Dizziness in the Outpatient Care Setting

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ABSTRACT

Purpose of Review: This article summarizes an approach to evaluating dizziness for the general neurologist and reviews common and important causes of dizziness and vertigo.

Recent Findings: Improved methods of diagnosing patients with vertigo and dizziness have been evolving, including additional diagnostic criteria and characterization of some common conditions that cause dizziness (eg, vestibular migraine, benign paroxysmal positional vertigo, chronic subjective dizziness). Other uncommon causes of dizziness (eg, superior canal dehiscence syndrome, episodic ataxia type 2) have also been better clarified. Distinguishing between central and peripheral causes of vertigo can be accomplished reliably through history and examination, but imaging techniques have further added to accuracy. What has not changed is the necessity of obtaining a basic history of the patient’s symptoms to narrow the list of possible causes.

Summary: Dizziness and vertigo are extremely common symptoms that also affect function at home and at work. Improvements in the diagnosis and management of the syndromes that cause dizziness and vertigo will enhance patient care and cost efficiencies in a health care system with limited resources. Clinicians who evaluate patients with dizziness will serve their patient population well by continuing to manage patients with well-focused workup and attentive care.

INTRODUCTION

Dizziness accounts for 5% of outpatient clinic presentations, making it a very common symptom in clinical medicine.1 Meanwhile, dizziness may be one of the most challenging symptoms to which physicians can ascribe a mechanism and cause.2

Dizziness is a term used commonly by patients and physicians alike that broadly refers to a misperception of spatial orientation. Because dizziness is so nonspecific a term, it may refer to vertigo (the illusion of movement, especially rotation), disequilibrium or imbalance, lightheadedness, or near faintness.

Unsteadiness or disequilibrium indicate a feeling of reduced balance when standing or walking, but without vertigo, lightheadedness, an illusion of movement, or a feeling of spatial misperception. That is, the patient senses that balance is impaired because of poorer-than-normal stability on his or her feet or a feeling that he or she might fall. This sensation may result from dizziness but may be caused by many other conditions, ranging from orthopedic to neurodegenerative disorders, which will not be discussed in this article.

APPROACH TO PATIENTS WITH DIZZINESS

The effective history and directed examination can determine the source of dizziness in many cases. The history should identify what is meant by “dizziness,” whether it is constant or intermittent, provoked or random, and how long it lasts if intermittent. The
history should also identify whether dizziness varies in certain circumstances and if other symptoms accompany the dizziness that might be clues to its nature and mechanism.

Clarifying “Dizziness”

Dizziness may be caused by both benign and dangerous conditions, so a careful history is important to improve the diagnostic accuracy. When the patient and clinician can come to a clear and mutual understanding of the patient’s description of dizziness, determining a clear cause is more straightforward. However, substantial limitations exist regarding how much the patient’s report of the quality of the sensation can be used to accurately determine the mechanism, particularly in the acute care setting. Whether the patient states that he or she is spinning or that the room is spinning is of little importance. If the patient can identify clear rotational sensations, the condition qualifies as vertigo, which implies a greater likelihood of a vestibular mechanism. Other descriptions are less helpful in narrowing the broad differential diagnosis.

The quality-of-symptom approach to characterizing dizziness leads to many errors in diagnosis. In one study by Hanley and O’Dowd, when patients at an ambulatory clinic were given options to describe their dizziness, 7.1% were unable to classify their symptoms at all, and many others chose multiple words simultaneously to describe their sensation. While getting the patient to elaborate on the quality of the sensation is helpful, more is needed to make a diagnosis (Case 2-1).

Narrowing the Differential Diagnosis

What other symptoms or signs can aid in narrowing the possible mechanisms and causes of the patient who experiences dizziness? As outlined in Table 2-1, the patient’s descriptions of the timing, triggering events, and associated symptoms are generally more reliable than their descriptions of the quality of the dizziness.

Case 2-1

A 46-year-old woman presented with a 4-month history of dizziness that she described as a constant feeling of instability; she sometimes felt as if she were spinning and at times felt that she could faint. Her symptoms had begun suddenly when at an out-of-town meeting while sitting at a table. She had begun to feel unstable and later became nauseated; she recalled a “topsy-turvy” spinninglike sensation that lasted the whole evening. She was alone and stayed in the hotel room, and eventually the spinning gave way to a constant swaying and floating feeling, which she continued to experience daily. She had not experienced any hearing loss, slurred speech, double vision, weakness, or numbness, and the nausea had not occurred since the initial onset of symptoms. Brain MRI was found to be normal on two occasions, and magnetic resonance angiography (MRA) of the head and neck, cardiac workup, and multiple blood tests were normal. At work, she had used all her vacation time and leave allowed by the Family and Medical Leave Act and was worried that she could no longer do her job as an information technology project manager. She was also reluctant to travel or drive. No abnormalities were found on examination.

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Useful Vestibular Examination Techniques

Several examination techniques are helpful in evaluating the patient with vertigo and dizziness. These include recognition of direction-fixed horizontal nystagmus and distinction from horizontal gaze-evoked nystagmus (Table 2-1), head impulse testing, and the Dix-Hallpike maneuver (described in the later section on benign paroxysmal positional vertigo [BPPV]).

The head impulse (or head thrust) test is a simple test for detecting impaired vestibular function related to the horizontal semicircular canal. When the head is turned quickly (1 Hz or more), the only eye movement system that can move so quickly and with such short latency is the vestibuloocular reflex. In accordance with Ewald’s second law, part of a set of principles by which the vestibuloocular reflex works, turning the head rapidly stimulates the vestibular function on the side toward which the head is turned more than it inhibits the other ear. The result is that if there is loss of vestibular function on the side to which the head is turned, the vestibuloocular reflex is not able to drive the eyes in an equal and opposite direction, so the eyes move with the head and then require a catch-up saccade to get back to the target the patient had intended to focus on.

The head impulse test is done by grasping the patient’s head firmly on both sides, explaining the test to the patient, and then making a very rapid but very small-amplitude turn of the head while the patient is instructed to look at the physician’s eye or some other fixed target. If the patient’s eyes never deviate from the target during or after the head impulse, then vestibular function related to the horizontal canal is normal or nearly normal in the ear on the side toward which the head is turned. If, however, the eyes moved with the head and then must take a quick excursion or catch-up saccade back to the target, then the test is abnormal on that side. Head impulse testing remains abnormal for a lifetime once permanent unilateral or bilateral vestibular loss of the horizontal canal has taken place.

Practical performance of the head impulse test takes a little experience, and so it is wise to perform the head impulse test in many patients, including healthy patients. One common mistake is to assume that a patient must make a large turn of the head, risking neck injury. While it would be unwise to do this test in someone with a cervical fracture, the risk of neck injury is negligible in most people. One problem with interpretation can be blinking or the inability of the patient to relax or to avoid moving his or her own head. Nevertheless, the head impulse test is easily evaluated and interpreted in most patients. A bit of gradation exists in how prominent and consistently the catch-up

**KEY POINTS**

- The head impulse test is a useful test of vestibular function that can aid in the diagnosis of vestibular disorders and requires no special equipment.
- The head impulse test is done by grasping the patient’s head firmly on both sides, explaining the test to the patient, and then making a very rapid but very small-amplitude turn of the head while the patient is instructed to look at the physician’s eye or some other fixed target.
- If the patient’s eyes never deviate from the target during or after the head impulse, then vestibular function related to the horizontal canal is normal or nearly normal in the ear on the side toward which the head is turned. If, however, the eyes moved with the head and then must take a quick excursion or catch-up saccade back to the target, then the test is abnormal on that side.

**Comment.** This case exemplifies a situation in which a patient probably experienced a transient peripheral vestibular disturbance that caused spinning vertigo, but later a different form of dizziness became the predominant cause. This case shows that multiple descriptions of dizziness may be given by the patient; in this case, it was clarified that initially the patient experienced true vertigo but later experienced more of a floating and swaying sensation. This could be due to poor compensation of unilateral vestibular loss but, more likely, the patient had a mild case of vestibular neuritis that created alarm and fear, and subsequently, chronic subjective dizziness related to anxiety ensued.
### Table 2-1: History and Examination of the Patient With Dizziness

<table>
<thead>
<tr>
<th>Feature</th>
<th>Response and Implied Mechanism or Cause</th>
</tr>
</thead>
</table>
| Quality of dizziness           | - Vertigo or clear spinning implies vestibular mechanism  
- Near faintness (not floating) as a sole description implies cardiovascular/hemodynamic cause  
- Other descriptions far less clear as to mechanism                                                                                                                                 |
| Timing and duration            | - Brief recurrent spells are seen with benign paroxysmal positional vertigo (BPPV), but do not totally exclude vascular, cardiac, psychological, or other mechanisms  
- Recurrent spells of dizziness lasting 1 to 15 minutes may be seen with vertebrobasilar transient ischemic attacks, vestibular migraine, or sometimes with panic attacks  
- Recurrent dizziness lasting hours may be seen with vestibular migraine or with Ménière disease, the latter more likely if associated unilateral hearing loss or fluctuating hearing occurs  
- Chronic ongoing dizziness nearly continuously for many weeks implies anxiety or migrainous mechanism but does not totally exclude other causes, including slow recovery from vestibular neuritis or brainstem/cerebellar lesions or drug toxicity |
| Triggering circumstances      | - Induced by rolling in bed, certain head positions, or looking up implies BPPV  
- Dizziness worsened by head movement implies vestibular mechanism, whether central or peripheral  
- Dizziness almost exclusively occurring when standing or walking and not when supine or recumbent implies hemodynamic mechanisms but is sometimes reported in those who have simple gait unsteadiness  
- Dizziness triggered by vibration or loud sounds (Tullio phenomenon) may suggest superior canal dehiscence syndrome, a bony abnormality of the labyrinth  
- Dizziness induced predominantly by seeing objects in motion implies visual vertigo (optokinetic motion sickness), a vestibular syndrome seen in several vestibular disorders |
| Other associated symptoms      | - Unilateral hearing loss with spinning vertigo implies labyrinthine cause  
- Diplopia, dysarthria, or focal weakness or numbness with vertigo is not peripheral vestibular and implies central nervous system (CNS) vestibular mechanism  
- Nausea is characteristic of vestibular causes (both central and peripheral) but may sometimes also occur with hemodynamic mechanisms  
- Autophony is an unusual symptom in which the patient reports hearing or feeling his or her own voice, breathing, eye movements, or footsteps |
| Dix-Hallpike maneuver          | - Paroxysmal upbeating and torsional nystagmus after the Dix-Hallpike maneuver is characteristic of BPPV of the posterior canal and thus is highly diagnostic |
| Nystagmus                      | - Spontaneous direction-fixed<sup>a</sup> nystagmus strongly supportive of a peripheral vestibular mechanism, particularly if it adheres to the Alexander law<sup>b</sup>  
- Spontaneous vertical or horizontal gaze-evoked nystagmus indicates a CNS process likely affecting the brainstem or cerebellum |

<sup>a</sup> Direction-fixed nystagmus means the nystagmus continues to beat in the same direction regardless of the direction of gaze. This distinguishes it from gaze-evoked nystagmus, where the nystagmus beats to the right in right gaze and to the left in left gaze, which indicates a CNS mechanism.

