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DDX: Fixed Vestibular Deficits

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For purposes of this chapter, a fixed vestibular deficit is defined as any disease process that results in damage, either partial or complete, to only one side of the peripheral vestibular system: the eighth cranial nerve or end organ. An additional requirement of this definition is that the damage is completed, and the vestibular input from that side is no longer changing. So, a patient with active Ménière's disease has a fluctuating disorder, and a patient with inactive Ménière's disease has a fixed deficit.

The multiple causes of fixed vestibular deficits can be placed together in one chapter not because they have similar symptoms at the onset but because they eventually lead to the same collection of symptoms. These symptoms are derived from poor or inadequate central compensation for this vestibular injury. That said, most patients with fixed vestibular deficits are essentially asymptomatic or may have minimal symptoms because most patients develop adequate compensation for these injuries.

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Some of the most common causes of fixed vestibular deficits are vestibular neuritis, advanced (inactive) Ménière's disease, trauma, postsurgical ablation of the vestibular system, ototoxicity, and acoustic neuroma. Patients who have fixed unilateral deficits and have had poor compensation for these injuries will display generally similar symptoms. The most notable of these is disequilibrium with head movements, with faster movements producing more notable symptoms. This is particularly the case when the patient turns the head to the side of the injury. Unlike acute vestibular

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injury patients, these patients do not have severe episodes of whirling vertigo. Their symptoms may have a vertiginous component, but it is much milder and usually lasts only for a few moments. Another distinction from acute vestibular dysfunction or fluctuating vestibular disease is that fixed vestibular deficits generally do not produce symptoms (or symptoms can be alleviated) when the patient is motionless. In fluctuating disorders, the patient is often completely motionless during the worst part of their vertiginous spell (Table 19.1).

Some symptoms of a fixed deficit are:

1. Asymptomatic (most patients)
2. Chronic disequilibrium
3. Exacerbation with movement
4. No symptoms when motionless

One common fluctuating disorder that may be difficult to distinguish from a fixed deficit with poor compensation is BPPV.

One common fluctuating vestibular disorder that may be difficult to distinguish from a fixed vestibular deficit with poor compensation is benign paroxysmal positional vertigo (BPPV), which may be differentiated by an abnormal Dix-Hallpike test. BPPV is described in detail in the preceding chapter.

VESTIBULAR NEURITIS

Acute vestibular neuritis has also been known by many other names: vestibular neuronitis, neurolabyrinthitis, viral labyrinthitis, epidemic vertigo, and acute vestibulopathy of unknown etiology. The clinical effects of this disorder are very dramatic and usually precipitate a visit to the emergency room. The patient experiences a sudden onset of severe whirling vertigo without a change in hearing. The vertigo is incapacitating and associated with nausea, vomiting, and diaphoresis. The physical examination will show a distressed patient with spontaneous nystagmus with the fast phase beating away from the affected ear. The vertigo is persistent, and the most severe symptoms will resolve slowly over several days to weeks after onset. The disease is mostly self-limited, and most patients have complete resolution of their symptoms. Treatment is supportive with use of vestibular suppressant and antinausea med-

Table 19.1 **Symptoms of Fixed Vestibular Deficits versus Fluctuating Vestibular Deficits**

	Motion Provoked	Duration	Hearing Loss, Tinnitus
Fixed deficit	Usually	Seconds	None or stable
Fluctuating deficit	Only in BPPV	Seconds–hours	May be fluctuant

ications and steroids for inflammation at the onset of the disorder and vestibular rehabilitation exercises to hasten central vestibular compensation.

Clinical Presentation

Vestibular neuritis has such a dramatic presentation that when you have seen it once, you will never forget it. This patient is severely ill. There may be a history of a prior cold or upper respiratory infection in the days or weeks prior to the onset of the acute vertigo. It has been noted by many to have a seasonal occurrence and occur in epidemic fashion. The characteristic symptom is a single, acute, severe episode of vertigo without any associated auditory or neurologic symptoms. The vertigo will generally progress to a severe spinning sensation associated with nausea, vomiting, and diaphoresis over the course of just a few minutes. Very often the patient (and often the physician) will be convinced that he is having a stroke or a heart attack, precipitating a visit to the emergency room.

