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Recent Updates of Clinical Guidelines in Diagnosis of Meniere's Disease: Review Article

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ABSTRACT

Background: Meniere's disease (MD) is a peripheral vestibular disorder presented presented with fluctuating aural symptoms (fullness, tinnitus and hearing loss). In addition to recurrent spontaneous vertigo lasting between few minutes to several hours. Based on these symptoms, there are two categories of MD: definite and probable.

Objective: The objective of this review article is to present an overview about recent methods for diagnosis of MD.

Methods: We looked for data on Meniere's disease, Tinnitus, Vertigo, and Electrocochleography in medical journals and databases like PubMed, Google Scholar, and Science Direct. However, only the most recent or extensive study was taken into account between February 2015 and January 2023. References from related works were also evaluated by the authors. There are not enough resources to translate documents into languages other than English, hence those documents have been ignored. It was generally agreed that documents such as unpublished manuscripts, oral presentations, conference abstracts, and dissertations did not qualify as legitimate scientific study.

Conclusion: MD is diagnosed clinically since it typically manifests as unilateral ear problems that might remain for decades. Attacks from MD are sporadic and episodic, with remissions in between. As there is no definitive vestibular testing for MD, these tests are typically carried out to rule out disorders with similar symptoms.

Keywords: Meniere's disease, Tinnitus, Vertigo, Electrocochleography.

INTODUCTION

Meniere's disease (MD) is an idiopathic peripheral audiovestibular disorder characterized by episodic vertigo that lasts between minutes to hours, unilateral fluctuating or permanent loss of hearing, tinnitus, and blocking of ear. In 1861, Prosper Meniere recognized that an insult within the inner ear could result in symptoms of vertigo and hearing loss [(1)].

The pathophysiology of MD is not fully understood, and over the years, several theories have been developed to explain the disease's well-known symptoms. Based on investigations on human temporal bones, it is clear that all MD subjects exhibit endolymphatic hydrops (EH), which is used to describe an accumulation of endolymph in the membranous labyrinth, frequently in cochlear duct, saccule before utricle, and semi-circular canals [(2)].

Increased endolymph production or impaired endolymph reabsorption are two mechanisms suggested to cause hydrops. Genetic, viral, traumatic, mechanical, autoimmune, allergy, hormonal, and vascular etiologies all predispose them. All of the clinical and pathological findings in MD cannot be explained by any of them [(3)]. According to theory of Schuknecht's rupture, the endolymphatic space enlarges and leading to rupture of Reissner's membrane. After being exposed to toxic and K-rich endolymph the hair cells of cochlea and the audiovestibular nerve experience episodic spinning vertigo and hearing abnormalities [(4)].

Strict diagnostic criteria based on symptom categories were developed by the American Academy of Otolaryngology—Head & Neck Surgery (AAO-HNS) 2015 to facilitate the diagnosis and management of MD. MD can be categorized as either defined or probable. For the diagnosis of MD, there is no definitive test [(5)].

A) Definite MD

The AAO-HNS stated that this condition is associated with two or more spells of vertigo lasting between twenty minutes and twelve hours, audiometric confirmation of low- to mid-frequency hearing loss concomitant with vertigo, and fluctuating aural symptoms in the affected ear. It's critical to discriminate MD from other prevalent causes of vertigo that may also be accompanied by hearing loss, tinnitus, or auditory fullness [(5)].

B) Probable MD

At least two attacks of dizziness or vertigo lasting for twenty minutes to several hours, varying aural symptoms (such as hearing loss, tinnitus, or fullness), and other reasons that have been ruled out by further testing. Although the condition often manifests as unilateral ear symptoms, they may last for many years. Around six to eleven attacks of MD occur episodically and randomly on average every year. Remission intervals can range from months to years. Hence, the diagnosis of MD is not usually made entirely at once. It could take a long time to

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experience all of the clinical symptoms necessary for a conclusive diagnosis [6].

