Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 4: Inner Ear

Chapter 49: Ménière's disease and Other Labyrinthine Diseases

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When a patient complains of dizziness, the physician first should ascertain whether the patient actually has vertigo. A patient with dizziness may be experiencing a sensation of giddiness, lightheadedness, disequilibrium, or imbalance. A patient with anemia occasionally describes a weak, faint feeling as a dizzy spell. Classically, vertigo is described as a sensation of twirling motion. It is unimportant whether the patient feels he or she is spinning clockwise or counterclockwise or has a rocking or an up-and-down sensation. It is also unimportant whether the environment or the patient seems to be spinning. Any hallucination of motion is vertigo.

Table 1. Types of Ménière's Disease and Other Labyrinthine Disorders

Ménière's Disease
Typical
Vestibular
Cochlear
Other Peripheral Diseases
Benign paroxysmal positional vertigo
Vestibular neuronitis
Labyrinthitis
Motion sickness
Otosclerosis
Syphilis
Cogan's syndrome
Posttraumatic vertigo
Perilymphatic fistula
Ototoxicity
Presbyvertigo
Intralabyrinthine hemorrhage
Central-Peripheral Diseases
Vascular loop syndrome
Vertebrobasilar insufficiency
Cervical vertigo
Tumors
Multiple sclerosis.

If vertigo can be proved, the cause is probably in the ear and not in the central nervous system or other organ systems. Eighty-five per cent of these patients have a peripheral disorder in the vestibular system; 15 per cent have a central disorder (Paparella, 1972). Ménière's disease is the most common cause of vertigo, followed by other, less common labyrinthine diseases. In the following sections of this chapter, we discuss Ménière's disease and other diseases, as outlined in Table 1. We believe that a detailed study of Ménière's disease is important, not only for better assessment of this common and often disabling pathologic condition but also as a key to improving the diagnosis and understanding of the whole group of peripheral diseases. Surgery for Ménière's disease and vertigo is discussed more fully in the next chapter (Chap 60).

Ménière's Disease

The term Ménière's disease refers to a group of symptoms including episodic vertigo, fluctuating sensorineural hearing loss, tinnitus, and a feeling of fullness in the affected ear. The pathologic correlate of this group of symptoms is endolymphatic hydrops. There are three important dates in the history of Ménière's disease - 1861, 1927, and 1938 - when outstanding progress was made in the description of this disorder and aspects of its pathology, pathogenesis, and treatment. In 1861 Prosper Ménière first described the disease in his series of six articles in the Medical Journal of Paris. His great contribution to understanding of the disease that bears his name was proving that the symptoms are of labyrinthine origin and not due to brain disease or "apoplectic cerebral congestion", the general term then in use of conditions with these symptoms.

In 1927, Guild clearly demonstrated longitudinal flow of endolymph, referring to the endolymphatic sac as a site of "outflow of endolymph" in guinea pigs. Portmann (1927) described surgery on the endolymphatic sac, for Ménière's disease. In the same year, Dandy popularized sectioning of cranial nerve VIII as treatment for vertigo in a surgical operation first successfully employed by Parry in 1904. The next most important advance was the description by Hallpike and Cairns (1938) of endolymphatic hydrops in two patients. It is of historical interest that both patients died as a complication of sectioning of the cranial nerve VIII, done to treat the Ménière's disease, thus providing the first opportunity to assess pathologic conditions in this entity. Subsequently, there appeared a number of excellent, well-documented reports on patients with Ménière's disease (Altmann and Fowler, 1943; Lindsay, 1946; Brunner, 1948).

In recent years the histopathologic findings in Ménière's disease have been described (Antoli-Candela, 1976; Fraysse et al, 1980; Paparella, 1984) as they occurred in large series of temporal bones. The most important of these findings was endolymphatic hydrops. Hydrops usually was found to involve both the scala media and otolithic organs. It was rarely seen segmentally in only the vestibular portion, but often in only the cochlear portion.

Natural History

Stahle and colleagues in Sweden (1978) estimated that Ménière's disease is four times more common than otosclerosis. Although extrapolating these proportions to figures for the USA is difficult, it is estimated that more than 2.4 million people in the USA have the disease. It is

likely that there are racial (genetic) as well as environmental factors that influence differences in incidence among countries and among various sections of countries. The disease is much more common in adults, with an average age of onset in the forties, the symptoms beginning usually between ages 20 and 60 years. We have seen typical Ménière's disease in many children as early as the age of 4 years, however, and in patients of all ages upward to the 90s as well. Ménière's disease is equally common in each sex (in some reports females outnumbered males by up to 1.3:1), and right and left ears are affected with fairly equal frequency (Meyerhoff et al, 1981).

The incidence of bilateral or unilateral Ménière's disease raises questions not only of pathogenesis but also of diagnosis and treatment. In a recent study, bilateral disease was definitely seen in one in three patients, and 78 per cent of all patients with Ménière's disease had a hearing loss in the contralateral ear (Paparella and Griebie, 1984). A peak audiometric configuration was seen in half the patients, and a slight peak (incipient Ménière's disease?) was commonly seen in the "normal" ear of those with unilateral cases. Other common audiometric patterns include low-frequency losses in early Ménière's disease and flat losses in advanced Ménière's disease. A certain subset of patients will progress to complete deafness. Less common audiometric patterns include high-frequency losses and trough-shaped losses.

These patients were younger (average age 40 years) and, if one could estimate incidence over full life-spans, it seems likely that at least half of these patients will develop difficulty in the opposite ear (Paparella, 1984). This is in keeping with reports by others (Balkany, 1980). In our study (Paparella and Griebie, 1984), half of the patients who developed Ménière's disease in the second ear did so 5 years after onset in the first ear. The frequency of bilaterality in Ménière's disease dictates that, when considering therapy, every attempt should be made at conservation.

The major categorical triad of symptoms of Ménière's disease includes vestibular symptoms, auditory symptoms, and aural pressure. Usually this triad occurs together, whereas in many patients vestibular symptoms alone or auditory symptoms alone may precede the development of other symptoms for many months or years. Episodic vertigo associated with vegetative symptoms is the most disabling symptom (96.2 per cent). In 25 per cent of patients, vertiginous attacks lasted less than 1 hour, in 50 percent 1 or 2 hours, and in 25 per cent more than 2 hours to a day or so. Hearing loss is found in 87.7 per cent of cases, tinnitus in 91.1 per cent, intolerance of loudness in 56 per cent, and diplacusis in 43.6 per cent. Aural pressure in one or both sides is related in 74.1 per cent of cases (Paparella, 1984).

As has been described by Jongkees (1983) and others, Ménière's symptom-complex was divided into two categories: Ménière's syndrome, in which a known cause exists, and Ménière's disease, in which the cause seems idiopathic. Recently, cases of Ménière's symptom-complex that had a likely or known cause have been described on the basis of clinical records and findings in the temporal bones; these cases evolved subsequent to syphilis (Paparella et al, 1980), infection (chronic otitis media, meningitis), trauma, and otosclerosis (Paparella and Sajjadi, 1987). Those findings have led us to our current classification of Ménière's disease: We now divide Ménière's disease into that with known causes or contributing factors (extrinsic) and that with multifactorial

inheritance, as in Table 2. We consider the concept "idiopathic" defeatist; it is clear that as we understand more about the etiologic factors in endolymphatic hydrops, "idiopathic" will, we hope, become a discarded term.

Table 2. Ménière's Symptom-Complex (Vestibular Symptoms, Cochlear Symptoms, and Aural Pressure)

Causes of Ménière's Disease*

Genetic causes
Infection (inflammation - bacterial, viral, etc)
Otosclerosis
Trauma (physical or acoustic)
Syphilis
Others, including allergy, tumors, leukemia, and autoimmune disorders

Subsets of Ménière's Disease

Typical (Classic)
Triad of vestibular symptoms, cochlear symptoms, and aural pressure

Atypical
Vestibular Ménière's disease (vestibular symptoms and aural pressure)
Cochlear Ménière's disease (cochlear symptoms and aural pressure).

* Cause = multifactorial inheritance; this can include extrinsic contributing causes and/or an underlying intrinsic (genetic, developmental) cause.

In approximately one-fourth of our patients, likely extrinsic cause of Ménière's disease were found. Up to 20 per cent of patients had a positive family history of symptoms suggestive of Ménière's disease in blood relatives (Paparella, 1985); thus, genetic predisposition to Ménière's disease remains a possibility for some patients. In accordance with findings of other authors (Wheel et al, 1974; Wilbrand and Stahle, 1981), we found that patients with Ménière's disease generally displayed hypocellular mastoids and deficient radiographic representation of the vestibular aqueduct. Trautmann's triangle was often reduced in size or distorted, primarily because of the nature of the lateral sinus, which was displaced anteriorly and more medially in almost all patients with Ménière's disease.

As shown in Table 2, Ménière's disease can be broken further into two subsets: typical Ménière's disease, in which a patient develops the full-blown symptom-complex; and atypical, in which either vestibular symptoms (vertigo) or cochlear symptoms (hearing loss) appear first and full-blown Ménière's disease does not develop. Cochlear Ménière's disease is recognized as a fluctuating, progressive, sensorineural hearing loss associated with aural pressure in the absence of vestibular symptoms or findings (Williams et al, 1947). Vestibula Ménière's disease (Harker

and McCabe, 1980; Paparella, 1985) is characterized as the occurrence of episodic vertigo and disequilibrium associated with pressure in one nystagmographic testing. Patients have presented with incapacitating vestibular Ménière's disease of more than 45 years' duration who never developed cochlear deafness.

Pathologic Findings

A detailed study was recently complete of the histopathology of cases of Ménière's disease that have been described in the literature since the classic description of endolymphatic hydrops in two cases by Hallpike and Cairns in 1938 (Paparella, 1985). The pathologic findings of 48 temporal bone cases were well documented and included clinical records. These findings can be summarized as follows.

Cochlear hydrops was seen in all cases and saccular hydrops in most, while utricular hydrops was rare. The endolymphatic space bulged in the helicotrema in approximately half of the cases, whereas a saccule bulging into the semicircular canal (usually the horizontal semicircular canal) was seen in approximately one-third of the cases. Endolymphatic hydrops obliterated and displaced perilymphatic spaces in the scala vestibuli and cisternae vestibuli in most cases. The saccule bulged against the footplate in approximately 60 per cent of cases. The end-organs (ie, the organ of Corti and vestibular end-organs) were described as intact and presumably normal under light microscopy. Ruptured membranes were described in 16 of 48 cases (33 per cent). Of these, severe were described in the cochlea (Reissner's membrane), five in the saccule, and seven in the utricle.

The pathologic findings in 16 temporal bones from 12 patients with clinical histories of Ménière's disease from the University of Minnesota's human temporal bone collection, which were reported by Paparella (1984), are generally similar to those described in the literature; ruptured membranes, however, were observed only in the group with severe hydrops. The series reported by Antoli-Candela (1976) included 19 temporal bones from 14 patients. Cochleosaccular hydrops was seen in all cases, and ruptured membranes in 13 of 19 cases. Another series described by Fraysse and colleagues (1980) included 23 temporal bones from 17 patients. Cochleo-saccular hydrops was present in most cases, and ruptured membranes were described in three of 23 cases.

