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Review

Ménière disease: diagnostic instrumental support

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Abstract The current guidelines of the American Academy of Otolaryngology—Head and Neck Surgery entrust the diagnosis of Ménière disease (MD) only to the clinical presentation and the pure tone audiometry. However, most otolaryngologists request a widened instrumental evaluation of the patients suspected of MD. The effective reliability of the further instrumental support for the diagnosis of MD is still debated in the literature because of nonstandardized procedures and sometimes incoherence among authors. New and more sophisticated diagnostic tests have been developed both in audiovestibology and in imaging in the last few years. A review of the recent literature on this controversial subject is provided.

1. Introduction

Ménière disease (MD) commonly presents with recurrent, spontaneous episodic vertigo, hearing loss, aural fullness, and tinnitus. The natural history of the disease is variable in intensity and frequency among individuals, possibly evolving over months or years [1]. The so-called idiopathic endolymphatic hydrops—an increase of the endolymphatic pressure leading to the distension and sometimes to the rupture of the membranous labyrinth—is strongly associated with MD, although the causal relation between endolymphatic hydrops and symptoms of MD remains unproven [1] and hydrops has been sometimes reported only as an epiphenomenon of the disease [2,3]. Indeed, the after-death histopathologic confirmation of hydrops is required for a certain diagnosis of MD, whereas the presence of possible, probable, or definite MD can be inferred during life by clinical suspicion and pure tone audiometry as stated in the current guidelines of the American Academy of Otolaryngology—Head and Neck Surgery [4]. Unlike the guidelines, a survey on the management of MD [5] revealed that most otolaryngologists pursue further diagnostic studies either to investigate the diseases that may mimic MD attacks or to assess the hypothesis of an endolymphatic hydrops. The most striking research of the last few years in this field has been directed toward the early detection of endolymphatic hydrops, in particular when the symptoms of MD are less definite or still absent [6,7]. An overview of the instrumental tests that are available to assess MD is provided.

2. Audiologic and vestibular tests

Some audiologic and vestibular tests used in the evaluation of patients suspected of MD were classified by de Sousa et al [11] in 2002: pure tone audiometry, speech audiometry, brainstem-evoked auditory responses (BEAR), immittance tests, and instrumental-aided study of nystagmus—in particular electronystagmography with caloric testing—were considered "routine" tests, whereas traveling-wave velocity (TWV) technique, electrocochleography (ECoG), and dehydration test with glycerol were included in the "extended" group [11]. Low-frequency masking (LFM), otoacoustic emissions (OAE), and vestibular-evoked myogenic potentials (VEMP) have been also reviewed.

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2.1. Audiologic tests

2.1.1. Pure tone and speech audiometry

The American Academy of Otolaryngology—Head and Neck Surgery current guidelines assert the essential role of pure tone audiometry in the diagnosis of MD [4]. The audiometric presentation of a patient with MD can be variable. In the early stages of the disease, a low-frequency sensorineural hearing loss is expected, usually with a peak pattern at the onset [8]; but different configurations are possible. Fluctuation is common, but it is not essential for the diagnosis [4]. The pure tone audiometric threshold is usually expected to increase in the later stages and the audiogram to become more flat [4], although Mateijsen et al [9] concluded that neither the shape of the pure tone audiogram nor the average hearing loss correlates with the duration of disease. Furthermore, no reduced speech discrimination relative to the expectations based on pure tone loss, as otherwise reported in the previous literature, was observed [9]. Occasionally, the presence of a low-frequency air-bone gap without middle ear disease is reported in early MD, probably as a result of the inner ear pressure exerted against the medial surface of the stapedial footplate during bouts of hydrops [10]. This conductive component may be a pitfall for patients with MD having a feeling of aural fullness because they may be mistakenly treated for middle ear disease [11].

