Vertigo: Some Uncommon Causes of a Common Problem

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ABSTRACT

Vertigo, an extremely common symptom, may be caused by numerous disorders affecting the central or peripheral vestibular systems. Patients can usually be categorized into four groups based on the clinical presentation: monophasic, prolonged episodes of vertigo due to acute unilateral vestibular hypofunction; recurrent episodes of vertigo, due to transient vestibular dysfunction; vertigo provoked by changes in head position with respect to gravity (positional vertigo); and bilateral vestibulopathies, which present with imbalance and oscillopsia. Although each clinical syndrome is usually caused by a limited number of disorders, many less common entities must be considered in the differential diagnosis. This article reviews the clinical presentation of the less common causes of vestibular syndromes and discusses their medical and nonmedical management.

Keywords: vertigo; vestibular dysfunction; positional vertigo; oscillopsia

Vertigo may result from numerous pathologic processes involving the inner ear, vestibulocochlear nerve, or brain. This article will review some less common causes of vertigo as well as unusual presentations of more common disorders (Table 1), but will not attempt to be all-inclusive. Before considering the individual disorders, I will briefly review the pathophysiology that underlies the symptom of vertigo and the symptoms and signs that should be evaluated in all patients presenting with vertigo.

PATHOPHYSIOLOGY

The vestibular labyrinth in the inner ear responds to head motion, both rotatory (transduced by the three semicircular canals) and linear (transduced by the otolith organs). When the head is stationary, the afferent signals from the two labyrinths are exactly matched. Head motion produces an imbalance between the afferent signals from the two sides, and this imbalance drives the normal compensatory eye movements that stabilize gaze (the vestibulo-ocular reflex) and the modulation of skeletal muscle activity that maintains posture (vestibulo-spinal output).

Pathologic processes involving the vestibular system also produce an imbalance between the afferent signals from the two sides, which the brain cannot differentiate from the afferent imbalance associated with head motion. Vestibular disorders therefore produce pathologic eye movements (nystagmus) and skeletal muscle activity (evident as ataxia and past-pointing), as well as a subjective sense of motion (vertigo). Vertigo is usually a feeling of rotation of the external environment or oneself, reflecting an imbalance between the semicircular canal afferents, but may also involve feelings of linear translation or tilting when the otolith signals are altered by the pathologic process.

CLINICAL EVALUATION

HISTORY

One must determine if vertigo is a recurrent or monophasic symptom, the duration of the episodes, and the presence of other otologic or neurologic symptoms. Commonly associated otologic symptoms are tinnitus, change in auditory acuity, and ear full-

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Table 1. Causes of vertigo

1. Monophasic vertigo
   - A. Common causes
     - Peripheral
       - Labyrinthitis
       - Vestibular neuritis
     - Central
       - Brainstem/cerebellar stroke
       - Multiple sclerosis
   - B. Uncommon causes
     - Labyrinthine infarct
     - Ramsay Hunt syndrome
     - Syphilitic labyrinthitis
     - Tuberculous labyrinthitis
     - Lyme disease
     - Sarcoid
     - Cholesteatoma
     - Acoustic neuroma
2. Recurrent vertigo
   - A. Common causes
     - Peripheral
       - Ménière's syndrome
     - Central
       - Migraine
       - Vertebrobasilar ischemia
   - B. Uncommon causes
     - Vestibular Ménière's
     - Perilymph fistula
     - Recurrent labyrinthine ischemia
     - Hyperviscous states
     - Cogan's syndrome
     - Otosclerosis
     - Vestibular atelectasis
     - Familial recurrent ataxia
     - Vestibular seizures
3. Positional disorders
   - A. Common causes
     - Peripheral
       - Benign positional vertigo (posterior canal)
   - B. Uncommon causes
     - Benign positional vertigo (anterior, lateral canal)
     - Alcohol intoxication
     - Waldenström's macroglobulinemia
     - Central
     - Central positional vertigo
4. Bilateral vestibulopathy
   - A. Common causes
     - Peripheral
       - Ototoxins (aminoglycosides)
   - B. Uncommon causes
     - Otosclerosis
     - Sequential vestibular neuritis
     - Bilateral acoustic neuromas (neurofibromatosis)
     - Bilateral Ménière's syndrome
     - Idiopathic bilateral vestibulopathy
     - Paget's disease
     - Central
     - Wernicke's syndrome

