CME

Recognizing vestibular migraine

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ABSTRACT

Vestibular migraine is a recently defined disorder that is underrecognized by clinicians across specialties. Acute attacks cause symptoms of migraine headaches as well as vestibular symptoms such as dizziness or vertigo. Further research is needed to determine the pathophysiology of vestibular migraine. No consensus treatment guidelines exist for this condition, and treatment is based on other migraine guidelines. Clinicians who are aware of vestibular migraine can speed diagnosis and treatment for patients and improve their quality of life.

Keywords: vestibular, migraine, headache, vertigo, migrainous vertigo, vestibulopathy

Learning objectives

- Define vestibular migraine.
- Recognize the clinical presentation of vestibular migraine.
- Discuss potential treatment options for vestibular migraine.
- Understand how vestibular migraine affects patients' quality of life.

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Vestibular migraine is a migraine disorder that causes episodes of vestibular symptoms not always temporally related to the migraine headache. The condition can be debilitating, but because it is not as widely recognized as other forms of migraine and vertigo, diagnosis often is delayed. The underlying pathophysiology of the diagnosis is not well understood and there are no clear consensus guidelines for treatment. This can leave patients and clinicians feeling frustrated and overwhelmed. Clinicians who include vestibular migraine in their differential diagnosis for patients with migraine and/or vertigo can help these patients get early appropriate treatment and experience improved quality of life.

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Many other terms have been used to describe vestibular migraine, including migrainous vertigo, migraine-associated vertigo/dizziness, vertiginous migraine, migraine-associated vestibulopathy, and benign recurrent vertigo.[1-3] In the 1980s, neurologists and otolaryngologists published findings about patients with migraine and vestibular symptoms that responded to medications typically used for migraine prophylaxis.[4] The term *vestibular migraine* along with its clinical diagnostic criteria were finally proposed in 2004.[1] However, a clear definition of vestibular migraine was not formally developed until 2012, when a consensus statement by the International Headache Society, the Bárány Society, ear-nose-throat (ENT) physicians, and other physicians was published.[3] It was first listed in the beta version of the International Classification of Headache Disorders, 3rd ed. (ICHD-3 beta) in 2013.[5] The development of diagnostic criteria for vestibular migraine was delayed in part because of the complexity and variety of symptoms associated with the disorder.[3] Even with the published diagnostic criteria, the lack of a standard knowledge base regarding vestibular migraine means that the condition continues to be underdiagnosed.[6]

EPIDEMIOLOGY

Using data from the 2008 National Health Interview Survey, Formeister and colleagues calculated the prevalence of vestibular migraine in adults in the United States to be 2.7%.[7] In children, benign paroxysmal vertigo of childhood is an early manifestation of migraine and has a prevalence

Key points

- Vestibular migraine is characterized by episodes of migraine headache and vertigo that do not always coincide.
- No consensus treatment guidelines exist, and vestibular migraines are managed based on other migraine recommendations.
- Prompt recognition and diagnosis by clinicians is essential to improving patient quality of life.

of about 3%.[1,4] Like other migraine subtypes, vestibular migraine is more common in women. The median reported age of onset is mid-30s to 40s.[1] A familial occurrence has been reported in some patients and an autosomal dominant pattern of inheritance with a decreased penetrance in men has been described.[1,2]

PATHOPHYSIOLOGY

The exact pathophysiology of vestibular migraine is not known. Many mechanisms have been proposed but further study in this area is necessary. Vestibular migraine is a migraine variant, so hypotheses for migraine pathophysiology generally are accepted to apply, in addition to a multisystem overlap of central vestibular pathways.[4] Cortical spreading depression is thought to cause migraine with aura and is considered to be part of the cause of vestibular migraine. When the cortical spreading depression reaches the vestibular areas of the cortex, it can cause vestibular symptoms in a brief attack. However, this does not explain all of the clinical symptoms of vestibular migraine. The reciprocal connections between brainstem vestibular nuclei and the structures that modulate trigeminal nociceptive inputs are thought to underlie the pathogenesis of vestibular migraine.[2] Patients with a history of migraine have a low threshold activation of the connections between the trigeminal and vestibular systems.[8] Current research suggests that a change in ion channel function results in altered neural activity in the trigeminovascular system, which results in release of neurotransmitters such as substance P and calcitonin gene-receptor peptide (CGRP).[9] Receptors for CGRP are expressed in the vestibular system and have been associated with motion sickness.[9] Activation of this system likely is a contributing factor for the pathophysiology of vestibular migraine. Further research is still needed in this area to determine the precise cause of migraine and vestibular migraine.

