REVIEW ARTICLE

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Vestibular Testing

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ABSTRACT

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

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

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

INTRODUCTION

V*estibular testing* is defined as the quantification of the function of the motion-sensing portions of the inner ear (semicircular canals and otoliths). Vestibular testing is generally performed in the context of an evaluation of the symptom of dizziness, and such evaluations often benefit from information about hearing. Accordingly, although this article mainly focuses on vestibular assessments, it also includes content about how hearing testing contributes to forming a diagnosis.

Vestibular testing has improved immensely over the past 30 years. Five motion sensors are located in each inner ear: three semicircular canals and two otolith organs (the utricle and saccule). Ideally, one should be able to quantify the function of all five.

In 1914, Robert Bárány was awarded the Nobel Prize in Physiology or Medicine for the development of a test of the lateral semicircular canal.[1] Since then, and especially in the past decade, new tests have come into clinical use that can quantify the remaining two canals (anterior/superior and posterior), as well as both otolith organs.

The main goal of vestibular testing is to determine whether vestibular function is normal or abnormal, testing in each sensor of the peripheral vestibular apparatus, which may identify when central vestibular and ocular motor pathways exhibit dysfunction. For example, if examination identifies subtle

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spontaneous downbeating nystagmus but testing shows normal canal and otolith function, then this would suggest that peripheral vestibular function is intact and the observed nystagmus must therefore be arising from a central vestibular abnormality.

BEDSIDE TESTING

Dizziness has numerous causes, many of which are unrelated to the peripheral vestibular system (eg, blood pressure fluctuations). A thorough neurologic history and general neurologic evaluation are desirable. The approach to the patient with dizziness when time does not permit a complete neurologic evaluation has been discussed elsewhere[2] and is discussed in the article "Approach to the History and Evaluation of Vertigo and Dizziness" by Terry D. Fife, MD, FAAN, FANS,[3] in this issue of *Continuum*.

In this section, the repertoire of relevant bedside tests is briefly described.

Balance Assessment

The goal of the assessment of balance is to quantify imbalance, look for inconsistency, and to separate vestibular patterns of imbalance from other neurologic problems, such as cerebellar ataxia, sensory loss in the feet, movement disorders, and simulated unsteadiness.

GAIT OBSERVATION. The assessment of the balance of a patient with dizziness starts when the patient is met in the waiting room and walked to the examination room. Informal observations should be made concerning how the patients arise from their chair, as well as how they lower themselves into the examination room chair, their speed of locomotion, whether they swing their arms, and whether they have a wide-based gait or use the wall or a caregiver's arm to steady themselves. To screen for a functional disorder, it is helpful to compare informal observation and formal balance testing such as the Romberg test, which is described in the following section. Most patients with acute vestibular problems are unsteady, and most patients with chronic vestibular problems are not unsteady. Inconsistencies should be noted.

EYES-CLOSED TANDEM ROMBERG TEST. The tandem Romberg test, also referred to as *sharpened Romberg*, is quick and useful, albeit also nonspecific and somewhat insensitive.[4] It is a test for sensory ataxia. Borderline normal performance consists of the ability to stand heel-to-toe, with eyes closed, for 6 to 30 seconds. The test can be made easier and thus quantified to some extent by allowing the eyes to be open or by allowing the feet to be in parallel but next to one another (standard Romberg test) rather than in tandem. Variants of the test involve standing on a foam pad, rather than in tandem, or on a narrow rail.[5,6] High-normal performance, defined as the time before a step is required to prevent a fall, is generally found in young adults, who can often perform the eyes-closed tandem variant of the Romberg test for 30 seconds. Performance declines greatly with age, especially in patients in their seventies and older.[6] Many middle-aged patients with chronic inner ear disorders will have no difficulty standing in tandem with their eyes open, but they may need to take a step before 6 seconds passes with eyes closed.

It is helpful to develop a judgment of how much impairment of the Romberg test is appropriate for a given degree of ear injury. Patients with bilateral

KEY POINTS

- The function of all five vestibular sensors in the inner ear, including the otolith organs (saccule and utricle) and all three semicircular canals, can now be tested.
- The assessment of the balance of a patient with dizziness starts when the patient is met in the waiting room and walked to the examination room.

vestibular loss have moderate ataxia; they rely heavily on their vision and are unsteady when their eyes are closed when standing with a narrow base, whether together in parallel or tandem. Most patients with substantial bilateral vestibular loss cannot stand in the eyes-closed tandem Romberg test for 6 seconds. Patients with bilateral vestibular deficits with an additional superimposed position sense deficit, such as peripheral neuropathy, lose balance when standing with a narrowed base even with eyes open. Patients with chronic unilateral vestibular loss are only mildly ataxic, and they usually perform normally on the eyes-closed tandem Romberg test. Patients with acute unilateral vestibular hypofunction with nystagmus may be much more off balance but can adapt in weeks to a few months to show fairly normal balance.

THE FUKUDA STEPPING TEST. The Fukuda stepping test (FIGURE 2-1) and assessment for past-pointing are measurements of vestibulospinal function. They are rarely used in contemporary clinical practice. In the Fukuda stepping test, the patient is asked to march in place with eyes closed for approximately 30 seconds; and the clinician then notes rotation and translation on a calibrated mat.[7] The Fukuda stepping test fell from popularity after it was shown that it has very wide variability in subjects without balance problems. Honaker and Shepard[8] concluded, "Overall, the [Fukuda stepping test] provides little benefit to clinicians when used in the vestibular bedside examination."

The past-pointing test is also called the *Quix test*.[9] During the test, the patient and examiner assume mirror-image postures with outstretched hands so that the fingers almost touch, and the examiner assesses whether the patient's fingers drift after their eyes are closed. The Quix test has not undergone rigorous scrutiny in the literature, and it is rarely used. Practically, much stronger tools are available to detect vestibular imbalance based on nystagmus, ie, Frenzel goggle testing.

FIGURE 2-1
The Fukuda test. Patients march in place for 30 seconds with their eyes closed. The amount of rotation and translation is documented and interpreted from markings on the mat.

Hearing Assessment at the Bedside

Because many vestibular conditions share an underlying pathology with the hearing apparatus, it is prudent to examine hearing and evaluate the status of the external ear and tympanic membrane. High-frequency hearing can be screened quickly with the rubbed-fingers test, during which examiners use their own hearing as a control. Young patients should be able to perceive the finger rubbing at an arm's length, whereas many older patients cannot hear the rubbing sound until it gets as close as 0.3 m (1 ft). Tuning fork tests can also be used for a similar purpose, but formal audiometry is the preferred next step if the patient fails the finger-rub test. Low-frequency hearing loss is not well assessed with the finger-rub test, and patients with Ménière disease, for example, may pass this bedside test. In other words, one cannot avoid ordering

audiometry even if the finger-rub test is normal if Ménière disease is a possible diagnosis.