2.1.2. Brainstem-evoked auditory responses and traveling-wave velocity

Brainstem-evoked auditory responses are commonly tested for the evaluation of asymmetric sensorineural hearing loss to exclude the presence of retrocochlear lesions that may also mimic the clinical features of MD. Indeed, BEAR were found to be poor in being a reliable screening test for possible retrocochlear disease when compared to magnetic resonance (MR) imaging. The sensitivity was 74% and the specificity was 71% in a large-scale, multi-institutional, prospective study [12]. Therefore, it is questionable how much BEAR may help in the differential diagnosis of MD. Brainstem-evoked auditory responses may show 3 different patterns that may suggest the presence of hydrops in MD-affected ears as reported by de Sousa et al [11]. In the first type, which is predominant, the affected ear has an equal or even shorter fifth-wave latency when compared with the healthy ear in unilateral MD. This could represent the manifestation of an electrophysiologic recruitment. In the second type, there is a delayed fifth-wave latency in the affected ear. In the third type, a shift of first-, third-, and fifth-wave latency is observed. This "conductive pattern" could be explained by the displacement of the footplate of the stapes toward the middle ear because of the hydrops [11].

An interesting application of BEAR technique is the measurement of TWV, which requires a slightly modified BEAR equipment. Traveling-wave velocity represents the different delay of activation between the basal and apical acoustic cells. The rationale of measuring TWV is that hydrops causes the stiffening of the basilar membrane because of the increased pressure in the scala media, leading to faster traveling waves, in particular in the apical portion. The responses of different regions of the Corti organ are obtainable by presenting properly high-pass and low-pass masked clicks to the tested ear. A difference in the latency of the fifth wave of less than 0.6 milliseconds is indicative of a raised scala media pressure [13]. However, wave V is not very easily identifiable in the masked condition. This problem, along with others, currently prevents the routine use of this testing technique [14].

2.1.3. Low-frequency masking

Low-frequency masking is an emerging method for the early diagnosis of endolymphatic hydrops in MD. Low-frequency tones are responsible for the displacement of the whole basilar membrane, which is most evident at the apex of the cochlea. A short, high-pass–filtered, acoustic stimulus and the LFM tone are applied to the same ear in an adjustable phase relationship; and subjective hearing thresholds are recorded [15]. A maximum masking threshold is usually found at 270° phase lag, and there is a mean maximal difference (*modulation depth*) of 28 dB in masking among phases (0°–270°) in healthy subjects. The phase dependence may be totally absent in patients with MD and may change as the disease progresses. Modulation depth is also often significantly reduced in the nonsymptomatic ears of patients with MD [16]. Moreover, Don et al [17] found that in symptomatic patients with MD the ipsilateral masking noise is insufficient so that an undermasked wave V of the BEAR is still present at a latency similar to that of wave V, whereas it is absent or significantly delayed in the same conditions in healthy subjects. The difference in delays between pathologic and normal populations varies among authors, resulting in a 31%-100% sensitivity and a 28%-100% specificity [17,18]. Considering the good conformity of LFM to ECoG findings, some authors recommend LFM for the detection of endolymphatic hydrops [19], but others report low reliability and high rates of non-interpretable tests (49%) [18].

2.1.4. Immittance tests

The first studies on immittance tests in MD were published in the 1970s and provided contradictory results [20,21]. Meanwhile, clinical immittance was gaining popularity for the analysis of the middle ear condition and the study of the cochleostapedial reflex, and it is now available as a routine audiologic test for this purpose. Only one preliminary study, which evaluates tympanometry results at selected frequencies, has been published recently on immittance tests in MD by Franco-Vidal et al [22]. Multifrequency tympanometry allows the finding of the resonance frequency of the middle ear, studying its immittance components (conductance and susceptance) for each selected frequency, being able to assess mass or stiffness loads. Multifrequency tympanometry parameters may be altered in MD with endolymphatic hydrops. The reason is probably the influence that the increased pressure has on the round window and the oval window. Resonance