes move together? Conjugate movements occur when both eyes move in the same direction with similar amplitude and frequency (eg, downbeat nystagmus). Disconjugate movements occur when both eyes move in the same direction but with different amplitude and frequency (eg, internuclear ophthalmoplegia). Disjunctive movements occur when the eyes move in opposite directions (eg, oculomasticatory myorhythmia).

5. What is the plane of oscillation? Peripheral vestibular nystagmus usually has a mixed horizontal–torsional component, whereas central vestibular nystagmus usually has a pure vertical or torsional component.

6. Are there additional signs? Does the direction of nystagmus change over time (eg, periodic alternating nystagmus)? Is there a gaze palsy (eg, vertical gaze palsy associated with convergence-retraction nystagmus in dorsal midbrain syndrome)? Is there a duction deficit (eg, adduction deficit associated with “abducting nystagmus” in internuclear ophthalmoplegia)? Are there signs of inner ear disease (eg, nausea, tinnitus, and hearing loss in peripheral vestibular nystagmus)? Are there other neurologic signs?

evaluating the patient with involuntary eye oscillations. Once these questions are answered, a correct diagnosis can usually be made through pattern recognition (Figure 2).

Categories of Nystagmus

1: INFANTILE-ONSET/CONGENITAL NYSTAGMUS

INFANTILE NYSTAGMUS. Infantile nystagmus (infantile idiopathic motor nystagmus, congenital nystagmus) is present from infancy, although it may not be recognized until later in life. It can have a jerk or pendular waveform. The oscillation is typically conjugate and horizontal, although there may be a small torsional component. It remains horizontal during upward and downward gaze. Infantile nystagmus increases with fixation. It decreases with convergence, in darkness, and behind closed lids. Patients usually have abnormal visual acuity but rarely experience oscillopsia. Infantile nystagmus may occur sporadically, or it may follow an autosomal-dominant, autosomal-recessive, or X-linked recessive inheritance pattern.

Most infantile nystagmus has a jerk waveform. When pendular, the nystagmus can change to a jerk waveform in right and left gaze. The nystagmus is punctuated by foveation periods, during which the eyes are transiently stationary and foveate on a target. In many patients, there is a null zone of minimum nystagmus intensity and optimal visual acuity. If the zone is eccentric, the patient often adopts a head turn to position the eyes in the null zone to achieve better visual acuity. Head oscillation may accompany the nystagmus.

Not all infantile-onset nystagmus is idiopathic. Abnormalities of the afferent visual system may present with infantile nystagmus, including:

- optic nerve disorders (eg, optic nerve hypoplasia)
- retinal disorders (eg, Leber congenital amaurosis, achromatopsia)
- foveal hypoplasia (eg, albinism, aniridia)
- cortical visual impairment (eg, hypoxic ischemic encephalopathy in term infants, periventricular leukomalacia in preterm infants, traumatic brain injury, infections, metabolic disorders)
- delayed visual maturation

Given the range of possible causes, infantile nystagmus is a diagnosis of exclusion requiring a thorough history, complete eye examination, neuroimaging, and electroretinogram and/or visual evoked potential.

The treatment of infantile-onset nystagmus includes optical, surgical, and medical approaches. Base-out prisms can be used to stimulate convergence and dampen the nystagmus to improve acuity. Prisms can also be used
Figure 2 Diagnostic algorithm for nystagmus and saccadic oscillations.
to shift the viewing position into the null zone. Surgical options include the Anderson-Kestenbaum procedure (to move the null zone to correspond to the primary position of the eyes), the Cuppers divergence procedure (to stimulate convergence and thereby improve acuity), recession of all 4 horizontal rectus muscles (to reduce head turn without reducing the range of gaze), or extraocular muscle tenotomy. Medical treatments for infantile nystagmus are less favored, since they require patients to be on life-long therapy and may produce significant side effects. Gabapentin, memantine, and carbonic anhydrase inhibitors, both oral and topical, have been shown to produce benefits.

LATENT FIXATION NYSTAGMUS. Latent fixation nystagmus (also called fusional maldevelopment nystagmus) is a type of infantile horizontal jerk nystagmus not evident during binocular viewing. It occurs when only one eye is used for fixation during monocular viewing. The quick phase of the nystagmus is directed conjugately away from the covered eye, and the direction reverses instantaneously when the fixating eye is switched. Patients with latent nystagmus often have infantile esotropia, dissociated vertical deviation, and inferior oblique overaction. Latent nystagmus typically persists into adulthood despite successful surgical realignment of the eyes. Thus, it is a biomarker of abnormal fusional development and can be seen in other unilateral congenital conditions, including congenital cataract and congenital glaucoma. Latent nystagmus can be a feature of trisomy 21 (Down syndrome). In some patients with unilateral visual impairment (including amblyopia), latent nystagmus may persist and beat away from the nondominant eye during binocular viewing; this persisting form is given the paradoxical name, manifest latent nystagmus.

SPASMUS NUTANS SYNDROME. Spasmus nutans usually appears in the first year of life. It consists of a triad of pendular nystagmus, head nodding, and abnormal head posture. Because infantile nystagmus can sometimes present with abnormal head posture and head nodding, differentiating it from spasmus nutans requires careful examination of the nystagmus. In infantile nystagmus, the oscillation is horizontal and conjugate with little variability, whereas the nystagmus in spasmus nutans is highly variable—it may be conjugate, disconjugate, disjunctive, or purely monocular over the course of a few seconds or minutes. Spasmus nutans is a benign condition and resolves spontaneously within 1 to 2 years after onset (up to 8 years). However, because tumors in the visual pathway (eg, optic glioma) may present with clinical features that are indistinguishable from spasmus nutans, neuroimaging should be performed on all patients with spasmus nutans.

2: NYSTAGMUS DUE TO DEFECTIVE GAZE-HOLDING

GAZE-EVOKED AND REBOUND NYSTAGMUS. Nystagmus that occurs when the eyes move to an eccentric position is called gaze-evoked nystagmus. It is not present in primary position. Gaze-evoked nystagmus has a jerk waveform, is conjugate, and occurs on lateral or up gaze (ie, right-beating on right gaze, left-beating on left gaze, upbeat on up gaze) but seldom on down gaze. Gaze-paretic nystagmus refers to a subtype of gaze-evoked nystagmus in which there is an associated gaze paresis from a cortical or brainstem abnormality, or weakness of the extraocular muscles.

