Chapter 38
Tinnitus and Ménière's Disease

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Keypoints

1. Ménière's disease is characterized by a triad of symptoms: fluctuating hearing loss, attacks of vertigo, and tinnitus. Some authors have included aural fullness.
2. Patients often seek treatment for the severe vertigo attacks, but may have had other otologic symptoms for some time prior to the onset of vertigo.
3. Tinnitus that occurs in Ménière's disease is best characterized as a low pitched, narrow band of noise, resembling a "roaring sound."
4. The tinnitus may change with the fluctuations in hearing loss, and tinnitus increases as hearing loss worsens with the progression of the disease.
5. During the active phase of Ménière's disease, the vertigo can be debilitating and dominating the symptoms.
6. As the disease stabilizes, the tinnitus can become a serious and a severe problem.
7. It is believed that endolymphatic hydrops (imbalance in volume of the fluid systems of the inner ear) is the cause of the symptoms of Ménière's disease, but there is still uncertainty regarding many aspects of the pathology of the disease.

Keywords Tinnitus • Cochlear implants • Promontory stimulation • Treatment

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Abbreviations

AAO-HNS	American Academy of Otolaryngology – Head and Neck Surgery
DHI	Dizziness Handicap Inventory
HHIA	Hearing Handicap Inventory for Adults
PA	Pure tone average
THI	Tinnitus Handicap Inventory

Introduction

Ménière's disease is defined by the presence of three symptoms (or four): intermittent vertigo, fluctuating hearing loss, and tinnitus with aural fullness occurring on one side. Some authors have added aural fullness as a fourth symptom. It was first described by Prosper Ménière in his original publication in 1861. Its diagnosis is largely based on the clinical history and hearing tests. Individuals with Ménière's disease experience incapacitating attacks of vertigo, associated with nausea and vomiting lasting for hours [1]. The sudden attacks of vertigo last anywhere from 30 min to several hours, with unilateral hearing loss occurring together with tinnitus; often aural fullness is present as well. Audiological findings include fluctuating low frequency and progressive sensorineural hearing loss with tinnitus. The course of Ménière's disease is unpredictable and highly variable among individuals and can be accompanied with periods of remission. Disequilibrium may persist for 24–72 h after the attack before resolving completely.

Tinnitus may be the first symptom of Ménière's disease and may precede the remaining symptoms by months or years. Fluctuating cochlear signs, such as tinnitus, hearing impairment, and/or fullness in the ear

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Table 38.1 Diagnosis of Ménière's disease

Certain Ménière's disease
Definite Ménière's disease, plus histopathologic confirmation
Definite Ménière's disease
Two or more definitive spontaneous episodes of vertigo 20 min or longer
Audiometrically documented hearing loss on at least one occasion
Tinnitus or aural fullness in the treated ear
Other causes excluded
Probable Ménière's disease
One definitive episode of vertigo
Audiometrically documented hearing loss on at least one occasion
Tinnitus or aural fullness in the treated ear
Other causes excluded
Possible Ménière's disease
Episodic vertigo of the Ménière type without documented hearing loss or
Sensorineural hearing loss, fluctuating or fixed, with disequilibrium but
without definitive episodes
Other causes excluded

Adapted from Committee on Hearing and Equilibrium guidelines for the diagnosis and evaluation of therapy in Ménière's disease. American Academy of Otolaryngology – Head and Neck Foundation, Inc. Otolaryngol Head Neck Surg. 1995; 113(3):181

were present prior to onset of the first vertigo attack in more than 50% of patients in a study by [2].

Criteria for the diagnosis of Ménière's disease can be divided into four categories as possible, probable, definite, and certain (Table 38.1). Furthermore, scales of dizziness [i.e., Dizziness Handicap Inventory (DHI)], hearing loss [i.e., Hearing Handicap Inventory for Adults (HHIA)], and tinnitus [i.e., Tinnitus Handicap Inventory (THI)] have been developed to quantify the symptoms associated with Ménière's disease.

The Committee on Hearing and Equilibrium of the American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNS) published its most updated guidelines for defining, reporting, and interpreting results of the treatment of Ménière's disease in 1995 [3]. (For detailed discussion see Chap. 60)

Epidemiology

Reports on incidence and prevalence vary among investigators [4]. The results of studies of the prevalence of Ménière's disease in the US vary from 218 per 100,000 people [5], thus approximately 0.2% of the population. The latter study was performed by the Mayo Clinic [6]. The reported prevalence of Ménière's disease throughout the world varies, in average it is about 1% CHECK). One study finds it to be more common in industrialized countries and in adult white populations [7]. Studies have reported the prevalence of Ménière's disease in England as 56/100,000 [8] and 157/100,000 [9]. In Japan, the reported prevalence of Ménière's disease is low, one study by Shojaku and Watanabe (1997) [10] found a prevalence of between 21.4 and 36.6/100,000. In Finland, the prevalence has been reported to be 43/100,000 and the incidence 4.3/100,000 [11]. In Sweden, Stahle reported an incidence of 45/100, using more stringent criteria than some other studies [12]. In Italy, the reported prevalence is low, 45/100,000, with an incidence of only 8/100,000 [13].