ness or pressure. Neurologic symptoms, which are generally referable to the posterior fossa, include diplopia, facial paresthesia or weakness, and dysarthria, as well as symptoms due to dysfunction of the motor and sensory tracts. Vertigo may occur randomly or may be precipitated or exacerbated by stimuli such as a change in head position with respect to gravity (as on looking up or turning in bed), rapid head movements about the earth-vertical axis, the Valsalva maneuver, or noise.

EXAMINATION

The oculomotor correlate of vertigo is nystagmus. When a patient is examined during an episode of vertigo, nystagmus will be present if the symptoms are vestibular in origin. In contrast, nystagmus is often absent if the patient is examined when asymptomatic. Spontaneous nystagmus (present with the head upright and the eyes straight) should be sought while the patient fixates a visual target and in the absence of fixation. Fixation may be prevented with Frenzel glasses, or by viewing the optic disc of one eye with an ophthalmoscope while the other eye is covered.[1] With peripheral vestibular disorders (due to disease in the labyrinth or eighth nerve), spontaneous nystagmus is usually horizontal-torsional in waveform, with slow phases directed toward the bad side, and is generally suppressed with fixation. In contrast, central vestibular disorders result in a variety of waveforms, including vertical and torsional nystagmus, which are not suppressed with fixation.

Positional nystagmus is provoked by moving the head with respect to gravity. This is usually accomplished by quickly placing the patient into a head-hanging position with either ear undermost (Hallpike-Dix maneuver). This maneuver is particularly important when patients report that their vertigo is provoked by changes in head position, as is the case with benign positional vertigo and other positional disorders (see later).

Gaze-holding nystagmus is present during eccentric gaze, with quick phases in the direction of gaze. This form of nystagmus frequently results from central (typically cerebellar) dysfunction, but may also occur with peripheral disorders. The gaze-holding nystagmus present with peripheral lesions is most prominent when gaze is directed away from the bad ear (Alexander's law).[2]

These three forms of nystagmus should be sought in all patients presenting with vertigo. Other useful clinical signs are described later in conjunction with the relevant vestibular disorders.

ANCILLARY TESTS

If an accurate diagnosis cannot be made following a careful history and physical examination, one should obtain an audiogram (pure tone and speech discrimination testing) and a quantitative test of vestibular function (caloric or rotatory chair). Magnetic resonance imaging studies should be performed if there is a suggestion of a structural lesion of the brain or vestibulocochlear nerve. Imaging studies should not be routinely obtained in patients with a normal neurologic examination and normal auditory and vestibular tests. Blood studies that are usually obtained in patients with unexplained vertigo include

thyroid function tests, erythrocyte sedimentation rate, antinuclear antibodies, and a serologic test for syphilis.

If the basic workup just outlined does not lead to a diagnosis, other diagnostic tests based on the specific features of the clinical presentation should be considered. These tests include electrocochleography (which is particularly helpful in diagnosing Ménière's disease and perilymph fistula), brainstem auditory evoked responses, spinal fluid examination, studies of the posterior circulation (such as magnetic resonance angiography or transcranial Doppler) and specialized blood tests (such as Lyme titer and serum protein electrophoresis).

CLINICAL DISORDERS

MONOPHASIC VERTIGO

Case 1

A 48-year-old man awakens with severe rotatory vertigo associated with nausea, ataxia, and loss of hearing on the left. Examination reveals a spontaneous horizontal-torsional nystagmus with slow phases directed to the left that is suppressed with fixation; absent hearing on the left; and a tendency to lean and past-point to the left. The patient remains persistently vertiginous for 5 days, but then notes a gradual decrease in his dizziness and ataxia. Three months later he is no longer dizzy or ataxic, but experiences transient dysequilibrium when he turns his head quickly. Vertigo does not recur, but his hearing loss does not recover.