Recently, Okuno and Sando (1987) described 22 temporal bones from 16 patients with Ménière's disease. Endolymphatic hydrops was more often observed in the pars inferior than in the pars superior of the temporal bone. Interestingly, in addition to hydrops, collapse of membranes was also seen in many cases with hydrops. Eight of 22 cases had no pathologic findings in the pars superior.

Most ears with Ménière's disease show no loss in population of hair-cells in the organ of Corti or in the vestibular sense-organ (Schuknecht and Igarashi, 1986). This study did describe loss in the population of cochlear neurons, occurring in the apical region of the cochlea in about 10 per cent of ears with Ménière's disease, but rarely a loss of neurons in the vestibular system.

Etiopathogenesis

What is the cause and pathogenesis of Ménière's disease? There are no definite answers to this question. However, many identical features are presented by the variety of forms of what we have formerly referred to as Ménière's syndrome and Ménière's disease - but herein refer to with one term, Ménière's disease, for cases with causes both known and unknown. This similarity in features suggests a common etiologic explanation.

Assuming that some forms of Ménière's disease have identifiable causative agents (most do not) and assuming developmental anomalies related to the endolymphatic duct and sac to be common findings in this disorder, then which one common explanation best accounts for all forms of Ménière's disease? It is our belief that a multifactorial inheritance is the best answer, and the conditions so created in turn lead to endolymphatic malabsorption and then to hydrops. There is clinical and laboratory evidence to support this concept.

Recently, cases in patients with Ménière's symptom-complex with a cause likely or known have been described on the basis of clinical records and findings in temporal bone; these cases had evolved subsequent to infections, chronic otitis media (both active and inactive), inflammation (eg, following involvement with viral labyrinthitis), otosclerosis, trauma, syphilis, allergy, leukemia, and autoimmune disease (see Table 2).

Endolymph comes primarily from the stria vascularis. The planum semilunatum and dark vestibular cells may contribute a small amount. Endolymph may also derive from perilymph across labyrinthine membranes. Gradually, endolymph is absorbed in the endolymphatic duct and sac - a biologically active transporting structure where absorption primarily occurs as well as, perhaps to a lesser degree, secretion. The evidence strongly suggests that the concepts of both longitudinal flow (slow process) and radial flow (rapid, ongoing process) are concurrently operational. Both hydrostatic and osmotic pressure gradients are also simultaneously operational.

The basic concept in the pathogenesis of Ménière's disease is that of the "lake-river-pond". In the normal condition, there is no disturbance of the rate of flow. If there is any obstruction ("dam") near the "river" or "pond", however, endolymphatic flow is obstructed due to physical or chemical causes, and hydrops develops. The fundamental problem in Ménière's disease of known or unknown cause appears to be endolymphatic *malabsorptive* dysfunction, the site being the endolymphatic duct or sac. In Ménière's disease, this is a slowly building process that takes many years to develop. Thus, all forms of Ménière's disease are developed after some inciting cause or causes that may have occurred years earlier, and thus all are "delayed". When the endolymphatic duct is mechanically obstructed, as in fracture following trauma to the head, the development of endolymphatic hydrops is hastened. Thus, endolymphatic hydrops in Ménière's disease is a primary dysfunction of longitudinal flow.

Studies in temporal bones have revealed perisaccular fibrosis (Altmann and Fowler, 1943; Zechner and Altmann, 1969), loss of epithelial integrity and atrophy of the sac (Arenberg et al, 1970), hypoplasia of the vestibular aqueduct (Sando and Ikeda, 1984), and narrowing of the

lumen in the endolymphatic duct (Ikeda and Sando, 1984). However, because blockage of the duct and hypoplasia of the sac are not seen in all cases of Ménière's disease, it seems that other mechanisms may contribute to the development of endolymphatic hydrops. Stahle and Wilbrand (1983) described characteristic features in patients with Ménière's disease as a lack of periaqueductal pneumatization, a lack of pneumatization medial to the arcuate eminence, a short vestibular aqueduct, a narrow external aperture of the vestibular aqueducts, and a reduction in size of the mastoid air-cell system. Sando and Ikeda (1985) described the poor pneumatization of the temporal bones of patients with Ménière's disease. Paparella and Sajjadi (1989) described the characteristic finding of an anteriorly and medially displaced lateral sinus in patients with Ménière's disease.

The long-term endolymphatic malabsorption likely relates to a developmental abnormality of the endolymphatic duct and sac in association with displacement of the lateral sinus. Gussen (1982) reported that unimpeded venous drainage of the vestibular organs via the paravestibular canaliculi vein is crucial to inner ear fluid mechanisms. Vascular anomalies, especially abnormality in venous drainage, may be associated. Thus, the quantity of endolymph is affected.

Insights into the pathogenesis of Ménière's disease have been achieved by observation of cases of revision of surgical procedures for enhancement of the endolymphatic sac (Paparella and Sajjadi, 1989). The most remarkable findings at the time of revision were osteoneogenesis, fibrosis, and granulation tissue resulting in an exceedingly tight and constricted dura, Trautmann's triangle, and sac region, with obvious pressure and obstruction in this region. In most instances, after correction of these lesions and decompression or loosening of the dura, the patient once again had elimination or control of symptoms of Ménière's disease. These findings appear to substantiate the observation that the pathogenesis of Ménière's disease is basically correlated with endolymphatic malabsorption.

Genetic Ménière's disease was seen in up to 20 per cent of 500 patients with Ménière's disease who, in response to a recent questionnaire, indicated a positive family history (Paparella, 1985). We believe that future research into the genome will lead to descriptions of developmental conditions predisposing to Ménière's disease. A recessively inherited condition or an incompletely penetrant dominant - as well as multifactorials - could explain the vagaries of structures found in this disease and could help lead to explanations of the developmental abnormalities of Trautmann's triangle and the endolymphatic absorptive system. It is likely, as accumulating information suggests, that we are dealing with a multifactorial inheritance that is a collaboration of multiple genetic and nongenetic (environmental) factors in the causation of this particular disorder.

Extrinsic Contributing Factors

Otitis Media and Ménière's Disease

Clinical observations of patients with fluctuant sensorineural hearing loss following or occurring with chronic otitis media led to the hypothesis that hydrops can result from chronic

otitis media (Paparella et al, 1979; Paparella et al, 1983). In animal models, endolymphatic hydrops occurring with otitis media was observed by Kimura (1982). Endolymphatic hydrops has been observed as a common occurrence in human cases of suppurative or serous labyrinthitis, whether of otogenic or meningogenic types. A study of 560 temporal bones held in our laboratory demonstrated 109 with endolymphatic hydrops; 194 evidenced otitis media. Of these 560 bones, 75 contained histopathologic evidence of both otitis media and endolymphatic hydrops. These numbers provide supporting evidence that the coincidence of otitis media and endolymphatic hydrops in the same temporal bone is sufficiently frequent to support the hypothesis that the two pathologic conditions may indeed be dependent on one another.

It is of interest that, in the classic description by Hallpike and Cairns in 1938, one of the first cases of Ménière's disease was associated with extensive pathologic findings of chronic otitis media. Indeed, those authors discussed ways in which hydrops could have evolved from infection passing through the round window membrane. In our opinion, the pathogenesis of Ménière's disease resulting from previous otitis media includes considerations of infectious products spreading through the round window membrane, as well as developmental aberrations of the endolymphatic duct and sac. Enzymes and other toxins from otitis media can invade the perilymphatic spaces, primarily through the round window, and then the endolymphatic spaces, through perforations in the basilar membrane or through Reissner's membrane. An imbalance in osmotic pressure due to alterations in electrolytes could explain the acute hydrops associated with otitis media.

In considering the pathogenesis of long-term endolymphatic hydrops after otitis media, one should consider how quality as well as quantity of endolymph can influence hydrops. Serous labyrinthitis of the perilymphatic spaces can readily change the chemical composition of endolymph across Reissner's membrane or, as is less likely, the basilar membrane. Such a change of chemical (electrolytic) or possibly physical composition (in the stria vascularis) and/or of absorbability of endolymph (in the endolymphatic sac) might contribute to hydrops.

Another means by which otitis media, especially early in life, might lead to Ménière's disease later in life is by influencing development of the endolymphatic duct and sac. Hypocellularity and development disturbances of Trautmann's triangle can theoretically disturb development of the endolymphatic duct and sac including its blood supply. Osteitis from mastoiditis can affect the adjacent developing sac and the blood supply in Trautmann's triangle, and thus this too could play a role.

Otosclerosis and Ménière's Disease

It is well documented that patients with otosclerosis may occasionally develop vestibular symptoms including fluctuating sensorineural hearing loss, in addition to the conductive hearing loss typical of otosclerosis plus aural pressure (Paparella and Chasin, 1966; McCabe, 1966; Sando et al, 1974; Cody and Baker, 1978; Ghorayeb and Linthicum, 1978; Igarashi et al, 1982; Liston et al, 1984). The pathogenesis that best explains how otosclerosis can lead to endolymphatic hydrops years later is related to long-term malabsorption of endolymph in the endolymphatic duct

and sac. Otosclerosis can envelop the vestibular aqueduct, resulting in malfunction of the endolymphatic duct and sac, or otosclerosis can invade the endosteum, altering chemical characteristics of perilymph and endolymph, thus affecting characteristics of radial as well as longitudinal flow of endolymph.

Trauma and Ménière's Disease

Trauma, either physical or acoustic, can play a role in the development of a certain number of cases of Ménière's disease (Paparella and Mancini, 1983). Although attempts have been made to exclude trauma as coincidental and not causative, we have some well-documented cases of Ménière's disease in which it is difficult to exclude the importance of the role of trauma when considering the chronological sequence of events. For consideration of the pathogenesis of endolymphatic hydrops associated with Ménière's disease, acoustic and physical trauma are considered together as one pathogenic factor. Trauma might in some way lead to biochemical dysfunction of cells producing endolymph or, more likely, cells involved in absorption of endolymph. We believe that trauma contributes to passage or dysfunctional absorption of endolymph. Trauma causing a shock to the membranous labyrinth may result in displaced epithelia of the sensory end-organs and other cellular elements including the otoconia of the saccule and utricle. Such cellular debris could mechanically or chemically cause decrease in endolymphatic absorption through the endolymphatic duct, cause ensuing endolymphatic hydrops, and thus cause Ménière's disease.

Syphilis and Ménière's Disease

Pulec (1972) reported that 7 per cent of his patients with Ménière's symptom-complex were found to have a syphilitic etiologic agent. Luetic hydrops may be seen in both congenital and acquired syphilis. Karmody and Schuknecht (1966) reported that 38 per cent of 123 congenital syphilitics had a hearing loss, and most of them had associated vestibular symptoms with episodic vertigo and diminished caloric responses. This report demonstrated a case of endolymphatic hydrops due to syphilitic osteitis of the otic capsule. Endolymphatic hydrops secondary to invasion of the labyrinthine capsule may have some resemblance to the changes reported in Ménière's disease and may account for the occasional similarity of symptoms. Schuknecht (1974) suggested that the inner ear's reaction to lues is characterized by progressive endolymphatic hydrops and degeneration of the membranous labyrinth, resulting in rupture of the membranes due to overaccumulation of endolymph.

