REVIEW ARTICLE

CONTINUUM AUDIO INTERVIEW AVAILABLE ONLINE

Episodic Spontaneous Dizziness

By Scott D. Z. Eggers, MD

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.**

CITE AS:
CONTINUUM (MINNEAP MINN) 2021;27(2, NEURO-OTOLOGY): 369–401.

Address correspondence to Dr Scott Eggers, Department of Neurology, Mayo Clinic, 200 First St SW, Rochester, MN 55905, eggers.scott@mayo.edu.

RELATIONSHIP DISCLOSURE:
Dr Eggers has received research/grant support from the National Institutes of Health/National Institute on Deafness and Other Communication Disorders (U01 DC13256), publishing royalties from UpToDate, Inc, and technology intellectual property royalty payments from the Mayo Clinic.

UNLABELED USE OF PRODUCTS/INVESTIGATIONAL USE DISCLOSURE:
Dr Eggers discusses the unlabeled/investigational use of amitriptyline, atenolol, betahistine, cinnarizine, cyproheptadine, diazepam, diltiazem, dimenhydrinate, flunarizine, gabapentin, lamotrigine, lomerizine, lorazepam, metoclopramide, metoprolol, nortriptyline, pizotifen, venlafaxine, and verapamil, none of which are approved by the US Food and Drug Administration (FDA) for the treatment of vestibular migraine; carbamazepine, lacosamide, and oxcarbazepine, none of which are FDA approved for the treatment of vestibular paroxysmia; and rizatriptan, which is not FDA approved for the treatment of motion sickness.

INTRODUCTION

As with any type of spell, recurrent episodes of dizziness or vertigo can be challenging to diagnose for several reasons. Because patients are often asymptomatic at the time of evaluation, diagnosis depends primarily on eliciting a detailed description of the episodes from the patient and observer. Commonly, the physical

examination is normal between events. Tests must be carefully chosen because overreliance on indiscriminate and costly investigations risks uncovering irrelevant or diagnostically misleading abnormalities. Conditions that cause episodic dizziness span several areas of practice, including neurology, otolaryngology, cardiology, physical therapy, psychiatry, and general medicine. This creates a challenge for clinicians to gain expertise in evaluating and managing the spectrum of vestibular disorders.

Ambiguous symptom terminology and excessive focus on symptom quality both contribute to diagnostic errors. Patients and clinicians use the terms *dizziness*, *vertigo*, *lightheadedness*, *faintness*, *presyncope*, *unsteadiness*, and *disequilibrium* in different and arbitrary ways. Many providers have been taught to first classify the character of the "dizziness" symptom into a rotational sensation, a sensation of impending faint, disequilibrium, or ill-defined "lightheadedness" with the rationale that the symptom quality will appropriately dictate the diagnostic categories and evaluation.[1] This approach is no longer advocated and results in premature disregard for many diagnostic possibilities because spinning vertigo does not always imply a vestibular disorder just as a sensation of impending faint does not rule one out.[2] Furthermore, patient reports of symptom quality have been shown to be both inconsistent (mismatched across different question formats) and unreliable (mismatched on test-retest of the same question).[3] Instead, the history should focus on the timing, triggers, and accompanying symptoms before placing too much significance in the symptom quality description.

Despite the risks of overreliance on symptom quality, establishing uniform symptom definitions is important to promote clear communication among clinicians. The International Classification of Vestibular Disorders[4] defines vertigo, dizziness, and unsteadiness as separate vestibular symptoms that do not necessarily predict the underlying cause or localization (TABLE 4-1). Often the term *dizziness* is used more broadly (as in this article's title) for any vestibular symptom, with *vestibular* also being used here in the broadest sense and not merely for the inner ear.

TABLE 4-1 **Definition of Vestibular Symptoms According to the International Classification of Vestibular Disorders[a]**

Term	Definition
Vertigo	(Internal) vertigo is the sensation of self-motion when no self-motion is occurring or the sensation of distorted self-motion during an otherwise normal head movement. This "internal" vestibular sensation is distinguished from the "external" visual sense of motion referred to as either *external vertigo* or *oscillopsia*. The term encompasses false spinning sensations (spinning vertigo) and also other false sensations such as swaying, tilting, bobbing, bouncing, or sliding (nonspinning vertigo).
Dizziness	(Nonvertiginous) dizziness is the sensation of disturbed or impaired spatial orientation without a false or distorted sense of motion.
Unsteadiness	Unsteadiness is the feeling of being unstable while seated, standing, or walking without a particular directional preference.

[a] Data from Bisdorff A, et al, J Vestib Res.[4]

This article focuses on conditions that cause a spontaneous "episodic vestibular syndrome," which the World Health Organization in the forthcoming *International Classification of Diseases, Eleventh Edition*[5] defines as the following:

> ...a clinical syndrome of transient vertigo, dizziness, or unsteadiness lasting seconds to hours, occasionally days, and generally including features suggestive of temporary, short-lived vestibular system dysfunction (e.g., nausea, nystagmus, sudden falls). There may also be symptoms or signs suggesting cochlear or central nervous system dysfunction. Episodic vestibular syndrome usually connotes multiple, recurrent events caused by an episodic disorder with repeated spells (triggered or spontaneous), but may initially present after the first event.

An episodic vestibular syndrome should always be specified as either spontaneous or triggered (eg, by position change or Valsalva maneuver) because the differential diagnosis and diagnostic evaluations are quite distinct. Triggers should be distinguished from occasional precipitants (eg, red wine) or exacerbating factors (eg, head movement worsening already-present dizziness). Many of the disorders herein now have formal consensus definition papers by the Bárány Society that are freely available at jvr-web.org/ICVD.html.

The majority of patients with episodic spontaneous (nontriggered) dizziness can be diagnosed with one of a handful of disorders (TABLE 4-2). In addition to the circumstances and accompanying symptoms, the duration of the episodes helps narrow the differential diagnosis:

- **Seconds: vestibular paroxysmia, cardiogenic dizziness**
- **Minutes: vertebrobasilar transient ischemic attack (TIA), vestibular migraine, panic disorder, delayed orthostatic hypotension, hypoglycemia**
- **Hours: vestibular migraine, Ménière disease, toxic/metabolic**
- **Days: vestibular migraine**

It is important to distinguish the duration of the core vestibular symptoms from any residual symptoms just as it is important to distinguish the overall duration of the condition from the duration of individual episodes.

VESTIBULAR MIGRAINE

Dizziness is a common symptom in patients with migraine. *Vestibular migraine* is a term used to specifically describe episodic vestibular symptoms attributed to migraine. It is the most common but underdiagnosed cause of recurrent spontaneous episodes of vestibular symptoms. Several factors have hindered the study of vestibular migraine, including the lack of standardized nomenclature, incomplete understanding of the pathophysiology, clinical manifestations that vary and overlap with other conditions, absence of specific tests or biologic markers, and only recently developed consensus diagnostic criteria. Prior terms include *migraine-associated vertigo*, *migraine-associated dizziness*, *migraine-related vestibulopathy*, and *migrainous vertigo*. Benign paroxysmal vertigo is considered a childhood precursor to migraine,[6] and benign recurrent vertigo is an entity with strong links to migraine.[7,8] Only a tiny fraction of patients with migraine and vertigo meet the criteria for migraine with brainstem aura.

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.**

Epidemiology and Associations

A large German study that screened a representative sample of the general population with a validated questionnaire found that vertigo and migraine co-occurred 3 times more often than would be expected by chance and estimated a lifetime vestibular migraine prevalence of 1%, making it the most common cause of spontaneous episodes of vertigo.[9] A subsequent nationwide US population-based survey found a 1-year vestibular migraine prevalence of 2.7%.[10] As with migraine headache, women are diagnosed with vestibular migraine more often than men, making up 60% to 83% of patients in case series.[10–12] The mean age of vertigo onset is around 40 years but ranges from the first to seventh decades. Migraine headache typically precedes vertigo onset by several years; although they may begin concurrently, headaches may have resolved decades before vertigo begins.[9,11,13,14]

Many clinical series report a high prevalence of vestibular symptoms and signs in patients with migraine but not in patients with other headache types. Similarly, the prevalence of migraine among patients being evaluated or treated for episodic vertigo has been estimated between 38% and 87%, compared with 10% to 24% in control groups.[7,14,15] Half to two-thirds of patients with migraine have motion sickness (2 to 5 times greater than the general population) and appear more susceptible to visually induced motion sickness.[13,16]

TABLE 4-2 Disorders and Key Features of Episodic Spontaneous Dizziness

Disorder	Key features
Vestibular migraine	Episodes of vestibular symptoms lasting 5 min to 72 hours (may be spontaneous, positional, visually induced, or head motion–induced), history of migraine, migraine features during episodes
Ménière disease	Episodes of spontaneous vertigo lasting 20 min to 12 hours accompanied by fluctuating sensorineural hearing loss, tinnitus, and aural fullness
Vertebrobasilar transient ischemic attack	Episodes of spontaneous vertigo lasting minutes to 1 to 2 hours, either isolated or accompanied by diplopia, dysarthria, dysphagia, limb dysmetria, or visual field defects; affects mainly older adults with vascular risk factors
Vestibular paroxysmia	Brief attacks of vertigo (seconds) multiple times per day with or without tinnitus; typically responsive to carbamazepine
Benign recurrent vertigo	Episodes of spontaneous vertigo without migrainous, neurologic, or otologic features that does not go on to cause any persistent vestibular or hearing loss
Panic disorder	Recurrent panic attacks lasting minutes that may have prominent dizziness, lightheadedness, or unsteadiness along with other symptoms
Delayed orthostatic hypotension	Dizziness, lightheadedness, or vertigo developing more than 3 minutes after assuming upright posture, associated with significant blood pressure drop, relieved by sitting or lying down
Cardiogenic dizziness	Dizziness or vertigo lasting seconds to minutes due to cerebral hypoperfusion from low cardiac output, most often from a paroxysmal arrhythmia (bradycardia less than 40 beats/min or tachycardia greater than 170 beats/min)
Hypoglycemia	Dizziness or vertigo due to transient drop in serum glucose; affects mainly patients with diabetes who are on insulin

Pathophysiology

The pathophysiology of vestibular migraine is incompletely understood. The wide spectrum of clinical manifestations and vestibular laboratory findings suggests heterogeneous mechanisms that are not mutually exclusive.

Vertigo in vestibular migraine could sometimes represent a migraine aura, either as a brainstem aura or as cortical spreading depression affecting the posterior parietal cortex and, via direct connections, influencing the brainstem vestibular nuclei.[17] However, only 2% to 30% of patients have vertigo meeting the aura criteria of 5 to 60 minutes preceding a migraine headache.[11,14,18]

Altered neural activity in the trigeminovascular system is implicated as a primary mechanism for migraine headache. Some of the same neurotransmitters that cause vasodilation and neurogenic inflammation (calcitonin gene-related peptide and serotonin) are expressed in the vestibular system and reciprocally connected to the trigeminal nuclei and from there to thalamocortical projections and nociceptive brainstem areas. Thus, longer-lasting vestibular symptoms could stem from processes paralleling the headache phase of migraine related to the trigeminovascular system.

Patients who have migraines have reduced thresholds to a variety of sensory stimuli, such as light, sound, smell, and tactile stimuli. This sensory hypersensitivity appears to extend to vestibular and motion stimuli. They have higher rates of optokinetic stimulation-induced motion sickness,[19] enhanced perceptual sensitivity to head motion in the roll plane,[20] and larger errors in spatial orientation during lateral head tilts,[21] implying abnormal integration of semicircular canal and otolith inputs. These and other findings suggest patients with vestibular migraine have alterations in sensory processing and integration of inputs contributing to the perception of spatial orientation.

The P/Q-type calcium channelopathy episodic ataxia type 2 manifests clinically with episodic vertigo, and half of the patients with that condition have migraine.[22] Other kindreds with apparently dominantly inherited syndromes that include both migraine and vertigo have been described, but a similar ion channel defect has not been identified for those or others with vestibular migraine.[23] It has been suggested that a channelopathy causing migraine could also cause an osmotic disequilibrium in the inner ear that results in endolymphatic hydrops, the pathologic process underlying Ménière disease. The vertigo attacks of vestibular migraine and Ménière disease have similar duration, and one study found that vertigo attacks in Ménière disease have migrainous features (headache, photophobia, or visual aura) in 45% of patients.[24]

Clinical Features

Numerous case series have described the clinical features of vestibular migraine. Evolving diagnostic criteria and referral bias in neuro-otology clinics likely influence some of the findings. Clinical features may be divided into ictal (episodic) symptoms and signs, interictal symptoms and signs, and other patient characteristics and comorbidities.

