SHORT ABSTRACT

MRI in Menière’s Disease
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The visualization of the morphologic substrate of Menière’s disease -the endolymphatic hydrops – can be performed using non-contrast as well as contrast-enhanced magnetic resonance imaging techniques. The non-contrast magnetic resonance imaging technique uses a heavily T2-weighted sequence; however, the reproducibility of this technique remains to be confirmed. The contrast-enhanced techniques most frequently use a 3-dimensional fluid-attenuated inversion recovery sequence or a real 3-dimensional inversion recovery sequence either after intratympanic gadolinium administration either 4 hours after intravenous gadolinium administration. The latter is the most frequently used technique and is able to detect definite Menière’s disease with a high sensitivity and specificity. It has been proven to be a reliable technique with a high diagnostic accuracy, enabling the visualization of endolymphatic hydrops.

Keywords: Magnetic Resonance Imaging; Endolymphatic hydrops; Temporal bone disease; Menière’s disease; Classification; Diagnosis

Introduction
Menière’s disease (MD) is a chronic disease with a reported prevalence of 17–513 patients per 100,000 [1]. It is characterized by spontaneous episodic attacks of vertigo, fluctuating low-frequency hearing loss, tinnitus, and aural fullness [1, 2]. More than 150 years ago, Prosper Menière was the first to recognize the inner ear as the site of origin for this disease [3]. Hallpike pointed out the endolymphatic hydrops (EH) as the pathological counterpart for MD [4]. EH is indeed a condition characterized by a distension of the structures filled with endolymph. The cochlear duct, the saccule, the utricle or ampullae contain endolymph. A change in volume of these structures has been proven to correlate with the symptoms of MD [5].

MR Methods for the Visualization of Endolymphatic Hydrops
Recent developments of high-resolution MR imaging of the inner ear have now enabled us to visualize in vivo EH in patients with suspected MD.

The data on the use of non-contrast MR techniques in the evaluation of patients with MD in literature are limited. So far, only two papers have documented the saccule measurement on coronal reformations of a heavily T2-weighted sequence [6, 7]. On these images, the saccule can be detected as a small oval hypo-intense lesion in the vestibule on a coronal reconstruction. Overall, the height of the saccule in patients with MD is reported to be over 1.6 mm. The advantage of this technique is that it requires no contrast administration. The disadvantage is that it only evaluates vestibular hydrops; cochlear hydrops is not evaluated. The reproducibility of measuring such small anatomical structures remains to be confirmed.

The contrast-enhanced hydrops MR imaging essentially exists out of two different contrast techniques, that is intratympanically gadolinium (Gd)-based contrast medium administration and intravenous Gd-based contrast administration with subsequent delayed MR imaging.

In 2007, Nakashima et al reported the clear visualization of EH in patients with MD by an intratympanic injection of a Gd-based contrast medium using a three-dimensional fluid attenuated inversion recovery (3D-FLAIR) sequence on a 3T machine [8]. Intratympanic injection of Gd, however, is considered an off-label use of Gd and moreover, only one ear can be evaluated at a time [8].

The intravenous (IV) administration of Gd has the advantage of being able to evaluate both ears at the same time. Moreover, it is an approved use of Gd [9, 10]. MRI four hours after a double dose of IV Gd administration gives the maximum peri-lymphatic contrast enhancement of both, symptomatic and asymptomatic, ears [9, 10]. To have the highest SNR ratio and to optimize the image quality, a 3 T magnet is required with a dedicated head and neck coil. In most reports, a 3D-FLAIR sequence is used. The disadvantage is that these sequences are time-consuming, so patient immobilization in order to avoid motion degradation is crucial [9, 10].

Diagnostic Imaging Criteria of Menière’s Disease
Various semi-quantitative grading criteria have been proposed. Baráth et al. [11] defined the normal situation as a hardly visible non-enhancing cochlear duct in the enhancing scala vestibuli and scala tympani (Figure 1a).
Grade I cochlear hydrops is defined as a mild dilation of the non-enhancing cochlear duct into the scala vestibuli with partial obstruction of the scala vestibuli (Figure 1b). In grade II cochlear hydrops, the scala vestibuli is uniformly obstructed by the maximally distended cochlear duct (Figure 1c). In the vestibule – in normal cases – one can clearly discriminate the non-enhancing saccule and utricle in the enhancing vestibule. The saccule is the smallest of both structures and is located anterior, inferior, and medial in the vestibule (Figure 2a). A grade 1 vestibular