Comment

Acute damage to the peripheral vestibular system (labyrinth or eighth nerve) results in an episode of vertigo that lasts for days to weeks and then gradually resolves. The resolution of symptoms reflects recovery of peripheral vestibular function or central adaptation to the persistent unilateral deficit. Central vertigo, due to dysfunction in the vestibular nuclei or their connections in the brainstem and cerebellum, is typically more persistent than vertigo of peripheral origin. This reflects both the permanence of central lesions such as infarcts and the fact that central lesions frequently damage the neural substrate that underlies the process of vestibular compensation.

Peripheral causes of monophasic vertigo are usually vascular or infectious. Vascular disease affecting the vertebrobasilar circulation may result in ischemia of central (brainstem and cerebellum) or peripheral (labyrinth and eighth nerve) components of the vestibular system. When vertigo is due to ischemia of central structures, it is almost always associated with other symptoms and signs referable to the posterior fossa. This has led to the dictum that isolated vertigo is not a manifestation of vertebrobasilar disease. The posterior circulation, however, also supplies the labyrinth and eighth nerve via the anteroinferior cerebellar artery (AICA). Ischemia in the AICA distribution that damages the peripheral vestibular system presents with isolated vertigo, vertigo associated with cochlear symptoms such as hearing loss, or vertigo associated with dysfunction of the dorsal lateral pons and cerebellum (which are also in the AICA distribution).[3] Angiography in these patients typically demonstrates atherosclerotic disease immediately proximal to the takeoff of the AICA.[4] Posterior circulation disease may therefore present with vertigo associated with central symptoms (due to involvement of the brainstem or cerebellum), isolated vertigo or vertigo with cochlear symptoms (due to labyrinthine or eighth nerve dysfunction), or a combination of central and peripheral abnormalities.

Common infectious causes of vertigo are labyrinthitis (which presents with vertigo associated with unilateral auditory symptoms) and vestibular neuritis[5] (which presents with isolated vertigo). These syndromes are presumed to be viral, although the specific viral agent is rarely identified.[6] One clearly defined viral labyrinthine syndrome is herpes zoster oticus (Ramsay Hunt syndrome), which presents with pain and vesicles in or near the external auditory canal, loss of auditory and vestibular function, and facial nerve paresis.[7]

Syphilitic labyrinthitis, which may be due to congenital or acquired syphilis, is an uncommon infectious cause of vertigo. Early presentations of syphilitic labyrinthitis are characterized by congenital or progressive deafness without prominent vestibular symptomatology. The adult-onset syndrome typically consists of a bilateral decline in vestibular and cochlear function, resulting in progressive but fluctuating vertigo and hearing loss.[8]

Lyme disease typically causes cranial neuropathies in its early disseminated stage, in combination with aseptic meningitis and radiculoneuritis.[9] Although the facial nerve is most frequently involved, the vestibulocochlear nerve may also be damaged, producing a syndrome of vertigo and hearing loss that is indistinguishable from labyrinthitis.[10] Central nervous system involvement may occur during the late disseminated state and can cause vertigo in association with other central symptoms and signs.[11]

Most infectious forms of meningitis, including bacterial, viral, fungal, and tubercular, can damage the eighth nerve, resulting in vertigo and hearing loss. Similarly, noninfectious forms of chronic meningitis such as sarcoid and carcinomatous meningitis may cause eighth nerve dysfunction. Sarcoid may also rarely cause vertigo by directly invading the temporal bone or by damaging the central vestibular system in the brainstem or cerebellum.[12]

Although acute lesions of the central or peripheral vestibular system invariably result in vertigo, chronic or congenital lesions generally do not. This is because central vestibular adaptive mechanisms can

usually compensate for a static or slowly progressive asymmetry in vestibular function. The chronic and congenital lesions that produce vertigo are usually those that are associated with fluctuations in vestibular activity. For example, the Mondini malformation, a congenital abnormality of the labyrinth that is frequently associated with a perilymph fistula (see later), results in fluctuating labyrinthine function and vertigo.[13] An example of fluctuation in central vestibular function is the vertigo provoked by the Valsalva maneuver in patients with Chiari malformations.