EPISODIC SYMPTOMS. Patients may describe vertigo that is internal (a false sensation of self-motion) or external (a false sensation that the visual surround is

KEY POINTS

- 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.

spinning or flowing). The character is often that of spinning but may also commonly be rocking, tilting, swaying, falling, or floating.[11,13] Frequently, accompanying postural unsteadiness or nonvertiginous lightheadedness is present. Although vestibular migraine most commonly causes spontaneous episodes of dizziness, patients frequently describe their vestibular symptoms as being triggered by (and aggravated by) head motion, visual stimuli (such as complex motion–rich visual environments, busy patterns, optic flow, or screen motion), or changing head positions such as to supine or side-lying.[13,25,26] Patients may also report episodes provoked by other common migraine triggers such as stress, sleep deprivation, menses, bright lights, specific foods, or weather changes.

The temporal relationship between headaches and vertigo is quite variable. Migrainous headaches are associated with at least some of the vestibular episodes in between 50% and 94% of patients.[13,14,27] Although some patients rarely experience vertigo and headache together, some report a consistent pattern with their episodes, with vertigo occurring before, during, or after the headache phase. Few patients experience vertigo consistently as a typical aura lasting 5 to 60 minutes before headache onset.[9,18]

Other migrainous symptoms appear to be more common than headache during vertigo attacks. Case series report photophobia in 70% to 90% of patients, phonophobia in 60% to 90% of patients, and migraine auras (usually visual) in 13% to 36% of patients.[13,14,18,28] Nausea with or without vomiting occurs in the majority of patients but is not specific for migraine because nausea is a symptom of most vestibular disorders. Auditory symptoms beyond phonophobia occur in 40% to 60% of patients.[9,13,28,29] These most often include tinnitus and aural pressure/fullness but may also include muffled hearing or other subjective hearing impairment. Usually binaural, these symptoms can be monaural and thus do not immediately discriminate conditions such as Ménière disease from vestibular migraine, although objective low-frequency sensorineural hearing loss is not a feature of vestibular migraine. Other nonspecific symptoms such as cognitive slowing, fatigue, visual blurring, word-finding difficulty, extrapersonal misperceptions, or visual distortions are reported not uncommonly.[13]

The duration of a vestibular migraine episode is variable, but most last minutes to hours. Roughly one-third of patients report episodes lasting minutes, one-third lasting hours, and one-third lasting longer than 1 day.[18,30] Occasionally, patients report brief paroxysmal vestibular symptoms lasting seconds, ranging from vertigo to directional pulsion to postural unsteadiness, without other accompanying symptoms. Those symptoms alone would not meet criteria for vestibular migraine. Like other migraine types, the frequency of vestibular migraine episodes is also highly variable, but most occur a few times per year to a few times per month.[11,12,27,31]

EXAMINATION FINDINGS DURING EPISODES. Examination findings during episodes of vestibular migraine are highly variable. Case series describing ocular motor signs typically use video-oculography with visual fixation removed, which is more sensitive at detecting subtle abnormalities than routine bedside examination. One series found 70% of patients to have pathologic nystagmus during episodes, which could be spontaneous, gaze-evoked, or positional and have either central or peripheral features.[32] An abnormal head impulse test can sometimes accompany spontaneous horizontal nystagmus, supporting a unilateral deficit in

the vestibulo-ocular reflex during episodes. Most patients have impaired gait and stance during episodes, showing progressively increasing rates of difficulty with standard gait, Romberg test, tandem gait, and tandem Romberg test. A study capturing ictal nystagmus in patients with episodic vertigo found 44 of 67 patients with vestibular migraine to have spontaneous horizontal (n = 28), upbeat (n = 6), or downbeat (n = 10) nystagmus; 16 had positional nystagmus only, and 7 had no nystagmus.[33] Strong spontaneous horizontal nystagmus (>12 degrees per second) was much more sensitive and specific for Ménière disease than vestibular migraine, but spontaneous vertical nystagmus was highly specific for vestibular migraine. As opposed to paroxysmal positional nystagmus with benign paroxysmal positional vertigo (BPPV), positional nystagmus in vestibular migraine persisted for the duration the head position was maintained. Another study found that a sustained, low-velocity positional nystagmus could be elicited in all 26 patients examined during vestibular migraine attacks with visual fixation blocked, and this could be horizontal, vertical, or torsional.[34] Spontaneous nystagmus was observed in only 19% but could be provoked by horizontal headshaking in 35%.

INTERICTAL SYMPTOMS. Although vestibular migraine is considered an episodic vestibular disorder, patients report a high prevalence of symptoms between attacks. It is sometimes unclear whether these should be considered within the spectrum of vestibular migraine or represent comorbid vestibular disorders. Most patients have a past or concurrent history of more typical headache-predominant migraine with or without aura. But beyond the episodic vestibular symptoms meeting the criteria for vestibular migraine, other vestibular symptoms are common. Motion sickness affects those with migraine much more commonly than those without. One study found 61% of 131 patients with vestibular migraine experience motion sickness,[13] although it is uncertain whether patients with vestibular migraine have an even higher susceptibility than other patients with migraine.[35,36]

Patients with vestibular migraine frequently report sensitivity to head motion and visual stimuli beyond that of classic motion sickness, even when such triggers do not provoke a full episode of vestibular migraine. One series reported visually induced dizziness in 89%, including sensitivity to supermarket aisles, busy visual patterns, motion on large movie screens (three-dimensional, IMAX), scrolling on electronic screens, video games, optic flow in vehicles, or visual motion (windshield wipers, ceiling fans). The same series reported dizziness induced by quick head movements in 66%, beyond the already high 13% of patients with vestibular migraine with coexisting BPPV.[13] In fact, 51% of this cohort with vestibular migraine reported persistent, almost constant dizziness. When these visually and motion-induced symptoms are so pervasive as to be present throughout most days, some authors have considered whether this may represent a form of chronic vestibular migraine. However, it may be more appropriate to make a coexisting diagnosis of persistent postural perceptual dizziness (PPPD) if criteria for that condition are met.[37] Vestibular migraine appears to be among the most common conditions leading to the development of comorbid PPPD, with up to 35% of patients with vestibular migraine ultimately developing PPPD by the time they reach a tertiary referral center for evaluation of dizziness.[18,38]

In addition to motion sickness and chronic dizziness of PPPD, patients with vestibular migraine have higher rates of psychiatric comorbidities and psychological strain compared with those with other vestibular disorders or

KEY POINTS

- 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.

controls. This appears independent of any measurable degree of vestibular dysfunction on laboratory testing. Anxiety or depression affects more than half of patients with vestibular migraine.[13,39,40] Rates of insomnia, phobic disorders, and functional disorders also appear to be increased.

INTERICTAL EXAMINATION AND LABORATORY TEST FINDINGS. The general neurologic examination of patients with vestibular migraine is normal between episodes, so any abnormalities suggest a comorbid condition or alternative diagnosis. Even a detailed bedside ocular motor and vestibular examination is typically normal. Among 131 patients assessed in a retrospective chart review study, none had any abnormalities of alignment, extraocular range, saccades, pursuit, dynamic visual acuity, vestibulo-ocular reflex by head impulse test, or vestibulo-ocular reflex suppression nor any spontaneous nystagmus, and only 3.8% had gaze-evoked nystagmus.[13] However, 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, that allow for eliminating visual fixation to examine nystagmus. Case series found that 33% to 70% of patients with vestibular migraine have at least one of the following test abnormalities: spontaneous, gaze-evoked, positional, or other triggered (head-shaking– or vibration-induced) nystagmus; impaired pursuit; or vestibulo-ocular reflex abnormalities.[11,12,41–45] Positional nystagmus is most often persistent, asymptomatic, and central appearing. Studies have also found increased rates of such abnormalities in patients with migraine without vestibular symptoms.[41,42]

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. Caloric testing is reportedly abnormal in about 20% of patients, most often a reduced unilateral response or abnormal directional preponderance but occasionally a bilateral reduced response.[11,12,41,43,46] Video head impulse testing may be abnormal in about 10% of patients.[46] Studies measuring vestibular-evoked myogenic potentials to test otolith pathway function have shown conflicting findings of various parameters in patients with vestibular migraine. Although the rate of vestibular-evoked myogenic potential abnormalities is higher in these patients than in controls, overall they are not specific to allow or refute a diagnosis of vestibular migraine nor can they discriminate between vestibular migraine and Ménière disease as the cause of episodic vertigo in individual patients.[47–49]

Differential Diagnosis

The differential diagnosis for vestibular migraine is broad and largely consists of other conditions that cause episodic dizziness (TABLE 4-2), particularly those causing vertigo with a similar temporal profile. Most of these disorders are discussed in subsequent sections, but a few comments about them are particularly relevant to the differential diagnosis of vestibular migraine.

- **The diagnosis of migraine with brainstem aura requires two or more brainstem symptoms acting as an aura lasting 5 to 60 minutes before a migraine headache. Only a small fraction of patients with vestibular migraine meets such criteria.[11,18]**
- **Ménière disease is about 1/10 as common as vestibular migraine but can be difficult to distinguish from vestibular migraine because of overlapping clinical features.[9,50] Just as**

auditory symptoms are common during attacks of vestibular migraine (see the earlier section Episodic Symptoms), migrainous symptoms such as headache are common during attacks of Ménière disease.[18,24,29,51] Occasionally, patients meet the criteria for both conditions.

- Vertebrobasilar TIAs can cause episodes of isolated vertigo lasting from minutes to 1 to 2 hours without other brainstem symptoms. These would typically not be accompanied by migrainous symptoms and would be an unlikely cause of vertigo attacks spread weeks or months apart.
- BPPV is the most common vestibular disorder (although uncommon in the teens and twenties) and is comorbid with vestibular migraine more often than would be expected by chance. Vestibular migraine can sometimes be mistaken for BPPV when it causes primarily positional-induced symptoms. Typically, the two can be distinguished by history based on the temporal profile. During episodes of vestibular migraine, patients may experience persisting vertigo for the duration the offending position is maintained, but the entire episode lasts hours to 1 or 2 days, with episodes weeks to months apart.
- PPPD is a chronic vestibular disorder of daily fluctuating nonvertiginous dizziness. As discussed earlier in the section Interictal Symptoms, it can sometimes be difficult to know whether to attribute vestibular symptoms to PPPD or vestibular migraine, particularly in patients with chronic migraine headaches who have long-standing episodic unsteadiness superimposed on a strong background of chronic nonvertiginous dizziness.[18]
- Vestibular paroxysmia causes spontaneous episodes of vertigo lasting less than 1 minute,[52] although patients with episodes lasting up to 5 minutes may meet criteria for probable vestibular paroxysmia but also reach the duration qualifying for vestibular migraine.

Diagnostic Process

In the absence of any specific clinical examination signs or diagnostic tests, vestibular migraine is diagnosed based on symptoms meeting diagnostic criteria, with consideration of the differential diagnosis mentioned earlier and sometimes investigations to exclude those conditions.

DIAGNOSTIC CRITERIA. Building on the criteria originally proposed by Neuhauser and colleagues[14] in criteria from 2001, the International Headache Society and the Bárány Society jointly developed and published diagnostic criteria in 2012 and 2013 (TABLE 4-3).[53,54] Essentially, these require having migraine with or without aura, recurrent episodes of vestibular symptoms lasting 5 minutes to 72 hours, and at least one migrainous feature besides nausea with more than half of vestibular episodes. The Bárány Society also published criteria for probable vestibular migraine for when the relationship between episodic vestibular symptoms and migraine is less clear.[54]

Unsurprisingly in practice, patients with recurrent spontaneous episodes of vestibular symptoms more commonly meet the criteria for probable vestibular migraine than vestibular migraine. In that case, it is appropriate to consider the differential diagnosis, perform any investigations needed to exclude other causes, identify and manage any coexisting vestibular disorders (eg, BPPV, PPPD), treat as vestibular migraine, and reconsider the diagnosis if the patient does not respond to treatment or if clinical features change. Over time, some patients with probable vestibular migraine will eventually meet criteria for vestibular migraine and some will not, but it is uncommon for patients with probable vestibular migraine to ultimately develop an alternative disorder such as typical Ménière disease.[55]

EVALUATION. For many or most patients, a characteristic symptom history meeting diagnostic criteria, a normal examination between episodes, and

KEY POINTS

- 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.

absence of any red flags to suggest an alternative diagnosis permit the clinician to diagnose vestibular migraine in the office without any special diagnostic tests, as is done with other forms of migraine. Some further evaluation may be necessary to investigate alternative conditions:

- **Positional testing in the office should routinely be performed to identify and treat BPPV, especially in patients reporting any positional vestibular symptoms. Without special equipment such as video goggles, such testing may trigger vertigo and nystagmus in patients with BPPV, but it is virtually always normal in patients with vestibular migraine between attacks.**
- **Audiometric evaluation should be obtained for patients with any auditory symptoms during or between attacks to look for characteristic features of Ménière disease or, less likely, superior canal dehiscence syndrome mimicking (or coexisting with) vestibular migraine.**
- **Neuroimaging should be considered for patients with recent-onset episodes lasting minutes, especially those with vascular risk factors, to exclude the possibility of vertebrobasilar TIAs presenting as isolated episodes of vertigo. MRI of the brain with diffusion-weighted imaging and magnetic resonance angiography (MRA) or CT angiography of the posterior circulation can exclude a completed stroke or critical stenosis. Vestibular schwannoma, best identified with gadolinium-enhanced MRI of the internal auditory canals, would rarely cause episodic vertigo mimicking vestibular migraine, especially in the absence of progressive monaural sensorineural hearing loss.**

TABLE 4-3 **Diagnostic Criteria for Vestibular Migraine and Probable Vestibular Migraine[a]**

Vestibular migraine

A At least five episodes with vestibular symptoms[b] of moderate or severe intensity,[c] lasting 5 minutes to 72 hours

B Current or previous history of migraine with or without aura according to the International Classification of Headache Disorders (ICHD)

C One or more migraine features with at least 50% of the vestibular episodes:

- Headache with at least two of the following characteristics: one-sided location, pulsating quality, moderate or severe pain intensity, aggravation by routine physical activity
- Photophobia and phonophobia
- Visual aura

D Not better accounted for by another vestibular or ICHD disorder

Probable vestibular migraine

A At least five episodes with vestibular symptoms[b] of moderate or severe intensity,[c] lasting 5 minutes to 72 hours

B Only one of the criteria B and C for vestibular migraine is fulfilled (migraine history or migraine features during the episode)

C Not better accounted for by another vestibular or ICHD disorder

[a] Reprinted from Lempert T, et al, J Vestib Res.[54] © 2012 IOS Press and the authors.
[b] Qualifying vestibular symptoms include spontaneous vertigo, positional vertigo, visually induced vertigo, head motion–induced vertigo, or head motion–induced dizziness with nausea.
[c] Vestibular symptoms are moderate or severe if they interfere with daily activities.

