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NEURO-OTOLOGY

ARTICLE 1: APPROACH TO THE HISTORY AND EVALUATION OF VERTIGO AND DIZZINESS

Terry D. Fife, MD, FAAN, FANS. Continuum (Minneap Minn). April 2021; 27 (2 Neuro-otology):306–329.

ABSTRACT

PURPOSE OF REVIEW:

This article reviews a method of obtaining the medical history of patients presenting with dizziness, vertigo, and imbalance. By combining elements of the history with examination, the goal is to identify patterns and an effective differential diagnosis for this group of patients to help lead to an accurate diagnosis.

RECENT FINDINGS:

Studies over the past dozen years have changed the historical approach to patients with dizziness from one based primarily on how the patient describes the sensation of dizziness. This older approach can lead to misdiagnosis, so a preferred method puts greater emphasis on whether the dizziness is acute or chronic, episodic or continuous, or evoked by or brought on by an event or circumstance so that a pattern may be derived that better narrows the differential diagnosis and focused examination can further narrow to a cause or causes.

SUMMARY:

Dizziness is a common symptom of many possible causes. This article will help clinicians navigate gathering the history and examination to formulate a working diagnosis in patients affected by dizziness.

KEY POINTS

- Dizziness is a common symptom that occurs at all ages but especially in patients aged between 41 and 70 years.
- Peripheral vestibular disorders are common, but half of patients with dizziness have a nonvestibular mechanism, and approximately one in six patients present with two different causes of dizziness at the same time.
- Many patients with dizziness see multiple health care providers in evaluation of the dizziness and feel frustrated, misdiagnosed, or misdirected.

- Overreliance on a patient's description of the dizziness and using it as the main piece of information to choose among causes leads to mistakes in the diagnosis.
- All aspects of history (symptom description, onset, frequency, duration, and provoking or aggravating circumstances) should be questioned until understood as well as possible because any part of the history can be miscommunicated by a patient or misunderstood by the health care provider.
- History-taking is best started with an open-ended question to allow patients to relay how the symptoms began and what they experience, although patients may need to be redirected in some cases.
- Excessive reliance on the patient's description of dizziness or vertigo leads to mistakes in diagnosis, but some patients accurately describe spinning, whirling, rotational sensations that do indeed imply a higher likelihood of a vestibular process.
- For patients who describe clear vertigo (spinning, whirling, rotation), if spells last less than 1 minute, then benign paroxysmal positional vertigo may be the cause. If the spells last minutes, transient ischemic attack or vestibular migraine should be considered. If the spells last hours, Ménière disease or vestibular migraine may be the cause.
- It is helpful to ask patients about the impact the dizziness or vertigo has on their quality of life and ascertain their goal for the visit and evaluation because some patients just want to be reassured that the cause is benign but can live with the symptom if need be, whereas others are desperate for treatment to relieve the symptoms.
- Spontaneous downbeat nystagmus should be considered a central finding that localizes to the cerebellar vermis or cervicomedullary junction.
- In peripheral vestibular horizontal nystagmus, the nystagmus stays in one direction, intensifying with gaze in the direction of the fast phase and diminishing or abating with gaze in the direction away from the fast phase of nystagmus.
- Delayed orthostatic hypotension (having onset beyond 3 minutes after standing or head-up tilt) may be missed by routine orthostatic vital signs but can be detected by a tilt-table test.

ARTICLE 2: VESTIBULAR TESTING

Timothy C. Hain, MD; Marcello Cherchi, MD, PhD, FAAN. Continuum (Minneap Minn). April 2021; 27 (2 Neuro-otology):330–347.

ABSTRACT

PURPOSE OF REVIEW:

Vestibular testing, both at the bedside and in the laboratory, is often critical in diagnosing patients with symptoms of vertigo, dizziness, unsteadiness, and oscillopsia. This article introduces readers to core concepts, as well as recent advances, in bedside and instrumented vestibular assessments.

RECENT FINDINGS:

Vestibular testing has improved immensely in the past 2 decades. While history and bedside testing is still the primary method of differential diagnosis in patients with dizziness, advances in technology such as the ocular vestibular-evoked myogenic potential test for superior canal dehiscence and the video head impulse test for vestibular neuritis have capabilities that go far beyond the bedside examination. Current vestibular testing now allows clinicians to test all five vestibular sensors in the inner ear.

SUMMARY:

Contemporary vestibular testing technology can now assess the entire vestibular periphery. Relatively subtle conditions, such as superior canal dehiscence or a subtle vestibular neuritis, can now be diagnosed with far greater certainty.

KEY POINTS

- The function of all five vestibular sensors in the inner ear, including the otolith organs (saccule and utricle) and all three semicircular canals, can now be tested.
- The assessment of the balance of a patient with dizziness starts when the patient is met in the waiting room and walked to the examination room.
- Frenzel goggles are critical to the rapid and efficient evaluation of patients with dizziness because they improve the clinician's ability to detect vestibular nystagmus.
- Vestibular spontaneous nystagmus is suppressed by fixation.
- Congenital nystagmus is enhanced by fixation.
- The ophthalmoscope can be used to assess spontaneous nystagmus if Frenzel goggles are not available.
- The Alexander law can be used to assess for spontaneous jerk nystagmus.
- The neck-vibration test is a sensitive and durable test of unilateral vestibular weakness.
- Hearing testing is critical to assess for Ménière disease and contributes greatly to the diagnosis of a vestibular schwannoma.
- The combination of a subjective hearing test such as the audiogram with an objective test such as otoacoustic emissions can help with the diagnosis of functional hearing loss.
- False-positive videonystagmography findings of bilateral vestibular weakness or central vestibular disturbance are common sources of referrals to neurologists.

