Practical Management of the Dizzy Patient,
edited by Joel A. Goebel.
Lippincott Williams & Wilkins,
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DDX: Fluctuating Vestibular Disease

Gerard J. Gianoli

Fluctuating vestibular disease represents the most common clinical entity you will encounter among your dizzy patients. The most common diagnoses that are identified for fluctuating disease are benign paroxysmal positional vertigo (BPPV), Ménière's disease (also known as endolymphatic hydrops), and perilymphatic fistula (PLF) (Table 18.1). Other disease processes that cause fluctuating vestibular input include autoimmune inner ear disease, superior semicircular canal dehiscence syndrome, ototoxicity, chronic suppurative otitis media/cholesteatoma, congenital inner ear malformations, syphilis, hypothyroidism, hyperlipidemia, and vascular loop compression syndrome. Other entities that less commonly cause fluctuating disease include disease processes that are traditionally considered to cause fixed deficits. These include such entities as viral vestibular neuritis, labyrinthitis, vestibular schwannoma (acoustic neuroma), labyrinthine concussion, and postsurgical disruption of the balance system on one side (e.g., after labyrinthectomy, vestibular nerve section, and acoustic tumor removal). The above list is not exhaustive.

After an insult of any kind to the inner ear the patient will immediately begin central compensation for the loss of vestibular function on that side. This can be noted clinically by the progression of a patient's symptoms. Initially, a patient will have symptoms of whirling vertigo and nausea. After the spinning sensation resolves, there is a period during which the patient notes a more subtle disequilibrium. This is generally described as lightheadedness and is

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Table 18.1 **Characteristics of Common Fluctuating Vestibular Disorders**

Diagnosis	Duration of Rotary Vertigo	Vertigo with	Hearing Loss	Aural Fullness	Fluctuating Hearing	Tinnitus	Trauma
BPPV	<Minute	Positional changes	None	None	None	None	Sometimes
Ménière's	Hours	No association	Usually, unilateral	Usually	Usually	Usually	No
PLF	Minutes–hours	Physical straining	Sometimes, unilateral	Sometimes	Sometimes	Sometimes	Usually
AIED	Hours	No association	Progressive, bilateral	Sometimes	Sometimes	Sometimes	None

most notable with quick head movements. Central compensation will gradually improve these symptoms if there are no further changes in the vestibular input from either ear. Eventually, in the great majority of patients, central compensation continues until the patient is essentially asymptomatic. The duration to completion of compensation depends on the severity of the injury, medical condition, and activity level of the individual patient. Central compensation can range anywhere from a few days to a year. In the case of fluctuating disease, the patient may not complete this process of central compensation before another insult to the vestibular system occurs. Consequently, patients with fluctuating vestibular disease will have the symptoms of the acute period (whirling vertigo) followed by days or weeks of the more subtle symptoms of disequilibrium. The process then repeats itself with each insult to the inner ear. In summary, symptoms of fluctuating vestibular disease are characterized by whirling vertigo followed by variable periods of more subtle disequilibrium.

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BENIGN PAROXYSMAL POSITIONAL VERTIGO (BPPV)

BPPV is the most common cause of vertigo. We are now only beginning to recognize how often this entity has been undiagnosed in the past. It is characterized by spontaneous remissions and exacerbations with symptoms of short lived episodic positional vertigo and chronic disequilibrium. It is common after colds, trauma, surgery, ear infections and is seen frequently in association with other inner ear disorders, but many cases are idiopathic. BPPV can be seen in all age groups, but it is particularly common in the elderly. Diagnosis is obtained by the Dix-Hallpike maneuver, which is easily performed as part of the office examination. The pathophysiology is thought to involve dislocated otoconia (from the utricle) migrating into the posterior semicircular canal causing abnormal stimulation. The Epley maneuver is a highly successful

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Clinical Presentation

A patient with BPPV will generally have two components to the vestibular symptoms. The first and most characteristic of these is positionally induced vertigo. The typical scenario is an intense but brief episode of rotary vertigo when rolling over in bed to the affected ear. The patient will also note this same vertigo, but perhaps to a lesser degree, when bending over and turning the head to look up. The duration of the vertigo is usually no more than 30 s, but to many patients it will feel like much longer. When taking the history, it is important to ask how long the rotational sensation lasted rather than asking how long the patient was dizzy. Many of these patients will say that they are dizzy for hours at a time, but if pressed, they will state that the "spinning spells" last for only a few seconds, and the rest of the time they feel off balance.

