Magnetic Resonance Imaging and the endolymphatic hydrops in Menière's disease

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Abstract
Background. Over the last decades, continuous efforts to improve the understanding of endolymphatic hydrops (EH) have progressively led to the integration of magnetic resonance imaging (MRI) into the diagnostic work-up of inner ear disorders. In recent years, imaging has gained increasing relevance in patients affected by Menière's disease (MD), as MRI enables in vivo visualization of fluid compartment alterations. Nevertheless, several critical issues remain unresolved, including technical limitations of MRI protocols, inter-observer variability in EH assessment, heterogeneity of hydrops distribution, and the lack of a clear prognostic or etiopathogenetic interpretation of imaging findings.
Methods. A systematic review of the literature was performed using PubMed, EMBASE, Web of Science, and Scopus. Eligible studies were English-language articles investigating the use of MRI for the assessment of EH in the inner ear. Study selection and data extraction were conducted according to PRISMA guidelines.
Results. A total of 455 articles were identified, with the earliest publication dating back to 1992. The present review focuses on the most recent evidence, including studies published from January 1st, 202, reflecting advances in imaging techniques and clinical applications. Overall, the included studies consistently reported a high capability of MRI to identify EH and related inner ear abnormalities.
Conclusions. Recent literature provides a confirmation of the clinical utility of MRI in MD. Key emerging topics include the evaluation of blood–labyrinth barrier (BLB) dysfunction, the extent and grading of EH, and their correlations with clinical symptoms, audiological test outcomes, as well as disease stage and progression. MRI appears to represent a valuable adjunctive tool in the clinical assessment of MD.

Keywords*: Menière's disease, endolymphatic hydrops, cochlear hydrops, vestibular hydrops, blood-labyrinthine barrier (BLB), Magnetic resonance imaging, diagnosis, classification.*

Introduction

Ménière's disease (MD) is characterised by episodic vertigo, fluctuating hearing loss, and tinnitus. The prevalence of MD in the United States is estimated at 190 per 100,000, with a female-to-male ratio of 1.89:1, and its incidence increases with age (Alexander TH, 2010). In 1861, Prosper Ménière reported that this triad of symptoms reflected an inner ear lesion (Baloh RW, 2001), which was later associated with endolymphatic hydrops (EH) nearly a century ago (Hallpike CS, 1938; Yamakawa K, 1938). It eventually became understood that EH should be considered a histological marker for MD rather than the direct cause of its symptoms (Merchant SN, 2005).

In the cochlea, EH typically presents as a distension of Reissner's membrane into the scala vestibuli (SV) (Kimura RS, 1995; Salt AN, 1995). Displacements of the bas-

ilar membrane in the apical segments of the cochlea have also been reported in some patients (Nageris B, 1996; Xenellis JE, 2004). The saccule, utricle, and ampullae of the semicircular canals can also be displaced to varying degrees based on the mechanical compliance of the inner ear's membranous components. Consequently, EH occurs more frequently in the cochlear duct, saccule, and utricle—which possess weaker membranes—and less frequently in the semicircular canals, which are structurally more resistant to hydrops (Kimura RS, 1976; Salt AN, 1995a, 1995b). In 2013, Foster et al. suggested that EH is unlikely to be a mere epiphenomenon of MD, as every temporal bone specimen with MD exhibited EH (Foster CA, 2013).

Historically, imaging studies were primarily used to exclude retrocochlear disorders, such as vestibular schwannomas. In the 1990s, Magnetic Resonance Imaging (MRI) techniques were proposed to study EH in humans (Tanioka H, 1992), and these methods were later improved using Gadolinium (Gd) contrast (Fitzgerald DC, 1996). Although image-based diagnosis of EH was considered key to understanding fluctuating sensorineural hearing loss, imaging was not fully established in patients until 2007, when Nakashima visualized EH via intratympanic (IT) Gd administration 24 hours post-injection (Nakashima T, 2007). This method utilized T2-weighted scans with a 3D Fluid-Attenuated Inversion Recovery (3D-FLAIR) algorithm to decrease the endolymph signal relative to the adjacent perilymph.