<sup>b</sup> The Alexander law states that nystagmus is stronger with gaze in the direction of the fast phase and diminishes or abates with gaze in the direction opposite of the fast phase. The Alexander law is characteristic of peripheral vestibular disorders<sup>6</sup> but may occasionally be seen where a vascular mechanism has caused peripheral vestibular ischemia as with the ischemia of the lateral pons affecting the vestibular nerve entry zone or the labyrinth itself (eg, the anterior inferior cerebellar artery syndrome).
saccade is, with milder vestibular loss being harder to detect than severe loss. When in doubt, the test can be repeated until the examiner feels comfortable that it is either normal or not.

**PRIMARY LABYRINTHINE CAUSES OF VERTIGO AND DIZZINESS**

This section describes causes of dizziness related to inner ear and peripheral vestibular mechanisms. Most commonly, the character of the dizziness in these disorders is spinning vertigo, but as described in the prior section, the clinician should not over rely on the quality of dizziness but also look at patterns that include timing, triggering events, associated features, and the examination for nystagmus to improve diagnostic accuracy.

**Benign Paroxysmal Positional Vertigo**

BPPV is the most common disorder causing recurrent vertigo. The lifetime prevalence of BPPV is 2.4%, and the 1-year prevalence is about 1.6%. Most spells of BPPV last a few weeks but may last much longer or become a recurrent condition lasting years. BPPV is also an important health issue in older people since the incidence increases with age and may contribute to falling and its associated morbidity.

BPPV is characterized by episodic vertigo usually lasting from 10 to 30 seconds. The spells of vertigo are evoked by certain tilting positions of the head, rolling over while supine, or straightening up after bending over. The Dix-Hallpike maneuver is a simple bedside examination technique that induces the vertigo and nystagmus of the most common form of BPPV (Figure 2-1). Most cases of BPPV (85%) are related to involvement of the posterior semicircular canal, but the horizontal (15%) or, far less commonly, the anterior semicircular canals (less than 1%) may be affected. The directional features of the nystagmus help to identify the side and semicircular canal affected (Table 2-2).

**Mechanism.** The semicircular canals of the membranous labyrinth of the inner ear on each side detect angular or turning movements of the head (Figure 2-2). Each labyrinth has three semicircular canals: anterior, posterior, and horizontal. The semicircular canals are roughly orthogonal, and this orientation maximizes their ability to detect all angular head acceleration movements. The ampulla of each canal is the motion sensor containing a bendable cupula that responds to any movement of endolymph as occurs during angular head acceleration. Stimulation (or inhibition) of the structures creates a volley of neural activity in the ampullary nerve that is carried via the vestibular nerves to the brain.

Each labyrinth also has a saccule and a utricle that are sometimes referred to as the otolith organs (because they contain otoconia), and these structures contain the sensory epithelium known as the macula. Their function is to detect linear or translational acceleration (Figure 2-2), including detection of forces such as gravity or accelerating or stopping in a car.

BPPV is caused when the calcium carbonate otoconia erode or parts of them become dislodged. This debris may then inappropriately end up within the lumen of one of the semicircular canals, where it inappropriately stimulates the ampulla, causing transient positional vertigo and nystagmus. The calcium carbonate crystals are more than twice as dense (specific gravity 2.7 g/cm³) as endolymph (specific gravity 1.0 g/cm³), so they sink or move in the fluid within the semicircular canal in response to gravity.

The triggering event and the direction of nystagmus will vary depending on the canal where the calcium carbonate...
material is located. That is, each canal has its own pattern of paroxysmal positional nystagmus. The posterior canal type is upbeating and torsional, with the top pole beating toward the downward ear. The nystagmus of horizontal (lateral) canal BPPV is horizontal and changes direction as the head position is changed (paroxysmal direction-changing positional nystagmus). For example, when the patient is supine with the head turned to the right, there is a burst of right-beating nystagmus, and, conversely, when the head is then turned to the left, the nystagmus changes to a burst of left-beating nystagmus. The rarest form is the anterior canal type, which is downbeating, sometimes with a minor torsional component (Table 2-2). In some instances, more than one canal may be simultaneously affected. The most common form is simultaneously bilateral posterior canal type, and the second most common is simultaneous involvement of the posterior and horizontal canals on the same side.

**Diagnosis.** BPPV is diagnosed by the presence of recurrent attacks of

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**KEY POINT**

- Benign paroxysmal positional vertigo is diagnosed by the presence of recurrent attacks of positional dizziness lasting less than 1 minute that are provoked by lying down or turning while supine and are associated with characteristic nystagmus.

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**FIGURE 2-1** Dix-Hallpike maneuver. The Dix-Hallpike maneuver for detection of right posterior canal benign paroxysmal positional vertigo (A) and for the left posterior canal (B). The patient is taken from the sitting position (1) to the head hanging on the respective side being tested (2).

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positional dizziness lasting less than 1 minute that are provoked by lying down or turning while supine and are associated with characteristic nystagmus (Table 2-2).\(^9\) The history is generally adequate to make a presumptive diagnosis of BPPV, but observing characteristic nystagmus upon examination confirms the diagnosis (Case 2-2). Occasionally, patients with strong symptoms or who are especially prone to motion sickness report mild nausea and floating dizziness for several hours after the positional vertigo. However, most patients are well between episodes. Vertigo that continues for more than 2 minutes or that does not seem consistently evoked by position changes should lead the clinician to consider alternative causes.

The Dix-Hallpike maneuver is used to diagnose the posterior canal type of BPPV (Figure 2-1). The maneuver is performed by moving the head from an upright position to a head-hanging position while the head remains turned 45 degrees to the side being assessed. Performing the Dix-Hallpike maneuver to the side affected by posterior canal BPPV results in a burst of torsional and upbeat nystagmus that ensues after a short latency, which is usually approximately 2 to 15 seconds. The latency can occasionally be long enough that the nystagmus can be missed if the patient is not observed in the head-hanging position for long enough. The direction of the nystagmus as well as the side of positioning help identify the affected canal. For example, with BPPV affecting the left posterior semicircular canal, the nystagmus seen with the Dix-Hallpike maneuver to the left is clockwise torsional admixed with upbeat nystagmus. Meanwhile, with BPPV affecting the right posterior semicircular canal, the nystagmus with Dix-Hallpike positioning to the right is counterclockwise torsional and upbeat nystagmus occurs.

### TABLE 2-2 Directional Features of Variants of Benign Paroxysmal Positional Vertigo by Canal Type

<table>
<thead>
<tr>
<th>Type of Benign Paroxysmal Positional Vertigo (BPPV)</th>
<th>Direction of Paroxysmal Nystagmus(^a) (Fast Phase Direction)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right posterior canal BPPV</td>
<td>Upbeating, counterclockwise torsional</td>
</tr>
<tr>
<td>Left posterior canal BPPV</td>
<td>Upbeating, clockwise torsional</td>
</tr>
<tr>
<td>Right horizontal canal BPPV(^b)</td>
<td>Geotropic direction-changing horizontal nystagmus</td>
</tr>
<tr>
<td></td>
<td>Apogeotropic direction-changing horizontal nystagmus</td>
</tr>
<tr>
<td>Left horizontal canal BPPV(^b)</td>
<td>Geotropic direction-changing horizontal nystagmus</td>
</tr>
<tr>
<td></td>
<td>Apogeotropic direction-changing horizontal nystagmus</td>
</tr>
<tr>
<td>Right anterior canal BPPV</td>
<td>Downbeating, slight torsional component</td>
</tr>
<tr>
<td>Left anterior canal BPPV</td>
<td>Downbeating, slight torsional component</td>
</tr>
</tbody>
</table>

\(^a\) The determination of clockwise or counterclockwise is made from the examiner’s viewpoint. The horizontal or vertical direction is determined as from the patient’s viewpoint (eg, right beating refers to nystagmus with fast phase toward the patient’s right side).

\(^b\) Determination of the side affected in horizontal canal BPPV is based in part on the side that produces the most intense nystagmus.\(^7\)
torsional and upbeating. By convention, the direction of the nystagmus is defined by its fast phase.

The horizontal canal form of BPPV occurs when the calcium material moves within the horizontal semicircular canal. During performance of the Dix-Hallpike maneuver, if the head is turned to the side enough, it is possible to elicit the horizontal direction-changing nystagmus of the horizontal form of BPPV. Nevertheless, the best

Case 2-2
A 67-year-old man came to the office for a neurologic consultation for vertigo that had begun 6 months prior to presentation. He recalled having a feeling of severe spinning that occurred when he would get out of bed but also occurred periodically when working on his car or around the house. The spells lasted less than 1 minute and never came on when he had been completely motionless. The spells initially abated after about 1 month after their onset, and he had been free of the vertigo until about 4 weeks ago, when it suddenly recurred upon trying to get up after working on the plumbing under his kitchen sink. Although he had not fallen, he periodically staggered and experienced intermittent dizziness when active.

Examination was normal, but the Dix-Hallpike maneuver to the right side revealed an upbeating and counterclockwise torsional paroxysmal positional nystagmus that developed after a several-second latency and reproduced his symptoms. A canalith repositioning maneuver was performed, and upon repeating the maneuver, no further nystagmus or symptoms were present.

Comment. This case is a classic example of benign paroxysmal positional vertigo of the posterior canal. Some patients may have an idea what side is more likely to cause the symptoms, but not all patients have deduced that it is triggered by position changes. Treatment is highly effective.
method for inducing the nystagmus of horizontal canal BPPV is to perform a supine head turn test, sometimes referred to as the Pagnini-McClure supine head turn test (Figure 2-3). While the patient is supine, quickly turning the head to the right results in robust right-beating nystagmus and, once abated, quickly turning the head to the left results in robust left-beating horizontal nystagmus. Thus, direction-changing positional nystagmus is revealed, and it is considered geotropic (toward the ground) because the nystagmus beats toward the downward ear and toward the ground. The geotropic form of horizontal canal BPPV is the most common, but a less common form in which the nystagmus is apogeotropic (nystagmus beats away from the ground) may also occur. The latter form of horizontal canal BPPV has been considered suggestive of cupulolithiasis, in which calcium material is adherent to the cupula rather than the more common canalolithiasis, in which the calcium freely moves within the lumen of the semicircular canal.7

The least common form of BPPV involves the anterior semicircular canal. When present, it produces paroxysmal downbeat positional nystagmus with the Dix-Hallpike maneuver. Because this form of BPPV is typically very short-lived, no treatment maneuvers have been established, although maneuvers for posterior canal BPPV are often employed. When identifying downbeat positional nystagmus in a patient, it is important to remember that this may also be seen in central nervous system (CNS) lesions near the cervicomedullary junction. Persisting and consistent downbeat positional nystagmus should raise suspicion for a brainstem or cerebellar process.

**Differential diagnosis.** Of course, it is important to distinguish central

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**KEY POINT**

- Three forms of benign paroxysmal positional vertigo occur (one form for each canal). There is one type of nystagmus for posterior canal benign paroxysmal positional vertigo (the most common type), two forms of positional nystagmus for horizontal canal benign paroxysmal positional vertigo (geotropic and apogeotropic), and one form that is usually very short lived for anterior canal benign paroxysmal positional vertigo. Recognizing these forms of nystagmus will make it easier to recognize an outlier form of positional nystagmus that could be central in origin.