- **Vestibular laboratory testing is not required to diagnose vestibular migraine and commonly reveals minor nonspecific central or peripheral abnormalities that create diagnostic confusion. Testing can be helpful in select circumstances when concern exists for an alternative cause of episodic vertigo such as Ménière disease or superior canal dehiscence syndrome or when additional persistent symptoms raise the question of a unilateral or bilateral vestibulopathy. Neurologists are generally skilled at detecting signs of central nervous system dysfunction at the bedside (downbeat or gaze-evoked nystagmus, impaired pursuit, dysmetric saccades), although vestibular testing can evaluate these ocular motor signs in a more sensitive and quantitative way.**

Treatment

Treatment for vestibular migraine has not been evaluated in large, well-designed, controlled trials. Most data come from numerous case series and retrospective reviews vulnerable to placebo effect, spontaneous improvement, or potentially biased un-blinded investigators. Thus, current recommendations represent a synthesis of these available studies, anecdotal experience, expert opinion, and adaptation from the much larger and scientifically solid migraine headache literature (TABLE 4-4).

NONPHARMACOLOGIC MANAGEMENT. Education and reassurance may be the most important aspects of management. Many patients with vestibular

Treatment Options for Vestibular Migraine

TABLE 4-4

Nonpharmacologic management

- Education and reassurance
- Sleep hygiene
- Trigger avoidance (eg, dietary restriction)
- Stress reduction
- Regular exercise

Acute symptomatic or abortive medications

- Antihistamines (eg, meclizine, dimenhydrinate)
- Benzodiazepines (eg, diazepam, lorazepam)
- Antiemetics (eg, promethazine, prochlorperazine, metoclopramide)
- Triptans

Prophylactic medications

- Antiepileptic drugs (valproic acid, topiramate, gabapentin, lamotrigine)
- Beta-blockers (propranolol, atenolol, metoprolol)
- Calcium channel blockers (flunarizine,[a] cinnarizine,[a] verapamil, diltiazem, lomerizine[a])
- Serotonin norepinephrine reuptake inhibitors (SNRIs) (venlafaxine)
- Tricyclic antidepressants (amitriptyline, nortriptyline)
- Cyproheptadine
- Pizotifen[a]

[a] Not approved by the US Food and Drug Administration (FDA) for use in the United States.

migraine have had unexplained attacks of vertigo for years or decades without a diagnosis or with an incorrect diagnosis, most often Ménière disease, BBPV, or a symptomatic label such as *recurrent vestibulopathy*. Educating patients that, although episodes can be temporarily disabling, they are not going to permanently lose hearing or vestibular function or have a stroke can help frame the treatment discussion toward symptom control and shared treatment decision making. Because patients often seek care when their attacks are most frequent or severe, the expected disease fluctuations over time commonly improve. Patients should examine modifiable lifestyle factors that could be contributing to migraine (eg, stress, insufficient/irregular sleep, triggering foods, insufficient exercise) and address those where possible to take an active role in management. If vestibular migraine episodes are mild, short, or infrequent, pharmacologic management may be unnecessary unless indicated for other migraine symptoms such as headache.

ACUTE EPISODE TREATMENT. Antihistamines (eg, meclizine, dimenhydrinate), benzodiazepines (eg, diazepam, lorazepam), or antiemetics (eg, promethazine, prochlorperazine, metoclopramide) can be used to treat significant vertigo and nausea when episodes tend to last more than 30 minutes (long enough for the medication to begin to work). Most patients prefer oral administration, although some medications are available in a rectal formulation if vomiting makes oral use impractical. In the acute care setting, the IV route is preferred.

Triptans have not been rigorously studied in vestibular migraine but should be considered when headache symptoms accompany vertigo attacks or when vertigo acts as a migraine aura. A pilot study of zolmitriptan for vestibular migraine found a nonsignificantly higher response rate with treatment versus placebo (38% versus 22%) but was underpowered because of low enrollment.[56] Rizatriptan pretreatment reduced vestibular-induced motion sickness in patients with migraine in one small trial.[57] A multicenter, randomized, double-blind, placebo-controlled trial of rizatriptan for treating vertigo attacks of vestibular migraine is currently underway. Noninvasive vagal nerve stimulation and external trigeminal nerve stimulation have also been reported in single-center retrospective case series as potentially shortening attack duration or reducing vertigo intensity.[58,59]

PROPHYLACTIC MEDICATIONS. Migraine prophylactic medications may be appropriate for patients with frequent episodes of vestibular migraine when acute treatments are insufficient. The approach is similar to that for migraine headache, with the goal of reducing the frequency and severity of the attacks. Because prospective trials are limited and almost every migraine preventative has been reported as potentially effective in some case series, medication choice is driven more by side effect profiles and comorbidities than efficacy data. A small randomized controlled trial found the calcium channel blocker flunarizine plus betahistine significantly reduced the frequency of vertiginous episodes in vestibular migraine compared with betahistine alone.[60] Another small randomized controlled trial compared venlafaxine to propranolol (without a placebo group) in vestibular migraine and found significant reductions in vestibular symptom severity and attack frequency at 4 months; the treatment outcome was similar in both groups, although venlafaxine also reduced

depression symptoms.[61] A single-blinded randomized trial compared venlafaxine, flunarizine, and valproic acid in 25 patients per group and found no difference among groups in total Dizziness Handicap Inventory score reductions (although only venlafaxine improved the emotional domain); valproic acid and venlafaxine but not flunarizine were associated with reduced vertigo attack frequency.[62] Metoprolol was compared with placebo over 6 months in a multicenter, randomized, controlled trial but found no difference (with both groups showing the same reduction in attack frequency over time) before being terminated prematurely because of poor participant accrual.[63] Case series have reported many other medications used to treat migraine can be helpful in vestibular migraine, including other calcium channel blockers (cinnarizine, verapamil, diltiazem, lomerizine), beta-blockers (propranolol, atenolol, metoprolol), antiepileptic drugs (topiramate, gabapentin, lamotrigine, valproic acid), tricyclic antidepressants (amitriptyline, nortriptyline), and medications from other classes (cyproheptadine, pizotifen).[12,17,64–72] Among these prophylactic medications studied, flunarizine, cinnarizine, betahistine, lomerizine, and pizotifen are not US Food and Drug Administration (FDA) approved in the United States.

VESTIBULAR THERAPY. Vestibular rehabilitation exercises probably have little role in the management of vestibular migraine for patients who only have spontaneous episodes of vestibular symptoms with no interictal symptoms or other vestibular comorbidities that would benefit from vestibular rehabilitation. A review of five retrospective and prospective studies examining the role of vestibular rehabilitation in patients with vestibular migraine (along with other conditions but generally without a control group) found the current evidence inconclusive.[73]

TREATING COMORBID CONDITIONS. Identifying and treating comorbid conditions are critical in the management of vestibular migraine.[38] In addition to considering and ruling out alternative diagnoses in the differential (discussed in the earlier Differential Diagnosis section), some conditions not infrequently coexist with vestibular migraine, including BPPV, PPPD, motion sickness, and anxiety (CASE 4-1). Some patients meet criteria for both vestibular migraine and Ménière disease and require careful medical management for both conditions, sometimes using headache behavior as an indicator of migraine treatment response, before considering any ablative procedure for Ménière disease.[29] Serotonin norepinephrine reuptake inhibitors (SNRIs), particularly venlafaxine, may be ideal when vestibular migraine coexists with PPPD, anxiety, or depression.

MÉNIÈRE DISEASE

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. Although histopathologically associated with endolymphatic hydrops within the labyrinth, it remains uncertain precisely how this relates to the clinical findings.[75] This article discusses the clinical features and diagnosis of Ménière disease. For more information about its pathophysiology and treatment, refer to the articles "Selected Otologic Disorders Causing Dizziness" by Gail Ishiyama, MD,[76] and

KEY POINTS

- 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.

"Tinnitus, Hyperacusis, Otalgia, and Hearing Loss" by Terry D. Fife, MD, FAAN, FANS, and Roksolyana Tourkevich, MD,[77] in this issue of *Continuum*.

Clinical Features

The prevalence of Ménière disease in the United States has been reported as 190 per 100,000, with slightly more women affected than men.[78] It most commonly begins in the fourth to sixth decades, with prevalence increasing with advancing age. It is rare in children (9 per 100,000 children). Most patients present with vertigo and cochlear symptoms together, although some may experience only vertigo or hearing loss alone initially. If sensorineural hearing loss predates the onset of episodic vertigo by many months or years, it is called *delayed endolymphatic hydrops* or *delayed Ménière disease*. At disease onset, the frequency of attacks averages 4 to 6 times per year but rapidly declines in the first 5 to 10 years to reach 1 to 2 times per year, slowly declining thereafter until eventually "burning out."[79] However, the clinical course can vary considerably among patients, with intervals of unrelenting attacks separated by long periods of only occasional attacks.

CASE 4-1

A 37-year-old woman presented with episodic and chronic vestibular symptoms. She had a 20-year history of occasional migraine headaches without aura. She had previously been treated for generalized anxiety disorder. Four years ago, she began having spontaneous episodes of rocking or spinning vertigo, aggravated by head movement or environmental motion and usually accompanied by nausea, photophobia, and phonophobia. She experienced binaural tinnitus with them but not aural fullness or hearing loss. With a few of the dozen episodes, she developed a unilateral throbbing headache. Episodes typically lasted half a day, although some lasted 2 days.

Last year, she also began experiencing a feeling of floating woozy lightheadedness and mild unsteadiness almost every day, aggravated by her own movements, walking through busy stores or streets, scrolling on her computer or smartphone, or watching action movies. She felt better when she was recumbent. She had repeatedly normal neuroimaging, audiograms, and vestibular laboratory testing. She worried about overlooked multiple sclerosis or brain tumor. She had restricted many activities because of her daily symptoms and concern that she may become incapacitated by an attack. Neurologic, ocular motor, and vestibular examinations were normal.

COMMENT

This case illustrates the challenge of coexisting episodic and chronic vestibular syndromes along with the diagnosis and management of relevant comorbidities. The patient's episodic vestibular symptoms along with long-standing migraine headaches are consistent with vestibular migraine. Although tinnitus might raise the possibility of Ménière disease, the binaural nature and absence of fluctuating or progressive hearing loss

Attacks of Ménière disease typically begin with cochlear symptoms such as roaring or rushing tinnitus, a sensation of aural fullness or pressure, and hearing loss (or distorted, muffled hearing). Within minutes to sometimes hours, vertigo (most commonly spinning) rapidly develops and quickly peaks in intensity before declining over the course of 20 minutes to several hours, averaging 2 to 3 hours. If examined during an attack, most patients have intense spontaneous horizontal jerk nystagmus that ultimately reverses direction during the attack. Generally, the nystagmus initially beats toward the affected ear (excitatory or irritative phase) before reversing direction within 12 hours (inhibitory or paretic phase). Subsequently, another reversal representing "recovery nystagmus" may occur.[33,80] During episodes, patients commonly experience nausea, vomiting, sweating, diarrhea, severe unsteadiness, and such head motion intolerance that they must lie perfectly still until the episode subsides. They often have some mild residual unsteadiness for 1 to 2 days.

Although the sensorineural hearing loss 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

after 4 years is inconsistent with Ménière disease. The diagnostic criteria for vestibular migraine require that at least half of the episodes be accompanied by migrainous symptoms, but they do not need to be headache. Probable vestibular migraine does not require a temporal relationship between episodic vestibular symptoms and migrainous symptoms.