Either consciously or subconsciously, many patients will avoid the position that produces this intense vertigo because it is such an unpleasant and sometimes terrifying feeling. Patients who are successful at avoiding the "provocative position" will more commonly complain of the second component of symptoms, a milder disequilibrium that is noted with movements. Patients will state that they feel drunk when trying to walk with a sensation they are going to fall to one side. They will also state that quick positional changes will induce mild disequilibrium. Some will even sound like they have orthostatic hypotension: "when I get up quickly, I feel dizzy." The most common symptoms are

1. Episodic positional vertigo lasting less than 30 s
2. Chronic disequilibrium
3. Spontaneous remissions and exacerbations

Pathophysiology

Several etiologies have been proposed for BPPV. The two most accepted theories at present are cupulolithiasis and canalithiasis. The cupulolithiasis theory proposes that otoconia from the utricle have migrated to the ampulla of the posterior semicircular canal where they have become attached. This results in abnormal stimulation of the posterior semicircular canal, changing the input from that of an angular acceleration detector to that of a linear acceleration or gravity detector. Consequently, every time the head is moved, the vestibular system produces abnormal input from the ampulla of the posterior semicircular canal. However, the ampulla

of the posterior semicircular canal is stimulated maximally when it is positioned in the most dependent position. This may explain why any type of head movement can induce mild disequilibrium, but the most severe symptoms occur with the affected ear placed in the down position.

The canalithiasis theory is similar to the cupulolithiasis theory in that the pathology, again, revolves around detached otoconia from the utricle. However, those who ascribe to this theory feel that the otoconia are floating freely in the endolymphatic fluid of the posterior semicircular canal. With approximation of the provocative position, these crystals then come into contact with the ampulla, resulting in the classic nystagmus noted with the Dix-Hallpike maneuver. This theory better explains the 2- to 5-s delay that you will note before the nystagmus is seen after the Dix-Hallpike maneuver. This latency is one of the very characteristic features of BPPV. Of course, these two theories are not mutually exclusive, and a combination of these two entities offers a better explanation for all of the symptoms and findings of BPPV.

Diagnosis

The diagnosis of BPPV is made with the Dix-Hallpike maneuver. BPPV is such a common entity that I personally feel that all patients who complain of dizziness should undergo a Dix-Hallpike maneuver. The physical exam in these patients is generally normal with the exception of this test. The Dix-Hallpike maneuver is performed by having the patient lie prone with the head turned approximately 45 degrees to the side being tested and slightly hanging over the edge of the examining table. During this, the eyes are examined for nystagmus. A normal exam should demonstrate no evidence of nystagmus, and the patient should experience no disequilibrium. The presence of BPPV will elicit vertigo and nystagmus with five major characteristics:

1. Latency of onset, usually 2–6 s
2. Short duration, usually less than 30 s
3. Reversibility
4. Fatigability
5. Direction

During an abnormal Dix-Hallpike maneuver the patient will be fine initially when getting to the prone position, but after a short latency period of 2–6 s, the nystagmus will occur. Coincident with the nystagmus will be the sensation of whirling vertigo. The nystagmus/vertigo may last anywhere from a few seconds to a minute but is usually no longer than 30 s. The nystagmus is a torsional movement of the eyes. With an abnormal Dix-Hallpike on the right side the patient will exhibit nystagmus with a fast phase in the counterclockwise direction (geotropic nystagmus, upper pole of the eye

moving toward the ground). With an abnormal Dix-Hallpike for the left ear, the fast phase of the nystagmus is in the clockwise direction (geotropic nystagmus). Bilateral BPPV can occur, but it probably represents fewer than 10% of all cases.

When brought back to the upright position, many patients will experience a milder degree of vertigo, and their eyes will demonstrate a milder degree of nystagmus in the direction opposite that of the pathologic nystagmus. This is the phenomenon of *reversibility*. For example, if your patient has right sided BPPV, the left Dix-Hallpike maneuver would be normal, but the right Dix-Hallpike would produce a counterclockwise nystagmus. When he is brought back to the upright position, he will have clockwise nystagmus of lesser magnitude.