Gaze-evoked nystagmus and gaze-paretic nystagmus must be distinguished from physiological end-point nystagmus, a normal phenomenon seen in extreme eccentric gaze. End-point nystagmus is typically not sustained, lasting only a few beats. When sustained, end-point nystagmus is normal if it is present symmetrically in both right and left gaze, and low in amplitude (under 4°). Sustained end-point nystagmus becomes more common with advanced age. Fatigue-induced end-point nystagmus occurs after eccentric fixation for about 30 seconds. Pathologic gaze-evoked nystagmus is distinguished from end-point nystagmus by its larger amplitude (over 4°), asymmetry on gaze to the right versus left, and association with other eye movement abnormalities, including abnormal smooth pursuit.

During a normal eye movement, a pulse of innervation moves the eye to the new position. This is followed by a step of innervation that holds the eye in its new position against orbital elastic recoiling forces (ie, gaze-holding). The step is generated by the neural integrator—the medial vestibular nucleus and nucleus prepositus hypoglossi in the medulla for horizontal movements, and the interstitial nucleus of Cajal (INC) in the midbrain for vertical and torsional movements. The cerebellar flocculus is also part of the neural integrator network that is critical for gaze-holding. When the neural integrator network becomes defective or “leaky,” the step cannot be sustained despite a normal pulse. After each eccentric gaze, the eye then drifts back toward the primary position followed by corrective quick phases back toward the intended eccentric position. This results in gaze-evoked nystagmus.

Gaze-evoked nystagmus is often associated with downbeat nystagmus.
Gaze-evoked nystagmus generally signifies disease of the cerebellum (particularly the flocculus) and its brainstem connections. It is often associated with downbeat nystagmus. It is most commonly caused by drugs including sedatives, anticonvulsants, and alcohol. Other causes include infarction, hemorrhage, demyelination, neoplasm, spinocerebellar degenerations, and Chiari malformation. Gaze-evoked nystagmus does not give rise to visual symptoms, and thus, no treatment is required.

Rebound nystagmus is a variant of gaze-evoked nystagmus. Upon refixation to the primary position, the nystagmus transiently “rebounds” to beat in the same direction as the refixation movement (eg, with fixation from right gaze to primary gaze, the eyes beat to the left) persisting for less than 30 seconds. This oscillation is a sign of cerebellar disease, and is commonly seen in cerebellar degenerations, Chiari malformation, multiple sclerosis, and anticonvulsant intoxication.

BRUN NYSTAGMUS. Brun nystagmus is typically caused by cerebellopontine angle tumors (eg, schwannoma of the eighth cranial nerve). It consists of a low-frequency, large-amplitude gaze-evoked nystagmus on gaze toward the side of the lesion (as a result of defective gaze-holding), and a high-frequency, small-amplitude vestibular nystagmus (see Category 4) on gaze to the contralateral side (as a result of vestibular imbalance).

INTERNUCLEAR OPHTHALMOPLEGIA. Lesions in the medial longitudinal fasciculus (MLF) in the brainstem result in an adduction deficit of the ipsilateral eye, referred to as internuclear ophthalmoplegia (INO). The adduction deficit can range from complete to a subtle decrease in saccadic velocity alone (ie, “adduction lag”). The MLF carries the signal for conjugate horizontal eye movements from the contralateral sixth nerve nucleus to the ipsilateral lateral rectus muscle (LR) and, via the medial longitudinal fasciculus (MLF), to the contralateral third nerve nucleus (III) and so on to the contralateral medial rectus (MR) to make both eyes turn to the side of the stimulating sixth nerve nucleus. A lesion blocking the path between the ipsilateral sixth nerve nucleus and the contralateral third nerve nucleus results in an internuclear ophthalmoplegia. (Reproduced, with permission, from Harper RA. Basic Ophthalmology, American Academy of Ophthalmology, 2010.)

3: ACQUIRED PENDULAR NYSTAGMUS

Acquired pendular nystagmus is one of the more common types of nystagmus, often associated with oscillopsia. It usually has horizontal, vertical, and torsional components with the same frequency, the combination of which may result in a diagonal, circular or elliptical trajectory. The nystagmus may be conjugate, disconjugate in amplitude and trajectories (sometimes appearing monocular), or disjunctive (convergent–divergent). The oscillations are rapid (frequency: 2–6 Hz), may vary in amplitude in different gaze positions, and subside under closed lids. Acquired pendular nystagmus is readily differentiated from infantile pendular nystagmus by an absence of oscillopsia, a predominantly horizontal, conjugate trajectory, and the presence of foveation periods in the latter.

Acquired pendular nystagmus in adulthood is typically a manifestation of multiple sclerosis. When
associated with visual loss, it is most commonly caused by optic nerve disease, and the nystagmus may resolve when vision returns. In unilateral optic nerve disease, the nystagmus is manifested in the affected eye (monocular nystagmus), with a prominent vertical, low-frequency, pendular waveform, and a less prominent horizontal jerk waveform. In bilateral optic nerve disease, the amplitude of the nystagmus is usually greater in the eye with poorer vision (ie, the Heimann-Bielschowsky phenomenon, which is also seen in severe amblyopia, dense cataract, and high myopia). Acquired pendular nystagmus can be seen in oculopalatal tremor, ocular-masticatory myorhythmia, brainstem stroke, cerebellar degeneration, and toluene abuse. In children, it is a feature of 2 rare leukodystrophies, Pelizaeus-Merzbacher disease and Cockayne syndrome.

Acquired pendular nystagmus is one of the more common types of nystagmus.

Acquired pendular nystagmus is believed to be caused by an abnormality in the internal feedback circuits between brainstem nuclei and the cerebellum (eg, the gaze-holding network), which are important for long-term recalibration using visual input. It may be suppressed by gabapentin and memantine in patients with multiple sclerosis.