ARTICLE 3: EPISODIC POSITIONAL DIZZINESS

Kevin A. Kerber, MD, MS. Continuum (Minneap Minn). April 2021; 27 (2 Neuro-otology): 348–368.

ABSTRACT

PURPOSE OF REVIEW:

This article provides a summary of the evaluation and treatment of patients presenting with episodic positional dizziness.

RECENT FINDINGS:

Positional components are nearly ubiquitous among diagnoses of dizziness, so it can be challenging to classify patients with episodic positional dizziness simply based on the history of present illness. Overreliance on the presence of a report of positional components has likely resulted in misapplication or misinterpretation of positional testing and negative experiences with maneuvers to treat positional dizziness. The prototypical episodic positional dizziness disorder is benign paroxysmal positional vertigo (BPPV). BPPV is caused by free-floating particles in a semicircular canal that move in response to gravity. The diagnosis is made by identifying the characteristic patterns of nystagmus on the Dix-Hallpike test. Particle repositioning for BPPV is supported by randomized controlled trials, meta-analyses, and practice guidelines. Other disorders that can present with episodic positional dizziness are migraine dizziness, central lesions, and light cupula syndrome.

SUMMARY:

Episodic positional dizziness is a common presentation of dizziness. Neurologists should prioritize identifying and treating BPPV; doing so provides an important opportunity to deliver effective and efficient care. Providers should also recognize that positional components are

common in most causes of dizziness and, therefore, should not over-rely on this part of the history of presentation when considering the diagnosis and management plan.

KEY POINTS

- Patients with any cause of dizziness typically report positional components. As a result, providers should be cautious in relying on the self-report of positional components to make a diagnosis of benign paroxysmal positional vertigo (BPPV).
- Patients with dizziness are often not reliable in their self-report of the type of dizziness. As a result, BPPV should be considered a diagnostic possibility even in patients who do not report vertigo.
- The ocular motor examination starts with observing the eyes in primary gaze for 5 to 10 seconds and specifically looking for any spontaneous movements such as nystagmus or saccadic intrusions.
- When impaired smooth pursuit is identified in patients who are cognitively intact and without other large motor deficits (eg, hemiplegia), it is a strong indicator of cerebellar dysfunction.
- Eye movement testing, including pursuit tracking, gaze testing, and saccadic eye movement observation, takes very little time but can be the key factor in identifying a cerebellar or vestibular disorder.
- The Dix-Hallpike test is designed to identify posterior canal BPPV but can also identify the horizontal and anterior canal variants and central causes of dizziness.
- Some patients with BPPV report a constant milder dizziness before and even for a time after treatment for unclear reasons.
- The gold standard test for BPPV is the Dix-Hallpike test. A positive finding is a triggered and transient nystagmus.
- *Direction-changing positional nystagmus* refers to nystagmus that changes direction with changes in head position and should not be confused with gaze-evoked nystagmus in which nystagmus changes direction with changes in the direction of gaze.
- Clinicians should not think of the Dix-Hallpike test as a test only for peripheral disorders; it is also a test for central positional dizziness.
- The most common patterns of central positional nystagmus are downbeat, apogeotropic horizontal, geotropic horizontal, and multiplanar.
- Vestibular migraine can closely mimic BPPV by presenting with purely positional dizziness.
- Patients with light cupula syndrome typically have profound positional vertigo and a constant sense of imbalance. The nystagmus is persistent geotropic direction-changing positional nystagmus without latency or fatigability.
- The Dix-Hallpike test should be performed by moving the patient from the sitting to head-hanging position at a pace a bit more quickly than he or she would ordinarily lie down and trying to tilt the head back approximately 20 degrees.
- A Cochrane collaboration meta-analysis of eight randomized controlled trials found that conversion from a positive to a negative Dix-Hallpike test significantly favored the Epley treatment group when compared with a sham maneuver or control.
- Horizontal canal BPPV can be treated by using the barbeque roll maneuver, the Gufoni maneuver, or the forced prolonged position.
- If a patient is suspected of having BPPV but the positional testing does not trigger nystagmus, it is possible that the patient had spontaneous resolution, the positional testing was not adequately performed, or the patient developed an anxiety response to previous BPPV.

ARTICLE 4: EPISODIC SPONTANEOUS DIZZINESS

Scott D. Z. Eggers, MD. Continuum (Minneap Minn). April 2021;
27 (2 Neuro-otology):369–401.

ABSTRACT

PURPOSE OF REVIEW:

Conditions causing recurrent spontaneous episodes of dizziness or vertigo span several medical specialties, making it challenging for clinicians to gain confidence in evaluating and managing the spectrum of episodic vestibular disorders. Patients are often asymptomatic and have normal examinations at the time of evaluation. Thus, diagnosis depends heavily on eliciting key features from the history. Overreliance on symptom quality descriptions commonly leads to misdiagnosis. The goal of this article is to provide the reader with a straightforward approach to the diagnosis and management of conditions that cause episodic spontaneous dizziness.

RECENT FINDINGS:

Consensus diagnostic criteria have been established for vestibular migraine, Ménière disease, vestibular paroxysmia, and hemodynamic orthostatic dizziness/vertigo. Vertigo has been recognized as a common symptom in vertebrobasilar ischemia, cardiogenic dizziness, and orthostatic hypotension. Treatment recommendations for vestibular migraine still lack high-quality evidence, but controlled trials are occurring.

SUMMARY:

The evaluation should start with a detailed description of the episodes from the patient and any observers. Rather than focusing first on whether the symptom quality is most consistent with vertigo, dizziness, lightheadedness, or unsteadiness, the clinician should clarify the timing (episode frequency and duration), possible triggers or circumstances (eg, position changes, upright posture), and accompanying symptoms. History should identify any auditory symptoms, migraine features, posterior circulation ischemic symptoms, vascular risk factors, clues for anxiety, and potentially relevant medications. Carefully selected testing can help secure the diagnosis, but excessive and indiscriminate testing can lead to more confusion. Treatments for these conditions are vastly different, so an accurate diagnosis is critical.

