Chapter 181: Ménière's Disease and Other Peripheral Vestibular Disorders

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Basic Principles of Peripheral Vestibular Disorders

The peripheral vestibular system consists of the following: (1) sensory receptor structures that are responsible for sensing the motion of the head in space and (2) the vestibular portion of the eighth nerve that carries sensory information from the receptors to the central nervous system in the form of neural activity. The system includes the semicircular canals, which sense nonlinear, or angular, accelerations of the head, and the otolith organs, the utricle and saccule, which detect linear accelerations of the head, including gravity. A simplified overview of vestibular function follows.

Vestibular receptors lie within the temoral bone. Each is precisely oriented to detect head movement in a specific direction or plane; that is, horizontal head turning produces a change in the activity of the horizontal semicircular canals. Changes in the pitch of the head are detected by the superior and posterior semicircular canals. The receptors are tonically active. There is always a resting, spontaneous outflow of action potentials from the vestibular portion of the eighth nerve. As the head is turned, this spontaneous activity is modulated up and down, a bidirectional response. For example, turning the head to the right in the upright position produces an increase in activity in the nerve coming from the right horizontal semicircular canal, whereas left turns decrease activity below the resting rate in the same canal. Paired, the two labyrinths, one in each ear, behave in a "push-pull" fashion. The central nervous system compares the input coming from each ear. When input is equal, the system is balanced and there is no sense of movement. When inputs are asymmetric, the central nervous system interprets this as a head rotation and as a result generates compensatory eye movements and postural adjustments, all of which lead to a conscious sensation of movement relative to the perceived movement of the head.

Very importantly, the central nervous system is capable of rebalancing itself; that is, compensating after an injury to the peripheral receptor. This is thought to occur as a result of the organism's being able to compare other sensory, visual, and kinesthetic inputs to vestibular input and, once recognizing the inaccuracy of the latter, readjusting the central response. As a consequence, the point of balance is reset (Fig. 181-1). Central nervous system compensation in an otherwise healthy individual results in near normal clinical recovery even if complete unilateral vestibular loss has occurred. However, compensation takes time, in the order of days. The abrupt vestibular imbalance created by a labyrinthectomy leads to severe vertigo, nausea, and ataxia that gradually improve over ensuing days. On the other hand, a slowly progressive lesion of the vestibular system, such as occurs with acoustic neuromas, rarely causes vertigo. Although vestibular function is destroyed, because it has occurred gradually, it has resulted in central compensation.

As opposed to acute unilateral peripheral vestibular loss, bilateral vestibular loss, exemplified in the patient who has suffered ototoxicity, results in permanent clinical disability. Because peripheral vestibular input is lacking, the central nervous system is unable to make the required adjustments to head movements. If the loss is simultaneous and symmetric, there is no significant vertigo. The patient complains of worsened balance, especially in the dark.

Fixed objects appear to jump with any head movement (oscillopsia). The symptoms are caused by a now inefficient vestibuloocular and vestibulospinal system. It is important to recognize that as long as there is no asymmetry in vestibular input, even in the case of its absence, there is no sensation of vertigo.

Clinical Relevance

The most important clinical feature in the diagnosis of peripheral vestibular dysfunction is its pattern of presentation. The importance of a well-taken history is paramount.

Vertigo is the hallucination of movement, either of self (subjective) or the environment (objective). Although this chapter will not address the thorough otoneurologic examination, a minimum vertigo history should address the following: (1) the duration of the individual attack, that is, hours versus days; (2) frequency, that is, daily versus monthly; (3) the effect of head movements, that is, worse, better, or no effect; (4) specific position induced, for example, rolling onto the right side in bed; (5) associated aural symptoms, that is, hearing loss and tinnitus; and (6) concomitant ear disease, that is, otorrhea, prior surgery, or trauma.

One of the most important features of the pattern of presentation of peripher vestibular disorders is the duration of vertigo. Based on this parameter, a classification of peripheral vestibular disease is put forward.

1. Vertio lasting minutes to hours
    a. Idiopathic endolymphatic hydrops (Ménière's disease)
    b. Secondary endolymphatic hydrops
        (1) Otic syphilis
        (2) Delayed endolymphatic hydrops
        (3) Cogan's disease
        (4) Recurrent vestibulopathy
2. Vertigo lasting seconds (benign paroxysmal positional vertigo)
3. Vertigo lasting days (vestibular neuronitis)
4. Vertigo of variable duration
    a. Inner ear fistula
    b. Inner ear trauma
        (1) Nonpenetrating trauma
        (2) Penetrating trauma
        (3) Barotrauma.

Vertigo Lasting Minutes to Hours

Ménière's disease (idiopathic endolymphatic hydrops)

Ménière's disease is defined as a disorder of the membranous labyrinth that consists of fluctuating sensorineural hearing loss, episodic vertigo, tinnitus, and less frequently aural fullness.

Background

History. In 1861 Prosper Ménière first described the symptom complex and proposed the pathologic site to be in the labyrinth. Interestingly, in his first description of the disease, vertigo was felt to originate in the central nervous system, "apoplectic cerebral congestion" (Ruben, 1989). It was following his later recognition of the work of Flourens and his evaluation of a patient with labyrinthitis that he recognized the direct association of vertiginous symptoms with the inner ear.

In 1926 Portmann, believing Ménière's was secondary to endolymphatic hypertension, performed the first drainage operation of the endolymphatic sac.

In 1938 Hallpike and Cairns, and independently, Yamakawa, demonstrated the histologic findings of "hydrops" in two patients with Ménière's disease.

In 1965 Kimura and Schuknecht produced hydrops in the guinea pig by destroying the endolymphatic duct and sac. Also in 1965 Dohlmann and Johnson caused vestibular paralysis by infusion of artificial endolymph into the perilymphatic compartment in animals.

Incidence. Wide variation exists in the published incidence of Ménière's disease. Incidence varies from 157 per 100.000 persons in England to 46 per 100.000 in Sweden and 7.5 per 100.000 in France (Pfaltz and Thomsen, 1986). The incidence has reportedly increased in Japan since World War II (Watanabe, 1981). Racial distribution is unestablished (Pfaltz and Thomsen, 1986). A sexual preponderance of the disease is also not clearly defined, with several reports suggesting opposing conclusions (Balkany et al, 1980; Paparella, 1985; Pfaltz and Thomsen, 1986; Watanabe, 1981). Ménière's disease is responsible for 10% of visits to the Dizziness Unit at Sunnybrook Health Science Centre, University of Toronto (Nedzelski et al, 1986). This reflects the prevalence of the disease among people complaining of dizziness in a tertiary referral center. The age distribution is widely variable with a noted peak in the 40- to 60-year age group (Paparella, 1985).

The frequency of bilateral disease is also unclear. Published reports cite ranges from 2% to 78% (Balkany et al, 1980) (Stahle, 1991). This disparity is ascribed to a lack of consensus as to what criteria constitute the entity (Balkany et al, 1980; Greven and Oosterveld, 1975). Paparella and Griebie (1984) noted a demonstrable hearing loss in 78% of contralateral ears of unilateral Ménière's patients. However, in only 32% of the patients was the diagnosis of bilateral Ménière's made based on the clinical picture. Several authors have stated that if bilateral involvement has not occurred within 5 years of onset of disease in the first ear, then there is less likelihood of bilateral involvement (Friberg et al, 1984). On the other hand, studies have concluded that the incidence of bilateral disease increases continuously over time (Friberg et al, 1984; Paparella and Griebie, 1984; Stahle, 1991).

Familial occurrence of Ménière's disease has been reported in 10% to 20% of patients in one study (Paparella, 1985). In a Swedish study, 14% of 91 patients studied were noted to have a familial distribution of the disease. It was concluded that genetic inheritance played a role, but the mode of transmission was variable (Birgerson et al, 1987).

Pathogenesis. Ménière's disease is characterized by recurring attacks of vertigo, hearing loss, and tinnitus. Acute attacks are superimposed on a gradual deterioration in hearing in the involved ear, typically in the low frequencies initially. Over time a reduction in responsiveness of the involved peripheral vestibular system occurs.

The pathologic basis felt to underscore these findings is a distortion of the membranous labyrinth. The hallmark of this is endolymphatic hydrops (Altmann and Hager, 1968; Anatoli-Candela, 1976; Hallpike and Carins, 1938; Schuknecht and Igarishi, 1986). This reflects the changes in the anatomy of the membranous labyrinth as a consequence of the overaccumulation of endolymph. This occurs at the expense of the perilymphatic space (Anatoli-Candela, 1976; Klis et al, 1990; Paparella, 1985; Schuknecht and Kitamura, 1981).

Normally, endolymph, which is produced by the stria vascularis in the cochlea and by the dark cells in the vestibular labyrinth, circulates ni both a radial and longitudinal fashion. Radial circulation is primarily a local phenomenon, whereas longitudinal flow is characterized by absorption within the endolymphatic sac. In the case of hydrops, inadequate absorption of endolymph by the endolymphatic sac is the prevalent theory (Paparella, 1985). Temporal bone histologic equivalents of hydrops have been produced in animals as a consequence of disruption of the endolymphatic sac by various means, thereby supporting the above theory of pathogenesis (Fukuda et al, 1988; Kimura, 1967; Sawada et al, 1987; Yazawa et al, 1985). Pathologic studies of the human sac in the hydrops patient have recorded perisaccular ischemia and fibrosis as well as accumulation of cellular and proteinaceous debris within the sac lumen (Takumida et al, 1989; Yazawa and Kitahara, 1989; Wackym et al, 1990). Other studies have implicated other processes in endolymph absorption such as duct obstruction (Franklin et al, 1990; Sismanis et al, 1986; Yoon et al, 1990; Yuen and Schuknecht, 1972). The possibility that endolymph overproduction is responsible has also been put forward (Henriksson et al, 1986).

Although it has been assumed that the clinical picture of Ménière's disease is correlated with the pathologic finding of endolymphatic hydrops, the carefully controlled histopathologic study of Rauch et al (1989) raised some questions. Of 19 cases with histopathologic findings of idiopathic endolymphatic hydrops, 6 had clinical histories that were incompatible with the diagnosis of Ménière's disease. The authors suggest that endolymphatic hydrops may be an epi-phenomenon that occurs with the vertigo and hearing loss but is not necessarily the cause of them.

Endolymphatic hydrops is most consistently found in the pars inferior (cochlea and saccule) (Altmann and Kornfeld, 1965; Schuknecht and Igarishi, 1986). It is typified by bowing of Reissner's membrane into the scala vestibuli and distention of the saccule (Schuknecht, 1974) (Fig. 181-2). These changes vary in degree. Saccular expansion can distort the utricle and semicircular canals as well as contact the undersurface of the stapes foot plate directly or via fibrous adhesions (Issa et al, 1983). These findings may be responsible for the observed vertigo produced by pressure-induced excursions of the foot plate, so-called Hennebert's sign (Nadol, 1977). Changes in the pars superior (utricle and semicircular canals) can be seen but are generally less dramatic. The utricle has been observed to herniate into the crus commune, and the cupula has been found displaced from its attachment to the roof of the ampulla (Birgerson et al, 1987; Rizvi, 1986; Sando et al, 1968).

Hair cells, along with the innervating nerves, are spared histologically in the Ménière's patient. It is only in the most severely affected cases that there are decreases in hair cells and loss of the neuronal population (Birgerson et al, 1987; Schuknecht, 1986).