Oculopalatal tremor, also called oculopalatal myoclonus, is a form of acquired pendular nystagmus with predominately vertical and torsional components at a frequency of 1–3 Hz. It is associated with tremor of the palate, and may be accompanied by synchronous rhythmic movement of the pharynx, face, tensor tympani, vocal cords, shoulders, or respiratory muscles.

The pathologic hallmark of oculopalatal tremor is hypertrophic degeneration of the inferior olivary nucleus in the medulla, readily seen on MRI (ie, the pimento sign). Common causes of oculopalatal tremor are infarct and hemorrhage. It is rarely caused by demyelination and trauma. The onset of oculopalatal tremor is usually delayed for months after the initial insult due to neural deafferentation. It may respond to gabapentin, memantine, and trihexyphenidyl.

Oculomasticatory myorhythmia. Oculomasticatory myorhythmia is a pathognomonic finding of cerebral Whipple disease, a rare relapsing disorder caused by the bacteria *Tropheryma whippelii*. It consists of pendular vergence nystagmus and synchronous repetitive contractions (1–2 Hz) of the masticatory, facial, and pharyngeal muscles, with or without limb involvement. The horizontal pendular oscillations of each eye are 180° out of phase, producing an alternating convergence–divergence nystagmus at a frequency of about 1 Hz. In addition, cerebral Whipple disease is associated with vertical saccadic palsy/gaze palsy. Neurologic symptoms of Whipple disease, including ocular-masticatory myorhythmia, are treated by prompt initiation of antibiotics (eg, tetracycline, penicillin, trimethoprim/sulfamethoxazole, chloramphenicol), but they may not reverse completely.

4: Vestibular Nystagmus

Peripheral vestibular nystagmus results from disease of the inner ear or vestibular nerve. Central vestibular nystagmus results from imbalance of vestibular projections from lesions in the brainstem or cerebellum.

Peripheral Vestibular Nystagmus. Diseases of the vestibular end-organ and nerve cause peripheral vestibular jerk nystagmus that beats away from the side of damage, with a mixed horizontal–torsional component. Peripheral vestibular nystagmus follows Alexander’s law and is suppressed by visual fixation or smooth pursuit. The slow phases are typically linear with a constant velocity, and the slow phase velocity increases when fixation is impaired or in darkness. Peripheral vestibular nystagmus is accompanied by signs of inner ear disease (eg, nausea, vomiting, vertigo, hearing loss, tinnitus). It subsides within a few days to weeks, due to central rebalancing of tonic activity in the vestibular nuclei. Central vestibular nystagmus, on the other hand, is typically confined to one plane (purely vertical or purely torsional), not inhibited by fixation, not associated with signs of inner ear disease, and is often accompanied by brainstem or cerebellar signs.

Downbeat Nystagmus. Downbeat nystagmus is the most common type of central vestibular nystagmus. It occurs in the primary position and is maximal in lateral gaze, down gaze, and on convergence. Many patients also have gaze-evoked and rebound nystagmus. Downbeat nystagmus is seen in diseases of the
cerebellum and in Chiari malformation, but a variety of structural lesions, anticonvulsant drugs, lithium intoxication, or magnesium depletion can be responsible. About 20% of cases are idiopathic.

The anterior semicircular canals activate the upward vestibulo-ocular reflex (VOR), and the posterior canals activate the downward VOR. Imbalances between the anterior and posterior canals that favor anterior canal activity lead to a tonic upward vestibular bias so that the eyes drift upward slowly, followed by corrective downbeating quick phases. Brainstem lesions that disrupt posterior canal projections bilaterally or cerebellar disease that causes disinhibition of anterior canal projections cause a tonic upward bias, resulting in downbeat nystagmus.

Downbeat nystagmus causes vertical oscillopsia. Both 3,4-diaminopyridine and 4-aminopyridine are effective treatments that are now available commercially in the United States. In those who do not respond, clonazepam may help.

**UPBEAT NYSTAGMUS.** Upbeat nystagmus occurs in the primary position and increases in up gaze, in accordance with Alexander’s law. The most common causes are multiple sclerosis, brainstem infarcts, tumors, and cerebellar degeneration. Lesions in the brainstem at the pontomedullary junction that damage the upward VOR pathway, or lesions of the superior cerebellar peduncle and vermis that disrupt anterior canal projections bilaterally, cause a tonic downward vestibular bias and upbeat nystagmus. Upbeat nystagmus produces vertical oscillopsia that often resolves spontaneously. If it persists, memantine, 4-aminopyridine, or baclofen may be effective.

**TORSIONAL NYSTAGMUS.** In torsional nystagmus, the eyes rotate around the anteroposterior axis. Pure torsional nystagmus indicates brainstem damage disrupting input from the anterior and posterior semicircular canals on one side. Lesions in the pontomedullary region produce torsional jerk nystagmus with the upper poles of the eyes beating away (ie, contralesional) from the side of the damage. With unilateral medullary lesions, this may be accompanied by an ipsilesional ocular tilt reaction (ie, ipsilesional head tilt, ipsilesional hypotropia, excyclotorsion of the ipsilesional eye, incyclotorsion of the contralesional eye). Lesions in the midbrain also produce contralesional torsional jerk nystagmus, but the torsional component is combined with a large upward or downward component. In contrast to medullary lesions, torsional jerk nystagmus from midbrain lesions is accompanied by contralesional ocular tilt reaction. Causes of torsional nystagmus include infarction, multiple sclerosis, and Chiari malformation. Gabapentin may be helpful in patients with oscillopsia.

**SEESAW AND HEMI-SEESAW NYSTAGMUS.** Seesaw nystagmus is a subtype of torsional nystagmus with a disjunctive vertical component. In a half-cycle, one eye incyclotorts and moves up, while the fellow eye excyclotorts and moves down; during the next half-cycle, the movement reverses in each eye (similar to the action of 2 balls sitting on opposite ends of a seesaw). If the waveform is pendular, it is called a *seesaw nystagmus*; if the waveform is jerk (ie, one half-cycle being quick phases), it is called a *hemi-seesaw or jerk seesaw nystagmus*. Most patients with seesaw nystagmus have bitemporal hemianopia caused by a parasellar tumor (eg, pituitary adenoma, craniopharyngioma), head trauma, or rarely, hydrocephalus. It can also be congenital or develop in the setting of progressive visual loss without chiasmal or brainstem disease. Hemi-seesaw nystagmus can result from unilateral midbrain hemorrhage, medullary infarcts, syringobulbia, and Chiari malformation. When seesaw nystagmus is caused by a tumor, the nystagmus usually resolves after tumor resection. Patients with oscillopsia may be treated with clonazepam, gabapentin, or memantine.