This patient developed daily nonvertiginous dizziness and unsteadiness consistent with the entity persistent postural perceptual dizziness (PPPD). Vestibular migraine and generalized anxiety disorder are among the most common conditions leading to the development of PPPD. For her, although episodes of vestibular migraine a few times per year were briefly debilitating, she was much more affected by daily symptoms of PPPD, catastrophic thinking, and avoidance behavior. Management must consider her migraine headaches, episodic vertigo, PPPD, and anxiety.

Acute treatment for vestibular migraine episodes may include vestibular suppressants, antiemetics, or triptans. Because her migraine episodes (vestibular and headache) occurred less than 1 time per month, they may not warrant prophylactic medication, although it could be considered if they were particularly severe, prolonged, or difficult to treat acutely. Her PPPD would be best treated with a selective serotonin reuptake inhibitor (SSRI) or serotonin norepinephrine reuptake inhibitor (SNRI). Although any such medication may also help treat her anxiety, venlafaxine may be an ideal choice because it has some supporting data for migraine and vestibular migraine, as well. Habituation-type vestibular therapy should be initiated to help desensitize her to head motion, visual motion, and complex visual environments.[74] Cognitive-behavioral therapy may help address aspects of illness anxiety and avoidance behavior.

less varying. Tinnitus also generally becomes persistent but may still fluctuate in intensity. Estimates of eventual contralateral ear involvement vary widely, but ultimately roughly 30% (10% to 50%) of patients develop some bilateral hearing involvement.[81] Bilateral Ménière disease may produce slightly more severe (and bilateral) hearing loss with a flatter audiometric curve.[82]

Sudden, unexplained falls without vertigo or loss of consciousness can occur in a minority (less than 10%) of patients with Ménière disease. These *otolithic crises of Tumarkin*, or drop attacks, are often described by patients as if they have been forcefully shoved or thrown to the ground or as if the floor has been pulled out from under them.[83] They often occur within the first few years of the disease, with most affected patients having two or three attacks over the course of a year or less before they remit. Similar drop attacks have been described in patients with migraine, cardiac, or cerebrovascular disorders.[84]

Diagnosis

Ménière disease is a clinical diagnosis. Diagnostic criteria include specific audiometric findings. Imaging and vestibular laboratory testing sometimes play a role.

DIAGNOSTIC CRITERIA. In 2015, the Bárány Society, in collaboration with other international societies, jointly developed and published updated international consensus diagnostic criteria for Ménière disease (TABLE 4-5).[50] These updates included four major changes from the widely used 1995 American Academy of Otolaryngology–Head and Neck Surgery criteria: (1) elimination of the histologically "certain" and the "possible" categories, (2) requirement for audiometrically documented low- to medium-frequency hearing loss in the definite category only,

TABLE 4-5 **Diagnostic Criteria for Ménière Disease**[a]

Definite Ménière disease

- **A** Two or more spontaneous episodes of vertigo, each lasting 20 minutes to 12 hours
- **B** Audiometrically documented low- to medium-frequency sensorineural hearing loss[b] in one ear, defining the affected ear on at least one occasion before, during, or after one of the episodes of vertigo
- **C** Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear
- **D** Not better accounted for by another vestibular diagnosis

Probable Ménière disease

- **A** Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours
- **B** Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear
- **C** Not better accounted for by another vestibular diagnosis

[a] Reprinted from Lopez-Escamez JA, et al, J Vestib Res.[50] © 2015 IOS Press and the authors.

[b] Low-frequency sensorineural hearing loss is defined as increases in pure tone thresholds for bone-conducted sound that are worse in the affected ear than in the contralateral ear by at least 30 decibels hearing loss (dB HL) at each of two contiguous frequencies below 2000 Hz, or if bilateral, hearing loss must be worse than 35 dB HL or more at each of two contiguous frequencies below 2000 Hz. Demonstrating recovery of hearing loss at some point in time further supports the diagnosis of Ménière disease.

(3) auditory symptoms should be fluctuating, and (4) vertigo attack duration with an upper limit of 12 hours (definite) or 24 hours (probable).[85]

AUDIOMETRIC TESTING. Audiometrically documenting low- to medium-frequency sensorineural hearing loss (FIGURE 4-1) is important for establishing the diagnosis early on, especially because tinnitus and even subjective hearing loss can occur with episodes of vestibular migraine,[29,51] and other vestibular disorders such as superior canal dehiscence and vestibular schwannoma are associated with different patterns of hearing loss. Later on, audiometry can help determine the progression of hearing loss, involvement of the contralateral ear, potential benefit of hearing amplification, or candidacy for cochlear implantation.

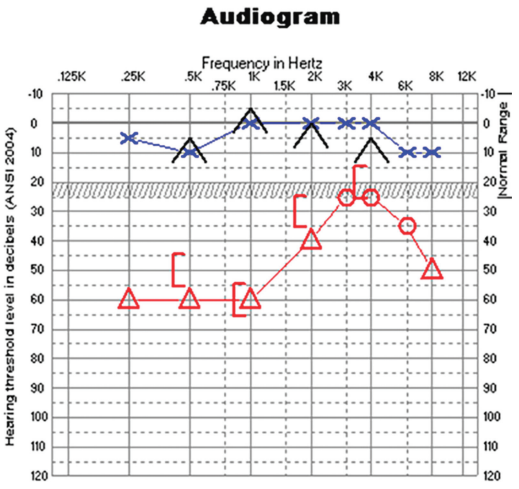

FIGURE 4-1
Typical audiometric pattern in Ménière disease. Pure tone thresholds for air conduction (*circles*), masked air conduction (*triangles*), and masked bone conduction (*brackets*) in the right ear indicate moderately severe low- and medium-frequency (250 to 1000 Hz) sensorineural hearing loss compared with normal air conduction hearing in the left ear (*X*).

ROLE OF IMAGING AND VESTIBULAR LABORATORY TESTING. Ménière disease remains a clinical diagnosis supported by audiometric findings (CASE 4-2). Given its overlapping symptoms with other conditions such as vestibular migraine, several tests have been investigated in hope of identifying markers that would discriminate patients with Ménière disease from those without or those with other vestibular disorders. Lack of a gold standard and a high frequency of some test abnormalities in other conditions have made this challenging.

MRI has been investigated to identify the presence of endolymphatic hydrops, particularly expansion of the saccule. However, conflicting results in patients with Ménière disease may be due to differences in MRI sequence parameters, hydrops grading methods, and patient inclusion criteria. It appears that saccular hydrops is associated with hearing loss and may be a function of disease duration, but endolymphatic hydrops on MRI may also be seen in some patients with vestibular migraine, tinnitus, autoimmune inner ear disease, or acute sensorineural hearing loss without vertigo, as well as frequently in the contralateral unaffected ear in patients with Ménière disease.[86] At this time, MRI does not yet have an established role in the diagnosis of Ménière disease, although MRI of the internal auditory canal with gadolinium may be used to exclude other retrocochlear causes of hearing loss and vertigo.

Vestibular laboratory testing may be normal or abnormal in patients with Ménière disease. Caloric testing is often normal early in the disease course, but in the first several years, caloric responsiveness in the affected ear frequently deteriorates, with a functional reduction of about 35% to 50%.[81] Video head

KEY POINTS

- 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.

impulse testing might provide a diagnostic complement to caloric testing in Ménière disease.[87,88] One study found that, among 606 subjects in a neuro-otology clinic with various vestibular disorders who underwent both tests, 27 of 73 patients with Ménière disease had discordant results, all with asymmetric caloric testing but normal symmetric video head impulse testing. This is in contrast to the 532 patients without Ménière disease, only 9 of whom

CASE 4-2

A 44-year-old man was evaluated for a 5-year history of episodic vertigo and auditory symptoms. Initially, 5 years ago he developed progressive right-sided hearing loss and intermittent tinnitus. Then 4 years ago, he began having spontaneous attacks 2 times per month of external spinning vertigo, generally lasting about 2 hours, accompanied by nausea and vomiting and preceded by right aural fullness and buzzing tinnitus, but he had no decrease in hearing during attacks. More recently, he had been experiencing episodic focal throbbing headaches accompanied by nausea about 2 times per month, but they were not temporally associated with his vertigo attacks. Neurologic examination, including detailed ocular motor and vestibular examination and otoscopy, was normal. With video-oculography and visual fixation removed, both horizontal headshaking and mastoid vibration produced left-beating nystagmus.

Audiometric testing was normal on the left but revealed moderate low- and medium-frequency sensorineural hearing loss on the right, with normal tympanogram and acoustic reflexes and 55% word recognition. Videonystagmography demonstrated normal ocular motor and gaze stability testing. A 39% right caloric weakness was observed. Video head impulse testing of both lateral canals was normal. Ocular and cervical vestibular-evoked myogenic potentials were normal and symmetric with 500-Hz tone bursts, but the right ear showed an abnormal upward shift in the most sensitive cervical vestibular-evoked myogenic potential threshold from 500 Hz to 1000 Hz.

COMMENT

In this patient with right-sided sensorineural hearing loss and episodic vertigo accompanied by tinnitus and aural fullness, evaluation demonstrated supportive findings for a diagnosis of Ménière disease. Diagnosing definite unilateral Ménière disease requires, in part, audiometrically documented low- to medium-frequency sensorineural hearing loss in the affected ear in which pure tone thresholds for bone-conducted sound are at least 30 decibels hearing loss (dB HL) higher at each of two contiguous frequencies below 2000 Hz compared with the contralateral ear. Function of the vestibulo-ocular reflex pathways serving the lateral canals was assessed two different ways. Dissociation between normal video head impulse test and abnormal ipsilesional caloric testing has been proposed as a potentially useful diagnostic finding in Ménière disease that may be the result of enlargement of the membranous duct causing a reduced response to caloric stimulation, as local convective flow dissipates hydrostatic pressure across the cupula.

showed dissociation. Thus, more than two-thirds of patients with caloric video head impulse testing dissociation had Ménière disease.[89] Vestibular-evoked myogenic potentials have been studied in Ménière disease. Asymmetrically reduced cervical and ocular vestibular-evoked myogenic potential amplitude to air-conducted sound but not bone-conducted vibration has been reported to be the most common profile in Ménière disease, although it is still abnormal in only 30% to 50% of patients compared with 10% to 20% of controls.[90] Other vestibular-evoked myogenic potential response metrics have been studied, including shifts in tuning frequency, that could ultimately be useful to discriminate Ménière disease from other causes of episodic vertigo or hearing loss.[91,92] Electrocochleography currently has little to no role in the diagnosis of Ménière disease.

Differential Diagnosis

The differential diagnosis of Ménière disease includes other conditions causing episodic vertigo or fluctuating progressive hearing loss (TABLE 4-6).

VERTEBROBASILAR TRANSIENT ISCHEMIC ATTACKS

Vertebrobasilar insufficiency from stenosis, embolism, or dissection may cause a stroke, producing an acute vestibular syndrome (discussed elsewhere in this issue). But vertebrobasilar ischemia may also manifest as TIAs of spontaneous vertigo. The notion that episodes of isolated vertigo can be the sole symptom of a TIA, preceding up to half of posterior circulation infarctions, has growing evidence in the literature.[93] Thus, although TIAs are an uncommon cause of episodic vertigo across the population, they are an important and dangerous cause to consider, especially in older patients with recent-onset symptoms and vascular risk factors.

Differential Diagnosis of Ménière Disease TABLE 4-6

- Autosomal dominant sensorineural hearing loss type 9 from the *COCH* gene or type 6/14 from the *WSF1* gene
- Autoimmune inner ear disease
- Cerebellopontine angle tumor (eg, meningioma)
- Cogan syndrome
- Endolymphatic sac tumor
- Neuroborreliosis
- Otosyphilis
- Paraneoplastic encephalitis with Kelch-like protein 11 antibodies and testicular cancer
- Perilymphatic fistula
- Superior canal dehiscence syndrome
- Susac syndrome
- Vertebrobasilar transient ischemic attack or stroke
- Vestibular migraine
- Vestibular paroxysmia (neurovascular compression syndrome)
- Vestibular schwannoma

Pathophysiology

The posterior circulation supplies both the central and peripheral structures of the vestibular system. The posterior inferior cerebellar artery branches from the vertebral artery and irrigates the caudal cerebellum (including the inferior vermis, nodulus, and uvula) and lateral medulla (including the caudal portion of the vestibular nuclei). The anterior inferior cerebellar artery (AICA) branches from the basilar artery to irrigate the anterior inferior cerebellum (including the flocculus), middle cerebellar peduncle, lateral pons, and inner ear. Transient ischemia of vestibular structures may arise from several mechanisms, such as atherosclerotic stenosis/thrombosis, artery-to-artery embolism, or arterial dissection. Cardioembolism would be less likely to cause multiple identical TIAs to the same vascular distribution. Very rarely, dynamic spondylotic compression of the vertebral arteries during extreme head rotation can produce vertebrobasilar ischemia with vertigo or near syncope, known as *rotational vertebral artery syndrome* or *bow hunter syndrome*.[94]

Clinical Features

Vertebrobasilar ischemia may produce a variety of brainstem, cerebellar, occipital lobe, medial temporal lobe, and thalamic symptoms, and such transient neurologic symptoms appear to be more common in the immediate days or weeks before vertebrobasilar territory strokes than carotid territory strokes. Although attacks of multiple simultaneous posterior circulation symptoms should alert the clinician to possible vertebrobasilar TIAs, a large population-based study showed that sudden spontaneous episodes of vertigo are the most common vertebrobasilar TIA symptom preceding a stroke and consist more often of isolated vertigo (preceding 51% of posterior circulation strokes) than accompanied by other posterior circulation symptoms, mimicking peripheral vertigo.[95] Of those isolated vertigo episodes, 52% lasted more than 1 hour, which is longer than typical TIAs.