Figure 1: Cropped axial 3D-FLAIR image of the right ear, four hours after intravenous administration of a double dose of Gd, at the level of the mid turn of the cochlea. (a) Note the clear delineation of the enhancing scala vestibuli and scala tympani (perilymphatic space) with in between the non-enhancing cochlear duct or scala media (endolymphatic space) visible as a thin hypo-intense line (arrowhead): normal findings. (b) The non-enhancing dilated cochlear duct (arrowheads) can be seen as a small non-enhancing nodule bulging into the enhancing scala vestibuli. Cochlear hydrops grade 1 according to the Baráth classification. (c) The enlarged scala media or cochlear duct is completely pushing away the scala vestibuli and can be seen as band-like hypo-intensities (arrowheads) in the mid and apical turn of the cochlea. Cochlear hydrops grade 2 according to the Baráth classification.

Figure 2: Cropped axial 3D FLAIR image of the right ear, four hours after intravenous administration of a double dose of Gd, at the level of the lower part of the vestibule. (a) The saccule (small arrowhead) and utricle (large arrowhead) can nicely be discriminated. There are no signs of a vestibular hydrops. (b) In this case, the saccule (large arrowhead), normally the smallest of the two vestibular sacs, has become equal or larger than the utricle (small arrowhead) but is not yet confluent. In the Baráth classification – using the three-stage grading system – this is regarded as normal. However, this should be regarded as a mild form of vestibular hydrops and should be considered abnormal: vestibular hydrops grade 1 in the four-stage grading system. (c) There is enlargement of the saccule and utricle (arrowhead), which have become confluent but still are surrounded by perilymphatic contrast enhancement (arrow). According to the Baráth classification (three-stage grading system), this is a grade 1 vestibular hydrops. However, using the four-stage grading system, this becomes a grade 2 vestibular hydrops. (d) Note the enlargement and confluence of saccule and utricle, without surrounding contrast (arrowhead). There is only some contrast visible in the base of the posterior semicircular canal (arrow). In the Baráth classification – using the three-stage grading system – this is considered a grade 2 vestibular hydrops. Using the four-stage grading system, this becomes a grade 3 vestibular hydrops.
hydrops presents as a distention of the endolymphatic space of the saccule, utricle, or both, with the enhancing perilymphatic space still visible along the periphery of the bony vestibule (Figure 2c) [11]. In a grade II vestibular hydrops, the saccule and utricle are extremely distended without any visible surrounding enhancing perilymphatic space (Figure 2d). By using this technique and classification, Baráth et al found a high interobserver agreement [11]. Ninety percent of clinically diseased ears had EH on MR imaging, whereas 78% of the clinically normal ears had no EH on MR imaging. However, 22% of clinically normal ears showed EH on MR imaging and 10% of ears with clinical diagnosis of MD did not show EH [11].

In a more recent study, the semi-quantitative grading system was questioned as the authors noted that saccular abnormalities were much more frequent than those of the utricle on temporal bone sections [12]. They proposed the inversion of the saccule to utricle ratio (SURI) on an oblique sagittal section as a marker of EH. SURI was only found in patients with MD (50%) and was felt to be a more reliable approach than conventional semi-quantitative methods for distinguishing subjects with MD from healthy subjects [12].

Our own data of a recent study [13] with delayed IV Gd-enhanced 3D-FLAIR MR imaging on 148 patients (296 ears) also confirm that adding an extra low-grade vestibular hydrops to the Baráth classification – in which the saccule, normally the smallest of the vestibular sacs, has become equal or larger than the utricle but not yet confluent – raises the sensitivity without loss of specificity for the diagnosis of definite MD (Figure 2b). By adding this extra low-grade vestibular hydrops, the classification for vestibular hydrops goes from a three-stage grading system [11] to a four-stage grading system resulting in a higher accuracy of MR imaging in detecting MD [13].

It has already been described that the disruption of the blood-perilymph barrier results in an asymmetrical enhancement of the membranous labyrinth in patients with MD [14, 15]. The enhancement in patients with MD is more pronounced on the affected side [14, 15]. Our study [13] confirms this asymmetrical perilymphatic enhancement (PE) in patients with MD (Figure 3). Moreover, our data show that adding this PE criterium to the EH criteria augments the specificity while maintaining the sensitivity [13].

In conclusion, delayed IV Gd-enhanced MR technique is the most frequently used technique and is able to visualize and grade endolymphatic hydrops in patients with definite MD with a high sensitivity and specificity. A four-stage EH grading system in combination with PE assessment gives the best diagnostic accuracy.

Competing Interests
The author has no competing interests to declare.

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