There are almost never any auditory symptoms to suggest that the process is related to the ear, and there are no other neurologic symptoms. The vertigo will be persistent and gradually improve over the course of several days to weeks. After this, the patient usually notes symptoms brought on only by quick head movements, particularly to the affected side. The patient may feel off balance for several months after the incident and in some cases may have symptoms persist for years. Additionally, it is not uncommon for patients to develop BPPV after a bout of vestibular neuritis. Symptoms include

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1. One severe, prolonged episode of vertigo, nausea, vomiting, and diaphoresis
2. No auditory symptoms, no other neurologic symptoms
3. May develop BPPV
4. No recurrence of vertigo

Pathophysiology

The pathophysiology is felt to be a viral inflammatory process of the superior division of the vestibular nerve or a vascular occlusive process resulting in sudden loss of superior division of the vestibular nerve function on the involved side. Inflammatory cell infiltrate and edema are present. Eventually there is neural degeneration and replacement of neural elements with fibrous tissue. The end result is a significant loss of neural innervation of the vestibular end organ. This denervation may be partial or complete. Because the superior division of the vestibular nerve is selectively affected, function of the end organs innervated by the inferior vestibular nerve are preserved, namely the saccule and the posterior semicircular canal. Additionally, there may be degeneration of the vestibular end organ, the utricle, the horizontal and the superior semicircular canal ampullae. Degeneration of the otolithic organ (utricle) can result in the dispersement of otoconia, which can result in secondary BPPV.

Diagnosis

The diagnosis is made by a typical history and exclusion of other causes of sudden loss of unilateral vestibular input. This can be accomplished by history taking. Physical examination during the acute phase of vestibular neuritis will demonstrate horizontorotary nystagmus with the fast phase beating to the uninvolved side. An otherwise normal neurologic exam is important to distinguish this from an acute cerebellar hemorrhage or infarct. Imaging studies are usually normal, but high resolution magnetic resonance imaging (MRI) with gadolinium may show enhancement of the eighth nerve. Electronystagmography (ENG) will demonstrate absent or significantly reduced caloric response on the involved side. Spontaneous nystagmus may or may not be seen on the ENG depending on how soon after the insult the ENG was performed. Diagnostic criteria include

1. Appropriate history and physical exam
2. Absent or significantly reduced caloric response on ENG
3. Normal imaging studies

ADVANCED MÉNIÈRE'S DISEASE

Ménière's disease is described in more detail in the preceding chapter because it is the prototypical example of a fluctuating disorder. However, Ménière's is also known for spontaneous remissions and exacerbations. Patient's with advanced Ménière's disease (also known as *burned out Ménière's*) will have different complaints from its more active phase. Very frequently after a patient has been affected by Ménière's for more than 10–15 years, there is a reduction in the acute vertiginous spells. These patients will complain of a more chronic form of imbalance. They have a stable, flat, sensorineural hearing loss, and vestibular testing will show a significant reduction in caloric function.

Clinical Presentation

Very often these patients will not give a past history of Ménière's disease or prior vertiginous attacks unless they are questioned specifically. Their symptoms are similar to those of other patients with fixed unilateral or bilateral deficits, namely chronic disequilibrium exacerbated by movement. In addition to the vestibular symptoms, these patients will have unilateral or bilateral hearing loss and tinnitus. The ear fullness noted in active Ménière's may or may not be present. It has been proposed that the stabilization of symptoms is caused by progressive endolymphatic hydrops resulting in fistulization of the endolymphatic space to the perilymphatic space thus eliminating an imbalance in fluid pressure between the endolymphatic and perilymphatic spaces.

Diagnosis

The diagnosis is most easily made in a patient who gives a history of prior active Ménière's disease who now has stable hearing loss, tinnitus, and chronic disequilibrium. It is important to rule out other concomitant vestibular disorders in this setting. In particular, BPPV is common in this patient population and may present with similar symptoms.

ACOUSTIC NEUROMA

Acoustic neuroma (AN) is a misnomer because it does not arise from the auditory division of the eighth cranial nerve, nor is it a neuroma. The more precise term, but less commonly used, is *vestibular schwannoma*. This lesion represents 80–90% of all cerebellopontine angle tumors. It is a benign neoplasm arising from the Schwann cells of the vestibular portion of the eighth cranial nerve. Interestingly, vertigo is not a common complaint with AN, occurring in fewer than one fifth of patients and most often with a very small AN. Disequilibrium, or the chronic, more subtle feeling of imbalance is a more frequent complaint. As opposed to vertigo, which is more common with smaller AN, disequilibrium becomes more common with an increasing tumor size. Nearly three fourths of patients with large (more than 3 cm) AN will complain of disequilibrium.