Acoustic neuroma is typical of the chronic vestibular lesions that rarely result in vertigo. Vertigo is the initial symptom of acoustic neuroma in only 1% of patients, is the presenting complaint in 3%, and is present at the time of diagnosis in 9%.[14] Acoustic neuromas typically present with a combination of unilateral hearing loss and tinnitus, which may be associated with headache, disequilibrium, and facial paresthesia. The other chronic structural lesions in the cerebellopontine angle that may damage the eighth nerve, such as meningiomas and epidermoid cysts, also rarely produce vertigo or nystagmus. A pure tone audiogram and a test of speech discrimination should be obtained when one suspects the presence of an acoustic neuroma. If these are abnormal, a contrast-enhanced magnetic resonance imaging study focusing on the internal auditory canal and cerebellopontine angle should be performed. Brainstem auditory evoked potentials may miss up to 7% of acoustic neuromas and should be performed in place of imaging studies only if the index of suspicion is fairly low.[15]

Therapy for unilateral vestibular dysfunction should be directed to the cause when possible. In the majority of cases, however, specific therapy is not available. In this circumstance, vestibular rehabilitative exercises are usually indicated. These exercises promote central compensation to the vestibular deficit and stimulate nonvestibular mechanisms, such as the cervico-ocular reflex, to help stabilize gaze and improve balance. As suggested earlier, vestibular exercises are more efficacious for peripheral than central lesions. Similarly, pharmacologic therapy with vestibular suppressants such as meclizine and clonazepam appears to be more effective for peripheral deficits. Vestibular suppressants may be used for a limited period of time following an acute vestibular lesion, but should not be used chronically, as they probably retard the process of central vestibular compensation. A recent report suggests that the antiemetic odansetron may be effective in treatment of vertigo associated with central vestibular lesions; this medication may warrant a trial in these patients.[16]

RECURRENT VERTIGO

Case 2

A 35-year-old woman is involved in an automobile accident that results in a concussion. When she wakes, she notes vertigo, imbalance, and tinnitus in the left ear. These symptoms resolve over several days but recur when she laughs, coughs, or hears an organ play at church. Examination is normal with the exception of a horizontal-torsional nystagmus with slow phases directed to the left that is provoked by the Valsalva maneuver and compression of the tragus against the external auditory canal of the left ear.

Comment

Many patients who complain of vertigo note discrete episodes of dizziness separated by periods of normalcy. There are numerous pathologic processes that may underlie this clinical presentation. Ménière's syndrome, which presents with a characteristic combination of vestibular and cochlear symptoms, is a common cause of recurrent vertigo. When the cochlea is spared, patients develop recurrent vertigo without associated auditory symptoms ('vestibular Ménière's').[17] This syndrome is both uncommon and difficult to diagnose. Patients with vestibular Ménière's may have an antecedent history of head trauma or ear infections, and typically develop episodes of vertigo, nausea, and ataxia that last for hours and are not accompanied by other neurologic, otologic, or systemic symptoms.[18] Examination during an episode of vertigo reveals a typical horizontal-torsional nystagmus of the peripheral type, but the quick phases may be directed toward or away from the involved ear. Interictal examination is usually normal, but may demonstrate evidence of a mild unilateral vestibular deficit. It is crucial to perform an audiogram in patients presenting in this manner, as a characteristic, subclinical low-frequency sensorineural loss may be present.[19] Electrocochleography may also be suggestive of Ménière's syndrome in patients lacking subjective auditory symptoms.[20] If the diagnosis of vestibular Ménière's is suspected, therapy with a low sodium diet (less than 2 g daily) and a diuretic such as acetazolamide or hydrochlorothiazide is usually warranted. If episodes of vertigo are frequent and severe, and do not respond to medical management, vestibular nerve section or labyrinthectomy should be considered if the abnormal ear can be conclusively determined.[21,22]