Ruptures in the membranous labyrinth of the Ménière's patient are felt to be significant to the pathophysiology of Ménière's disease (Kimura, 1984; Koskas et al, 1983; Lawrence and McCabe, 1959; Schuknecht, 1963, 1968). Membranous ruptures have been found in nearly all parts of the inner ear. Healed scars, presumably after rupture, have also been identified (Anatoli-Candela, 1976; Schuknecht, 1986). Their presence has supported one of the more prominent theories of the pathogenesis of Ménière's disease. Ruptures in the membranous labyrinth allow leakage of the potassium-rich endolymph into the perilymph, bathing the eighth nerve and lateral sides of the hair cells. High concentratons of extracellular potassium depolarize the nerve cells and cause their acute inactivation. This results in a decrease in auditory and vestibular neuronal outflow, consistent with the hearing loss and features of acute vestibular paralysis seen in the typical Ménière's attack. Healing of the membranes is presumed to allow restitution of the normal chemical milieu, with termination of the attack, and improvement in vestibular or auditory function. The chronic deterioration in inner ear function is presumably the effect of repeated exposure to the effects of the potassium (Thomsen and Bretlan, 1986).

This hypothesis is supported by the observed ability of the labyrinth to heal after rupture (Kimura and Schuknecht, 1975) and the production of acute, reversible, vestibular paralysis with perfusion of artifical endolymph into the perilymphatic space (Dohlmann and Johnson, 1965; Silverstein, 1970). Others are of the opinion that maldistribution of ions occurs as a consequence of a leaky perilymph-endolymph barrier rather than frank rupture (Jahnke, 1981; Sziklai et al, 1989).

A biochemical hypothesis of Ménière's attacks is enticing, given that the receptor hair-cell population is not severely reduced in the Ménière's patient. This theory is disputed by others who feel that ruptures in the endolymph-limiting membranes occur rarely and cnostitute catastrophic events (Thomsen and Bretlau, 1986; Tonndorf, 1983). Rather, they suggest that mechanical tensing and distortion of the membranous inner ear may be contributing factors (Tondorf, 1975, 1986).

Etiology of Ménière's disease. The triad of hearing loss, tinnitus, and vertigo constitutes Ménière's syndrome. If a cause is unknown, it is defined as Ménière's disease (Paparella, 1984). However, if a disease entity that is known to cause endolymphatic hydrops is associagted with the syndrome, the diagnosis is one of secondary endolymphatic hydrops, for example, syphilitic endolymphatic hydrops. Numerous mechanisms have been implicated as the cause of the disease.

Obstruction of the endolymphatic duct is the basis for development of hydrops in animals. This is accomplished by mechanical blockage (Beal, 1968; Horner et al, 1989a; Kimura, 1967), chemical fibrosis of the endolymphatic duct (Yazawa et al, 1985), viral inoculation of the endolymphatic sac (Fukuda et al, 1988), and immunologically induced inflammation (Sawada et al, 1987; Yoo et al, 1982). Animal models only serve as histologic equivalents. To date these models do not mirror the clinical entity experienced by humans.

Mechanical blockage of the endolymphatic duct in humans from otosclerotic foci or tumor, can precipitate hydrops and a Ménière's-like syndrome (Franklin et al, 1990; Yoon et al, 1990).

Study of the human endolymphatic sac has suggested that it is an immunocompetent structure capable of processing antigen, synthesizing antibodies, and raising a cellular immune response (Harris, 1989; Rask-Anderson and Stahle, 1980; Veldmann et al, 1984). Autoimmune mechanisms have been implicated as being responsible for the inflammatory response. Circulating immune complexes were found to be elevated in Ménière's patients (Brooks, 1986; Hsu et al, 1990). The response of some patients to steroids and electrophoresis implicates autoimmunity as a possible cause in selected cases (Brookes, 1986; Futaki et al, 1988; Hughes et al, 1988a). Antibodies directed against mesenchymal elements of normal inner ear have been found in the serum of Ménière's patients (Arnold et al, 1985). Immunoglobulins deposited in the endolymphatic sac tissues as well as in increased concentrations in the endolymph in Ménière's patients have been described (Dickens and Graham, 1990; Futaki et al, 1988; Yazawa and Kitahara, 1989). Antibodies specific to type II collagen have been found in the serum of Ménière's patients. Type II collagen autoimmunity is also believed related to otosclerosis. Induction of type II collagen immunoreactivity in mice produces an otosclerosis-like lesion as well as hydrops, with auditory and vestibular dysfunction (Yoo et al, 1982). Hughes et al (1988b) suggested that autoimmune hydrops should be suspected in middle-aged women with concomitant autoimmune disease.

Viral infection causing damage to the endolymphatic duct and sac has been suggested as a cause of Ménière's disease (Schuknecht, 1986). The observed occurrence of symptomatic hydrops many years after unexplained deafness, so-called delayed endolymphatic hydrops, suggests that subclinical viral infection could cause hydrops many decades later (Schuknecht et al, 1990).

Small vestibular aqueducts have been implicated as a cause of Ménière's disease. Radiologic studies of human temporal bones suggest this possibility (Clemis and Valvassori, 1968). However, histologic studies do not support these observations (Sackett et al, 1980; Yuen and Schuknecht, 1972).

Diagnosis

Clinical presentation. There is no test that conclusively establishes the diagnosis of Ménière's disease. A combination of the history of illness coupled with appropriate audiologic and vestibular investigations is required. The typical history consists of recurring attacks of vertigo (96.2%) with tinnitus (91.1%) and ipsilateral hearing loss (87.7%) (Paparella and Mancini, 1985). Attacks are often preceded by an aura consisting of a sense of fullness in the ear, increasing tinnitus, and a decrease in hearing. They may, however, be sudden in onset, with little or no warning. Attacks may awaken the patient from sleep (Barber, 1983). Acute attacks typically last from minutes to hours, most commonly 2 to 3 hours (Barber, 1983; Oosterveldt, 1980). Attacks lasting more than a day are unusual and should be considered inconsistent with the diagnosis (Friberg et al, 1984; Stahle and Klockohoff, 1986).

The classic presentation described above is not always detailed by the patient. This is particularly true in the early phases of the disease. Often in retrospect, the disease seems to present with a predominance of either vestibular or auditory complaints (Cawthorne and Hewlett, 1954; Friberg et al, 1984; Paparella, 1985; Thomas and Harrison, 1971). As an example, in Kitahara et al's series (1984), 50% of patients presented with vertigo and hearing loss together, 19% with vertigo only, and 26% with deafness. These variable initial presentations have led to use of the terms *cochlear* or *vestibular Ménière's disease.* We believe, as do others including the American Academy of Otolaryngology - Head and Neck Surgery Committee on Hearing and Equilibrium (AAO-HNS, 1985)), that this is an inappropriate application of the diagnosis (Kitahara et al, 1984; Stahle and Klockhoff, 1986; Nadol, 1990). Studies that have observed patients with "cochlear and vestibular" Ménière's have found that 80% of the "cochlear Ménière's" patients go on the have classic Ménière's disease. In the case of the "vestibular Ménière's" patient, only 10% to 20% will develop classic Ménière's disease (Kitahara et al, 1984; Paparella and Mancini, 1985). This suggests that the majority of these patients do not have Ménière's disease. In addition, there is no pathologic correlation for a vestibular Ménière's (Kitahara et al, 1984). For these reasons the term *recurrent vestibulopathy* is used to describe such individuals. It will be discussed subsequently. Patients who present with the audiologic features of Ménière's disease but without the clinical triad of symptoms may be described as having atypical Ménière's disease.

The clinical course of Ménière's disease is highly variable. Patients often have a cluster of attacks separated by long remissions. Silverstein et al (1989) found that vertigo ceased spontaneously in 57% of patients in 2 years and in 71% after 8.3 years. There may be single, sporadic attacks or periods of unrelenting, recurring attacks. Patients may be minimally inconvenienced or completely incapacitated. Although few long-term studies of large groups of Ménière's patients have been carried out, there are several that indicate the average expected outcome of persistent disease (Friberg et al, 1984). Friberg et al's study reported an average pure-tone hearing loss of 50 dB, a mean speech discrimination score of 53%, and an average caloric response reduction of 50%. The mean frequency of attacks was 6 to 11 per year for the first 20 years, with wide ranges, and 3 to 4 per year after 20 years. Bilateral disease occurred in 47% of patients observed for 20 years or more. Similar patient profiles have been noted by Stahle (1976).

History and physical findings

Vertigo. Incapacitating, spinning vertigo, usually in the horizontal axis, is the most distressing complaing of the affected patient (Paparella, 1984). It characteristically peaks early in the attack. As is typical of peripheral vestibular dysfunction, the symptoms are exacerbated with any head movement. There is often accompanying nausea, vomiting, diarrhea, and sweating. However, the description and severity of attacks vary over time and between patients (Barber, 1983). Between attackes, patients may be entirely asymptomatic or describe periods of disequilibrium, light-headedness, or tilt (Paparella, 1984).

Sudden unexplained falls without loss of consciousness or associated vertigo are occasionally described. Tumarkin (1936) attributed these to acute utriculosaccular dysfunction, so-called "Tumarkin crises". It is felt that as a consequence of an abrupt change in otolithic input, an erroneous vertical gravity reference occurs. This in turn generates an inappropriate postural adjustment via the vestibulospinal pathway, resulting in a sudden fall (Barber, 1983;

Odkvist and Bergenius, 1988). Drop attacks occur in less than 2% of Ménière's afflicted individuals.

An unusual pattern of clinical presentation is that described by Lermoyez (1919). Increased tinnitus and hearing loss precede the vertiginous episode and dramatically resolve with onset of vertigo (Schmidt and Schoonhaven, 1989).

An acute attack is rarely physician observed. Horizontal nystagmus is the cardinal finding during such an episode. The direction varies over the time course of the attack. Early in the attack, nystagmus is often noted beating toward the affected ear, so-called "irritative nystagmus" (Bance et al, 1991). Later, the nystagmus beats toward the healthy ear, "paralytic nystagmus". As the attack subsides and vestibular function improves, nystagmus often reverses toward the affected ear, so-called "recovery nystagmus" (Brown et al, 1988; McClure et al, 1981). The reliability of determining the involved ear on the basis of the direction of nystagmus is questionable (Stahle and Klockhoff, 1986).

Hearing loss and tinnitus. The sensorineural hearing loss in Ménière's disease is typically fluctuating and progressive. It often occurs coincident with the sensation of fullness or pressure in the ear. A pattern of low-frequency, fluctuating loss and a conincident nonchanging, high-frequency loss is described, a so-called "peaked" or "tentlike" audiogram. This peak classically occurs at 2 kHz. Over time the hearing loss flattens and becomes less variable. Friberg et al (1984) on initial examination found a flat audiogram in 21% of patients and a peaked one in 28%; an isolated high-frequency loss was rare. The authors further note that the flat audiogram was found in 74% of the same group of patients after 15 years of follow-up. Peaked and low-frequency patterns of loss were the most prevalent in other studies (Barber, 1983; Paparella, 1984). The Ménière's patient rarely becomes profoundly deaf. This occurs in 1% to 2% of severely affected patients (Stahle, 1976). It has been suggested that the rate of hearing deterioration may predict the future trend in hearing loss (Sakurai et al, 1991). However, further confirmation is needed.

Additional features include diplacusis, a difference in the perception of pitch between the ears (43.6%), and recruitment (56%) (Paparella, 1985).

Tinnitus tends to be nonpulsatile and variously described as whistling or roaring. It may be continuous or intermittent. Tinnitus often begins, gets louder, or changes pitch as an attack approaches. Following the attack there is frequently a period of improvement.