The prevalence increases linearly with age up to 60 years. There is a slight female preponderance, and the typical age of onset is 30–60 years. Bilateral Ménière's disease incidence ranges from 10 to 70%, increasing the frequency with time [14]. Genetic predisposition has been reported in families with Ménière's disease [15, 16]. Factors such as diet, weather changes, as well as emotional and physical stress can also precipitate vertigo attacks and make any symptom complex and worse. Some of the variance in the incidence reported by different investigators may have been caused by differences in the definition of Ménière's disease.

The symptoms of Ménière's disease are signs of an imbalance in the volumes of the fluids in the inner ear, known as endolymphatic hydrops [17]. Cochlear hydrops causing fluctuating low-frequency hearing loss, tinnitus, and aural fullness without associated vertigo may precede Ménière's disease with eventual development of the full syndrome occurring in 37–42% of patients [18]. Lermoyez's syndrome is a variant of Ménière's disease, where hearing loss and tinnitus precede an attack of vertigo by days or months, with improvement of hearing after vertigo episodes. Patients with severe and long-term Ménière's disease are at risk of developing Tumarkin otolithic crisis or drop attacks of falling because of loss of lower-extremity muscle tone without loss of consciousness. The incidence is reported as 7% of patients with Ménière's disease to as high as 72% in one report [19]. The cause is thought to be a sudden stimulation

of the vestibular end organs by shift of the utricular macula or rupture of inner ear membranes, but the exact cause is unknown [20].

The frequency of vertigo attacks varies widely with a mean of 6–11 episodes per year [21]. Spells tend to change in severity over a period of time, becoming milder but still unpredictable. The decrease in hearing and tinnitus can occur before or during the vertigo attack. Typically, the disease eventually "burns out" with the decline and cessation of vertigo, but there is progressive deterioration of hearing. Hearing fluctuates in the early course of disease but eventually becomes progressively worse, stabilizing at about 50 dB pure tone average (PA) and 50% word discrimination score [18]. The hearing loss in Ménière's disease begins at low frequencies, thus, different from many other causes of hearing loss such as noise induced hearing loss, hearing loss caused by ototoxic substances, and presbycusis that mainly affect hearing at high frequencies. Eventually, hearing loss in individuals with Ménière's disease involves all frequencies. Tinnitus that occurs in Ménière's disease is often described as a harsh roaring machine-like sound that is more pronounced during vertigo attacks. Hyperacusis (decrease tolerance to sounds, see Chap. 3) and distortion of sound in the affected ear can also be present. Tinnitus and aural fullness prevail during life in the majority of individuals with Ménière's disease [22].

Pathophysiology

Despite the long history of Ménière's disease, the etiology and pathophysiology of this condition are still unknown. Most hypotheses of the pathogenesis of Ménière's disease include anatomical abnormalities of the endolymphatic fluid system, but other hypotheses involve viral infection, autoimmune disease, allergy, and activation of neural plasticity. That endolymphatic hydrops is the cause of the symptoms of Ménière's disease has been supported by histopathological findings, although not all patients with the histopathology have the typical symptoms [23, 24]. The hydrops are believed to be caused by mechanical obstruction to endolymphatic flow or by intrinsic malfunction of the endolymphatic system, resulting in an overabundance of endolymphatic volume and/or pressure [25]. Dysfunction of the spiral ligament fibrocytes, which interferes with the recycling of K^+ ions and results in osomotic imbalance, can cause expansion of the endolymphatic compartment [26].

Schuknecht developed a theory of gradual distention of the endolymphatic system that leads to a rupture of membranous labyrinth and sudden release of a large volume of endolymph into perilymphatic space [20]. It is suggested that sensory and neural structures are injured from exposure to the potassium-rich endolymph, resulting in vertigo and hearing loss. As the rupture heals and hemostasis is restored in the inner ear compartments, the symptoms subside. Dohlman (1980) [27] suggested that increase of potassium occurs in the perilymph during a Ménière's attack and that potassium-rich fluid surrounding the vestibular nerve is the cause of the experienced vertigo. Zenner (1987) [28] found that perilymphatic potassium intoxication leads to a longitudinal contraction of the outer hair cells. This results in their decoupling from the tectorial membrane. Dulon et al. (1987) [29] demonstrated that small changes in the osmolarity of the surrounding in vitro medium induce fast contractions (hypo-osmotic solution) or elongations (hyper-osmotic solution) in isolated outer hair cells. However, the hypotheses that assume that Reissner's membrane ruptures before an attack occurs have been questioned.

Another theory proposed by Gibson and Arenberg (1997) [30] is a disturbance in longitudinal flow of endolymph from the cochlear duct to the endolymphatic sac due to a narrow vestibular aqueduct, resulting in hydrops.

It has been suggested that longitudinal flow was involved in maintaining endolymph homeostasis. However, measurements of the dispersal of markers in the endolymph [17] have failed to support these hypotheses. These measurements were interpreted to suggest that the normal state of the endolymph is maintained without a significant involvement of volume flow at all [17]. The situation is different in abnormal states, such as is assumed to be present in Ménière's disease where the volume of inner ear fluid is abnormal. In such situations, the longitudinal volume flow of the endolymph may contribute to homeostasis.