While IT-Gd injection provides detailed EH visualization through perilymph subtraction (Wesseler A, 2019) using low doses, the technique has limitations. Most notably, the puncture is limited to the affected ear, leaving the contralateral ear uninvestigated; furthermore, the procedure is considered off-label in many countries, including Italy. Additionally, individual differences in round window membrane permeability can affect results (Yoshioka M, 2009). Consequently, the intravenous (IV) route for Gd has been proposed (Naganawa S, 2010). This method is less invasive, offers comparable resolution, and reduces operating times (approximately the highest peak of perilymph enhancement is at 4 hours for a comprehensive study). IV injection involves a weight-based dose of Gd (0.1–0.2 ml/kg) that diffuses into the perilymph via the blood-labyrinth barrier (BLB) as well as to the fluid within the anterior portion of the eye, the subarachnoid space surrounding the optic nerve, Meckel's cave, and the fundus of the internal auditory canal but does not enter the endolymph (Tagaya M, 2011; Conte G, 2018a). IV administration is now widely preferred due to its feasibility and its similarity to standard MRI protocols used to exclude retrocochlear pathology.

Regarding contrast agents, most reports utilise gadodiamide or gadoteridol, with no significant differences in perilymph contrast observed after four hours (Sano R, 2012; Naganawa S, 2013, 2014). Non-contrast visualization of EH has also been described in specific cases, such as haemorrhage or vestibular schwannoma, where high protein content or compression provides natural perilymphatic contrast (Naganawa S, 2009; Conte G, 2018b).

The first grading system for EH, proposed by Nakashima et al., divided EH into three levels: none, mild, and significant (Nakashima T, 2009). According to these criteria, a mild EH is within 34–50 % of the ratio of the endolymphatic area over the vestibular scala (sum of the endolymphatic and perilymphatic area) and significant EH covers the ratio of more than 50 % in the vestibule. Although it graded the cochlea and vestibule separately, it lacked precise definitions for the slice levels of evaluation. Subsequently, other subjective classifications (Gurkov R, 2011; Liu F, 2011) and hallmarks (Attyé A, 2017; Conte G, 2018c) were proposed until the current classification was established in 2019 (Bernaerts A, 2019). This scale assesses cochlear and vestibular hydrops separately based on the endolymph-to-perilymph ratio.

For the cochlear duct, the scale identifies three grades:

- **Grade 0:** Normal/no EH; the scala media is not detectable, and the interscalar septum is recognizable.
- **Grade I:** The scala media is visible with a reduction of the SV.
- **Grade II:** The SV is completely occupied by the scala media.

For vestibular hydrops, the Bernaerts scale offers increased sensitivity for utricular involvement:

- **Normal:** The saccule and utricle are visible separately and occupy less than half of the vestibular surface.
- **Grade I:** The saccule (normally the smaller sac) is equal to or larger than the utricle but not yet confluent.
- **Grade II:** Confluence of the saccule and utricle is observed, though a peripheral rim of perilymphatic enhancement remains visible.
- **Grade III:** Perilymphatic enhancement is no longer visible. An utricular protrusion into the non-ampullated part of the lateral semicircular canal may also be present.

In 2021, we reviewed the literature on EH imaging in MD and concluded that MRI with 3D-FLAIR sequences and delayed (4-hour) Gd uptake was a promising tool. We emphasised the need for further investigation into the prevalence and role of EH in various inner ear disorders, as well as the relevance of other findings such as BLB breakdown, methaemoglobin, and inflammatory deposits in the endolymphatic space (Zanetti D, 2021). MRI-detected EH correlates strongly with the symptomatic side in MD, allowing for the confirmation of "certain" EH in "definite" MD cases—a diagnosis that was previously only possible post-mortem. The AAO-HNS (1995) criteria, in fact, includes EH as landmark to define certain MD. As Gurkov (2016) noted, a causal relationship is highly probable given the consistency of the findings. Furthermore, literature indicates that the severity of EH on MRI correlates with the degree of hearing loss (Kahn, 2020), that it is useful for identifying "probable" MD (Gurkov R, 2022), and can differentiate EH compartments using both 3T and 1.5T MRI (Naganawa S, 2014; Gurkov R, 2016). Even if iv Gd- 4 hour MRI have been considered a crucial procedure in the audiological workflow in MD since 2013, (Pyykkö I, 2013), the reported studies vary regarding Gd administration, acquisition timing, and resolution. Since the interest in this field has grown significantly, with over 200 publications released since our last review, we aim to review the current literature to better define advances in imaging analysis and methodology, evaluate its value as a diagnostic tool for MD, and clarify the role of EH imaging in clinical decision-making.