Investigations

Electronystagmography. Electronystagmographic recording of eye movements following caloric and rotational stimulation constitutes the most reliable method of assessing vestibular function. The caloric test is the most reliable for determining the involved ear. A significant caloric response reduction is found in 48% to 73.5% of Ménière's patients (Black and Kitch, 1980; Stahle and Klockoff, 1986). Complete loss of vestibular function, as elicited by the caloric test, is reported in 6% to 11% of patients.

Dehydrating agents. The assumption that an increase in endolymph volume, with its effect on labyrinthine membrane behavior, produces in part the hearing loss and vestibular deficit in Ménière's disease has lead to the administration of dehydrating agents (that is, urea, glycerol, and furosemide). The goal is to reduce the volume abnormalities in the inner ear and produce a measurable change in response; that is, improvement in behavioral audiometric test scores, reduction in summating potential negativity, as recorded with electrocochleography, and a change in the directional preponderance of the vestibuloocular response to rotational stimulation (Daumann et al, 1988; Kobayashi et al, 1989; Morrison et al, 1980).

The standard glycerol test consists of the administration of a loading dose of glycerin (1.5 g/kg mized with equal volume of juice) followed by serial audiograms over 3 hours (Klockoff et al, 1974). A positive test result consists of a 16% improvement in speech discrimination or a total pure-tone threshold shift of 25 dB at three consecutive frequencies (Weit et al, 1981).

Electrocochleography. The summating potential (SP), as recorded by electrocochleography in Ménière's patients, is larger and more negative. This is felt to reflect the distension of the basilar membrane into the scala tympani, causing an increase in the normal asymmetry of its vibration (Ferraro et al, 1983; Morrison et al, 1980). Because of the variability in absolute amplitude, the measure of the SP amplitude is not useful. However, the ratio of the amplitudes of the summating potential to the action potential (AP), SP:AP ratio, reduces the intertest variability, and a neal linear response results. The summating potential becomes larger in hydrops; thus the SP:AP ratio increases (Goin et al, 1982). Goin et al found elevated ratios in 62% of Ménière's patients and 21% of controls. The difficulty in obtaining reproducible recordings and the variability of the wave amplitudes noted with patient age, hearing loss, and stage of disease, as well as the availability of reliable, less invasive diagnostic methods, have resulted in electrocochleography rarely being used for this purpose (Ferraro et al, 1983).

Treatment

In a review of the available treatments for Ménière's disease, Torok (1977) found that all treatments, both medical and surgical, reported significant improvement in 60% to 80% of cases. The author interpreted this to indicate a common factor among diverse treatment modalities and suggested the possibility of a significant placebo effect. This is further supported by others who report 71% resolution of symptoms in patients refusing treatment (Silverstein et al, 1989). Many treatment modalities have been proposed for Ménière's disease. However, because of its extreme clinical variability, difficulty exists in quantifying treatment effectiveness. To this end, a set of guidelines for the assessment of treatments for Ménière's disease has been developed (see box) (see AAO-HNS, 1985; Pearson and Brackmann, 1985).

To date there is no proven cure for Ménière's disease. Current therapy is aimed at the reduction of its associated symptoms. The optimal treatment should stop vertigo, abolish tinnitus, and reverse hearing loss. Currently almost all proven therapy is directed at relieving the most distressing aspect of Ménière's disease, namely, vertigo.

Box: 1985 AAO-NHS criteria - summary

I. VERTIGO: Requires 6 months pre-treatment observation and 24 months post-treatment follow-up.

A. Vertigo control - numerical value:

(Average No of spells per month post-treatment (24 mos) x 100)/
(Average No of speells per month pre-treatment (6 mos))

Numerical value	Verbal score
= 0	complete control
= 1-40	substantial control
= 41-80	limited control
= 81-120	insignificant control
= > 120	patient worse

B. Disability status:

0 = No disability
1 = Mild disability - mild unsteadiness/dizziness that precludes working in a hazardous environment
2 = Moderate disability - unsteadiness/dizziness that results in necessity for a sedentary occupation
3 = Severe disability - symptoms exclude gainful employment.

III. HEARING: Pure tone average (PTA) = average threshold over 0.5, 1, 2, 3 kHz

Pre-treatment audiogram = worst PTA and Speech Discrimination (SD) over the 6 months before therapy

Post-treatment audiogram = worst PTA and SF over 24 months post-treatment

Hearing

Unchanged	±	10 dB PTA or ± 15% SD
Improved	>	10 dB PTA decrease or 15% SD increase
Worse	<	10 dB PTA increase or 15% SD decrease.

Medical therapy

Prophylaxis. Medical regimens aimed at prevention of vertigo employ the strategy of reduction in endolymph accumulation. Such protocols include dietary modification; intermittent dehydration (Beck, 1986a); diuretics (see below); enhanced microcirculation of the ear - that is, adenosine triphosphate, isosorbide, histamine, and betahistine (see below); and reduction in immune reactivity with steroid, immunoglobulin, and allergy therapy (Futaki

et al, 1988; Hughes et al, 1988b; Weit et al, 1981).

Dietary modification and diuretics. Salt restriction and diuresis are believed by many to be the best medical therapy for Ménière's disease (Jackson et al, 1981). At the Dizziness Unit at Sunnybrook Hospital, Toronto, a trial of a low-sodium diet along with a diuretic (hydrochlorothiazide) is routinely prescribed before more invasive therapies. The goal of salt restriction, diuretics, and hyperosmolar dehydration is to reduce endolymph volume by fluid removal or reduced production. The ability of this form of therapy to reduce the symptoms of Ménière's has been reported. In one representative study, diuretics controlled vertigo in 58% of patients and stabilized hearing in 69% (Klockoff and Lindblom, 1967; Klockoff et al, 1974). The authors also reported that there was no permanent cessation of disease progression, especially with respect to hearing loss. In contrast, double-blind studies have shown no effect of diuretics (Horner et al, 1989b; Van Deelen and Huizing, 1986). Carbonic anhydrase inhibitors, such as acetazolamide, were recommended based on the localization of carbonic anhydrase in the dark cells and the stria vascularis. However, their use has not proven to be clinically more effective than other diuretics (Beck, 1986b; Corvera and Corvera, 1989; Jackson et al, 1981; Klockoff and Lindblom, 1967; Shinkawa and Kimura, 1986).

Vasodilators. In the belief that Ménière's disease is the result of strial ischemia, vasodilating agents have been used (Wilmot and Menon, 1976). Betahistine, an oral preparation of histamine, is one such medication (Hommes, 1970; Wilmot and Menon, 1976). Quantitative improvement has not been shown consistently using these medications. Subjective improvement in hearing and vertigo was reported by non-Ménière's patients using betahistine (Oosterveld, 1984). Other agents utilized include the papaverine analog eupavarine, nicotinic acid, adenosinte triphosphate, and dipyridamole (Beck, 1986a; Mizukoshi et al, 1988a).

Symptomatic relief. Antivertiginous medications, antiemetics, sedatives, antidepressants, and psychiatric treatment have been reported to be beneficial in reducing the severity of vertigo and vegetative symptoms and improving tolerance of Ménière's symptoms (Beck, 1986b; Mizukoshi et al, 1988a; Pyykko et al, 1988; Weit et al, 1981).

Other therapies. Other therapies include holistic treatments such as acupuncture and herbal remedies (Eichner et al, 1986), as well as hypobaric pressure chamber therapy (Tjernstrom, 1986; Tjernstrom et al, 1980). Many of these therapies have been empirically applied and poorly documented. The rationale and results of some of the more conventional regimens will be discussed below.

Surgical treatment. Surgical treatment is reserved for those patients who have failed medical management and is estimated to be necessary in 10% of individuals (Brown, 1983).

Although vestibular ablative procedures cure the acute attacks of vertigo associated with Ménière's disease, it is important to appreciate that central adaptation is crucial for clinical recovery. In our experience only one third of patients are ultimately asymptomatic with respect to head movement-associated instability. Fully two thirds of patients continue to experience some degree of motion-induced instability. In a small percentage of patients, especially the elderly, central adaptation may be woefully inadequate, resulting in significant handicap. In those patients undergoing vestibular ablation, it is important that early

ambulation and head exercises be part of the treatment protocol (Fetter et al, 1988a and b).

Surgical treatment can be broadly classified into those procedures that spare hearing and those that do not. Within the former are those whose goal is the restoration of normal endolymph volume. These include the endolymphatic decompression procedures; that is, sac decompression (see below), the tack procedure (Cody, 1973; Cody and McDonald, 1983), and the Fick procedure (Fick, 1964). Other hearing conservative procedures are designed to ablate vestibular function in the affected ear without damage to the cochlea; that is, vestibular neurectomy (see below), intratympanic gentamicin and inner ear streptomycin (see below), cryosurgery (Horowitz et al, 1989), ultrasound (Almann and Hager, 1968), and cochlear dialysis (Morris and Morrison, 1989).

Hearing conservative nonvestibular ablative surgery

Endolymphatic sac decompression. The first endolymphatic sac procedure was performed by Geogre Portmann in 1926 (Portmann, 1987). Subsequent modifications of endolymphatic sac surgery have been developed. All such operations have a similar objective, namely, the reduction of endolymph volume via increased drainage or increased absorption. Variations include exposure (decompression) of the sac and insertion of materials such as Silastic sheeting, tubing, gelatin, or tissue stents to keep the mastoid drainage pathway patent (Futaki and Nomura, 1989; Kitahara et al, 1987; Glasscock, 1984; Morrison, 1979; Paparella and Sajjadi, 1987; Shea, 1966). The drainage pathway has been routed to the subarachnoid space by incision of the back wall of the sac (Glasscock et al, 1989c; Naito, 1962); by stenting of the subarachnoid drainage route with various tubes, including the one-way valve used by Arenberg (1987); and by the tubed sponge developed by Austin (1984; Huang and Lin, 1989).

The results of the above procedures are quite similar, both between studies as well as within studies where several techniques are compared. Complete resolution of vertigo is reported in about 50% to 75% of patients (Arenberg, 1987; Brackmann and Nissen, 1987; Monsell and Wiet, 1988). In a study comparing several methods of endolymph shunting. Glasscock et al (1989c) reported complete control of vertigo in 65% of patients, which fell to 50% in 10 years. Improvement or stabilization of hearing is reported in about 55% of patients across studies. Complications of surgery, excluding vertigo, are low and consist primarily of postoperative hearing loss in 1% to 2% of cases (Arenberg, 1987; Brackmann and Nissen, 1987; Paparella and Sajjadi, 1987). Wound infection, CSF leak, meningitis, and facial paralysis (permanent and transient) have been reported.

Other therapies. Temporal bone studies suggest that longitudinal flow of endolymph may be obstructed before the endolymphatic duct as a consequence of mechanical blockage of the utricular and saccular ducts. With intent to bypass this obstruction and improve the efficacy of endolymph drainage procedures, the cochleosacculotomy was developed to create a permanent fistula in the membranous labyrinth. This was done by introduction of a pick through the round window membrane with perforation of the saccule behind the oval window (Schuknecht, 1982b). Vertigo was reportedly controlled completely in 70% of patients. However, recurrence of symptoms and an unacceptable incidence of hearing loss have been major drawbacks (Brackmann, 1990; Giddings et al, 1991). The Fick and Cody tack procedures, previously mentioned, were similarly designed to create fistulas in the saccule via

the oval window. Results have not been consistent with these procedures, and they have not been generally accepted.