Otoscopy can determine if the ear canal is occluded by cerumen, if a perforation or scarring of the tympanic membrane has occurred, or if a mass is present as may be seen with a cholesteatoma or glomus tumor.

Finally, some bedside maneuvers are useful in patients with tinnitus, which often accompanies dizziness. Rarely, objective tinnitus deriving from the tensor tympani muscle of the tympanic membrane can be diagnosed by careful observation for intermittent dimpling of the eardrum during otoscopy. In addition, rarely in patients with pulsatile tinnitus, the examiner will be able to hear the high-flow bruit of a dural arteriovenous malformation with a stethoscope. For more information about tinnitus, refer to the article, "Tinnitus, Hyperacusis, Otalgia, and Hearing Loss" by Terry D. Fife, MD, FAAN, FANS, and Roksolyana Tourkevich, MD,[10] in this issue of *Continuum*.

KEY POINTS

- Frenzel goggles are critical to the rapid and efficient evaluation of patients with dizziness because they improve the clinician's ability to detect vestibular nystagmus.
- Vestibular spontaneous nystagmus is suppressed by fixation.

Nystagmus Testing

Evaluation of nystagmus is very useful in a patient with dizziness. Optimally, this requires the use of Frenzel goggles (FIGURE 2-2), which are worn by the patient to reduce fixation, as well as to magnify the examiner's view of the patient's eyes. Frenzel goggles are useful because most inner ear causes of dizziness produce nystagmus that can be suppressed by fixation. To see nystagmus roughly 1 week after onset of an acute vestibular syndrome such as vestibular neuritis, the patient's eyes must not be allowed to fixate when being viewed.

Of the two available variants of Frenzel goggles (optical and infrared video), the infrared video goggles are far superior, but the optical goggles are more affordable. Without a method of viewing the eyes without fixation, some types of nystagmus may not be observable. The ophthalmoscope can be used for making inferences about spontaneous nystagmus if Frenzel goggles are not available; this is discussed in more detail in the following sections.

FIGURE 2-2
Frenzel goggles. *A*, Optical Frenzel goggles, which reduce fixation through the use of +20 diopter lenses over the eyes. *B*, Video Frenzel goggles, which use infrared illumination to prevent fixation; the image of the eye can be made very large.

NYSTAGMUS ASSESSMENT TESTS THAT DO NOT REQUIRE FRENZEL GOGGLES. Some types of nystagmus may be observable even without Frenzel goggles.

SPONTANEOUS NYSTAGMUS. The assessment for spontaneous nystagmus is important for diagnosing conditions characterized by vestibular imbalance, such as vestibular neuritis. It should not be omitted in the assessment of a patient with dizziness. With either

KEY POINTS

- Congenital nystagmus is enhanced by fixation.
- The ophthalmoscope can be used to assess spontaneous nystagmus if Frenzel goggles are not available.
- The Alexander law can be used to assess for spontaneous jerk nystagmus.

Frenzel goggles placed on the patient or using the ophthalmoscope, the eyes are observed for spontaneous nystagmus. FIGURE 2-3 illustrates the two most common types of spontaneous nystagmus. The nystagmus typical of inner ear dysfunction is jerk nystagmus in primary gaze in which the eyes deviate slowly (slow phase) in one direction followed by a rapid correction (fast phase) in the opposite direction. The direction of nystagmus is named for the fast phase (VIDEO 2-1). The more rapidly the eyes deviate off center (the slow phase), the more frequently the corrective jerks (the fast phase) occur, making the nystagmus appear faster. Most nystagmus of other patterns (eg, pendular or saccadic) are of central origin (VIDEO 2-2). For the most part, if a jerk-type nystagmus can be detected without the use of some method of blocking fixation, the patient has either acute dizziness or a disorder that impairs fixation, such as a cerebellar or brainstem problem. These central cases are relatively rare compared with inner ear types of dizziness.

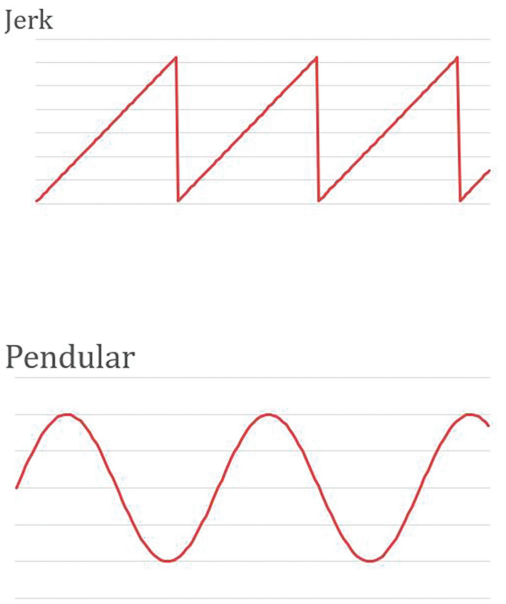

FIGURE 2-3
Illustration of how the eyes move from side to side. By convention, the upward direction on the tracing corresponds to rightward eye movement, while the downward direction on the tracing corresponds to leftward eye movement. Pendular nystagmus is less common but may be seen in some central nervous system causes or congenital nystagmus. The far more common jerk nystagmus (in this case, to the left because it is named after the fast phase) is typical of vestibular nystagmus.

An exception to the preceding general rule is congenital nystagmus. Unlike vestibular nystagmus, congenital nystagmus is often reduced by removal of fixation, and this is one of the ways it can be recognized. The most common congenital nystagmus is latent nystagmus, generally found in people with congenital strabismus (CASE 2-1). Latent nystagmus is a jerk nystagmus that changes direction according to the viewing eye (VIDEO 2-3). Patients with latent nystagmus also have no stereovision and will fail bedside tests for stereopsis, such as the Titmus Fly test.