CLINICAL FEATURES

A detailed history is essential for diagnosing vestibular migraine. Diagnosis is based on the patient's reported symptoms and the exclusion of other causes of vertigo. Patients often have difficulty describing their symptoms and clinicians should try to determine the character of the vertigo or dizziness, headaches, as well as associated symptoms. Clinicians also need to ask patients about the duration of symptoms and their association with each other temporally.[4] Vestibular migraine has the same triggers as common migraine, including menstruation, insufficient sleep, stress, specific foods, and sensory stimuli.[3,8] Most patients experience migraine headache first, with a delay of years before the onset of vestibular symptoms.[1] Patients with vestibular migraine are more susceptible to motion sickness.[8] As patients age, migraine can become less frequent and independent vertigo, dizziness, or disequilibrium can become more common.[2] This is seen especially in postmenopausal women.[2] Auditory symptoms including hearing loss, tinnitus, and ear fullness occur in 38% of patients.[2] More than half of patients with vestibular migraine have comorbid psychiatric disorders, most commonly depression and anxiety.[10]

The terms *dizziness* and *vertigo* often are used synonymously but are different. Dizziness is a sensation of disturbed spatial awareness; vertigo is a false sensation of self-motion or a false sensation that the visual surrounding is spinning or flowing.[4] Complicating the diagnosis further, 30% to 50% of patients with migraine experience dizziness or vertigo without having vestibular migraine.[6] The dizziness or vertigo component of vestibular migraine can take on various forms in the same patient and can be hard for patients to describe.[3] Patients can even experience vertigo transformations from one form to another during episodes.[3,8] The most frequent vertigo symptoms in vestibular migraine are spontaneous rotatory vertigo and positional vertigo but patients also may experience visually induced vertigo and head motion-induced vertigo.[3,9] Patients are most likely to describe a sense of spinning, floating, rocking, tilting, swaying, feeling off-balance, lightheaded, or foggy.[1] Vestibular symptoms are often triggered or aggravated by changing position, movement, or visual motion within the patient's environment.[1]

Table 1 outlines the diagnostic criteria for vestibular migraine and probable vestibular migraine. The criteria show the importance of recognizing that the timing of vestibular symptoms and migraine headaches is quite variable; they do not always overlap. In fact, some patients never experience vertigo and headache at the same time.[3,8] To fulfill diagnostic criteria, patients must have migraine features including headache, phonophobia, photophobia, or visual aura in at least half of the episodes.[4] In about 30% of vestibular migraine attacks, patients experience vertigo without a headache; only 25% of patients experience dizziness or vertigo with every headache episode.[2] Patients may experience different combinations of symptoms during each episode. The associated symptoms may occur before, during, or after an acute episode. The duration of attacks is seconds for 10% of patients, minutes for 30%, hours for 30%, and days for 30%.[3] Episodes rarely exceed 72 hours but patients can take weeks to fully return to baseline after an episode.[5]

DIAGNOSIS

Primary care providers should perform a basic neurologic examination, including cranial nerve and extraocular eye movement assessment, when evaluating a patient with vertigo. Depending on patient presentation, special testing may include the Dix-Hallpike maneuver, head-impulse test (HIT), and cerebellar testing. In most patients, the basic primary care physical examination will be normal between attacks.[8] Some patients will have signs of central vestibular dysfunction between attacks, including positional nystagmus and saccadic pursuit. During attacks, patients may have central spontaneous or positional nystagmus.[2,8]

No diagnostic tests can confirm vestibular migraine; instead, tests serve to rule out other potential causes of the symptoms. Common testing to evaluate vertigo includes MRI of the brain and internal auditory canal, CT of the temporal bone, videonystagmogram, vestibular-evoked myogenic potentials, video head impulse testing, rotatory chair testing, and audiogram. The specific tests done are determined by patient presentation and the differential diagnosis.