The efficacy of various endolymph drainage procedures has been seriously questioned. Foremost in this controversy is the sham study performed by Thomsen et asl (1981). In this study the authors performed a simple mastoidectomy with or without an endolymphatic shunting procedure. Follow-up was double blinded in that neither the patients nor the examining physician were aware of which procedure had been performed. Results were identical for the two operations. There was improvement in 73% of the patients undergoing a shunt procedure and in 80% of the sham group. These results have remained unchanged with subsequent follow-up (Bretlau et al, 1984; Thomsen et al, 1986). Further, these results compare with the average outcome of all treatments identified by Torok (1977), 70% to 80%.

Hearing conservative vestibular ablative surgery

Vestibular neurectomy. Eighth nerve and selective eight nerve section for the treatment of intractable vertigo secondary to unilateral vestibular disorders was first performed in the early part of the 1900s (Barber and Ireland, 1952; Dandy, 1928; Frazier, 1912; McKenzie, 1936). Although this procedure was very effective in controlling vertigo, it was replaced in popularity by the peripheral destructive procedures, such as labyrinthectomy. Selective vestibular neurectomy was introduced by House in 1961 via the middle fossa approach. Applying this procedure or modifications of it has resulted in complete resolution of vertigo in approximately 95% of patients (Fisch, 1984; Smyth et al, 1976). Although hearing was initially reported to stabilize or improve in 51% to 83% of patients (Brackmann, 1990), subsequent long-term study has demonstrated hearing loss consistent with the natural history of the disease process (Glasscock et al, 1989a). Complications of the middle fossa approach include temporary facial weakness (3% to 44%), total hearing loss (5%), subdural hematoma, CSF leak, and meningitis (Brackmann, 1990; Glasscock et al, 1991; Smyth et al, 1976). It has been suggested for technical reasons that the procedure be used only in patients under 60 years of age or in cases where retrolabyrinthine vestibular neurectomy has been unsuccessful (Brackmann, 1990).

Based on the anatomic studies by Streeter in 1907, selective section of the vestibular nerve from a posterior, suboccipital approach was first performed in 1931 by McKenzie (McKenzie, 1936). Silverstein reintroduced a modified version of the above, the retrolabyrinthine vestibular neurectomy (Silverstein et al, 1987, 1990). Complete remission of vertigo was reported in 82% of patients. Results were less than expected presumably because of inability to section vestibular fibers intermixed in the cochlear nerve. Investigators have modified the approach, enabling the surgeon to acess the vestibular nerve in the lateral portion of the internal auditory canal. Both the retrosigmoid and suboccipital approaches are used. Vertigo remission is now comparable to the middle fossa approach, with a 93% complete response (Kemink and Hoff, 1986; Silverstein et al, 1990). Complications of the retrolabyrinthine approach include CSF leaks (3% to 10%), facial paralysis (0% to 2%), and hearing loss (Brackmann, 1990; Glasscock et al, 1991; Silverstein et al, 1987). The retrolabyrinthine approach should carry approximately the same risk of postoperative infection (meningitis) and bleeding (subdural hematoma) as the middle cranial fossa and the suboccipital or retrosigmoid approach. Although the suboccipital approach has a very low incidence of CSF leak, it is associated with a significant incidence of postoperative headache

(Glasscock et al, 1991; Silverstein et al, 1990).

Chemical vestibular ablation. Aminoglycoside ototoxicity is well recognized. Streptomycin and gentamicin, by virtue of their more selective vestibular toxicity, have been instilled locally into the middle ear as a treatment for unilateral Ménière's disease (Beck, 1986; Beck and Schmidt, 1978; Kozaki et al, 1988; Lange, 1976; Odkvist, 1988; Schuknecht, 1956). The initial studies utilized streptomycin. Vertigo was abolished in 60% of patients; however, hearing was lost in the majority (Schuknecht, 1956). Gentamicin has been used subsequently. The drug is instilled through the tympanic membrane into the middle ear via a small needle or an indwelling catheter. Protocol for drug administration has varied. Several investigators have described daily instillation until signs of vestibular or cochlear loss are evident. Vertigo control has been reported at approximately 90%, with hearing loss in 6% to 30% of patients (Odkvist, 1988). In an ongoing study at Sunnybrook Health Science Centre using a fixed protocol of intratympanic gentamicin (17 mg per dose given three times daily with a 12-dose total), 90% vertigo control has been attained based on 2-year follow-up according to the AAO-NHS 1985 guidelines. Treatment has been carried out on an outpatient as well as an inpatient basis. Profound hearing loss was precipitated in 8.9% of patients (Nedzelski et al, 1992a, 1992b).

Results of treatment consisting of direct injection of aminoglycosides into the perilymph/endolymph via an opening created in the horizontal semicircular canal, first described by Shea (1988), have recently been published (Morris et al, 1990). Good control of vertigo is reported; however, hearing loss appears significant. One recent preservation reported substantial hearing losses after infusion in more than one third of the patients, most to the severe or profound level, which prompted several of the participants in the study to abandon the procedure (Monsell et al, 1991).

Other procedures. Other hearing preservation procedures include cryotherapy (Horowitz et al, 1989; Wolfson, 1984), and ultrasound treatment (Kossoff et al, 1967). Although these treatment modalities are not commonly performed, some physicians continue their use in clinical treatment (Dickens and Graham, 1990; Peron et al, 1983).

Nonhearing conservative vestibular ablative therapy (labyrinthectomy). Labyrinthectomy is an effective treatment for vertigo in Ménière's patient when serviceable hearing is absent (pure-tone average > 60 dB and discrimination < 50%) (Graham and Kemink, 1984). Several techniques are described: ablation with hypertonic saline (Colletti et al, 1989), as well as transcanal, transmastoid, and translabyrinthine cochleovestibular neurectomy (Beneke et al, 1986; Graham and Cotton, 1980; Graham and Kemink, 1984; Schuknecht, 1957). Some authors advocate destruction of Scarpa's ganglion on the basis of possible development of a neuroma after labyrinthectomy (Linthicum et al, 1979). This has been questioned by others (Schuknecht, 1982a). Excellent results are reported using all the aforementioned techniques, with resolution of symptoms in greater than 90% of cases.

In view of the proven effectiveness of cochlear implants and the possibility of bilateral occurrence of Ménière's disease, labyrinthectomy as treatment should be questioned.

Bilateral Ménière's disease (parenteral streptomycin). The objectives of treatment in the patient with symptomatic bilateral Ménière's disease are relief of vertigo and auditory preservation. Streptomycin sulfate is primarily vestibulotoxic. It can be administered parenterally with resultant ablation of peripheral vestibular function while leaving hearing largely unchanged (Hanson, 1951; Wilson and Schuknecht, 1980; Schuknecht, 1956). Although early use included instances of unilateral disease, the treatment is now indicated for incapacitating bilateral Ménière's disease only (Glasscock et al, 1989c).

Whereas patients initially received streptomycin until the ice water response was absent (that is, 1.5 to 3.5 g/day for 17.5 days average and 39 g mean total drug dose) (Wilson and Schuknecht, 1980), others have suggested a lesser total dose, that is, 20 g total (Glasscock et al, 1989b; Moretz et al, 1987; Graham et al, 1984; Silverstein et al, 1984). The rationale for the latter is based on preserving some degree of vestibular function such that the disabling oscillopsia and vestibular ataxia are less severe. Further, should symptoms recur, retreatment with a lesser dose is an option.

Otologic syphilis

Syphilis has been estimated to account for up to 6.5% of all cases of unexplained sensorineural hearing loss. It was found to be the cause of symptoms in 7% of patients said to have Ménière's disease (Becker, 1979; Darmstadt and Harris, 1989). Hearing loss is a major symptom in 38% of patients suffering from congenital syphilis and 80% of symptomatic neurosyphilis patients (Darmstadt and Harris, 1989; Nadol et al, 1975; Tamari and Itkin, 1951).

Otologic involvement can be grouped into two categories: early syphilis, with the symptoms occurring within 2 years of exposure, and late otologic syphilis, when the disease becomes symptomatic after 2 years (Darmstadt and Harris, 1989; Durham et al, 1984; Karmody and Schuknecht, 1966). Vestibular symptoms are noted in both. The pattern of presentation varies (Becker, 1979).

Vestibular symptoms are less frequent in early syphilis. When present, they vary from mild imbalance to symptoms of protracted vertigo with vegetative features lasting days. Hearing loss is rapid, bilateral, and frequently profound (Darmstadt and Harris, 1989). Late-stage otologic syphilis, whether acquired or congenital, can present up to 50 years or more after exposure (Patterson, 1968). This later form of the disease has cochleovestibular symptoms indistinguishable from Ménière's disease (Becker, 1979; Karmody and Schuknecht, 1966). Episodic vertigo lasting minutes to hours and fluctuating, asymmetric, sensorineural hearing loss, with recruitment and tinnitus, are described (Darmstadt and Harris, 1989; Durham et al, 1984; Morrison and Booth, 1988).

Temporal bone findings in syphilis vary with the stage of disease. In early syphilis a treponeme-induced meningoneurolabyrinthitis occurs that incites an inflammatory response within the vestibular and cochlear nerve, as well as the labyrinth (Morrison and Booth, 1988; Nadol et al, 1975). Systemic symptoms frequently overshadow the otologic and include signs of meningitis. In the case of early congenital syphilis, systemic involvement may prove fatal (Darmstadt and Harris, 1989). Late-onset syphilis, whether acquired or congenital, is a distinct entity (Becker, 1979). Inner ear symptoms are a result of otic capsule involvement.

Obliterative endarteritis and mononuclear infiltration, producing a periostitis (endosteal inflammation found especially in the semicircular canals) and gummatous osteitis/periostitis (a gumma has central necrosis with surrounding lymphocytic infiltration and vascular occlusion), have been documented. These in turn cause atrophy and fibrosis of the membranous labyrinth, narrowing of the endolymphatic duct and sac, and subsequent development of endolymphatic hydrops (Becker, 1979; Karmody and Schuknecht, 1966; Linthicum and El-Rahman, 1987; Morrison and Booth, 1988). Osteitis within the ossicles can result in fixation leading to an additional conductive hearing loss (Darmstadt and Harris, 1989; Nadol, 1987).

Interstitial keratitis is a feature of 90% of patients with late-onset otologic syphilis, whether congenital or acquired (Yanoff and Fine, 1989). The triad of sensorineural hearing loss, interstitial keratitis, and notched incisors, so-called Hutchinson's triad, is an exclusive feature of late congenital syphilis (Darmstadt, 1989).

Hearing loss can be unilateral, bilateral, sudden, of gradual progression, or fluctuating (Hughes and Rutherford, 1986; Karmody and Schuknecht, 1966). Hennebert's sign (nystagmus and vertigo produced by pressure change on the ear, felt to be caused by deformation of a softened, gummatous otic capsule) and Tullio phenomenon (nystagmus caused by loud noise), although often encountered in syphilis, are not pathognomonic of the disease (Hughes and Rutherford, 1986; Morrison and Booth, 1988).

Although the treponeme has been isolated from sampled perilymph (Weit and Milko, 1975), serologic testing is the only method by which syphilis is diagnosed (Hughes and Rutherford, 1986).