When Frenzel goggles are not available, the ophthalmoscopic examination can help with obtaining some information about spontaneous nystagmus. The examiner monitors movement of the back of the eye while obscuring vision by covering the other eye. As the back of the eye moves in the direction opposite of the front of the eye, horizontally and vertically, the examiner must be sure to invert the direction of the nystagmus when making notes. If an ophthalmoscope is also not available, the presence of a unidirectional nystagmus can sometimes be inferred from an evaluation of the effect of gaze nystagmus. According to the Alexander law, vestibular nystagmus nearly always increases when the patient looks in the direction of the fast phase, so an asymmetric frequency or intensity of nystagmus

will be seen depending on whether the patient looks to the right or left (although the nystagmus does not change direction) in peripheral vestibular nystagmus.[11]

DIX-HALLPIKE TEST. Ideally, the Dix-Hallpike test is performed with Frenzel goggles, optical or video (FIGURE 2-4), although in many cases, the nystagmus of benign paroxysmal positional vertigo (BPPV) is seen readily with the naked eye. The sensitivity of the test improves when performed with the goggles. For the Dix-Hallpike test to the right (VIDEO 2-4), the patient's head is turned 45 degrees to the right and then the patient is moved quickly from the sitting position to this head-hanging position. If the patient feels no dizziness or if nystagmus is not appreciated after 15 seconds, then the patient is moved back up to the sitting position. For the Dix-Hallpike test to the left (VIDEO 2-5), the patient's head is turned 45 degrees to the left and then the patient is quickly laid back to the head-hanging left position. After 15 seconds, the patient is again moved to a sitting position. This may be repeated if BPPV is still suspected based on history. In a positive test, a burst of nystagmus is provoked by either the head-right or the head-left position. Further information on BPPV is described in the article "Episodic Positional Dizziness" by Kevin A. Kerber, MD, MS,[12] in this issue of *Continuum*.

CASE 2-1

A 50-year-old man was born with congenital esotropia; as a child, he frequently squinted and eventually was able to describe that he was experiencing double vision, so at age 5 years he underwent surgery to correct the esotropia.

Videonystagmography recordings (with the camera placed over the right eye while the patient had initially viewed out of the left eye) documented a spontaneous left-beating nystagmus; however, caloric tests were normal. Because he had spontaneous nystagmus but no caloric weakness, the audiologist concluded that the patient had a central vestibular disorder.

In the office, the patient had no depth perception (stereopsis) as determined by the Titmus Fly test in which the patient wore polarized glasses while looking at a specially formatted picture of a housefly that should appear to pop out from the page if depth perception is intact. The patient also had amblyopia in the right eye and weak left-beating horizontal nystagmus in the light. When the left eye was covered, the nystagmus reversed direction and became right beating. The nystagmus stopped in complete darkness (with the use of video Frenzel goggles). Close inspection of the videonystagmography pursuit traces showed that the eye actually tracked faster than the target to the left, and backup saccades were present.

COMMENT

This case illustrates some of the examination findings in a patient with a type of congenital nystagmus called *latent nystagmus*, which often results from congenital esotropia, and points out how some of the findings taken in isolation can lead to an incorrect diagnosis.

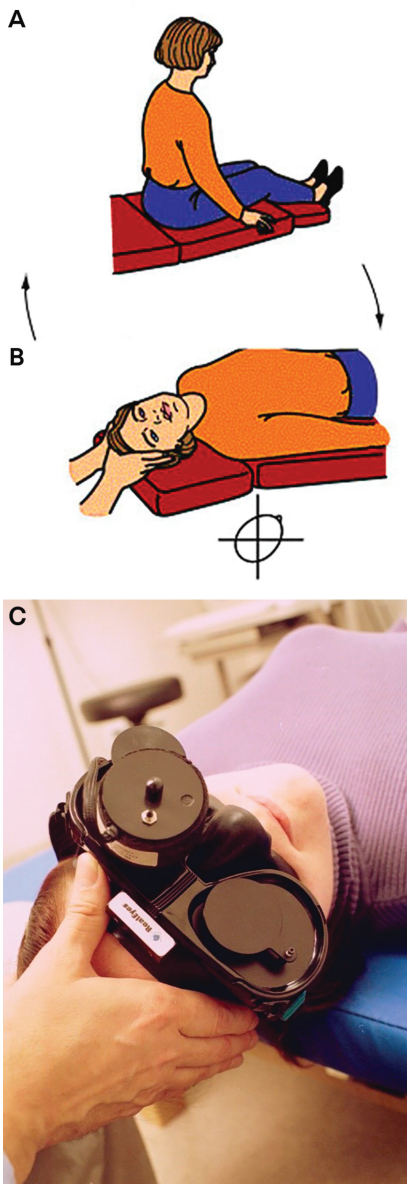

FIGURE 2-4
Dix-Hallpike positional maneuver. To precipitate the characteristic nystagmus of benign paroxysmal positional vertigo, the patient is rapidly brought from sitting (*A*) into a head-hanging position (*B* and *C*) that makes the posterior canal vertical and brings it through a large angular displacement.

The most common type of positional nystagmus, posterior canal BPPV, beats upward and has a rotatory component, such that the top part of the eye beats toward the undermost ear (VIDEO 2-4 and VIDEO 2-5). The nystagmus typically has a latency of 2 to 5 seconds and lasts 5 to 60 seconds, and an unwinding downbeat/rotatory nystagmus may be seen when the patient is sat up again (VIDEO 2-6). Variations of BPPV have different vectors. The lateral canal variant of BPPV is typified by a strong horizontal nystagmus that reverses direction between head left and right (VIDEO 2-7 and VIDEO 2-8). The rare anterior canal variant is associated with a downbeating nystagmus elicited by the Dix-Hallpike test. The Dix-Hallpike test is a high-yield portion of the dizziness bedside assessment and should almost never be omitted. For more information on positional vertigo, refer to the article, "Episodic Positional Dizziness" by Kevin A. Kerber, MD, MS,[12] in this issue of *Continuum*.

NYSTAGMUS ASSESSMENT TESTS THAT REQUIRE FRENZEL GOGGLES. In general, the following additional maneuvers should not be used if Frenzel goggles are not available. More detail about these maneuvers can be found elsewhere.[13]

NECK-VIBRATION TEST. The neck-vibration test (FIGURE 2-5) is very useful as a bedside test because it is a robust and durable test for unilateral vestibular weakness.[14] In the neck-vibration test, the patient's eyes are observed in complete darkness (ie, with video, not optical, Frenzel goggles) while vibration (typically from a massaging device) is applied to the sternocleidomastoid muscle for 10 seconds. The vibration is applied first on one side and then on the other (VIDEO 2-9). One looks for direction-fixed nystagmus (that is, it does not change direction with changes in the direction of gaze) with fast-phase beating to the side opposite the ear with vestibular hypofunction. The neck-vibration test requires less expertise than the head impulse test (see the Head Impulse Test section), requires almost no subjective judgment, and is not greatly affected by

FIGURE 2-5
Neck-vibration test. While the examiner applies vibration to the sternocleidomastoid muscle, video Frenzel goggles are used to detect nystagmus. A positive test elicits strong unidirectional horizontal nystagmus from both sides.

the passage of time since the onset of the lesion. Provided that video Frenzel goggle equipment is available, the neck-vibration test can be helpful in patients with suspected unilateral vestibular hypofunction.

