Operative Techniques in Otolaryngology (▮▮▮▮) ▮, ▮▮▮–▮▮▮

ELSEVIER

Operative Techniques in Otolaryngology

Meniere's disease an overview

Hetal H. Patel, MD, Huseyin Isildak, MD

From the Division of Otolaryngology, Penn State Hershey Medical Center, Hershey, Pennsylvania

KEYWORDS
Meniere's Disease;
endolymphatic hydrops;
vertigo attacks

The following is a brief overview of Meniere's Disease (MD). It is important to recognize that MD is a diagnosis of exclusion. Although there are established histopathologic changes identified in MD, the exact pathophysiology remains controversial. There are various treatment options including diet modification and medical management that can help control symptoms. For patients who have intractable symptoms there are a number of surgical interventions that can be offered.

Introduction

Prosper Meniere first described the entity known today as Meniere's Disease (MD) in 1861 as the clinical combination of the following symptoms: aural fullness, tinnitus, and fluctuating hearing loss associated with episodes of vertigo.[1] Today it is known that the disease is characterized by recurrent attacks of these symptoms associated with a progressive hearing loss over time. Presentation and clinical course can be variable with some patients presenting with both auditory and vestibular symptoms and others with either symptom alone. Usually patients will experience clusters of attacks followed by long periods without vertigo. Symptoms may continue for many years, but many patients will have resolution of vertigo attacks by 2-8 years.[2] Patients generally present in the third to seventh decades with MD and there is a slight female predominance. The incidence of MD seems to vary based on geographic location, with highest rates reported in Finland (513/100,000), the reported incidence in the United states is approximately 1 in 500.[2,3]

Pathophysiology

It has been well described that endolymphatic hydrops is found in temporal bones of patients with MD. There remain many controversies regarding the etiology of the hydrops. The theories are centered on explaining the histologic findings noted in temporal bones of experimentally induced endolymphatic hydrops. These findings consist of "ruptures of the membranous labyrinth, fistulas of the membranous labyrinth, collapse of the membranous labyrinth, obstruction of longitudinal flow, vestibular fibrosis, sensory lesions, and neural lesions."[4]

In a healthy cochlea there are 3 chambers comprised of the scala tympani, scala vestibuli, and scala media. The scala media is a part of the membranous labyrinth and contains endolymph. The other parts of the membranous labyrinth are the semicircular canals (SCCs), saccule, utricle, and endolymphatic duct and sac; these structures are all in continuity and contain endolymph. The electrolyte balance of the endolymph differs from other extracellular fluid and is potassium rich with a low sodium concentration. This gradient is the foundation for function of the hair cells of the inner ear. The theory proposed by Schuknecht is that ruptures occur in Reissner membrane that allow the potassium rich endolymph to enter the perilymph. These potassium levels may be toxic to the hair cells and the cochlear nerve exposed in this area.[5] It is theorized that the sudden increase in potassium levels causes the episodic symptoms of tinnitus, and vertigo. Eventually the membrane heals and the exposure stops until the membrane ruptures again in a potentially different location. Over time and after many such ruptures there are changes to the cytoarchitecture of the sensory

Address reprint requests and correspondence: Huseyin Isildak, MD, Division of Otolaryngology, Penn State Hershey Medical Center, 500 University Dr, Hershey, PA 17033.
E-mail address: hisildak@hmc.psu.edu

http://dx.doi.org/10.1016/j.otot.2016.10.001
1043-1810/

cells and their supporting cells. These changes can be correlated to the progressive hearing loss and loss of vestibular function on the affected side.[6] Other theories suggest there is a role in the disruption of radial flow of the endolymph resulting in the endolymphatic overflow in to the saccule. The result is distension of the saccule and altered cochlear and vestibular function via traveling waves and contact within the crista ampullaris. Yet another theory is that of obstruction within the endolymphatic duct. The blockage results in secretion of osmotic agents within the endolymphatic sac to help draw fluid and clear the debris. The sudden release of the fluid is thought to result in the symptoms of Meniere attacks.[4] There are also theories that focus on poor cochlear perfusion, and immunologic dysfunction as potential etiologies of MD.