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Clinical Presentation

There is a tremendous amount of variability in the clinical presentation of AN, and it has been known to mimic several other auditory and vestibular syndromes. Consequently, AN remains in the differential diagnosis of anyone with unilateral symptoms relating to auditory dysfunction (hearing loss, tinnitus) or vestibular dysfunction (vertigo, disequilibrium). Additionally, many AN patients may be completely asymptomatic. However, the "classic" clinical presentation of a small AN is initially unilateral tinnitus and mild unilateral hearing loss. As the tumor grows the patient will note progressive hearing loss in the involved ear and may experience vertigo. The vertigo, however, is usually short lived and eventually resolves or is replaced by a much milder disequilibrium. As the tumor becomes large, the trigeminal nerve may become compressed with variable symptoms, numbness, facial pain, and trigeminal neuralgia. As the brainstem becomes compressed, hydrocephalus ensues with associated headaches and visual loss. If untreated, persistent growth results in lower cranial nerve dysfunction, ataxia, hemiparesis, and respiratory arrest. Early symptoms include hearing loss, tinnitus, and disequilibrium. Late symptoms include these as well as headache, trigeminal symptoms, and hydrocephalus.

Pathophysiology

Acoustic neuromas have a variable growth rate with four distinct stages: internal auditory canal (IAC), cisternal, brainstem compressive, hydrocephalic. The tumor is felt to arise from the junction of central and peripheral myelin in the vestibular portion of the eighth cranial nerve. This is in the internal auditory canal. Growth of the tumor progresses from the IAC to the cerebellopontine angle cistern and attains the appearance of an ice cream cone. During the IAC and cisternal stages of growth, there is progressive compression of the vestibular and cochlear nerves. This correlates with progressive hearing loss, tinnitus, and disequilibrium. The facial nerve, although quite compressed during these stages, very rarely demonstrates dysfunction. Continued growth is in the direction of the cerebellopontine angle cistern and results in compression of the brainstem and may also compress the lower cranial nerves. Further compression of the brainstem eventually results in collapse of the fourth ventricle and development of hydrocephalus. The terminal stage ends with herniation of the cerebellar tonsils. To summarize, the stages are:

1. Internal auditory canal
2. Cisternal
3. Brainstem compressive
4. Hydrocephalic

Diagnosis

As mentioned, ANs have a quite variable presentation and should be in the differential diagnosis of any process of vestibular or cochlear nerve dysfunction. Although many tests are advocated as screening tools for AN, the gold standard for diagnosis is a thin section MRI of the internal auditory canals with gadolinium contrast. The AN enhances markedly with gadolinium on a T1-weighted image (Fig. 19.1). Other studies used for screening for AN, but less sensitive than MRI, include computed tomographic (CT) cisternography,