Allergy and Ménière's Disease

Allergy is reported to be an etiologic factor in Ménière's disease by some investigators (Duke, 1923; Endicott and Stucker, 1977). The possible role of IgE in Ménière's disease has been proposed. However, Stahle and colleagues (1974) reported normal serum IgE levels in patients with Ménière's disease.

Leukemia and Ménière's Disease

Leukemia is also reported to have a relationship to endolymphatic hydrops (Lindsay, 1946; Paparella et al, 1973). Sando and Egami (1977) reported endolymphatic hydrops in the cochlea and saccule, and a relatively narrowed vestibular aqueduct and endolymphatic sac, in a leukemic patient. This suggests that leukemic infiltration might be considered as a possible cause of Ménière's disease.

Autoimmune Disease and Ménière's Disease

Autoimmune inner ear disease was first reported by McCabe (1979). Studies in animals showed that autoimmunity to type II collagen could be hypothesized as an etiopathogenetic agent in Ménière's disease (Yoo et al, 1982; Yoo, 1984). Harris and associates (1986), however, failed to induce type II collagen autoimmunity in an animal study. Hughes and coworkers (1983) reported that some cases of presumed idiopathic Ménière's disease are autoimmune in origin.

Pathophysiology

Any explanation for the clinical symptoms of Ménière's disease should account for all of the symptoms, including rapid or prolonged attacks of vertigo, disequilibrium, positional vertigo during and between attacks, fluctuating progressive sensorineural hearing loss, tinnitus, aural pressure, loudness intolerance, and diplacusis. The attacks of Ménière's disease (vertiginous, cochlear, and aural pressure) have been explained throughout the literature on both physical and chemical grounds. Membranous ruptures have been described as a cause of all attacks of Ménière's disease (Schuknecht, 1968; Schuknecht and Igarashi, 1986). A recent review of human temporal bones with endolymphatic hydrops from our laboratory, and of other well-documented cases of human temporal bones in Ménière's disease, demonstrated, as mentioned, that some cases show no evidence of rupture.

Ruptures, when present, are not necessarily seen in the scala media and in the saccule or utricle simultaneously. Furthermore, while considering theories of longitudinal (slow) and radial (fast) dispersal of fluid, if a rupture occurred - for example, in Reissner's membrane in the middle turn - it is hard to explain how endolymph rich in potassium could bypass the saccule to reach the crista ampullaris of the semicircular canal and cause vertigo instantaneously. In a typical attack of Ménière's disease there would have to be simultaneous ruptures in the cochlear duct and the saccule for both cochlear deafness and vertigo to occur together and each time, but pathologic studies do not substantiate this. It should be recalled that patients enjoy relief of the attack when the pressure subsides and when the vertigo and cochlear dysfunction improve, suggesting that ruptures most usually alleviate and do not incite vertiginous episodes. Except for isolated examples, the evidence from many well-described histopathologic studies shows no ruptures or, when they are present, such ruptures are more often seen in the cochlear duct and not in the saccule or other vestibular structures. Vosteen and Morgenstern (1986) explained that the disturbance in balance of ions between production and resorption of endolymph is primary in generating hydrops and its symptoms.

Saccular hydrops can be so severe as to extend into the semicircular canal. Such distention can physically alter function of the crista ampullaris, causing instantaneous vertigo. The presence of such distention can also help explain positional vertigo in patients with Ménière's disease. Hydrops can mechanically alter traveling waves, and this would explain certain aspects of cochlear dysfunction (Tonndorf, 1968). Tonndorf (1983) has described the available evidence that suggests that vestibular symptoms in Ménière's disease can be caused by mechanical factors, as are their auditory counterparts. Thus, both mechanical and chemical factors appear to provide the best explanations for the pathogenesis of the symptoms of Ménière's disease.

Characteristically, in advanced Ménière's disease, endolymphatic hydrops is seen in the pars inferior - that is, in the scala media and in the saccule, which fills the vestibule and scala vestibuli in many cases. Because of the displacement (loss) of perilymph in advanced (decompensated) Ménière's disease, radial flow diminishes and longitudinal flow becomes dominant. Less often the utricle or cochlear duct will extend to occupy the vestibule. The saccule in Ménière's disease seems to function as a reservoir for endolymph trying to reach the endolymphatic duct and sac for the process of outflow. Stagnation of endolymphatic outflow can occur and such distention can mechanically interfere with traveling waves, thus altering cochlear function.

In typical Ménière's disease, cochlear and vestibular structures are similarly involved, due to a dysfunctional endolymphatic duct and sac. In atypical Ménière's disease, secondary obstructive sites can occur in addition. In cochlear Ménière's disease, for example, it is possible that obstruction occurs at the ductus reuniens. In an animal study, Kimura and colleagues (1980) obliterated the ductus reuniens and found that cochlear hydrops developed. Although malabsorption of endolymph relating to dysfunction of the endolymphatic duct and sac is the most likely underlying cause, excessive production of endolymph may play a role. For example, the planum semilunatum and dark vestibular cells may be more active than the stria vascularis in producing endolymph in vestibular Ménière's disease. In vestibular Ménière's disease, the utriculoendolymphatic valve may be deficient, leading to utricular hydrops to the exclusion of hydrops of the pars inferior (Schuknecht and Belal, 1975; Paparella and Mancini, 1985).

Clinical Manifestations

Episodic vertigo associated with vegetative symptoms such as nausea and vomiting is the most disabling symptom of Ménière's disease and the one for which medical attention is usually sought. Vertiginous episodes lasting 20 minutes, hours, or a day as definitive spells punctate an otherwise normal state of equilibrium. The pattern of occurrence of these spells varies, but commonly there is a gradual increase in frequency until a maximum is reached over a period of years; then the frequency decreases as the disease begins to run its course and destroy the inner ear. Some patients exhibit long periods of remission between attacks, with or without therapy. Some show a clustering of attacks with long symptom-free periods in between. Tension and anxiety tend to precipitate attacks, and excessive intake of salt will occasionally do the same. Imbalance is very common between attacks.

A definitive spell may be preceded by an aura that usually consists of tinnitus and/or aural fullness or stuffiness. The spell begins suddenly with a severe whirling sensation accompanied by pallor, diaphoresis, prostration, nausea, and frequently vomiting. The slightest motion of the head accentuates the symptoms. During the attack, the patient is completely oriented to his or her surroundings and has no neurologic deficit such as paresthesia, diplopia, loss of consciousness, weakness or paralysis, or dysarthria. Vision may be blurred because of the nystagmus.

Over the ensuing minutes or hours there is a gradual abatement in symptoms and the patient often falls asleep. Frequently, upon awakening, the spell is over and the feels entirely normal (unless there has been considerable vomiting). In other cases the vertigo while at rest is gone, but intolerance for motion, and unsteadiness, persist for a day or two. As years pass, if the disease remains active, function of more and more of the membranous labyrinth is destroyed, and the spells become less severe.

Positional vertigo is common in these patients not only during an attack but also between attacks. There may be adjunctive spells between the definitive spells. These only last a few seconds and are characterized by a momentary sensation of loss of balance, often described as "missing a step". They may be uncommon or occur several times a day. Unlike the definite spells, these adjunctive spells are usually provoked - by quick motions of the head, looking up, or cornering. This aspect of the disease may bother the patient psychologically because it is commonly assumed that the patient is inebriated.

Rarely, patients also have attacks of such explosive onset and severity that they fall violently to the ground. When this happens there is no preceding aura and no loss of consciousness; it is as though something or someone had propelled the subject to the ground. The duration of these drop-attacks is very short. Without much objective evidence, they have been termed "utricular crises" or "falling spells of Tumarkin" (Tumarkin, 1936).

Auditory symptoms include hearing loss, tinnitus, intolerance of loudness, and diplacusis. The decrease in auditory acuity is typical of the cochlear type of sensorineural loss. Sounds are distorted in the affected ear and are perceived as tinny by the patient. This distortion is always present during a spell and frequently occurs between them. The same sound frequently elicits a different pitch-percept in the two ears (diplacusis binauralis dysharmonica), which is usually higher in the affected ear. Loud sounds are intolerable or even painful to the patient, due at least in part to recruitment. The almost universal sensation of fullness or pressure during the spells is sometimes relieved by termination of the spell.

The hearing loss may fluctuate considerably during the course of this disease, especially in the first year or two. If the disease remains active over the years, fluctuation becomes less and hearing level gradually declines. During a definitive spell, acuity is always decreased and may remain so afterward, although dramatic improvement may occur occasionally with the passing of a spell, especially a falling spell of Tumarkin. This paradoxical improvement in hearing following an acute attack is sometimes referred to as Lermoyez's syndrome (Mawson and Ludman, 1979).

Tinnitus is described in a variety of ways; it can be exasperating and generally accompanies hearing loss. Continuous or intermittent, it is nonpulsatile and not affected by carotid pressure. Tinnitus may be the first symptom of Ménière's disease, preceding all others by years, or its first appearance may be with the first spell. It is always present during a spell if the patient is able to listen for it, and usually is present to some degree between spells. While little objective information is available, certain generalizations can be made: (1) the pitch tends to be related to the region of the most severe hearing loss - most commonly, low-frequency tinnitus is associated with low-frequency hearing loss; (2) its magnitude is roughly proportional to the severity of the hearing loss, although in the late stages of the disease the tinnitus may be less bothersome to the patient; (3) prognosis for amelioration is better if treatment is instituted early in the course of the disease.

Pressure is a remarkable symptom. Most times it is an aural pressure, but the patient may feel it in any part of the head or even the neck, during an attack. Headache is another symptom that sometimes can be picked up in the patient's history. In our experience, symptoms in decreasing order of importance and according to the frequency with which they register as patients' chief complaints are (1) vertigo, including typical attacks of vertigo as well as positional vertigo, (2) progressive sensorineural hearing loss, (3) tinnitus, (4) aural pressure, (5) loudness intolerance, and (6) diplacusis. For clinical use and comparison of pre- and post-treatment conditions, a system based on subjective observation, while imperfect, probably represents the best available method for reporting disability (Pearson and Brackmann, 1985) (Table 3).

Table 3. Classification of Disability Resulting from Ménière's Disease

Grade I

No or mild disability with intermittent or continuous dizziness/unsteadiness that precludes working in a hazardous environment.

Grade II

Moderate disability - intermittent or continuous dizziness/unsteadiness that results in a sedentary occupation.

Grade III

Severe disability - disabling symptoms that exclude gainful employment.

Clinical History and Diagnosis

Vertigo is almost always the result of a peripheral labyrinthine disturbance. Even if intracranial pathologic conditions exist, vertigo can result from a peripherally mediated response. For diagnosis of its origin, therefore, even in this day of high technology, the "art" of medicine still reigns; this is particularly true for Ménière's disease, for which the art of history-taking will,

more than anything else, provide the diagnosis of Ménière's disease or, at the least, the differential diagnosis of Ménière's disease with other peripheral labyrinthine disorders. A careful, detailed history is the most important guide to correct diagnosis and, indeed, in 90 per cent of cases with a good and complete history, one can make the diagnosis. Audiograms first and vestibular studies next are secondarily helpful. For this reason, in this section we describe aspects of how we personally apply the history-taking process and our preliminary workup.