The many pathophysiologic theories suggest that MD is multifactorial and there seems to be genetic and environmental factors that play a role in the development of MD.

Diagnosis

Unfortunately, patients often present with variable signs and symptoms and a biological marker for diagnosis is lacking. To develop an international consensus, the Classification Committee of the Bárány Society, The Japan Society for Equilibrium Research, the European Academy of Otology and Neurotology, the Equilibrium Committee of the American Academy of Otolaryngology—Head and Neck Surgery and the Korean Balance Society have jointly published diagnostic criteria. There are 2 categories identified, definite and probable MD. Definite MD is defined as 2 or more episodes of spontaneous vertigo lasting from 20 minutes to 12 hours, documented low-medium frequency sensorineural hearing loss in the identified affected ear, and fluctuating aural symptoms. Additionally, the symptoms must not be explained by another vestibular diagnosis. Probable MD is defined as 2 or more episodes of spontaneous vertigo lasting from 20 minutes to 12 hours and fluctuating aural symptoms in the affected ear not accounted for by another diagnosis.[3]

Despite published guidelines it can be challenging to establish the diagnosis of MD. In evaluating patients suspected of MD, it is important to have a broad differential and rule out other potential etiologies of the patient's symptoms. According to the recently published diagnostic criteria important considerations on the differential diagnosis include hereditary nonsyndromic sensorineural hearing loss, stroke, Cogan syndrome, endolymphatic sac tumor, meningioma, third window syndromes, vestibular schwannoma, and a few others.

Although MD is a clinical diagnosis there are vestibular studies that may aid in establishing the diagnosis. The most commonly used adjunctive study is electrocochleography (ECoG), which is thought to indicate elevated endolymphatic pressure when the summating potential or action potential ratio of the auditory nerve is elevated.[7] Unfortunately, the specificity of the test can be limited by similarly elevated ratios noted in superior semicircular canal dehiscence. Additionally, ECoG values may not be altered in between symptomatic episodes and patients with definitive MD may not always have elevated summating potential/action potential ratios. Electronystagmography can also be used an adjunctive study. Patients with MD should exhibit weakness of the vestibular response on the side that is affected. Unfortunately, the sensitivity of the study has only been reported at 50% and testing is not accurate if antivertiginous medications have been taken within 2 weeks.[8] In a similar manner, vestibular evoked myogenic potential (VEMP) from the cervical muscles (cVEMP) and extraocular muscles (oVEMP) can be decreased in MD. The use of these tests may help in determining the extent of progression because of the correlation of cVEMP with the saccule and the oVEMP with the utricle.[8] In the future there may be a more prominent role for imaging in MD. Studies have demonstrated decreased enhancement in the perilymph in patients with MD in comparison to normal controls after intratympanic contrast injection. Additionally, the severity of imaging findings have been correlated to the severity of symptoms the patient is experiencing. Overall, it remains difficult to establish a definitive diagnosis of MD because of the variability of symptoms. It is important to follow patients over time and monitor symptoms to have a clear understanding of their disease process.

Treatment

The ideal treatment for MD would result in fewer and less severe attacks of vertigo with associated hearing loss and tinnitus. Additionally, optimal treatment would ameliorate the potential chronic symptoms of tinnitus and imbalance and prevent further progression. Unfortunately, no such medication or treatment exists; regardless there are a number of interventions that are recommended for patients with some potential benefits.[9]

Diet modification

Generally, diet modification is considered a primary treatment modality for MD. Excessive consumption of salt is thought to exacerbate endolymphatic hydrops by some otologists. Additionally, there are some patients who experience MD attacks after salt binges.[10] There are also patients who experience significant improvement in symptoms and a decreased frequency of episodes with low salt diets.[11,12] There are varying recommendations on the maximum salt intake advised between 1 and 2 g/d.[10] Although there is no consensus many providers also recommend discontinuing use of caffeine, tobacco, and alcohol products.[12]

Medications

Diuretics are a popular initial treatment medication for MD. Theoretically, diuretic therapy changes the electrolyte balance of the endolymph and could cause improved drainage or decreased production of the endolymph. A recently published Cochrane review verifies that there