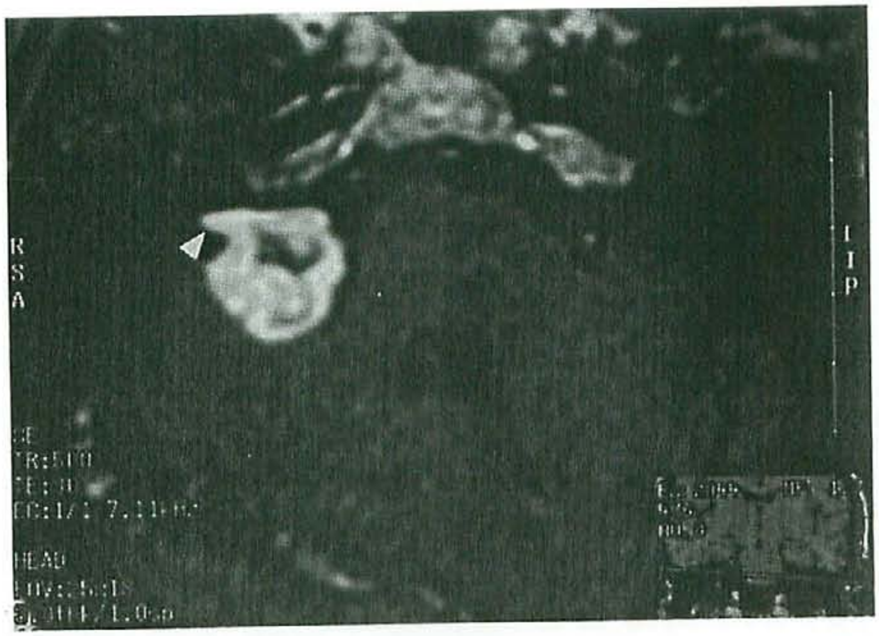

FIGURE 19.1 Axial, T1-weighted MRI through the internal auditory canal with gadolinium contrast demonstrates the typical appearance of an acoustic neuroma. The *arrowhead* points to the internal auditory canal portion. The rounded portion of the tumor is the cisternal component.

acoustic brainstem response, ENG, and retrocochlear audiometric studies. Other tumors in this region which may produce similar symptoms but are less common include meningiomas, epidermoids, arachnoid cysts, schwannomas of the other lower cranial nerves, and lipomas.

TRAUMA

Trauma to the inner ear may result in labyrinthine concussion, perilymphatic fistula (PLF), BPPV, endolymphatic hydrops, or a combination of these. (PLF, BPPV, and endolymphatic hydrops have been discussed in the preceding chapter on fluctuating unilateral disorders.) The most severe trauma to the inner ear would result in a labyrinthine fracture (i.e., temporal bone fracture with labyrinthine involvement) with total loss of vestibular and auditory function.

The typical history of labyrinthine concussion is vertigo, which begins immediately after the trauma. The duration of the vertigo is quite variable and may last only for an hour or may last several weeks depending on the severity of the injury and degree of vestibular function lost. Most patients will have complete resolution of vestibular symptoms, but some will have persistent disequilibrium resulting from incomplete central vestibular compensation. Long term treatment is vestibular rehabilitation exercise and elimination of vestibular suppressant medication.

Clinical Presentation

Initially, these patients will have acute vertigo immediately after the trauma; however, if the patient had lost consciousness, this part of the history may not be recalled. After the initial vertigo resolves, the patient will have chronic disequilibrium until central compensation occurs. In the patient who fails to compensate, continued motion intolerance and chronic disequilibrium are noted. Additional symptoms may include tinnitus and hearing loss in the affected ear. If a temporal bone fracture had occurred, evidence of tympanic membrane disruption or hemotympanum may be noted. Other complications from a temporal bone fracture can include ipsilateral facial paralysis and cerebrospinal fluid otorrhea.

Diagnosis

The diagnosis is usually suspected in the face of a history of trauma followed by a short period of severe vertigo and then chronic disequilibrium. Physical examination may show evidence of trauma, or it may be completely normal. Supportive diagnostic tests include audiometry, which may show a conductive, sensorineural, or mixed hearing loss. ENG will often show spontaneous nystagmus and a reduced caloric response in the involved ear. A high resolution CT scan of the temporal bone may demonstrate a fracture and evidence of blood accumulation in the mastoid/middle ear (Figs. 19.2 and 19.3). The most important entities to consider in the differential diagnosis are BPPV and PLF. These can sometimes be excluded by history, but Dix-Hallpike and fistula tests should supplant the exam.

1. History of trauma
2. Unilateral hearing loss and tinnitus may be present
3. Chronic disequilibrium
4. Exclusion of BPPV, PLF

POSTSURGICAL VESTIBULAR LOSS

For patients who have incapacitating fluctuating unilateral disease and fail medical and/or nonmedical therapy, ablation of all remaining vestibular input from the diseased ear is often desirable. This changes the disorder from a fluctuating unilateral disorder to a fixed unilateral deficit.