KEY POINTS

- **Patients with episodic spontaneous dizziness are often asymptomatic at the time of evaluation and most often have normal examinations. The diagnostic history should focus on the timing, triggers, circumstances, and accompanying symptoms rather than placing too much emphasis on the patient's description of the quality of dizziness.**
- **Dizziness is a common accompaniment of migraine that is not associated with other headache types.**
- **Vestibular migraine is the most common cause of episodic spontaneous vertigo, affecting between 1% and 2.7% of the population.**
- **Migraine headache typically precedes vertigo onset by several years; although they may begin concurrently, headaches may have resolved decades before vertigo begins.**
- **The wide spectrum of clinical manifestations and vestibular laboratory findings in vestibular migraine suggests heterogeneous pathophysiologic mechanisms.**

- The character of vestibular symptoms varies widely in vestibular migraine. Vertigo may be external or internal spinning, rocking, tilting, swaying, falling, or floating. Symptoms may be spontaneous or may be triggered or aggravated by position changes, head movements, or visual stimuli.
- The temporal relationship between headaches and vertigo is quite variable in vestibular migraine, but few patients experience vertigo consistently as a typical aura.
- Auditory symptoms occur during episodes in about half of patients with vestibular migraine and can create diagnostic confusion with Ménière disease.
- Most patients with vestibular migraine have nystagmus during episodes, although tools to block visual fixation may be needed to appreciate it. It may be present in the upright position or only during positional testing, may look central or peripheral, and may be horizontal, vertical, or torsional. However, very intense horizontal nystagmus is more suggestive of Ménière disease.
- Between episodes, patients with vestibular migraine experience higher rates of motion sickness, head motion–induced dizziness, and visually induced dizziness with complex or moving visual stimuli.
- Patients with vestibular migraine have higher rates of other coexisting vestibular disorders such as benign paroxysmal positional vertigo and persistent postural perceptual dizziness, as well as higher rates of anxiety and depression than the general population.
- The general neurologic, ocular motor, and vestibular examinations are typically normal between episodes of vestibular migraine, although minor nonspecific peripheral or central vestibular and ocular motor findings are common during the symptom-free period when carefully evaluated with quantitative tools such as videonystagmography.
- A variety of vestibular laboratory tests are more commonly abnormal in patients with vestibular migraine than in controls, but none of them is specific for the diagnosis.
- Vestibular migraine is diagnosed based on symptoms meeting diagnostic criteria, with consideration of the differential diagnosis and sometimes investigations to exclude those conditions.
- Diagnosing vestibular migraine requires a patient meets International Headache Society criteria for migraine with or without aura, have recurrent episodes of vestibular symptoms lasting 5 minutes to 72 hours, and have at least one migrainous feature besides nausea with more than half of vestibular episodes.
- Patients can often be diagnosed with vestibular migraine based on a characteristic history meeting diagnostic criteria, a normal examination between episodes, and an absence of any red flags. Further investigations to exclude alternative diagnoses may include positional testing, audiometric evaluation, neuroimaging, or vestibular laboratory testing.
- Treatment recommendations for vestibular migraine come largely from case series, retrospective reviews, expert opinion, a few small controlled trials, and adaptation from the much larger migraine headache literature.
- Identifying and treating comorbid conditions is critical in the management of vestibular migraine.
- Ménière disease is an inner ear disorder whose clinical syndrome consists of spontaneous episodes of vertigo associated with typically unilateral fluctuating sensorineural hearing loss, tinnitus, and aural fullness.
- Ménière disease attacks typically produce severe spinning vertigo lasting 2 to 3 hours on average. If examined during an attack, most patients have intense spontaneous horizontal jerk nystagmus that ultimately reverses direction during the attack.
- Although the sensorineural hearing loss in Ménière disease fluctuates and initially recovers after attacks, it progressively worsens over time, generally still affecting lower frequencies but eventually flattening out to affect all frequencies and becoming less varying.
- Audiometrically documenting low- to medium-frequency sensorineural hearing loss is important for establishing the diagnosis of Ménière disease early on, especially because tinnitus and even subjective hearing loss can occur with episodes of vestibular migraine.
- Transient ischemic attacks are an uncommon cause of episodic vertigo across the population, but they are an important and dangerous cause to consider, especially in older patients with recent-onset symptoms and vascular risk factors.

- Isolated vertigo is the most common warning symptom before vertebrobasilar stroke. Most such vertebrobasilar TIAs last minutes to 1–2 hours.
- New hearing loss accompanied by vertigo can occur in lateral pontine or inner ear stroke.
- *Vestibular paroxysmia* refers to recurrent spontaneous or sometimes triggered episodes of vertigo lasting seconds to 1 minute that can occur up to dozens of times per day. It is most often attributed to neurovascular cross-compression of the vestibulocochlear nerve. Sometimes time-locked tinnitus aids localization.
- Most patients with vestibular paroxysmia respond to carbamazepine or oxcarbazepine.
- Panic attacks commonly cause dizziness, unsteadiness, or lightheadedness, but intense vertigo is uncommon.
- Anxiety disorders, including panic disorder, can be the cause of vestibular symptoms, the result of a vestibular disorder, or a comorbidity that is necessary to identify and manage simultaneously. Psychiatric disorders may also trigger functional vestibular disorders such as persistent postural perceptual dizziness.
- Patients with delayed orthostatic hypotension have a gradual fall in blood pressure that takes more than 3 minutes of upright posture to develop. Thus, the relationship between the trigger (upright position) and dizziness may be less clear, and patients may present with what appears to be episodic spontaneous dizziness.
- Patients with orthostatic hypotension may describe symptoms of vertigo or unsteadiness rather than lightheadedness or faintness.
- Dizziness is a prominent symptom in patients with bradycardia, tachycardia, or other low cardiac output states, and it is commonly experienced as vertigo lasting seconds to minutes.