**PERIODIC ALTERNATING NYSTAGMUS.** Periodic alternating nystagmus (PAN) is a rare disorder characterized by horizontal jerk nystagmus in primary gaze that reverses direction periodically. One 90-second half-cycle of right-beating nystagmus is followed by a 90-second half-cycle of left-beating nystagmus. Between each half-cycle there is a 0–20-second transition period during which there may be no nystagmus (null period), vertical nystagmus, or saccadic movements. Because of this periodicity, the diagnosis may be missed unless the clinician observes the nystagmus for several minutes. PAN follows Alexander’s law, and patients may adopt an alternating head turn to minimize oscillopsia (eg, right head turn to put the eyes in left gaze during right-beating cycles, and vice versa). Thus, PAN should be included in the differential diagnosis of abnormal head posture, specifically periodic alternating head turn.

PAN may be congenital or acquired. The congenital form is much less regular in timing of direction reversal. Acquired PAN has been reported in association with Chiari malformation, cerebellar diseases (degeneration, mass, infection), multiple sclerosis, and anticonvulsants (phenytoin). It may rarely occur following visual loss...
(vitreous hemorrhage and cataract) and may resolve when vision is restored.

Lesions of the nodulus and uvula in the cerebellum have been implicated in PAN. Pharmacologic evidence suggests that the nodulus and uvula maintain inhibitory control on vestibular responses by using GABA. Thus baclofen, a GABA agonist, is considered first-line treatment for acquired PAN. Memantine may be helpful in patients who do not respond to baclofen.

5: SACCADEIC INTRUSIONS AND OSCILLATIONS

SQUARE WAVE JERKS. Square wave jerks (SWJ) are the most common ocular intrusion. They are sporadic, horizontal, conjugate saccades taking the eyes from 0.5° to 5° off fixation. After an intersaccadic interval of about 200 msec, a saccade returns the eyes back to fixation. Many normal subjects have SWJ with low frequency (4–6 per minute) and low amplitude (less than 2°). SWJ with frequencies exceeding 15 per minute and amplitudes over 2° are pathologic.

The causes of SWJ are myriad. They are prominent in neurodegenerative conditions, including Friedreich ataxia, olivopontocerebellar atrophy, spinocerebellar ataxia (SCA) type 3, and in multiple sclerosis with cerebellar involvement. They also occur in Parkinsonism; they are more frequent in progressive supranuclear palsy and multiple system atrophy than in idiopathic Parkinson disease. SWJ also occur in patients with focal cerebral lesions and Huntington disease.

Square wave jerks are the most common ocular intrusion.

SWJ result from damage of projections from the frontal eye field, the rostral pole of the superior colliculus, and the central mesencephalic reticular formation to the omnipause cells in the pons. SWJ are usually not associated with visual symptoms and usually do not require treatment. If symptomatic, they may be treated with methylphenidate, diazepam, phenobarbital, and amphetamines.

MACRO SQUARE WAVE JERKS. Macro SWJ are similar to SWJ but with amplitudes ranging from 5° to 50° and shorter intersaccadic intervals. They often occur in bursts that are directed to one side of fixation. They are associated with cerebellar or brainstem diseases affecting cerebellar outflow, including spinocerebellar degeneration, Friedreich ataxia, olivopontocerebellar atrophy, Chiari malformation, multiple sclerosis, and focal cerebellar hemispheric lesions. Macro SWJ result from defective GABA inhibition of the superior colliculus by the substantia nigra pars reticulata. Diazepam or phenobarbital may be used for treatment.

SQUARE WAVE OSCILLATIONS. Square wave oscillations are similar to SWJ, but they occur continuously rather than sporadically. They are seen in Parkinson disease, alcoholic cerebellar degeneration, and progressive supranuclear palsy.

MACROSACCADIC OSCILLATIONS. Macrosaccadic oscillations consist of a series of large saccades, up to 60° in amplitude, that straddle fixation (i.e., passing from one side to the other side of the intended position of fixation, overshooting it each time without foveation). The amplitude of the oscillation often increases and then decreases in a crescendo-decrescendo pattern within a burst. These rare oscillations occur with deep cerebellar lesions affecting the vermis and paramedian nuclei, including hemorrhage, tumor, multiple sclerosis, and paraneoplastic syndromes. Macrosaccadic oscillations are caused by abnormal calibration of saccadic size by the cerebellum with excessive “gain,” resulting in hypermetric primary and corrective saccades that oscillate around the desired eye position. Macrosaccadic oscillations often result in difficulty with reading. Memantine may be used for treatment.

OPSOCLONUS AND OCULAR FLUTTER. Opsoclonus consists of arrhythmic, chaotic, multidirectional saccades, without intersaccadic intervals. The movements are of large amplitude and high frequency (10–15 Hz), causing visual blur and oscillopsia. Opsoclonus is present during fixation, smooth pursuit, convergence, and persists during sleep or eyelid closure. It is often accompanied by myoclonic jerks of the limbs and trunk, and cerebellar ataxia. Opsoclonus occurs in 4 general clinical settings: parainfectious encephalitis, paraneoplastic, idiopathic, and miscellaneous (toxic-metabolic states, multiple sclerosis, pontine and thalamic hemorrhage). In paraneoplastic opsoclonus, the most common underlying cancers are small-cell lung, breast, and ovarian in adults, and neuroblastoma in children.