TABLE 1. Vestibular migraine diagnostic criteria[5,17]

Vestibular migraine

A. At least five episodes fulfilling criteria C and D
B. A current or past history of migraine without aura or migraine with aura
C. Vestibular symptoms of moderate or severe intensity, lasting 5 minutes to 72 hours
D. At least 50% of episodes are associated with at least one of the following three migrainous features:
   a. Headache with at least two of these four characteristics: unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity
   b. Photophobia and phonophobia
   c. Visual aura
E. Not better accounted for by another ICHD-3 diagnosis or by another vestibular disorder

Probable vestibular migraine

A. At least five episodes with vestibular symptoms of moderate to severe intensity, lasting 5 minutes to 72 hours
B. Only one of the criteria B or D for vestibular migraine is fulfilled (migraine history or migraine features during the episode)
C. Not better accounted for by another vestibular or ICHD diagnosis

QUALITY OF LIFE

Neuhauser and colleagues found that about two-thirds of patients with vestibular migraine visited a physician because of their symptoms but only 20% of these patients were diagnosed with vestibular migraine at the time of their visit.[11] Millen and colleagues found a significant difference in recognition and diagnosis of vestibular migraine between ENT physicians and neurologists, with higher recognition among neurologists.[6] This contributed to patient confusion and affected patient outcomes.[6] The discrepancy shows the importance of all clinicians being aware of vestibular migraine and including it when forming differential diagnoses for patients with vertigo, dizziness, and migraine.

Stovner and colleagues used data from the 2016 Global Burden of Diseases, Injuries, and Risk Factors (GBD) studies to calculate the years of life lived with disability for patients with migraine-type headaches.[12] They determined that globally, 1.04 million people are affected by migraine, leading to 45.1 million years of life lived with disability.[12] The group most heavily affected were women ages 15 to 49 years. Within this group, migraine headaches accounted for 11.2% of all years of life lived with disability.[12] In the study done by Neuhauser and colleagues in Germany, 40% of patients with vestibular migraine reported needing sick leave from work due to their symptoms.[11] This study also found that the age-adjusted health-related quality of life scores were lower in patients with vestibular migraine than in patients without dizziness. This was true for men and women.[11]

The vestibular symptoms associated with vestibular migraine often are more debilitating than the actual headache pain. These symptoms can last for hours or days and lead to severe impairment of daily activities.[1] Acute attacks can force patients to stay in bed for a day or more as they try to avoid even the slightest head movement.[8] Patients with vestibular migraine commonly have poor sleep quality, high levels of depression, and an overall low quality of life.[2] Over time, spatial misperceptions can become chronic and further limit daily activities. Spatial misperceptions are unusual sensitivity to head motion or visual stimuli or sudden feelings of imbalance or tilt. Overall, this illness has a considerable effect on patients.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for vestibular migraine should include other vestibular disorders complicated by superimposed migraine attacks, including Ménière disease, benign paroxysmal positional vertigo, migraine with brainstem aura (previously basilar migraine), vertebrobasilar transient ischemic attack (TIA), psychiatric dizziness syndromes, vestibular paroxysmia, persistent postural-perceptual dizziness, and episodic ataxia type 2.[1,3,5,8] Differentiating vestibular migraine from Ménière disease, benign paroxysmal positional vertigo, and migraine with brainstem aura is most challenging (**Table 2**).

Symptoms of Ménière disease and vestibular migraine overlap significantly. Patients with Ménière disease can have migraines and patients with vestibular migraine can experience cochlear symptoms.[2,5] Both diseases can cause aural fullness, tinnitus, low-frequency hearing loss, and vertigo. The characteristics of the symptoms can be used to help differentiate the disorders. Vertigo episodes that are very short (seconds to less than 15 minutes) or prolonged (more than 24 hours) are more likely to be due to vestibular migraine.[2] The vertigo attacks in Ménière disease tend to last 20 minutes to 12 hours.[8] Hearing loss in both disor-

TABLE 2. Differential diagnosis of vestibular migraine[2,5,8]

	Vestibular migraine	Ménière disease	Benign paroxysmal positional vertigo	Migraine with brainstem aura
Vertigo episode length	Less than 15 minutes or longer than 24 hours	20 minutes to 12 hours	Several seconds to a few minutes	5-60 minutes
Positional vertigo frequency	Several times per month or year	N/A	Typically not more than a couple of times per year	N/A
Association with headache	Migraine headaches occur	Can have superimposed migraines	Can have superimposed migraines	Migraine headache following aura symptoms
Acoustic symptoms	Mild, low-frequency, bilateral nonprogressive hearing loss	Low-frequency, unilateral progressive hearing loss; tinnitus	None	Aura can cause hypacusis and tinnitus