ARTICLE 5: ACUTE VESTIBULAR SYNDROME

Kristen K. Steenerson, MD. Continuum (Minneap Minn). April 2021;
27 (2 Neuro-otology):402-419.

ABSTRACT

PURPOSE OF REVIEW:

This article provides a practical approach to acute vestibular syndrome while highlighting recent research advances.

RECENT FINDINGS:

Acute vestibular syndrome is defined as sudden-onset, continuous vertigo lasting longer than 24 hours with associated nausea and vomiting, all of which are worsened with head movement. Acute vestibular syndrome is provoked by a variety of central and peripheral causes, the most common of which are vestibular neuritis and acute stroke (posterior circulation). A clinical approach focusing on timing, associated history, and ocular motor findings can improve diagnostic accuracy and is more sensitive and specific than early neuroimaging. Because of the shared neurovascular supply, both peripheral and central vestibular disorders can manifest overlapping signs previously considered solely peripheral or central, including vertical skew, nystagmus, abnormal vestibular ocular reflex, hearing loss, and gait instability. Although acute vestibular syndrome is typically benign, stroke should be considered in every person with acute vestibular syndrome because it can act as a harbinger of stroke or impending cerebellar herniation. Treatment is focused on physical therapy because the evidence is minimal for the long-term use of medication.

SUMMARY:

The diagnosis of acute vestibular syndrome first requires the elimination of common medical causes for dizziness. Next, underlying pathology must be determined by distinguishing between the most common causes of acute vestibular syndrome: central and peripheral vestibular disorders. Central vestibular disorders are most often the result of ischemic stroke affecting the cerebellar arteries. Peripheral vestibular disorders are assumed to be caused mostly by inflammatory sources, but ischemia of the peripheral vestibular apparatus may be underappreciated. By using the HINTS Plus (Head Impulse test, Nystagmus, Test of Skew with *Plus* referring to hearing loss assessment) examination in addition to a comprehensive neurologic examination, strokes are unlikely to be missed. For nearly all acute vestibular disorders, vestibular physical therapy contributes to recovery.

KEY POINTS

- Acute vestibular syndrome describes the sudden onset of continuous vertigo lasting longer than 24 hours and associated with nausea, head motion intolerance, and unstable balance.
- Nausea, vomiting, and unsteadiness are symptoms of acute vestibular syndrome that are common to several causes, so additional measures are needed to narrow the diagnostic possibilities.
- The posterior circulation (vertebral arteries, posterior inferior cerebellar artery, anterior inferior cerebellar artery, and less often superior cerebellar artery) supplies the brainstem and cerebellar regions causing stroke that might present with acute vestibular syndrome.
- Cerebellar strokes that can present with vertigo, nystagmus, and imbalance if left unrecognized and untreated could lead to brain edema that can rarely lead to herniation and death.
- CT may miss posterior fossa strokes because of considerable bony artifact from the skull, and MRI may be diffusion negative up to the first 48 hours of symptoms.
- Strictly defined acute vestibular syndrome is most commonly due to acute unilateral vestibulopathy (vestibular neuritis or ischemic labyrinthopathy). If all types of acute dizziness are included, about one-third are due to vestibular causes.
- Orthostatic hypotension and other cardiac causes of dizziness can classically cause near-syncope and possibly brief bouts of vertigo but rarely sustained vertigo as is classic for acute vestibular syndrome.
- Vitamin B_1 deficiency causing Wernicke encephalopathy can rarely present as acute vestibular syndrome, although often with additional neurologic findings.
- Phenytoin toxicity may present with acute vertigo, nausea, ataxia, and gaze-evoked nystagmus.
- Acute intoxication with alcohol, phencyclidine, opiates, marijuana, and barbiturates can cause nystagmus and vertigo.
- Vertigo is a disorder of motion perception and encompasses false spinning sensations (spinning vertigo) and other false sensations such as swaying, tilting, bobbing, bouncing, or sliding (nonspinning vertigo).
- Spontaneous vertigo classic to acute vestibular syndrome continues even when the patient is motionless but worsens with any kind of head movement; in contrast, the vertigo of benign paroxysmal positional vertigo ensues after only certain provocative head maneuvers that evoke vertigo lasting less than 1 minute rather than continuously for 24 hours.
- Recurrent attacks that are new and increasing may rarely be a sign of stuttering transient ischemic attack of the posterior circulation.
- In patients with acute vertigo, nystagmus may be obvious with the naked eye, but when it is not, removal of fixation by some means (eg, Frenzel goggles, Fresnel lenses, magnifying sheet, bright penlight, or fundoscopy) improves detection of more subtle nystagmus.
- Although ideally hearing should be assessed in every patient with suspected acute vestibular syndrome, emergency settings are rarely equipped for formal audiometry. Bedside testing with hearing test smartphone applications or finger-rub testing followed by an eventual outpatient formal audiogram can provide valuable vascular risk factor information.