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**FIGURE 2-3** Supine head roll test (Pagnini-McClure maneuver) to determine the presence of horizontal canal benign paroxysmal positional vertigo. The patient’s head is turned rapidly from the straight-supine position (1) to the head right position (2) while observing for nystagmus. The patient is then taken back to the straight-supine position (1), and then from there, the head may be turned quickly to the left (3), again observing for nystagmus.

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Vestibular Neuritis

Vestibular neuritis (also referred to as vestibular neuronitis, epidemic vertigo, vestibular paralys, labyrinthitis, and neurolabyrinthitis) is an acute peripheral vestibular process most commonly due to reactivation of latent herpes simplex virus in the vestibular ganglion. Degeneration of peripheral vestibular nerve fibers and of the neuroepithelium of peripheral receptors has been observed on histopathologic examination.

By one commonly used definition, the term vestibular neuritis causes only unilateral vestibular dysfunction and does not affect hearing. On the other hand, la byrinthitis refers to acute unilateral loss of both hearing and vestibular function usually attributed to a viral infection.

Vestibular neuritis most commonly affects the superior division of the vestibular nerve, less commonly affects both the superior and inferior divisions together, and yet less often affects only the inferior division. The superior vestibular nerve division innervates the anterior and horizontal semicircular canals and the utricle and, if severely affected, results in caloric vestibular loss on the affected side. The inferior vestibular nerve division innervates the saccule and the posterior semicircular canal, and isolated involvement does not result in reduced caloric vestibular responses on the affected side. The selective vulnerability of the superior vestibular nerve division may result in recurrent vertigo.

Prognosis.

The recurrence rate of BPPV after successful treatment at 4 years is about 30%, but most recurrences occur within the first 6 months of symptom onset. Women are more prone to developing BPPV and having recurrences, but some patients are likely more innately susceptible to BPPV and prone to more recurrences.

KEY POINTS

- The canalith repositioning maneuver is intended to move, by the effect of gravity, the calcium carbonate material out of the affected semicircular canal and back in the main vestibule/utricle region from which it originated.
- Two effective treatments exist for posterior canal benign paroxysmal positional vertigo: the canalith repositioning maneuver and the Semont liberatory maneuver. These treatments are safe and effective for the most common (posterior canal) form of benign paroxysmal positional vertigo.
- The recurrence rate of benign paroxysmal positional vertigo after successful treatment at 4 years is about 30%, but most recurrences occur within the first 6 months of symptom onset.
- Vestibular neuritis is most commonly due to reactivation of latent herpes simplex virus in the vestibular ganglion.
- Vestibular neuritis (also referred to as the term vestibular neuritis) causes of vertigo from BPPV. Knowing and recognizing the patterns of nystagmus characteristic of BPPV will aid in recognizing when a type of positional nystagmus “does not fit” with BPPV and, thus, may be of central origin. Central lesions may not necessarily be triggered by positioning on one side only. The nystagmus may persist or be dissociated (ie, both eyes are not moving in the same way) or may be accompanied by slower, irregular eye movements. A general rule is that if the nystagmus is typical of posterior canal BPPV and resolves with treatment, the diagnosis is correct. If the nystagmus is atypical of BPPV (eg, spontaneous, present with gaze testing, and if nystagmus fails to resolve with treatment of BPPV), then central causes should be considered.

Treatment.

The canalith repositioning maneuver is intended to move, by the effect of gravity, the calcium carbonate material out of the affected semicircular canal and back in the main vestibule/utricle region from which it originated. The canalith repositioning maneuver (also referred to as the Epley maneuver) is the most commonly used (Figure 2-4), but a second type—the Semont liberatory maneuver (Figure 2-5)—is also highly effective. If properly done, these treatment methods have nearly the same success rate of about 90% in eliminating symptoms and nystagmus by the same or next day. Generally, antivertiginous medications such as meclizine are unnecessary unless the patient is exceptionally prone to motion sickness. Anxiolytic medications such as benzodiazepines do not detract from the success of treatment and may have a role when patients feel too anxious to proceed with treatment. Rarely, BPPV can be refractory to repositioning maneuvers and requires surgical occlusion of the affected semicircular canal.

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from its longer course in a more narrowed pathway in the temporal bone that makes it susceptible to the effects of swelling and entrapment.

The time course of vestibular neuritis and labyrinthitis are similar to one another. Vertigo usually begins suddenly or may evolve over a period

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**FIGURE 2-4** A stepwise method of performing the canalith repositioning maneuver for right posterior canal benign paroxysmal positional vertigo. Step 1, The patient is positioned so he or she may be taken back en mass to avoid neck strain or forced hyperextension to the head-hanging position with the neck in slight extension. This is essentially the Dix-Hallpike maneuver. Step 2, Observe the eyes, holding them open if necessary. Wait for all the nystagmus to stop and then continue to wait about 15 additional seconds. Step 3, Keeping the head back with the neck slightly hyperextended, turn the head toward the opposite side and wait 20 seconds. Step 4, Roll the patient all the way in such a way to allow the head to be turned to face the ground and wait 20 seconds. Step 5, From this side-lying face-to-the-ground position, have the patient sit up. Hold the patient to stabilize him or her for 15 seconds or so. The procedure may be repeated once or twice.

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of 30 minutes to a few hours or, less commonly, in a stuttering course over several weeks. Once patients have been having vertigo for 20 to 30 minutes, most people experience nausea and vomiting, although this depends also on the severity of the vestibular asymmetry. Vertigo is present in all head positions but is aggravated by head motion of any kind and even by seeing things in motion; it gradually abates in the days and weeks that follow.

Examination can be very helpful and usually confirms the diagnosis and reveals the side affected. Acutely, and if significant vestibular asymmetry is present, spontaneous horizontal nystagmus may be seen that has a fast phase beating away from the affected ear. Importantly, the direction of nystagmus does not change with changing the direction of gaze. The nystagmus fast phase increases during gaze in the direction of the fast phase and diminishes or abates entirely with gaze away from the fast phase of nystagmus. This is referred to as the Alexander law and is characteristic of acute peripheral vestibular loss, as mentioned in a previous section. The nystagmus of acute unilateral vestibular loss decreases within the first 12 to 36 hours in most cases, although in some patients this may last as long as 1 or 2 months. Generally, however, the spontaneous nystagmus subsides within 1 to 2 days after the onset, and then the nystagmus can be seen only with gaze in the direction of the fast phase. Eventually, the nystagmus is no longer visible or elicitable as the patient compensates. At this point, the clinician needs to rely on caloric

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**KEY POINT**

- Gaze-evoked nystagmus is nystagmus that changes direction with alterations in the direction of gaze and is a central nervous system finding.

- Direction-fixed nystagmus is nystagmus that beats in the same direction regardless of the direction of gaze and is more typical of peripheral vestibular loss, particularly if it intensifies with gaze in the direction of the fast phase of nystagmus (ie, it follows the Alexander law).

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**FIGURE 2-5**

Steps in performing the Semont liberatory maneuver to treat right posterior canal benign paroxysmal positional vertigo. Step 1, Start with the patient sitting on a table or flat surface with the head turned away from the affected side. Step 2, Quickly put the patient into the side-lying position toward the affected side, with the head turned up. Nystagmus will occur shortly after arriving at the side-lying position. Keep the patient here until at least 20 seconds after all nystagmus has ceased. Step 3, Rapidly move the patient back up and through the sitting position without stopping and continue moving the patient to the opposite side-lying position with the head facing down (head should not turn relative to the shoulders during the position change). Keep the patient in this position for about 30 seconds (some recommend up to 10 minutes). Step 4 (not depicted), From the position shown as Step 3, have the patient sit back up to the sitting position.

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testing or the head impulse test to confirm unilateral vestibular loss. Recall that horizontal nystagmus in vestibular neuritis results from hypofunction of the horizontal semicircular canal. However, occasionally vestibular neuritis affects only the inferior vestibular nerve, which does not innervate the horizontal canal; therefore, there may be dizziness but no nystagmus.

The differential diagnosis of acute vestibular neuritis is limited but important. The most concerning issue is an isolated infarct of the posterior circulation.\(^\text{18}\) These syndromes are discussed later in this article.

While small isolated cerebellar infarcts can mimic the history of vestibular neuritis, the examination\(^\text{19}\) can separate these clinical entities fairly well, obviating the routine need for CT angiography or magnetic resonance angiography (MRA) for clinicians who learn how to carefully examine this patient population. The first attack of Ménière disease might easily be mistaken for vestibular neuritis, particularly in the absence of unilateral loud tinnitus or mention of hearing loss. Attacks of Ménière disease, however, usually subside within 8 hours, whereas residual symptoms usually continue for days to weeks in vestibular neuritis. If the patient reports a prior history of a similar attack, Ménière disease might be more highly suspected since recurrences of vestibular neuritis are low, at about a 2% over a lifetime.\(^\text{20}\)

In most cerebellar or brainstem strokes, other neurologic signs occur, such as unilateral dysmetria, slurred speech, hemibody numbness, diplopia, or nystagmus that changes direction when changing the direction of gaze (gaze-evoked nystagmus). The head impulse test is typically abnormal in peripheral causes and not abnormal in most central disorders. Nystagmus of peripheral origin follows the Alexander law, as previously mentioned, and remains unidirectional (Case 2-3). Meanwhile, central nystagmus may

**Case 2-3**

A 76-year-old man presented to the emergency department because of acute vertigo that began 3 hours earlier. He had not been recently ill and recalled no similar vertigo in the past. On evaluation, the patient sat quietly on a gurney with an empty emesis basin by his side. He denied hearing loss, slurred speech, numbness, or double vision but reported feeling intense spinning and nausea. Examination revealed spontaneous right-beating nystagmus that appeared more intense when he looked to the right and that lessened in severity but continued to beat toward the right when he looked to the left. Head impulse testing appeared abnormal with quick turns to the left, but this testing made him feel more nauseated. Finger-nose coordination was normal, and his speech articulation was normal. No diplopia or evident ocular malalignment was seen on examination. He could walk a few steps but was cautious and had to hold on for balance; after reassuming his place on the gurney, he began to vomit.

**Comment.** This case shows many of the characteristic features of acute left peripheral vestibular loss as would be seen with acute left vestibular neuritis. The nystagmus beats away from the hypofunctioning ear and remains fixed in terms of its direction, always beating to the right. Furthermore, the nystagmus follows the Alexander law, being more intense with gaze in the direction of the fast phase and less intense or gone with

Continued on page 372
change direction with changes in the direction of gaze or may be purely vertical or cyclotorsional.

Diagnostic vestibular testing using videonystagmography with caloric testing can identify unilateral peripheral vestibular loss, which can be a clue to the affected side long after vestibular loss has occurred. When more than 24% asymmetry in caloric vestibular nystagmus occurs, pathologic loss of vestibular function may be present that is related to the horizontal semicircular canal function. Videonystagmography need not be carried out in the first days of the vertigo, as unilateral vestibular loss remains detectable for the rest of the patient’s life if permanent vestibular loss is present. As mentioned previously, the head impulse test is another method for determining unilateral vestibular loss of the horizontal canal.