Ocular flutter, in contrast, consists of back-to-back saccades that are confined to the horizontal plane. It may be seen in patients with opsoclonus, or occur as an isolated oscillation in patients with spinocerebellar degeneration (eg, Friedreich ataxia and olivopontocerebellar atrophy). Less common causes include multiple sclerosis, parainfectious encephalitis, paraneoplastic syndrome, and various drugs.
Ocular flutter and opsoclonus can produce disabling oscillopsia. When caused by brainstem encephalitis, treatment with intravenous immunoglobulin (IVIg), corticosteroids, azathioprine, or anti-CD20 monoclonal antibodies (rituximab) directed against B-lymphocytes can hasten recovery. In adults with paraneoplastic opsoclonus, treatment of the tumor itself leads to improvement. Corticosteroids, adrenocorticotropic hormone (ACTH), plasmapheresis, and IVIg are sometimes effective. Opsoclonus in children with neuroblastoma often responds to corticosteroids, ACTH, and sometimes to IVIg. Treatment with rituximab may also prove effective.

VOLUNTARY FLUTTER (VOLUNTARY NYSTAGMUS).
Voluntary flutter is a benign condition present in about 8% of the normal population, and may occur as a familial trait. It consists of horizontal, conjugate, saccadic oscillations that can be initiated at will, with frequency and amplitude similar to those seen in opsoclonus and ocular flutter. Unlike the sustained oscillations seen in opsoclonus and ocular flutter, voluntary flutter can be maintained at most for 30 seconds. It is usually initiated by convergence and is accompanied by fluttering of the eyelids (not in synchrony with the eye oscillations), pupillary constriction, and contraction of facial muscles in an expression of effort. Distinguishing voluntary from pathologic flutter may sometimes require long-term observation to ensure absence of other neurologic findings, including ocular dysmetria, cerebellar ataxia, and myoclonus.

SUPERIOR OBLIQUE MYOKYMIA. Superior oblique myokymia is caused by spontaneous discharge of the trochlear nerve. It consists of intermittent bouts of tiny monocular torsional–vertical microtremor causing diplopia, monocular oscillopsia, or tremulous sensation of the eye. It lasts for seconds and occurs in clusters at unpredictable intervals. It may be triggered by blinking, by looking down then back to primary position, or by head tilt toward the side of the affected eye. It is usually not associated with any underlying disease, but is rarely reported after trochlear nerve palsy, mild head trauma, brainstem stroke or demyelination, cerebellar tumor, and vascular compression at the root exit zone of the trochlear nerve by a branch of the superior cerebellar artery or posterior cerebral artery. It may resolve spontaneously, and often responds to carbamazepine, gabapentin, baclofen, or beta blockers (topical or systemic). In persistent cases, superior oblique tenotomy with ipsilateral inferior oblique recession is effective. When caused by vascular compression at the root exit zone, vascular decompression may be useful.

Treatment
A variety of treatments for nystagmus and saccadic oscillations have been proposed, including medical, optical, surgical, and other miscellaneous approaches, but few have been evaluated in prospective masked clinical trials. Table 1 summarizes dosing of medical treatments for various forms of nystagmus.
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Conclusion

This module has categorized involuntary eye movements as follows:

- infantile-onset/congenital nystagmus
- nystagmus due to defective gaze-holding
- acquired pendular nystagmus
- vestibular nystagmus
- saccadic intrusions and oscillations
- other involuntary eye movements

By answering the 6 key questions highlighted in this module, the clinician can usually make a correct diagnosis through pattern recognition.

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Clinicians’ Corner

Clinicians’ Corner provides additional viewpoints on the subject covered in this issue of Focal Points. Consultants have been invited by the Editorial Review Board to respond to questions posed by the Academy’s Practicing Ophthalmologists Advisory Committee for Education. While the advisory committee reviews the modules, consultants respond without reading the module or one another’s response. –Ed.

1. Is it necessary to perform a workup for square wave jerks or macro square wave jerks? If so, what is the workup?

>>> Dr. Rucker: In order to determine if further diagnostic evaluation for the incidental discovery of square wave jerks (SWJ) and macro SWJ is needed, it is essential to ensure that the eye movements are accurately characterized and differentiated from nystagmus and other types of saccadic intrusions, such as macrosaccadic oscillations and ocular flutter. Ideally, quantitative eye movement recordings should be obtained to confirm the presence of an intersaccadic interval (a rest interval between back to back saccades) in SWJ and macro SWJ. Recordings should also ensure that the movement consists of a removal from and a return to central fixation, rather than a crescendo/decrescendo oscillation about fixation with an intersaccadic interval (as in macrosaccadic oscillations) or an oscillation about fixation with no intersaccadic interval (as in ocular flutter).

SWJ may occur in healthy, especially elderly, individuals. Thus, infrequent (less than 10–15 per minute) SWJ in an older, visually and neurologically asymptomatic person do not warrant further evaluation. In younger persons, a few SWJ per minute (less than 4 or 5) may be a normal incidental finding, but care should be taken to ensure the absence of neurologic disease. Excessive or nearly continuous SWJ often signify underlying neurologic disease such as progressive supranuclear palsy or Friedreich ataxia. In contrast to SWJ, macro SWJ are uncommon and generally signify underlying neurologic disease, such as multiple sclerosis, cerebellar structural lesions, or multiple system atrophy. Further workup is warranted with macro SWJ.

Workup for excessive SWJ or the presence of macro SWJ should include magnetic resonance imaging (MRI) of the brain and neurologic consultation to assess for signs of parkinsonism, ataxia, peripheral neuropathy, or other neurologic pathology. It is worth noting that several of the diseases mentioned above (progressive supranuclear palsy and multiple system atrophy, for example) have few to no obvious MRI findings and are neurologic clinical diagnoses.

>>> Dr. Thurtell: Square wave jerks (SWJ) are pairs of small (usually <2°) involuntary horizontal saccades, the first of which takes the eye away from the object of interest and the second of which brings it back to the object of interest shortly after. Macro SWJ are larger (usually >5°) and more variable in amplitude. SWJ can be seen in normal humans, especially the elderly, and with certain spinocerebellar degenerations (eg, Friedreich ataxia), extrapyramidal disorders (eg, progressive supranuclear palsy), and cerebral hemisphere lesions. Macro SWJ can be seen in patients with multiple sclerosis and multiple system atrophy. I do not pursue any workup in patients with SWJ (or macro SWJ), unless they have other neurologic symptoms or signs. When other neurologic symptoms or signs are present, I start with MRI of the brain and neurologic consultation to assess for underlying neurologic disease.

