

Magnetic resonance imaging of the inner ear in the diagnostics of Ménière's disease

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Data Interpretation
E – Manuscript Preparation
F – Literature Search
G – Funds Collection

Agnieszka Jasińska^{1ABDEF}, Magdalena Lachowska^{1AEF}, Emilia Wnuk^{2ABD}, Kazimierz Niemczyk^{1E}

¹Department of Otorhinolaryngology, Head and Neck Surgery, Medical University of Warsaw, Poland;

Head: Prof. Kazimierz Niemczyk MD PhD

²^{2nd} Department of Clinical Radiology, Medical University of Warsaw, Poland; Head: Prof. Olgierd Rowiński MD PhD

Article history: Received: 18.12.2020 Accepted: 18.12.2020 Published: 28.12.2020

ABSTRACT:

Ménière's disease is characterized by sudden episodes of vertigo accompanied by tinnitus and/or feeling of fullness in the ear as well as fluctuating sensorineural hearing loss. Despite numerous studies, the etiology of this disease remains unknown. However, the enlargement of the inner ear's endolymphatic spaces, referred to as endolymphatic hydrops, is considered the underlying condition. Thanks to recent advances in magnetic resonance (MR) technology, it is now possible to obtain in vivo imaging of endolymphatic hydrops in patients presenting with Ménière's disease symptoms. Visualization of the inner ear fluid compartments is achieved after gadolinium contrast is administered into the tympanic cavity or via the intravenous route. Evaluation of endolymphatic hydrops is possible as the contrast agent selectively penetrates the perilymph, and endolymph is visualized as contrast defects. The currently used radiological hydrops grading systems include qualitative, semi-quantitative, and volumetric scales. The methods are subject to ongoing modifications to increase their sensitivity and specificity. Numerous studies describe correlations between clinical symptoms and audiological and otoneurological examination results with the endolymphatic hydrops grade. MRI is also applicable in patients' diagnostics with an incomplete or atypical course of the Ménière's disease. In the course of the treatment, follow-up MRI scans enable assessing individual treatment modalities' efficacy in terms of the severity of lesions and the further course of the disease within the inner ear.

KEYWORDS:

endolymphatic hydrops, hearing loss, magnetic resonance, Ménière's disease, vertigo

ABBREVIATIONS

AAO-HNS – American Academy of Otolaryngology-Head and Neck Surgery

AP – action potential amplitude

CVEMP – cervical vestibular evoked myogenic potentials

MR – magnetic resonance

OVEMP – ocular vestibular evoked myogenic potentials

SP – summating potential amplitude

SURI – saccule to utricle ratio inversion

VNG – videonystagmography

INTRODUCTION

Ménière's disease's clinical presentation consists of vertigo episodes with nausea and vomiting, fluctuating low-frequency hearing loss, tinnitus, and/or feeling of congestion within the ear [1–3]. The natural history of the disease varies between individuals and includes periods of exacerbations and spontaneous remission. Ménière's disease affects patients of all ages, with incidence peaking in the fifth and the sixth decade of life [4]. In most cases, symptoms are unilateral at the time of diagnosis. Still, the risk of symptoms within the contralateral ear increases with the disease's duration and affects 2–73% of patients, according to literature sources [5–9]. Ménière's disease's etiology remains unknown, with autoimmune processes, infections, allergies, cardiovascular disorders, chronic middle and inner ear

diseases being proposed as potential causes [5, 10–12]. In 5 to 15% of cases, the disease is inherited within families following an autosomal dominant pattern with incomplete penetrance [13].

Ménière's disease has been a subject of interest for researchers for more than 150 years since Prosper Ménière first linked the presence of vertigo to a pathology of the inner ear [14]. More than 70 years later, Hallpike and Cairns [15] and Yamakawa [16] analyzed the temporal bones of patients presenting with symptoms corresponding to Ménière's disease to observe the enlargement of endolymphatic spaces within the inner ears of these patients. This presentation was subsequently termed the endolymphatic hydrops.

For many years, endolymphatic hydrops has been considered the morphological origin of Ménière's disease symptoms, although confirmation was possible only in post-mortem histopathological examinations [1].

The introduction of MRI offered new possibilities for the diagnostics of endolymphatic hydrops. Previously, MRI was used in patients presenting with Ménière's disease symptoms to exclude other vertigo causes, such as nerve VIII tumors. In 2007, Nakashima et al. [17] were the first to observe that gadolinium contrast, when introduced into the tympanic membrane, is accumulated in perilymphatic spaces. A similar effect was observed later by Naganawa et al. [18] following intravenous administration of a double dose of the contrast agent.

Tab. I. Semi-quantitative MR-based scale for endolymphatic hydrops grading according to Nakashima et al. [19] and Naganawa et al. [18].