HEAD-SHAKE TEST. With the patient wearing Frenzel goggles and sitting, the patient's head is rotated by the examiner in the horizontal plane for 20 cycles. The examiner should aim for a 30- to 45-degree turn of the head to either side (if the patient is safely able) and a frequency of 2 cycles per second. A nystagmus that lasts 5 seconds or longer indicates an organic disorder of the ear or central nervous system and indicates further investigation is warranted. The head-shake test is neither as reliable nor as durable as the neck-vibration test. False positives are common.

VALSALVA TEST. The Valsalva test is optional and mainly performed if a pressure or sound sensitivity symptom is elicited from the history. While wearing the Frenzel goggles, the patient inhales a deep breath and strains for 10 seconds while the examiner observes for nystagmus. Nystagmus at the onset and release of pressure indicates a positive test. The glottis can be open or closed. The Valsalva test is used mainly to assess for superior canal dehiscence. Because far more sensitive laboratory tests for superior canal dehiscence (ie, vestibular-evoked myogenic potential) are available, performing this test at the bedside has little benefit. It is also possible, in a small subset of patients with superior canal dehiscence, to elicit nystagmus in response to sound. This is called the Tullio test, but it is also highly insensitive and rarely performed.

HYPERVENTILATION TEST. The hyperventilation test is also optional. It is mainly performed if the examination has been entirely normal or if a vestibular schwannoma or other partial lesion of the vestibular nerve is suspected. The patient takes 30 deep, forceful breaths. Immediately after hyperventilation and with the use of the Frenzel goggles, the patient's eyes are inspected for nystagmus. Hyperventilation-induced nystagmus suggests a partially conducting eighth nerve or central vestibular pathways caused by, for example, a tumor of the eighth cranial nerve, gamma knife radiosurgery for vestibular schwannoma, or multiple sclerosis. The hyperventilation test is both insensitive and nonspecific.

VESTIBULO-OCULAR REFLEX GAIN TESTING

The vestibulo-ocular reflex mediates eye movements in response to head rotation, such that eyes move equally in amplitude but in the direction opposite

KEY POINT

- **The neck-vibration test is a sensitive and durable test of unilateral vestibular weakness.**

of the head rotation, which serves to stabilize the image of a viewed target on the fovea during movement.

Head Impulse Test

The head impulse test is a bedside vestibular test, first described in 1988.[15] It is useful for detecting vestibular damage and most helpful in documenting vestibular neuritis. The test requires no special equipment. Standing directly in front of the patient, the examiner holds the patient's head firmly on each side and instructs the patient to look at a fixed point, usually the examiner's nose. The examiner then abruptly rotates the patient's head rapidly but only a small distance to the left and right (approximately 10 degrees); this brisk rotation (the impulse) should be in a pattern that is unpredictable in the timing and direction of the head turn; several impulses toward each side should be assessed. In a person with an intact vestibular system, the vestibulo-ocular reflex will keep the eyes on target and the patient will still be looking at the examiner's nose after the impulse. In a patient with a recent unilateral vestibular deficit, especially involving the vestibular nerve, the eyes move with the head (due to an impaired vestibulo-ocular reflex on the side to which the head was turned), and this is followed by a corrective rapid eye movement to bring the eyes back to the target (the examiner's nose) (CASE 2-2). The head impulse test is most useful when video Frenzel goggles are not available. The next two maneuvers are aimed at documenting bilateral vestibular loss.

Dynamic Illegible E Test

This is a test of dynamic visual acuity. Using an eye chart positioned at least 3 meters (10 feet) from the patient, preferably calibrated in LogMAR units (FIGURE 2-6), the examiner records visual acuity while the patient's head is still. Then, the examiner gently turns the patient's head 30 degrees to the left and right at approximately 1 to 2 Hz and again records the visual acuity. Patients with no loss of vestibular function drop from 0 to 2 lines per LogMAR of acuity with turning of the head. Patients with complete or partial bilateral loss of vestibular

CASE 2-2

A 40-year-old man reported acute dizziness and unsteady gait and had been vomiting for several hours. Although he was ambulatory, he was unsteady and preferred to hold onto his wife's shoulder as he was taken from the waiting room to the examination room.

On examination, he could not stand with his eyes closed in a tandem Romberg stance. He had a left-beating spontaneous nystagmus readily seen in both eyes. The nystagmus increased on left gaze and stopped on right gaze. His head impulse test was positive to the right and normal to the left.

COMMENT

This is the presentation of acute right-sided vestibular neuritis. This case illustrates how a unilateral vestibular weakness presents with spontaneous unidirectional horizontal nystagmus that obeys the Alexander law.

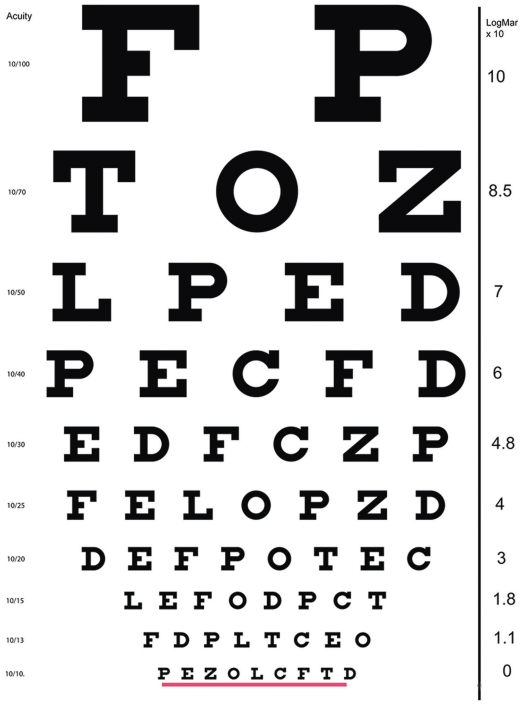

FIGURE 2-6

Dynamic illegible E test. This test of dynamic visual acuity is performed with the examiner oscillating the patient's head side to side about the vertical axis at approximately 1 to 2 Hz; the lowest line that can be read is ascertained. A decline in visual acuity before and during movement of at least three lines is abnormal, indicating bilateral vestibulopathy.

function drop from 3 to 7 lines per LogMAR of acuity. Patients with acute complete bilateral loss generally can read only the top line (ie, the F and P).