TABLE 3. Medications for migraine treatment[2,4,9,14,16]

Medication	Routes of administration	Uses	Common adverse reactions
CGRP receptor antagonists (ubrogepant, rimegepant)	tablet, orally disintegrating tablet	Acute migraine	Nausea, sedation
Triptans (most commonly zolmitriptan or sumatriptan)	tablet, nasal spray, orally disintegrating tablet, injectable	Acute migraine	Sedation, dry mouth, nausea, dizziness, diarrhea
Antihistamines (diphenhydramine, dimenhydrinate, meclizine)	oral, IV, rectal	Antiemetic; nausea due to motion sickness	Sedation, constipation, blurred vision, dry mouth
Antidopaminergic (metoclopramide, prochlorperazine, promethazine)	IV, IM, oral	Nausea and vomiting associated with migraine	Extrapyramidal effects, hypotension, QT prolongation, sedation
Benzodiazepines	IM, IV, oral, sublingual, intranasal, rectal	Vestibular suppressant	Sedation, light-headedness, unsteadiness, dizziness
Methylprednisolone	IV high dose	Continuous severe episode, intractable headache	Nausea, headache, dizziness, insomnia, depression, anxiety

ders is typically low frequency but is bilateral in vestibular migraine and typically unilateral in Ménière disease.[2,8]

Differentiating benign paroxysmal positional vertigo from vestibular migraine also is challenging because both cause positional vertigo. Having the patient perform diagnostic provocation maneuvers during an acute episode may be the only way to differentiate the two disorders clinically.[2] In patients with vestibular migraine, positional vertigo typically lasts from a few minutes to several days and occurs several times per month or year. In patients with benign paroxysmal positional vertigo, positional vertigo lasts from several seconds to a few minutes and occurs less frequently, typically occurring only a couple of times per year.[2,8]

Migraine with brainstem aura is a rare form of migraine that meets criteria of migraine with aura and also causes at least two fully reversible brainstem symptoms, including dysarthria, vertigo, tinnitus, hypacusis, diplopia, ataxia, and decreased level of consciousness. The aura symptoms typically last 5 to 60 minutes.[5] Unlike migraine with brainstem aura, vestibular migraine does not typically cause other symptoms related to brainstem involvement.[1]

TREATMENT

No specific treatment guidelines exist for vestibular migraine. Although several retrospective clinical trials have been performed, only a few randomized controlled trials have sought to identify effective treatment regimens.[9] Clinicians rely on treatment recommendations for migraine headaches, anecdotal experience, expert opinion, and observational studies.[4,8] Studies evaluating prophylactic treatment combined with lifestyle changes showed patient symptom improvement.[4] The goal of treatment is to reduce symptom frequency and severity.[1]

Nonpharmacologic lifestyle changes include avoiding known triggers, eating regularly, getting adequate sleep, and exercising.[2,4,8] Regular exercise seems to reduce intensity and frequency of vestibular migraine symptoms.[8] Stress is a common trigger for vestibular migraine and finding ways to reduce stress can help prevent attacks.[4] Patients also may benefit from vestibular physical therapy or rehabilitation, especially if they experience chronic dizziness and imbalance.[2,4,8] Vestibular rehabilitation has been shown to improve vestibular migraine patient outcomes and is underused by clinicians.[13] Referral to a neurologist or

TABLE 4. Prophylactic medications for migraine[2-4,9,14,16]