As we have seen from study of its natural history, Ménière's disease is usually typical, with deafness and vertigo, or it can be an atypical subvariety having vertigo alone (vestibular Ménière's disease) or hearing loss and cochlear symptoms alone (cochlear Ménière's disease). Symptoms, and therefore questions, relating to Ménière's disease fall into three categories: vestibular, cochlear, and aural pressure.

Sometimes it is difficult to take a useful history from a patient with vertigo. The patient may have anxiety, may have seen many doctors, may have used many medications, and so forth. The patient is carefully instructed that specific questions will be asked and that a one- or two-word answer is best. The patient is advised that after these specific questions he or she will have ample opportunity to discuss everything about his or her problem. This method abbreviates the process and helps provide relevant diagnostic information. It is interesting that after these directed questions are asked and specific information is provided, the patient often has little or nothing to add to the history. The patient's chief complaint is sought for; it is usually vertigo and/or deafness, but it might be tinnitus, pressure/headache, or, rarely, loudness intolerance or diplacusis. The history starts with the chief complaint and follows with the patient's next problem, and next, and so forth. Questions are asked in each of the three categories of symptoms.

Vestibular Symptom Category

Let us assume that dizziness is the chief complaint, as is often true. The patient is asked when he or she experienced the first episode during his or her lifetime. Then the patient is asked about episodes of dizziness following the initial episode: are they paroxysmal, do they come and go? Is the dizziness vertigo - that is, is there any sensation of motion? Often there will be a spinning or twirling motion, although motion can consist of an up-and-down or sideways or a dropping sensation or "drop-attack", as described by Tumarkin (1936). The length of the vertiginous episode is defined.

Although there is variability, the frequency as well as the severity of these spells is assessed by specific questions. Typically the patient will say the vertigo will last for an hour or so and may be accompanied by nausea and vomiting. The patient may also experience imbalance or disequilibrium, which can continue to incapacitate him or her for hours or days afterwards. Sometimes the patient will say that he or she was incapacitated for a week or more, and by specific questioning we find that the patient had true vertigo in the beginning of that period of time, followed by a prolonged period of imbalance. These periods of imbalance or disequilibrium often may last for a day or so, but usually the entire episode lasts a matter of several hours. It is invariably true that motion or changing the position of the head during an attack will make the

vertigo or vestibular upset even more severe. Patients are also routinely asked if they have positional vertigo in between attacks, and it is surprising that many of them do indeed describe positional vertigo between attacks. Thus positional vertigo is almost always present during the attack and frequently in between episodes of vertigo, as well.

Some patients will describe a sense of imbalance or a sense of disequilibrium and not absolute vertigo with a twirling sensation. This too can result from Ménière's disease. Some patients will describe nausea or a chronic state of nausea, and this also can be a part of Ménière's disease. In addition, the patient is always asked whether there are any factors such as anxiety, allergies, and so forth that seem to trigger an attack; this happens in many patients. Frequently, patients will notice that visual stimuli, such as looking at moving objects such as in a shopping center, will help stimulate their vertigo, as will changing the position of the head. Patients will often have a sense of pressure and a chronic sense of imbalance and state that they are "always dizzy"; however, when asked specifically if there are periods or peaks when the dizziness is more severe, one can often extract a history of vertigo that is superimposed on top of the feeling of having constant imbalance. This is particularly noticeable in patients who have vestibular Ménière's disease.

To summarize the vestibular symptoms in Ménière's disease, first is episodic paroxysmal vertigo; second is a feeling of imbalance or disequilibrium, which can last for a longer period of time; third is positional vertigo, which exacerbates an attack of vertigo or can occur in between attacks; and last is the sensation of nausea, which can be intermittent or more chronic and can be accompanied by vomiting when the attack of vertigo is at its peak.

Cochlear Symptom Category

Classic cochlear symptoms in Ménière's disease consist of (1) progressive fluctuating sensorineural hearing loss, (2) tinnitus, (3) loudness intolerance, and (4) diplacusis.

The patient is asked when he or she first noticed the onset of hearing loss. Did it precede, follow, or accompany the onset of vertigo? Does the hearing loss worsen when there is vestibular upset? Has it gotten worse over time? Has the problem of hearing loss and vertigo together been more of a problem in recent months and years in spite of the medication? Was the hearing loss sudden in onset or gradual? Sometimes it is difficult to differentiate perilymphatic fistulae from Ménière's disease because of the sudden onset which some patients describe. Typically the hearing loss will be more profound when there is an attack of vertigo, but this is not always the case. The patient, of course, is asked if there is a hearing loss on one or both sides.

The next line of questioning has to do with tinnitus: when did the patient first notice the tinnitus? How does it correlate with the other symptoms in terms of frequency and intensity? What are the nature, characteristics, and severity of the tinnitus? Often the tinnitus will be constant, and even though it is constant there will be peak periods of exacerbation, often occurring with episodes of Ménière's attacks. Is the tinnitus in one or both ears?

Does the patient notice that he or she cannot tolerate loudness? For example, will loud sounds cause actual pain or discomfort in one or both ears? Is this more noticeable during an attack of Ménière's disease?

Finally, questions are asked regarding diplacusis. Is there any noticeable sensation of change in quality of hearing, such as change in pitch? Is there an octave difference (higher) in one ear (that is, the diseased ear) compared with the other? This often is true. Some patients will describe a sensation of hollow sound or a variety of other unusual auditory sensations. Do all of these auditory symptoms correlate with episodes of vertigo? Sometimes they will and sometimes they will not.

Aural Pressure

From pathologic studies, we see that hydrops of the pars inferior (the saccule and the cochlear duct) is the most significant feature. Presumably, therefore, this is the source of pressure. Pressure is almost always present in patients who have Ménière's disease and should be specifically asked for. When did the patient first notice pressure? Is it in the region of the ear? Is it on the side of the head? Does it coincide with the cochlear and vestibular episodic attacks of Ménière's disease? Is the pressure there all the time? Very often, patients will have such severe pressure that they will actually describe it as a dull headache. It also should be asked for.

Often synonyms need to be used when asking questions: does your ear feel plugged up? Do you have a headache? Patients often will have pressure all over the head: in the forehead, in the region of the eyes, and located generally in the region of the head, which seems to worsen with episodes of vertigo. Patients often will have a sensation of pressure in the neck, especially on the involved side. Aural pressure usually comes and goes and characteristically is present during episodes of vertigo, but sometimes it is present in between episodes as well. Being specific about the presence of aural pressure and documenting this in the history is particularly important in diagnosing vestibular Ménière's disease, which may ultimately evolve into typical Ménière's disease with cochlear symptoms.

Having asked the specific questions in the three categories concerning duration and time, one asks about the various medications the patient has used; what has benefited the patient in the past will guide our choice of medications to recommend in the future.

Also, we ask specifically if the patient has any family history of Ménière's disease - that is, dizziness and deafness. As many as 20 per cent of patients will give a positive family history of other members having symptoms suggestive of Ménière's disease. From the studies of pathogenesis, we believe there is evidence to suggest there is a genetic predisposition to the so-called "idiopathic" endolymphatic hydrops form of Ménière's disease.

We always ask whether the patient has had any other injuries to the ears prior to the onset of the Ménière's disease: have there been any severe blows to the head, any skull fractures? Have there been any severe infections? Have there been any other ear diseases? Are there any other

metabolic diseases or are there any other systemic diseases, such as syphilis, that may be relevant? We know that some patients will have trauma and some patients will have infection preceding Ménière's disease, and in these patients there may be a contributing cause-and-effect relationship. These various aspects of the history are documented in abbreviated fashion on the chart and constitute an important part of the diagnostic workup.

Examination

If a patient presents with dizziness with or without a hearing loss, our preliminary workup includes a history, an otologic and head-and-neck examination, audiologic workup, caloric vestibular tests, and routine mastoid x-rays. Routine audiologic workup is first done, including pure-tone air- and bone-conducted frequencies, tympanometry, and discrimination of speech. More sophisticated audiologic studies are done later, if necessary. Auditory brain-stem response (ABR) is often used to differentiate labyrinthine from central diseases (for example, acoustic tumors) in lieu of more costly computed tomography (CT) scans or magnetic resonance imaging (MRI). Initially, caloric studies are done; electronystagmographic studies are usually pursued as follow-up studies. Vestibular hypofunction, as measured by caloric testing, is the most important finding of all vestibular tests in Ménière's disease; however, as seen in many centers including our own, up to 50 per cent of patients with Ménière's disease will have normal findings, even in the presence of incapacitating vestibular symptoms. Electronystagmography (ENG) provides a better method of recording caloric dysfunction and/or other abnormalities.

In diagnosing Ménière's disease, the history is most important, audiologic findings second-most important, and caloric (vestibular) and x-ray studies next in importance. Full otoneurologic assessment is necessary for confirmation and exclusion of other possible causes. General examination between attacks will reveal normal stance and gait without spontaneous nystagmus and with or without other ENG abnormalities. Otoscopy is almost always normal and tuning-fork and live-voice testing are used to corroborate the audiologic findings.

Auditory Tests

Pure-tone testing demonstrates a low-tone sensorineural hearing loss early in the course of the disease which may progress to a flat or occasionally a falling or through-shaped pattern later. The fluctuation in level can usually be observed by serial audiometry over weeks or months, and improvement in the audiometric threshold is a useful diagnostic feature. In many patients we noted a common configuration to comprise a peak audiometric pattern with comparatively good hearing at 2000 Hz, but poorer hearing below and above that point. In a series of 211 patients and 236 ears, the most common audiologic pattern was a flat audiogram (41 per cent), followed by a peak audiogram (31 per cent) (Meyerhoff et al, 1981).

Special audiometric tests can typify hair-cell disease. Acoustic reflex thresholds demonstrate recruitment, as does the test for alternate binaural balance in loudness (when recruitment is unilateral). The pattern of decay in acoustic reflex will fall between normal and the pattern caused by acoustic neuromas. Scores for discrimination of speech vary widely but

usually fall between those seen in normal individuals and those in patients with retrocochlear disorders.

A glycerol test may be used, and positive results confirm the histopathologic diagnosis of endolymphatic hydrops and may indicate better postoperative results regarding improvement in hearing and vertigo, if decompressive surgery is being considered. A positive result could also be a contraindication for destructive surgical procedures, since such a patient has the potential for some hearing. Whereas we, and others, formerly used glycerol tests, in recent years such tests have not appeared efficacious and are largely abandoned.