Vestibular neuritis is not associated with slurred speech, diplopia, headache, paresthesia, focal weakness, or limb ataxia, so the presence of these symptoms should prompt further investigation. When vestibular neuritis is due to herpes zoster oticus (Ramsay Hunt syndrome), pain in or around the affected ear may occur, as described in the following section.

Treatment of vestibular neuritis in the first 1 or 2 days can be simply supportive and incorporate use of vestibular suppressants as needed for nausea (Table 2-3). These medications should be discontinued within days to a week as they may delay or limit CNS adaptation to the acute vestibular loss.

Acute use of prednisone 60 mg/d for 1 week may help reduce the severity of vestibular neuritis if given in the first few days, but insufficient evidence exists for greater functional improvement based on currently published data.

Herpes zoster oticus is due to reactivation of latent varicella-zoster virus in the geniculate ganglion and may cause hearing loss or ipsilateral peripheral facial paresis. If varicella zoster is suspected, the patient should be treated with corticosteroids and either acyclovir, famciclovir, or valacyclovir in high oral doses. With herpes zoster oticus, the clinician should look carefully for painful vesicles on the posterior auricle of the external ear or within the back of the external ear canal. If the patient has significant localized ear pain and the tympanic membrane appears normal, it is probably prudent to treat with one of the antiviral agents even in the absence of visible vesicles.

Once nausea has subsided sufficiently to allow it, the patient should
begin moving his or her head from side to side, extending the activity each day until the patient can undergo more extensive vestibular exercises. Table 2-4 gives an example of some general exercises for patients to promote CNS adaptation, but it is probably better to have a therapist knowledgeable of techniques of vestibular rehabilitation to tailor an exercise program to the patient’s needs based on a hands-on assessment. Vestibular rehabilitation promotes CNS adaptation to the unilateral loss and improves functional outcome in patients with unilateral vestibular loss.

**Ménière Disease**

Ménière disease is an inner ear disorder characterized by recurrent, spontaneous attacks of vertigo usually described as spinning, unilateral hearing loss, ear fullness, tinnitus. The prevalence of Ménière disease is about 1 in 150,000 people, affecting men and women about equally. The peak incidence is between 40 and 60 years of age.

The role for the neurologist in this disorder is both to make the correct diagnosis and to exclude any neurologic disorders that might account for the symptoms while also initiating treatment with a low-sodium diet and possibly the addition of a mild diuretic. As long as patients are controlled acceptably, the patients can be followed, but if recurrent attacks of vertigo continue, then referral to an otolaryngologist is indicated.
The history of Ménière disease is one in which at least one attack of vertigo has occurred, usually lasting 20 minutes up to 12 hours (Case 2-4). If the patient is aware of fluctuating hearing or recent unilateral hearing loss and changes in the pitch and loudness of tinnitus, the history is very suspicious for Ménière disease. New diagnostic criteria were recently published for Ménière disease. The criteria importantly add documentation of characteristic low-frequency hearing loss (Figure 2-6), even if it is only transient and resolves on a subsequent audiogram. When patients experience an attack of Ménière disease, they may exhibit this low-frequency hearing loss for up to a few days afterward, and with repeated attacks, eventual permanent hearing loss occurs.
in ensuing months and years that may affect all the frequencies. Infrequently, patients may experience sudden unexpected falls without loss of consciousness. These drop attacks are referred to as otolithic crises of Tumarkin and may lead to serious injury. They may occur presumably from either mechanical deformation or from sudden surges of nerve activity related to the utricle or saccule. This, in turn, distorts the patient’s sense of vertical, resulting in the reflexive loss of tone and a fall.

The main differential diagnosis is between Ménière disease and vestibular migraine. The key distinction is a greater likelihood of migraine in the personal history and the absence of characteristic hearing loss in vestibular migraine. Interestingly, Ménière disease and migraine occur together more than expected based on the epidemiology of the two conditions. The first attack of Ménière disease could mistakenly be diagnosed as vestibular neuritis, but if multiple vertigo attacks occur, the diagnosis of vestibular neuritis should be abandoned.

Ménière disease is idiopathic but appears to be associated with a process of endolymphatic hydrops.

KEY POINTS
- Although the classic triad of hearing loss, tinnitus, and vertigo is seen with Ménière disease, it is important to know that it causes characteristic unilateral low-frequency hearing loss with tinnitus and attacks of vertigo, with nausea and vomiting usually lasting hours.
- Ménière disease is idiopathic but appears to be associated with a process of endolymphatic hydrops.

Case 2-4
A 42-year-old woman presented for evaluation after experiencing five prolonged attacks of vertigo during the prior year. Each vertigo spell came on randomly and consisted of severe spinning with nausea and vomiting that lasted 2 to 3 hours. During these spells, she could not get up, walk, or move, and she tried to remain as motionless as possible. Just prior to the onset of the spinning she felt fullness in the left ear and heard a roaring tinnitus in the left ear, and her hearing became muffled on the left side. The vertigo abated gradually, but after each event she continued to feel a little off balance for the rest of the day. The first episode occurred 1 year ago, and she did not have another until about 6 months ago, but at the time of presentation they seemed to be occurring more often and were very disruptive to her life since she worked as an intensive care unit nurse.

Her examination was normal, and her hearing had improved since her last attack. Her brain MRI was normal. An audiogram showed about 35 dB of sensorineural hearing loss on the left only at the low frequencies, and her hearing on the right was completely normal.

Comment. A patient’s hearing may fluctuate in Ménière disease, and unilateral fluctuation is characteristic of the diagnosis, as is unilateral low-frequency hearing loss. Early in the course of Ménière disease, vestibular function as determined by videonystagmography with caloric testing may be normal so that the audiogram is more helpful and sensitive. Initial treatment includes a sodium-restricted diet with the possible addition of a diuretic.
function occurs, leading to vertigo, nausea, vomiting, and acute asymmetry of hearing, which causes louder tinnitus (often described as roaring) and muffled hearing in the affected ear.

When endolymphatic hydrops is caused by a known cause such as syphilitic otitis, the late effects of trauma, or a viral infection or autoimmune mechanisms, it is often referred to as Ménière syndrome and delayed or secondary endolymphatic hydrops.

Treatment during vertigo can consist of antivertiginous medications, as clinicians might consider for any cause of acute vertigo (Table 2-3). Management of Ménière disease with a goal to stop the recurrent vertigo and hearing symptoms should include a sodium-restricted diet to about 1500 mg/d and possibly the addition of a thiazide or another diuretic. Betahistine is not approved for interstate transport in the United States but may have a role in some cases of Ménière disease. It is dosed at 16 mg 2 times a day up to 32 mg 3 times a day and can be obtained at compounding pharmacies throughout the United States. Betahistine is also available in most countries of the world. Betahistine is a potent histamine H3 receptor antagonist and weak histamine H1 receptor agonist, but the mechanism of betahistine in Ménière disease is unknown. Some evidence suggests that this medication affects histaminergic neurons of the tuberomammillary nucleus in the posterior hypothalamus and also in the vestibular nuclei. It should be noted that no modern high-quality randomized clinical trials show that betahistine, sodium restriction, or diuretic use are beneficial in prevention of vertigo attacks in Ménière disease.
For patients with disabling vertigo attacks from Ménière disease despite adhering to a sodium-restricted diet and using a diuretic, other approaches should be considered by otolaryngologists, including administration of intratympanic corticosteroids or gentamicin and endolymphatic mastoid shunt procedures. In cases not otherwise responsive, vestibular neurectomy or labyrinthectomy are options, the latter only if the patient is already deaf in the affected ear. Ménière disease may also become a bilateral process, but most commonly does so within the first several years of symptom onset. Estimates on the incidence of bilateral Ménière disease vary widely, but are generally between 15% and 20% when strict diagnostic criteria are used. When present, bilateral involvement limits some of the destructive procedure options since treating both ears with a vestibular ablation procedure can result in bilateral vestibular loss.

**Bilateral Vestibular Loss**

Bilateral vestibular loss, sometimes referred to as Dandy syndrome, denotes bilateral peripheral vestibular loss. Recognizing bilateral vestibular loss is important to neurologists for three reasons: (1) bilateral vestibular loss can be very debilitating to the patient and is often but not always iatrogenic (Case 2-5), (2) physicians must have some suspicion for the presence of bilateral vestibular loss in order to even test for it and make the diagnosis, and (3) bilateral vestibular loss is partially remediable with proper physical therapy.

Bilateral vestibular loss may have several causes, including ototoxic medication exposure, bilateral Ménière disease, neurosarcoidosis, bilateral ear

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**Case 2-5**

A 53-year-old man with severe diabetes mellitus and diabetic neuropathy presented because of severe imbalance. He had developed pneumonia 7 weeks prior, and he had been admitted to the intensive care unit, was intubated, and received IV antibiotics for the subsequent 14 days. After he was extubated and started to move about, he noticed that he could not keep his balance well. Ever since he had been hospitalized, and despite outpatient physical therapy to regain his strength, he had been falling. His symptoms were especially severe in the dark such that he nearly always fell over. Sometimes, while watching television, he had noticed that the image oscillated with his own heartbeat, and sometimes he could not tell who was walking toward him unless he stopped walking himself and held still. Further history indicated that one of the antibiotics that had been administered for the treatment of his pneumonia was gentamicin.

His examination showed a consistently positive Romberg sign and absent knee and ankle reflexes due to severe diabetic polyneuropathy (no worse than in the past). Head impulse testing was abnormal in both directions, and videonystagmography with caloric testing confirmed the absence of peripheral vestibular responses on both sides. Brain MRI was normal.

**Comment.** This case illustrates one of the common causes of bilateral vestibular loss, which is due to exposure to an aminoglycoside. In this particular case, the patient had a preexisting severe diabetic polyneuropathy that is now compounded by the vestibular loss. In a sense, his vision is the only good sensory input he still has for maintaining balance.
surgery, some congenital disorders, and autoimmune inner ear disease. Among these, the most commonly encountered cause of acquired bilateral vestibular loss is use of a vestibulotoxic medication, and gentamicin is the most common of these. Nevertheless, nearly one-fourth of older patients with an imbalance of unknown origin have significant bilateral vestibular loss, presumably on a degenerative basis.

The symptoms may include some history of vertigo, but it is often minor, and the more vexing symptom for the patient is unsteadiness that is worse when walking, especially in darkness or on uneven ground. However, if patients make hand contact with a wall while walking, their balance significantly improves as they gain sensory information about their body position through the upper limb, which can help in the absence of useful input from the vestibular system. Oscillopsia, the perception of bouncing vision, is a characteristic symptom of severe bilateral vestibular loss but is only notable in severe bilateral vestibular loss, so not all patients have this symptom. Oscillopsia occurs because each step or movement causes minor head oscillations that are not countered well by the vestibuloocular reflex.