- Grading truncal ataxia in acute vestibular syndrome patients can increase anterior inferior cerebellar artery stroke detection sensitivity to 100%.
- If the HINTS Plus (Head Impulse test, Nystagmus, Test of Skew plus hearing loss assessment) examination demonstrates a central pattern consisting of no catch-up saccade on head impulse testing, central pattern nystagmus (direction-changing, vertical, unaffected by fixation), vertical skew deviation, and/or new, sudden asymmetric hearing loss, stroke is likely.
- The internal auditory artery, also known as the labyrinthine artery, supplies the cochlea, saccule, and posterior semicircular canal. Greater than 90% of anterior inferior cerebellar artery infarctions affect hearing and have evidence of peripheral vestibulopathy superimposed on central vestibulopathy.
- Antivirals have not been found to be effective medications and are not recommended in acute vestibular syndrome or acute unilateral vestibulopathy in isolation. Ramsay Hunt syndrome, which may cause acute vestibular syndrome in addition to vesicles around the ear and multiple cranial neuropathies, requires immediate antiviral therapy.
- Peripheral-pattern nystagmus for the majority of patients with acute unilateral vestibulopathy is direction-fixed, horizontal nystagmus that beats away from the affected side. It intensifies with gaze in the direction of the fast phase and diminishes or abates with gaze in the direction of the slow phase (known as the Alexander law).
- A catch-up saccade on head impulse testing is an abnormality that indicates ipsilateral peripheral vestibular hypofunction. It is a cortically generated response to the loss of the normal vestibular ocular reflex. A catch-up saccade with a quick turn to the right indicates the right is hypofunctional, and a catch-up saccade with a quick turn to the left indicates the left is hypofunctional.
- Abnormalities in the pathways, peripheral to central, of the ocular tilt reaction can result in skew deviation; although skew deviation is usually a central finding, it can sometimes be present with acute unilateral vestibular loss.
- HINTS Plus in acute vestibular syndrome due to vestibular neuritis consists of an abnormal head impulse test with a catch-up saccade toward the side affected, spontaneous unidirectional nystagmus with fast phases away from the affected ear, and the absence of skew eye deviation on cover-uncover testing.
- Mood disorders and inactivity can prolong or cause incomplete recovery from symptoms of acute unilateral vestibulopathy.
- Evidence-based guidelines support the use of vestibular physical therapy alone in the treatment of classic acute unilateral vestibulopathy resulting from acute vestibular syndrome.
- Physical and barotrauma can cause central and peripheral vestibular dysfunction.
- An empiric steroid trial for autoimmune inner ear disease should be considered in patients with aggressive, subacute, fluctuating, bilateral vestibular, and cochlear symptoms.

ARTICLE 6: CHRONIC DIZZINESS

Yoon-Hee Cha, MD, FAAN. Continuum (Minneap Minn). April 2021;
27 (2 Neuro-otology):420–446.

ABSTRACT

PURPOSE OF REVIEW:

Determining the etiology of disorders that manifest with chronic dizziness can seem a daunting task, but extracting some basic elements of the patient's history can reduce the differential diagnosis significantly. This includes determining initial triggers, timing of symptoms, associated features, and exacerbating factors. This article covers distinct causes of chronic dizziness including persistent postural perceptual dizziness, mal de débarquement syndrome, motion sickness and visually induced motion sickness, bilateral vestibulopathy, and persistent dizziness after mild concussion.

RECENT FINDINGS:

To date, none of the disorders above has a cure but are considered chronic syndromes with fluctuations that are both innate and driven by environmental stressors. As such, the mainstay of therapy for chronic disorders of dizziness involves managing factors that exacerbate symptoms and adding vestibular rehabilitation or cognitive-behavioral therapy alone or in combination, as appropriate. These therapies are supplemented by serotonergic antidepressants that modulate sensory gating and reduce anxiety. Besides expectation management, ruling out concurrent disorders and recognizing behavioral and lifestyle factors that affect symptom severity are critical issues in reducing morbidity for each disorder.

SUMMARY:

Many syndromes of chronic dizziness can be diagnosed by recognition of key features, although many symptoms overlap between these groups. Symptoms may be manageable and improve with time, but they are often incompletely relieved.

KEY POINTS

- Persistent postural perceptual dizziness is a chronic disorder of postural instability that lasts at least 3 months but can have fluctuations that are both innate as well as driven by environmental stimuli such as passive or active motion and visual stimuli.
- Persistent postural perceptual dizziness may be triggered by any severe homeostatic perturbation such as a vestibular disorder or medical, neurologic, or psychological process. Symptoms may continue despite resolution of the initial trigger or can coexist with an ongoing trigger.
- A slowly progressive disorder without a clear precipitant is not consistent with persistent postural perceptual dizziness.
- The mainstays of therapy for persistent postural perceptual dizziness include vestibular rehabilitation, cognitive-behavioral therapy, and serotonergic antidepressants.
- Mal de débarquement syndrome is a disorder of post–motion-induced persistent oscillating vertigo lasting more than 48 hours.
- The perception of motion in mal de débarquement syndrome is usually described as rocking, bobbing, or swaying. This perception decreases when the individual is back in motion such as when driving.
- Symptoms associated with mal de débarquement syndrome include chronic fatigue, visually induced dizziness, headaches, tinnitus, and anxiety. It is not typical for mal de débarquement syndrome to be associated with nystagmus, extraocular movement abnormalities, hearing loss, or spinning vertigo.
- Clinically available treatments for mal de débarquement syndrome include serotonergic antidepressants and benzodiazepines; vestibular therapy is generally not helpful.
- Motion sickness and visually induced motion sickness are generally self-limited processes that end when the stimulus is over. Symptoms may include nausea/vomiting, stomach awareness, headache, sweating/pallor, dizziness, drowsiness, or eyestrain.
- Motion sickness susceptibility peaks at the ages of 7 to 12 years, is stable through adult years, and declines after age 60 years. Visually induced motion sickness generally worsens with age.
- Certain disorders such as migraine, vestibular migraine, and Ménière disease can increase susceptibility to motion sickness. Motion sickness susceptibility can increase with vestibular neuritis but return to normal if the vestibular paresis is compensated. Individuals with bilateral vestibulopathy have very low motion sickness susceptibility.
- Habituation exercises, medications (antimuscarinic, anticholinergic, antihistaminergic, or diazepam), controlled breathing, music, or pleasant smells can modify motion sickness severity.
- Core symptoms of bilateral vestibulopathy include gait unsteadiness, postural instability, visual blurring with head movement, and sometimes oscillopsia.
- Bilateral vestibulopathy can be diagnosed by rotational chair testing, caloric irrigation, or video head impulse testing; the most reliable method is rotational chair testing.