Electrocohleography (ECoG) is not absolutely necessary for diagnosis, but recent work with ECoG has suggested its possible usefulness in the evaluation of patients with Ménière's disease. This methodology allows measurement of the electrophysiology of the inner ear including the auditory nerve's action potential, cochlear microphonics, and summating potential. Characteristic changes in the ECoG are found in patients whose clinical history and examination are compatible with Ménière's disease. Since monitoring by ECoG reveals electrophysiologic changes in the inner ear, use of ECoG in the operating room (eg, during surgery on the endolymphatic sac), as well as in outpatient settings, may be useful and may add considerably to our understanding of Ménière's disease and its treatment (International Ménière's Disease Institute, 1988).

Especially promising is the possibility of detecting by ECoG those patients in whom Ménière's disease may eventually become bilateral. Features of ECoG characteristic of these patients include a broad waveform, steep input-output curves, and normal amplitude-latency relationships. Brain-stem evoked response is of value in separating patients with acoustic neuroma from those with Ménière's disease. In Ménière's disease, the early low-frequency sensorineural hearing loss is difficult to detect with this technique, which measures primarily the high-frequency response of the cochlea.

Vestibular Tests

It is unusual to be able to examine a patient during an acute spell, but labyrinthine beat- or jerk-nystagmus is always present during such a spell; it may be demonstrable at other times as latent nystagmus recorded electronystagmographically with the eyes closed, hours or days after a severe spell. During such a period, the brain is still compensating for the sudden vestibular insult.

The most common observation during testing of vestibular-ocular reflex in Ménière's disease is hypoactivity of the involved vestibular labyrinth to caloric stimulation. In general, the longer the duration of the disease, the less the caloric response is, and more nearly like the response of the normal side, earlier in the disease. Subjectively, caloric irrigation duplicates the patient's spell, although the perceived intensity of the response may be less than the patient's own spells. Caloric responses may be confusing if the caloric stimulation closely follows a major spell. This is also true for electronystagmography during positional and gaze-testing with eyes

closed. Vestibulospinal testing by posturography may prove to be a valuable method, not only to distinguish central from peripheral disorders but also to gain further insight into the effects of certain peripheral diseases. Nadol (1977) described a positive fistula sign in 30 per cent of patients with Ménière's disease; the deflection of the eye were induced by negative pressure on the external auditory canal much more frequently than by positive pressure, and occasionally by either positive or negative pressure.

Radiographic Findings

Routine mastoid x-rays or a lateral or Law's view is used to screen any abnormalities in the mastoid and the temporal bone. The patient may have had otitis media in childhood, resulting in a cloudy mastoid - chronic silent (subclinical) otitis media and mastoiditis. We wish to know if the mastoid is sclerotic, diploic, or pneumatic. Moreover, a number of authors have described mastoid and periaqueductal hypocellularity in Ménière's disease (Stahle and Wilbrand, 1983; Sando and Ikeda, 1985). Furthermore, recent studies in our clinic, in which measurements were made of the location of the lateral sinus in patients with Ménière's disease, indicate a decided statistical correlation of an anterior and medical displacement of the lateral sinus in patients with Ménière's disease (Paparella and Sajjadi, 1988), and this also is looked for and noted. CT scans are occasionally ordered to diagnose pathologic conditions in the temporal bone or retrocochlear space, whereas MRI is the most definitive test to diagnose and rule out intracranial pathologic conditions and is occasionally ordered as indicated. Consultation with other specialists, especially with neurologists, is sometimes necessary to aid in diagnosis and management.

No significant radiologic findings occur in the labyrinth per se, either in the cochlear or semicircular canal systems. However, varying degrees of abnormality may be seen in polytomographic examination of the vestibular aqueduct. Plane mastoid x-rays can show an anterior displacement of the lateral sinus and sometimes hypopneumatization of the mastoid air-cell system; both are common findings in patients with Ménière's disease. Radiographs of the petrous apex and other sophisticated examination should be reserved for patients suspected of having acoustic neuroma.

Laboratory Tests

In our opinion, the treponemal antigen test for syphilis is indicated and cost-effective in evaluating some patients with Ménière's disease. Studies of glucose metabolism and thyroid function, however, are probably not indicated in patients without a history suggestive of a metabolic disorder. Auto-immune studies are sometimes helpful.

Differential Diagnosis

Differential diagnosis of Ménière's disease is extremely important. Ménière's disease can mimic, or may be mimicked by, a number of diseases of the peripheral and central nervous system. The misdiagnosis of these conditions can result in potentially serious risks to the patient's integrity. Some of these diseases are easily recognizable, and the diagnosis is quite obvious.

Many times, however, the characteristics of the disorder are not typical and confusion may arise. In these cases, a careful history and physical examination with other diagnostic examinations as indicated will help to uncover the correct diagnosis. Table 4 summarizes the most important differential diagnosis of Ménière's disease. For further details, the reader is referred to the second part of this chapter and other chapters in this volume.

Table 4. Differential Diagnosis of Ménière's Disease

Psychogenic vertigo	Cogan's syndrome
Central nervous system causes	Ototoxicity
Idiopathic postural vertigo	Intermittent tubotympanitis
Benign paroxysmal positional vertigo	Barotrauma with sudden hearing loss
Benign paroxysmal vertigo of childhood	Intralabyrinthine hemorrhage
Vestibular neuronitis	Vertebrobasilar ischemia
Labyrinthitis	Vascular diseases
Labyrinthine fistula	Multiple sclerosis
Herpes zoster	Tumors of the cerebellopontine angle (meningioma, intracanalicular cranial nerve VIII schwannoma)
Trauma - labyrinthine concussion or fracture of the temporal bone	Hereditary ataxia
Otosclerosis	Vestibular epilepsy
Paget's disease	Postural imbalance of the elderly.
Latent congenital syphilis	

Central Causes

Causes traceable to the central nervous system do not generally produce such clear-cut paroxysmal attacks that typify Ménière's disease, and there may be persisting imbalance - rarely found in Ménière's disease. Examination may expose disturbance of gait and spontaneous nystagmus. Often, other neurologic abnormalities will be found. Unconsciousness suggests epilepsy.

Other Forms of Hydrops

Lermoyez's Syndrome

Lermoyez's syndrome is a rare variant of Ménière's disease in which the symptoms arise in the reverse order. Progressive deterioration of hearing is followed by episodic vertigo, at which time the hearing recovers. Patients with Lermoyez's syndrome often have a history of migraine. The course of the condition resembles that of Ménière's disease proper (Mawson and Ludman, 1979).

Cochlear Ménière's Disease

This relatively uncommon disorder is characterized by the gradual development of unilateral hearing loss and tinnitus. The impairment of hearing is found to be due to a low-tone, recruiting sensorineural deafness. The level of hearing fluctuates, and indeed the symptoms are those of endolymphatic hydrops affecting only the cochlea. Some patients recover spontaneously, whereas others later experience episodic vertigo and develop typical Ménière's disease (Mawson and Ludman, 1979; Williams et al, 1947).

Vestibular Ménière's Disease

A useful definition of vestibular Ménière's disease, a subvariety of this disease, was made by The Committee on Hearing and Equilibrium (Paparella and Mancini, 1985; Pearson and Brackmann, 1985). Vestibular Ménière's disease or Ménière's disease without deafness is characterized solely by the definitive spells of vertigo. This is more difficult to diagnose, as there are no objective findings between spells. The diagnosis may be accepted upon exclusion of other disease. Some patients with vestibular Ménière's disease may develop hearing loss or typical Ménière's disease subsequently. On follow-up, 20 per cent of our patients with vestibular Ménière's disease had their diagnoses changed to typical Ménière's disease. In other patients, vestibular Ménière's disease may persist for many years (30 or more). Three patients in the group that we studied had vestibular Ménière's disease for 20 years or longer.

A comparison of the group of patients with vestibular Ménière's disease with the group with the typical Ménière's disease may provide useful information regarding the natural history of vestibular Ménière's disease. It is interesting that in our clinic the incidence of vestibular Ménière's disease in women is higher (3 female:1 male) than is regular Ménière's disease. Also, the incidence of bilaterality in this group (women with vestibular Ménière's disease seen in our clinic) is much lower (14 per cent) than in those with typical Ménière's disease.

A clinical profile of a typical patient with vestibular Ménière's disease can be described. The patient can be male or (more likely) female. There may be a history of physical trauma or infection in the ear (vestibular Ménière's syndrome), but usually there is not (vestibular Ménière's disease). The diagnosis of bilateral vestibular Ménière's disease is somewhat more difficult to make than in unilateral Ménière's disease, since the clinical diagnosis is based chiefly on the history. Aural pressure, present on both sides, will usually be more severe on one side, and electronystagmographic findings tend to be similar on the two sides. In both Ménière's disease and vestibular Ménière's disease, the patient's chief complaint is vertigo, which is recurrent and typical of Ménière's disease. Other vestibular symptoms may occur, such as positional vertigo during or between episodes.

The patient does not complain of hearing loss, and the audiographic findings are normal. Nevertheless, the patient often can describe tinnitus and may note loudness intolerance in the involved ear in unilateral vestibular Ménière's disease, along with increased aural pressure, especially accompanying vertiginous attacks, in bilateral vestibular Ménière's disease. Aural

pressure is second in importance to vestibular symptoms, especially when localizing the side of involvement. Electronystagmographic changes are often present. Other otologic and neurologic diseases are absent.

In the case of vestibular Ménière's disease, the utriculo-endolymphatic valve may play an important role in pathogenesis. Patients with intractable vestibular Ménière's disease undergoing endolymphatic sac enhancement may have a better index of control and cure of vertigo than patients with typical Ménière's disease.

Therapy

A significant advance in Ménière's disease has been the development of fixed criteria for the evaluation of therapy and the reporting of results, devised by the American Academy of Otolaryngology's Subcommittee on Hearing and Equilibrium and Their Measurements (Alford, 1972; Pearson and Brackmann, 1985). Criteria for determining whether hearing is improved, stable, or worsened by treatment are defined. Control of vertigo means that the patient is free of definitive spells for an interval equal to or exceeding ten times the average interval between spells before treatment. This suggested method for classifying results is shown in Table 5.

Table 5. Classified Levels of Results in Ménière's Disease

Class	Results
A	Absence of definitive spells for described period; hearing improved
B	Absence of definitive spells for described period; hearing unchanged
C	Absence of definitive spells for described period; hearing worse
D	Failure to control definitive spells.

In 1985, a new formula was adopted for the reporting of results of treatment for Ménière's disease: Change in the definitive spells of vertigo is expressed by a single numerical score (summarized in Table 6). In addition, hearing loss was formularized: Improvement in hearing of 10 dB in the pure-tone average of 500, 1000, 2000, and 3000 Hz, or an improvement of 15 per cent in the speech discrimination score, is reported as hearing improvement. If the hearing is worse by 10 dB or 15 per cent, the hearing is reported as poorer. Lesser changes are reported as "no change". Categories of degree of change in status of disability (improved, unchanged, or worse) are also reflected in these formulations (see Table 6).

Table 6. Numerical Values Derived From Formula Adopted for Reporting Results of Treatment for Ménière's Disease*

0	Complete control of definitive spells
1-40	Substantial control of definitive spells
41-80	Limited control of definitive spells
81-120	Insignificant control of definitive spells
120	Worse (poorer) control of definitive spells.