The VDRL (Venereal Disease Research Laboratory) and RPR (rapid plasma reagin) nontreponemal tests are easily performed and relatively inexpensive. They are the most commonly used screening tests for syphilis. Sensitivity varies with the stage of disease. In view of the fact that otologic symptoms occur in late-stage disease, nontreponemal tests detect only 70% of patients (Hughes and Rutherford, 1986). They are useful in observing therapeutic response by change in titer and loss of reactivity. On the other hand, treponemal tests - for example, FTA-ABS (free treponemal antigen absorption) - that detect the presence of the organism are far more more sensitive, detecting 95% or more of patients with late disease. The FTA and its variations are the tests of choice for the detection of otologic syphilis (Becker, 1979; Hughes and Rutherford, 1986; Morrison and Booth, 1988). In view of the fact that neurosyphilis is rarely noted in the presence of otologic disease, lumbar puncture is not necessarily warranted (Becker, 1979).

Treatment of otologic syphilis varies. Some authors feel that in the early stages of the disease, treatment with antibiotics alone; that is, benzathine penicillin, 2.4 million units IM, provides a cure, frequently resulting in hearing improvement (Darmstadt and Harris, 1989). Additional use of a 10-day course of steroids is advocated by others (Balkany and Dans, 1978; Morrison and Booth, 1988).

Although consensus is lacking as to exact drug treatment for late-stage disease, it is generally agreed that there is a need for protracted therapy with both antibiotics and steroids. Difficulty in eradication of the infection despite adequate antibiotic therapy is ascribed to a prolonged dividing time for the treponeme in late-stage disease (Darmstadt and Harris, 1989; Mack et al, 1969; Weit and Milko, 1975). Antibiotic regimens of varying duration have been suggested; that is, 3 weeks to 1 year. One treatment used for neurosyphilis has been 1.8 million units of procaine penicillin IM daily with 500 mg of probenecid by mouth four times daily for 17 to 21 days. It is suggested that otic syphilis and neurosyphilis are comparable with respect to treatment (Darmstadt and Harris, 1989). Prednisone, 40 to 60 mg/day, is given for at least 2 weeks before tapering. Maintenance steroid therapy may be necessary. Improvement in hearing, particularly of speech discrimination, is noted in 35% to 50% of cases, along with resolution of vestibular symptoms (Becker, 1979; Darmstadt and Harris, 1989).

Delayed endolymphatic hydrops

Delayed endolymphatic hydrops was first described independently by Nadol et al and by Wolfson in 1975. It is characterized by attacks of vertigo identical to those of Ménière's disease in a patient who has had a previous, profound loss of hearing in one or both ears. The reported time between the loss of hearing and the development of episodic vertigo varies widely (1 to 74 years) but is invariably separate from the initial hearing loss (Hicks and Wright, 1988; LeLiever and Barber, 1980; Paparella and Mancini, 1983; Schuknecht et al, 1990).

Causes of the initial hearing loss vary. Head trauma (acoustic and physical), viral labyrinthitis (mumps, influenza), mastoiditis, meningitis, diphtheria, measles, and early childhood deafness of unknown etiology are all reported (Hicks and Wright, 1988; Lambert, 1985; LeLiever and Barber, 1980; Paparella and Mancini, 1983; Schuknecht et al, 1990; Ylikoski, 1988).

The presence of hydrops is supported by several studies. Analysis of fluid aspirated via a stapedotomy has been confirmed to be endolymph. This is felt to be a consequence of the distended saccule being adherent to the underside of the stapes footplate (LeLiever and Barber, 1980). Dehydrating agents, such as Lasix, improve the vestibular response of the affected ear, the so-called Lasix test (Futaki et al, 1984; Hicks and Wright, 1988).

Temporal bone studies demonstrate endolymphatic hydrops in ipsilateral and contralateral forms of the disorder. In the case of ipsilateral disease, hydrops is superimposed on evidence of prior cochlear damage; that is, from a virus (Lindsay et al, 1954; Paparella and Mancini, 1983).

The cause of delayed endolymphatic hydrops is unknown; however, it is assumed that it develops as a consequence of a preceding insult to structures necessary for endolymph homeostasis (Paparella and Mancini, 1983). The delay possibly reflects the time required for humans to develop hydrops after an insult (LeLiever and Barber, 1980).

Diagnosis is based on a Ménière's-like pattern of presentation of vertigo and the history of prior severe hearing loss. The opposite ear is normal in every respect.

There are reported cases of Ménière's syndrome developing in patients who have experienced a profound contralateral hearing loss years previously (Futaki et al, 1984; Schuknecht et al, 1990). It has been suggested that such individuals may have experienced a form of delayed endolymphatic hydrops. It is postulated that the agent causing deafness in one ear, perhaps viral, was responsible for initiating a series of events in the contralateral, healthy ear, which, although not causing deafness at the time of infection, resulted in hydrops years later (Schuknecht et al, 1990).

Labyrinthectomy has been used with excellent results (Schuknecht et al, 1990). In those instances where delayed hydrops has occurred in the only hearing ear, conservative medical management is recommended (Hicks and Wright, 1988; LeLiever and Barber, 1980).

Cogan's syndrome

Cogan's syndrome was first described in 1945 and is characterized by interstitial keratitis, Ménière's-like hearing loss, and vestibular symptoms, as well as nonreactive tests for syphilis (Cogan, 1949; Cogan and Williams, 1945). The syndrome is divided into typical and atypical forms based on specific ocular findings. The typical form includes interstitial keratitis (IK), whereas the atypical form has ocular findings of scleritis, episcleritis, papilledema, and retinal detachment (Haynes et al, 1980). Ten percent of patients with Cogan's syndrome have the atypical form (Benitez et al, 1990). Multisystem involvement, including the central nervous system and inflammatory vascular disease, is a feature of Cogan's disease. Of the two forms, systemic features are noted more commonly in atypical Cogan's disease (Haynes et al, 1980). In the typical form the heart and lungs are the usual site of systemic involvement, including aortitis, aortic insufficiency, pleuritis, pericardial effusion, coronary arteritis, and myocardial infarction (Haynes et al, 1980; Hughes et al, 1983). In atypical Cogan's, systemic features arise from a systemic vasculitis; that is, polyarteritis nodosa, arthritis, glomerulonephritis, and gastrointestinal problems.

Ocular and inner ear findings occur concurrently or within 6 months of each other (Haynes et al, 1980). Ocular complaints include photophobia, blurred vision, lacrimation, and pain. These symptoms may be unilateral or bilateral (Peeters et al, 1986). Onset is usually sudden, with gradual resolution. There may be recurrences for years. Ocular pathologic findings include infiltration of the cornea with lymphocytes and plasma cells as well as corneal neovascularization (Haynes et al, 1980).

Balance-related symtoms are similar to those of Ménière's disease and consist of sudden attacks of true vertigo, with ataxia and vegetative symptoms. Progression to complete absence of vestibular function, manifested by ataxia and oscillopsia, is common (Haynes et al, 1980; Peeters et al, 1986).

The initial configuration of the hearing loss is similar to that of hydrops (a peak-shaped audiogram) (Benitez et al, 1990; Haynes et al, 1980). In addition, administration of glycerol has been shown to result in a transient improvement in hearing. However, unlike Ménière's, hearing loss is bilateral and progressive, without spontaneous improvement and

frequently becoming profound.

Endolymphatic hydrops and abnormal bone formation have been demonstrated in temporal bone studies (Wolff et al, 1965). Inflammatory infiltration of the spiral ligament of the cochlea and cellular debris within the endolymphatic spaces are also noted.

Cogan's syndrome is thought to be of autoimmune basis. This suspected etiology is based on lymphocyte transformation tests, migration inhibition to inner ear tissues, the associated incidence of rheumatologic diseases, and presence of lymphocytes and plasma cells in the ear and eye, as well as the clinical response to systemic steroids (Haynes et al, 1980; Hughes et al, 1983).

Clinically, patients often report having had an upper respiratory infection within 7 to 10 days of initial onset of Cogan's syndrome (Haynes et al, 1980). Raised IgG and IgM titers to *chlamydia* are reported in association with active Cogan's disease. Further, antibody titers are noted to decrease with remission of disease. For this reason it has been suggested that an acute infection, perhaps chlamydial, promotes sensitization of the immune system to "self" and leads to immune-mediated disease (Haynes et al, 1980).

Corticosteroids, both systemic and topical, are the accepted treatment. In addition, cyclophosphamide has been recommended by some authors (Hughes et al, 1983). Generally, systemic steroid treatment leads to hearing improvement, as opposed to near uniform deafness without treatment (Haynes et al, 1980, 1981). Hearing improvement is best realised if therapy is instituted within the first few weeks of onset (see also Chapter 164).

Recurrent vestibulopathy

The clinical term *recurrent vestibulopathy* was first used by LeLiever and Barber in 1981 to describe a particular clinical entity. Typically, individuals describe attacks of vertigo similar to those of Ménière's disease without audiologic symptoms such as tinnitus, hearing loss, or a sensation of fullness in the ear. Focal neurologic features are absent. The patient typically describes the onset of vertigo as sudden, lasting minutes to hours (typically less than 24 hours) and recurring at variable intervals. Before the introduction of this descrptive diagnosis, patients presenting with this pattern of disease were classified as having "vestibular Ménière's disease". However, clinical evidence tends to support the conclusion that "vestibular Ménière's disease" is not a distinct entity (Harker, 1989).

Vertigo was found to be the presenting symptom of Ménière's disease in 50% of 318 patients studied retrospectively (Thomas and Harrison, 1971). A second symptom, either hearing loss or tinnitus, began within 2 years in 76% of patients and within 5 years in 97%. Of 83 patients with recurrent vestibulopathy who were observed for a mean of 9.5 years (LeLiever and Barber, 1981; Rutka and Barber, 1986; Wallace and Barber, 1983), the diagnosis of recurrent vestibulopathy was changed to Ménière's disease in only 14% of the cases. The natural history of recurrent vestibulopathy is distinctly different from that of Ménière's disease (LeLiever and Barber, 1981).

No cause for recurrent vestibulopathy is known at this time. A viral etiology has been postulated on the grounds of its similarities to vestibular neuronitis (Rutka and Barber, 1986). In addition, there is a case report describing two cases of recurrent vestibulopathy in a family with a reference case of Ramsay Hunt syndrome (Longridge, 1989).

Recurrent vestibulopathy is relatively common. It is diagnosed in 9.3% of patients seen at the Sunnybrook Health Science Centre Dizziness Unit (Nedzelski et al, 1986). The mean age of onset is 37 years versus 45 years for Ménière's. Sex distribution is equal.

A unilateral caloric response reduction was found in 22% of these patients. This is significantly less than the 50% incidence noted in the Ménière's patient population.

The prognosis for uneventful recovery is good. Seventy percent of patients initially diagnosed as having recurrent vestibulopathy in the study went on to spontaneous resolution of vertigo over the follow-up period of 9.5 years (LeLiever and Barber, 1981). Eight percent continued to experience disease symptoms. Nine percent noted resolution of recurrent vestibulopathy; however, they realized symptoms or signs of benign type paroxysmal positional vertigo. None of the patients initially diagnosed as having recurrent vestibulopathy developed symptoms indicative of central nervous system disease.

Symptomatic and supportive treatment, when required, proved sufficient. A small number of patients were given diuretics with no improvement. Vestibular neurectomy was carried out on one patient, which proved curative.

Vertigo Lasting Seconds

Benign paroxysmal positional vertigo (BPPV)

BPPV is generally agreed to be one of the most common of peripheral vestibular disorders. It represents 17% of patients seen in the Dizziness Unit at Sunnybrook Health Science (Nedzelski et al, 1986). The hallmark of this disorder is the onset of brief (seconds) spells of often severe vertigo, which are experienced only with specific head positions.