- Bilateral vestibulopathy may occur after sequential inner ear injury such as from Ménière disease, vestibular neuritis, or vestibular schwannomas, from extension of intracerebral processes such as meningitis, carcinomatosis, or other processes in the subarachnoid space into the inner ear, or secondary to vestibulotoxic medications such as aminoglycoside antibiotics.
- Vestibular rehabilitation, protecting vision, and avoiding deconditioning are helpful in reducing morbidity from bilateral vestibulopathy. Safety measures should emphasize care in low-light settings.
- Postconcussion dizziness includes categories such as positional vertigo, exertional dizziness, vestibular migraine, spatial disorientation, and visual disorders.
- Concurrent ocular motor dysfunction and visual processing disorders may occur with postconcussion dizziness and can significantly add to morbidity.
- It is important to rule out structural injury to the inner ear after head trauma, particularly if severe vertigo or concurrent hearing loss is present.
- Graded vestibular rehabilitation is generally required for postconcussive dizziness along with a multipronged approach to address concurrent cognitive slowing, headache, anxiety, and mood dysregulation.

ARTICLE 7: VERTIGO RELATED TO CENTRAL NERVOUS SYSTEM DISORDERS

Kamala Saha, MD. Continuum (Minneap Minn). April 2021;
27 (2 Neuro-otology):447–467.

ABSTRACT

PURPOSE OF REVIEW:

This article provides an overview of the numerous causes of vertigo and dizziness that are due to central nervous system (CNS) pathology and guides clinicians in formulating a differential diagnosis and treating patients with CNS causes of vertigo.

RECENT FINDINGS:

Specific autoimmune vestibulocerebellar syndromes may now be tested for, and this article discusses the antibodies known to cause such syndromes. Superficial siderosis can be more accurately diagnosed with imaging studies, and treatment using iron chelation has recently been studied but has not yet been established as an effective treatment. Central autonomic network damage in the brain can cause central orthostatic hypotension in some neurodegenerative diseases, and medication has been approved for treatment.

SUMMARY:

CNS causes of vertigo are numerous and important for clinicians to recognize. Examination findings are still an extremely valuable way to diagnose central vertigo; therefore, learning how to differentiate central from peripheral vertigo based on examination is an important skill. CNS causes of vertigo often have available treatments.

KEY POINTS

- Multiple sclerosis lesions causing vertigo occur most frequently in the root entry zone of cranial nerve VIII and the medial vestibular nucleus.
- Treatment of vertigo as part of a multiple sclerosis exacerbation is usually with steroids, plus a very short course of a vestibular suppressant.
- Although known to experience central positional vertigo, patients with multiple sclerosis are much more likely to be experiencing benign paroxysmal positional vertigo if positional vertigo is the presenting symptom.

- Anterior inferior cerebellar artery territory infarcts can cause vertigo due to a peripheral lesion or central lesion, or both.
- Central vertigo in vestibular schwannoma often results from brainstem compression.
- Cavernous malformations are seen in the posterior fossa 25% of the time, and posterior fossa cavernous malformations have higher rates of hemorrhage than supratentorial cavernous malformations.
- Hemangioblastoma is typically associated with von Hippel-Lindau disease.
- Medulloblastoma causes vertigo and increased intracranial pressure from fourth ventricle involvement.
- *Listeria monocytogenes* is the most common infectious cause of rhombencephalitis.
- Chiari malformation type 1 is a radiographic diagnosis usually made by measuring cerebellar tonsil herniation greater than or equal to 5 mm below the foramen magnum.
- Downbeat nystagmus in patients with Chiari malformations localizes to the cervicomedullary junction.
- Infratentorial superficial siderosis most commonly causes hearing loss, but ataxia and vertigo are often also present.
- Imaging, usually MRI with gradient recalled echo and susceptibility-weighted imaging sequences, shows the findings of hemosiderin damage in superficial siderosis but does not necessarily correlate with clinical symptoms in a patient.
- Treatment of superficial siderosis is symptomatic, but identifying any possible underlying structural lesion causing the superficial siderosis is imperative. Surgery and iron chelators are being investigated but have not yet been established as effective treatments.
- Patients with cerebellar ataxia often have paroxysmal vertigo along with central nystagmus findings on examination.
- The central autonomic network is damaged in some neurodegenerative diseases and can lead to central orthostatic hypotension.
- Diagnosis of autoimmune vestibulocerebellar disorders depends on both a clinical syndrome that is characteristic and a positive antibody result.
- To improve test yield, both serum and CSF samples should be obtained for antibody testing for autoimmune vestibulocerebellar disorders.
- Identifying the specific antibody causing an autoimmune vestibulocerebellar disorder can help prognosticate and determine the likelihood of a malignancy eventually being found.

ARTICLE 8: SELECTED OTOLOGIC DISORDERS CAUSING DIZZINESS

Gail Ishiyama, MD. Continuum (Minneap Minn). April 2021;
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ABSTRACT

PURPOSE OF REVIEW:

This article details updated clinical presentations and current treatment paradigms of the common otologic disorders that may present to the neurologist for vertigo, including Ménière disease, superior semicircular canal dehiscence syndrome, perilymphatic fistula, barotrauma, cholesteatoma, Ramsay Hunt syndrome, enlarged vestibular aqueduct syndrome, and autoimmune inner ear disease including Cogan syndrome.