Otolithic crisis of Tumarkin is a very rare manifestation of Ménière's syndrome characterized by sudden drop attacks.[23] These attacks are presumably due to rapid changes in vestibulospinal tone resulting from pathologic fluctuations in otolithic activity. Ablative procedures, such as gentamicin or surgical labyrinthectomy, eliminate these episodes.[24]

Perilymph fistula, an uncommon cause of vertigo, is due to fracture of the labyrinth, usually in the region of the oval or round window. Fistulas are typically caused by head trauma or barotrauma, which may result from changes in the ambient pres-

sure (while flying or diving, for example) or from extreme Valsalva maneuvers (such as occur during weight lifting or blowing on a tuba).[25] When a fistula develops, patients typically experience an acute onset of vertigo and ataxia, which may be associated with unilateral tinnitus and decreased hearing. These symptoms often resolve spontaneously but may recur during Valsalva maneuvers (such as coughing or laughing) or changes in ambient pressure. Vertigo induced by noise (Tullio's phenomenon) may occur with perilymph fistulas. Examination typically reveals hearing loss and nystagmus of the peripheral type. Nystagmus and vertigo may be provoked by the Valsalva maneuver, or by increasing the pressure in the middle ear (Hennebert's sign), which can be accomplished by compressing the tragus against the external auditory canal.[26] Fistulas are frequently difficult to diagnose with certainty. An audiogram should be obtained whenever this diagnosis is considered, and electrocochleography may also be useful.[27] The majority of perilymph fistulas heal spontaneously; patients should be advised to rest in bed with the head elevated, and to avoid Valsalva maneuvers during the convalescent period. If symptoms persist, the fistula can be surgically plugged with fascia,[28] although this frequently does not affect the vestibular and cochlear deficits.

Familial recurrent ataxia type 2 is an autosomal dominant disorder of uncertain etiology that may present with episodic ataxia or vertigo. Episodes typically begin in early adult life, occur with a variable frequency, and last for several hours. Examination during an episode reveals prominent truncal ataxia in association with pathologic eye movements of vestibulocerebellar origin, such as downbeat and gaze-holding nystagmus. Interictal examination may reveal a mild truncal ataxia, which is often slowly progressive, and subtle oculomotor abnormalities.[29] Diagnosis is based on the characteristic clinical presentation, positive family history, and the magnetic resonance imaging scan, which may demonstrate atrophy of the cerebellar vermis. Other forms of familial intermittent ataxia, such as Hartnup disease and maple syrup urine disease, should be excluded with appropriate laboratory tests (including serum and urine amino acid analysis and serum biotin level), although these disorders are all autosomal recessive. Familial recurrent ataxia is highly responsive to therapy with acetazolamide, which should eliminate the episodes of ataxia or decrease their frequency.[30] Other drugs that may be efficacious are valproate and calcium channel blockers. These agents probably do not alter the course of the underlying progressive ataxia.

Vascular disease affecting the vertebrobasilar circulation may present with recurrent episodes of vertigo. Although atherosclerotic disease in the posterior circulation is the most common vascular cause of vertigo, hyperviscous states such as Waldenström's macroglobulemia and polycythemia vera may also result in vestibular symptoms. These disorders frequently produce recurrent or fluctuating vertigo associated with tinnitus and hearing loss.[31] These symptoms are thought to result from ischemia in the peripheral labyrinth and respond to measures that lower serum viscosity.[32]

POSITIONAL VERTIGO

Case 3

A 60-year-old man goes to see a popular movie and is forced to sit in the first row of the theater. When he tilts his head back to look at the screen he becomes acutely vertiginous. The vertigo lasts for 20 seconds and is associated with nausea but no other neurologic, audiologic, or systemic symptoms. When he stands he feels slightly unsteady for several minutes. That night he notes a recurrence of the vertigo when he lies in bed. Physical examination is notable for nystagmus associated with vertigo, provoked when the patient lies on the right side. The quick phase of the nystagmus is directed downward and torsionally counterclockwise.