Repeated Dix-Hallpike maneuvers will result in progressively milder degrees of both nystagmus and vertiginous symptoms until the exam becomes "normal." This is the phenomenon of *fatigability*. For example, in a patient with right BPPV, you will note less intense nystagmus with a repeat Dix-Hallpike, and the patient will experience less severe vertigo. Perhaps by the third or fourth repetition, the Dix-Hallpike maneuver will demonstrate no evidence of nystagmus, and the patient will no longer note vertigo. Because of the phenomenon of fatigability, the Dix-Hallpike maneuver is very specific but not sensitive. In other words, an abnormal Dix-Hallpike is indicative of BPPV, but a normal Dix-Hallpike does not rule out BPPV. This is because some patients will have "fatigued" their response before coming to see you in the office. Additionally, the presence of BPPV does not rule out other concomitant pathologies. BPPV has been noted in patients with Ménière's disease, acoustic neuromas, after vestibular neuritis, chronic suppurative otitis media, and others. Said in another way, just because someone has another inner ear disorder, does not mean he cannot also have BPPV.

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MÉNIÈRE'S DISEASE (ENDOLYMPHATIC HYDROPS)

Endolymphatic hydrops is the progressive retention of fluid in the endolymphatic space. There are many causes for endolymphatic hydrops, and a predictable collection of symptoms results from endolymphatic hydrops. This collection of symptoms is termed Ménière's syndrome. When no identifiable cause can be found for Ménière's syndrome, it is termed Ménière's disease. So, Ménière's disease is idiopathic Ménière's syndrome. Ménière's can be seen in all age groups but is relatively rare in the pediatric population and common in the 30- to 50-year-old group. Most patients present with unilateral disease, but 30–50% will develop bilateral disease. The characteristic symptoms are unilateral aural fullness, fluctuating hearing loss, tinnitus, and vertigo. Spontaneous remissions and exacerbations also characterize this disorder. The physical examination is typically normal. The workup should include a search for

possible underlying causes. Most patients respond to dietary changes and medical therapy. Fewer than 20% of patients will require surgical intervention.

Clinical Presentation

Ménière's disease is characterized by

1. Fluctuating hearing loss
2. Tinnitus
3. Aural fullness
4. Vertigo

A typical Ménière's attack will start with the sensation of unilateral aural fullness, which is often described as similar to the feeling the ear gets during a cold or an airplane flight. Coincident with this is often a low pitch roaring tinnitus in the affected ear. Hearing loss will develop in the affected ear, and then a spell of rotary vertigo will occur. The vertigo will typically last for 30 min to 4 h. During the vertigo spell, the patient will frequently experience nausea, vomiting, and diaphoresis. After the vertigo spell, the hearing loss, tinnitus, and aural fullness may resolve, and the patient may experience disequilibrium that will last for several days. The patient will then have resolution of symptoms and is typically free of symptoms when visiting the physician.

A fairly rare, but disquieting symptom of Ménière's disease is the drop attack or *otolithic crisis of Tumarkin*. In this situation the patient will suddenly lose all postural tone and drop to the floor without any warning. It can be distinguished from a cardiac event or stroke because there is no loss of consciousness during the drop attack and, although quite unsteady, immediately after falling to the floor the patient can often get right back up.

Pathophysiology

The pathophysiology of endolymphatic hydrops is felt to be most likely inadequate resorption of endolymph. The endolymphatic sac is purportedly the organ where endolymph is resorbed. Experimental disruption of the endolymphatic sac or blocking the endolymphatic duct, which leads to the sac, has been shown to result in endolymphatic hydrops. Progressive dilation of the endolymphatic space will eventually result in intralabyrinthine membrane ruptures, which may be the cause of vertiginous spells and hearing loss. After these ruptures fluid equilibrium returns, and the patient often has a quiescent period until endolymphatic pressure builds up again. Progressive destruction of intralabyrinthine structures results in what has been termed *burned out Ménière's*.

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Diagnosis

Patients with Ménière's disease will typically have a normal physical exam unless they are seen during an acute attack. During an acute attack, the patient will be nauseated, diaphoretic, and will demonstrate nystagmus. The nystagmus is typically beating with the fast phase toward the normal ear. They will also note a feeling like the affected ear is blocked by wax or is full of fluid in spite of a normal otoscopic exam. Audiometric examination will often demonstrate a low frequency sensorineural hearing loss that may improve to normal once the attack has resolved.