RECENT FINDINGS:

The recent data on modern imaging techniques with three-dimensional delayed IV contrast in Ménière disease, findings on the clinical and testing parameters to diagnose semicircular canal dehiscence and barotrauma, and clinical findings in Ramsay Hunt syndrome, cholesteatoma, and enlarged vestibular aqueduct syndrome are discussed in the article. The most recent

findings on the treatment and evaluation of autoimmune inner ear disease and Cogan syndrome are also covered.

SUMMARY:

This article discusses the common clinical otologic entities in patients who may present to the neurologist for vertigo, and it can be used as a guide in the diagnosis of these conditions with the use of auditory, vestibular, and imaging results.

KEY POINTS

- Ménière disease may be caused by oxidative damage of the microvasculature resulting in degeneration of the blood-labyrinthine barrier.
- Tumarkin attacks occur in some patients with Ménière disease and are important to recognize because the falls are unpredictable and may lead to serious injury and are nearly always an indication for ablative treatment.
- The normal acoustic reflex helps distinguish the conductive hearing loss of superior semicircular canal dehiscence from that of otosclerosis, which is associated with an absent acoustic reflex.
- The demonstration of a thinning or dehiscence of the superior semicircular canal on CT of the temporal bone does not necessarily indicate the presence of superior semicircular canal dehiscence syndrome.
- All cases of superior semicircular canal dehiscence should have radiographic evidence, but CT alone overestimates the diagnosis by 6-fold to 20-fold. Patients diagnosed with superior semicircular canal dehiscence should meet criteria based on clinical presentation and audiologic and vestibular testing.
- The absence of a fistula sign at bedside testing does not rule out the diagnosis of a perilymphatic fistula.
- In trauma associated with hearing loss and/or vertigo, the CT should be evaluated in the coronal view for air bubbles (pneumolabyrinth), which is evidence of a traumatic perilymphatic fistula. Pneumolabyrinth, ossicular fracture or dislocation, or a temporal bone fracture through the otic capsule may be indications for urgent surgical exploration to preserve inner ear function.
- Clinicians should have a high level of suspicion in children presenting with hearing loss and should rule out perilymphatic fistula in the setting of inner ear anomaly as etiology.
- Mild symptoms consistent with perilymphatic fistula may be treated conservatively with avoidance of a Valsalva maneuver or with rest. However, conservative treatment is not recommended for traumatic perilymphatic fistula secondary to penetrating inner ear injury, temporal bone fracture, or ossicular damage.
- Acoustic hyperacusis with bone conduction thresholds less than 0 dB, autophony, and abnormal cervical vestibular-evoked myogenic potentials can help distinguish superior semicircular canal dehiscence from perilymphatic fistula.
- Barotrauma related to scuba diving is often associated with hearing loss (90%) and variably associated with vertigo (averaging 50%).
- High-resolution CT of the temporal bone is always indicated in audiovestibular loss in the setting of diving to rule out anatomic risk factors, ossicular disruption, hemorrhage, or pneumolabyrinth.
- Vestibular symptoms and vertigo in inner ear barotrauma should be referred to an otologic surgeon because surgical correction results in a high rate of symptom relief.
- The distinction between barotrauma and decompression inner ear syndromes in diving is critical, as inner ear barotrauma can be managed with an observational period. In contrast, decompression inner ear disease, which presents with a predominance of vestibular symptoms, should be treated with hyperbaric oxygen within 5 hours of injury as any further delay usually results in permanent inner ear damage.
- Early recognition of chronic otitis media and invasive cholesteatoma is critical. Because of the proximity of the middle ear canal to the facial nerve, the horizontal semicircular canal, and overlying dura, invasive cholesteatoma can cause hearing loss, vertigo, facial paresis, meningitis, and intracranial abscess.
- Surgical eradication of cholesteatoma is indicated and aims to prevent the extension through the dura membrane and the associated intracranial complications.

- Evaluation for fistulization of the bony labyrinth may be tested at the bedside with the fistula test using a pneumatic otoscope (the Hennebert test). However, the fistula test may be falsely negative in the case of a cholesteatoma abundantly filled with keratin debris.
- The patient presenting with a gradual onset of facial weakness, often incomplete in the setting of otalgia and otorrhea, likely has chronic otitis media with cholesteatoma causing dehiscence of the facial nerve canal.
- The classical signs of meningitis may be masked in the patient with invasive cholesteatoma due to antibiotics.
- A diffusion-weighted imaging sequence on MRI can be used to follow a cholesteatoma, which will be hyperintense on diffusion-weighted imaging.
- Chronic otitis media with headache, nausea, and vomiting should trigger a workup for meningitis.
- Areas of vesicular rash eruption in Ramsay Hunt syndrome involve the sensory distribution of the facial nerve, which can include vesicles in the ipsilateral ear (concha and antihelix, antitragus, and a portion of the lobule and adjacent mastoid), ipsilateral hard palate, and anterior two-thirds of the tongue, which has the special taste sensory fibers.
- In addition to evaluation of the external ear for vesicles in Ramsay Hunt syndrome, the clinician should evaluate the back of the ear and conduct an otoscopic examination of the external canal and the tympanic membrane.
- Facial paralysis occurs in nearly all and hearing loss in up to half of patients with Ramsay Hunt syndrome. About 30% to 50% have vestibular neuritis–like vertigo and subsequent imbalance.
- A key factor in the recovery of cranial nerve function after Ramsay Hunt syndrome appears to be an earlier onset of treatment. Treatment within 1 to 3 days of symptom onset ensures 75% of patients recover function, but only 30% recover if treated after 7 days.
- Up to 15% of children with sensorineural hearing loss have an enlarged vestibular aqueduct.
- An enlarged vestibular aqueduct and pathogenic sequence alteration in the *SLC26A4* gene are associated with bilateral hearing loss and recurrent spells of episodic vertigo.
- An enlarged vestibular aqueduct, like superior semicircular canal dehiscence, can present with cervical vestibular-evoked myogenic potentials with low thresholds and high amplitudes, but the Tullio phenomenon and the Hennebert sign are generally less prominent. A Ménière disease–like presentation with recurrent spells of vertigo can be associated with an enlarged vestibular aqueduct.
- An enlarged vestibular aqueduct can be diagnosed by using either CT of the temporal bone or MRI of the internal auditory canal.
- Many patients with an enlarged vestibular aqueduct have been advised to avoid contact sports, but current data may not support an association with minor head trauma and drops in hearing. Also, cochlear implantation in these patients is safe and effective.
- Deafness associated with autoimmune inner ear disease occurs over weeks and not 1 or 2 days, as in sudden sensorineural hearing loss. One-third of the patients with autoimmune inner ear disease have or will develop a systemic autoimmune disease.
- Up to 50% of patients with autoimmune inner ear disease report vertigo or dizziness or have tinnitus and aural fullness, mimicking Ménière disease.
- Corticosteroids are the mainstay of treatment for autoimmune inner ear disease, with serial audiograms to evaluate hearing with taper. Referral to rheumatology for consideration of steroid-sparing medications and evaluation for systemic autoimmune disease should be considered.
- The vasculitis and audiovestibular dysfunction of Cogan syndrome usually respond to high-dose corticosteroids, with the expectation of a beneficial response within 2 to 3 weeks. In intractable Cogan syndrome, the progression to deafness occurs in more than half of patients.
- In both autoimmune inner ear disease– and Cogan syndrome–associated deafness, cochlear implantation is often restorative of hearing with good to excellent results.
- The finding of immunoglobulin deposition in the stria vascularis and spiral ganglia in Sjögren syndrome associated with hearing loss indicates that the hearing loss in autoimmune inner ear disease and systemic autoimmune disease may be mediated in part by immunoglobulin deposition in the inner ears.