Clinical presentations of MD

A) Vertigo

Vertiginous attacks frequency varies widely across MD subjects. These attacks are typically not positional in nature and are described as spinning or motion feelings. Attacks typically range from a few minutes to several hours and are complemented with nausea and vomiting. Most patients experienced attacks that were of a moderate degree. About half of patients with follow-up over ten years reported total clearance of vertiginous symptoms, with the majority of patients experienced some degree of symptom resolution. While, drop attacks (Tumarkin's Otolithic Crisis) are sudden falls accompanied by MD episodes that occur without prior notice and it doesn't cause the patient to lose consciousness. These attacks can manifest in the advanced stages of MD, although not all patients experience them [7].

B) Hearing Loss

In the early stage of MD, it is classically associated with a sensorineural hearing loss at low frequencies**.** Hearing may improve between episodes, but as the disease progresses, permanent losses are apparent leading to inverted V audiogram. Almost 50% of patients totally lose their hearing in the affected ear fourteen years following their diagnosis [8]. According to the severity of hearing loss between attacks, there are four stages of hearing loss in MD (500 HZ-4 KHZ average): stage one ≤ 25 dB HL), stage two (26-40 dB HL), stage three (41-70 dB HL), and stage four (>71 dB HL) [9].

C)Tinnitus

Tinnitus presents in the majority of MD patients. Early in the disease, tinnitus and vertigo coexist, with tinnitus increasing in severity before attacks. Tinnitus worsens over time and becomes chronic, even when there are no attacks, with the progression of the disease. It described as "roaring" or "ringing" and corresponds to hearing loss [8].

D) Aural fullness

Aural fullness in the affected ear is another common symptom of MD patients. Patients may define a "pressure" or "clogged sensation" in their ears. Before to an attack, this sense is frequently demarcated as moderate to severe with a sudden peak in severity. Normal resolution of aural fullness with time is rare [10].

Differential diagnosis of MD:

The exclusion of other common disorders is necessary for the diagnosis of MD. The most frequent reason of episodic vertigo is vestibular migraine (VM), which is diagnosed when a patient reports having recurrent episodes of vertiginous attacks with accompanying migrainous symptoms. It may be difficult to distinguish VM from MD in the early phases of MD since cochlear symptoms may not exist and hearing may completely improve in between spells. Hearing loss can assist distinguish between MD and VM since it is less likely in VM patients than in MD patients for persistent unilateral hearing loss [11].

Other disorders, such as autoimmune inner ear disease, vestibular schwannoma, otosyphilis, vestibular neuritis, and acute labyrinthitis are further differential diagnoses. According to the patient's medical history, audiometry, and imaging, these disorders are typically simpler to distinguish from MD [12].

Role of electrocochleography (ECOG) as a documented test for diagnosis of MD

In MD, it is believed that ears with hydrops show an increase in SP amplitude due to the basilar membrane distention to the scala tympani. Elevated SP amplitude in comparison to AP amplitude may be a reliable predictor for EH [13]. A level of more than 0.5 is regarded as abnormal in all clinics and typically denotes the existence of an inner ear disorder such as MD. The normal values varied from 0.35 to 0.5 [14].

For the MD diagnosis, an increased SP/AP amplitude ratio is considered specific but not sensitive. ECOG sensitivity has been reported to range from 30% to 70%, while specificity for MD has been reported to range from 92% to 100% [15]. In more recent research, higher sensitivity has been attained by combining the SP/AP amplitude ratio with other ECOG data, as the SP/AP area ratio or with audiological measurements (sensitivity 80%). Early helpful findings have been obtained from integrating the SP/AP amplitude ratio with other audiological testing [16].

As MD progresses, the sensitivity of ECOG increases because people with milder disease might not have advanced cochlear changes that induce abnormal ECochG, which reduces the sensitivity to detect pathology [17].

Role of videonystagmography (VNG) and video head impulse (VHIT) in MD

In VNG, eye movements are recorded while a series of tests are performed to assess vestibular function. One of these techniques is caloric testing, which is considered most effective at identifying unilateral peripheral vestibular hypofunction. Using temperature-driven, low-frequency lateral semicircular canal stimulus, the caloric test provides information specific to the ear. Based on caloric testing, 42% to 84% of MD patients in cross-sectional studies and case series exhibit unilateral weakness. However, normal caloric testing does not exclude MD [18]. The remaining VNG results in MD

patients are often normal and include saccades, smooth pursuit, gaze, optokinetic, and positioning tests [19].

