

Meniere's disease – A century old mystery

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Abstract

Ménière's disease, first described by the French physician Prosper Ménière, is one of the most common causes of dizziness. It can have a debilitating effect on patients, significantly impairing their ability to carry out normal daily activities. Despite been described over a century and a half ago, the disease is still poorly understood and there is no cure. However, it can be controlled with nonsurgical or surgical treatments. This review of Ménière's disease is based on a literature survey and focuses on the pathophysiology, clinical features, and management options available.

Key words- Vertigo, Tinnitus, Hearing impairment, Meniere's disease

Introduction

Meniere's disease is a debilitating chronic inner ear disorder that is characterised by clinical triad of episodes of vertigo, tinnitus, and hearing loss. The disease was first described by French physician Prosper Ménière in 1861.¹ Meniere's disease can have a significant impact on both physical and psychological well-being. Vertigo can make it difficult to drive, work, and perform other daily activities. Tinnitus and hearing loss can also be debilitating, particularly in their impact on social interactions. The fear of another vertigo attack can lead to anxiety and social isolation.² Clinical symptoms and audiometric tests are the basis for diagnosing of Ménière's disease, but differential diagnosis can be challenging, as most of the findings are subjective and nonspecific. Therefore misdiagnosis is likely.³

Some people with Ménière's disease experience long periods of remission between episodes. The classic full-blown picture of Ménière's disease may only be seen in advance disease.³ In the early stages, various forms of the disease may be present, with symptoms arising from either the vestibular or cochlear system. The disease also tends to relapse episodically.³

The prevalence of Ménière's disease in the Western world is approximately 13 per 100,000 people⁴. Data on the prevalence of Ménière's disease in Sri Lanka is not readily available. Women are more likely to develop Ménière's disease than men, and the disease is most common in people over the age of 50.⁴

Pathophysiology

Despite decades of research, the pathophysiology of Ménière's disease remains elusive. It has noted that those who has disease has a distension of membranous

labyrinth by the endolymph, called endolymphatic hydrops. Numerous causative factors have been proposed for endolymphatic hydrops, including excessive endolymph production, endolymphatic sac dysfunction, ion imbalance, genetic causes, viral infections, autoimmune diseases, blood flow disorders, allergies, and dietary habits.⁵

Clinical features

Diagnosis of Meniere's disease is primarily clinical, based on the patient's medical history and symptoms. Classical Meniere's disease is diagnosed with a triad of symptoms: vertigo, hearing loss, and tinnitus. However, there can be atypical symptoms depending on the stage of the disease. Vertigo typically lasts for 20 minutes to 2 hours. They can be unpredictable or follow a feeling of ear fullness and tinnitus and occur frequently. Tinnitus and ear fullness are often the first symptoms to appear during an episode and can precede or accompany vertigo, lasting for about 20 minutes after the episode of vertigo. Vertigo can be associated with vomiting, and symptoms may improve after vomiting. The symptoms can recur again within the days to few weeks.⁶ Clinical examination findings are usually normal except for hearing impairment. At the onset of disease hearing too may be normal, but hearing threshold goes down progressively. Hearing loss may be very mild at early stages, and patients may not complain about it at all, especially if tinnitus and vertigo are more pronounced. Initially the low frequencies are affected, but when the disease progressed audiogram shows low threshold for all the frequencies and finally a patient can ends up with severe to profound hearing loss. However, a comprehensive neurological, and cardiovascular examination may be needed for those with atypical symptoms of Meniere's disease to exclude other possible causes of dizziness and imbalance.⁷

Investigations

There is no definitive test to confirm Meniere's disease, as endolymphatic hydrops can only be demonstrated in postmortem specimens. However, audiograms, glycerol

dehydration tests, and electrocochleography can be used as subjective tests. Audiograms show a sensorineural hearing impairment pattern. In the early stage of the disease, the lower frequencies are usually affected.

Table 1. “Diagnostic criteria for Meniere’s disease” (Van RompaeyWatkinson and Clarke, 2018).