2. Which ocular motility abnormalities can be manifestations of a paraneoplastic syndrome? What is the recommended workup, including specific antibody testing, when seeing a patient with one of these motility abnormalities?

>>> Dr. Rucker: Many ocular motility abnormalities are reported in the setting of paraneoplastic disorders, but the most common are cerebellar eye movement...
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abnormalities (downbeat and gaze-evoked nystagmus, impaired smooth pursuit, saccadic dysmetria, skew deviation) with subacute cerebellar degeneration and ocular flutter/opsoclonus, typically accompanied by myoclonus. Supranuclear vertical gaze paresis is increasingly recognized with paraneoplastic brainstem encephalitis. Examples of abnormalities that may rarely be paraneoplastic include horizontal gaze paresis, abducens palsies, internuclear ophthalmoplegia, and upbeat nystagmus. Given the difficulties inherent in recognizing these as paraneoplastic, the diagnosis should be considered in any older patient with an unexplained ocular motility abnormality accompanied by rapidly progressive neurologic illness—especially illness affecting cognition, peripheral nerves, or cerebellar function.

In any patient in whom the differential includes a paraneoplastic disorder, brain MRI is the initial diagnostic test to rule out a structural cause (eg, primary brain tumor, metastases, infarction). MRI is often normal in subacute cerebellar degeneration and ocular flutter/opsoclonus, but may show T2 hyperintensities in the brainstem with paraneoplastic brainstem encephalitis. Lumbar puncture is often obtained to rule out neoplastic or infectious meningitis and often shows nonspecific low-level inflammatory changes in paraneoplastic syndromes. Specific antibody-paraneoplastic syndrome relationships exist, as described below; however, paraneoplastic antibodies (Ab) may be much more strongly linked with the type of underlying malignancy than with the specific paraneoplastic syndrome and a full paraneoplastic Ab panel may be ordered. Pure subacute cerebellar degeneration is most often linked with Yo Ab (with ovarian and breast cancer) and Tr Ab (with Hodgkin lymphoma). Subacute cerebellar degeneration with co-existing peripheral neuropathy or limbic encephalitis is linked with Hu Ab (with small cell lung cancer). In adult women with ocular flutter/opsoclonus, Ri Ab (with ovarian and breast cancer) are most common.

Paraneoplastic Ab are often negative in paraneoplastic ocular flutter/opsoclonus. This should not dissuade a search for underlying malignancy, especially for small-cell lung carcinoma. A special situation is the development of opsoclonus-myoclonus in children. In this setting, regardless of the presence or absence of paraneoplastic Ab, diagnostic evaluation for neuroblastoma with urine vanillymandelic acid and homovanillic acid and MRI of the neck, chest, abdomen, and pelvis should occur. Ma2 Ab (with testicular germ cell tumors) are linked with paraneoplastic brainstem encephalitis and vertical gaze paresis.

Malignancy is often occult and undiagnosed at time of presentation of the paraneoplastic syndrome. Co-registered body PET-CT scan has the highest sensitivity in detecting underlying malignancy. In addition, patients should undergo sex-appropriate neoplastic screening, including mammography and breast examination or testicular ultrasound and testicular examination. Serum tumor markers, such as alpha-fetoprotein (germ cell tumors), human chorionic gonadotropin (germ cell tumors), and CA125 (ovarian cancer), may also be helpful.

>>> Dr. Thurtell: A variety of ocular motor abnormalities can occur with paraneoplastic neurologic syndromes. These include ocular flutter (horizontal saccadic oscillations without an intersaccadic interval), opsoclonus (multidimensional saccadic oscillations without an intersaccadic interval), saccadic dysmetria, slow saccades progressing to complete saccadic palsy, internuclear ophthalmoplegia, external ophthalmoplegia, downbeat nystagmus, gaze-evoked nystagmus, impaired smooth pursuit, skew deviation, and ductional deficits. Since there are many other causes for these ocular motor abnormalities, my level of concern for a paraneoplastic syndrome depends on the clinical presentation.

My concern is greatest when the patient reports a subacute onset of symptoms that are suggestive of a specific paraneoplastic syndrome with relentless subsequent progression. In such cases, I investigate with MRI of the brain with contrast, and I order the entire antineuronal antibody panel (on serum) rather than check for specific antibodies. Unfortunately, a negative antineuronal antibody panel does not exclude an underlying paraneoplastic disorder. Therefore, with the guidance of an oncologist, I usually obtain other investigations to look for an occult malignancy, which may include computed tomography (CT) of the neck, chest, abdomen, and pelvis, a mammogram, a testicular ultrasound, and whole-body positron emission tomography (PET).

My selection of more specialized investigations depends on the clinical presentation (eg, I check for urinary catecholamines in a child with opsoclonus, I obtain electrophysiology in a patient with suspected Lambert-Eaton myasthenic syndrome). If the patient has other neurologic symptoms and signs, I would consult a neurologist to ensure that other possible etiologies have been excluded.

3. What is the appropriate workup for an adult with new-onset nystagmus? If neuroimaging is normal, what other testing do you recommend? Does the workup depend on the age of the adult patient?

>>> Dr. Rucker: Neuroimaging with brain MRI is the initial workup in almost any adult with new-onset nystagmus—especially if the nystagmus is present with fixation in central position. MRI will rule out most structural brainstem and cerebellar causes of new-onset nystagmus, including stroke, tumor, vascular malformation, Arnold-Chiari malformation, and demyelination. Beyond MRI, the workup is dependent on the type of
According to pediatric neurology, treatment is crucial for preventing long-term disability. Correct recognition of the clinical syndrome and timely diagnosis can be easily overlooked, but untreated Wernicke encephalopathy can result in persistent neurologic symptoms and signs. MRI changes in Wernicke encephalopathy may be highly suggestive of bilateral abduction deficits, ataxia, and confusion in a patient with upbeat nystagmus. MRI may show increased T2 signal in the dorsal midbrain and along the third ventricle with Wernicke, but may also be normal. Anti-GAD (glutamic acid decarboxylase) antibodies should be considered with MRI-negative, new-onset downbeat nystagmus.