Using high-frequency stimulation and high-speed records of eye movements before and after rapid head impulses, the video head impulse test (vHIT), additional vestibular test, determines the independence of the function of each six semicircular canals. A functional canal deficiency causes vHIT to produce a pattern of corrective saccades and a measurement of vestibulo-ocular reflex (VOR) gain [20].

AAO-HNS criteria revealed that patients with definite MD had aberrant caloric values, of which forty five% had normal vHIT results [18]. There are a few explanations for this mismatch, one of which is that MD damages type II hair cells specifically, impairing their ability to produce the crista's low-frequency response during caloric testing while protecting their ability to produce the high-frequency response during vHIT [21].

Role of Vestibular evoked myogenic potentials (VEMPs) in Meniere's disease

The functionality of the otolith organs and their afferent vestibular pathway is assessed using VEMPs. cVEMPs assess the saccule and inferior vestibular nerve, whereas oVEMPs assess the utricle and superior vestibular nerve. The research on the application of cVEMP and/or oVEMP for the diagnosis of MD was changed for the 2017 American Academy of Neurology practice recommendation. Findings showed that no research demonstrated that the utility of VEMP as a gold standard test for the diagnosis of MD [22].

By comparing the two ears, VEMP's diagnostic usefulness in cases with unilateral MD may be increased. Smaller cVEMP amplitudes are observed in the affected ear in patients with unilateral MD as a result to 500 Hz tone burst, according to VEMP data [23]. Moreover, the cVEMP 1,000/500 Hz amplitude ratio is increased in the ears of MD patients, demonstrating dynamic modifications in the mechanics of the saccular membrane motion in hydrops. The saccular membrane becomes distended as a result of EH, which reduces low frequency responses. Also, the degree of membrane distention and membrane electrical resonance can alter depending on how severe the hydrops is, which means that variations in the cVEMP tuning may correspond to how severe the saccular hydrops is [24].

Moreover, VEMPs might help in predicting the progression of bilateral MD. A potential role in diagnosing asymptomatic or presymptomatic MD is suggested by the fact that unaffected ears showed a different cVEMP frequency tuning compared to that of ears with a hydrops [25]. In acute episodes and later stages of disease, cVEMP testing's sensitivity, specificity, and accuracy improved [26].

CONCLUSION

MD is diagnosed clinically since it typically manifests as unilateral ear problems that might remain for decades. Attacks from MD are sporadic and episodic, with remissions in between. As there is no definitive vestibular testing for MD, these tests are typically carried out to rule out disorders with similar symptoms.

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REFERENCES

1. **Christopher L, Wilkinson E (2021):** Meniere's disease: Medical management, rationale for vestibular preservation and suggested protocol in medical failure. American Journal of Otolaryngology, 42 (1):3-28
2. **Merchant S, Adams J, Nadol J (2005):** Pathophysiology of Meniere's syndrome: are symptoms caused by endolymphatic hydrops? Otology & Neurotology, 26 (1): 74-81.
3. **Luryi A, Morse E, Michaelides E (2019):** Pathophysiology and diagnosis of Meniere's disease. Diagnosis and treatment of vestibular disorders. https://doi.org/10.1007/978-3-319-97858-1_13
4. **Oberman B, Patel V, Cureoglu S *et al*. (2017):** The aetiopathologies of Ménière's disease: a contemporary review. Acta Otorhinolaryngologica Italica, 37 (4): 241-250.
5. **Basura G, Adams M, Monfared A *et al.* (2020):** Clinical practice guideline: Ménière's disease executive summary. Otolaryngology–Head and Neck Surgery, 162 (4): 415-434.
6. **Ihler F, Stoycheva I, Spiegel J *et al*. (2021):** Diagnosis of Meniere's disease according to the criteria of 2015: Characteristics and challenges in 96 patients. Journal of Vestibular Research, 32(3): 271-283.
7. **Sumi T, Watanabe I, Tsunoda A *et al*. (2012):** Longitudinal study of 29 patients with Meniere's disease with follow-up of 10 years or more (In commemoration of Professor Emeritus Isamu Watanabe). Acta oto-laryngologica, 132 (1): 10-15.
8. **Ueberfuhr A, Wiegrebe L, Krause E *et al*. (2017):** Tinnitus in normal-hearing participants after exposure to intense low-frequency sound and in Ménière's disease patients. Frontiers in neurology, 239 (7):8-11.
9. **Zhang Y, Liu B, Wang R *et al*. (2016):** Characteristics of the cochlear symptoms and functions in Meniere's disease. Chinese Medical Journal, 129 (20): 2445-2450.
10. **Levo H, Kentala E, Rasku J *et al.* (2014):** Aural Fullness in M3ni2re's Disease. Audiology and Neurotology, 19 (6): 395-399.
11. **Dieterich M, Obermann M, Celebisoy N (2016):** Vestibular migraine: the most frequent entity of episodic vertigo. Journal of neurology, 263 (1): 82-89.