Background

History. The disorder was first described in 1921 by Barany. The realization that vertigo occurred only when the sufferer lay down on a particular side led him and others to associate the problem with the inner ear (Schuknecht, 1969). In 1952 Dix and Hallpike reported this entity in a large group of patients. They described the Hallpike maneuver for eliciting the classic pattern of nystagmus and its associated symptoms.

Early temporal bone studies from afflicted patients were all noted to demonstrate degeneration of the utricle. In 1969 Schuknecht reported the presence of basophilic debris adherent to the cupula of the posterior semicircular canal of the affected ear, while the opposite ear had none (Fig. 181-3). He suggested that the debris consisted of remnants of displaced utricular otoconia, and he introduced the term *cupulolithiasis* to describe this entity. Schuknecht postulated that utricular degeneration liberated otoconia, which then floated downward into the inferior-most region of the vestibule, the ampulla of the posterior

semicircular canal (Katsarkas and Kearney, 1990). As a result, these deposits alter the specific gravity of the cupula, changing its response characteristics from a purely angular acceleration detector to one that is stimulated by linear movements and gravity.

The suggested pathogenesis is consistent with observed clinical findings, experimental studies of peripheral vestibular function, and most importantly, results of therapeutic manipulations directed at modifying the response of the posterior semicircular canal (Gacek, 1991).

Nystagmus generated when a patient is placed in the Hallpike position, as viewed by the observer, is torsional. The top of the eye beats toward the undermost ear, so-called geotropic (earth-seeking) nystagmus (Baloh et al, 1987; Barber, 1984; Katsarkas and Outerbridge, 1983). The nystagmus is counterclockwise, as viewed by the observer, when the head is hanging to the right, and clockwise in the head-hanging-left position. Isolated stimulation of the posterior semicircular canal in animals results in contraction of the ipsilateral superior oblique and contralateral inferior rectus muscles. The resultant pattern of nystagmus is similar to that seen with the Hallpike maneuver if the undermost ear is stimulated (Cohen et al, 1964; Gacek, 1985).

Incidence. BPPV is the most common cause of vertigo seen by the otolaryngologist. The incidence is difficult to estimate given the benign course of the disease. It is thought to vary from 10.7 per 100.000 to 17.3 per 100.000 population in Japan (Mizukoshi et al, 1988a). BPPV as a diagnosis is almost twice as frequent as is Ménière's disease at the Sunnybrook Health Science Centre Dizziness Unit.

In studies of large groups of patients, the mean age of onset was in the fourth and fifth decades with a wide range, 11 to 84 years in one study (Baloh et al, 1987; Mizukoshi et al, 1988a). Overall, the incidence increases with age (Bloom and Katsarkas, 1989). There appears to be a slightly increased incidence of BPPV among women, approximately 1.6:1 (Bourgeois and Dehaene, 1988).

Diagnosis

History. The patient with BPPV experiences severe vertigo associated with change in head position. The most frequently cited occurrence of this symptom follows rolling over or getting into bed and assuming a supine position. Frequently a specific side is volunteered; for example, "the dizziness comes when I role over to my right side but not to the left". Patients will also experience similar symptoms on rising from a bending position, looking up to take an object off a shelf, tilting their heads back to shave, or turning rapidly.

Symptoms occur suddenly and last in the order of seconds, but never in excess of a minute (Baloh et al, 1987). The subjective impression of attack duration reported by the patient is frequently longer.

These single bouts of vertigo are frequently clustered in time and separated by remissions lasting months or more (Barber, 1973). The patient may also report that periods of active disease may be associated with constant feelings of light-headedness, worsened by head movement (Epley, 1980). Individuals also describe chronic balance problems (Black and

Nashner, 1984; Katsarkis, 1991; Katsarkas and Kearney, 1990). These may be worse on awakening in the morning.

Most cases of BPPV have no identifiable etiology. The most common identifiable cause of BPPV is felt to be closed head injury. Other cited predisposing events include vestibular neuronitis, infections, and following surgery; that is, stapedectomy (Baloh et al, 1987). In our experience, nearly 15% of patients suffering from vestibular neuronitis will later develop BPPV. It is noteworthy that we, as well as others, have found the entity to rarely follow Ménière's disease and recurrent vestibulopathy (Baloh et al, 1987).

Physical findings. A clear-cut diagnosis of this disease entity is made by (1) observing the classic eye movements in association with the Hallpike maneuver and (2) a suggestive history. The Hallpike maneuver is carried out as follows. The patient is positioned on an examination table such that when placed supine the head extends over the edge. The patient is lowered with the head supported and turned 45 degrees to one or the other side. The eyes are carefully observed. If no abnormal eye movements are seen, the patient is returned to the upright position in 30 seconds. The maneuver is repeated with the head in the opposite direction and finally with the head extended supine (Bourgeois and Dehaene, 1988; Gacek, 1985). The patient's symptoms are carefully noted should they occur.

The nystagmus pattern that is pathognomonic of BPPV consists of the following: (1) nystagmus is rotational and geotropic; (2) there is a latency of onset (seconds); (3) duration of nystagmus is short (< 1 minute); (4) vertiginous symptoms are invariably associated; (5) the nystagmus disappears with repeated testing (fatigable); and (6) nystagmus reverses its direction on return of the head to the upright position (Barber, 1973; Epley, 1980).

There are some observed exceptions to the classic nystagmus detailed above (McClure, 1985). Studies have identified a small percentage of BPPV sufferers (17%) who have elicitable nystagmus of the above type in both lateral head positions (Baloh et al, 1987; barber, 1978; Gacek, 1991). These individuals are felt to have bilateral BPPV. Because symptoms and physical findings can independently vary, caution should be exercised when considering treatment. It is thus suggested that all such patients be examined on several occasions before any surgical intervention.

Test results. Electronystagmography (ENG) is incapable of recording the rotational eye movements associated with BPPV. The resultant eye tracings solely reflect the associated vertical and horizontal components of the torsional eye movements (Katsarkos, 1991; Katsarkos and Outerbridge, 1983). The above factors highlight the need for careful clinical observation.

Treatment

Current treatment available for BPPV varies from an expectant attitude to surgical intervention. The majority of patients experience spontaneous resolution of BPPV within several months of onset (Gacek, 1985). This is particularly true of BPPV occurring subsequent to head injury. A small number will have persistent, near disabling symptoms lasting more than 1 year. These individuals are less likely to experience spontaneous recovery and therefore are considered candidates for operative intervention. Many patients experience

recurring bouts of positional vertigo, lasting weeks to months, with remissions lasting months to years. Suggested treatment is therefore tailored to each individual patient (Longridge and Barber, 1978).

Medical treatment

Drugs. Vestibular suppressants are generally ineffective, given the short-lasting, intense nature of the vertigo (McClure and Willett, 1980).

Liberatory maneuvers. In an effort to "displace" debris from the ampulla of the posterior semicircular canal, several head movement protocols have been described (Brandt and Daroff, 1980; Semont et al, 1988, 1989). Abolition of vertiginous symptoms has been reported in over 85% of individuals.

The Brandt maneuvers are performed by the patient, whereas the Semont maneuver is performed by the physician. In our opinion the latter should not be performed unless the patient has been accompanied by an escort. We find that it is not unusual for significant instability, nausea, and vertigo to be noted for as long as 24 to 48 hours following the maneuver.

Surgical treatment. Surgical procedures devised to definitely rid the patient of the vertigo associated with this entity include singular neurectomy, vestibular neurectomy, and, most recently, posterior semicircular canal occlusion. Labyrinthectomy is only considered in a coexisting deaf ear and a relatively youthful patient.

The singular neurectomy has been elegantly described by Gacek (1974). Complete resolution of symptoms has been reported in 80% to 97% of patients. Sensorineural hearing loss varies from 4% to 6% (Gacek, 1991; Meyerhoff, 1985; Silverstein and White, 1990).

Based on animal models, Money was first to determine that semicircular canal occlusion successfully abolished all responses to stimulation. Posterior semicircular canal occlusion has resulted in abolition of symptoms in humans with BPPV (Parnes and McClure, 1990). Vertigo control has been noted in all patients of a small series recently reported (Parnes and McClure, 1991). Transient sensorineural hearing losses have been reported, which generally return to preoperative acuity over ensuing weeks. This is believed to be a consequence of a reactive inflammatory serous labyrinthitis. A recent report using an argon laser to produce fibrous occlusion of the membranous canal has been published (Anthony, 1991).

Vertigo Lasting Days to Weeks

Vestibular neuronitis

Vestibular neuronitis typically presents with dramatic, sudden onset of vertigo and attendant vegetative symptoms (Silvoniemi, 1988). Typically, the dizziness lasts days, with gradual, definite improvement throughout the course. Balance-related complaints, particularly caused by/or related to rapid head movements, may be present for months after resolution of the acute disease. Paroxysmal positional vertigo occurs subsequently in a small percentage

of patients (Bance and Rutka, 1990; Lindsay and Hemingway, 1956; Schuknecht and Kitamura, 1981). Vestibular neuronitis has been known to recur, with some patients describing similar, usually less intense, attacks for years (Schuknecht and Kitamura, 1981). Bilateral disease has been described and must be considered in the differential diagnosis of bilateral vestibular loss. Vestibular neuronitis is not associated with subjective changes in hearing or any focal neurologic complaints (Dix and Hallpike, 1952). Previously this clinical entity was referred to as labyrinthitis. Given the uniform absence of auditory symptoms, a more accurate clinical term is *vestibular neuronitis.*

A documented caloric response reduction is frequently the only way to identify the side of involvement (Corvera and Davalos, 1985; Silvoniemi, 1988).

Pathologic studies of temporal bones from patients believed to have experienced this clinical entity reveal vestibular nerve degeneration with sparing of the peripheral receptor structures (Friedman and House, 1980; Schuknecht and Kitamura, 1981).

Cause of the degeneration is not established. Infection with one of the neurotropic viruses, such as herpes virus, is felt to be responsible.

The term *epidemic vertigo* is historically synonymous with vestibular neuronitis. Published reports of vestibular neuronitis within households or following exposure to patients with vertigo or upper respiratory infection are responsible for this term (Dalsgaard-Nielsen, 1953; Schuknecht and Kitamura, 1981). The reported incidence of preceding infection varies from 23% to 100% (Silvoneimi, 1988). Herpes virus has been implicated as the causative agent based both on the findings of increased serum titers (Matsuo, 1986) and on animal experiments (Davis and Johnsson, 1983; Falser et al, 1987). Vertigo coincident with recurrence of cutaneous zoster has been reported (Bance and Rutka, 1990). The similarity between the temporal bone pathologic condition induced by zoster and that of vestibular neuronitis has been noted (Lindsay and Hemingway, 1956; Schuknecht and Kitamura, 1981).

A complete or partial loss of caloric responsiveness of the involved ear is the most consistent finding. Nystagmus, if present, is direction appropriate for the involved ear (Silvoniemi, 1988). Not infrequently, recovery of the caloric response is noted (Schuknecht and Kitamura, 1981).

Subjectively, hearing remains normal. However, in a study of such patients with normal conventional audiograms, a hearing loss was found in the higher frequencies; that is, 10 to 15 kHz (Rahko and Karma, 1986). On the basis of this finding, the authors suggest that vestibular neuronitis is a feature of a more generalized polyneuritis. Further evidence of the more general nature of the disease is provided by auditory brain-stem response testing, with results suggesting central nervous system involvement (Corvera and Davalos, 1985). Such findings have not been uniformly confirmed (Bergenius and Borg, 1983).

Treatment is supportive and symptomatic for vertigo and related vegetative symptoms. As in all acute peripheral vestibular losses, early ambulation is encouraged.

