

Ménière's Disease:

Diagnosis *in the Balance*

Ménière's disease is defined as an idiopathic syndrome of endolymphatic hydrops, which is an abnormal increase in the volume of fluid (endolymph) in the inner ear. This excessive increase of fluid in the inner ear, or hydrops, can leak and disrupt the function of the inner ear, leading to bouts of vertigo, as well as fluctuating sensorineural hearing loss, ringing in the ears (tinnitus) and a sensation of fullness in the ear. It is not known what causes hydrops, but hypotheses include allergies, genetic flaws, autoimmune disorders and systemic viral infections. It is estimated that approximately 600,000 individuals in the United States suffer from Ménière's disease, or nearly eight people per 1,000. Although Ménière's is not a life-threatening disease, it

can be frustrating and the acute bouts of vertigo can be frightening and debilitating.

In an attack of dizziness from Ménière's disease, the patient generally experiences a spontaneous rotational vertigo lasting at least several minutes to several hours. The feeling of intense motion when sitting or standing still can cause the patient to fall to the ground and is often accompanied by nausea and vomiting. The natural history of Ménière's disease is poorly understood and highly variable, making diagnosis difficult. The extent of the economic impact on Ménière's disease patients who are unable to work is unknown,

in addition to the health care costs, but it is clear that Ménière's patients can experience costly and severe disability. The first step to extending treatment to these patients is an accurate evaluation of their symptoms.

Research scientist Manuel Don, Ph.D., who heads the Electrophysiology Department at the House Ear Institute, recently patented a diagnostic test for Ménière's disease and cochlear hydrops using