“Definite Meniere’s Disease”	“≥ Two definitive spontaneous episodes of vertigo lasting 20 min to 12 h + Audiometrically documented low- to medium-frequency sensorineural hearing loss in the affected ear on at least one occasion before, during, or after one of the episodes of vertigo + Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear”
“Probable Meniere’s Disease”	“≥ Two episodes of vertigo or dizziness, each lasting 20 min to 24 h + Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the reported ear” (8)

In the glycerol dehydration test, a test dose of glycerol is given orally. Serial audiograms are performed prior to the dose of glycerol, and at 90 minutes and 3 hours after. A positive finding is considered when there is a greater than 10-decibel improvement in more than two frequencies after glycerol. Glycerol dehydrates the patient and reduces the endolymphatic sac pressure hence the temporary improvement of hearing.^{9,10} Electrocochleography (ECoG) is considered a useful test for diagnosing Ménière’s disease. In an ECoG test, a needle electrode is placed either through the tympanic membrane on the promontory, on the tympanic membrane, or simply in the ear canal. The components measured are: Cochlear microphonics, Summating potentials (SP) and Action potentials (AP). The cochlear microphonics and the summating potentials reflect the cochlear bioelectric activity, while the action potentials reflect the activity of distal afferent fibers of the eighth nerve. The amplitude of the SP and the AP is determined from a common baseline. The ratio SP/AP is calculated and reported as a percentage. The cut-off criterion for the normal SP/AP amplitude ratio is 50% (0.5) for the ear canal electrode type, 40% (0.4) for the tympanic membrane electrode, and 30% (0.3) for the trans-tympanic electrode type. An increased level of SP/AP amplitude ratio points to the diagnosis of Ménière’s disease. Other useful test includes brain stem evoked response (BSER) and vestibular evoked myogenic potential (VEMP) mainly to differentiate other form of vestibular disorders.^{11,12,13}

Blood tests for sugar, electrolytes, and thyroxine can be used to exclude diabetic and other endocrine disorders. Antinuclear antibody testing can be used to exclude autoimmune diseases. An electrocardiogram can be used to exclude cardiac arrhythmias.

Treatment

While there is no definitive cure for Meniere’s disease, the treatment options available are mainly to manage the symptoms and improve quality of life. The main modality of treatment options widely practised includes dietary advice – particularly a low-salt diet, medication to treat and prevent attacks, treatment for tinnitus and hearing loss, vestibular rehabilitation therapy to cope with balance problems, treatment for stress, anxiety and depression and surgical treatment.

One of the most important treatments for Meniere’s disease is a low-salt diet. Salt can cause the body to retain fluid, which can worsen the symptoms of the disease. Aim to consume no more than 2,000 milligrams (mg) of sodium

per day. Betahistine can be recommended for the treatment of Ménière’s disease. It has been shown to improve vertigo, but only when taken regularly and as a preventive measure. Diuretics, such as hydrochlorothiazide and triamterene, have been anecdotally suggested to slow hearing loss by reducing fluid pressure in the inner ear, but there is limited evidence to support this claim³. Short-term use of oral prednisone can reduce the severity of vestibular symptoms by reducing inflammation and autoimmune reactions in the vestibular nucleus. However, due to the significant systemic risks of prednisone, it is not typically recommended. Benzodiazepines, such as diazepam, can be used cautiously to suppress vestibular symptoms during acute attacks. Tinnitus retraining therapy (TRT) is a type of counselling that can help to reduce the awareness of tinnitus. Sound therapy can be used to mask the sound of tinnitus. Cognitive behavioural therapy (CBT) can be used to help people cope with the emotional distress caused by tinnitus. There are a number of devices that can help to improve hearing. Hearing aids can amplify sound and make it easier to hear. Cochlear implants can be used to bypass the damaged inner ear and transmit sound directly to the brain.

Vestibular rehabilitation is a type of therapy that can help people with Meniere’s disease to cope with balance problems. Vestibular rehabilitation exercises can help to strengthen the balance system and improve coordination.

Ménière’s disease can be a stressful and frustrating condition. It is important for people with Meniere’s disease to seek treatment for stress, anxiety, and depression.

Surgery

Surgery is an option for people with severe Meniere’s disease that does not respond to medical treatments.