The role of the endolymphatic sac is complex and poorly understood. It seems to act as a "bidirectional overflow" system that responds to the endolymph volume disturbance [17].

In other hypotheses, the sac is postulated to actively regulate the flow by maintaining an osmotic gradient and secreting glycoproteins that attract movement of the endolymph toward the sac. The sac produces saccin, a hormone thought to increase the volume of endolymph, which may promote faster flow [30].

It has been suggested that the endolymphatic sac is primarily responsible for the immuno-defense of the inner ear [31]. It is hypothesized that a viral infection leads to an inflammatory immune and microvascular-mediated injury. Circulating immune complexes and serum auto-antibodies to inner ear antigens are greater in Ménière's disease patients than in controls [32–34]. This suggests that circulating immune complexes may be involved in the pathogenesis of Ménière's disease, either as a direct cause of damage or as a by-product of an underlying autoimmune abnormality [33]. This hypothesis is supported by the clinical experience of beneficial effect of treatment with corticosteroids [35].

As early as in 1923, Duke (1923) [36] proposed an allergic theory for Ménière's disease. However, it was not until 1970s that studies showed an improvement in the symptoms of this disease after desensitization to inhalant allergens and an elimination diet for allergies to food.

> Pulec (1973) [37], in discussing Ménière's disease, reported allergies were related to the sensorineural hearing loss and symptoms of Ménière's disease among 36% patients. Fourteen percent of his Ménière's disease patients responded to allergy treatment. In a case-control study, Derebery and Valenzuela (1992) [38] found an inhalant allergy in 41.6% and a food allergy in 40.3% of patients with Ménière's disease answering a self-reported questionnaire, in comparison with rates of 27.6 and 17.4% in their control population.

Furthermore, a significant percentage of patients with Ménière's disease and allergy showed improvement in both allergy and Ménière's symptoms when treated with desensitization and diet control [39]. Hence, symptoms of food allergy should be questioned for patients with endolymphatic hydrops and fluctuant hearing loss, as suggested by Shambaugh and Wiet (1980) [40].

Inspired by the benefits from treatment with air-pressure (pressure chamber) [41–43] by applying air-puffs to the inner ear (using the Meniett device), it has been suggested that activation of neural plasticity may be involved in creating the symptoms of Ménière's disease [44].

Tinnitus Associated with Ménière's Disease

Tinnitus in Ménière's disease is best characterized as a low pitched, narrow band of noise, usually described as a "roaring sound", corresponding to the low-frequency sensorineural hearing loss [45–47]. In the early stages of the disease, tinnitus may be intermittent. As the disease progresses, tinnitus becomes permanent, but its intensity fluctuates. Hearing loss and tinnitus normally increase over time. After a long time, the end state of Ménière's disease "burnt-out" where the effects of vertigo attacks have ceased, tinnitus may become the most disturbing complaint.

As Stahle (1988) [48] described in his results of an epidemiologic study in Sweden, the tinnitus quality fluctuated in its intensity and paralleled the control of vertigo symptom and ear blockage. In a separate group of patients with control of their chief vertigo complaint, the ear blockage persisted, as did the tinnitus. Herraiz et al. (2006) [49] found a statistical association between tinnitus intensity and worse hearing loss or hyperacusis in 102 patients with Ménière's disease, uninfluenced by the number of vertigo spells. In the initial phases of Ménière's disease, tonal-tinnitus is usually not present, as opposed to in the later stages of the disease where tonal-tinnitus is described by a number of patients [50].

Pathophysiology of Tinnitus in Ménière's Disease

Tinnitus in Ménière's disease may be caused by similar mechanisms as other forms of tinnitus that are related to injuries of the cochlea (see Chap. 10). Hearing loss may cause tinnitus through the effect of deprivation of input to the auditory system that activates neural plasticity (see Chap. 12).

Management and Treatments

There is no known cure for Ménière's disease, and treatments are aimed at reduction of its symptoms. Vertigo is often the most debilitating symptom of

Ménière's disease, and most treatments focus on relieving this symptom. The tinnitus of Ménière's disease may well remit with improvement in low-frequency hearing as a result of medical or surgical treatment. For a detailed discussion of treatment of Ménière's disease, see Chap. 60.

Treatments specifically directed toward tinnitus in Ménière's disease are similar to treatments of other forms of tinnitus described in the chapters in Part VI of this book.

Neural Plasticity

The reason that overpressure can relieve symptoms of Ménière's disease is not known, but it has been hypothesized that neural plasticity is involved in at least one or more of the symptoms of Ménière's disease [51]. These symptoms are assumed to be caused by an imbalance of the volumes of the fluid in the inner ear [17], the causes of which are unknown. The finding that applying air-puffs to the inner ear can ameliorate the symptoms, thus stimulating the vestibular sensory cells, indicates that functional abnormalities may be involved in causing the symptoms of Ménière's disease and an activation of neural plasticity may be involved.

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