In a patient with new unsteadiness following a hospitalization and in whom no other cause is apparent, one should inquire whether the patient might have received aminoglycoside antibiotics, which could have caused bilateral vestibular loss. Unsteadiness may occur within 1 to 3 weeks of administration of an aminoglycoside. Some cases can be due to overdosage of an aminoglycoside, but some individuals are particularly susceptible and may develop bilateral vestibular loss even when the dosage is administered and monitored properly.

The examination of the patient with bilateral vestibular loss often includes a positive Romberg sign, abnormal head impulse test responses related to both sides, and a dynamic visual acuity decline of three lines on a Snellen chart or Rosenbaum card during rapid side-to-side head shaking. The symptoms may include some history of vertigo, but it is often minor, and the more vexing symptom for the patient is unsteadiness that is worse when walking, especially in darkness or on uneven ground. However, if patients make hand contact with a wall while walking, their balance significantly improves as they gain sensory information about their body position through the upper limb, which can help in the absence of useful input from the vestibular system. Oscillopsia, the perception of bouncing vision, is a characteristic symptom of severe bilateral vestibular loss but is only notable in severe bilateral vestibular loss, so not all patients have this symptom. Oscillopsia occurs because each step or movement causes minor head oscillations that are not countered well by the vestibuloocular reflex.

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The examination of the patient with bilateral vestibular loss often includes a positive Romberg sign, abnormal head impulse test responses related to both sides, and a dynamic visual acuity decline of three lines on a Snellen chart or Rosenbaum card during rapid side-to-side head shaking. Some mild nystagmus may be evident, but this is generally not prominent if the vestibular loss is symmetric and chronic. Videonystagmography with caloric testing shows bilaterally reduced nystagmus, and rotational chair testing would also be confirmatory of bilateral vestibular loss if it shows reduced vestibuloocular reflex gain responses during chair rotations at all frequencies.

Treatment consists of stopping any offending cause or medication. The next step in treatment is vestibular physical therapy to promote adaptation and substitution, with the goal of improving balance. Vestibular physical therapy can be helpful in achieving this in part by employing plastic properties of the CNS and partly by using “substitution” to better use vision and proprioceptive cues to stabilize gait and mobility. Vestibular rehabilitation improves dynamic visual acuity, diminishes oscillopsia, and lessens asymmetry of vestibuloocular function.

Superior Canal Dehiscence Syndrome
Superior canal dehiscence syndrome is an uncommon disorder caused by an abnormal opening (dehiscence) in the thin bony roof of the superior semicircular canal (Figure 2-7). The tendency for this may be related to congenital thinning of the temporal bone, which occurs in about 0.1% of the population. Once a bony opening has “eroded” through, this opening renders the membranous labyrinth unusually susceptible to changes in sound and pressure. Patients commonly report autophony (hearing one’s own voice and internal body sounds), ear
fullness or pressure, and various sensations of dizziness affected by certain sounds, vibrations, or straining. A Tullio phenomenon (becoming dizzy with exposure to some sounds) may also be reported in some patients. Neurologic examination is usually unrevealing in patients with this condition. The audiogram may show mild conductive hearing loss, but it is nonspecific. If the history prompts enough suspicion, the diagnosis is clarified by a thin-cut temporal bone CT and, if possible, cervical or ocular vestibular-evoked myogenic potential (VEMP) testing may serve as a physiologic confirmation. VEMP is an evoked potential related to saccule and utricle function, but the waveform of response in superior canal dehiscence syndrome is uniquely markedly increased and evoked with less stimulation. Since some temporal bone CT studies appear to show an apparent dehiscence but actually are just very thin (and there is a difference), VEMP may serve as another way to ensure it is not a false-positive CT of the temporal bone.

This condition, when severe enough to warrant it, is managed surgically and should be referred to an otolaryngologist for ongoing management. In order to consider this disease as a diagnosis and to make the proper referral, a certain awareness of this condition is important.

CENTRAL NERVOUS SYSTEM CAUSES OF VERTIGO AND DIZZINESS

CNS causes of vertigo are, of course, of great interest and concern to neurologists and other clinicians because of a potentially more ominous prognosis if the cause is in the brainstem or cerebellum. The potential conditions and lesions that may cause central vertigo are substantial, but fortunately, these causes are far less common than inner ear causes. When due to structural...
causes, most lesions are located within the medulla, cerebellum, or pontine tegmentum and middle cerebellar peduncle regions. Some central causes, such as anxiety, phobic dizziness, and vestibular migraine, are not due to structural lesions but are caused by more complicated mechanisms that are still poorly understood.

**Vestibular Migraine**

Migraine may be associated with vestibular symptoms such as episodic vertigo and chronic motion sensitivity. Vertigo and motion sickness unaccompanied by headache may also occur with migraine and together are referred to as migrainous vertigo, migraine-associated vertigo, or vestibular migraine. The pathophysiology of vestibular migraine is not known. Evidence suggests that certain brainstem nuclei may become hypersensitized, causing excessive sensitivity for pain reception (headache, allodynia), hearing (phonophobia), vestibular stimulation (vertigo, motion sickness, visual vertigo), vision (photophobia), and the sense of smell (osmophobia). Both environmental and genetic factors participate in migraine-associated vertigo mechanisms.

The dizziness of vestibular migraine varies both in duration and in description from patient to patient and even sometimes in the same patient. Most otologic vestibular disorders have stereotypic duration: episodes of BPPV last 10 to 30 seconds, attacks of Ménière disease last hours, and vestibular neuritis goes on for days to weeks. Migraine may behave quite variably in terms of spell duration. Most often, patients have either recurrent random spells of dizziness ranging from a few minutes to several weeks, and patients often have a general motion sensitivity and sometimes visual vertigo (Case 2-6). Visual vertigo, also called optokinetic motion sickness, is a syndrome in which seeing objects in motion (e.g., moving ceiling fans, moving scenery, general visual commotion, walking down grocery store aisles) causes dizziness and

**Case 2-6**

A 38-year-old woman presented with a several-year history of recurrent spells of dizziness, which most commonly consisted of a rocking sensation that lasted for 30 minutes to half of the day and occasional sensations of spinning that lasted for 2 to 10 minutes. These symptoms were not provoked reliably by any particular circumstance, but rather seemed to occur randomly. Even between episodes of dizziness, she became motion sick easily when riding in a car or whenever exposed to motion or even when seeing things move around her. The patient experienced nausea that varied in severity along with the dizziness. She had migraine headaches, but most of the time they occurred at times separate from the dizziness. Her dizziness was worse when she felt fatigued or tired and when she was under more stress. She also noticed that drops in barometric pressure were associated with an increase in her dizziness. Her quality of life was severely impacted by these ongoing symptoms.

Her examination was normal. Brain MRI and magnetic resonance angiography (MRA) of her head and neck had been recently performed and were normal. Videoystagmography testing and an audiogram were both normal.

**Comment.** This patient had a typical presentation of vestibular migraine with mixed symptoms including rocking dizziness, spinning, and dizziness when exposed to motion and visual commotion. Because of the impact on the patient’s life, a migraine prophylactic should be considered.
sometimes nausea even if the individual is not in motion. Migraine headaches are also common in vestibular migraine, although, in most cases, they do not occur at the same moment as the vertigo spell.

Recently published diagnostic criteria indicate that the diagnosis of vestibular migraine is made by at least five spells of dizziness, vertigo, and motion sensitivity that last between 5 minutes and 72 hours and are associated with unilateral headache, photophobia or phonophobia, or a migraine visual aura that occurs in some of the dizzy spells. The criteria are not perfect but at least are a good start. Diagnostic testing is generally not helpful except to exclude other causes. Hearing is unaffected, brain and vascular imaging is normal, and videonystagmogram testing is often normal or nonspecific, showing only some minor degree of static positional nystagmus in a minority of patients.

Vestibular migraine is not life-threatening nor does it shorten life span; however, it may have a dramatic impact on quality of life and may, in some cases, be disabling. Mild cases require no specific treatment and can be managed with education, dietary and lifestyle changes where appropriate, and vestibular suppressant medications (Table 2-3) if needed.

When the symptoms are frequent or severe and interfere with quality of life, treatment with migraine prophylactic medication may be needed. These medications may include the daily use of propranolol or similar β-adrenergic blockers (timolol, nadolol), tricyclic antidepressants (nortriptyline, imipramine, amitriptyline), topiramate, divalproex sodium, or verapamil. Unfortunately, no randomized controlled trials have been performed to guide treatment or compare medications to one another for best effectiveness, and we do not know if vestibular migraine responds to migraine prophylactic medications in the same way as migraine headaches. Although anecdotal, this author prefers platform treatment with either a tricyclic antidepressant, titrating from 50 mg/d to 75 mg/d at bedtime, or a serotonin norepinephrine reuptake inhibitor (SNRI) such as venlafaxine, titrating to about 75 mg/d, especially when anxiety, panic attacks, or depression are prominent and nausea is less prominent. For those with lesser or no anxiety overlay and with more nausea or more headaches, verapamil, titrating from 120 mg/d to 240 mg/d, is a well-tolerated medication, dosed as once daily if in the extended release form, and dosed at 3 times a day if in the regular formulation, with seemingly good efficacy. Topiramate dosed between 50 mg/d and 200 mg/d (started as a dose every night at bedtime, but which may be split to twice daily if it is better tolerated as the daily dosage is increased) is often effective. Topiramate is difficult for many patients because of its side effects, but it is reasonable to offer the medication when outlining choices for patients to consider. Using a combination treatment with two or even three migraine prophylactic medications has been a helpful strategy in many patients with severe vestibular migraine. In addition to the medications listed above, other medications to consider include zonisamide and occasionally acetazolamide.

Chronic Subjective Dizziness

Chronic subjective dizziness refers to a form of unexplained dizziness ongoing for longer than 3 months. This name was aptly coined as it implies chronicity and the subjective nature of the symptom; the name does not imply knowledge of its mechanism, which is appropriate since the mechanism is unknown. Recent ongoing efforts are underway to rename this

KEY POINTS

- Vestibular migraine is a variant of migraine in which vestibular symptoms predominate. Vestibular migraine should be considered in a patient with multiple recurrent episodes of vertigo or dizziness with a prior history of migraine headaches and no hearing loss or other neurologic deficits on examination.

- Currently, no randomized controlled trials exist to guide treatment of vestibular migraine, so management currently consists of the same medications used for migraine headache prophylaxis.
condition or some variation of it as “persistent postural-perceptual dizziness.”

It is discussed here as a CNS cause of dizziness since it is highly likely to be a complex syndrome related to behavior, maladaptive adjustment to symptoms, neurotransmitter function, and as yet unclear additional CNS mechanisms.

As noted above, the syndrome of chronic subjective dizziness is undergoing a possible renaming to persistent postural-perceptual dizziness. By whatever name, this condition affects females about five to one over males and is often described as a rocking or floating sensation without nausea and is not worsened by head motion nearly to the degree seen with most vestibular disorders. Symptoms are often worsened by sleep deprivation and stress (Case 2-7). Interestingly, some patients have visual vertigo, and considerable but not complete overlap exists with some presentations of vestibular migraine. Cervical pain, migraine headaches, anxiety, and depression are common comorbidities. Patients note aggravation of dizziness by complex stimulation including visual commotion, flickering and sometimes fluorescent lights, scrolling on computer monitors, object motion in general, and not.