The natural history of Ménière's is one of remissions and exacerbations. Many will experience several years quiescent of any symptoms only to see them return. The natural history is also quite variable in that many patients will never progress beyond mild disease regardless of treatment and others will develop severe disease despite aggressive treatment. In the early onset of the disease, the patient may have only one or two of the classic symptoms, such as fullness and/or fluctuating hearing. As the disease progresses, the symptoms become more classic with the patient experiencing all of the classic symptoms: unilateral fluctuating hearing loss, tinnitus, aural fullness, and episodic vertigo. As the disease progresses further, the fluctuating hearing loss will become a permanent hearing loss, and the symptoms of tinnitus and aural fullness will become more constant than episodic. In burned out Ménière's, the patient will have chronic disequilibrium but no acute vertiginous episodes and will typically have stable, moderate to severe hearing loss.

Diagnostic tests that are helpful in the evaluation of Ménière's disease can be grouped into two categories: those helping to confirm Ménière's disease and those helping to rule out other causes. In the category of helping to confirm are audiometry, electronystagmography (ENG), and electrocochleography (ECoG). None of these tests needs to be done to diagnose Ménière's disease, but the information derived from them can provide information about the severity of disease and sometimes help in the diagnosis. Audiometry may be normal or demonstrate low frequency sensorineural hearing loss. Repeat testing may show objective evidence of fluctuation of the hearing loss. ECoG is an electrophysiologic test similar to evoked response testing, but one of the electrodes is placed either on top of the eardrum itself or through the eardrum and on top of the promontory of the inner ear. The tracings that are measured represent the inner ear response to auditory stimuli. The important variable being measured is the ratio of the amplitudes of the summating potential and the action potential. This is felt to be a fairly specific test but lacking somewhat in sensitivity. ENG may demonstrate normal findings, particularly in the early onset of disease, or a caloric weakness in the involved ear as the disease progresses. Additionally, spontaneous nystagmus may be noted if ENG testing is performed shortly after an acute vertiginous episode.

Among the tests that are useful in excluding causes for Ménière's syndrome include auditory brainstem evoked responses (ABR), radiographic imaging, and blood tests. Computed tomography (CT) and ABR have been used as a screen for acoustic neuromas and cerebellopontine angle tumors. However, a thin slice magnetic resonance imaging (MRI) with gadolinium contrast is currently the gold standard for anatomically evaluating this area for such lesions. Blood tests that may be useful include a thyroid panel, syphilis serology, and an autoimmune panel.

In summary, the diagnosis of Ménière's disease is made in a patient who has

1. Characteristic history: unilateral fluctuating hearing loss, tinnitus, aural fullness, and episodic vertigo
2. Supportive diagnostic tests: audiometry, ECoG, ENG
3. Exclusion of other causes by MRI and blood tests

The differential diagnosis of Ménière's syndrome is large. Entities that are known causes of Ménière's syndrome, include

Trauma: acoustic trauma, temporal bone fracture, surgical trauma to the inner ear
Autoimmune: autoimmune inner ear disease, Cogan's disease, lupus
Infectious: chronic otitis media, syphilis, labyrinthitis (viral or bacterial)
Metabolic: otosclerosis, Paget's disease, hyperlipidemia
Neoplastic: acoustic neuroma, leukemia
Congenital/developmental: Mondini's dysplasia (Fig. 18.1), large vestibular aqueduct syndrome, congenital deafness, jugular bulb diverticulum
Endocrine: thyroid disease, diabetes
Idiopathic: perilymphatic fistula, Ménière's disease

Treatment of Ménière's disease can be categorized into a four-step approach: dietary changes, medical therapy, conservative surgical therapy and ablative surgical therapy. Dietary changes involve the elimination of caffeine and reduction of sodium intake. Medical therapy usually includes control of any identified underlying medical problems (such as hypothyroidism or diabetes), maintenance diuretic therapy, and PRN use of vestibular suppressant medication during acute spells. Approximately four out of five patients will respond quite nicely to the above treatment. However, for those who have progressive disease in spite of maximal medical therapy, conservative surgical therapy may be employed, endolymphatic sac surgery or transtympanic steroid therapy. Ablative surgical therapy is usually reserved for the most severe cases that have failed all other treatments and includes transtympanic aminoglycosides, labyrinthectomy, or vestibular nerve section. Using this stepwise treatment approach, virtually all patients can be freed from vertiginous spells. Treatment is discussed more fully in a later chapter. A