* The following formula is used to express change in definitive spells of vertigo: (average number of definitive spells per month (24 months after therapy) x 100) / average number of definitive spells per month (6 months before therapy) = numerical value.

Medical Therapy

It is of crucial importance in the care of patients with Ménière's disease to establish a good doctor-patient relationship. Following the initial attack, the patient is extremely apprehensive and requires considerable support and reassurance. Psychological support is perhaps the most important part of medical management. It should include the explanation of the disease to the patient, with emphasis on its non-life-threatening nature, and of the various expectations regarding the natural history of the disease and/or its therapy (Blair, 1984). It is important to stress that there is no cure for Ménière's disease.

With medical therapy, we treat the symptoms, thus circumstantially improving conditions for the patient. Since most drugs act on an empirical basis, our policy is not to discourage the use of any of them if the treatment employed minimizes the symptoms and improves the quality of life for the patient. Ménière's disease may be progressive (PMD), leading to intractability, or more often Ménière's disease can be nonprogressive (NPMD), in which case progressive downhill labyrinthine damage and intractable vertigo and deafness will not ensure. Medical management, particularly that including psychological supportive therapy, should always be used first. If Ménière's disease with vertigo and/or deafness progresses and becomes intractable in spite of prolonged medical treatment, then surgery is considered.

While taking the history, one should search for contributory factors that may aggravate the course of the disease. Conditions such as fluid and hormonal imbalances, allergies, or stress must be detected and concomitantly treated. Currently a number of drugs from various pharmacologic groups are in use for alleviation of symptoms and in an attempt to alter the course of the disease. At this point there is little evidence that medical treatment can alter the course of Ménière's disease, although it may provide a buffering period, during which those patients destined for spontaneous remission can achieve it. There are also patients for whom surgical treatment is undesirable or who are unsuitable risks.

Diet and Diuretics

In the early 1930s, Mygind and colleagues (Mygind and Dederding, 1932) recognized the significance of water and sodium metabolic abnormalities in the etiology of Ménière's disease. In 1934, Furstenberg and coworkers noted the "salt sensitivity" of many patients suffering from Ménière's disease. This led to the concept of salt-restricted diets with diuretic therapy and potassium supplementation as a mode of treatment for this condition. While there may be some rationale for restriction of salt, and while it may help control the major spells, there is no evidence that it alters the progression of hearing loss. Perlman and colleagues (1953) were unable to induce or alleviate spells in patients with Ménière's disease by manipulating serum sodium levels. Hydrochlorothiazide was the only drug used for Ménière's disease that exhibited a salutory effect on the symptoms, when subjected to a controlled, double-blind study. Chlorthalidone has also been shown to be beneficial, over a 7-year trial period, but neither of these drugs arrests the degenerative course of the disease, although the number and severity of the definitive spells are improved so that the patient can frequently live with the problem.

Vestibular Sedatives

Drugs of the phenothiazine group with antihistamine properties (H_1-blocking agents) are strong vestibular repressants and also have antiemetic activity. They include cinnarizine and prochlorperazine. Antihistamines such as promethazine hydrochloride (Phenergan) are also useful as vestibular sedatives, and Meclizine (piperazine group) is another drug used to control vertigo and motion sickness. There is an important psychological component in Ménière's disease, and diazepam (Valium), a drug from the group of benzodiazepams, has proved to be useful in treatment, either because of its sedative effect, which may relieve some of the attendant anxiety, or because it may exert some selective sedative effect on vestibular nuclei. A drug recently added to the arsenal of medical therapy is transdermal scopolamine, an anticholinergic vestibular inhibitory agent.

Vascular Agents

Based on the supposition that Ménière's disease is in some way caused by labyrinthine ischemia, many agents have been advocated as labyrinthine vasodilators. It is extremely doubtful whether any of the so-called labyrinthine vasodilators significantly influence blood flow to the inner ear. Indeed, nicotinic acid, reportedly the most commonly used agent in the medical management of Ménière's disease, is known to have no effect on cochlear blood flow. Carbon dioxide, in the form of Carbogen or even resulting from breathing into a paper or plastic bag, is known to produce labyrinthine vasodilatation. Histamine and histamine analogues such as betahistine hydrochloride also have this effect, but their value has been questionable (Blair, 1984).

Vestibulotoxic Drugs

Streptomycin is sometimes used as an ablative agent in Ménière's disease, primarily when the disease is bilateral and it is essential to preserve hearing in a relatively young and otherwise healthy patient. It is not utilized until an adequate trial of conventional medical therapy has failed. Streptomycin primarily damage the type I hair-cells in the ampullae of the semicircular ducts and the striae of the otolithic organs. Treatment is continued until there is no response to caloric irrigation of either ear. The hearing is not significantly impaired.

Pressure Chamber

Thernstrom and coworkers (1980) reported the use of pressure chamber therapy in acute attacks of Ménière's disease. They exposed patients to a low-pressure environment and reported long-term relief of vertigo. Fundamental to this treatment is the concept that inner ear function can be affected by changes in pressure in the middle ear. Both vertigo and nystagmus were induced by changes in middle ear pressure.

Therapy for Acute Attacks of Ménière's Disease

Most patients with acute attacks of Ménière's disease are seen in the emergency room, not in the office of the ear-nose-and-throat doctor, because the attack causes frightening sensations in the patient. Acute attacks are best treated with bed rest and diazepam, orally or intravenously in appropriate doses. In severe attacks, droperidol can be used intravenously in a titrated administration under careful monitoring in the hospital. Atropine, 0.4 mg subcutaneously or sublingually, is effective for some patients during an acute attack (Alford, 1987).

Summary

We wish to emphasize the following topics concerning medical treatment of Ménière's disease:

1. The most important medical management is psychological support, which includes explaining the disease to the patient and describing the various expectations regarding the natural history of the disease and its therapy.
2. The primary purpose of medical management is to treat the patient and his or her symptoms - not only the disease per se.
3. In the long-term management of Ménière's disease, control of underlying etiologic and contributory factors such as stress, allergy, and hormonal disturbances is important.
4. A trial of medical treatment is important in determining whether Ménière's disease is progressive or non-progressive. Fortunately, most patients (an estimated 9 out of 10) have the nonprogressive form.
5. Although there is no cure for Ménière's disease, with conservative therapy we treat the symptoms and circumstantially improve conditions for the patient.
6. Most medical treatment is done on an empirical basis. Medical treatment cannot reverse the pathogenesis of progressive Ménière's disease.

Surgical Therapy

Whenever a patient complains of progressive Ménière's disease with symptoms that are disabling despite all efforts with medical therapy, he or she should be considered as a candidate for surgical treatment. There are many different surgical approaches to the treatment of Ménière's disease and vertigo, and they are discussed in detail in the following chapter (Chap 50).

Peripheral Labyrinthine Disorders

Ménière's disease is one of several diseases that may affect the peripheral labyrinthine system. Numerous other pathologic entities must be considered in the differential diagnosis. Vertiginous disorders of the central nervous system should be ruled out as well. Descriptions of and recommendations for management of other vertiginous entities are also discussed elsewhere in this volume.

Benign Paroxysmal Positional Vertigo

Among vertiginous disorders, benign paroxysmal positional vertigo (BPPV) is common. Furthermore, paroxysmal positional vertigo often accompanies other vestibular disorders such as endolymphatic hydrops or the poststapedectomy condition. Schuknecht (1974) has proposed cupulolithiasis as the principal mechanism. Calcium carbonate crystals from saccular or utricular otoconia impinge on the cupula of the posterior semicircular canal, the most dependent portion of the vestibular labyrinth.

Diagnosis is chiefly on the basis of history and physical examination. Vertigo and nystagmus are elicited upon Hallpike positioning; classically, there is a period of latency up to 5 seconds before nystagmus begins. Nystagmus is usually of the rotatory type, beating toward the underlying ear, although horizontal nystagmus may be present. Vertigo increases in intensity and subsequently disappears within 40 seconds, so long as the provoking position is maintained. Nystagmus is fatigable and should reverse direction when the upright position is assumed. Ideally, positional testing should reproduce the patient's distress exactly. Absence of latency should alert the physician to possible central nervous system pathology; lesions of the posterior fossa have been found to account for this type of malignant positional vertigo.

Positional vertigo of the benign paroxysmal type is distinguished by the usually brief duration of each attack and the absence of auditory symptoms. Jannetta and colleagues (1984), however, describe a syndrome of disabling positional vertigo. These patients had unremitting positional vertigo, accompanied by nausea that disabled them. Microvascular decompression of cranial nerve VIII led to symptomatic relief in all nine patients.

BPPV can follow head trauma, but commonly there is no obvious inciting etiologic agent. Symptoms can be recurrent over many years, but chronic BPPV is a rarity. Treatment, for the most part, consists of giving education and reassurance to the patient. Vestibular suppressants are used as adjunctive therapy, although there is no proof that they hasten recovery. Physical therapy

or Cawthorne-Hallpike exercises have also been attempted. Exercise helps in fatiguing vertiginous symptoms as well as probable central nervous system compensation.

Basser (1964) originally described benign paroxysmal vertigo of childhood. This is felt to be pathologically distinct from cupulolithiasis. Characteristically it occurs in children under the age of 4 years, but it may also occur in adolescence. Attacks consist of short-lived episodes of vertigo accompanied by nystagmus. The child may seem to be immobilized at times, while at other times he or she cries out in terror. Between attacks, the child appears to be completely normal. Children suffering attacks may experience nausea and sometimes vomiting. There are no auditory symptoms or loss of consciousness.

Laboratory, radiologic, and audiometric evaluations are unremarkable. Electroencephalograms are generally normal also. Vestibular testing has occasionally shown unilateral or bilateral paresis of the semicircular canal. Although the etiologic agent for benign paroxysmal vertigo of childhood is unknown, it has been postulated that there is a transient ischemic disturbance of the central vestibular system. The natural course of the disease is toward spontaneous remission over several years (Harker and Mccabe, 1980).

Vestibular Neuronitis

Ruttin (1909) provided the first description of vestibular neuronitis, characterizing it as sudden unilateral loss of vestibular function, with no auditory symptoms. Numerous reports have since followed. Aschan and Stahle (1956; see also Cawthorne, 1964) limited their definition to a single, severe vertiginous episode with permanent unilateral dysfunction upon vestibular testing. Adour and associates (1981) demonstrated hypesthesia of cervical nerve II, the glossopharyngeal nerve, and the trigeminal nerve, with vestibular vertigo. They postulated the cause as a viral cranial polyneuritis. Other terms used to refer to this disease include vestibular neuritis, acute labyrinthitis, acute epidemic vertigo, and vestibular paralysis.