Case 2-7

A 54-year-old woman presented with a 3-year history of dizziness that initially was intermittent but had become constant over the past year. She described a feeling of floating and lightheadedness and was mentally unable to focus. These symptoms were not provoked by anything but bothered her more when she was tired. She felt the sensation every day. She had no associated nausea, headache, prior motion sickness, or focal neurologic symptoms. She felt that she was at the end of her rope with the dizziness and had to stop working as a legal assistant 6 months ago, as she could not continue the stress of work, and she drove only very little. She periodically experienced panic attacks and felt that her life was spiraling out of control.

She had sought medical advice extensively and had had a brain MRI, magnetic resonance angiography (MRA) of the head and neck, CT head and temporal bone, videonystagmography, audiogram, tilt table testing, extensive blood work, cervical spine MRI, and extensive cardiologic workup, all of which were normal. She had seen an endocrinologist, and she had tried lorazepam, which had helped slightly. In addition, she had been prescribed gabapentin, diuretics, meclizine, buproprion, and buspirone; had been advised to start a low-sodium diet; and went through 8 weeks of vestibular rehabilitation, all without clear benefit. She was desperate for relief. Her examination was completely normal, and she displayed a good ability to turn, to balance on one leg, and to walk quite normally.

Comment. This case shows how some patients with chronic subjective dizziness have self-imposed restrictions on what they feel they can do, and how these restrictions seem to be out of proportion to the degree of expected impairment based on their examination and mobility. In this case, her extensive history indicated that she was very eager to find relief and demonstrated a willingness to try the recommendations of her physicians. In this kind of case, a selective serotonin reuptake inhibitor (SSRI) or serotonin norepinephrine reuptake inhibitor (SNRI), along with education about the nature of chronic subjective dizziness may be most helpful. Sometimes vestibular rehabilitation therapy can be helpful for restoring confidence and mobility.
infrequently by having too many sensory or cognitive inputs to juggle. Chronic subjective dizziness should be suspected in a patient who feels severely disabled from chronic dizziness and yet appears normal on examination and in whom vestibular and cardiovascular evaluations have been unrevealing.

Chronic subjective dizziness may develop de novo or may evolve after some kind of emotionally impactful event (e.g., automobile accident, panic attack), especially if associated with dizziness or vertigo. Chronic subjective dizziness may linger following minor head injury and become entwined with some of the other symptoms of post-concussive syndrome.

The differential diagnosis of this common syndrome includes vestibular migraine, incompletely compensated unilateral vestibular loss, superior canal dehiscence syndrome, postural hypotension, postural orthostatic tachycardia syndrome, and many other potential causes. Many possibilities could underlie such symptoms at the outset, but the examination remains normal. The presence of an anxious affect is not itself proof of cause, and Table 2-5 describes differences between chronic subjective dizziness and vestibular migraine, as these syndromes may look similar. Indeed, aside from nausea and migraine headaches, considerable overlap exists when the symptom is not spinning vertigo but simply rocking or floating. Brain and cerebrovascular imaging, vestibular testing, and audiometry are all normal in this group of patients.

Because of the common co-occurrence of chronic subjective dizziness and generalized anxiety, it is tempting to consider this a cause and effect phenomenon. This is possible but not yet substantiated by the literature.

Treatment should first and foremost consist of validation and education.

### Table 2-5: Comparison of Clinical Features in Vestibular Migraine and Chronic Subjective Dizziness

<table>
<thead>
<tr>
<th>Feature</th>
<th>Chronic Subjective Dizziness</th>
<th>Vestibular Migraine</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Abrupt (especially if after an emotionally impactful event), less often gradual</td>
<td>Abrupt or gradual onset</td>
</tr>
<tr>
<td>Description</td>
<td>Rocking, swaying, or floating</td>
<td>Spinning, rocking, tilting, floating, general motion intolerance, and motion sickness</td>
</tr>
<tr>
<td>Migraine headaches</td>
<td>Present in about 25% of cases</td>
<td>Present in more than 80% of cases</td>
</tr>
<tr>
<td>Anxiety</td>
<td>Present in two-thirds of cases</td>
<td>Present in nearly one-half of cases</td>
</tr>
<tr>
<td>Effect of vestibular rehabilitation therapy</td>
<td>Modest improvement in some</td>
<td>Minimal if any improvement</td>
</tr>
<tr>
<td>Examination</td>
<td>Normal</td>
<td>Normal, possibly cautious gait</td>
</tr>
<tr>
<td>Nausea</td>
<td>Uncommon</td>
<td>Common</td>
</tr>
<tr>
<td>Visual vertigo</td>
<td>Common</td>
<td>Common</td>
</tr>
</tbody>
</table>

**KEY POINTS**

- Chronic subjective dizziness (persistent postural perceptual dizziness) is a heterogenous group of conditions with a common theme of nonvertiginous dizziness without nausea that may be chronic or intermittent, may occur spontaneously or after a condition that causes vertigo, and may respond to some selective serotonin reuptake inhibitors or serotonin norepinephrine reuptake inhibitors.

- Chronic subjective dizziness is a common cause of chronic nonvertiginous dizziness that may sometimes occur spontaneously but commonly develops after an emotionally impactful situation (e.g., trauma, car accident, attack of vertigo, chronic stress at work or home).
KEY POINT

- Mal de débarquement syndrome refers to the persistence of a rocking sensation after a boating excursion or cruise and returning to land. Symptoms may last months to years, and no treatment has been found to be effective.

about the best understanding of this perplexing condition. In many cases, establishing rapport with the patient, explaining the interplay between anxiety and dizziness, and treating any remediable otogenic vestibular comorbidities goes a long way in allaying the patient’s anxiety. In some cases, working with a vestibular physical therapist can help patients regain confidence, hope, and improve anxiety.38

Stress reduction, improved quality of sleep, and possibly pharmacotherapy should be discussed if the patient is severely troubled and wishes to try treatment. Some patients improve well on selective serotonin reuptake inhibitors (SSRIs), and some do not. This author prefers, based purely on anecdotal experience, the use of an SNRI such as venlafaxine or duloxetine or possibly tricyclic antidepressants. These medications usually require about 4 to 5 weeks to assess effectiveness, so patients must be encouraged not to stop taking them too early to reap the benefit. Vestibular suppressants seem to rarely be helpful, and vestibular rehabilitation is helpful mainly in those afflicted by avoidance behavior and excessively cautious gait. Chronic subjective dizziness remains a common and challenging condition.

Mal de Débarquement Syndrome

Mal de débarquement syndrome is an uncommon vestibular disorder characterized by a persistent sense of rocking that continues after disembarking a boating excursion or cruise. Many individuals experience transient “sea legs” after returning to land after being on a boat but, in a small percentage of people, the feeling persists sometimes for weeks, months, or years. The syndrome is most common in women and has its onset soon after getting off a boat, at which time patients feel they are still swaying or rocking. Usually, no associated nausea or motion sickness occurs, and if they return to the motion stimulus, such as getting back on the boat, the symptoms again abate. Its cause is unknown but some have speculated it to be a disorder of the vestibular velocity storage mechanism with some limited ability to adapt.

Table 2-639 lists proposed diagnostic criteria for this disorder. Examination in these patients is normal, and their gait typically appears normal. No nystagmus is seen, and no abnormality on brain MRI or vestibular testing is present. The differential diagnosis for such dizziness includes vestibular migraine and chronic subjective dizziness. Unlike vestibular migraine, no nausea occurs, and a history of migraine may not necessarily be present. Unlike chronic subjective dizziness, the onset is typically following disembarkment from a boat.

No established treatment is known, but a nonblinded nonrandomized trial suggests that inducement of readaptation of the vestibuloocular reflex may result in improvement.40 In this approach, patients are exposed periodically to a rocking that mimics the pace and direction of their perceived sway while in a full-field optokinetic stimulus. Time will tell if this approach is truly effective in this group of patients.

CENTRAL NERVOUS SYSTEM STRUCTURAL LESIONS CAUSING DIZZINESS

Lesions that disrupt the vestibular pathways of the CNS vestibular system can, not surprisingly, lead to vertigo and imbalance. Any central (or peripheral) process that leads to an acute vestibular asymmetry can lead to vertigo. The brain can adapt to this asymmetry over time, but imbalance may still occur if vestibulocerebellar pathways are impacted. For example, a stroke may lead to sudden
onset of vertigo, whereas an acoustic neuroma (vestibular schwannoma) grows so slowly that adaptation to the asymmetry occurs gradually and imperceptibly. The result is that an acoustic neuroma is much less likely to present with vertigo than an acute event such as a stroke.

A CNS lesion that is anatomically related to vestibulocerebellar pathways may lead to vertigo, dizziness, or imbalance. Too many possible lesions can occur to enumerate here, but the location is the key to properly ascribing the lesion to the report of dizziness or vertigo. Examples are outlined in Table 2-7.

**Epileptic Vertigo**

Epileptic vertigo is an ictal condition in which a seizure involving the vestibular cortex is the cause of dizziness or vertigo. This is a rare condition even among patients known to have epilepsy, but it is even rarer for isolated vertigo to be the presenting feature of epilepsy. In most cases, other clinical features suggest seizures such as automatisms, loss of awareness, and convulsions that follow the aura of vertigo. Of course, a brain MRI should be performed when this diagnosis is strongly considered. EEG monitoring may be necessary to confirm an ictal process when no structural lesion is identified on brain imaging. Differential diagnostic considerations could include transient ischemic attacks, migrainous vertigo, and an episodic ataxia syndrome such as episodic ataxia type 2. Treatment would include standard antiepileptic medications as with any other seizure disorder.

**Craniocervical Junction Syndromes**

Within the region of the craniocervical junction are the vestibulocerebellum and posterior medulla, which carry vestibular pathways. Lesions in this region may lead to vertigo, dizziness, or imbalance. The most prototypical of such lesions is a Chiari malformation, but other disorders affecting this area

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**TABLE 2-6 Diagnostic Criteria of Persistent Mal de Débarquement Syndrome**

| A. Onset of symptoms after exposure to passive motion, such as following travel by boat/cruise ship or airline |
| B. Main symptom is illusory rocking motion |
| C. Symptoms improve with reexposure to passive travel, such as boat or car travel |
| D. Nausea is not a prominent symptom |
| E. Minimum of 3-month duration of symptoms |
| F. Not better accounted for by another International Classification of Headache Disorders, Third Edition diagnosis or by another vestibular disorder |

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**KEY POINTS**

- Structural, ischemic, or degenerative central nervous system processes that affect the vestibular nuclei, flocculonodular lobe (vestibulocerebellum) of the cerebellum, the cerebellar peduncles, and, very rarely, the cerebral cortex (as may occur with epileptic vertigo) may lead to central vertigo, sometimes with accompanying nystagmus.

- Isolated vertigo as a manifestation of epilepsy is rare, but when it occurs, is usually in individuals with known seizure disorders and is exceptionally rare as the presenting manifestation of new-onset epilepsy.
include atlantoaxial subluxation, and, more rarely, basilar invagination.