Among patients in an emergency department whose acute transient isolated vertigo had resolved within 24 hours and who were asymptomatic when evaluated, other investigators found that 32% had a posterior circulation stroke based on diffusion-weighted imaging (55%) or only by cerebellar hypoperfusion on perfusion-weighted imaging (45%).[96] Although vertebrobasilar ischemia still represents a minority of patients presenting to the emergency department with vertigo, a population-based study found a 9.3-times increased relative risk of 30-day stroke in patients discharged with a diagnosis of peripheral vertigo compared with those with renal colic, with the highest relative risk of 50 at 7 days.[97] A large cohort study found that patients hospitalized for isolated vertigo from all causes had a threefold greater risk of stroke in the subsequent 4 years than a hospitalized control group, and patients with vertigo with three or more vascular risk factors had a 5.5-fold higher stroke risk than those without risk factors.[98] Among patients with vertebrobasilar TIAs before stroke, up to 21% may have isolated episodes of vertigo for at least 4 weeks as the only presenting symptom.[99]

Because the AICA supplies the internal auditory artery to irrigate the cochlea and vestibular labyrinth, sudden or fluctuating hearing loss in the context of vertigo does not automatically imply a benign peripheral vestibular disorder such as Ménière disease or labyrinthitis. Sudden hearing loss occurring in proximity to vertigo is associated with a significantly higher subsequent stroke

risk compared with hearing loss or vertigo alone, based on a large population-based study.[100] Recurrent transient hearing loss and tinnitus alone can be the prodrome days before an AICA infarction producing hearing loss, tinnitus, vertigo, facial palsy, and hemiataxia.[101,102] In fact, mounting evidence suggests that new hearing loss occurring with an acute vestibular syndrome favors a stroke as the most likely cause.[103]

Evaluation

Identifying the small number of patients with a cerebrovascular cause among all patients with spontaneous episodes of dizziness or vertigo is a challenge. The possibility of vertebrobasilar ischemia should be considered in any patient presenting with a recent onset of spontaneous episodes of vertigo with or without auditory symptoms, especially with episodes lasting minutes to 1 or 2 hours in older patients or those with vascular risk factors. Eliciting other posterior circulation symptoms may be diagnostically helpful, but they are often absent. Vertebrobasilar TIAs are an unlikely cause of recurrent vertigo that has been occurring for more than 6 months. The patient group at greatest risk of an overlooked cerebrovascular cause is young patients without vascular risk factors, although that group also has the highest prevalence of vestibular migraine, a much more common benign cause. New headache, neck pain, or recent neck trauma with vertigo may suggest the possibility of vertebral artery dissection.[104]

If the patient is still symptomatic at the time of evaluation, the HINTS Plus (Head Impulse, Nystagmus, and Test of Skew plus bedside hearing test by finger rub) battery can be used to examine the acute vestibular syndrome and differentiate a central from peripheral cause to guide further workup and management.[93,105]

If the patient with possible vertebrobasilar TIAs is not symptomatic at the time of examination and the neurologic, ocular motor, and vestibular examinations including positional testing are normal, then the diagnostic approach must be based on the index of suspicion for a vascular cause. Patients with probable TIAs based on symptom features or risk factors (eg, ABCD^2 [age, blood pressure, clinical features, duration, presence of diabetes mellitus] score greater than 3) should be admitted or undergo rapid outpatient evaluation with MRI (including diffusion-weighted imaging) and MRA or CT angiography of the head and neck vessels. If available, perfusion-weighted MRI may increase the yield of demonstrating ischemia in the acute transient vestibular syndrome.[96]

VESTIBULAR PAROXYSMIA

Occasionally patients report very brief (1 to a few seconds) episodes of vertigo occurring up to dozens of times per day. These can be a diagnostic mystery because often no other accompanying symptoms are present, and the examination between episodes is normal. Originally reported by Jannetta[106] as *disabling positional vertigo* attributed to neurovascular cross-compression of the vestibulocochlear nerve, this clinical syndrome is now referred to as *vestibular paroxysmia*.[52]

Clinical Features

Vestibular paroxysmia represents a clinical syndrome that is most often attributed to microvascular compression of the eighth cranial nerve, although identical symptoms can occur from other compressive lesions or without

KEY POINTS

- **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.**

evidence of a lesion at all. As with trigeminal neuralgia, the mechanism is thought to be ephaptic transmission, which refers to coupling of the action potentials via current flow through the extracellular space between adjacent axons. The temporal profile is similar to that of trigeminal neuralgia. The frequency of episodes varies widely, from one every few months to dozens per day. Most attacks occur spontaneously, although they can sometimes be triggered by position changes, head turns, loud noise, or hyperventilation, requiring differentiation from BPPV, rotational vertebral artery syndrome, superior canal dehiscence syndrome, or vestibular schwannoma, respectively. Attacks generally last only a few seconds or up to 1 minute. Patients often report an individually stereotyped type of spinning or nonspinning vertigo, fine shimmering binocular oscillopsia, and/or directional pulsion that can be associated with abrupt unsteadiness and falls. The simultaneous occurrence of monaural tinnitus or hyperacusis, or rarely hemifacial spasm, can occasionally localize the affected side.

Diagnosis

The vestibular and ocular motor examination is typically normal between attacks, although hyperventilation (while removing visual fixation) can sometimes induce nystagmus beating toward the affected side, suggesting an excitatory nystagmus.[107] In patients with such frequent attacks that they are occurring during the office examination, brief bursts of spontaneous horizontal-torsional nystagmus beating toward the affected ear may be seen coinciding with the patient's spontaneous episodes. Often, the nystagmus amplitude is so low that it is best seen by patiently waiting to observe fine intermittent jiggling of the eye's conjunctival vessels or shimmering of the optic disc during ophthalmoscopy time-locked to the patient's reported symptoms.

Diagnostic criteria for vestibular paroxysmia were established in 2016 by the Bárány Society based on clinical features and treatment response (TABLE 4-7).[52]

TABLE 4-7 Diagnostic Criteria for Vestibular Paroxysmia[a]

Vestibular paroxysmia (each point needs to be fulfilled)

- **A** At least 10 attacks of spontaneous spinning or nonspinning vertigo
- **B** Duration less than 1 minute
- **C** Stereotyped phenomenology in a particular patient
- **D** Response to a treatment with carbamazepine or oxcarbazepine
- **E** Not better accounted for by another diagnosis

Probable vestibular paroxysmia (each point needs to be fulfilled)

- **A** At least five attacks of spinning or nonspinning vertigo
- **B** Duration less than 5 minutes
- **C** Spontaneous occurrence or provoked by certain head movements
- **D** Not better accounted for by another diagnosis

[a] Reprinted from Strupp M, et al, J Vestib Res.[52] © IOS Press and the authors.

MRI with high-resolution constructive interference in steady-state (CISS) or fast imaging employing steady-state acquisition (FIESTA) sequences through the brainstem should be performed in patients with vestibular paroxysmia, in part to exclude a cerebellopontine angle tumor, arachnoid cyst, demyelinating lesion, or other structural cause for the episodes. Demonstrating neurovascular cross-compression is not necessary for the diagnosis, and the precise role of imaging in this regard requires further study. Although high-resolution MRI appears very sensitive at visualizing eighth cranial nerve compression, most often from the AICA near the nerve root entry zone, that finding is not specific for vestibular paroxysmia, as one study found 35% of unaffected control patients had similar neurovascular compression and another study found 42% of patients with vestibular paroxysmia had bilateral neurovascular compression despite presumably unilateral symptoms.[108,109]

The differential diagnosis for short attacks includes BPPV, otolithic crises of Tumarkin (in Ménière disease), paroxysmal brainstem attacks (from multiple sclerosis or stroke), or superior canal dehiscence syndrome. In longer-lasting attacks (longer than 5 minutes), vestibular migraine, Ménière disease, vertebrobasilar TIAs, and panic attacks should be considered

Treatment

Medical therapy is generally effective at reducing or eliminating attacks of vestibular paroxysmia. Treatment responsiveness is one of the diagnostic criteria. The majority of patients respond to carbamazepine 200 mg/d to 800 mg/d or oxcarbazepine 300 mg/d to 900 mg/d in divided doses.[110] Lacosamide may be another treatment option, particularly if contraindications to or side effects with carbamazepine or oxcarbazepine are present.[111] Microvascular decompression of the eighth cranial nerve could be considered for patients in whom the affected side is clearly established when they have responded to medical treatment (further supporting the diagnosis) but cannot tolerate an effective dose (CASE 4-3).

BENIGN RECURRENT VERTIGO

The entity known as *benign recurrent vertigo* is a condition of spontaneous episodes of vertigo without migrainous, neurologic, or otologic features that do not go on to cause any persistent vestibular or hearing loss.[112] No consensus diagnostic criteria exist, so the literature is quite sparse and variable. The temporal profile is generally similar to that of vestibular migraine, with recurrent spontaneous episodes of vestibular symptoms lasting minutes to 1 or 2 days.[113] Strong links to migraine exist, and today many such patients would meet the criteria for probable vestibular migraine if a current or past history of migraine exists.[7,8,15]

Clinical features and outcomes were examined in 66 patients (73% women) with benign recurrent vertigo.[113] Vertigo attack duration varied from minutes to 72 hours. Family history of migraine or recurrent vertigo was reported in 51%. Four of the 66 patients developed vestibular migraine at a median 32.5-month follow-up. Among 338 patients with benign recurrent vertigo, other investigators identified a subgroup of 35 with a new diagnostic entity they called *recurrent spontaneous vertigo with interictal headshaking nystagmus*.[114] These patients, with robust post–head-shaking nystagmus as an interictal marker of central vestibular dysfunction within the velocity storage mechanism, had a higher susceptibility to motion sickness than the other patients with benign recurrent vertigo.

KEY POINTS

- ***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.**

CASE 4-3

A 57-year-old woman was evaluated for a 2-year history of brief stereotyped paroxysms of tinnitus and oscillopsia. Her spells would begin with sudden static-like left tinnitus. Within 5 seconds, she developed fine shimmering binocular horizontal oscillopsia, without a clear sense of spinning, pulsion, or unsteadiness. The visual symptoms and then the tinnitus subsided within 20 seconds. Spells occurred spontaneously, without any clear trigger, hundreds of times per day.

She experienced several spells during examination, but it was difficult to appreciate any nystagmus upon direct inspection or even with infrared video-oculography. However, with fundoscopy, within a few seconds of her indicating the onset of tinnitus, she developed fine shimmering of the optic disc indicative of nystagmus lasting 10 seconds that correlated with the room oscillopsia experienced in the other eye. The tinnitus subsided a few seconds after the nystagmus stopped. The only other abnormal examination finding was an abnormal leftward head impulse test.

Caloric testing revealed a 54% left caloric weakness. Left ocular vestibular-evoked myogenic potential was absent. Mastoid vibration produced right-beating nystagmus. Previous MRI of the brain and internal auditory canals had been interpreted as normal. However, scrutinizing the T2-weighted images suggested signal abnormality within the prepontine cistern and cerebellopontine angles, with the left vestibulocochlear nerve taking a distorted path as it traversed through the cisternal segment. Repeat imaging confirmed what was felt to represent an epidermoid cyst in the prepontine cistern and cerebellopontine angles encasing and displacing the left seventh and eighth cranial nerves.

COMMENT

This case illustrates several important points. The syndrome of vestibular paroxysmia, although most commonly attributed to microvascular compression of the eighth cranial nerve, may be associated with normal neuroimaging or caused by other structural lesions, such as this epidermoid cyst. The temporal profile of this case is typical, with very brief but frequent spells. The character of vertigo is sometimes not one of spinning but rather of a fine shimmering binocular oscillopsia from very rapid, low-amplitude nystagmus (not unlike the spells of monocular oscillopsia seen in superior oblique myokymia). Sometimes, sudden directional pulsion can lead to falls. Although isolated vestibular symptoms are most common, concurrent monaural tinnitus during spells can add localizing value. Tests of vestibular function are generally normal in vestibular paroxysmia, although this patient with a structural lesion had evidence of accompanying vestibular dysfunction.