Patients experience the onset of sudden, severe vertigo, often accompanied by nausea and vomiting. Spontaneous nystagmus beats toward the normal ear. Vertigo is sustained, with gradual improvement over a period of days. Unsteadiness or disequilibrium may continue for weeks to months. Coates (1969) has provided the following diagnostic criteria:

1. Acute unilateral peripheral vestibular dysfunction without associated hearing loss.

2. Occurrence predominantly in middle age.

3. One single episode of prolonged vertigo.

4. Decreased electronystagmographic caloric response in the involved ear.

5. Spontaneous and complete resolution of symptoms within 6 months.

Chronic forms of vestibular neuronitis are also believed to occur. Repeated episodes may take place over long periods of time. The initial episode is generally the most severe. Disease often occurs in clusters. Anttinen and colleagues (1983) found that 36 per cent of their patients had had a preceding viral infection. They further noted that 15 of 37 patients had an abnormal EEG and five of 12 patients had abnormal responses on testing for auditory brain-stem evoked response, suggesting possible involvement of the brain stem. Schuknecht and Kitamura (1981) also believed the etiologic agent to be viral, possibly herpes zoster. Pathologic findings in the temporal bones showed atrophy of one or more vestibular nerve trunks, with or without their associated sense organs. This study failed to reveal any evidence of occlusive vascular disease.

Treatment is symptomatic, consisting of bed rest and labyrinthine suppressants. Patients may require hospitalization. Adour and coworkers (1981) advise prednisone, 40 mg initially followed by 20 mg bid for 5 days. Chronic symptoms can be treated by sectioning the vestibular nerve.

Labyrinthitis

Both acute and chronic labyrinthitis can present with auditory and vestibular dysfunction. Labyrinthitis may be classified according to infectious (bacterial, viral, syphilitic, protozoal, fungal) or noninfective (toxic agents, autoimmune disease, labyrinthitis ossificans) origins. Further details may be found in Chapter 42 in this volume. Symptoms may be indistinguishable from those of Ménière's disease or perilymphatic fistula. Initial treatment is based on control of the initiating pathologic agent, if possible.

Motion Sickness

Motion sickness, or seasickness, has been described through the ages. Clinical symptoms include dizziness and/or vertigo accompanied by autonomic reactions such as sweating, pallor, and vomiting. Although abnormal vestibular stimulation may provoke motion sickness, it is not essential. These symptoms may be provoked as well by moving visual stimuli when these are in conflict with normal vestibular stimulation. Many authors (Sjoberg, 1929; Wang and Chinn, 1956; Money and Friedberg, 1964; Kennedy and Graybiel, 1965; Graybiel, 1965) have reported that motion sickness cannot be induced following bilateral labyrinthectomy. Symptoms seem to be dependent on activity in the vestibulocerebellum and on secretion of humoral mediators to the emetic trigger zone in the area postrema. Reason and Brand (1975) believe that sensory input to the vestibular system is somehow distorted by unusual stimuli.

Various drugs have been used in treatment and prevention of motion sickness. These include hyoscine, cyclizine, diphenhydramine, promethazine, and meclizine. Babin and coworkers (1984) have shown the efficacy of both transdermal scopolamine and meclizine over placebos.

Otosclerosis

In its early stages, otosclerosis may be accompanied by vertigo, thus leading frequently to an erroneous diagnosis of Ménière's disease. There is, however, rare fluctuation of hearing in otosclerosis (the otospongiotic state) when the hearing loss is less than 25 dB and the air-bone gap is minimal. It should not be forgotten that both pathologic conditions may coexist in the same patient.

Three major mechanisms have been postulated whereby an otosclerotic focus may produce vertigo. First, the otosclerotic focus could produce end-organ and/or neural degeneration. Second, otosclerosis may produce vertigo by virtue of the otosclerotic focus coming into contact with the perilymph within the vestibular labyrinth and changing the biochemistry of the perilymph. Finally, there may be a relationship between otosclerosis and endolymphatic hydrops. Johnson and colleagues (1978) and Liston and coworkers (1984) have described several temporal bones with otosclerosis and endolymphatic hydrops, and they believe that extensive capsular otosclerosis can cause labyrinthine hydrops. It seems reasonable to assume that otosclerotic involvement of the vestibular wall may produce endolymphatic hydrops owing to changes in the labyrinthine fluids in a manner somewhat analogous to that whereby cochlear otosclerosis produces a sensorineural hearing loss.

Medical therapy for otosclerosis and Ménière's disease consists of sodium fluoride and calcium gluconate. Paparella and coworkers (1984) have described successful surgical treatment with a stapedectomy-sacculotomy procedure. The rationale for this method, described in Chapter 50 in this volume, is to provide a yielding oval window membrane and to decompress a hydropic saccule.

Syphilis

Otologic syphilis must always be kept in mind as a possible cause of labyrinthine dysfunction. Symptoms may be present as a result of either congenital or acquired disease. The causative organism is *Treponema pallidum.* Manifestations of congenital syphilis may be present at birth or appear much later in life (Hendershot, 1973). In both congenital and acquired forms, symptomatology may mimic Ménière's disease.

Histopathologic findings consist of chronic osteomyelitis, along with miliary gummata and obliterative endarteritis (Schuknecht, 1974; Belal and Linthicum, 1980). Careful examination in suspicious cases should include an attempt to elicit Hennebert's sign or Tullio's phenomenon. History and physical examination are often unrewarding in the diagnosis of luetic disease of the inner ear; therefore, proper serologic testing is of the utmost importance.

The VDRL has great sensitivity in secondary syphilis but is only positive in approximately 70 per cent of cases of latent or tertiary syphilis. On the other hand, the fluorescent treponemal antibody-absorption test (FTA-ABS) is exquisitely sensitive: early latent syphilis is 99 per cent, late latent 96 per cent, and late (tertiary) syphilis 95 per cent detected

with this test (US Public Health Service, 1968). Other treponemal diseases such as *Treponema pertenue* (yaws) and *T. carateum* (pinta) may be responsible for false-positive results; so may autoimmune or collagen vascular disease (Anderson and Stillman, 1978). The microhemagglutination assay for *T. pallidum* (MHA-TP) is another diagnostic test with sensitivity and specificity similar to those of the FTA-ABS test (Jaffe et al, 1978). Of course a positive test result has no bearing on whether or not the patient has been adequately treated.

Penicillin and steroids remain the drugs of choice in treating luetic otologic disease. In order for treatment to be effective, the drug must penetrate perilymph. Differential penetration of penicillin into bodily fluid compartments may account for persistence of organisms after adequate therapy (Vrabec et al, 1965). Hughes and Rutherford (1986) recommended a regimen of 2.4 million units of penicillin G benzathine intramuscularly for each of 3 consecutive weeks, followed by a similar injection every 2 weeks for a total of 3 months. Prednisone, 20 mg four times daily, was given orally for 10 days, followed by doses tapering down to 10 mg every second day. For those patients allergic to penicillin, erythromycin, or tetracycline, 500 mg four times daily for 30 days, is a viable alternative (US Center for Disease Control, 1976). The effect of treatment on hearing is generally beneficial, with greater improvement in discrimination of speech than in pure-tone thresholds (Harker and McCabe, 1980). There is a more variable effect on vertigo.

Paparella and colleagues (1980) reported the results of eight endolymphatic sac decompressive procedures on six patients with refractory luetic vertigo and hearing loss. All patients had previously undergone medical therapy with no improvement in vertigo. Postoperatively, vertigo remained under control or was eliminated in all patients, although hearing was improved in only one patient.

Cogan's Syndrome

Cogan (1945) described four patients with nonsyphilitic interstitial keratitis and audiovestibular dysfunction. The syndrome is a rare clinical entity with fewer than one hundred cases reported in the world's literature. Incidence is equal for both sexes. Median age of onset is 25 years. A significant number of patients have had a preceding infection of the upper respiratory tract. Within 6 months, at least three-quarters of patients have involvement of both ocular and audiovestibular systems.

Common ophthalmologic symptoms include redness, photophobia, sensation of having a foreign body in the eye, and changes in visual acuity. Optic signs include interstitial keratitis, conjunctivitis, excessive lacrimation, and uveitis. Sensorineural hearing loss is an almost invariable component of the disease, with many patients suffering bilateral deafness. Severe vertigo accompanied by nausea and vomiting is common, as well as roaring tinnitus. Ataxia and nystagmus have also been observed. Various systemic illnesses have been noted in most patients, including nonspecific findings such as fever, loss of weight, and headache, as well as arthralgia, gastrointestinal bleeding, vasculitis, aortic valvular insufficiency, pleuritis, lymphadenopathy, splenomegaly, hepatomegaly, cranial neuropathy, and encephalitis.

Laboratory data frequently show an elevated erythrocytic and sedimentation rate, leukocytosis, anemia, and thrombocytosis. Cryoglobulinemia has also been reported (Hayes et al, 1980). Pathologic findings correlate with those of generalized vasculitis.

High-dose steroids are the therapy of choice. Medications that have been used without proven benefit include cyclophosphamide, azathioprine, histamine, antihistamines, vitamins, and antibiotics. Serious sequelae are common in Cogan's syndrome, and several deaths have been attributed to the disease.

Posttraumatic Vestibular Dysfunction

Vertigo and dizziness following closed trauma to the head are among the most common reasons for otolaryngologic consultation (Wiet et al, 1985). Mild trauma is believed to cause vertigo secondary to concussion to the inner ear; however, severe head trauma resulting in fracture of the temporal bone will cause direct damage to the cochlea and semicircular canals (Cannon and Jahrsdoerfer, 1983). This is especially true in cases of transverse fractures of the temporal bone. A study by Tuohimaa (1978) has shown posttraumatic vertigo or dizziness to occur in about 80 per cent of patients.

Complete otolaryngologic and neurologic examination, including cerebellar testing and cranial nerve function, should be performed as soon as possible. Complaints of decreased hearing or tinnitus must be elicited. Audiometric evaluation is essential in all patients with traumatic dizziness. Nystagmus should be documented during physical examination. Directional, fixed, spontaneous nystagmus or benign paroxysmal positional nystagmus may be present. Nystagmus of central origin may be enhanced by visual fixation. No response at all on electronystagmographic testing may be indicative of total peripheral labyrinthine destruction. Infective erosion of the labyrinth should be recognizable under otoscopic examination. If there is any suspicion of cholesteatoma, provoked, say, by the appearance of attic crust, cholesteatoma should be the presumed caused until it can definitely be excluded by an exploratory operation.

There are several hypotheses regarding the etiology of posttraumatic vestibular dysfunction, besides simply labyrinthine concussion. Disturbance is most probably a combination of central and peripheral causes. Positional vertigo is likely to be based on cupulolithiasis. Ménière's syndrome following trauma has also been shown. Perilymphatic fistula has been seen secondary to fracture of the stapes or subluxation of the stapedial footplate; these leaks are often amenable to surgical therapy.

Arragg and Paparella (1964) described a case of an emergency stapedectomy in a 12-year-old boy following injury by a twig of a branch entering his right ear. The child had unremitting vomiting and vertigo with third-degree horizontal rotatory nystagmus. Surgical exploration revealed fracture of the neck of the stapes with depression of the footplate into the vestibule. Stapedectomy relieved all symptoms and restored normal hearing.

Iatrogenic injuries of the labyrinth come in many forms. Perilymphatic fistula following stapedectomy is a well-recognized complication. Vestibular dysfunction may also occur without any technical surgical error. Endolymphatic hydrops may be present in an otosclerotic ear. Vestibular contents may mix with blood or enzymes of the middle ear.

