Editorial

Ménière's Disease

In over a half-century as an academician and practitioner, I have had a fascination with Ménière's disease (MD). Entering the discipline when some "authorities" were teaching and writing that sensorineural hearing loss was a condition that either, and hopefully, stayed the same or got worse, MD showed characteristic fluctuations often for the better in pure-tone air and bone conduction and word recognition. It appeared to me to represent a *series* of disorders of the sensorineural mechanism showing audiologic reversals, for example, noise-induced hearing loss, sudden idiopathic sensorineural hearing loss, allergies of the inner ear, multiple sclerosis (Miller et al, 1957). But MD with its paroxysmal character caused such distress and anguish for the sufferer that it stood in marked contrast to the "typical" patient with sensorineural hearing loss whose problem, so often associated with the aging process, was a stealthy process that could be, and was, ignored for years before anything was done about it. As a young practitioner, long before the era of electroacoustic and electrophysiologic measures had arrived, I did crude tolerance testing with pure-tone stimuli to evoke a sensation of discomfort, and one of the first patients on whom I did this procedure vomited on the equipment, the floor of the audiometric booth, and on me, when levels of 90+ dB HTL were reached for frequencies of 250 and 500 Hz in the affected ear. Even in the quiescent stage of the disease, fitting patients with the "new" transistorized hearing aids was a formidable challenge for the patients with fluctuating hearing loss, reduced dynamic range, and impaired speech discrimination.

Ménière's disease or syndrome does not occupy the same prevalence role in the panorama of sensorineural hearing loss as noise-induced hearing loss, presbycusis, or ototoxic medications. (The prevalence of MD in the United States is reported as 218 cases per 100,000) (Wladislabosky et al, 1984). It does, however, in the opinion of many practitioners, result in more suffering and anguish than all other otic and auditory diseases combined. Its paroxysmal character and patterns of remission and exacerbations create a research conundrum in efforts to evaluate the efficacy of various therapeutic interventions, both medical and surgical. For the audiologist, some of these patients suffer from extremely reduced intermittent inability to discriminate speech during acute attacks and accompanying severe vertigo, which present among the most formidable and difficult challenges in order to provide successful amplification and audiologic rehabilitation. A roaring tinnitus during the acute stages of the disease presents further problems to the sufferer. Between attacks, auditory function may be normal or near normal, but the fear of the recurrent attack often leaves the patient psychologically compromised.

This symptom complex was first described by Prosper Ménière in 1861, although he may have been describing a disease other than what we now know as MD. We now consider the disease as involving the membranous inner ear, whose symptoms consist of vertigo, hearing loss, tinnitus, and according to some, a sensation of aural fullness in the affected ear (Alfaro, 1959).

This disease is attributed to excessive endolymphatic fluid pressure in the membranous labyrinth causing Reissner's membrane to become distended (Gelfand, 2001). But what factor or factors cause the endolymphatic hydrops? Because of the excess production and/or absorption of endolymph, the condition is often referred to as "endolymphatic hydrops." It has been postulated that the membrane may perforate during the acute attack and reattach itself between attacks. Attacks may last from 20 minutes to as long as several hours or days. The patient is often acutely ill with severe dizziness and vomiting during attacks. There is usually some warning that an attack is imminent allowing the patient to pull over to safe area if driving a car or cease the operation of potentially dangerous equipment.

The hearing loss in MD is unilateral in about 70–85% of cases. However, the incidence of bilaterality increases with the duration of the disease, reaching about 40% after 15 years (Morrison, 1976). The incidence of bilateral disease has significant implications for choice of therapy in intractable cases, that is, protection of residual hearing in efforts to target vertiginous attacks.

In the early stages of MD, the characteristic audiometric configuration is a rising curve; that is, as the frequency increases, the hearing loss decreases. This contour has also been called a "reverse slope" audiogram, since high-frequency losses are by far the most frequent pattern in the hearing-impaired population. Reverse slope and abnormal PI-PB functions present extreme challenges for success in audiologic rehabilitation. Word recognition (discrimination) has been reported in some studies as poor as 32%. Although mid- and high-frequency sensitivity tend to be good in the early stages, as the disease progresses, these frequencies become involved, leaving the patient with a "flat" audiometric configuration. The hearing loss may progress to a profoundly impaired degree as the disease process continues. However, the degree of hearing loss seldom exceeds a 70 dB average.

The tinnitus may with vertiginous attacks be among the most disabling aspects of the disease. The tinnitus, as mentioned earlier, is of often of a roaring character with a strong low-frequency component. MD shows a prevalence of .05 to .15% of the population. It is considered by some (Schwaber, 1998) to be the third most common inner ear disorder after presbycusis and noise-induced hearing loss, although I consider hearing loss secondary to ototoxic medications a far more frequent cause of cochlear hearing loss than MD. Additionally, presbycusis does not affect only the cochlea. The onset of MD is at 40–60 years of age in 50% of affected persons. The sensorineural hearing loss is accompanied by loudness recruitment (often hyper-recruitment), a narrowed range of comfortable loudness and severe acoustic distortions. This combination of acoustic factors plus the paroxysmal nature of the disease creates major problems in otic and audiologic management of the MD sufferer.

In this special issue of *JAAA*, we have assembled a distinguished panel of experts on different aspects of Ménière's disease. George Gates, one of otology's leading figures, who has devoted considerable efforts to develop treatments that work, contributes a section from the historical identification of the syndrome to the recently introduced Meniett device. Drs. Ghossaini and Wazen of the Department of Otolaryngology—Head and Neck Surgery of Columbia University College of Physicians and Surgeons review and evaluate the pros and cons of various surgical approaches to provide relief for patients with incapacitating forms of the disease. Both Dr. Gates and Drs. Ghossani and Wazen review transtympanic injections of gentamycin, a less invasive surgical approach to relieve the acute vertiginous attacks but that places residual hearing at some risk. ECochG, one of the premium techniques for diagnosing and monitoring the disease, is described by John Ferraro and John Durrant, who bring to the assignment years of research and clinical experience with the technique as applied to the MD population. Since MD is considered by most to be a disease of the vestibular labyrinth, we are pleased to have Neil Shepard to review the vestibular evaluation of the MD patient with some consideration of the role of vestibular rehabilitation in the management of these patients. Michael Valente and associates write of audiologic rehabilitation and provide a positive upbeat tilt on what amplification can do for sufferers of the disease—an optimistic view that stands in sharp contrast with much that has been written and taught about the ability of these patients to derive benefits from amplification.

Still no cure, still no etiology, but a wealth of improved diagnostic and therapeutic procedures to offer these patients at least relief. We extend appreciation to all the contributors to this special issue for sharing their wisdom and experience on what remains a major unresolved challenge for all who encounter patients with MD.

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