12. **Pyykkö I, Nakashima T, Yoshida T *et al.* (2013):** Meniere's disease: a reappraisal supported by a variable latency of symptoms and the MRI visualisation of endolymphatic hydrops. BMJ open, 3 (2): 7-11.
13. **Eggermont J (2017):** Ups and downs in 75 years of electrocochleography. Frontiers in Systems Neuroscience, 11 (2): 3-21.
14. **Chung W, Cho D, Choi J *et al.* (2004):** Clinical usefulness of extratympanic electrocochleography in the diagnosis of Meniere's disease. Otology & neurotology, 25 (2): 144-149.
15. **Kim H, Kumar A, Battista R *et al.* (2005):** Electrocochleography in patients with Meniere's disease. American journal of otolaryngology, 26 (2): 128-131.
16. **Gerenton G, Giraudet F, Djennaoui I *et al.* (2015):** Abnormal fast fluctuations of electrocochleography and otoacoustic emissions in Menière's disease. Hearing Research, 327: 199-208.
17. **Ziylan F, Smeeing D, Stegeman I *et al.* (2016):** Click stimulus electrocochleography versus MRI with intratympanic contrast in Ménière's disease: A systematic review. Otology & Neurotology, 37 (5): 421-427.
18. **Cordero-Yanza J, Arrieta E, Hernaiz J *et al.* (2017):** Comparative study between the caloric vestibular and the video-head impulse tests in unilateral Menière's disease. Acta Oto-Laryngologica, 137 (11): 1178-1182.
19. **Lee S, Kim H, Koo J *et al.* (2017):** Comparison of caloric and head-impulse tests during the attacks of Meniere's disease. The Laryngoscope, 127 (3): 702-708.
20. **McGarvie L, Curthoys I, MacDougall H *et al.* (2015):** What does the dissociation between the results of video head impulse versus caloric testing reveal about the vestibular dysfunction in Ménière's disease? Acta oto-laryngologica, 135 (9): 859-865.
21. **McCaslin D, Rivas A, Jacobson G *et al.* (2015):** The dissociation of video head impulse test (vHIT) and bithermal caloric test results provide topological localization of vestibular system impairment in patients with "definite" Ménière's disease. American journal of audiology, 24 (1): 1-10.
22. **Fife T, Colebatch J, Kerber K *et al.* (2017):** Practice guideline: Cervical and ocular vestibular evoked myogenic potential testing: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. Neurology, 89 (22): 2288-2296.
23. **Tabet P, Saliba I (2017):** Meniere's disease and vestibular migraine: updates and review of the literature. Journal of clinical medicine research, 9 (9): 727-733.
24. **Angeli S, Goncalves S (2019):** Cervical VEMP tuning changes by Meniere's disease stages. Laryngoscope investigative otolaryngology, 4 (5): 543-549.
25. **Lin M, Timmer F, Oriel B *et al.* (2006):** Vestibular evoked myogenic potentials (VEMP) can detect asymptomatic saccular hydrops. The Laryngoscope, 116 (6): 987-992.
26. **Zhang S, Leng Y, Liu B *et al.* (2015):** Diagnostic value of vestibular evoked myogenic potentials in endolymphatic hydrops: a meta-analysis. Scientific reports, 5 (1): 1-10.