Comment

Positional vertigo is defined as vertigo occurring when the head position is changed with respect to gravity. Common scenarios are the provocation of vertigo when looking up, bending forward, lying down or sitting up in bed, and turning while lying in bed. Positional vertigo may be due to peripheral or central abnormalities in the vestibular system. The sensory organs of the semicircular canals (the hair cells) are embedded in a gelatinous mass, the cupula. Normally, the cupula has the same density as the surrounding endolymph. The cupula therefore remains stationary when the position of the head is changed with respect to gravity. Positional vertigo of peripheral origin is usually due to a change in the relative densities of the cupula and endolymph, which leads to abnormal movement of the cupula (and activation of the hair cells) when head position is altered.

The most common form of positional vertigo is benign positional vertigo (BPV). This disorder is caused by the abnormal accumulation of otoconial debris in one of the semicircular canals.[33] Posterior canal BPV is by far the most common type. When the patient is placed in the head-hanging position with the involved ear undermost (Hallpike-Dix maneuver), after a brief latency the patient becomes vertiginous and develops nystagmus with quick phases directed upward and torsionally toward the affected ear (that is, clockwise torsion with left BPV, counterclockwise with right BPV). The vertigo and nystagmus resolve in less than a minute, and recur when the subject sits up, although the direction of the nystagmus reverses on sitting.[34]

The uncommon forms of BPV are anterior and lateral canal BPV. The symptoms are the same with these forms as with the posterior canal type, although

lateral canal BPV more commonly becomes symptomatic when the patient turns in bed, rather than when lying down or sitting up, as is the case with the vertical canal forms. The nystagmus associated with anterior canal BPV is the same as with posterior canal, except that the quick phase of the vertical component is directed downward.[35] Lateral canal BPV produces a horizontal nystagmus, which reverses direction when the patient is placed with the opposite ear undermost.[36]

Posterior canal BPV is best treated with otoconial repositioning methods such as the Epley maneuver. This maneuver generally cures this disorder in 50 to 80% of patients when it is performed correctly.[37] It is also used to treat anterior canal BPV, although its effectiveness is less certain with this form. There is no clearly efficacious repositioning maneuver for lateral canal BPV. All forms of BPV may also be treated with the Brandt-Daroff exercises, which frequently terminate the BPV episode over a period of several weeks.[38]

Two other types of positional vertigo of peripheral origin are probably due to changes in the relative densities of the cupula and endolymph. Waldenström's macroglobulinemia may be associated with positional vertigo and nystagmus. It has been suggested that the positional syndrome is due to an increase in the specific gravity of the cupula, resulting from the elevation in gamma globulin weight.[39] Alcohol intoxication also results in positional vertigo, presumably due to the different rates at which alcohol diffuses into and out of the endolymph and cupula. Following alcohol consumption, the patient experiences positional nystagmus that beats toward the ground (geotropic) when placed with either ear undermost. After several hours, positional vertigo and nystagmus resolve, but then recur and may be remain for many hours. The direction of the positional nystagmus is reversed during the recurrent phase, beating away from the ground (ageotropic).[40]

Microvascular compression of the vestibulocochlear nerve, sometimes referred to as "disabling positional vertigo," is a controversial syndrome that is believed to result from vascular compression of the eighth nerve root entry zone. The symptoms associated with this syndrome are not clearly defined, but often include brief episodes of positional vertigo, prolonged nonvertiginous dizziness, and progressive tinnitus and hearing loss.[41] Abnormal prolongation of the interval between peaks I and III on brainstem auditory evoked response testing may suggest the presence of this disorder.[42] Treatment with surgical decompression is carried out in some centers,[43] although its utility remains controversial. Carbamazepine (Tegretol) may also reduce the symptoms associated with this syndrome, presumably by suppressing abnormal activity in the eighth nerve that results from localized demyelination.