Neurologic consultation should be obtained for any patient with new-onset nystagmus, as detailed neurologic examination is needed to detect if the nystagmus is neurologically isolated. If it is not, further testing for genetic, degenerative, paraneoplastic, or other neurologic disease may be warranted. Evaluation of new-onset nystagmus is not greatly dependent on age in adults.

Dr. Thurtell: Acquired nystagmus in an adult has a broad differential diagnosis. My workup for these patients is not influenced by the patient’s age, but does depend on the type of nystagmus. For example, downbeat nystagmus occurs with lesions affecting the flocculonodular lobe of the cerebellum, including cerebellar degenerations (eg, inherited or paraneoplastic), hindbrain malformations, mass lesions, stroke, demyelinating disease, and toxins (eg, lithium). In contrast, acquired pendular nystagmus can occur with lesions affecting the afferent visual system (eg, retinal degenerations), demyelinating disease (eg, multiple sclerosis), or as part of the syndrome of oculopalatal tremor.

Although my workup would usually include MRI of the brain with contrast in most cases, it is otherwise tailored according to the known etiologies for the type of nystagmus in question. I carefully check for other neurologic symptoms and signs, as they often help to narrow the differential diagnosis and prioritize investigations or management. For example, the presence of bilateral abduction deficits, ataxia, and confusion in a patient with upbeat nystagmus is highly suggestive of Wernicke encephalopathy. The MRI changes in Wernicke encephalopathy can be easily overlooked, but correct recognition of the clinical syndrome and timely treatment is crucial for preventing long-term disability.

Dr. Rucker: According to pediatric neuroophthalmology experts, neuroimaging is not necessary in neurologically normal infants or children with characteristic infantile nystagmus. Brain MRI should be obtained when the diagnosis is in question, such as when the clinical characteristics do not adhere to those of classic infantile nystagmus (eg, predominantly horizontal oscillations—maintained as horizontal in upgaze, dampens with convergence, association with null zone and head turn or head shaking, absence of oscillopsia). Neuroimaging should be obtained when spasmus nutans is suspected to rule out structural brain lesions. The presence of optic atrophy, optic nerve hypoplasia, or see-saw nystagmus in association with infantile nystagmus should prompt brain MRI. Optic atrophy may indicate hydrocephalus or an underlying congenital suprasellar tumor infiltrating or compressing the anterior visual pathways, such as a chiasmal glioma or craniopharyngioma. Optic nerve hypoplasia is associated with abnormalities of the midline septum pellucidum, corpus callosum, or pituitary infundibulum and with abnormalities of the cerebral hemispheres. A see-saw nystagmus component in infantile nystagmus may be accompanied by achiasmia.

Dr. Thurtell: Nystagmus in a child is usually congenital. Most children with congenital nystagmus do not require further investigations, including neuroimaging, unless other visual, neurologic, or systemic abnormalities are identified or suspected. Neuroimaging is not trivial to obtain, since the child will usually need to be placed under general anesthesia for the test. However, it should be obtained if the child has optic nerve hypoplasia, optic atrophy, or other neurologic problems (eg, developmental delay), to assess for abnormalities of the anterior visual pathways, brain, and pituitary gland. Ideally, MRI of the brain with contrast should be obtained. Children with congenital nystagmus may also require electrophysiology testing (eg, electroretinogram) to assess for retinal disease and an endocrine evaluation if there is suspected pituitary dysfunction (eg, with optic nerve hypoplasia).

Neuroimaging should be obtained when the diagnosis of congenital nystagmus is in question. It is also required for children with seesaw nystagmus, presumed spasmus nutans syndrome, and monocular nystagmus, especially if the eye examination does not reveal a cause (eg, cataract or retinoblastoma) or there are signs to suggest an optic neuropathy. Neuroimaging is also required for the investigation of acquired forms of nystagmus that are suggestive of central disease (eg, downbeat nystagmus). When should neuroimaging be ordered in children with nystagmus?
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nystagmus, upbeat nystagmus, torsional nystagmus), although these are uncommon in children.

5. When do you treat congenital nystagmus? What has been your experience with extraocular muscle surgery and with medication in improving visual acuity in individuals with congenital nystagmus?

>>> Dr. Rucker: Treatment for infantile nystagmus should be considered for improvement in torticollis and/or for visual function improvement, though it may not result in improvement in visual acuity. Kestenbaum-Anderson surgical procedures are often utilized to reposition the nystagmus null zone into a central position and to correct for abnormal head position. In addition to repositioning the null zone, the surgery results in expansion of the null zone and a decrease in nystagmus intensity outside of the null zone. Tenotomy has been reported to broaden the field of the null zone and decrease the latency to visual recognition, thus leading to potential improvements in visual function despite minimal change in visual acuity. This procedure has strong proponents, but remains controversial. Medical therapy with gabapentin or memantine has been shown in a prospective, randomized clinical trial to reduce nystagmus intensity. It is appropriate to offer medical therapy as a therapeutic option; however, many parents decide against it due to the long-term commitment for ongoing therapy and the potential risk of side effects. Surgical intervention is often preferred by parents, due to the one-time nature of the treatment.

>>> Dr. Thurtell: The waveform of congenital nystagmus is often punctuated by brief “foveation” periods, during which the eyes are transiently still. Since these patients can see clearly during foveation periods, their visual acuity is often much better than expected. Indeed, patients with well-developed foveation periods can have no visual symptoms and excellent visual acuity, and therefore do not require treatment. For those who do have visual symptoms or decreased visual acuity, a number of treatment options are available, including optical, medical, and surgical treatments. I start with a refractive intervention, to ensure that the correction of any refractive error is optimized. If the patient has refractive error, I will suggest using contact lenses, if possible, since they can dampen the nystagmus in addition to providing optical correction. When the nystagmus dampens with convergence, I will instead suggest adding base-out prism to spectacles to induce convergence; I always ensure that minus correction be combined with a divergence procedure (which forces the patient to converge while viewing distant targets), if the nystagmus dampens with convergence. Since optical, medical, and surgical therapies are thought to act via different mechanisms, I consider combining therapies if one therapy alone is inadequate.

