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AJNR Am J Neuroradiol 2020, 41 (11) 1964-1965

doi: <https://doi.org/10.3174/ajnr.A6818>

<http://www.ajnr.org/content/41/11/1964>

This information is current as
of May 17, 2025.

Imaging for Menière Disease

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Menière disease is an inner ear problem that manifests with the symptoms of ear fullness, hearing loss, tinnitus, and vertigo attacks. Typically, the fullness, hearing loss, and tinnitus manifest first and are followed by vertigo attacks. The typical episode of vertigo lasts a few hours and is usually accompanied by nausea and vomiting but may persist for days. Menière disease is most commonly unilateral but can affect patients in both ears. The pathognomonic anatomic finding in Menière disease relates to increased endolymphatic fluid volume resulting in distension of the Reissner membrane into the scala vestibuli of the cochlea. Presently, endolymphatic hydrops can only be confirmed with postmortem examination of temporal bone specimens.

Thus, the diagnosis of definite Menière disease is based on clinical criteria and requires the observation of an episodic vertigo syndrome associated with low- to midfrequency sensorineural hearing loss and fluctuating aural symptoms (hearing loss, tinnitus, and/or fullness) in the affected ear. Probable Menière disease is a broader concept defined by episodic vestibular symptoms (vertigo or dizziness) associated with fluctuating aural symptoms occurring for 20 minutes to 24 hours.¹ With astute history-taking and clinical observation, Menière disease can be differentiated from other inner ear or neurologic disorders that cause similar symptoms.

As neurologists with a large practice in a metropolitan area, we treat many patients with endolymphatic hydrops. We have often requested MR imaging of the cerebellar pontine angle when we are contemplating a more aggressive intervention in patients who do not obtain relief with medical intervention, such as endolymphatic sac decompression, vestibular nerve section, and labyrinthectomy. We request imaging to rule out other pathologies on the affected and the contralateral sides. We do not consider imaging to be absolutely necessary for the initial diagnosis of Meniere disease.

Other physiologic tests have been used to evaluate and diagnose the patient with Menière disease. The audiogram is important because Menière disease preferentially affects the lower and middle frequencies and is one of the only inner ear pathologies to cause fluctuating low-frequency sensorineural hearing loss. With progression of disease, the patient may also lose high-frequency hearing and even completely lose hearing. Electrocochleography (ECoG) has been used as a diagnostic tool. Clinical evaluation of cochlear function using ECoG has focused on the amplitude ratio of the summating potential (SP) and action potential (AP). An elevated SP/AP ratio is thought to reflect endolymphatic hydrops in patients with suspected Menière disease. Results from a survey of otologist and neurotologist professional society members suggested low clinical utility of ECoG in diagnosis/management of Menière disease. For approximately half of respondents, ECoG had no role in their

clinical practice. ECoG was used routinely by only 1 in 6 respondents. Those who used ECoG differed widely in electrode placement and type of stimulus paradigm used.² Vestibular-evoked myogenic potentials have also been investigated as a means of diagnosing Menière disease, but their clinical accuracy and utility are not evident at this time.³

Imaging of the inner ear can now detect endolymphatic hydrops.⁴ Different techniques such as delayed imaging after intravenous administration of a double dose of gadolinium and imaging after intratympanic gadolinium administration have been used. Several studies, including one of our own, have validated a strong correlation with a symptomatic ear and positive imaging findings of dilated endolymphatic spaces in the inner ear.⁵

Although no cost analysis has been performed evaluating MR imaging in the diagnosis of Menière disease, its use certainly incurs higher costs. As stated, these studies can be performed with either intratympanic (IT) or IV injection of gadolinium. The IT route requires an individual qualified in injecting into the middle ear through the tympanic membrane. Furthermore, the MR imaging is performed 24 hours after the IT injection, involving 2 hospital or clinic visits. If bilateral Menière disease is suspected, the patient requires bilateral injections. There are risks to IT injections of any sort, including further hearing loss, tympanic membrane perforation, and infection. The IV route is less labor-intensive; however, patients are still required to wait 4–6 hours before MR imaging.

Other inner ear findings may mimic Meniere symptoms, such as a dural-based tumor adjacent to the endolymphatic sac. Some reports support a high-riding jugular bulb, impinging on the endolymphatic sac, that can be associated with a higher incidence of Menière disease along with other causes of “secondary” endolymphatic hydrops.⁶ In addition, other diagnoses such as vestibular migraine and vestibular neuronitis, for example, may mimic symptoms associated with endolymphatic hydrops. MR imaging shows promise in helping to better differentiate these entities.⁷

However, can imaging absolutely rule out Menière disease? In complex situations in which Workers' Compensation insurance or malpractice litigation are involved, can imaging be of use? Imaging of the inner ear, with the ability to reliably detect dilation of the endolymphatic spaces, might satisfy those who are making legal and financial decisions. Additionally, we do not know the incidence of positive radiographic findings in ears of patients without symptoms. Is an ear with the appearance of hydrops on imaging likely to develop into a symptomatic ear? Can imaging predict the severity of the associated hearing loss or the likelihood of recovery? Can we use imaging to regulate medical management, such as knowing when to stop or reduce a medication that has been effective? Can imaging predict which patient might develop bilateral Menière disease and therefore help determine the aggressiveness of interventions?⁸

In summary, we think that imaging of the inner ear with advanced MR imaging techniques, while interesting, is not essential for the diagnosis and management of the overwhelming majority of patients with Menière disease. There are some questions that might be answered with well-designed clinical studies in

asymptomatic patients, in unilateral symptomatic patients, in patients in whom the diagnosis of Menière disease is questioned, and in patients in whom the cessation of medical therapy is under consideration. Furthermore, patients with so-called “cochlear hydrops” may manifest only auditory symptoms with characteristic low-frequency sensorineural hearing loss but without vertigo. It is unclear whether this symptom complex represents a spectrum of Menière disease with the same histopathologic findings or a distinct entity. There is the potential for imaging to clarify this question. Undoubtedly, high-resolution imaging of the inner ear is of great interest and importance to neurotologists for many other reasons. Pursuing Menière imaging technology might lead eventually to visualizing the spiral ganglion cell count or accurately measuring the cross-sectional area of the cochlear nerve.

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