Ophthalmoscope Test

The ophthalmoscope test is used to obtain objective corroboration when a patient has a positive dynamic illegible E test. The examiner focuses on the optic disk and then gently moves the patient's head as described for the illegible E test. The vestibulo-ocular reflex gain is abnormal if the disk moves with the head, ie, moves back and forth from the examiner's perspective.[16] The ophthalmoscope test is less sensitive than the dynamic illegible E test and should be performed with the patient's spectacles on to avoid interaction with the effects of spectacles on vestibulo-ocular reflex gain.

Hearing Testing

TABLE 2-1 lists the indications for common laboratory procedures used for evaluating hearing in patients with vertigo and dizziness. Not all of these tests are useful for every patient. To be time- and cost-efficient, tests should be chosen according to each patient's specific set of symptoms (TABLE 2-2). For more information on hearing loss, refer to

Hearing-Related Laboratory Testing for Dizziness and Vertigo TABLE 2-1

Test	Indications
Audiogram	Hearing symptoms, dizziness, vertigo, Ménière disease suspected, superior canal dehiscence suspected
Otoacoustic emissions	Hearing symptoms, functional hearing loss
Brainstem auditory evoked response	Suspicion of vestibular schwannoma with no access to MRI
Electrocochleography	Secondary test for Ménière disease

MRI = magnetic resonance imaging.

the article, "Tinnitus, Hyperacusis, Otalgia, and Hearing Loss" by Terry D. Fife, MD, FAAN, FANS, and Roksolyana Tourkevich, MD,[10] in this issue of *Continuum*.

When hearing symptoms are present or when a disorder such as Ménière disease or vestibular schwannoma is reasonably suspected, then an audiogram is the most useful initial test. An audiogram is recommended even for patients who have few symptoms of hearing loss because some patterns of hearing loss cannot be determined at the bedside (eg, low-frequency hearing loss) and may not be readily noticed by the patient. TABLE 2-2 outlines four common hearing patterns that may be documented on audiometry. FIGURE 2-7 shows the typical low-tone hearing loss seen in early Ménière disease.

It is incumbent for the clinician seeing patients with dizziness to recognize each of the abnormal patterns above because they should trigger important actions on the part of the clinician.

AUDIOGRAM. The audiogram is a subjective test (requiring cooperation from the patient) that measures hearing and mainly tests the cochlea. Certain abnormalities suggest otologic vertigo (TABLE 2-2). It is nearly always indicated in patients with dizziness. Hearing declines symmetrically in both ears with age, mainly at high pitches. In some cases, it is helpful to combine the audiogram (a subjective test) with the otoacoustic emissions test (an objective test) to look for inconsistency when factitious hearing loss is a concern.

OTOACOUSTIC EMISSIONS. Otoacoustic emissions testing is an objective test based on registration of sound elicited from the inner ear itself in response to an external sound. Otoacoustic emissions are a quick and simple automated procedure. In newborns or others who cannot cooperate with formal audiometry, otoacoustic emissions are valuable because, when present, they show that cochlear function is present. Otoacoustic emissions are usually not helpful in people older than 60 years old because otoacoustic emissions are reduced with age. In adults younger than 60 years, otoacoustic emissions as objective tests are useful in detecting functional hearing loss through the inconsistency between otoacoustic emissions and audiometry. For example, if a patient claims to be deaf on one side but has a robust otoacoustic emission in that ear, functional hearing loss is a reasonable possibility. Otoacoustic emissions can also be normal in central hearing deficits, such as a brainstem or cortical site of hearing loss, but these cases are very rare. Regarding functional hearing loss, a large assortment of audiologic procedures can detect psychogenic hearing loss, including the excellent Stenger test. The Stenger effect refers to the psychoacoustic phenomenon in which a tone, presented simultaneously to both ears but with a greater intensity in one ear, will only be perceived in the ear receiving the louder stimulus; the Stenger effect is leveraged by the Stenger test to detect functional unilateral hearing loss.

TABLE 2-2 **Common Audiometric Findings in Selected Disorders Causing Dizziness**

- Ménière disease: unilateral low-frequency sensorineural loss at the onset of disease
- Vestibular schwannoma: unilateral progressive high-frequency sensorineural loss
- Superior canal dehiscence (conductive hyperacusis): bone conduction better than air conduction at 500 Hz

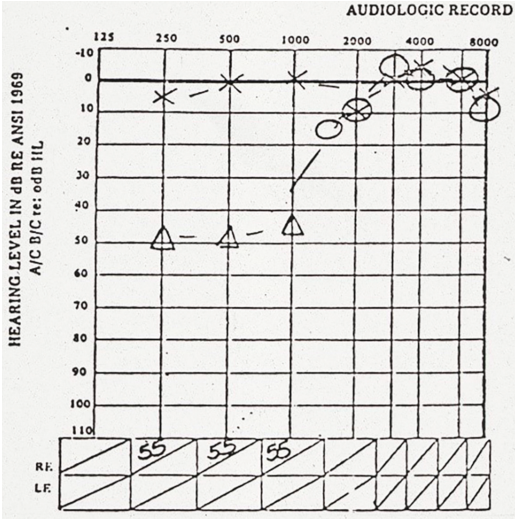

FIGURE 2-7
An audiogram showing right-sided low-frequency sensorineural hearing loss. Low-frequency sensorineural hearing loss on the right side is depicted, which is fairly specific for right-sided Ménière disease. Here, *X* depicts hearing thresholds for the left ear, and *O* and the *triangle* indicate thresholds for the right ear.

BRAINSTEM AUDITORY EVOKED RESPONSE TESTING. Brainstem auditory evoked response, also referred to as *auditory brainstem responses* among audiologists, is an evoked potential test measuring brainstem responses to sound. In patients with dizziness, it is most useful to detect vestibular schwannomas. Brainstem auditory evoked response testing has fallen out of favor because MRI testing has far superior diagnostic sensitivity and specificity.

ELECTROCOCHLEOGRAPHY. Electrocochleography is an evoked potential test in which the recording electrode is positioned on the eardrum to get a better definition of the cochlear potential from the inner ear. An abnormal electrocochleogram may suggest Ménière disease in patients with a clinical history that is consistent with Ménière disease. Electrocochleography is technically difficult and relatively unrewarding in diagnostic power and should not be considered a useful screening test in all patients with vertigo. For these reasons, electrocochleography has fallen out of favor and is not widely available.