ENDOLYMPHATIC HYDROPS GRADING SCALE ACCORDING TO NAKASHIMA AND NAGANAWA	
Cochlea	
Healthy condition	No cochlear duct enlargement
Moderate grade	Cochlear duct enlargement Cochlear duct narrower than the scala vestibuli
Significant grade	Enlarged cochlear duct extending into the scala vestibuli
Vestibule (Endolymphatic space vs vestibular fluid space area ratio)	
Healthy condition	≤33.3%
Moderate grade	>33.3% but ≤50%
Significant grade	>50%

Tab. II. Morphological assessment of endolymphatic hydrops in magnetic resonance (MR) imaging scans based on the SURI value proposed by Attyé et al. [21]. *SURI – saccule to utricle ratio inversion.

MORPHOLOGICAL ASSESSMENT OF ENDOLYMPHATIC HYDROPS IN MRI SCANS AS PROPOSED BY ATTYÉ ET AL.		
Hydrops grade	Saccule	SURI*
Grade 0	not enlarged	<1
Grade 1	enlarged	≥1
Grade 2	invisible	not applicable

Tab. III. Descriptive MR-based scale for endolymphatic hydrops grading according to Barath et al. [22] as modified by Bernaerts et al. [23].

COCHLEA – THE ASSESSMENT OF ENDOLYMPHATIC HYDROPS IN MRI SCANS		
Barath scale (identical to Bernaerts scale)	Hydrops presentation in MRI	
Healthy condition	No cochlear duct enlargement	
Grade I	Partial enlargement of the cochlear duct, visible obstruction of scala vestibuli	
Grade II	Enlarged cochlear duct extending into the vestibular duct	
Vestibule – the assessment of endolymphatic hydrops in MRI scans		
Barath scale	Bernaerts scale	Hydrops presentation in MRI
Healthy condition	Healthy condition	Saccule and utricle not enlarged, saccule smaller than the utricle
	Grade I	Saccule larger than the utricle, boundary between the two is maintained
Grade I	Grade II	Enlargement and overlapping of saccule and utricle, peripheral contrast enhancement
Grade II	Grade III	significant enlargement of saccule and utricle, no contrast enhancement within the vestibule

MRI PROTOCOL IN THE DIAGNOSTICS OF ENDOLYMPHATIC HYDROPS

The most common modality for the imaging of inner ear fluid compartments consists of T2-3D fluid-attenuated inversion recovery (FLAIR) images requiring previous administration of a contrast agent. The evaluation is based on the finding that the contrast agent selectively penetrates the perilymph while the endolymph remains uncontrasted. Gadolinium contrasts may be administered via the transtympanic [17] and intravenous [18] routes. When administered via the transtympanic route, the contrast agent penetrates the inner ear in the round window and the oval window region and across the medial walls' continuity. It is estimated that the accumulation of gadolinium within the perilymphatic space takes approximately 24 hours. If a double dose of the gadolinium agent is administered intravenously, scans can be acquired as early

as after 4 hours. Other benefits of intravenous contrast administration include assessing both inner ears simultaneously and avoiding the puncturing of the tympanic membrane.

METHODS FOR EVALUATION OF ENDOLYMPHATIC HYDROPS IN MAGNETIC RESONANCE SCANS

Endolymphatic hydrops is observed as enlargement of endolymphatic spaces presenting as contrast defects against contrast-enhanced perilymph. Since the first reports on MR's use in Ménière's disease's diagnostics, numerous authors have proposed various scales for endolymphatic hydrops grading. The currently used radiological hydrops grading systems include qualitative, semi-quantitative, and volumetric scales. The selected classifications are summarized in Tab. I.–III.

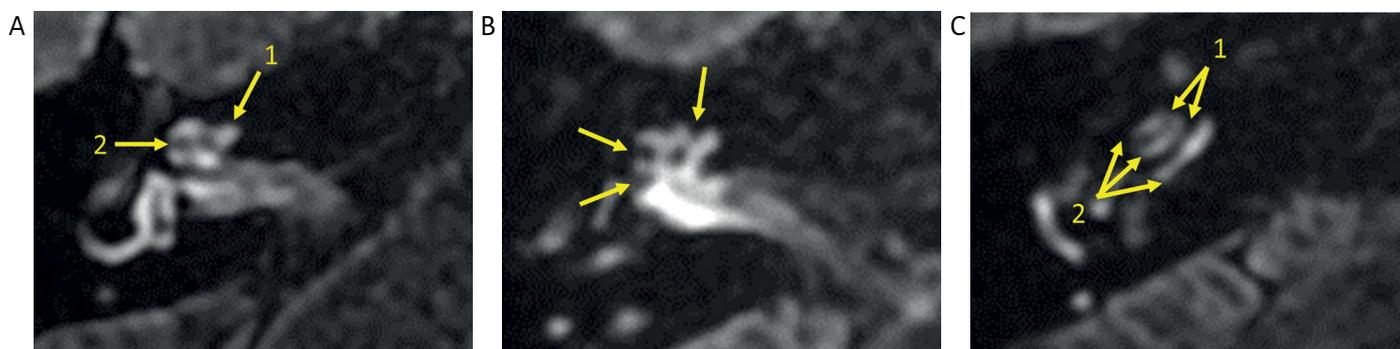


Fig. 1. MRI of the inner ear—a 3D-FLAIR T2-weighted sequence performed 4 hours after intravenous administration of a double dose of the contrast agent—assessment of the cochlear hydrops grade, according to Barath et al. [22] and Bernaerts et al. [23]. (A) The typical image of the cochlea without enlargement of endolymphatic spaces—arrow #1 points to contrast-enhanced scala vestibuli and scala tympani, arrow #2 points to the osseous spiral lamina and the narrow cochlear duct visible as a slight defect in contrast enhancement; (B) grade I cochlear hydrops—arrows point to partial enlargement of the cochlear duct, obstruction of scala vestibuli is also visible; (C) grade II cochlear hydrops—arrows #1 point to the cochlear hydrops, significantly enlarged along the entire length and extending into the scala vestibuli area; arrows #2 point to the contrast-enhanced scala tympani.