Medication	Dosage	Uses	Common adverse reactions
CGRP antagonists (erenumab, galcanezumab, fremanezumab, eptinezumab)	Variable	Migraine prevention	Nausea, sedation
CGRP receptor antagonist (rimegepant)	75 mg every other day	Migraine prevention	Nausea
Topiramate	50-100 mg/day	Frequent migraine attacks (more than 15 per month); obesity	Paresthesia, somnolence, weight loss, cognitive dysfunction
Acetazolamide	250-750 mg/day	Patients with visual aura	Paresthesia, nausea, sedation, hypokalemia
Lamotrigine	25-100 mg/day	Predominant vertigo or dizziness without frequent migraine	Balance disturbance, double or blurred vision, difficulty concentrating, headache, sedation
Amitriptyline	50-100 mg/day	Frequent migraine attacks (more than 15 per month); sleep disturbance	Sedation, orthostatic hypotension, dry mouth, weight gain, constipation, urinary retention, conduction block
Nortriptyline	27-75 mg/day	Frequent migraine attacks more than 15 per month); sleep disturbance	Sedation, orthostatic hypotension, dry mouth, weight gain, constipation, urinary retention, conduction block
Metoprolol	50-200 mg/day	Long lasting or disabling attacks; hypertension	Fatigue, hypotension, erectile dysfunction, depression, bronchial constriction
Propranolol	40-240 mg/day	Long lasting or disabling attacks; hypertension	Fatigue, hypotension, erectile dysfunction, depression, bronchial constriction
Valproic acid	600-900 mg/day	Long lasting or disabling attacks	Weight gain, sedation, fetal malformation
SSRIs, SNRIs	Variable	Comorbid anxiety/ depression	Headache, nausea, sleep disturbance, dizziness, sexual dysfunction, dry mouth
OnabotulinumtoxinA	155 units every 3 months	Migraine prevention	Redness/soreness/swelling at injection site, bruising, chills, fatigue, dry mouth, neck stiffness

headache clinic is appropriate; this disorder can be very complicated to manage.

A newer class of migraine prevention and treatment medication, calcitonin gene-related peptide (CGRP) antagonists and CGRP receptor antagonists, is being used in patients with insufficient response to triptan therapy and in those who cannot use triptans.[14] A small retrospective study by Hoskin and Fife demonstrated a trend toward improvement in vestibular migraine patients treated with some of these medications.[9] Further study is needed to determine their full effectiveness for treatment and prevention of vestibular migraine.

A small study by Beh showed improvement in patients' vertigo and migraine severity following treatment with an external trigeminal nerve stimulator.[15] This represents another potential treatment for acute vestibular migraine that needs further study.

Abortive therapy consists of antivertiginous and antiemetic medications.[13] Triptans such as zolmitriptan and sumatriptan are used as abortive medications for migraine headache (**Table 3**).[4] Using a triptan that is available in nasal or injectable form can be helpful if the patient experiences nausea or vomiting associated with the episode. Benzodiazepines also can be used for patients with prolonged acute symptoms of vertigo and nausea.[8] Antiemetics and antivertiginous medications can be used to suppress vestibular symptoms. Promethazine combines antivertiginous, antiemetic, and sedating properties. Metoclopramide helps control nausea and vomiting associated with headache and vertigo. Metoclopramide also increases gastric motility and thus response to oral antimigraine medications.[16] Its effectiveness in migraine-associated nausea has been well established.[10] Antihistamines can be used to treat milder episodes of vertigo and for nausea associated with motion sickness. They do not have strong antivertiginous action.[10]

Patients who have frequent and/or severe attacks should be started on preventive medication (**Table 4**). These medications typically are chosen based on the patient's comorbid conditions and expected adverse reactions. In general, dosing should be started low and then gradually increased. Patients should keep a diary of their symptoms and their response to treatment should be evaluated after 2 to 3 months.[8] Prophylactic medications used are essentially the

same as those used for other migraines and include CGRP antagonists and CGRP receptor antagonists, topiramate, acetazolamide, lamotrigine, amitriptyline, nortriptyline, metoprolol, propranolol, valproic acid, and onabotulinintoxinA.[1,3,4,8,13] For patients with comorbid psychiatric conditions, especially depression and anxiety, selective serotonin reuptake inhibitors (SSRIs) commonly are used.[1] Effective preventive treatment reduces attack frequency by 50%.[8] If this reduction is not seen at reassessment, consider stopping the medication and trying a different one.[10] A retrospective study by Power and colleagues found that only a minority of patients responded adequately to the first agent chosen.[13]

CONCLUSION

Vestibular migraine is underrecognized and underdiagnosed by clinicians. This can lead to expensive and uncomfortable testing for patients, inaccurate diagnoses, and inappropriate and ineffective treatments. The symptoms of vestibular migraine attacks are debilitating and lead to significant loss of productive time at work and home for patients. Over time, this disorder can take a heavy psychologic toll. Patients may need to try several different medications or medication combinations before finding an effective treatment regimen that they tolerate. Patients may miss work and family obligations and have difficulty with activities of daily living because of symptoms and acute attacks. Those with this disorder often have concurrent anxiety and/or depression, which can further affect their activities of daily living. Clinicians who understand vestibular migraine can help patients be accurately diagnosed and effectively managed. **JAAPA**

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