KEY POINTS

- **Hearing testing is critical to assess for Ménière disease and contributes greatly to the diagnosis of a vestibular schwannoma.**
- **The combination of a subjective hearing test such as the audiogram with an objective test such as otoacoustic emissions can help with the diagnosis of functional hearing loss.**

Vestibular Laboratory Testing

Vestibular laboratory testing can now assess all five sensory organs in the inner ear. Three tests—videonystagmography (VENG), rotary chair, and video head impulse—assess the semicircular canals. VENG and rotary chair test only horizontal canal function, whereas the video head impulse test can test all three semicircular canals on each side. These tests are mainly helpful in patients with dizziness when no clear diagnosis is evident after history and bedside examination (TABLE 2-3). Two vestibular tests, cervical vestibular-evoked myogenic potential and ocular vestibular-evoked myogenic potential, assess the otolith organs (saccule and utricle, respectively). Vestibular tests are sometimes unreliable and, to confirm abnormal results, it is often helpful to obtain several independent measurements, usually entailing two different techniques (ie, video head impulse test and VENG or rotary chair). One can also use a combination of a good bedside examination (with video Frenzel goggles) and one or more laboratory tests to increase reliability of the diagnostic process. More detail about the older vestibular tests (ie, VENG and rotary chair) can be found in an American Academy of Neurology (AAN) assessment of vestibular testing.[17]

VIDEONYSTAGMOGRAPHY. VENG is a battery of eye-movement recordings that can identify vestibular asymmetry, such as that caused by vestibular neuritis, and document spontaneous or positional nystagmus, such as that caused by BPPV.

VENG includes a caloric test, wherein the vestibular system is stimulated by warming or cooling the eardrum with water or air. VENG is a long and difficult test, both for the patient and the technician, and patients sometimes call it the "water torture test" because part of the test may induce vertigo and, in some people, motion sickness.

Laboratories sometimes attempt to avoid the strong dizziness that can be encountered in the caloric test by replacing water irrigation with air, resulting in less stimulating and messy procedures but lower-quality data. False-positive interpretations, especially "bilateral vestibular weakness," are common if the person performing the air caloric testing does not point the stream of warm or cool air directly at the eardrum (CASE 2-3). The interpretation of "central vestibular disturbance," is sometimes applied to patients with a VENG finding outside of the usual distribution found in inner ear disorders. For example, patients with weak but measurable spontaneous upbeating or downbeating nystagmus may be given the diagnosis of central vestibular disturbance and referred for neurologic evaluation. Another source of testing error can be a partial cerumen impaction of the ear being tested, which results in the false-positive diagnosis of unilateral vestibular weakness. VENG results are usually interpreted by audiologists, whose training mainly covers inner ear disorders; consequently, some audiologists characterize as a "central finding" any results they do not recognize deriving from an inner ear disturbance, leading to a neurology referral.

As is the case with other vestibular tests, an abnormal result that does not fit the clinical picture should be supplemented by rotary chair testing or video head impulse testing, ideally, in combination with vestibular-evoked myogenic potential testing (see the following section). VENG remains the vestibular test of choice but can be associated with some discomfort, including nausea and/or headache, especially in patients with motion sickness or migraine. Similar clinical information can be gained by combining the video head impulse test and bedside nystagmus testing with video Frenzel goggles. It should be noted that, currently, Centers for Medicare & Medicaid Services and many other insurance plans do not cover video head impulse test procedures (TABLE 2-4).

VESTIBULAR-EVOKED MYOGENIC POTENTIAL. Vestibular-evoked myogenic potential testing measures the function of the otolith organs (the utricle and saccule) and

TABLE 2-3 **Vestibular Laboratory Tests (in Order of Usefulness)**

Test	Indication(s)
Videonystagmography (VENG)	Vertigo
Video head impulse test (vHIT)	Vertigo
Vestibular-evoked myogenic potential (cervical [cVEMP] and ocular [oVEMP])	Vertigo, sound sensitivity, pulsatile tinnitus
Rotary chair test (Rchair)	Bilateral vestibular loss suspected, secondary test to confirm abnormal caloric responses or video head impulse test suggesting unilateral or bilateral vestibular loss
Posturography (CDP)	Assess for functional disorder, assess fall risk

was mentioned briefly in the 2000 AAN vestibular assessment,[17] but now it is mainstream and fairly widely available. Vestibular-evoked myogenic potential testing is sensitive to the superior semicircular canal dehiscence syndrome[18] and has a minor role in supporting the diagnosis of bilateral vestibular loss, vestibular neuritis, and conductive hearing loss.

The two variants of the vestibular-evoked myogenic potential test most commonly used in clinical practice are the ocular vestibular-evoked myogenic potential and cervical vestibular-evoked myogenic potential. The ocular vestibular-evoked myogenic potential quantifies utricular function, whereas the cervical vestibular-evoked myogenic potential quantifies saccular function. Vestibular-evoked myogenic potential testing is sensitive to the superior semicircular canal dehiscence syndrome. Either the ocular or cervical test may show larger than normal responses and lower than normal sound thresholds on the affected side(s). While thin-slice CT scans of the temporal bone images document dehiscence, this procedure both exposes the patient to radiation as well as may be falsely positive, as asymptomatic dehiscence is found in about 1% of the population.[19] The combination of a visible dehiscence (abnormal opening of the bony canal) on CT of the temporal bone with abnormal vestibular-evoked myogenic potential responses (either cervical or ocular) strongly points to the diagnosis of superior semicircular canal dehiscence syndrome. For more information on superior canal dehiscence syndrome, refer to the article, "Selected Otologic Disorders Causing Dizziness" by Gail Ishiyama, MD,[20] in this issue of *Continuum*.

CASE 2-3

A 70-year-old woman underwent videonystagmography for evaluation of chronic dizziness. The air caloric testing technique was used. Warm air caloric stimulation was applied in each ear sequentially, then cool air caloric stimulation was applied in each ear sequentially; the total response (sum of the peak slow-phase velocity responses from all four caloric stimuli, warm and cool in each ear) was only 8 degrees per second whereas normal is between 20 and 100 degrees per second. The patient was referred for a neurologic evaluation for potential bilateral vestibular loss.

On examination, she could stand in the tandem Romberg stance with her eyes closed and had no loss of visual acuity on the dynamic illegible E test. Her ophthalmoscope and head impulse tests were normal.

COMMENT

This case illustrates a discrepancy between clinical examination and an inadequate vestibular test. On physical examination, the facts that the patient could maintain a tandem Romberg stance with her eyes closed and she had normal performance on dynamic illegible E testing are incompatible with a diagnosis of bilateral vestibular loss. In contrast, air caloric testing is very dependent on pointing the column of air directly at the tympanic membrane to properly evoke good caloric vestibular responses. The conclusion of "bilateral vestibular loss" was a false-positive result.