A Chiari malformation occurs when the lower part of the cerebellum does not fully migrate rostrally during fetal development, so that the cerebellar tonsils remain below the foramen magnum. This may lead to symptoms early or later in life due to compression of the brainstem or spinal cord or by disrupting the normal flow of CSF with the subsequent development of a cervical syrinx. The Chiari malformation may occur due to disorders of fetal development of the brain or skull. The compression of the cerebellum from this condition may lead to occipital headaches, vertigo, nystagmus, and ataxia in early childhood or later in life. Some Chiari malformations are found incidentally on brain MRI without ever causing symptoms, whereas other cases have a slowly progressive course. One commonly used radiographic definition defines the presence of Chiari malformation if the cerebellar tonsils are more than 5 mm below the foramen magnum.42

Treatment is based on symptom severity and consists of suboccipital decompressive surgery. Because of the prevalence of Chiari malformations as incidental findings on routine brain MRIs,

### TABLE 2-7 Central Nervous System Regions That May Be Associated With Vertigo, Nystagmus, or Ataxia

<table>
<thead>
<tr>
<th>Anatomic Location</th>
<th>Structures Affected</th>
<th>Signs or Symptoms</th>
<th>Lesion Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dorsolateral medulla</td>
<td>Vestibular nuclei, root entry zone cranial nerve VIII</td>
<td>Nausea, nystagmus, vertigo, ataxia</td>
<td>Medulloblastoma, metastasis, multiple sclerosis, medullary infarction</td>
</tr>
<tr>
<td>Floor of fourth ventricle</td>
<td>Vestibular nuclei, especially superior vestibular nucleus</td>
<td>Nausea, nystagmus, vertigo, ataxia</td>
<td>Medulloblastoma, cysts</td>
</tr>
<tr>
<td>Anterior cerebellar vermis</td>
<td>Cerebellar connections</td>
<td>Ataxia</td>
<td>Alcohol-related cerebellar ataxia</td>
</tr>
<tr>
<td>Dorsal cerebellar vermis</td>
<td>Flocculus, nodulus, uvula connections; dorsal vermis; fastigial nucleus</td>
<td>Nystagmus, ataxia, possibly vertigo, saccadic dysmetria</td>
<td>Cerebellar degeneration</td>
</tr>
<tr>
<td>Superior cerebellar peduncle</td>
<td>Cerebellar efferents</td>
<td>Positional vertigo</td>
<td>Multiple sclerosis</td>
</tr>
<tr>
<td>Middle cerebellar peduncle</td>
<td>Pontocerebellar fibers</td>
<td>Ataxia, dysarthria, ipsilateral limb clumsiness</td>
<td>Pontine infarct, cavernous malformation</td>
</tr>
<tr>
<td>Inferior cerebellar peduncle</td>
<td>Vestibulocerebellar afferents</td>
<td>Isolated vertigo, possible ipsilateral limb clumsiness</td>
<td>Cavernous malformation, arteriovenous malformation</td>
</tr>
<tr>
<td>Anterior cerebellum</td>
<td>Flocculus, nodulus, uvula</td>
<td>Vertigo, ataxia, gaze-evoked nystagmus</td>
<td>Basilar meningitis</td>
</tr>
<tr>
<td>Vestibular cortex</td>
<td>Right temporal perisylvian cortex</td>
<td>Vertigo</td>
<td>Partial epilepsy</td>
</tr>
<tr>
<td>Dorsal midbrain</td>
<td>Rostral interstitial nucleus of medial longitudinal fasciculus, interstitial nucleus of Cajal</td>
<td>Disconjugate vertical/torsional nystagmus, vertical gaze disorders</td>
<td>Cavernous malformation, pinealoma, arteriovenous malformation</td>
</tr>
</tbody>
</table>

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**KEY POINT**

- The incidental discovery of a Chiari malformation on routine brain MRI occurs frequently, and physicians should consider the more common causes of headache and dizziness such as migraine or inner ear causes before attributing these symptoms to the Chiari malformation.
care should be taken before ascribing dizziness or headaches necessarily to the Chiari malformation without considering more common conditions such as migraine or inner ear conditions.

Atlantoaxial subluxation is a serious medical condition that is usually due to erosion or malformation of the ligaments holding C1 and C2 together. Possible causes include traumatic rupture of the ligaments, Down syndrome, and rheumatoid arthritis. Patients with atlantoaxial subluxation may experience severe headaches, neck pain, and vertigo and should additionally be evaluated with flexion and extension cervical spine radiographs since routine CT spine or brain MRI do not necessarily show laxity in these ligaments.

VASCULAR CAUSES OF VERTIGO AND DIZZINESS

Vertebrobasilar insufficiency occurs when there is inadequate blood flow through the vertebral and basilar arteries that supply the brainstem and cerebellum. Momentarily insufficient blood flow may cause symptoms because of ischemia of the brainstem or cerebellar structures, resulting in dizziness. A focal area of vascular narrowing is the usual cause.

If stenosis of the right or left subclavian artery is just proximal to the origin of the vertebral artery, it may cause reversal of blood flow in the ipsilateral vertebral artery. This rare situation is referred to as subclavian steal syndrome because the subclavian artery “steals” blood flow from the vertebrobasilar system to supply the upper limb. In such cases, vertigo and other symptoms of vertebrobasilar insufficiency may be caused or exacerbated by exercise of the arm on the affected side. Vascular imaging may localize the site of the narrowing and the reversal of blood flow.

Transient ischemic attacks in the vertebrobasilar arterial system may produce vertigo, imbalance and ataxia, slurred speech, nystagmus, diplopia, drop attacks, and visual hallucinations or visual field defects. Vertigo may be an isolated initial symptom of vertebrobasilar insufficiency or may be accompanied by other brainstem symptoms. In most cases, treatment of vertebrobasilar insufficiency consists of controlling vascular risk factors and using antiplatelet drugs. Table 2-8 delineates the clinical features of several brainstem vascular syndromes that can be associated with vertigo.

Cerebellar strokes affecting the distribution of the vertebral artery, posterior inferior cerebellar artery, anterior inferior cerebellar artery, or superior cerebellar artery can result in isolated cerebellar infarction (Case 2-8). If the infarction affects portions of the cerebellum that carry vestibular fibers, vertigo and ataxia may occur. Small cerebellar infarctions in the cerebellar hemispheres are more likely to result in vertigo or substantial ataxia than are midline or parasagittal lesions such as those of the flocculus or nodulus.

Spontaneous intraparenchymal cerebellar hemorrhage may present with vertigo, nausea, vomiting, headache, the inability to stand or walk, and unilateral limb clumsiness. Involvement of the cerebellum can often be distinguished from peripheral vestibular vertigo because the former results in dysmetria and limb clumsiness, neither of which occurs with peripheral vestibular lesions. Brain imaging by CT or MRI accompanied by vascular imaging by CT angiography or MRA are essential in documenting the status of the cerebral vasculature.

Conditions that can produce focal and transient neurologic findings that could cause or mimic a transient ischemic attack but that do not show occlusion

KEY POINT

- Vascular narrowing of the vertebral arteries can lead to episodes of inadequate blood flow to the brainstem and cerebellum (vertebrobasilar insufficiency) and may cause isolated vertigo or ataxia in episodes lasting several minutes.
of the vessels include cardiogenic thromboemboli (including paradoxical emboli), hyperviscosity syndromes, complicated migraine, and, rarely, mitochondrial cytopathies.

Specifics of the diagnosis and management of patients with transient ischemic attacks and stroke are outside of the scope of this discussion; for more information, refer to the February 2017 Continuum issue, Cerebrovascular Disease.\(^5\) For patients with completed infarction, the addition of physical therapy may speed and improve return of vestibular function.

**Multiple Sclerosis**

Multiple sclerosis (MS) may present with dizziness, vertigo, or imbalance, particularly as a result of lesions affecting the brainstem or cerebellum that affect the vestibular system; such lesions may also result in nystagmus, ataxia, slurred speech, and diplopia. Despite the increased risk of CNS causes in patients with MS, non-MS causes such as BPPV and vestibular migraine may also occur, and the clinician should be careful about assuming that dizziness or vertigo are directly related to MS. However, in patients with known or possible MS, the presence of gaze-evoked nystagmus, spontaneous upbeat or downbeat nystagmus, periodic alternating nystagmus, ocular bobbing, seesaw nystagmus, pendular nystagmus, dissociated nystagmus with internuclear ophthalmoplegia, or upgaze restriction with convergence-retraction nystagmus are strong indicators of MS involvement, since none of these forms of nystagmus can be of peripheral vestibular origin.

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**TABLE 2-8** Selected Vascular Syndromes Associated With Vertigo and Ataxia

<table>
<thead>
<tr>
<th>Vascular Syndrome</th>
<th>Blood Supply</th>
<th>Common Cause</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vertebrobasilar transient ischemic attack</td>
<td>Vertebral arteries, basilar artery, posterior inferior cerebellar artery or anterior inferior cerebellar artery, rarely superior cerebellar artery</td>
<td>Atherosclerosis</td>
<td>Vertigo, clumsiness, ataxia, dysarthria, diplopia, drop attacks</td>
</tr>
<tr>
<td>Lateral medullary infarction (Wallenberg syndrome)</td>
<td>Posterior inferior cerebellar artery</td>
<td>Atherosclerosis or vertebral artery dissection</td>
<td>Vertigo, ipsilateral facial numbness, limb clumsiness, Horner syndrome, diplopia, lateral pulsion of saccades, asymmetric-gaze nystagmus, slurred speech, falling to one side, contralateral loss of pain and temperature sensation</td>
</tr>
<tr>
<td>Lateral pontine stroke</td>
<td>Anterior inferior cerebellar artery</td>
<td>Atherosclerosis</td>
<td>Vertigo, ipsilateral facial numbness, facial paresis, unilateral hearing loss, limb dysmetria, Horner syndrome, diplopia, lateral pulsion of saccades, asymmetric-gaze nystagmus, slurred speech, falling to one side, contralateral loss of pain and temperature sensation</td>
</tr>
</tbody>
</table>

**KEY POINT**

- Patients with multiple sclerosis are certainly predisposed to central nervous system causes of vertigo, but peripheral vestibular disorders are also common in this population.
Case 2-8
A 75-year-old man with a history of hypertension, hyperlipidemia, and coronary artery disease presented for acute vertigo and clumsiness on the right side with falling to the right side. He had experienced about 6 weeks of intermittent vertigo and had a pending appointment with an otolaryngologist for suspected benign paroxysmal positional vertigo, although he had not actually been examined yet. The recent spells of vertigo were random and would last for 2 to 5 minutes or so, and he would feel spinning and a bit “drunk,” but then they would pass. He ordinarily took aspirin but had stopped it over the past few months because of gastrointestinal upset that he thought might be due to the aspirin. On the day of presentation, he awoke from bed in the morning severely off balance, felt some spinning, and fell to the floor before he was brought to the emergency department by paramedics.

Examination showed that his speech was mildly slurred, he had horizontal gaze-evoked nystagmus that was most prominent when looking to his right, and a right Horner syndrome was present. He had marked right hemiataxia, poor balance, and some reduced sensation to pinprick on the left side of the body when compared to the right side. Brain MRI showed a large cerebellar infarction on diffusion-weighted imaging, with some additional involvement of the right lateral medulla (Figure 2-8).