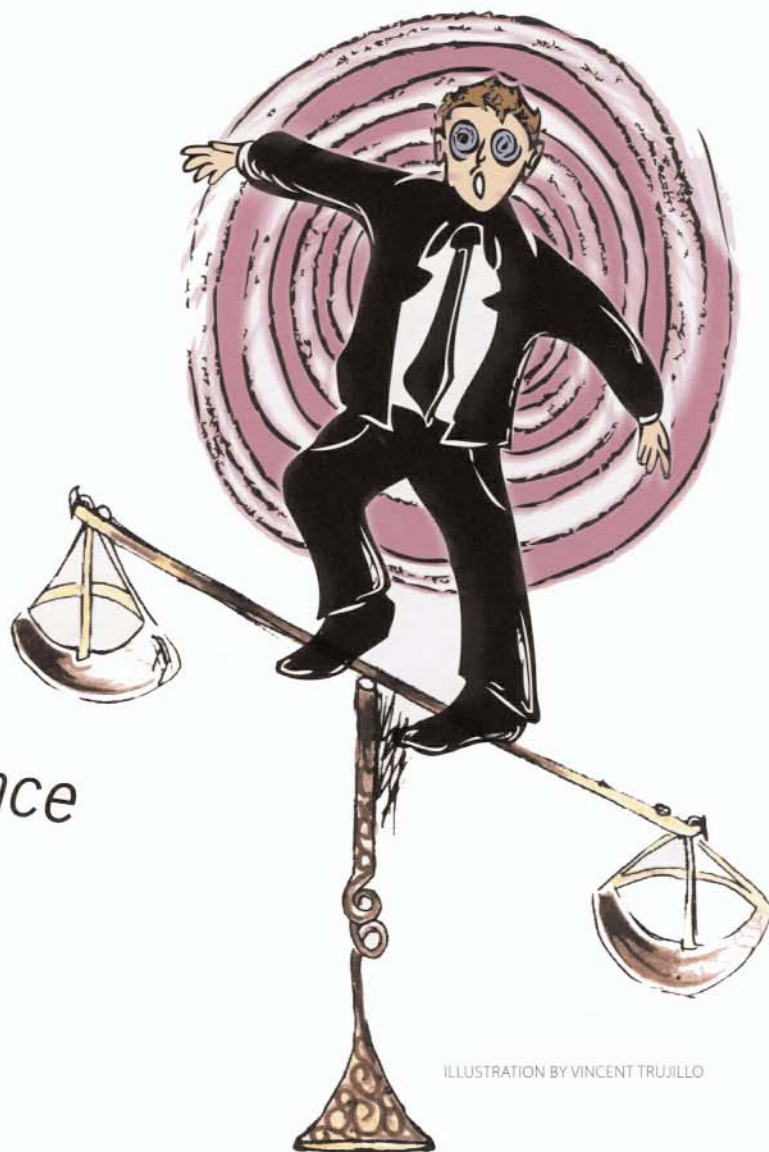


ILLUSTRATION BY VINCENT TRUJILLO

impaired high-pass noise masking of auditory brainstem responses (ABR). Dr. Don and colleagues understood the frustration for both patients and physicians surrounding the difficulty in arriving at an unequivocal diagnosis of Ménière's disease. This difficulty was the impetus to finding a diagnostic test that would have high sensitivity and specificity in an individual patient seeking treatment. Unfortunately, a definite identification of Ménière's disease requires observation of the affected membranes postmortem (during an autopsy). Since postmortem verification is not an option (!) for patients actively seeking relief from their symptoms, this diagnostic study of Ménière's, like others, relied on the physician's confident assessment that a patient is correctly classified as having Ménière's disease. A physician relies on a thorough evaluation of patient history and observation of four of the classic symptoms of Ménière's: vertigo, head noise (tinnitus), fluctuating hearing loss, ear fullness.

In Dr. Don's study, he and his team only included patients who had been confidently diagnosed with Ménière's

disease and, at the time of ABR testing, still reported three to four of the classic symptoms of the disease. There were many other patients who had been initially diagnosed with Ménière's disease but at the time of recruitment and testing in Dr. Don's lab had two or fewer symptoms. The reduction in symptoms may have been the result of treatment or part of the natural course of the disease. The strict criteria of testing patients diagnosed with Ménière's disease and having three to four of the classic symptoms means that these patients probably had a full-blown, active case of Ménière's disease that was not in the early stages.

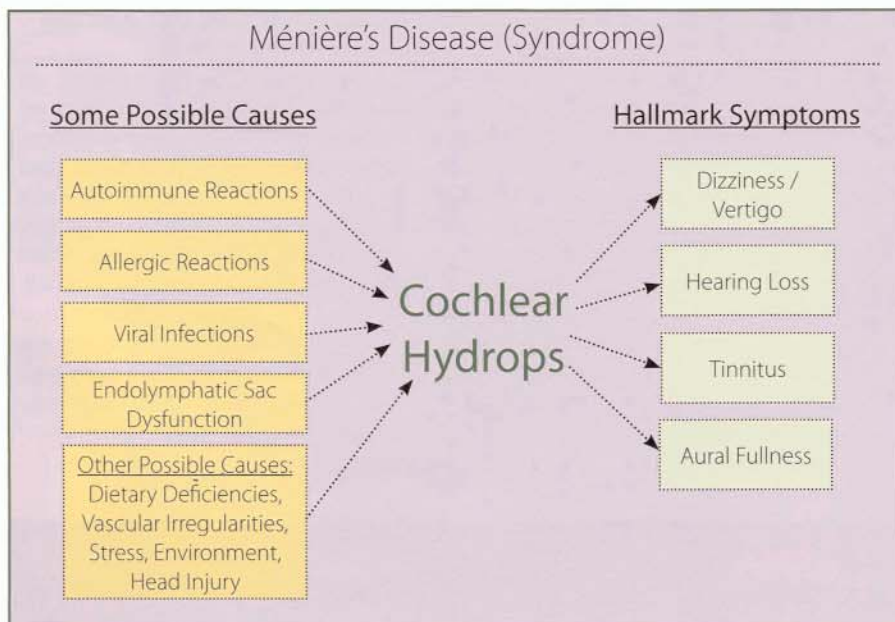
The findings of Dr. Don's study that led to his diagnostic test for Ménière's disease are encouraging. Dr. Don's ABR diagnostic test is able to distinguish active Ménière's disease in individuals already diagnosed with Ménière's. The diagnostic test also shows promise

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for tracking changes in the severity of the disease caused by progression or treatment. Many physicians are now using this test prospectively to identify Ménière's disease in cases where patients may not have a history of the traditional symptoms associated with Ménière's but have some symptoms that point towards Ménière's or cochlear hydrops. The test may also be utilized as a clinical screening to detect the underlying cause of Ménière's symptoms, that is cochlear hydrops, even for patients whose symptoms from Ménière's may have subsided.

"The real goal of this diagnostic test is to identify the presence of cochlear hydrops, the underlying cause of Ménière's symptoms, which are triggered by the physiological changes made by cochlear hydrops to the basilar membrane," says Manny Don, Ph.D. "The importance of identifying cochlear hydrops is that it brings us closer to developing treatments for the underlying cause of Ménière's rather than simply providing relief of ongoing symptoms." ❖