The consulting neurosurgeon estimated a 50% chance of helping her symptoms with resection of the cyst and a 10% to 20% chance of injuring her hearing or vestibular function. She was instead treated with carbamazepine 300 mg 2 times daily and quickly noted complete resolution of the spells of oscillopsia and a 10-fold reduction in the frequency of the now-isolated tinnitus spells.

PANIC ATTACKS

Panic attacks commonly cause dizziness, unsteadiness, or lightheadedness. Such vestibular symptoms are second only to cardiopulmonary symptoms (eg, dyspnea or chest pain) as a manifestation of panic attacks. Vertigo occurs less commonly and is usually less dramatic than the intense spinning of acute peripheral vestibular disorders. Panic attacks may be identified by their characteristic features listed in TABLE 4-8.[115] They may occur spontaneously or be triggered by identifiable fear-provoking stimuli. Although panic attacks are part of panic disorder, they may also occur in other anxiety, trauma-related, and obsessive-compulsive

Panic Disorder Diagnostic Criteria From the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition[a] TABLE 4-8

A Recurrent unexpected panic attacks. A panic attack is an abrupt surge of intense fear or intense discomfort that reaches a peak within minutes, during which time four (or more) of the following symptoms occur:

- Palpitations, pounding heart, or accelerated heart rate
- Sweating
- Trembling or shaking
- Sensations of shortness of breath or smothering
- Feelings of choking
- Chest pain or discomfort
- Nausea or abdominal distress
- Feeling dizzy, unsteady, light-headed, or faint
- Chills or heat sensations
- Paresthesias (numbness or tingling sensations)
- Derealization (feelings of unreality) or depersonalization (being detached from oneself)
- Fear of losing control or "going crazy"
- Fear of dying

B At least one of the attacks has been followed by 1 month (or more) of one or both of the following:

- Persistent concern or worry about additional panic attacks or their consequences (eg, losing control, having a heart attack, "going crazy")
- A significant maladaptive change in behavior related to the attacks (eg, behaviors designed to avoid having panic attacks, such as avoidance of exercise or unfamiliar situations)

C The disturbance is not attributable to the physiologic effects of a substance (eg, a drug of abuse, a medication) or another medical condition (eg, hyperthyroidism, cardiopulmonary disorders).

D The disturbance is not better explained by another mental disorder (eg, the panic attacks do not occur only in response to feared social situations, as in social anxiety disorder; in response to circumscribed phobic objects or situations, as in specific phobia; in response to obsessions, as in obsessive-compulsive disorder; in response to reminders of traumatic events, as in posttraumatic stress disorder; or in response to separation from attachment figures, as in separation anxiety disorder).

[a] Reprinted from the American Psychiatric Association.[115] © 2013 American Psychiatric Association.

disorders, in medical conditions such as hyperthyroidism, and during substance intoxication or withdrawal. In late adolescence and early adulthood, panic attacks are among the most common causes of episodic dizziness.

Patients with panic disorder are much more likely than control subjects to have uneasiness with heights, space and motion discomfort, and constant or fluctuating dizziness.[116] The dizziness from panic attacks is not related to underlying vestibular dysfunction, although patients with panic disorder are more likely than controls to have one or more nonspecific, nondiagnostic abnormalities on tests of basic vestibular reflexes that do not point toward any consistent pattern of central or peripheral vestibular dysfunction.[117] However, patients with panic disorder do sway more than controls on static and dynamic posturography that assesses higher-order integrated balance function, and the degree of postural instability correlates with the severity of phobic avoidance and anticipatory anxiety of situations associated with dizziness.[118] Patients with anxiety disorders and space and motion discomfort have more difficulty integrating visual, vestibular, and somatosensory cues, becoming overreliant on visual cues that are problematic in busy complex visual environments.[119]

Psychiatric and vestibular disorders interact in complex ways. Anxiety disorders, including panic and phobic disorders, are estimated to be the primary cause of vestibular symptoms in 8% to 10% of patients evaluated in specialty neuro-otology centers[120]; rates of psychiatric comorbidity in patients with structural or functional vestibular disorders are much higher still. Just like panic attacks can manifest with dizziness, acute vestibular syndromes frequently cause high levels of anxiety including panic attacks. High anxiety during and after acute vestibular episodes is associated with poor long-term outcomes and is more predictive of development of chronic dizziness than is the degree of peripheral vestibular injury.[121] Sometimes, preexisting psychiatric disorders that were not associated with dizziness or unsteadiness increase in severity with onset of a vestibular disorder and interact to contribute to overall morbidity.[122] Psychiatric disorders may also trigger functional vestibular disorders such as PPPD.

The most important part of the evaluation for patients with spontaneous episodes of dizziness is a careful history, which includes listening for symptoms or triggers that might suggest panic disorder as a cause. Clinicians should inquire about diagnoses or previous symptoms suggesting an anxiety disorder, particularly generalized anxiety disorder, panic disorder, or specific phobias. Such conditions can be the cause of vestibular symptoms, the result of a vestibular disorder, or a comorbidity that is necessary to identify and manage simultaneously. Evaluation of suspected panic attacks may need to rule out drug intoxication or withdrawal as well as medical conditions mimicking panic attacks, such as arrhythmia, angina, hyperthyroidism, pheochromocytoma, hypoglycemia, or temporal lobe epilepsy. When episodic dizziness is the prominent symptom, other vestibular disorders must be considered and potentially investigated. Panic disorder is typically managed with cognitive behavioral therapy or medications (mainly selective serotonin reuptake inhibitors [SSRIs]), or both.

DELAYED ORTHOSTATIC HYPOTENSION

Orthostatic hypotension typically presents as a triggered cause of dizziness, defined as a drop in blood pressure of at least 20 mm Hg systolic or 10 mm Hg diastolic when going from lying to sitting, lying to standing, or sitting to

standing. Classic orthostatic hypotension develops within 3 minutes of assuming upright posture, whereas initial (immediate) orthostatic hypotension is associated with a transient decrease in blood pressure within 15 seconds of standing. However, patients with delayed orthostatic hypotension have a gradual fall in blood pressure that takes more than 3 minutes of upright posture to develop.[123] In this case, 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. Of course, with orthostatic hypotension, the symptoms should resolve on sitting or lying down.

Patients experience the symptom quality of orthostatic dizziness in various ways. Although clinicians often expect a description of lightheadedness or faintness (ie, presyncope), patients may describe the symptoms instead or additionally as vertigo, unsteadiness, generalized weakness, tiredness, difficulty in thinking or concentrating, tremulousness, fading vision, or posterior head or neck discomfort, as well as experiencing tachycardia or palpitations.[124,125] Thus, clinicians should focus on the timing after the patient assumes an upright posture rather than the quality of symptom to appropriately suspect and diagnose delayed orthostatic hypotension.

In addition to orthostatic hypotension, the differential diagnosis for dizziness, vertigo, or unsteadiness with upright posture is broad and includes the following:

- postural tachycardia syndrome (POTS)
- PPPD
- bilateral vestibulopathy
- orthostatic tremor/myoclonus
- sensory neuropathy
- other gait disorders
- dizziness/vertigo due to cardiac problems
- anxiety and depressive disorders

Evaluation of suspected delayed orthostatic hypotension begins by querying for other symptoms that might suggest autonomic impairment (urinary incontinence, erectile dysfunction, anhidrosis), review of potentially causative medications (alpha-blockers, beta-blockers, diuretics), and office measurement of orthostatic blood pressure and heart rate with prolonged standing or while symptomatic. The head-up tilt test can be used to further evaluate forms of orthostatic hypotension, although reproducibility is suboptimal.[126] Additional autonomic testing may be required.

CARDIOGENIC DIZZINESS

Syncope, along with presyncope of various qualities (dizziness, vertigo), can be caused by several cardiac disorders. Bradycardia, tachycardia, or low cardiac output from heart failure or outflow obstruction can all lead to dizziness from global cerebral hypoperfusion. Dizziness is a prominent or presenting symptom in about 10% of patients with acute myocardial infarction.[127] A review of the limited literature found that, among patients with primary cardiovascular disorders who experience dizziness, actual vertigo is quite common (63%, including 37% for whom vertigo was the only dizziness type).[128] Paroxysmal

KEY POINTS

- 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.

arrhythmias typically cause episodic dizziness/vertigo lasting seconds to minutes, which may be accompanied by fading vision, bilateral tinnitus or hearing loss, palpitations, chest pain, or dyspnea and progress to syncope. Unlike with orthostatic hypotension, episodes may occur in any position. Arrhythmias may be suspected by the presence of an underlying cardiac disorder, medical condition promoting arrhythmias (thyrotoxicosis, hyperkalemia), or arrhythmogenic drugs. Evaluation is guided by a suspected cause but typically begins with an ECG and Holter monitor.

HYPOGLYCEMIA

Acute hypoglycemia may cause dizziness or vertigo along with confusion, anxiety, drowsiness, tremor, palpitations, sweating, and paresthesia. Hypoglycemia occurs most commonly in patients being treated for diabetes, particularly with insulin. Symptomatic hypoglycemia is rare in otherwise healthy individuals, although insulinomas or other causes of endogenous hyperinsulinism, as well as other drugs or hormone deficiencies, can produce hypoglycemia. Evaluation first requires demonstrating low serum glucose while symptomatic and relief of those symptoms when the serum glucose is raised. Further evaluation seeks to identify the specific cause.

CONCLUSION

Diagnosing patients with spontaneous episodes of dizziness or vertigo does not need to be a daunting task. Only a handful of conditions are commonly encountered in clinical practice. Because most patients are asymptomatic when evaluated in the office and have normal examinations, diagnosis relies heavily on the history. More important than the vestibular symptom quality are the timing, triggers, accompanying symptoms, and associated comorbidities. Many disorders are diagnosed based on clinical diagnostic criteria alone. Carefully selected tests including audiometric evaluation, neuroimaging, vestibular laboratory testing, or cardiovascular evaluation may be helpful, but overreliance on indiscriminate and costly investigations risks uncovering irrelevant or diagnostically misleading abnormalities. Although most of the conditions are treatable, the treatments are vastly different, and some, such as vertebrobasilar TIAs or arrhythmias, are potentially deadly. Thus, it is important to strive for a specific diagnosis to guide ongoing care.

USEFUL WEBSITE

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS
This web page presents a collection of freely available formal consensus definition articles by the Bárány Society.
jvr-web.org/ICVD.html

REFERENCES

1 Drachman DA, Hart CW. An approach to the dizzy patient. Neurology 1972;22(4):323-334. doi:10.1212/wnl.22.4.323

2 Edlow JA, Gurley KL, Newman-Toker DE. A new diagnostic approach to the adult patient with acute dizziness. J Emerg Med 2018;54(4):469-483. doi:10.1016/j.jemermed.2017.12.024

3 Newman-Toker DE, Cannon LM, Stofferahn ME, et al. Imprecision in patient reports of dizziness symptom quality: a cross-sectional study conducted in an acute care setting. Mayo Clin Proc 2007;82(11):1329-1340. doi:10.4065/82.11.1329

4 Bisdorff A, Von Brevern M, Lempert T, Newman-Toker DE. Classification of vestibular symptoms: towards an international classification of vestibular disorders. J Vestib Res 2009;19(1-2): 1-13. doi:10.3233/VES-2009-0343

5 Episodic vestibular syndrome. International classification of diseases 11th revision. Accessed December 17, 2020. icd.who.int/browse11/l-m/en#/http://id.who.int/icd/entity/1402706403

6 Olesen J. International classification of headache disorders. Lancet Neurol 2018;17(5):396-397. doi:10.1016/S1474-4422(18)30085-1

7 Cha YH, Lee H, Santell LS, Baloh RW. Association of benign recurrent vertigo and migraine in 208 patients. Cephalalgia 2009;29(5):550. doi:10.1016/S1474-4422(18)30085-1

8 Oh AK, Lee H, Jen JC, et al. Familial benign recurrent vertigo. Am J Med Genet 2001;100(4): 287-291.doi:10.1002/ajmg.1294

9 Neuhauser HK, Radtke A, von Brevern M, et al. Migrainous vertigo: prevalence and impact on quality of life. Neurology 2006;67(6):1028-1033. doi:10.1212/01.wnl.0000237539.09942.06

10 Formeister EJ, Rizk HG, Kohn MA, Sharon JD. The epidemiology of vestibular migraine: a population-based survey study. Otol Neurotol 2018;39(8):1037-1044. doi:10.1097/MAO.0000000000001900

11 Dieterich M, Brandt T. Episodic vertigo related to migraine (90 cases): vestibular migraine? J Neurol 1999;246(10):883-892. doi:10.1007/s004150050478

12 Johnson GD. Medical management of migraine-related dizziness and vertigo. Laryngoscope 1998;108(1 pt 2):1-28. doi:10.1097/00005537-199801001-00001

13 Beh SC, Masrour S, Smith SV, Friedman DI. The spectrum of vestibular migraine: clinical features, triggers, and examination findings. Headache 2019;59(5):727-740. doi:10.1111/head.13484

14 Neuhauser H, Leopold M, von Brevern M, et al. The interrelations of migraine, vertigo, and migrainous vertigo. Neurology 2001;56(4):436-441.