Endolymphatic sac surgery which involves draining the excess fluid from the inner ear is an option. Vestibular nerve section can relieve vertigo, but it also results in permanent hearing loss.

Non-ablative therapies: (procedures with hearing preservation) For patients whose symptoms are not well controlled by conservative therapy, intratympanic steroids can be offered for control of vertigo. Endolymphatic sac shunt surgery is another option for control of vertigo attacks. The effectiveness of this procedure has been debated. The physiologic rationale for this procedure is to drain excess endolymph, reducing the possibility of endolymphatic hydrops. This surgery is typically offered to patients whose symptoms are debilitating and who have failed conservative therapy and intratympanic steroid injections.

Ablative therapies (procedures with hearing compromised). Ablative therapies include intratympanic gentamicin, vestibular neurectomy, or labyrinthectomy. They are typically to control the vertigo but can have other consequences like permanent hearing loss.

References

1. Meniere P. Memoire sur des lesions de l'oreille interne donnant lieu a des symptomes de congestion cerebrale apoplectiforme. *Gaz Med (Paris)* 1861;16:597–601. [Article in French]
2. Cunha F, Settanni FA, Ganança FF. What is the effect of dizziness on the quality of life for patients with Meniere's disease? *Rev Laryngol Otol Rhinol (Bord)* 2005;126:155–8.
3. Anderson JP, Harris JP. Impact of Ménière's disease on quality of life. *Otol Neurotol*. 2001;22:888–94.
4. Saskia G. Bruderer, Daniel Bodmer, Nadja A. Stohler; Susan S. Jick; Christoph R. Meier Population-Based Study on the Epidemiology of Ménière's Disease; *Audiol Neurotol* (2017) 22 (2): 74-82.
5. Sung-Il Nam, MD. The Pathophysiology of Meniere's Disease, *Research in Vestibular Science* 2011;10(0): 95-98
5. Frykholm C, Larsen HC, Dahl N, Klar J, Rask-Andersen H, Friberg U. Familial Ménière's disease in five generations. *Otol Neurotol*. 2006;27:681–6.]
6. Saeed S, Penney S. Diagnosis and management of Meniere's Disease. *ENT News*. 2004;13:32–4.
7. de Sousa LC, Piza MR, da Costa SS. Diagnosis of Meniere's disease: routine and extended tests. *Otolaryngol Clin North Am*. 2002;35:547–64.]
8. Vincent WFM, Van Rompaey, Meniere's disease, (eighth ed.) J.C. Watkinson, R.W. Clarke (Eds.), Scott Brown's Otolaryngology Head & Neck Surgery, vol. 2, CRC Press, Taylor and Francis Group (2018), pp. 817-827
9. Di Girolamo S, Picciotti P, Sergi B, D'Ecclesia A, Di Nardo W. Postural control and glycerol test in Ménière's disease. *Acta Otolaryngol*. 2001;121:813–7.
10. Zhao R, Zhu W, Liu H. The control study of glycerol test in different stage of Meniere's disease patients. *Lin Chuang Er Bi Yan Hou Ke Za Zhi*. 2005;19:543–4.
11. A. Vassiliou, P. V. Vlastarakos,¹ P. Maragoudakis,² D. Candiloros,² and T. P. Nikolopoulos. Meniere's disease: Still a mystery disease with difficult differential diagnosis, *Ann Indian Acad Neurol*. 2011 Jan-Mar; 14(1): 12–18
12. Hall JW, Antonelli PJ. Assessment of peripheral and central auditory function. In: Bailey BJ, Jackler RK, Pillsbury HC 3rd, Lambert PR, editors. *Head and Neck Surgery-Otolaryngology*. 3rd ed. Lippincott, Philadelphia: Williams and Wilkins; 2001. p. 1666.
13. Chung WH, Cho DY, Choi JY, Hong SH. Clinical usefulness of extratympanic electrocochleography in the diagnosis of Ménière's disease. *Otol Neurotol*. 2004;25:144–9.
14. Vincent Wu¹, Edward A Sykes, Michael M Beyea, Matthew T W Simpson, Jason A Beyea Approach to Ménière disease management *Can Fam Physician*. 2019 Jul;65(7):463-467.