ARTICLE 9: TINNITUS, HYPERACUSIS, OTALGIA, AND HEARING LOSS

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ABSTRACT

PURPOSE OF REVIEW:

This article reviews the causes of tinnitus, hyperacusis, and otalgia, as well as hearing loss relevant for clinicians in the field of neurology.

RECENT FINDINGS:

Important causes of unilateral and bilateral tinnitus are discussed, including those that are treatable or caused by serious structural or vascular causes. Concepts of hyperacusis and misophonia are covered, along with various types of neurologic disorders that can lead to pain in the ear. Hearing loss is common but not always purely otologic.

SUMMARY:

Tinnitus and hearing loss are common symptoms that are sometimes related to a primary neurologic disorder. This review, tailored to neurologists who care for patients who may be referred to or encountered in neurology practice, provides information on hearing disorders, how to recognize when a neurologic process may be involved, and when to refer to otolaryngology or other specialists.

KEY POINTS

- Among the most serious causes of unilateral pulse-synchronous tinnitus are a dural arteriovenous fistula, arteriovenous malformation, or a glomus tumor arising from the jugular foramen or middle ear.
- Many forms of unilateral and bilateral tinnitus are more bothersome to patients with coexistence of depression, insomnia, stress, anxiety, and excessive caffeine use.
- Otologic causes of unilateral pulse-synchronous tinnitus include ceruminous impaction and middle ear disorders leading to conductive hearing loss such that bone-conducted sounds from vascular and other internal structures are heard more loudly.
- Unilateral pulse-synchronous tinnitus may be caused by sounds transmitted from the carotid artery (bruit) or the heart (murmur), increased intracranial pressure, or conditions that cause high-flow states such as pregnancy, anemia, and hyperthyroidism.
- "Typewriter tinnitus" is a term for a pulse-asynchronous tapping or Morse code–like staccato tinnitus that may respond to treatment with carbamazepine, oxcarbazepine, gabapentin, or pregabalin.
- Myoclonus of the stapedius or tensor tympani may cause a benign type of unilateral fluttering or thumping tinnitus that is rhythmic but not synchronous with the heart rate.
- Bilateral high-pitched subjective tinnitus that is constant but varies with ambient noise is the most common form of tinnitus and is often associated with some degree of sensorineural hearing loss.
- Palatal myoclonus and oculopalatal myoclonus cause an objective clicking sound audible to the patient and others and that persists during sleep.
- While phonophobia is an aversion to loud sounds as may occur during migraine headaches, hyperacusis is a rare disorder with constant intolerance to sounds of ordinary loudness that do not bother most people.
- Unilateral continuing otalgia without an evident structural cause can sometimes be attributable to hemicranial headache disorders, herpes zoster oticus, or postherpetic neuralgia, giant cell arteritis, or neuralgia of cranial nerves V, VII, IX, and X or of the sphenopalatine or greater occipital nerves.

- In a patient presenting with hearing impairment, the first step is to distinguish conductive from sensorineural hearing loss.
- Vestibular schwannoma can occasionally present with sudden unilateral hearing loss, although it is usually more gradual and progressive.
- When acute unilateral hearing loss is present, labyrinthine ischemia should be considered, but a viral/inflammatory cause is considered to be more common.
- Occlusion of the posterior inferior cerebellar artery can rarely cause acute audiovestibular symptoms.
- If any part of HINTS (Head Impulse, Nystagmus, Test of Skew) suggests central localization, the etiology must be assumed to be central until proven otherwise.
- Isolated labyrinthine stroke is unlikely to be visible on standard MRI.
- Ménière disease presents with an abrupt onset of recurrent vertiginous attacks with associated fluctuating unilateral low-frequency sensorineural hearing loss.
- Intracranial hypotension can present with tinnitus, altered hearing, dizziness, or vertigo.
- Among the more prevalent etiologies of chronic bilateral sensorineural hearing loss are age-related hearing loss, heritable factors, and noise exposure.