Methods

Literature search strategy

According to the PRISMA guidelines (Moher D, 2009), the scientific literature from the last 5 years was reviewed to identify studies focusing on the use of MR for the analysis of MD and EH in the inner ear.

Multiple combinations of "hydrops" AND "magnetic resonance" AND "inner ear" of the following search queries were used in Medline, Cochrane, Scopus and Clarivate database (from 1st January 2022 to Present). The search was restricted to articles that provided at least an abstract in English. Only human studies were retrieved. References of the selected publications were also examined to extract any further relevant articles. The retrieved articles were considered eligible if they provided results of MR in MD patients with EH. Two of the co-Authors (FDB and VMDPF) separately reviewed all articles and excluded those with unclear clinical diagnostic criteria.

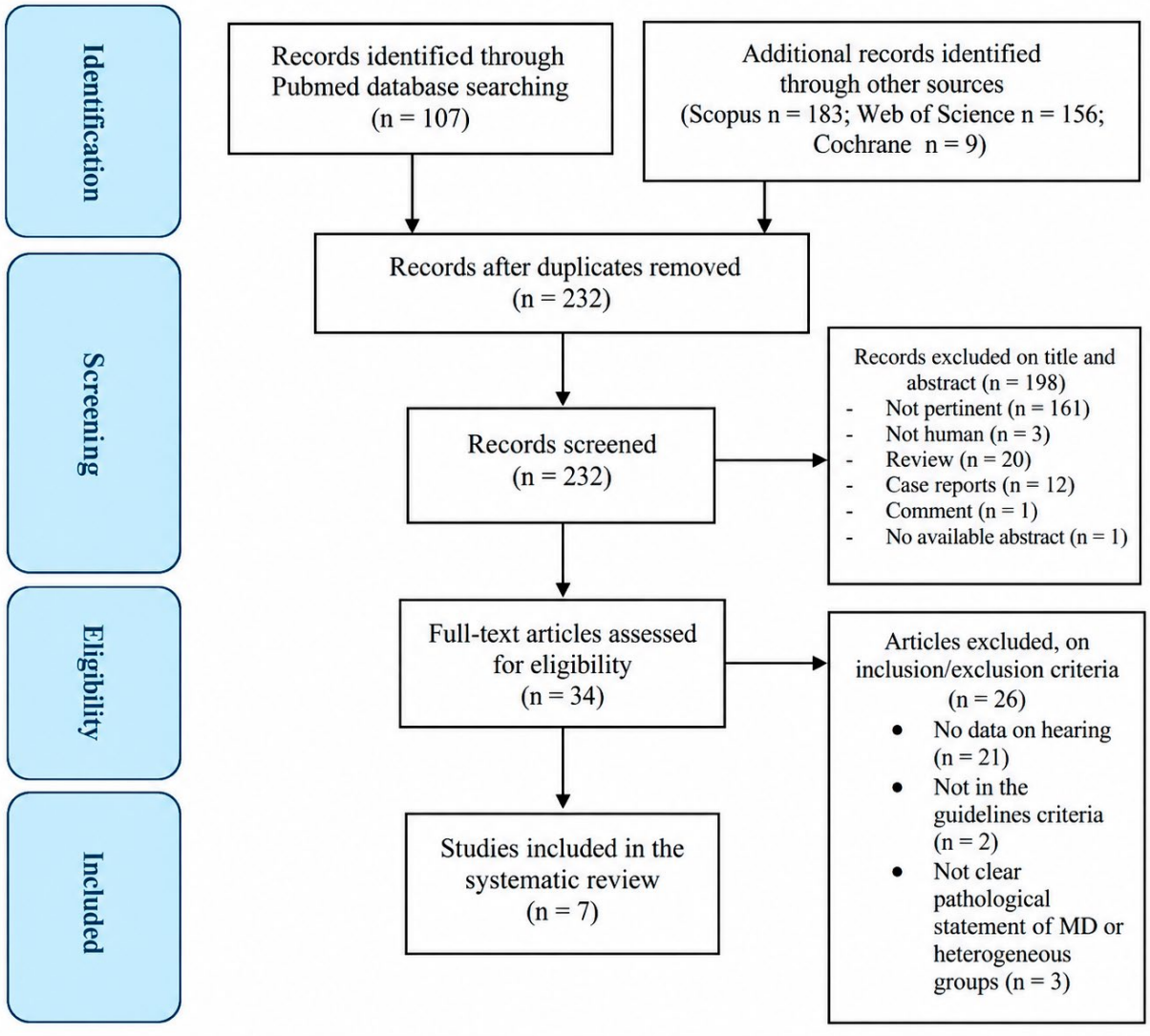
Other exclusion criteria were studies on animals, case reports, personal (expert) opinions, meta-analyses, different diagnostic criteria, studies where diagnosis of MD was lacking; studies not assessing hydrops by accepted MRI techniques. The following information were outsourced from the selected studies: first author, year of publication, total number of subjects enrolled,