Central positional vertigo is due to lesions in the vestibulocerebellum or brainstem. Unlike positional vertigo of peripheral origin, central forms often demonstrate prominent nystagmus without marked vertigo, lack a latency period between change in head position and nystagmus onset, and do not habituate with repeated positioning maneuvers. The nystagmus may be of various waveforms, but generally does not replicate that seen with peripheral abnormalities. One well defined type of central positional vertigo, due to lesions of the cerebellar nodulus, is associated with prominent downbeat nystagmus elicited by a change in head position.[44] Although this disorder may respond to baclofen, medical therapy for central positional vertigo syndromes is usually of limited utility.

BILATERAL VESTIBULOPATHY

Case 4

A 40-year-old man is admitted to the hospital with pneumonia and is treated with several antibiotics, including gentamicin. After several days of therapy, when he attempts to stand he notes imbalance and some type of visual disturbance which he cannot characterize. He is not vertiginous, lacks tinnitus, and demonstrates no nystagmus or hearing change on examination; therefore the gentamicin is continued. His symptoms are ascribed to his "debilitated state," but remain permanently. Physical examination is notable for ataxia, a decrease in visual acuity when the head is oscillated, movement of the optic disc during head oscillation, and the presence of compensatory saccades when the patient is asked to fixate an object and the head is rapidly rotated to one side. No nystagmus is present.

Comment

Bilateral loss of vestibular function is not uncommon. It does not result in vertigo. It is included in this review because the symptoms and signs of bilateral vestibular loss are not well understood by many clinicians and because this condition is frequently iatrogenic and irreversible. As discussed earlier, the vestibular system functions to keep the eyes still in space when the head moves and to maintain balance by modulating activity of the skeletal musculature. With a bilateral vestibular deficit, the symptoms and signs that result are directly referable to an absence of normal vestibulo-ocular and vestibulospinal function. When the head moves, the movement of the eyes in space results in movement of visual images on the retina. This produces symptoms of blurred vision and oscillopsia. These complaints are usually most prominent during ambulation. As suggested earlier, patients frequently do not complain of these specific symptoms, but rather note that their vision is "abnormal." Loss of vestibulospinal function produces symptoms of imbalance and ataxia, which are typically worse when the patient walks in the dark or on uneven surfaces such as gravel. It is important to note

that bilateral loss of vestibular function is usually not associated with vertigo.

The abnormal findings on examination include truncal ataxia and the signs associated with loss of the vestibular-ocular reflex. These signs include a decrease in visual acuity when the head is oscillated,[45] movement of the optic disc (as visualized with the ophthalmoscope) during head oscillation,[1] and the presence of compensatory saccades when the patient is asked to fixate the examiner's nose and the head is quickly rotated to either side.[46] Nystagmus is not present when the loss of vestibular function is symmetrical, and auditory symptoms and signs such as tinnitus and hearing loss may also be absent.

Ototoxic drugs are the most common cause of bilateral loss of vestibular function. Aminoglycoside antibiotics such as gentamicin frequently damage the vestibular labyrinth while sparing the cochlea, and the absence of auditory symptoms may lead to misdiagnosis.[47] Other ototoxic drugs include salicylates and loop diuretics, which cause prominent auditory symptoms, and certain cancer drugs such as cisplatinum. Bilateral loss of peripheral vestibular function may have numerous other causes, including the syndrome of "sequential bilateral vestibular neuritis"[48] and advanced, bilateral Ménière's syndrome. Progressive bilateral vestibulopathy may also occur in the absence of a clear underlying cause.[49] The most common central cause of bilateral vestibular loss, resulting from symmetrical damage to the vestibular nuclei in the brainstem, is the thiamine deficiency syndrome that typically occurs in alcoholics (Wernicke's syndrome).

Therapy for bilateral loss of vestibular function is limited. It is most important to recognize this syndrome early when it is due to the use of ototoxins, so that the offending agent can be stopped. Vestibular damage from aminoglycosides is generally irreversible, and many patients are permanently and severely disabled because the correct diagnosis was not made early in their course of treatment. Vestibular physical therapy, which attempts to promote nonvestibular mechanisms of gaze stabilization and balance, may be beneficial and is usually indicated. There is anecdotal evidence that chronic therapy with acetazolamide may lead to a gradual improvement in vestibular function, although this finding has not been demonstrated in a controlled study.

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