Surgical fenestration is no longer a common procedure, but labyrinthitis was a common accompaniment, although it usually resolved spontaneously. Suppurative labyrinthitis leading to a dead ear or meningitis has also been reported.

Myer and Cotton (1982) reported erosion of the stapedial footplate leading to a perilymphatic fistula secondary to placement of a total ossicular replacement prosthesis (TORP). Jensen (1986) reported two cases of traumatic perilymphatic fistula caused by myringotomy alone. The mechanism was felt to be due to indirect damage secondary to violent force acting on the tympanic membrane and ossicles.

Acoustic trauma is a well-documented cause of sensorineural hearing loss. However, effects on the vestibular system are less well known. Vestibular dysfunction following excessive exposure to noise is usually of short duration and followed by complete recovery.

Perilymphatic Fistula

Goodhill (1971) described rupture of the round and/or oval window as a cause of sudden sensorineural hearing loss. He postulated increased pressures of fluid in the middle ear or of cerebrospinal fluid as possible mechanisms. Perilymphatic fistula may occur in several clinical situations including barotrauma, congenital malformation of the inner ear, post-stapedectomy, head trauma, and increases in cerebrospinal fluid pressure transmitted to the perilymphatic space by way of the cochlear aqueduct. There is also agreement that perilymphatic fistula may occur with no obvious etiologic agent (Harker and McCabe, 1980; Goodhill, 1971).

Diagnosis is presumptive, as definite proof can only be obtained at surgical exploration of the ear. Symptoms are variable and may be vestibular, cochlear, or mixed. Hearing may be normal or there may be sudden or fluctuant sensorineural hearing loss. It may simulate Ménière's disease with tinnitus, aural pressure, diplacusis, and loudness intolerance (Healy et al, 1976). A positive Hennebert's sign may be present.

Hearing loss can be greatest at high frequencies (8000 Hz), or the audiogram may show a trough pattern felt to correspond to sites of pathologic conditions on the basilar membrane. Hearing loss also may be slowly progressive over years, as has been shown in younger age groups. Simmons (1978) showed hearing loss to be most common after patients had made only partial recovery. Outlook for spontaneous restoration of hearing is more guarded with high-frequency loss, compared with losses in the mid or low frequencies. Other clinical phenomena include episodic vertigo, positional vertigo or nystagmus, constant disequilibrium, ataxia, and disturbance of gait (Kohut et al, 1986).

Regular audiometric evaluation as well as vestibular examinations are essential. Tests may reveal a stable pattern, complete recovery, or gradually declining function. Once a tentative diagnosis of perilymphatic fistula is made, initial treatment is usually conservative. This consists of restricting physical activity as much as possible and of frequent clinical and audiometric follow-up. If there is no improvement in vestibular symptoms, then exploratory tympanotomy is warranted. This is a minor surgical procedure that can be done under local anesthesia at an outpatient basis.

Progressive, fluctuating sensorineural hearing loss or vestibular dysfunction in the pediatric age group in whom perilymphatic fistula is suspected also deserves surgical exploration. Paparella and coworkers (1987) recommend patching of the round and/or oval window, even if no definite fistula is seen. Arenberg (1987) normally performs round window grafting in all patients undergoing surgery on the endolymphatic sac for intractable Ménière's disease. Seltzer and McCabe (1986) also perform round window grafting in those patients with symptoms compatible with Ménière's disease. Perilymphatic leakage may be on an intermittent basis or be so slow as not to be visible at surgery. Hearing loss and vestibular disturbance often show dramatic improvement. Collagen tissue was felt to be superior to adipose tissue in patching a visible fistula.

Paparella and colleagues (1987) described a series of patients with sudden sensorineural hearing loss and bulging of the round window membrane noted at exploratory tympanotomy. Vestibular symptoms, however, were noted in only one patient. Paparella termed the entity "perilymphatic hypertension". Perilymphatic fistula may be a result of perilymphatic hypertension subjected to increases of either volume or pressure of perilymph. Paracentesis of the round window was safely performed by insertion of a 0.5-mm or 1-mm hook into the scala tympani, thereby reducing perilymphatic pressure. A connective tissue graft was then applied to the round window.

Ototoxicity

Ototoxicity results from the action of certain drugs on chemicals, causing hair-cell injury to the labyrinth. Vestibular dysfunction is more apt to present clinically as disequilibrium rather than as true vertigo. All age groups, including neonates, are susceptible to exposure to ototoxic drugs. Spontaneous nystagmus is not usually a cardinal feature. Electronystagmography with caloric testing is often the most sensitive determinant of vestibular dysfunction. Bilateral ablation of vestibular function results in Dandy's syndrome or Bobbins' oscillopsia. For a more detailed discussion of ototoxic drugs, the reader is referred to Chapter 46 in this volume.

Aminoglycoside antibiotics are well known to be vestibulotoxic, especially gentamicin, tobramycin, and streptomycin. Selected patients with bilateral Ménière's disease and unremitting vertigo have been treated with streptomycin as a means of vestibular ablation.

There are innumerable patients taking over-the-counter medications, abusing drugs, or taking inappropriate doses of properly prescribed medications. Anticonvulsants such as dilantin

or phenobarbital may cause dizziness with toxic levels. Therefore, a thorough drug history is mandatory in any patient complaining of vestibular dysfunction.

Presbyvertigo

Presbyvertigo is believed to be a degenerative disease of the vestibular labyrinth similar to presbycusis and degenerative cochlear changes. Pathologic evidence shows increased levels of degeneration of primary vestibular afferent neurons as well as of sensory hair-cells. Anniko (1983), while studying guinea pigs, described "aggregation of lipofuscin pigments, multivesiculated bodies, disintegration of the cuticular plate, and rod-shaped inclusions from the cortical area into the hair cell". He believed these changes to be less apparent in type II hair-cells compared with type I. Other studies using vestibular function tests have shown evidence of increasing vestibular dysfunction with advancing years. Disorders of carbohydrate metabolism may be a possible etiologic factor. Many elderly patients also suffer from visual problems, thus compounding their situation. Adaptation is difficult, as the cause is related to decreased sensory input.

Intralabyrinthine Hemorrhage

Temporal bone studies have documented intralabyrinthine infarction or hemorrhage. This is especially likely in patients with underlying coagulation defects such as leukemia. It has also been known to occur secondary to blunt trauma to the head or fracture of the temporal bone. The usual clinical presentation is severe acute vertigo, nausea and vomiting, and nystagmus. Electronystagmography will show unilateral vestibular paralysis.

Central-Peripheral Diseases

Not all causes of vertigo lend themselves to being easily distinguished as central or peripheral sources. They may have features in common with both or progress from one to another. As an example, underlying circulatory disturbances may affect either peripheral vestibular circulation or central nuclei. Symptoms often appear to be the result of strictly peripheral pathologic conditions, until subjected to closer scrutiny.

Vascular Loop Syndrome

Jannetta (1975) first described neurovascular cross-compression as a cause of cranial nerve VIII dysfunction. Patients may present with either cochlear or vestibular symptoms. These may include tinnitus, diplacusis, vertigo, and hearing loss. Traditional treatment (Jannetta, 1981) has consisted of direct visualization of the vascular loop and nerve at the cerebellopontine angle, with placement of a sponge between as a form of decompression. In McCabe and Harker's series (1983), they noted that this was not a viable alternative, in view of the depth of extension of the vascular loop into the internal auditory canal; therefore vestibular nerve section provided another option for symptomatic relief.

McCabe and Harker (1983) described a series of eight patients with episodic vertigo and intolerance for motion, believed to be caused by the vascular loop syndrome. They resorted to sectioning the vestibular nerve as therapy, although the possibility of the vascular loop syndrome had been considered preoperatively in only two patients. At surgery, they noted the presence of a large arterial loop in the lateral section of the internal auditory canal, which produced indentations of the vestibular nerve. According to Mazzoni and Hansen (1970), in only about 5 per cent of individuals will loops of the cerebellolabyrinthine and anterior inferior cerebellar arteries extend into the lateral half of the internal auditory canal.

No definitive diagnostic tests can prove the diagnosis with certainty. Jannetta and colleagues (1984) attempted auditory brain-stem evoked responses in their patients. They believed that vascular compression of cranial nerve VIII might lead to delayed conduction times.

Vertebrobasilar Insufficiency

Vascular compromise is a frequent cause of vertigo and/or disequilibrium in the elderly population. The vertebrobasilar circulation encompasses the vestibular nuclei, the entry zones of the vestibular nerve, and the vestibular labyrinth. Vertigo secondary to vertebrobasilar insufficiency may be of sudden or insidious onset. Vertigo has been reported as an isolated symptom in up to 20 per cent of cases. Accompanying nystagmus may be either vertical or horizontal. Other associated symptoms include slurring of speech, facial numbness, hemiparesis, headache, diplopia, and drop-attacks. Patients may be affected anywhere from a few seconds to hours.

Eighty per cent of patients with symptoms of vertebrobasilar insufficiency are more than 50 years old. Suspicion should be aroused in patients with evidence of other systemic vascular disease, hypertension, history of cerebrovascular accidents, diabetes, or hyperlipidemia. This lesion often presents a diagnostic challenge. Computed tomographic scans, nuclear magnetic resonance imaging, and arteriograms are helpful in establishing the diagnosis. Diagnosis is important, as patients may go on to develop basilar arterial thrombosis. Treatment is variable and may consist of anticoagulant therapy or surgical endarterectomy.

Cervical Vertigo

Cervical vertigo is a somewhat controversial topic. It is most often secondary to motor vehicle accidents and the whiplash type of injury. Animal studies have shown linkage between cervical sensory input and vestibular nuclei. Proprioception is felt to arise from the paravertebral joints and capsules. Disequilibrium in these patients may be extremely long-lasting.

Tumors

Neoplastic disease may produce vestibular symptoms directly or indirectly. Vestibular nerves may be subjected to disease both in the temporal bone and in the cerebellopontine angle. Tumors may cause compression of the cerebellum or brain stem also. Direct infiltration of the

labyrinthine structures can produce vestibular and auditory dysfunction. A classification of tumors affecting this area appears in Table 7.

Table 7. Classification of Tumors of the Posterior Fossa and Temporal Bone

Benign

Acoustic schwannoma
Meningioma
Primary cholesteatoma
Glomus tumor
Facial nerve schwannoma
Arachnoid cysts
Hemangioma
Osteoma
Cholesterol granuloma

Malignant

Squamous cell carcinoma
Chondrosarcoma
Chordoma
Rhabdomyosarcoma
Medulloblastoma

Metastatic (from)

Breast carcinoma
Lung carcinoma
Prostatic carcinoma
Kidney.

Multiple Sclerosis

Multiple sclerosis most commonly affects patients after the age of 20 years. Pathologically, it is a demyelinating disease of the central nervous system. About 10 percent of patients will complain initially of vertigo or imbalance. Eventually about two-thirds will experience these symptoms. Positional or spontaneous nystagmus is often present. Electronystagmography frequently shows signs of central nystagmus such as vertical nystagmus. There is usually a lack of visual fixation suppression. Nuclear magnetic resonance imaging is capable of showing the plaques characteristic of multiple sclerosis.