Vestibular-evoked myogenic potential responses, like balance, normally decline with advancing age but generally are repeatable and stable. Cervical vestibular-evoked myogenic potentials (whose pathway travels through the inferior division of the vestibular nerve) are often normal in patients with vestibular neuritis (which more commonly affects the superior division of the vestibular nerve), and thus can be helpful in making this diagnosis.[21] Vestibular-evoked myogenic potential testing now has a *Current Procedural Terminology* (*CPT*) code (TABLE 2-4).

ROTARY CHAIR. Rotary chair testing measures vestibular function of both inner ears together. It is sensitive to bilateral loss of vestibular function and performs better than VENG for this purpose.[17] In unilateral vestibular loss, rotary chair testing is sensitive but nonspecific because it is poor at identifying the side of the lesion.

TABLE 2-4 Common Vestibular and Auditory Tests and Their Corresponding *CPT* Codes[a]

Test	*CPT* code
Comprehensive audiometry threshold evaluation and speech recognition (92553 and 92556 combined)	92557
Auditory evoked potentials; screening of auditory potential with broadband stimuli, automated analysis neurodiagnostic, with interpretation and report	92635
Caloric vestibular test with recording, bilateral; bithermal (ie, one warm and one cool irrigation in each ear for a total of four irrigations)	92537
Electrocochleography	92584
Distortion product evoked otoacoustic emissions; limited evaluation (to confirm the presence or absence of hearing disorder, 3-6 frequencies) or transient evoked otoacoustic emissions, with interpretation and report	92587
Distortion product evoked otoacoustic emissions; comprehensive diagnostic evaluation (quantitative analysis of outer hair cell function by cochlear mapping, minimum of 12 frequencies), with interpretation and report	92588
Computerized dynamic posturography sensory organization test (CDP-SOT), 6 conditions (ie, eyes open, eyes closed, visual sway, platform sway, eyes closed platform sway, platform and visual sway), including interpretation and report	92548
Vestibular-evoked myogenic potential (VEMP) testing, with interpretation and report; cervical (cVEMP)	92517
Vestibular-evoked myogenic potential (VEMP) testing, with interpretation and report; ocular (oVEMP)	92518
Vestibular-evoked myogenic potential (VEMP) testing, with interpretation and report; cervical (cVEMP) and ocular (oVEMP)	92519
Video head impulse test	None; bill as 92700 and have patient sign an advanced beneficiary notice
Sinusoidal vertical axis rotational testing	92546

[a] *Current Procedural Terminology (CPT)* codes maintained by the American Medical Association and approved by the Centers for Medicare and Medicaid Services in the United States as of January 1, 2021.

Rotary chair testing is most useful when caloric vestibular testing done as part of VENG falsely suggests that vestibular responses are reduced or absent on both sides because of small ear canals, cerumen in the external canal, or inadequate irrigation technique. Rotary chair testing can clarify matters because it does not determine vestibulo-ocular reflex function by caloric stimulation via the external ear canal but rather by actual movement of the head. Rotary chair testing is almost always covered by health insurance, but it requires costly equipment. In addition, rotary chair testing requires considerable technician and patient time to perform the test.

KEY POINT

- False-positive videonystagmography findings of bilateral vestibular weakness or central vestibular disturbance are common sources of referrals to neurologists.

VIDEO HEAD IMPULSE TEST. This recently available vestibular test can quickly diagnose both severe bilateral vestibular loss and complete unilateral vestibular loss, especially when due to vestibular nerve injury. The video head impulse test is less sensitive to vestibular damage due to hair cell disease, such as Ménière disease, whereas VENG testing is more accurate. The video head impulse test is more resistant to false-positive findings in patients with functional symptoms than tests of nystagmus such as VENG and the rotary chair.

Variants of the video head impulse test purport to measure function of the posterior and anterior semicircular canals, but they should not be relied on for this because current commercially available eye-movement monitoring technology is not able to quantify the entire three-dimensional vector of eye movement. As a consequence, the results of video head impulse tests done for the anterior or posterior canal planes are often puzzling. Despite the high utility of video head impulse testing, many insurances do not currently cover it.

POSTUROGRAPHY. This discussion specifically refers to the computerized dynamic posturography device. Posturography attempts to evaluate vestibular, proprioceptive, and visual contributions to balance and is similar to the Romberg test. It requires cooperation from the patient. Posturography involves a series of six testing conditions, progressing through permutations of normal/absent/misleading visual input and normal/misleading proprioceptive input. The series of conditions become gradually more difficult, and a patient's performance should decline with progressively more difficult conditions. However, if the conditions are presented in a random order, and if the patient performs normally on difficult conditions but poorly in easy conditions, then such inconsistency[22] may raise suspicion for a functional disorder. Posturography is also a method of assessing fall risk and is usually reimbursed by Medicare insurance (TABLE 2-4), but often it is not covered by other types of insurance.

SUMMARY OF THE ROLE OF VESTIBULAR TESTING. In patients with dizziness, vestibular testing assists in the diagnostic process by supplementing the clinical history and examination. Vestibular tests can usually identify unilateral vestibular loss, bilateral vestibular loss, and superior canal dehiscence. When combined with hearing testing, they can assist in the identification of Ménière disease. When comprehensive vestibular testing is all normal, it goes a long way toward excluding primary peripheral vestibular disorders that leave persisting dysfunction. Most other conditions, including vestibular migraine, dizziness related to anxiety, dizziness from cardiovascular disturbances (eg, orthostatic hypotension), and conditions such as persistent postural perceptual dizziness have normal results on vestibular assessments at the bedside and in the laboratory. In these cases, vestibular tests may assist the clinician in avoiding

fruitless management strategies, such as vestibular-suppressant medication or vestibular rehabilitation for patients who have no vestibular disturbance.

CODING OF VESTIBULAR TESTING. Bedside testing (without instrumentation and recording), such as the Dix-Hallpike test for BPPV or the bedside head impulse test for unilateral and bilateral vestibular loss, has no billing code that is reimbursed in the United States. Instrumented testing, often involving the same basic process as bedside testing, but with a recording, is usually reimbursed by insurance (TABLE 2-4).

CONCLUSION

For a clinician to diagnose a patient with dizziness, a careful history and an examination that includes specific bedside vestibular tests are crucial. In some instances, this may need to be supplemented with audiometric tests and with instrumented vestibular testing, the latter of which has seen significant advances in the past decade as it is now possible to evaluate the entire labyrinth (all semicircular canals and otolith organs).

VIDEO LEGENDS

VIDEO 2-1
Weak jerk nystagmus in a patient with resolving vestibular neuritis. The eyes are first in primary position of gaze, and modest spontaneous left-beat nystagmus is present. When the patient directs gaze rightward, no nystagmus is present. When the patient directs gaze leftward, left-beat nystagmus is more pronounced than it had been on primary position of gaze. This pattern is in accordance with the Alexander law for a right-sided vestibular weakness.