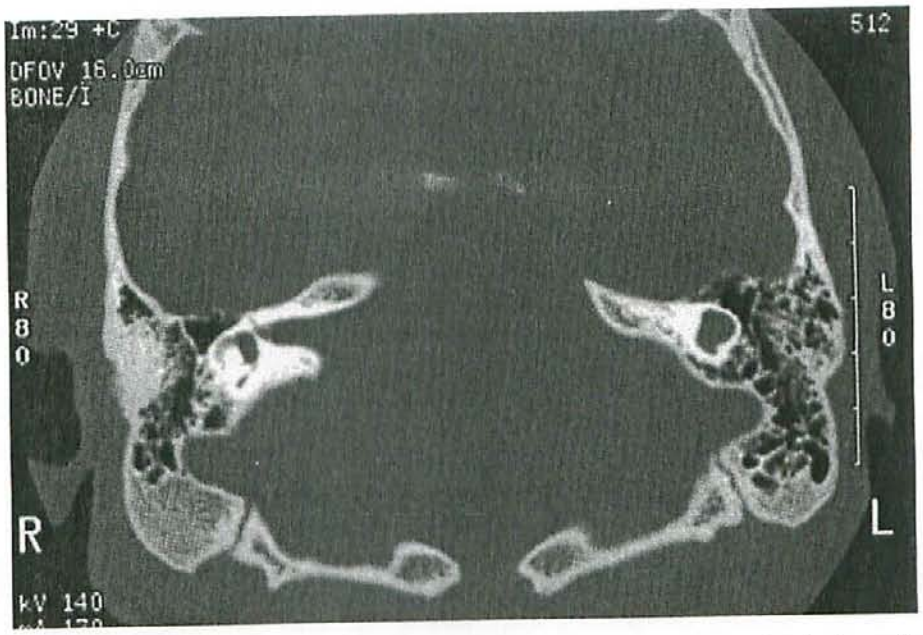

FIGURE 18.1 Axial CT scan of the temporal bone demonstrating Mondini's dysplasia on the left side. Note the absence of cochlear partitioning. These patients are at higher risk for PLF.

detailed review of Ménière's disease was recently published in the Otolaryngology Clinics of North America.

PERILYMPHATIC FISTULA

Perilymphatic fistula (PLF) is the abnormal opening of the bony capsule or round/oval window membranes of the inner ear. Symptoms are variable but can include episodic vertigo, fluctuating and progressive hearing loss, tinnitus, and aural fullness. The most common causes of PLF are trauma, surgery, cholesteatoma, and congenital abnormalities. The diagnosis of PLF can be very challenging even for the most experienced neurootologist and may still be in doubt even after surgical exploration. Initial treatment is usually bed rest with head elevation for acute PLF in hopes of spontaneous healing. For chronic PLF, surgical exploration with repair is usually indicated.

The most common causes of PLF are trauma, surgery, cholesteatoma, and congenital abnormalities.

Classification

The presentation for a PLF may be varied depending on the cause of the PLF. PLF can be classified by its cause:

1. Iatrogenic (usually poststapedectomy)
2. Traumatic

3. Erosive
4. Congenital
5. Spontaneous

For traumatic PLF, the trauma may be direct, implosive, or explosive. Direct trauma would include blunt and penetrating injuries to the temporal bone. Implosive trauma is any that would cause deflection of the round or oval windows inward and result in a subsequent membrane disruption. An example of this would be an open hand slap to the ear, water skiing accidents, barotrauma from scuba diving, and violent nose blowing. Explosive trauma is seen when internal pressure results in outward disruption of the round or oval window membranes. This can be seen with forms of Valsalva such as physical exertion as in weight lifting, violent coughing/vomiting, blunt abdominal/thoracic trauma, and childbirth. A classic presentation for this would be a weight lifter who, while straining at the bench press, notes a sudden pop in his ear followed by hearing loss and vertigo. Those patients with congenital inner ear abnormalities (Fig. 18.1) or who have undergone stapes surgery are at particular risk of developing a PLF under these scenarios.

Another classification of PLF is the spontaneous variety, those with no antecedent trauma or predisposing factors. The spontaneous PLF is a controversial entity and probably represents patients who have had some inciting event that they have forgotten or not correlated with their vestibular symptoms.

The usual location for a PLF is the round or oval window and usually is the result of trauma; however, erosive processes can result in PLF of the bony otic capsule. The most common clinical entity that causes an erosive PLF is cholesteatoma, and the most common place for it to erode into is the horizontal semicircular canal. Other erosive processes that can cause PLF are syphilis and tumors.