Complete loss of vestibular function is the desired or anticipated result of some surgical procedures. For patients who have incapacitating fluctuating vestibular disease and fail medical and/or nonmedical therapy, ablation of all remaining vestibular input from the diseased ear is often desirable. This changes the disorder from a fluctuating disorder to a fixed deficit. As mentioned previously, most patients will eventually become completely asymptomatic (or nearly so) from a fixed deficit. However, as many as 25% of patients will have persistent symptoms from incomplete central compensation after an ablative vestibular surgical procedure. (Many patients with fluctuating disorders will be quick to point out that the symptoms of a fixed deficit are much more tolerable.) These

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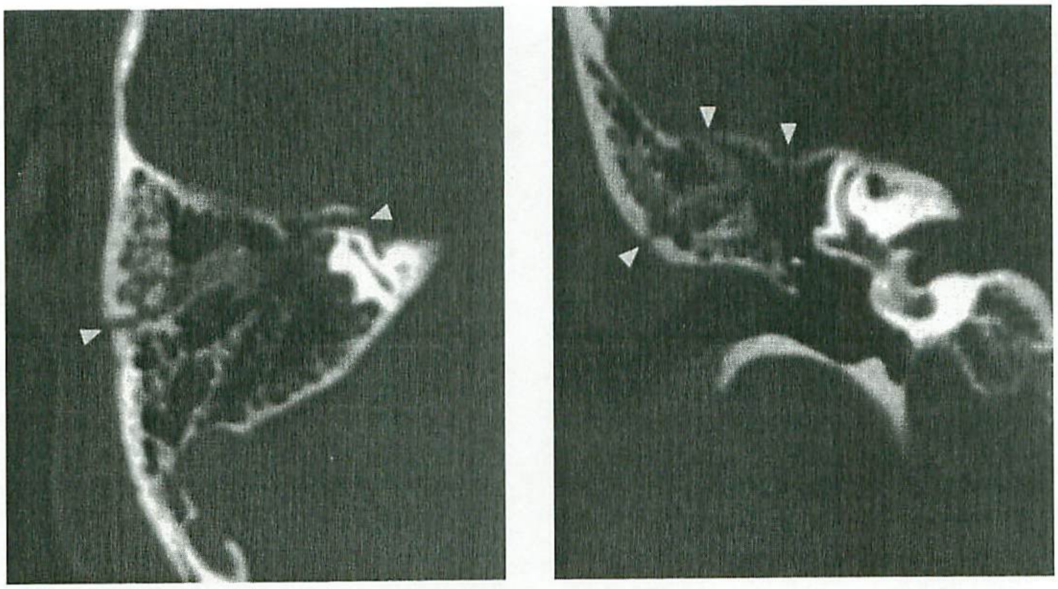

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FIGURE 19.2 Axial (**A**) and coronal (**B**) CT scan of the temporal bone demonstrating a longitudinal temporal bone fracture.

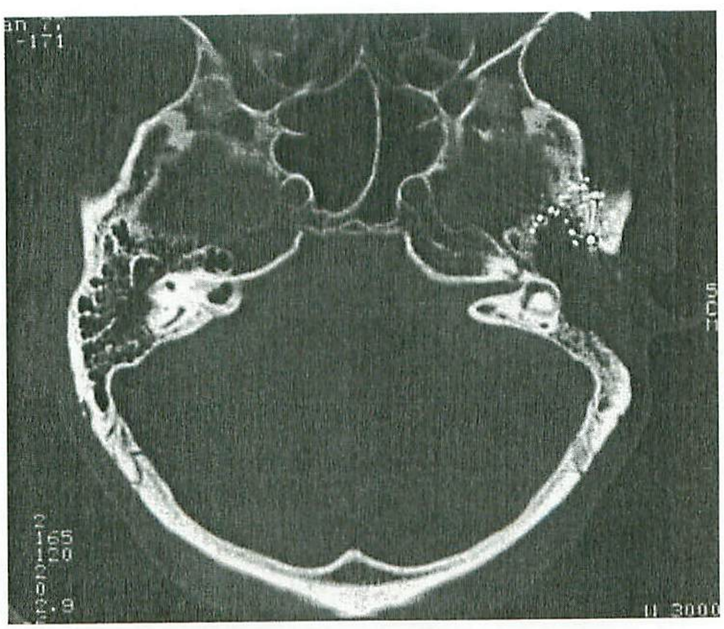

FIGURE 19.3 Axial CT scan of a temporal bone after a gunshot wound to the left mastoid demonstrating massive destruction to the area and residual metallic fragments.

procedures include labyrinthectomy, selective vestibular nerve section, and acoustic neuroma removal. The important differential diagnosis for these patients is determining whether complete elimination of vestibular function has indeed been accomplished and whether there is any disorder of the contralateral ear (most commonly seen in Ménière's disease). Treatment is directed at weaning the patient from vestibular suppressant medications and instituting vestibular rehabilitation exercises.