MRI techniques and peculiar MRI findings related with the presence of hydrops in the affected vs. unaffected ears.

Literature Search Results

The initial search yielded a total of 455 articles: 107 articles in PubMed, 156 in Web of Science, 183 in Scopus and 9 in the Cochrane Library. After excluding duplicates and those not respecting the inclusion criteria, 232 articles were included by relevance. (Figure 1). 20 of them were review studies and were only considered for the general overview of the topic and 9 were excluded by the exclusion criteria.

Figure 1. The PRISMA flow chart diagram reporting the inclusion process of the studies considered in the present review.



Results

Administration of contrast

Out of 232 screened studies, 27 performed an IT administration of Gd, 205 only an IV delivery and 3 studies both were per-

formed simultaneously. Table 1 reports all the papers reporting data on the audiological condition of MD evaluated in the considered studies.

The studies summarised in Table 1 collectively point out a strong and consistent association between MRI-detected EH and auditory dysfunction in patients with MD. Despite heterogeneity in imaging protocols, study design, and grading systems, most investigations converge on the conclusion that both cochlear and vestibular hydrops are clinically meaningful biomarkers of hearing impairment. Across large retrospective cohorts (Zhao, Yin, Liu, Xiao), EH severity showed a robust positive correlation with PTA and a clear dose–response relationship between hydrops grade and hearing thresholds. Liu et al. provided one of the most compelling quantitative demonstrations, showing progressive worsening of PTA from grade 0 to grade 3 hydrops in both the cochlea (38 → 68 dB) and vestibule (37 → 61 dB), and further documenting that ears with combined cochlear and vestibular hydrops had the poorest hearing outcomes. Similarly, Yin et al. found that both cochlear and vestibular EH were independent predictors of hearing loss across low, mid, and high frequencies, reinforcing the notion that EH is not only linked to low-frequency loss—the classical audiometric hallmark of early MD—but also to pan-tonal deterioration as the disease progresses. Xiao et al. corroborated this finding by demonstrating frequency-wide correlations between hydrops degree and hearing thresholds in a large cohort evaluated with delayed 3D-FLAIR imaging. Prospective data support these observations. Teggi et al. reported very high detection rates of EH (86% vestibular, 90% cochlear) and showed that the degree of cochlear hydrops correlated with hearing level, with more advanced EH associated with pantonal and bilateral audiometric patterns. Abdulla et al. likewise demonstrated a positive correlation between both vestibular and cochlear EH and hearing loss, further validating the clinical relevance of MRI-based hydrops assessment. An important outlier is the study by Lin et al., which found no correlation between cochlear hydrops grade and PTA. This discrepancy likely reflects methodological differences, including the small sample size (n = 25), the use of HYDROPS-Mi2 imaging limited to the cochlea, and the relatively narrow distribution of PTA values between hydrops grades. These factors may have reduced statistical power and sensitivity to detect a true association. Taken together, the evidence indicates that MRI-detected EH, particularly when involving both the cochlea and vestibule, is strongly associated with the severity and extent of hearing loss in Ménière's disease. The consistent dose–response relationship between hydrops grade and PTA across multiple large cohorts supports the concept that EH is not merely an epiphenomenon, but a structural correlate of cochlear dysfunction and disease progression.