A recently described variation of PLF is superior semicircular canal dehiscence. Some patients with symptoms of PLF have been noted on high resolution CT scan (Fig. 18.2) to have an absence of bone covering the superior aspect of the superior semicircular canal. Clinical experience with this entity is limited at this time, but repair of the defect appears to be curative.

Pathophysiology

The pathophysiology of PLF is poorly understood. The leading theory to explain the symptoms is the double membrane break theory. This theory proposes that for hearing loss and vertigo to result from a PLF, there must be a concomitant break of an inner ear membrane. It is generally accepted that mechanisms to explain audiovestibular dysfunction attributed to PLF include direct damage to the end organ, disruption of the fluid hydrodynamics of the inner ear, and electrolyte abnormalities caused by the mixture of perilymph and endolymph. A secondary endolymphatic hydrops

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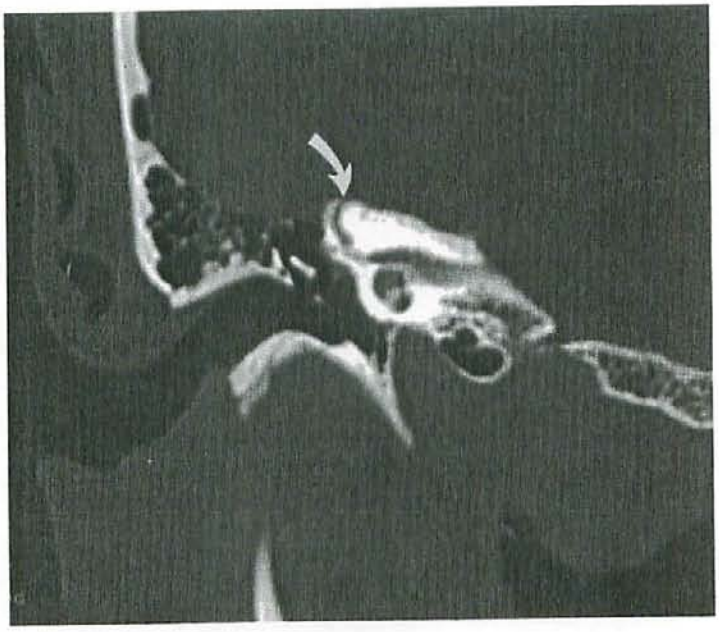

FIGURE 18.2 Coronal CT scan of the temporal bone demonstrating bony dehiscence of the superior semicircular canal (*curved arrow*). In this disorder, the dura of the temporal lobe is in direct contact with the membranous superior semicircular canal. Changes in intracranial pressure result in stimulation of the superior semicircular canal.

caused by the relative excess of endolymph has been demonstrated in experimental PLF; air in the inner ear may also contribute to the possible pathophysiology of audiovestibular impairment of PLF.

Diagnosis

The patient's history is the most important key in making the diagnosis of PLF. The hallmark of PLF is its variability of symptoms, none of which is pathognomonic. The patient may have auditory symptoms but no vestibular symptoms or vestibular symptoms but no auditory symptoms. Most commonly, however, there is usually evidence of both auditory and vestibular dysfunction. The auditory symptoms include fluctuating hearing loss, progressive hearing loss, and no hearing loss. Tinnitus may be present. The vestibular symptoms may be described as episodic vertigo with or without nausea/vomiting, positional vertigo, intermittent disequilibrium, or chronic disequilibrium. The vertiginous spells, when present, may last as long as several hours or be as short as a few seconds. Worsening of vestibular symptoms with physical straining is an important historical finding. Tullio's phenomenon is the finding of sound-induced nystagmus/vertigo and has been noted

with PLF. Probably the most helpful part of the history is whether there has been a predisposing factor (i.e., prior ear surgery, congenital inner ear abnormality) or an inciting traumatic event. There is no constellation of symptoms which is diagnostic for PLF, and this may be a difficult entity to distinguish from other vestibular disorders, particularly Ménière's disease.