15 Lee H, Sohn SI, Jung DK, et al. Migraine and isolated recurrent vertigo of unknown cause. Neurol Res 2002;24(7):663-665. doi:10.1179/016164102101200726

16 Drummond PD. Triggers of motion sickness in migraine sufferers. Headache 2005;45(6): 653-656. doi:10.1111/j.1526-4610.2005.05132.x

17 Furman JM, Marcus DA, Balaban CD. Migrainous vertigo: development of a pathogenetic model and structured diagnostic interview. Curr Opin Neurol 2003;16(1):5-13. doi:10.1097/01.wco.0000053582.70044.e2

18 Eggers SD, Staab JP, Neff BA, et al. Investigation of the coherence of definite and probable vestibular migraine as distinct clinical entities. Otol Neurotol 2011;32(7):1144-1151. doi:10.1097/MAO.0b013e31822a1c67

19 Drummond PD. Motion sickness and migraine: optokinetic stimulation increases scalp tenderness, pain sensitivity in the fingers and photophobia. Cephalalgia 2002;22(2):117-124. doi:10.1046/j.1468-2982.2002.00332.x

20 King S, Wang J, Priesol AJ, Lewis RF. Central integration of canal and otolith signals is abnormal in vestibular migraine. Front Neurol 2014;5:233. doi:10.3389/fneur.2014.00233

21 Winnick A, Sadeghpour S, Otero-Millan J, et al. Errors of upright perception in patients with vestibular migraine. Front Neurol 2018;9:892. doi:10.3389/fneur.2018.00892

22 Jen J, Kim GW, Baloh RW. Clinical spectrum of episodic ataxia type 2. Neurology 2004;62(1): 17-22. doi:10.1212/01.wnl.0000101675.61074.50

23 Baloh RW, Foster CA, Yue Q, Nelson SF. Familial migraine with vertigo and essential tremor. Neurology 1996;46(2):458-460. doi:10.1212/wnl.46.2.458

24 Radtke A, Lempert T, Gresty MA, et al. Migraine and Ménière's disease: is there a link? Neurology 2002;59(11):1700-1704. doi:10.1212/01.wnl.0000036903.22461.39

25 Huang TC, Wang SJ, Kheradmand A. Vestibular migraine: an update on current understanding and future directions. 2020;40(1):107-121. doi:10.1177/0333102419869317

26 von Brevern M, Radtke A, Clarke AH, Lempert T. Migrainous vertigo presenting as episodic positional vertigo. Neurology 2004;62(3):469-472. doi:10.1212/01.wnl.0000106949.55346.cd

27 Brantberg K, Trees N, Baloh RW. Migraine-associated vertigo. Acta Otolaryngol 2005;125(3): 276-279. doi:10.1080/00016480510003165

28 Zhang Y, Kong Q, Chen J, et al. International Classification of Headache Disorders 3rd edition beta-based field testing of vestibular migraine in China: demographic, clinical characteristics, audiometric findings and diagnosis statues. Cephalalgia 2016;36(3):240-248. doi:10.1177/0333102415587704

29 Neff BA, Staab JP, Eggers SD, et al. Auditory and vestibular symptoms and chronic subjective dizziness in patients with Ménière's disease, vestibular migraine, and Ménière's disease with concomitant vestibular migraine. Otol Neurotol 2012;33(7):1235-1244. doi:10.1097/MAO.0b013e31825d644a

30 Lempert T, Neuhauser H. Epidemiology of vertigo, migraine and vestibular migraine. J Neurol 2009;256(3):333-338. doi:10.1007/s00415-009-0149-2

31 Cutrer FM, Baloh RW. Migraine-associated dizziness. Headache 1992;32(6):300-304. doi:10.1111/j.1526-4610.1992.hed3206300.x

32 von Brevern M, Zeise D, Neuhauser H, et al. Acute migrainous vertigo: clinical and oculographic findings. Brain 2005;128(pt 2):365-374. doi:10.1093/brain/awh351

33 Young AS, Lechner C, Bradshaw AP, et al. Capturing acute vertigo: a vestibular event monitor. Neurology 2019;92(24):e2743-e2753. doi:10.1212/WNL.0000000000007644

34 Polensek SH, Tusa RJ. Nystagmus during attacks of vestibular migraine: an aid in diagnosis. Audiol Neurootol 2010;15(4):241-246. doi:10.1159/000255440

35 Jeong SH, Oh SY, Kim HJ, et al. Vestibular dysfunction in migraine: effects of associated vertigo and motion sickness. J Neurol 2010;257(6): 905-912. doi:10.1007/s00415-009-5435-5

36 Murdin L, Chamberlain F, Cheema S, et al. Motion sickness in migraine and vestibular disorders. J Neurol Neurosurg Psychiatry 2015;86(5): 585-587. doi:10.1136/jnnp-2014-308331

37 Staab JP, Eckhardt-Henn A, Horii A, et al. Diagnostic criteria for persistent postural-perceptual dizziness (PPPD): consensus document of the Committee for the Classification of Vestibular Disorders of the Barany Society. J Vestib Res 2017;27(4):191-208. doi:10.3233/VES-170622

38 Eggers SD, Neff BA, Shepard NT, Staab JP. Comorbidities in vestibular migraine. J Vestib Res 2014;24(5-6):387-395. doi:10.3233/VES-140525

39 Best C, Eckhardt-Henn A, Tschan R, Dieterich M. Psychiatric morbidity and comorbidity in different vestibular vertigo syndromes. Results of a prospective longitudinal study over one year. J Neurol 2009;256(1):58-65. doi:10.1007/s00415-009-0038-8

40 Best C, Tschan R, Eckhardt-Henn A, Dieterich M. Who is at risk for ongoing dizziness and psychological strain after a vestibular disorder? Neuroscience 2009;164(4):1579-1587. doi:10.1016/j.neuroscience.2009.09.034

41 Boldingh MI, Ljostad U, Mygland A, Monstad P. Comparison of interictal vestibular function in vestibular migraine vs migraine without vertigo. Headache 2013;53(7):1123-1133. doi:10.1111/head.12129

42 Casani AP, Sellari-Franceschini S, Napolitano A, et al. Otoneurologic dysfunctions in migraine patients with or without vertigo. Otol Neurotol 2009; 30(7):961-967. doi:10.1097/MAO.0b013e3181b4e780

43 Cass SP, Furman JM, Ankerstjerne K, et al. Migraine-related vestibulopathy. Ann Otol Rhinol Laryngol 1997;106(3):182-189. doi:10.1177/000348949710600302

44 Radtke A, von Brevern M, Neuhauser H, et al. Vestibular migraine: long-term follow-up of clinical symptoms and vestibulo-cochlear findings. Neurology 2012;79(15):1607-1614. doi:10.1212/WNL.0b013e31826e264f

45 Teggi R, Colombo B, Bernasconi L, et al. Migrainous vertigo: results of caloric testing and stabilometric findings. Headache 2009;49(3): 435-444. doi:10.1111/j.1526-4610.2009.01338.x

46 Kang WS, Lee SH, Yang CJ, et al. Vestibular function tests for vestibular migraine: clinical implication of video head impulse and caloric tests. Front Neurol 2016;7:166. doi:10.3389/fneur.2016.00166

47 Inoue A, Egami N, Fujimoto C, et al. Vestibular evoked myogenic potentials in vestibular migraine: do they help differentiating from Ménière's disease? Ann Otol Rhinol Laryngol 2016;125(11):931-937. doi:10.1177/0003489416665192

48 Taylor RL, Zagami AS, Gibson WP, et al. Vestibular evoked myogenic potentials to sound and vibration: characteristics in vestibular migraine that enable separation from Ménière's disease. Cephalalgia 2012;32(3):213-225. doi:10.1177/0333102411434166

49 Zuniga MG, Janky KL, Schubert MC, Carey JP. Can vestibular-evoked myogenic potentials help differentiate Ménière disease from vestibular migraine? Otolaryngol Head Neck Surg 2012; 146(5):788-796. doi:10.1177/0194599811434073

50 Lopez-Escamez JA, Carey J, Chung WH, et al. Diagnostic criteria for Ménière's disease. J Vestib Res 2015;25(1):1-7. doi:10.3233/VES-150549

51 Lopez-Escamez JA, Dlugaiczyk J, Jacobs J, et al. Accompanying symptoms overlap during attacks in Ménière's disease and vestibular migraine. Front Neurol 2014;5:265. doi:10.3389/fneur.2014.00265

52 Strupp M, Lopez-Escamez JA, Kim JS, et al. Vestibular paroxysmia: diagnostic criteria. J Vestib Res 2016;26(5-6):409-415. doi:10.3233/VES-160589

53 Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalalgia 2013;33(9):629-808. doi:10.1177/0333102413485658

54 Lempert T, Olesen J, Furman J, et al. Vestibular migraine: diagnostic criteria. J Vestib Res 2012; 22(4):167-172. doi:10.3233/VES-2012-0453

55 Radtke A, Neuhauser H, von Brevern M, et al. Vestibular migraine–validity of clinical diagnostic criteria. Cephalalgia 2011;31(8):906-913. doi:10.1177/0333102411405228

56 Neuhauser H, Radtke A, von Brevern M, Lempert T. Zolmitriptan for treatment of migrainous vertigo: a pilot randomized placebo-controlled trial. Neurology 2003;60(5):882-883. doi:10.1212/01.wnl.0000049476.40047.a3

57 Furman JM, Marcus DA, Balaban CD. Rizatriptan reduces vestibular-induced motion sickness in migraineurs. J Headache Pain 2011;12(1):81-88. doi:10.1007/s10194-010-0250-z

58 Beh SC. External trigeminal nerve stimulation: potential rescue treatment for acute vestibular migraine. J Neurol Sci 2019;408:116550. doi:10.1016/j.jns.2019.116550

59 Beh SC, Friedman DI. Acute vestibular migraine treatment with noninvasive vagus nerve stimulation. Neurology 2019;93(18):e1715-e1719. doi:10.1212/WNL.0000000000008388

60 Lepcha A, Amalanathan S, Augustine AM, et al. Flunarizine in the prophylaxis of migrainous vertigo: a randomized controlled trial. Eur Arch Otorhinolaryngol 2014;271(11):2931-2936. doi:10.1007/s00405-013-2786-4

61 Salviz M, Yuce T, Acar H, et al. Propranolol and venlafaxine for vestibular migraine prophylaxis: a randomized controlled trial. Laryngoscope 2016; 126(1):169-174. doi:10.1002/lary.25445

62 Liu F, Ma T, Che X, et al. The efficacy of venlafaxine, flunarizine, and valproic acid in the prophylaxis of vestibular migraine. Front Neurol 2017;8:524. doi:10.3389/fneur.2017.00524

63 Bayer O, Adrion C, Al Tawil A, et al. Results and lessons learnt from a randomized controlled trial: prophylactic treatment of vestibular migraine with metoprolol (PROVEMIG). Trials 2019;20(1):813. doi:10.1186/s13063-019-3903-5

64 Bikhazi P, Jackson C, Ruckenstein MJ. Efficacy of antimigrainous therapy in the treatment of migraine-associated dizziness. Am J Otol 1997; 18(3):350-354.

65 Bisdorff AR. Treatment of migraine related vertigo with lamotrigine an observational study. Bull Soc Sci Med Grand Duche Luxemb 2004; 2(2):103-108.

66 Carmona S, Settecase N. Use of topiramate (topamax) in a subgroup of migraine-vertigo patients with auditory symptoms. Ann N Y Acad Sci 2005;1039:517-520. doi:10.1196/annals.1325.057

67 Iwasaki S, Ushio M, Chihara Y, et al. Migraine-associated vertigo: clinical characteristics of Japanese patients and effect of lomerizine, a calcium channel antagonist. Acta Otolaryngol Suppl 2007(559):45-49. doi:10.1080/03655230701596491

68 Maione A. Migraine-related vertigo: diagnostic criteria and prophylactic treatment. Laryngoscope 2006;116(10):1782-1786. doi:10.1097/01.mlg.0000231302.77922.c5

69 Waterston J. Chronic migrainous vertigo. J Clin Neurosci 2004;11(4):384-388. doi:10.1016/j.jocn.2003.08.008

70 Brodsky JR, Cusick BA, Zhou G. Evaluation and management of vestibular migraine in children: experience from a pediatric vestibular clinic. Eur J Paediatr Neurol 2016;20(1):85-92. doi:10.1016/j.ejpn.2015.09.011

71 Gode S, Celebisoy N, Kirazli T, et al. Clinical assessment of topiramate therapy in patients with migrainous vertigo. Headache 2010;50(1): 77-84. doi:10.1111/j.1526-4610.2009.01496.x

72 Taghdiri F, Togha M, Razeghi Jahromi S, Refaeian F. Cinnarizine for the prophylaxis of migraine associated vertigo: a retrospective study. Springerplus 2014;3:231. doi:10.1186/2193-1801-3-231

73 Alghadir AH, Anwer S. Effects of vestibular rehabilitation in the management of a vestibular migraine: a review. Front Neurol 2018;9:440. doi:10.3389/fneur.2018.00440

74 Thompson KJ, Goetting JC, Staab JP, Shepard NT. Retrospective review and telephone follow-up to evaluate a physical therapy protocol for treating persistent postural-perceptual dizziness: a pilot study. J Vestib Res 2015;25(2): 97-103; quiz 103-104. doi:10.3233/VES-150551

75 Rauch SD, Merchant SN, Thedinger BA. Ménière's syndrome and endolymphatic hydrops. Double-blind temporal bone study. Ann Otol Rhinol Laryngol 1989;98(11):873-883. doi:10.1177/000348948909801108

76 Ishiyama G. Selected otologic disorders causing dizziness. Continuum (Minneap Minn) 2021; 27(2, Neuro-otology):468-490.