OTOTOXICITY

This is an entity more commonly known to occur as a bilateral process from systemic administration of ototoxic medications, but you will likely see this more often as the practice of intratympanic aminoglycoside perfusion (ITAP) becomes more popular. ITAP is becoming a popular minimally invasive method of ablating vestibular function of one inner ear with a relatively small risk to hearing. Additionally, aminoglycosides in ototopical medications administered in the face of a tympanic membrane perforation have been reported to have similar results, although this is a very rare occurrence.

Clinical Presentation

These patients will have a history of topical ototoxic medication administration and a defect in their tympanic membrane, or they will have a history of ITAP performed to ablate vestibular function. They will note varying degrees of disequilibrium and less commonly may have a history of vertigo during the onset of therapy. Hearing loss and tinnitus may also be present. Their history will include ITAP or ototopical medication use with perforated tympanic membrane and chronic disequilibrium.

Pathophysiology

The ototoxic medication is thought to pass through the round window membrane and become dispersed throughout inner ear fluid. Aminoglycosides are the primary offending medication. They initially produce a reversible blockage of transduction channels from the endolymph side of the inner ear hair cells. This is then followed by irreversible damage to the hair cell itself. Most ototoxic medications are more selectively toxic to the hair cells of the vestibular end organ than the cochlear hair cells, and consequently, vestibular dysfunction typically occurs before any clinically significant hearing loss is found. However, given in high enough doses and for a prolonged period of time, hearing loss and vestibular function will both occur. Topical aminoglycosides administered to an ear with a tympanic membrane perforation in the face of active infection very rarely show evidence of ototoxicity. It has been proposed that inflammation results in thickening of the round window membrane, which impedes absorption of the medication into the inner ear.

Diagnosis

The key to the diagnosis is obtaining a history of your patient receiving topical ototoxic medication. The patient will usually complain of vague disequilibrium that is exacerbated with quick head movements. Examination may show a perforation of the tympanic membrane and residue of prior infection. Audiometric examination may show a mixed or sensorineural hearing loss. ENG may demonstrate caloric weakness in the involved ear and may demonstrate spontaneous nystagmus. Treatment is discontinuation of the ototoxic medication and vestibular rehabilitation therapy.

SUPPURATIVE LABYRINTHITIS

Suppurative labyrinthitis is usually the result of otitis media but may also be seen as a complication of meningitis. The patient will present with an acute vestibular syndrome, similar to vestibular neuritis; however, in suppurative labyrinthitis, there is usually an associated sensorineural hearing loss and evidence of a bacterial infection such as fever, otalgia, or abnormal otoscopy. Confirmation of a middle ear infection and evaluation of other possibly associated complications (coalescent mastoiditis, epidural abscess, sigmoid sinus thrombosis) should be studied by radiographic imaging. It is imperative to make this diagnosis early because progression of the infection could result in meningitis and other ill sequelae. Additionally, loss of auditory and vestibular function may be averted by timely institution of appropriate medical and/or surgical intervention. Once the acute process has been stabilized, these patients may develop the symptoms of a chronic fixed unilateral lesion.

PSYCHOGENIC DIZZINESS

Psychogenic dizziness includes dizzy complaints that represent severe exaggeration of symptoms or are completely nonorganic in origin. Psychogenic dizziness probably represents 5% of all patients presenting to a vestibular center. This includes malingerers, patients with anxiety-driven symptoms, and Munchhausen's syndrome. It is listed under fixed deficits because the complaints of these patients often sound similar to the symptoms of a fixed deficit. Many of these patients will complain of chronic disequilibrium, tinnitus, and hearing loss, and most will have an obvious reason to exaggerate their symptoms. Often the motive behind their secondary gain is monetary–pending lawsuit, workman's compensation, or disability claim–but other motives may be present such as drugs and attention. Examination will show normal findings except for Romberg and Fukuda testing. Invariably, these patients will show wavering or falling on their Romberg and will stumble all about the room during the Fukuda stepping test. Their history and physical exam will usually defy diagnosis. ENG and audiometry will show normal or symmetric findings. Computerized dynamic platform posturography is most helpful in evaluating these patients and will demonstrate aphysiologic sway patterns in the majority of these patients. You must take care in labeling someone as psychogenic before you make an exhaustive search for physiologic abnormalities.

Suggested Reading

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