VIDEO 2-2
Vertical pendular nystagmus in a patient with pontine bleed and palatal myoclonus (*not shown*).

VIDEO 2-3
Latent nystagmus, a variety of congenital nystagmus. At first, the camera is over the right eye, the patient is viewing out of the left eye, and left-beat nystagmus is present. Then, the camera is switched from the right eye to the left eye (noted as *Switch Cover* in the video), and the patient is viewing out of the right eye, and right-beat nystagmus is present. The fast phase of the nystagmus is ipsiversive to the eye out of which the patient is viewing. This pattern is characteristic of latent nystagmus.

VIDEO 2-4
Dix-Hallpike test and right benign paroxysmal positional vertigo from the posterior canal.

VIDEO 2-5
Dix-Hallpike test and nystagmus from left benign paroxysmal positional vertigo from the posterior canal.

VIDEO 2-6
Left-posterior canal benign paroxysmal positional vertigo (BPPV). The patient is in the left Dix-Hallpike position, and upbeat and left-torsional nystagmus is present. This pattern is consistent with left-posterior canal BPPV. The patient then sits up and is put in the right Dix-Hallpike position, and downbeat and right-torsional nystagmus is present. This pattern is compatible with "unwinding" of left-posterior canal BPPV.

VIDEO 2-7
Geotropic direction-changing positional nystagmus. When the patient is first in the left Dix-Hallpike position, left-beat nystagmus is present. When the patient is moved to the right Dix-Hallpike position, right-beat nystagmus is present. This pattern is geotropic direction-changing positional nystagmus.

VIDEO 2-8
Apogeotropic direction-changing positional nystagmus. When the patient is first in the left Dix-Hallpike position, right-beat nystagmus is present. When the patient is moved to the right Dix-Hallpike position, left-beat nystagmus is present. This pattern is apogeotropic direction-changing positional nystagmus.

VIDEO 2-9
Vibration-induced nystagmus in a patient with a vestibular schwannoma that was removed 40 years ago. Vibration is applied on the right mastoid process and then on the left mastoid process. Vibration on either side elicits left-beat nystagmus, consisting of rightward drift (slow phase of nystagmus toward the side of vestibular weakness) and leftward saccade (fast phase of nystagmus away from the side of vestibular weakness).

REFERENCES

1 Baloh RW. Robert Bárány and the controversy surrounding his discovery of the caloric reaction. Neurology 2002;58(7):1094-1099. doi:10.1212/wnl.58.7.1094

2 Hain T. Approach to the patient with dizziness and vertigo. In: Biller J, editor. Practical neurology. 4th ed. Philadelphia, PA: Wolters Kluwer, Lippincott, Williams & Wilkins 2017.

3 Fife TD. Approach to the history and evaluation of vertigo and dizziness. Continuum (Minneap Minn) 2021;27(2, Neuro-otology):306-329.

4 Longridge NS, Mallinson AI. Clinical Romberg testing does not detect vestibular disease. Otol Neurotol 2010;31(5):803-806. doi:10.1097/MAO.0b013e3181e3deb2

5 Graybiel A, Fregly AR. A new quantitative ataxia test battery. Acta Otolaryngol 1966;61(4):292-312.

6 Agrawal Y, Carey JP, Hoffman HJ, et al. The modified Romberg balance test: normative data in US adults. Otol Neurotol 2011;32(8):1309-1311. doi:10.1097/MAO.0b013e31822e5bee

7 Fukuda T. The stepping test: two phases of the labyrinthine reflex. Acta Otolaryngol 1959;50(2):95-108. doi:10.3109/00016485909129172

8 Honaker JA, Shepard NT. Performance of Fukuda stepping test as a function of the severity of caloric weakness in chronic dizzy patients. J Am Acad Audiol 2012;23(8):616-622. doi:10.3766/jaaa.23.8.6

9 Hart CW. The Quix test. Laryngoscope 1983;93(9):1160-1161. doi:10.1288/00005537-198309000-00010

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

11 Robinson DA, Zee DS, Hain TC, et al. Alexander's law: its behavior and origin in the human vestibulo-ocular reflex. Ann Neurol 1984;16(6):714-722. doi:10.1002/ana.410160614

12 Kerber KA. Episodic positional dizziness. Continuum (Minneap Minn) 2021;27(2, Neuro-otology):348-368.

13 Cherchi M, Hain T. Provocative maneuvers for vestibular disorders. In Eggers S, Zee DS, eds. Vertigo and imbalance: clinical neurophysiology of the vestibular system. Amsterdam, Netherlands: Elsevier, 2010.

14 Dumas G, Perrin P, Ouedraogo E, Schmerber S. How to perform the skull vibration-induced nystagmus test (SVINT). Eur Ann Otorhinolaryngol Head Neck Dis 2016;133(5):343-348. doi:10.1016/j.anorl.2016.04.002

15 Halmagyi GM, Curthoys IS. A clinical sign of canal paresis. Arch Neurol 1988;45(7):737-739. doi:10.1001/archneur.1988.00520310043015

16 Kheradmand A, Zee DS. The bedside examination of the vestibulo-ocular reflex (VOR): an update. Rev Neurol (Paris) 2012;168(10):710-719. doi:10.1016/j.neurol.2012.07.011

17 Fife TD, Tusa RJ, Furman JM, et al. Assessment: vestibular testing techniques in adults and children: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology 2000;55(10):1431-1441. doi:10.1212/wnl.55.10.1431

18 Fife TD, Colebatch JG, Kerber KA, et al. Practice guideline: cervical and ocular vestibular evoked myogenic potential testing: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. Neurology 2017;89(22):2288-2296. doi:10.1212/WNL.0000000000004690

19 Klopp-Dutote N, Kolski C, Biet A, et al. A radiologic and anatomic study of the superior semicircular canal. Eur Ann Otorhinolaryngol Head Neck Dis 2016;133(2):91-94. doi:10.1016/j.anorl.2015.11.001

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

21 Oh SY, Kim JS, Yang TH, et al. Cervical and ocular vestibular-evoked myogenic potentials in vestibular neuritis: comparison between air- and bone-conducted stimulation. J Neurol 2013;260(8):2102-2109. doi:10.1007/s00415-013-6953-8

22 Cevette MJ, Puetz B, Marion MS, et al. Aphysiologic performance on dynamic posturography. Otolaryngol Head Neck Surg 1995;112(6):676-688. doi:10.1016/s0194-5998(95)70175-3