Discussion

These findings reinforce the role of delayed post-contrast inner ear MRI as a valuable tool for phenotyping MD and stratifying patients according to the burden of inner ear pathology. MRI performed with intravenous gadolinium injection and delayed imaging at 4 hours has been confirmed as the gold standard for the non-invasive visualization of endolymphatic hydrops.

Patients are generally willing to undergo this examination, as it confirms the clinical diagnosis and allows assessment of the contralateral ear.

MRI technique and EH grading

3-Tesla MRI and IV rout of administration of Gd remained the preferred choice. The imaging protocol has been refined to improve contrast-to-noise ratio (CNR) and signal-to-noise ratio (SNR) using 3D-FLAIR and optimized 3D real inversion recovery (IR) sequences (Zhao M., 2023). Recent articles have confirmed the previous reports (SURI, VESCO parameters etc) reporting the relevance of a quantitative volumetric measurement of vestibular and cochlear EH and

Table 1. Article included in the systematic review

#	1	2			3		4		5		6	7		
First author	Zhao	Teggi			Abdulla		Yin		Liu		Lin	Xiao		
Year	2025	2025			2025		2025		2024		2023	2022		
Country	China	Italy			Turkey		China		United Kingdom		Taiwan	China		
Type of study	Retrospective	Prospective			Retrospective		Retrospective		Retrospective		Prospective	Retrospective		
Sample size	155	44			49		47		154		25	97		
MRI sequences	3D-real IR	3D-FLAIR			3D-real IR		3D SPACE FLAIR		3D-real IR		HYDROPS-Mi2	3D FLAIR		
Contrast delay time	4-5 h	4-5 h			4 h		4 h		4 h		4 h	24 h		
Analyzed area	Vestibule	Vestibule	Cochlea		Vestibule	Cochlea	Vestibule	Cochlea	Vestibule	Cochlea	Cochlea	Vestibule	Cochlea	
EH		Vestibular 86%	Cochlear 90%		49 (100%)		Vestibular: grade 1, 28 (59.6%); grade 2, 12 (25.5%); grade 3, 3 (6.4%).	Cochlear: grade 1, 21 (44.7%); grade 2 27 (55.3%)	Vestibular: grade 1, 29; grade 2, 85; grade 3, 13.	Cochlear: grade 1, 66; grade 2, 67; grade 3, 8.	100%	Vestibule: 22 (22.7%)	Cochlea: 3 (3.1%)	Cochlea and vestibule: 34 (34.1%)
PTA	55.6 (24.5)*	-			-		PTA:41.0 (33.8,59.0)*		Vestibular hydrops degree: grade 0, 37 dB; grade 1, 40 dB; grade 2, 53 dB; grade 3, 61 dB.	Cochlear hydrops degree: grade 0, 38 dB; grade 1, 43 dB; grade 2, 52 dB; grade 3, 68 dB.	Grade I, 51 ± 19 dB; grade II, 53 ± 19 dB.	45.7 (18.6)*		
Type of hearing loss	-	12 unilateral, low-frequency	20 unilateral, pantonal	10 bilateral, pantonal	-		-		-		-	-		
Tinnitus	113 (60%)	-			-		-		-		-	-		
Aural fullness	169 (89%)	-			-		-		-		-	-		
Comment	Degree of EH correlates with PTA (p<0.001)	Degree of cochlear EH correlates with hearing level.			Degree of cochlear and vestibular EH positively correlates with hearing level.		Cochlear and vestibular hydrop is significant predictor of hearing loss in low, mid and high frequencies.		the level of cochlear and vestibular hydrops positively correlates with the hearing levels. Ear with both vestibular and cochlear hydrops have worse hearing levels.		Cochlar hydrops degree does not correlate with PTA.	Hearing level at low, mid and high freqeuncy positively correlates with inner ear hydrops degree.		

not only a qualitative analysis. (Suárez-Vega V, 2023) However, the major advancement has been the introduction of the Barnaetz (Bernaerts) classification, which has gained universal acceptance and is now the most frequently cited grading system in the literature.

Recent studies have explored the role of MRI as an adjunct diagnostic tool in patients with probable Ménière's disease, demonstrating that endolymphatic hydrops can be visualized on delayed MRI, thereby supporting earlier diagnosis and management.