6. What is the appropriate workup for superior oblique myokymia?

>>> Dr. Rucker: Most patients with superior oblique myokymia (SOM) will not be found to have underlying neurologic disease. However, brain MRI with contrast should be done to rule out structural pathology along the course of the trochlear nerve (the ipsilateral nerve, originating from the dorsal midbrain contralateral to the side of the SOM) and to rule out structural brainstem or cerebellar pathology, as cases have been reported with brainstem stroke, cerebellar tumor, and multiple sclerosis. SOM may develop after an acute trochlear nerve palsy, in which case appropriate work up for the trochlear nerve palsy should occur. Thinly sliced, high-resolution MRI of the brainstem region may disclose vascular compression of the trochlear nerve; however, it is not imperative to seek this, as surgical microvascular decompression is not generally a recommended treatment for SOM and radiographic detection of neurovascular compression will not likely change patient management.

>>> Dr. Thurtell: Superior oblique myokymia is characterized by brief attacks of monocular oscillations or visual blurring. The oscillations are often difficult to see, but irregular low-amplitude vertical-torsional oscillations of the affected eye may be detected with careful observation of the conjunctival vessels when the patient is symptomatic. The pathogenesis of this condition is thought to be similar to that of other paroxysmal cranial nerve disorders (eg, trigeminal neuralgia and hemifacial spasm). I therefore obtain MRI of the brain with contrast to assess for compression of the fourth nerve by a vascular loop. I request fine cuts through the fourth
nerves using the constructive interference in steady state (CISS) sequence, which gives excellent contrast between the CSF and cranial nerves. With the exception of eye movement recordings, I do not find other investigations to be helpful and I do not routinely recommend them. Incidentally, the discovery of vascular compression of the fourth nerve does not usually change my management plan. Many patients respond extremely well to medical therapy with carbamazepine, gabapentin, phenytoin, or beta-blockers. In those with vascular compression of the fourth nerve who do not respond to medical therapy, decompression of the nerve root entry zone can be effective. However, I only consider it as a last resort in patients with intractable symptoms, given the potential for significant complications.

7. How do you determine if new-onset nystagmus has a vestibular etiology? If nystagmus is vestibular in origin, what is the appropriate management?

>>> Dr. Rucker: Vestibular nystagmus may be central or peripheral. Diagnostic considerations for central vestibular nystagmus, such as downbeat or upbeat nystagmus in central fixation position, are discussed in question 3. Peripheral vestibular nystagmus (PVN) is considered here. Disease in the peripheral vestibular pathways (eg, the labyrinth, vestibular nerve, or its root entry zone) causes unidirectional nystagmus due to imbalance between the vestibular nuclei. Typically, PVN “beats away from the lesion.” For example, if disease on the left side results in reduced activity in the left vestibular nuclei, the normal right vestibular nuclei will drive the eyes slowly toward the left. Corrective fast movements toward the right result in right-beating nystagmus. PVN has mixed horizontal, vertical, and/or torsional components. It is almost never purely vertical or purely torsional. For example, complete labyrinthine destruction on one side results in mixed horizontal–torsional nystagmus and benign paroxysmal positional vertigo (BPPV) of the posterior semicircular canal, a very common cause of acute positional vertigo, results in mixed upbeat-torsional nystagmus that may only be detectable by the Dix-Hallpike maneuver. Additional characteristics of PVN include worsening of nystagmus in the direction of its fast phases (right-beating nystagmus is worse upon right gaze, for example) and substantial reduction or elimination of nystagmus during visual fixation. Thus, the nystagmus in a patient with vertigo from a peripheral vestibular process may not be seen when the patient is in a well-lit room, fixating on a visual target. Observing for nystagmus during ophthalmoscopy as the patient covers the opposite eye is helpful in detection of PVN. PVN may also worsen with hyperventilation or vibration applied to the mastoid bone.

Most PVN resolves spontaneously and does not require long-term medical therapy. Posterior semicircular canal BPPV and its associated nystagmus and vertigo are effectively treated with the Epley repositioning maneuver, which can be applied by most neurologists. Ongoing vertigo and PVN require further neurologic evaluation and may warrant neuroimaging with MRI of the internal auditory canals, CT scan of the temporal bones, electronystagmography, and/or audiologic testing.

>>> Dr. Thurtell: I suspect a peripheral vestibular etiology for nystagmus on the basis of the patient’s symptoms and the characteristics of the nystagmus. The patient will not often report oscillopsia, but will report vertigo (an illusion of self or environmental movement), autonomic symptoms (eg, nausea, vomiting, sweating), and sometimes auditory symptoms (eg, hearing loss, tinnitus, aural fullness). Peripheral vestibular nystagmus is always a jerk nystagmus that is unidirectional; it does not change direction depending on gaze direction. It obeys Alexander’s law, such that it is more intense when looking in the direction of the quick phases and less intense when looking in the opposite direction, and is increased with removal of visual fixation (eg, when looking at the eyes through Frenzel goggles).

Peripheral vestibular nystagmus can be spontaneous or provoked (eg, by a change in head position, loud sound, or Valsalva maneuver). Spontaneous peripheral vestibular nystagmus is most often due to a unilateral lesion affecting the labyrinth or vestibular portion of the eighth nerve and is characteristically a mixed horizontal–torsional nystagmus, with the quick phases directed away from the side of the lesion. Provoked peripheral vestibular nystagmus is usually due to inappropriate stimulation of a part of the labyrinth, typically one semicircular canal. The direction of the nystagmus depends on which portion of the labyrinth is being stimulated. For example, with the posterior canal variant of benign paroxysmal positional vertigo, the nystagmus is upbeat-torsional and elicited by a backward head movement. When I see nystagmus that is pendular, purely vertical, purely torsional, or changing in direction (either spontaneously or with different gaze positions), without associated vestibular, autonomic, or auditory symptoms or with associated neurologic symptoms, I suspect a cause other than peripheral vestibular disease. I usually consult an ENT surgeon or neuro-otologist for further workup and management. I recommend that the management be directed toward the underlying vestibular disorder rather than to the nystagmus itself.

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