Vertigo of Variable Duration

Inner ear fistula

Inner ear fistula is defined as an abnormal communication between the perilymphatic space and the middle ear or an intramembranous communication between endolymphatic and perilymphatic spaces. However, controversy exists with respect to this entity. The inability to reliably predict the presence of a fistula before surgical exploration, as well as the lack of a standard criteria for recognizing a fistula intraoperatively, has resulted in confusion and even doubt as to the existence of symptomatic fistulization of the membranous labyrinth (Shelton and Simmons, 1988; Singleton and Weider, 1987). Because fistulas have been identified intraoperatively and their repair resulted in symptomatic and objective improvement, this diagnosis must be kept in mind in the evaluation of the vertiginous patient (Rizer and House, 1991; Ruben and Yankelowitz, 1989).

The clinical presentation of inner ear fistula may vary from mild and inconsequential to severe and incapacitating. Inner ear fistulas mimic other disorders, such as Ménière's disease, infection, acoustic neuroma, and lesions of the central nervous system (House et al, 1991; Meyerhoff and Yellin, 1990). Therefore inner ear fistula should always be considered as a possibility following trauma to the inner ear. The mechanism of trauma can vary (that is, barotrauma, penetrating trauma, and so on), but the result is a rupture of the limiting membranes of the labyrinth. The resultant leakage of perilymph into the middle ear or an abnormal communication between the endolymphatic and perilymphatic spaces results in vestibular or auditory symptoms (Seltzer and McCabe, 1986; Simmons, 1967). Surgery is reported to be the most common cause, that is, poststapedectomy (Seltzer and McCabe, 1986). Other mechanisms include head trauma, explosive blast, barotrauma, and following physical exertion. There is little likelihood of a fistula occurring spontaneously. However, congenital malformations, including Mondini's deformity, may predispose individuals to fistula formation (Reilly, 1989). Others have suggested that fistula should be ruled out in cases of congenital hearing loss (Reilly, 1989; Ruben and Yankelowitz, 1989).

Reports vary as to the site, oval versus round window (Glasscock et al, 1987; Seltzer and McCabe, 1986). In studies that have included poststapedectomy fistulas, the oval window site has naturally been higher than in studies that have not.

Attempts to identify the prevalence and to characterize auditory or vestibular symptoms have been inconclusive. Some studies report vestibular symptoms as the major presenting complaint (Shelton and Simmons, 1988), whereas others indicate hearing loss equal to or more common than balance-related symptoms (Rizer and House, 1991).

Sensorineural hearing losses vary from an isolated high-frequency loss to a low frequency or flat one. Speech discrimination test results are not characteristic. Both the pure-tone threshold and speech discrimination scores have been noted to fluctuate. Isolated mild conductive losses have been noted (Singleton and Weider, 1987). Glasscock et al (1987), in a series of patients who had undergone exploration for fistulas, were unable to identify an audiometric profile in patients with proven fistulas from those without.

Audiologic tests felt to be helpful in the diagnosis include electrocohleography. This demonstrates a larger summating potential from endolymph/perilymph disequilibrium. However, the test is not sensitive or specific for perilymph fistula (Meyerhoff and Yellin, 1990). Improvement in pure-tone threshold or speech discrimination after the patient has been in Trendelenburg position for 30 minutes, the so-called Fraser test, is considered useful by some (Singleton and Weider, 1987).

Vestibular symptoms are also variable and include episodic incapacitating vertigo, equivalent to a Ménière's attack, positional vertigo, motion intolerance, or occasional dysequilibrium. Disequilibrium following increases in CSF pressure such as nose blowing or lifting (so-called Hennebert's phenomenon) has been noted, as has vertigo following exposure to loud noises (Tulio's phenomenon) (Healy et al, 1976; Meyerhoff and Yellin, 1990).

Results of vestibular testing are nondiagnostic. The most consistent abnormality seen is a unilateral reduced caloric response in the affected ear (Glasscock, 1987; Rizer and House, 1991). Other authors report variable ENG abnormalities (Seltzer and McCabe, 1986).

A very useful examination is the fistula test, which should be done in every suspected individual. The test is done as follows: positive pressure is introduced into the suspect ear, either by rapid pressure on the tragus, compressing the external canal, or via a pneumatic otoscope, while observing the eyes. A positive fistula sign consists of conjugate contralateral slow deviation of the eyes followed by three or four ipsilaterally directed beats of nystagmus. In those patients who present with a clinically suspicious history and a positive fistula sign, surgical exploration is recommended. It should be recognized that the specificity and sensitivity of the above test are variable (Rizer and House, 1991; Weider and Johnson, 1988). Some feel posturography is useful in detecting this entity, whereas others find it inconclusive (House et al, 1991).

It has recently been suggested that the determination of the free amino acid content of fluid, sampled from the middle ear, could be used to differentiate the presence of perilymph from mucosal secretion. The clinical application of this technique has yet to be established (Schweitzer et al, 1990; Silverstein, 1991).

Fiberoptic exploration of the middle ear, either by way of a myringotomy or with flexible endoscopic examination via the eustachian tube, is also under evaluation.

Treatment of patients with suspected inner ear fistulas should consist of the following: (1) bed rest, (2) head elevation, (3) laxatives to reduce risk of increased intracranial pressure, and (4) monitoring of both hearing and vestibular function. In those instances where hearing loss worsens or vestibular symptoms persist, surgical exploration is warranted. The intraoperative criteria used to define the presence of a fistula vary. The presence of an obvious communication with accumulating fluid in either window is demanded by some. Others accept as proof of a leak repeated pooling of fluid in the round or oval window while observing under high magnification. Last, microfissures with nonvisualizable leakage of fluid have been implicated in cases where no fistula is seen. Intraoperative identification of a fistula, regardless of criteria used, is reported in about 50% of individuals explored. At the time of surgery, the oval and round windows are patched with tissue, such as perichondrium, fat, or temporalis fascia.

The outcome of surgical repair varies. Reduction in vestibular-related complaints has been reported in 49% to 100% of treated patients (Glasscock et al, 1987; House et al, 1991). Hearing is less often improved, with reports of 24% to 49% improvement in the above studies. Glasscock, in the 1987 study reported above, compared the rate of improvement in vestibular symptoms in patients with positive fistula explorations and repairs to those with negative explorations. In the latter group the incidence of improvement was less.

Trauma

Vertigo and ataxia are common sequelae of head trauma. Vestibular-related complaints may arise from cervical trauma or central nervous system trauma, as well as peripheral vestibular damage. For the sake of this discussion, only the latter will be considered. There are numerous mechanisms by which damage can be inflicted, that is, blunt concussive trauma, penetrating trauma, explosive blast, and barotrauma. It is also important to recognize that similar lesions can occur following different mechanisms of injury. Hence, labyrinthine concussion can follow blunt trauma as well as barotrauma.

Nonpenetrating trauma

Labyrinthine concussion. Labyrinthine concussion refers to those injuries of the labyrinth that do not result in violation of the otic capsule or the intralabyrinthine limiting membranes. Labyrinthine concussion is believed to be a sequela of a number of traumatic events, including blunt head trauma and inner ear barotrauma. The symptom complex ascribed to this entity includes both auditory and vestibular complaints.

Vestibular-related symptoms, although variable and not well characterized, are generally felt to include mild vertigo, imbalance, visual confusion, and vegetative symptoms, that is, nausea and vomiting. These are typically short lived and gradually subside over days to weeks. Examination of the patient with active symptoms may demonstrate nystagmus directed toward the side of the lesion acutely (that is, hours) followed by more typical contralateral beating nystagmus. Past pointing and falling in the direction of the slow phase of nystagmus have been reported (Snow, 1988). Vestibular testing may demonstrate a reduction in the caloric response and abnormality of vestibuloocular reflex gain and phase following rotation testing.

Hearing loss and tinnitus are often reported. When present, the most common pattern of hearing loss is similar to that of a noise-induced hearing loss, with a loss most apparent at 4 kHz (Schuknecht, 1951). Alternatively, it can be profound. Total hearing loss in the involved ear has been reported.

Both the vestibular and auditory complaints are usually transient, with spontaneous resolution occurring over a matter of days to weeks. However, symptoms have been reported to persist and worsen over time (Lindsay and Zajtchuk, 1970). Vestibular complaints may persist for years, often consisting of bouts of movement-associated imbalance or even episodes of severe vertigo with nausea and vomiting. In the latter, one explanation offered has been the development of secondary hydrops (see discussion of delayed endolymphatic hydrops). Benign paroxysmal positional vertigo is also noted to occur following trauma (see discussion of benign paroxysmal positional vertigo).

Pathologic findings in cases of labyrinthine concussion include intralabyrinthine hemorrhage and exudation of fluid into the endolymph and perilymph. The presence of these substances and the inflammatory reaction they engender are felt to produce the symptoms seen in concussive trauma. Resolution of the above serous labyrinthitis is clinically manifested by resolution of symptoms. Other studies have demonstrated that the pathologic changes can be permanent and varying in severity. In these instances the acute lesion is felt to initiate a process of fibrosis and ossification that gradually destroys the membranous labyrinth. This is reminiscent of the pathologic findings seen in ischemic injury to the ear and may be responsible for the progressive hearing loss and persistent vestibular symptoms seen occasionally after trauma (Lindsay and Zajtchuk, 1970).

The reader is reminded that in cases of blunt trauma, as in the patient struck with a bat, there is a dissociated rotation of the brain within the skull (Makishima et al, 1976). This rotation is primarily about the axis of the brain stem and has been noted to produce damage to the cranial nerves, these structures being fixed between the rotating brain and their exit foramina in the skull. Such trauma has been shown to produce hemorrhage, contusion, and partial laceration or total transection of the eighth nerve and may explain transient or permanent deficits seen in some trauma patients. Clearly such a mechanism would not be operative in cases of trauma caused by blast, pressure, or penetrating trauma to the ear.

Blast trauma. Explosive blasts can produce pressure waves greater than 200 dB sound pressure level (SPL) (Roberto et al, 1989). The ear, which exists to capture and amplify sound energy, is frequently injured. Blast trauma includes such occurrences as the open-handed slap to the ear as well as actual explosions. Perforation of the tympanic membrane or ossicular disruption occurs, however, inner ear damage is more likely (Snow, 1988). Interestingly, inner ear damage is most severe when the conductive mechanism is not damaged. Clearly the destructive energy is best transmitted into the labyrinth by an intact conducting mechanism (Molvaer et al, 1978). Auditory loss is most common in the high frequencies and frequently recovers spontaneously (Chait et al, 1989). However, immediate, permanent, profound losses have occurred. Peripheral vestibular damage has been infrequently reported (Philips et al, 1989). Pathologic findings in animals subjected to explosive blast while in enclosed spaces have revealed extensive destruction of the cochlea, with shearing of the cochlea/hair cells off the basilar membrane (Roberto et al, 1989).

Penetrating trauma (violation of otic capsule/temporal bone)

In every instance that the inner ear or eighth nerve is disrupted there is associated loss of function. This occurs for a number of reasons. Blunt trauma that fractures the temporal bone can lacerate nerves or produce fistulas. Fracture of the otic capsule and injury to the eighth nerve and facial nerve are more commonly seen in transverse than in longitudinal fractures (Snow, 1988) (see discussion of temporal bone fracture). Penetrating trauma can result from a variety of foreign bodies; such as a twig, hair pin, or bullet. Injuries depend directly on the path taken through the temporal bone. In the case of an introduced hair pin, the middle ear sustains the majority of damage. However, the inner ear may be damaged by subluxation of the stapes into the vestibule.