Clinical Presentation

1. Inciting event such as trauma, straining, erosive process (cholesteatoma, syphilis, tumor)
2. Chronic disequilibrium, episodic vertigo
3. Auditory symptoms including hearing loss, fluctuating hearing, progressive hearing loss, tinnitus
4. Exacerbation of dizziness with physical exertion

The general physical examination is usually normal in PLF patients. Otoscopy should be performed carefully to evaluate the possibility of recent trauma, prior surgery, or evidence of cholesteatoma. Pneumatic otoscopy has been used to perform the office *fistula test*. While varying positive and negative pressure to the ear canal with the pneumatic otoscope, examine the patient's eyes for nystagmus and ask the patient to relay any symptoms of vertigo/disequilibrium. An abnormal test indicative of a PLF occurs when nystagmus is noted. The subjective sensation of dizziness noted by the patient is usually a less accurate indicator of PLF. The sensitivity of the fistula test can be improved by more objective means of evaluating the physiologic response. This can be done with ENG monitoring of the eyes while the pressure is applied to the ear, or postural sway can be measured on computerized dynamic platform posturography while pressure is applied. However, even with such objective monitoring, there is still a fair false positive and false negative rate for the fistula test.

Audiometry may demonstrate normal hearing or a unilateral hearing loss. Unfortunately, there is no characteristic hearing loss associated with PLF. ENG testing may show reduced caloric function in the affected ear, positional nystagmus, spontaneous nystagmus, or it may be completely normal. There is no completely accurate diagnostic test for PLF, but the workup should include a search for other possible causes. The differential diagnosis includes Ménière's disease and all of the entities included in the Ménière's differential. Tests generally used to exclude other etiologies are radiographic imaging (especially MRI), syphilis serology, chemistry panel, thyroid panel, and an autoimmune panel.

Initial treatment of an acute traumatic PLF is strict bed rest for 4–7 days with head elevation and avoidance of straining for an additional 6 weeks. Treatment of chronic round or oval window PLF generally requires middle ear exploration with repair of the defect. PLF associated with erosive processes such as cholesteatoma and

tumors requires more involved treatment of the underlying process with appropriate repair of the defect.

AUTOIMMUNE INNER EAR DISEASE

Autoimmune inner ear disease (AIED) is characterized most prominently by bilateral asymmetric progressive/fluctuating sensorineural hearing loss. The progression of hearing loss is typically over the course of several months but may progress over several years time or be more rapid in progression. Vertigo is usually not a prominent symptom of AIED, but a Ménière's syndrome may be present in some patients. The most important historical clues are the presence of other systemic autoimmune disorders and improvement of hearing loss with immunosuppressive medication. Audiometry will demonstrate the hearing loss, which will progress and/or fluctuate over several months to years. Vestibular testing will often show bilateral reduced vestibular function. Supportive blood tests include nonspecific tests such as sedimentation rate, antinuclear antibody titers, and rheumatoid factor. Blood tests specifically designed for AIED include the lymphocyte transformation test against pooled inner ear antigen, Western blot analysis against fresh bovine inner ear antigen, and anti-heat shock protein 70 antibodies. These tests are specific but are somewhat lacking in sensitivity. The best diagnostic test may be a trial of immunosuppressants. Treatment is typically high dose steroids, and some patients may require long term therapy.

Diagnostic Clues

1. Progressive asymmetric bilateral hearing loss
2. Ménière's syndrome may be present
3. Hearing improved with immunosuppressants
4. Supportive blood tests

FIXED VESTIBULAR DEFICITS

Fixed vestibular deficits will be covered in detail in the next chapter but are worth mentioning here. Although the classic presentation of a fixed vestibular deficit is quite different from fluctuating vestibular disease, the distinction in the clinical setting can sometimes be blurred in the early stages of the disease. During the onset of the disease process, there is often some fluctuation of vestibular input until the process is complete. Consequently, it may be impossible to distinguish a fixed vestibular process from a fluctuating vestibular process in the early stages of disease. Time and clinical observation will usually help distinguish between these types of process.

One lesion that generally is considered a fixed vestibular deficit which may have recurrence is vestibular neuritis. (This disorder is described in detail in the next chapter.) Considered to be a viral inflammation of the vestibular nerve (superior division), it is typically characterized by a single episode of severe vertigo with slow resolution over the course of weeks to months. A small percentage of patients with vestibular neuritis have chronic recurrent episodes of vertigo. In many instances, patients recovering from neuritis develop benign positional vertigo. One possible mechanism is utricular damage from the neuritis causing release of otoconia and stimulation of an intact posterior canal ampulla. Some patients have recurrent vestibular neuritis. Distinguishing recurrent vestibular neuritis from other fluctuating vestibular disorders may be difficult.

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