Comment. Transient ischemic attacks may present with isolated spells of dizziness lasting minutes. In such cases, the spells are not positionally provoked, and in between spells no examination findings are seen. This patient was likely having transient ischemic attacks in the form of spells of isolated dizziness, and eventually he had a posterior inferior cerebellar artery occlusion that led to a large cerebellar and lateral medullary stroke (Wallenberg syndrome).
For patients with MS with vertigo, a standard examination for neurologic signs related to dizziness should be undertaken. This should include the Dix-Hallpike maneuver and possibly additional vestibular testing if no other new CNS signs or MRI-evident lesions are present.

Specifics of the diagnosis and management of patients with MS are outside of the scope of this discussion; for more information, refer to the June 2016 Continuum issue, Multiple Sclerosis and Other Demyelinating Diseases. However, for patients with central vertigo and nystagmus due to MS, medications such as gabapentin, 4-aminopyridine, memantine, and occasionally baclofen can sometimes help diminish the nystagmus and, to a lesser degree, the dizziness, if any occurs.

Episodic Ataxia Syndromes

Episodic ataxia type 2 is a rare sporadic or inherited ataxia disorder characterized by recurrent episodes of vertigo and ataxia. The onset of symptoms may occur in childhood or early adulthood and may continue undiagnosed well into late adulthood. This disorder is due to various mutations in the P/Q-type voltage-sensitive calcium channel (CACNA1A) in the case of episodic ataxia type 2.

Episodic ataxia type 2 manifests with recurrent vertigo and ataxia often associated with downbeat nystagmus, both during and even between attacks of vertigo and ataxia. Some patients exhibit no abnormalities between attacks, but often downbeat nystagmus is seen in the supine position even between spells. During spells, patients report dizziness, loss of balance, and sometimes slurred speech that may last from minutes to a day or two. The episodes may begin in childhood and continue into adulthood. No specific diagnostic gene tests are commercially available for this condition. The diagnosis should be suspected in a patient with a long history of recurrent bouts of unexplained vertigo associated with slurred speech and ataxic gait. Downbeat spontaneous or positional nystagmus even between spells is very suggestive of the diagnosis but is not present in all cases. Brain MRI may show midline cerebellar vermis atrophy in some but not all patients.

The differential diagnosis includes spinocerebellar ataxia type 6 and vestibular migraine, but in the case of the latter, downbeat nystagmus would not be expected. The diagnosis is made by history and response to acetazolamide. Transient ischemic attacks from verteobasilar insufficiency are also a differential consideration.

This condition is an important treatable cause of vertigo because ataxia and vertigo attacks may subside with the use of acetazolamide in daily doses usually ranging from 375 mg/d to 1500 mg/d. Responsiveness to acetazolamide is further confirmation of the diagnosis of episodic ataxia type 2.

Episodic ataxia type 1 is a separate disorder related to disturbances in potassium channels and differs clinically because the attacks of vertigo and ataxia are shorter in duration and may be associated with myokymia even between attacks. If the patient has dysarthric speech during spells or interictal myokymia, episodic ataxia type 1 should be considered. Treatment for episodic ataxia type 1 and other subtypes is not well established.

HEMODYNAMIC CAUSES OF DIZZINESS

Orthostatic hypotension is common in older people and may cause dizziness without documented syncope based on transient reductions in global cerebral blood flow. Periodic hypotensive episodes may also occur even after the
patient has been upright for a period of time or when orthostatic vital signs have been normal. Orthostatic hypotension may occur 5 to 30 minutes after being asymptomatic in the upright position, so it can be missed as a cause of periodic syncope or presyncope dizziness. For a discussion of syncope and presyncope, refer to the article “Syncope” by William P. Cheshire Jr, MD, FAAN, in this issue of Continuum.

Disability and Impairment from Dizziness or Vertigo

In ambulatory neurology, it is not uncommon for the treating neurologist to be asked to address the work capabilities of a patient with vertigo or dizziness. When clear neurologic deficits exist, as may be seen in the aftermath of a brainstem or cerebellar stroke, the treating neurologist is well equipped to judge the patient’s limitations. Communication of these limitations to the workplace, if requested by the patient, is usually straightforward and sufficient to satisfy any questions of work capacity. However, in cases where the symptoms are entirely subjective and the severity is self-reported, it is more challenging for the physician, patient, and employer to reach an agreement that the patient can work some or not at all.

When a patient is affected by dizziness that is lacking objective findings, several questions should be considered (Table 2-9). Having a clear and credible diagnosis that explains symptoms is important, although understandably, in some cases, a clear diagnosis is what is lacking.

In such cases, awareness of whether or not the occupation in question has components considered to be “safety sensitive” may influence the approach. Safety sensitive refers to any activity where impaired performance could result in significant harm to self or others. For example, a commercial airline pilot who appears normal but reports periodic blackouts or spells of disorientation cannot be cleared to return to his occupation from a safety standpoint. Other jobs may have safety-sensitive components that are not as obvious or are less subject to regulation. In those

<table>
<thead>
<tr>
<th>TABLE 2-9 Questions to Consider in Determining Work Capability in Patients With Subjective Dizziness</th>
</tr>
</thead>
<tbody>
<tr>
<td>▶ Is there a diagnosis that explains the symptoms, and is the diagnosis certain?</td>
</tr>
<tr>
<td>▶ What is the nature of the patient’s occupation?</td>
</tr>
<tr>
<td>▶ Are there objective findings that support the patient’s diagnosis, and are such findings expected?</td>
</tr>
<tr>
<td>▶ Is there congruence between the severity and limitations reported and actual activity limitations?</td>
</tr>
<tr>
<td>▶ Is there consistency between the severity and the examination findings and observations?</td>
</tr>
<tr>
<td>▶ Is the patient’s report and symptom description credible?</td>
</tr>
<tr>
<td>▶ Is there consistency between the expected response to treatment and the actual response?</td>
</tr>
<tr>
<td>▶ Does the patient appear compliant and eager to improve and return to work?</td>
</tr>
</tbody>
</table>

KEY POINT

- Delayed orthostatic hypotension may occur 5 to 30 minutes after the patient has been upright and is thus not identified by routine orthostatic vital signs but can be a cause of recurrent transient dizziness (or syncope).
cases, clear communication between the treating physician and the employer will be necessary for all parties to arrive at correct decisions regarding work.

In cases of dizziness not involving safety-sensitive issues, verification of the presence and severity of disease may be the more immediate concern. In these cases, any objective findings that support the diagnosis and the patient’s reported limitation can be important, but some conditions by their nature (e.g., vestibular migraine) do not exhibit examination or test abnormalities. Consistencies between reported symptoms and observations add to credibility, whereas embellishment or exaggeration by the patient can detract from it. The impact of symptoms on nonwork-related activities is also important to consider since being a little uncomfortable but still functional will likely not meet a reasonable threshold for complete impairment from work.

In some cases, accommodations or advice to avoid specific kinds of activities may be more appropriate than simply not working at all. For example, limitations in the number of hours worked per week or in a given time span may make sense in particular cases. Finally, assessment of how hard the patient is trying to overcome reported impairments, such as compliance with recommended treatment, may be helpful in the assessment of work capacity. Ordinarily, when a person is significantly impaired with regard to personal comfort, function, or both, a physician would expect to see evidence of reasonable interest in and effort toward improving. The absence of such evidence may call into question the severity of the reported symptoms. In addition, responses to treatment successes and failures can be weighed in terms of whether or not they are consistent with a significantly impaired person who is attempting to improve, or more like an individual with little or no concern regarding his or her condition (in the absence of any neurologic basis for indifference).

When dizziness is considered to be significant by the treating neurologist, typical restrictions (advice from a physician on what not to do) for patients would be to avoid working at heights greater than 6 feet without safety harnessing or a railing. Other restrictions might include avoidance of climbing ladders to rungs greater than 6 feet above the ground. Driving restrictions are sometimes necessary, but most forms of dizziness occur with enough forewarning that driving can be accomplished, and if vertigo ensues, the patient has time to pull over. This is generally true for vestibular migraine and for Ménière disease. Patients with BPPV rarely get dizzy while driving since the provocative positioning is unlikely to occur when driving. Beyond that, in the case of BPPV, treatment is so highly effective that if a patient continues to have dizziness, one should question whether the diagnosis is correct, and if correct, consider whether the treatment maneuver has been done correctly.

Ménière disease can sometimes cause work impairment because attacks of vertigo last hours, occur at unpredictable times, and may have lingering nausea and dizziness for 1 to 2 days even after the full vertigo attack subsides. When evaluating a patient with Ménière disease, however, the physician should determine if the diagnosis appears correct since it is often misdiagnosed or listed as a potential cause. Unilateral hearing loss is almost a sine qua non of Ménière disease that is severe enough to cause impairment, so if audiometry is normal, the physician should question this as the basis for any work impairment.

Vestibular migraine can sometimes cause impairment, but this is uncommon.
Most patients can improve substantially with treatment using one or more migraine prophylactic medication combined with lifestyle adjustments.

Work restrictions and accommodations should not be required for acute vertigo for more than a temporary period of time because it is a self-limited problem. However, individuals with chronic vertigo might require ongoing work restrictions or accommodations. Positional vertigo could require adaptation of work areas to assure the individual is not forced to move the head at certain angles. Individuals may need to avoid tasks requiring especially good balance or physical maneuvering, such as working at heights or around moving machinery or driving.

Overall, the assessment of work capacity in patients with dizziness must be considered on an individual basis, with important factors including the nature and severity of the underlying condition, response to treatment, and whether or not special circumstances may be involved, such as safety-sensitive job duties.

**TRENDS AND FUTURE DIRECTIONS**

New developments are refining our definitions and the conditions that can cause dizziness and vertigo. Randomized controlled trials in the treatment of conditions such as vestibular migraine and improved diagnostic criteria for many vestibular disorders will improve the effectiveness of our treatments.

Meanwhile, advances in vestibular testing appear to now provide the ability to test the function of all parts of the labyrinth, including the long elusive utricle and saccule. Ultimately, the ability to diagnose causes of dizziness and vertigo will depend on the history and examination and recognition of the patterns that help separate important causes of vastly differing origins.

**CONCLUSION**

Dizziness and vertigo are very common symptoms, and determining the cause of dizziness and vertigo requires a combination of a careful history as well as consideration of the quality of the symptoms, triggers, duration of symptoms, and accompanying clinical features. Examination includes assessment of cerebellar and cranial nerve function, nystagmus, and, in some cases, the Dix-Hallpike maneuver and head impulse testing. Once this database of information is acquired, it is a matter of pattern recognition and searching for overlapping patterns. Additional narrowing of the cause can be obtained by CT and MRI studies and quantitative vestibular and audiometric testing.

Many causes of dizziness exist, ranging from mechanical, infectious, inflammatory, demyelinative, degenerative, and complex or unknown mechanisms. The challenge for all clinicians evaluating patients with dizziness and vertigo is to develop their own approach and algorithm suitable to their interest and background.

**REFERENCES**


Dizziness


46. Krieger SC. Multiple sclerosis and other demyelinating diseases. Continuum (Minneap Minn) 2016;22(3).