Acute vertigo and sensorineural hearing loss herald inner ear damage. Nystagmus beats in the direction of the healthy ear, and the patient will fall and demonstrate past pointing in the direction of the slow phase of nystagmus. Vegetative symptoms are typically present. Vertigo will gradually subside over the ensuing days to weeks. Hearing loss, when present, can vary from mild and transient to profound and permanent.

Barotrauma

Trauma to the inner ear as a consequence of atmospheric pressure alteration is a relatively uncommon entity. However, it can be problematic for individuals working underwater, such as divers, submariners, and bridge builders, as well as those working at high altitudes, such as pilots (Kennedy, 1974; Lundgren, 1965). Inner ear dysfunction can be produced by rapidly changing air pressure, as in atmospheric barotrauma; by elevated or asymmetric middle ear pressure, as in alternobaric trauma; or by bubble formation within the labyrinth or its blood supply as in the case of inner ear decompression sickness and isobaric gas counterdiffusion sickness. It should be recognized that during flying and especially during diving in deep water, visual and proprioceptive cues are less effective. These environments place an inordinate premium on vestibular input. Therefore acute vestibular dysfunction leading to disequilibrium, disorientation, nausea, and vomiting can be devastating in such a setting.

Alternobaric trauma. Alternobaric trauma is described as a transient vestibular or auditory dysfunction believed to occur as a result of elevated and probably asymmetric middle ear pressure. This notion is derived from its description by patients, as well as being documented in experiments. As many as 26% of divers and 10% to 17% of pilots have admitted to experiencing alternobaric-like vertigo (Lundgren, 1965; Wicks, 1989).

Alternobaric vertigo is most prominent in divers while in ascent to the surface and in fliers during ascent of the aircraft. Both are conditions of decreasing ambient pressure. Vertigo is relieved by equilibration of middle ear/ambient pressure differences, as well as repressurization; for example, by descending several meters in the case of divers (Molvaer and Albrekson, 1988).

Susceptibility to alternobaric vertigo is thought to be a consequence of individual variations in the pressure required within the middle ear to force open the eustachian tube. Although passive air movement from the middle ear to the nasopharynx occurs exclusively in response to decreasing atmospheric pressure, it has been observed that variations exist in the required pressure for this to occur. Conditions that alter eustachian tube patency, such as mucosal turgescence, are known to contribute. Such individuals are more liable to experience alternobaric vertigo. Conditions such as an upper respiratory infection would exacerbate the situation.

Several studies of humans have demonstrated that vertigo/nystagmus could be elicited when middle ear pressure was increased relative to atmospheric pressure (Inglestedt et al, 1974; Tjernstrom, 1974a, 1974b; This was done either by lowering ambient pressure in a chamber or by increasing pressure in the middle ear with a transtympanic catheter. The latter was done to rule out the possible influence of tympanic membrane/ossicular displacement as a cause of vestibular stimulation. It was noted that positive middle ear pressure stimulated the

vestibular system and caused nystagmus toward the ear with the higher middle ear pressure. Further, nystagmus and vertigo were observed only in those individuals in whom elevated middle ear pressure was required to passively open the eustachian tube.

It has been suggested that an asymmetric middle ear pressure between ears is necessary. Although it was noted that a pressure asymmetry was always present when alternobaric vertigo was elicited by the experimental paradigm, it is not clear that it is a necessary part of the pathogenesis.

The pathogenesis for labyrinthine dysfunction caused by alternobaric pressure is unknown. It has been suggested that pressure differences between the middle ear and the intracranial/intralabyrinthine space may play a role (Tjernstrom, 1974b).

The effects of alternobaric trauma on the inner ear are not felt to be permanent (Lundgren, 1965). In most cases, vertigo, hearing loss, and tinnitus are described as resolving in 10 to 15 minutes. The occurrence of alternobaric trauma can be minimized by frequent equilibration of middle ear pressure during diving, the use of topical decongestants before the dive, and the avoidance of diving during periods of increased upper respiratory obstruction; that is, upper respiratory infection and sinusitis (Farmer, 1977; Molvaer and Albrekson, 1988).

Atmospheric inner ear barotrauma. Extremes of pressure or abrupt changes in middle ear pressure are capable of damaging middle ear or inner ear structures. The latter is reflected by auditory and vestibular dysfunction. As opposed to alternobaric trauma, barotraumatic injury is frequently long lasting or permanent. Hearing loss and tinnitus are universal complaints, whereas vertigo tends to be less common (30%) and is rarely the sole complaint (Farmer and Gillespie, 1988).

Barotrauma to the inner ear is suffered most frequently by divers, both scuba and breath holding. However, it has also been described during forceful sneezing with a closed mouth and nostrils (Schuknecht and Witt, 1985).

A number of mechanisms have been proposed. All are based on a sudden pressure differential transmitted into the inner ear. Perhaps the most commonly cited theories are the "explosive and implosive" ones (Goodhill, 1971, 1972).

The implosive theory states that, as a consequence of increased middle ear pressure, the round window or oval window is displaced into the labyrinth and can rupture into the inner ear. The explosive theory states that the increased intracranial pressure is transferred into the inner ear, resulting in an outward rupture of the round or oval window into the middle ear.

Although the above have been cited as plausible explanations for sudden hearing loss, the underlying pathophysiologic mechanisms have been suggested as an explanation for inner ear dysfunction associated with atmospheric barotrauma.

We suggest a slightly different but very plausible explanation as the mechanism for atmospheric inner ear barotrauma (IEBT). In our experience, atmospheric IEBT has almost exclusively been a consequence of diving. Further, it has usually been experienced by divers at depths varying from 10 to 30 feet. Typically, in an effort to equilibrate pressure between the middle ear and the increasing ambient pressure in the external ear, a Valsalva or similar maneuver is performed. It is following such a maneuver that signs and symptoms of acute inner ear dysfunction become apparent.

We believe that the following events occur and are likely responsible for the clinical entity. Successful opening of the middle ear as a result of the Valsalva maneuver results in a sudden transient increase in middle ear pressure while rapidly displacing the tympanic membrane laterally. Given that the eustachian tube is now open, a rapid outflow of high-pressure air from the middle ear follows. The combined effect of an increased ambient pressure (surrounding water), a rapidly diminished volume of the middle ear, and most importantly the inherent fibroelastic nature of the tympanic membrane results in a dramatic slingshot like displacement of the eardrum and attached ossicles. The resultant rapid medial displacement of the stapes foot plate into the vestibule is responsible for the varying degrees of inner ear damage.

Support for such a pathophysiologic mechanism is gained by the observation that the associated hearing loss is typically high frequency. Such hearing losses are in keeping with transmission of traumatizing energy via the ossicular chain; that is, noise or a blast. Additionally, hearing loss following Valsalva maneuver at atmospheric pressure does not produce a stereotyped hearing loss (see discussion of perilymphatic fistula).

Atmopsheric IEBT results in damage of varying degree to the inner ear. Labyrinthine concussion, intralabyrinthine membrane tears, or damage to receptor structures, as seen in blast trauma, are reported (Molvaer et al, 1978; Parell and Becker, 1985). At its extreme there may be associated oval and, perhaps more likely, round window fistulas (Farmer and Gillespie, 1988).

The patient history should include a complete description of the inciting event, including details of the particular dive. Divers usually report that symptoms occur during descent. There is usually a report of difficulty clearing the negative pressure in one ear (Freeman and Edmonds, 1972). It must be recognized that barotrauma is not exclusive to scuba divers. It can occur in a few feet of water and consequently affect breath-holding divers as well.

Symptoms typically consist of dizziness, tinnitus, and sensorineural hearing loss (Rubenstein and Summit, 1971). A history of unsteadiness and physical findings of nystagmus and ataxia should alert the physician of complicating inner ear barotrauma. Joint pain and CNS findings, typically associated with systemic decompression sickness, are notably absent. On physical examination, findings consistent with middle ear barotrauma may be seen that is, hemotympanum or tympanic membrane perforation (Farmer and Gillespie, 1988; Reissman et al, 1990; Shupak et al, 1991).

The associated hearing loss varies but typically demonstrates an isolated deficit in the 4 to 8 kHz range. Total deafness has been noted (Freeman and Edmonds, 1972).

Suggested treatment of IEBT is similar to that recommended for perilymph fistula. Bed rest, head elevation, and close monitoring of hearing- and balance-related symptoms are suggested. Symptoms usually spontaneously resolve in a matter of hours to days using conservative management (Parell and Becker, 1985). Tympanotomy and middle ear exploration are reserved for those cases with progressive hearing loss or failure of resolution of vestibular symptoms in 3 to 5 days. Some surgeons advocate more immediate exploration (Farmer and Gillespie, 1988; Parell and Becker, 1985). Recompression is to be avoided in cases of barotrauma. It is therefore important to differentiate atmospheric IEBT from inner ear decompression sickness in view of the necessary use of recompression in the treatment of the latter.

Individuals suspected of having sustained IEBT and realized complete recovery should be advised to refrain from diving for at least 3 months. If permanent damage to either the vestibular or auditory system results, the patient should be cautioned against diving. In all instances, the patient should be counseled to perform gentle Valva maneuvers at frequent intervals during descent and to abort the dive if middle ear pressure equilibration cannot be achieved easily (Farmer and Gillespie, 1988).

Inner ear decompression sickness and isobaric gas counterdiffusion sickness. Inner ear decompression sickness (IEDS) and isobaric gas counterdiffusion sickness (IGCS) have become common as a consequence of the increased use of mixed gas, oxyhelium, for deep-water diving. Although damage produced by these disorders is not restricted to the inner ear, they now represent the most common decompression injuries experienced by individuals diving to depths greater than 100 m (Shupak et al, 1991). Otic symptoms are often the sole feature of decompression sickness, particularly in cases where mixed gases - that is, helium and oxygen - are used (Farmer, 1977). When the inner ear is affected, vestibular dysfunction and auditory dysfunction are often permanent, particularly if treatment is delayed. In contradistinction to atmospheric inner ear barotrauma, vertigo is a prominent complaint and often the sole complaint of individuals experiencing IEDS (50%).

IEDS and IGCS are both caused by the formation of gas bubbles in the body. In IEDS these bubbles occur because of a rapid reduction in atmospheric pressure, such as experienced by the diver who ascends too rapidly. In IGCS, the bubbles are a consequence of too rapid an exchange of inspired gas mixtures under isobaric conditions (D'Aoust et al, 1977, 1979; Farmer et al, 1976, 1988; Graves et al, 1973; Lambertsen and Idicula, 1975). The analogy to bubbles that form in carbonated beverages when the bottle is opened is appropriate. The symptoms and damage to various body parts caused by these occluding bubbles are termed *decompression sickness* or *the bends.*

Air bubbles within the intralabyrinthine fluids as well as blood vessels cause the damage to the inner ear (Farmer et al, 1976). It has also been suggested from animal studies that expanding bubbles can generate microfissures in the otic capsule (Fraser et al, 1983).

Pathologic findings in animals and humans reflect a process of intralabyrinthine fibrosis and new bone formation. These findings are very similar to those following ischemic injury (Landolt et al, 1980; Money et al, 1985).

Symptoms are identical to those of atmospheric barotrauma, consisting of tinnitus, sensorineural hearing loss, and vertigo in any combination. The history and associated findings support the diagnosis of this disorder. Symptoms usually begin during ascent. Rapid surfacing after deep, prolonged underwater dives may be reported. Disregard for specified decompression times is the cause. Treatment is immediate recompression. Delay in treatment is associated with permanent damage to the vestibular and auditory system.