MRI findings

The most intriguing aspect is the emergence of studies investigating different MRI patterns of endolymphatic hydrops and attempting to determine whether the extent or anatomical distribution of EH may have clinical relevance. Initial evidence suggests that evaluation of blood–labyrinth barrier (BLB) alterations may provide indirect information on acute disease phases or disease progression of a vestibular deficit. A prospective study published in 2023 demonstrated that patients experiencing acute vertigo attacks or hearing deterioration showed more pronounced perilymphatic enhancement compared with patients in remission, suggesting that delayed MRI is able to detect more symptomatic cases (Suzuki M., 2023). These findings are clinically significant, as they may help guide therapeutic strategies, particularly in identifying candidates for targeted intratympanic therapy.

Another very recent study (Vaudelin C., 2025) demonstrated that, in patients with unilateral acute vestibular syndrome, detection of blood–labyrinth barrier (BLB) impairment on delayed MRI was associated with poorer recovery on the video head impulse test, suggesting that BLB enhancement may serve as a predictor of vestibular recovery. (Eliezer M, 2019)

Some authors have previously demonstrated that 3D-FLAIR sequences obtained 4 hours after double-dose intravenous gadolinium administration can reveal enhancement of the vestibular nerve, vestibule, and semicircular canals, suggesting that this finding reflects gadolinium distribution within the perilymphatic space of the vestibule and early vestibular nerve inflammation (Byun H., 2018).

Conversely, it should be noted that similar enhancement may also be observed on conventional traditional MRI sequences (not delayed), particularly in the presence of increased protein levels in the inner ear, due to various causes, (Conte G., 2019) or in the haemorrhagic conditions (T1 sequences). Instead, an impairment of the BLB in the cochlea, as evaluated with 3D-FLAIR sequences performed 4 h after an IV administration, provides a good correlation with the results of neurotologic evaluation, determined by the extent of vestibular deficit, timing of imaging acquisition, and possibly by the underlying etiology (Kim KT, 2025).

BLB modifications have also observed to be associated with EH in MD cases; BLB alterations in MD occour with an higher frequencies than in sudden sensorineural hearing loss and have been detected also in the contralateral ear, inducing to think that BLB modification might predict a progression to bilateral of MD and confirming the hypothesis of a cause of systemic origin of MD. (Tagaya M, 2011; Pakdaman MN, 2016)

EH detection differentiating vestibular migraine and MD

Patients with vestibular migraine exhibited EH far less frequently than patients with MD (32% vs. 80%, respectively). In the few vestibular migraine cases showing EH, which were bilateral, no differences in EH grade were observed between the two ears; the EH was mainly involving the vestibule, and was generally of very low grade. In contrast, patients with MD showed a clear predominance of EH in one ear—typically the more severely or previously affected ear—with EH in the whole intralabyrinthine spaces but predominantly involving the cochlea (Kirsch V, 2025).

In addition, integrating intravenous contrast–enhanced MRI findings—beyond EH grading alone—with audiological data appears to better characterize cochlear pa-

thology, thereby enabling the development of tailored therapeutic strategies. A very recent study by Yin et al. (2025) demonstrated that MRI can reliably differentiate vestibular migraine from MD, confirming a significantly higher and unilateral EH in MD, along with greater signal asymmetry measured by the signal intensity ratio of the basal turn of the cochlea and the cochlear signal intensity ratio asymmetry index between the two ears. While contrast enhancement correlates with the degree of hearing loss, the combined assessment of EH and increased signal intensity achieves excellent diagnostic accuracy in MD (area-under-the-curve = 0.954).

In some cases, isolated saccular EH has also been identified; although its clinical significance remains debated, this condition may be associated with the full spectrum of MD, suggesting that saccular hydrops could represent the first manifestation of MD. (Maxwell AK, 2021)

Conclusion

Further studies are required to better characterize different MRI patterns of EH, including its anatomical distribution—such as selective involvement of the utricle and/or saccule—herniation into the semicircular canals, non-visualization of the saccule, alterations of the endolymphatic sac, and early or delayed BLB dysfunction, this last item has been better clarified by the last articles.

Future research should focus on imaging and their correlations with clinical symptoms, audiological outcomes, and not only on hearing and vestibular impairment, disease stage or progression.

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