77 Fife TD, Tourkevich R. Tinnitus, hyperacusis, otalgia, and hearing loss. Continuum (Minneap Minn) 2021;27(2, Neuro-otology):491-525.

78 Harris JP, Alexander TH. Current-day prevalence of Ménière's syndrome. Audiol Neurootol 2010; 15(5):318-322. doi:10.1159/000286213

79 Perez-Garrigues H, Lopez-Escamez JA, Perez P, et al. Time course of episodes of definitive vertigo in Ménière's disease. Arch Otolaryngol Head Neck Surg 2008;134(11):1149-1154. doi:10.1001/archotol.134.11.1149

80 Eggers SDZ, Bisdorff A, von Brevern M, et al. Classification of vestibular signs and examination techniques: nystagmus and nystagmus-like movements. J Vestib Res 2019;29(2-3):57-87. doi:10.3233/VES-190658

81 Huppert D, Strupp M, Brandt T. Long-term course of Ménière's disease revisited. Acta Otolaryngol 2010;130(6):644-651. doi:10.3109/00016480903382808

82 Belinchon A, Perez-Garrigues H, Tenias JM, Lopez A. Hearing assessment in Ménière's disease. Laryngoscope 2011;121(3):622-626. doi:10.1002/lary.21335

83 Baloh RW, Jacobson K, Winder T. Drop attacks with Ménière's syndrome. Ann Neurol 1990; 28(3):384-387. doi:10.1002/ana.410280314

84 Ishiyama G, Ishiyama A, Baloh RW. Drop attacks and vertigo secondary to a non-Ménière otologic cause. Arch Neurol 2003;60(1):71-75. doi:10.1001/archneur.60.1.71

85 Goebel JA. 2015 Equilibrium Committee Amendment to the 1995 AAO-HNS Guidelines for the Definition of Ménière's Disease. Otolaryngol Head Neck Surg 2016;154(3):403-404. doi:10.1177/0194599816628524

86 Lopez-Escamez JA, Attyé A. Systematic review of magnetic resonance imaging for diagnosis of Ménière disease. J Vestib Res 2019;29(2-3): 121-129. doi:10.3233/VES-180646

87 McCaslin DL, Rivas A, Jacobson GP, Bennett ML. The dissociation of video head impulse test (vHIT) and bithermal caloric test results provide topological localization of vestibular system impairment in patients with "definite" Ménière's disease. Am J Audiol 2015;24(1):1-10. doi:10.1044/2014_AJA-14-0040

88 McGarvie LA, Curthoys IS, MacDougall HG, Halmagyi GM. What does the dissociation between the results of video head impulse versus caloric testing reveal about the vestibular dysfunction in Ménière's disease? Acta Otolaryngol 2015;135(9):859-865. doi:10.3109/00016489.2015.1015606

89 Hannigan IP, Welgampola MS, Watson SRD. Dissociation of caloric and head impulse tests: a marker of Ménière's disease. J Neurol 2019. doi:10.1007/s00415-019-09431-9

90 Taylor RL, Wijewardene AA, Gibson WPR, et al. The vestibular evoked-potential profile of Ménière's disease. Clin Neurophysiol 2011;122(6): 1256-1263. doi:10.1016/j.clinph.2010.11.009

91 Noij KS, Herrmann BS, Guinan JJ Jr, Rauch SD. Cervical vestibular evoked myogenic potentials in Ménière's disease: a comparison of response metrics. Otol Neurotol 2019;40(3):e215-e224. doi:10.1097/MAO.0000000000002092

92 Rosengren SM, Colebatch JG, Young AS, et al. Vestibular evoked myogenic potentials in practice: methods, pitfalls and clinical applications. Clin Neurophysiol Pract 2019; 4:47-68. doi:10.1016/j.cnp.2019.01.005

93 Saber Tehrani AS, Kattah JC, Kerber KA, et al. Diagnosing stroke in acute dizziness and vertigo: pitfalls and pearls. Stroke 2018;49(3):788-795. doi:10.1161/STROKEAHA.117.016979

94 Choi KD, Choi JH, Kim JS, et al. Rotational vertebral artery occlusion: mechanisms and long-term outcome. Stroke 2013;44(7):1817-1824. doi:10.1161/STROKEAHA.113.001219

95 Paul NL, Simoni M, Rothwell PM, Oxford Vascular Study. Transient isolated brainstem symptoms preceding posterior circulation stroke: a population-based study. Lancet Neurol 2013; 12(1):65-71. doi:10.1016/S1474-4422(12)70299-5

96 Choi JH, Park MG, Choi SY, et al. Acute transient vestibular syndrome: prevalence of stroke and efficacy of bedside evaluation. Stroke 2017;48(3): 556-562. doi:10.1161/STROKEAHA.116.015507

97 Atzema CL, Grewal K, Lu H, et al. Outcomes among patients discharged from the emergency department with a diagnosis of peripheral vertigo. Ann Neurol 2016;79(1):32-41. doi:10.1002/ana.24521

98 Lee CC, Su YC, Ho HC, et al. Risk of stroke in patients hospitalized for isolated vertigo: a four-year follow-up study. Stroke 2011;42(1): 48-52. doi:10.1161/STROKEAHA.110.597070

99 Gomez CR, Cruz-Flores S, Malkoff MD, et al. Isolated vertigo as a manifestation of vertebrobasilar ischemia. Neurology 1996;47(1): 94-97. doi:10.1212/wnl.47.1.94

100 Chang TP, Wang Z, Winnick AA, et al. Sudden hearing loss with vertigo portends greater stroke risk than sudden hearing loss or vertigo alone. J Stroke Cerebrovasc Dis 2018;27(2):472-478. doi:10.1016/j.jstrokecerebrovasdis.2017.09.033

101 Lee H, Cho YW. Auditory disturbance as a prodrome of anterior inferior cerebellar artery infarction. J Neurol Neurosurg Psychiatry 2003; 74(12):1644-1648. doi:10.1136/jnnp.74.12.1644

102 Park JH, Kim H, Han HJ. Recurrent audiovestibular disturbance initially mimicking Ménière's disease in a patient with anterior inferior cerebellar infarction. Neurol Sci 2008; 29(5):359-362. doi:10.1007/s10072-008-0996-0

103 Newman-Toker DE, Kerber KA, Hsieh YH, et al. HINTS outperforms ABCD2 to screen for stroke in acute continuous vertigo and dizziness. Acad Emerg Med 2013;20(10):986-996. doi:10.1111/acem.12223

104 Gottesman RF, Sharma P, Robinson KA, et al. Clinical characteristics of symptomatic vertebral artery dissection: a systematic review. Neurologist 2012;18(5):245-254. doi:10.1097/NRL.0b013e31826754e1

105 Choi KD, Kim JS. Vascular vertigo: updates. J Neurol 2019;266(8):1835-1843. doi:10.1007/s00415-018-9040-3

106 Jannetta PJ. Neurovascular cross-compression in patients with hyperactive dysfunction symptoms of the eighth cranial nerve. Surg Forum 1975;26:467-469.

107 Ward BK, Gold DR. Tinnitus, oscillopsia, and hyperventilation-induced nystagmus: vestibular paroxysmia. Open J Clin Med Case Rep 2016; 2(7):1100.

108 Best C, Gawehn J, Krämer HH, et al. MRI and neurophysiology in vestibular paroxysmia: contradiction and correlation. J Neurol Neurosurg Psychiatry 2013;84(12):1349-1356. doi: 10.1136/jnnp-2013-305513

109 Hüfner K, Barresi D, Glaser M, et al. Vestibular paroxysmia: diagnostic features and medical treatment. Neurology 2008;71(13):1006-1014. doi: 10.1212/01.wnl.0000326594.91291.f8

110 Bayer O, Brémová T, Strupp M, Hüfner K. A randomized double-blind, placebo-controlled, cross-over trial (Vestparoxy) of the treatment of vestibular paroxysmia with oxcarbazepine. J Neurol 2018;265(2):291-298. doi:10.1007/ s00415-017-8682-x

111 Strupp M, Elger C, Goldschagg N. Treatment of vestibular paroxysmia with lacosamide. Neurol Clin Pract 2019;9(6):539-541. doi:10.1212/ CPJ.0000000000000610

112 Slater R. Benign recurrent vertigo. J Neurol Neurosurg Psychiatry 1979;42(4):363-367. doi:10.1136/jnnp.42.4.363

113 Pan Q, Zhang Y, Zhang S, et al. Clinical features and outcomes of benign recurrent vertigo: a longitudinal study. Acta Neurol Scand 2020; 141(5):374-379. doi:10.1111/ane.13214

114 Lee SU, Choi JY, Kim HJ, Kim JS. Recurrent spontaneous vertigo with interictal headshaking nystagmus. Neurology 2018;90(24):e2135-e2145. doi:10.1212/WNL.0000000000005689

115 American Psychiatric Association. Diagnostic and statistical manual of mental disorders, fifth edition (DSM-5). Washington, DC: American Psychiatric Association, 2013:947.

116 Jacob RG, Redfern MS, Furman JM. Space and motion discomfort and abnormal balance control in patients with anxiety disorders. J Neurol Neurosurg Psychiatry 2009;80(1):74-78. doi:10.1136/jnnp.2007.136432

117 Tecer A, Tukel R, Erdamar B, Sunay T. Audiovestibular functioning in patients with panic disorder. J Psychosom Res 2004;57(2): 177-182. doi:10.1016/s0022-3999(03)00568-3

118 Perna G, Dario A, Caldirola D, et al. Panic disorder: the role of the balance system. J Psychiatr Res 2001;35(5):279-286. doi:10.1016/s0022-3956(01)00031-0

119 Redfern MS, Furman JM, Jacob RG. Visually induced postural sway in anxiety disorders. J Anxiety Disord 2007;21(5):704-716. doi:10.1016/j.janxdis.2006.09.002

120 Staab JP, Ruckenstein MJ. Expanding the differential diagnosis of chronic dizziness. Arch Otolaryngol Head Neck Surg 2007;133(2):170-176. doi:10.1001/archotol.133.2.170

121 Godemann F, Siefert K, Hantschke-Brüggemann M, et al. What accounts for vertigo one year after neuritis vestibularis - anxiety or a dysfunctional vestibular organ? J Psychiatr Res 2005;39(5):529-234. doi:10.1016/j.jpsychires.2004.12.006

122 Staab JP, Ruckenstein MJ. Which comes first? Psychogenic dizziness versus otogenic anxiety. Laryngoscope 2003;113(10):1714-1718. doi:10.1097/00005537-200310000-00010

123 Gibbons CH, Freeman R. Delayed orthostatic hypotension: a frequent cause of orthostatic intolerance. Neurology 2006;67(1):28-32. doi:10.1212/01.wnl.0000223828.28215.0b

124 Kim HA, Yi HA, Lee H. Recent advances in orthostatic hypotension presenting orthostatic dizziness or vertigo. Neurol Sci 2015;36(11): 1995-2002. doi:10.1007/s10072-015-2363-2

125 Low PA, Opfer-Gehrking TL, McPhee BR, et al. Prospective evaluation of clinical characteristics of orthostatic hypotension. Mayo Clin Proc 1995; 70(7):617-622. doi:10.4065/70.7.617

126 Ward C, Kenny RA. Reproducibility of orthostatic hypotension in symptomatic elderly. Am J Med 1996;100(4):418-422. doi:10.1016/S0002-9343(97)89517-4

127 Culić V, Mirić D, Eterović D. Correlation between symptomatology and site of acute myocardial infarction. Int J Cardiol 2001;77(2-3):163-168. doi:10.1016/s0167-5273(00)00414-9

128 Newman-Toker DE, Dy FJ, Stanton VA, et al. How often is dizziness from primary cardiovascular disease true vertigo? A systematic review. J Gen Intern Med 2008;23(12):2087-2094. doi:10.1007/s11606-008-0801-z