The semi-quantitative scale described by Nakashima et al. [19] in 2009 (Tab. I.) is commonly used for the grading of endolymphatic hydrops visualized in MRI scans. For this purpose, a single cross-sectional image of the inner ear is analyzed. Within the cochlea, the assessment is based on the contrast defects within the scala vestibuli corresponding to the cochlear duct being enlarged due to the Reissner's membrane dislocation. Moderate-grade endolymphatic hydrops appears enlarged but still narrower than the scala vestibuli. Significant-grade endolymphatic hydrops is characterized by the cochlear duct being wider than the scala vestibuli. The assessment consists of comparing the contrast-defective area of endolymph to the total area or endo- and perilymph within the vestibule. Endolymph accounting for <33% of the vestibular area is considered normal, whereas moderate endolymphatic hydrops is diagnosed for endolymph accounting for 33–50% of the total area. When the non-enhanced area accounts for >50% of the total vestibular area, the endolymphatic hydrops is considered advanced.

In 2018, Conte et al. [20] published a review of studies using the grading scale proposed by Nakashima. Significant differences in sensitivity and specificity of endolymphatic hydrops detection were observed for the method used in 17 studies analyzed in the review. Patients with Ménière's disease symptoms presented with endolymphatic hydrops in 36–100% of symptomatic ears and 46% of asymptomatic ears. On the other hand, in healthy volunteers, cochlear duct enlargement was observed in 13–33% of subjects. Vestibular location of moderate-to-significant endolymphatic hydrops was reported for 94–100% of symptomatic ears in patients with Ménière's disease, albeit some studies reported its also being present in 53–100% of asymptomatic ears as well as in healthy subjects. Attyé et al. [21] analyzed results obtained for patients with Ménière's disease in 30 symptomatic ears and 30 healthy ears. They reported that endolymphatic hydrops was present within the cochlea and/or the vestibule in 100% of cases. For this reason, Attyé et al. [21] undertook a detailed analysis of vestibular MRI images. They proposed a new parameter, referred to as the saccule to utricle ratio inversion (SURI), determined using a single reference sagittal cross-section (Tab. II.). Under physiological conditions, the saccule is smaller than the utricle, and the SURI is < 1. The authors of the study found that 50% of

Ménière's disease patients presented with SURI of ≥ 1 , whereas elevated values of the parameter were absent from the control group (specificity of 100%).

As proposed by Barath et al. [22] in 2014, the three-grade scale evaluates the enlargement of endolymphatic spaces separately within the cochlea and the vestibule (Tab. III., Figs. 1. and 2.). For the cochlea, grade 0 corresponds to no cochlear enlargement, grade I corresponds to partial enlargement of the cochlear duct and visible obstruction of scala vestibuli. In contrast, grade II corresponds to Reissner's membrane's significant dislocation, making it impossible to visualize the perilymph within the scala vestibuli. Similarly, concerning the vestibule, grade 0 corresponds to no enlargement of the saccule or utricle, grade I corresponds to the enlargement and overlapping of the saccule and utricle with peripheral contrast enhancement. In contrast, grade II corresponds to no contrast enhancement being detected within the vestibule as its area is fully encompassed by endolymphatic space. The authors analyzed the results of the MRI scans in 53 patients with symptoms indicative of the Ménière's disease and found the presence of endolymphatic hydrops in 90% of symptomatic ears, with the sensitivity of the method being as high as 95% when only the group of patients with the definite disease was included in the analysis. Endolymphatic hydrops was also detected in 22% of asymptomatic, contralateral ears.

In 2019, Bernaerts et al. [23] modified Barath's scale by adding a „lower” grade I where the saccule is greater or equal in size to the utricle, but the boundary between the two is still visible (Tab. III., Figs. 1. and 2.). Thus, in the Bernaerts modification, the scale for the assessment of vestibular hydrops is four-grade. In addition, the authors compared the degree of contrast enhancement within the cochlea on the symptomatic side and the contralateral side. They reported an asymmetrical increase in contrast enhancement to be characteristic for symptomatic ears (specificity of 97.4%). The algorithm for the MRI assessments of the inner ear, as proposed in this study, assumes that the higher enhancement of the cochlea on the symptomatic side is equivalent to the presence of endolymphatic hydrops. The reported results suggest the involvement of the blood-perilymph barrier disorders in the pathogenesis of Ménière's disease.

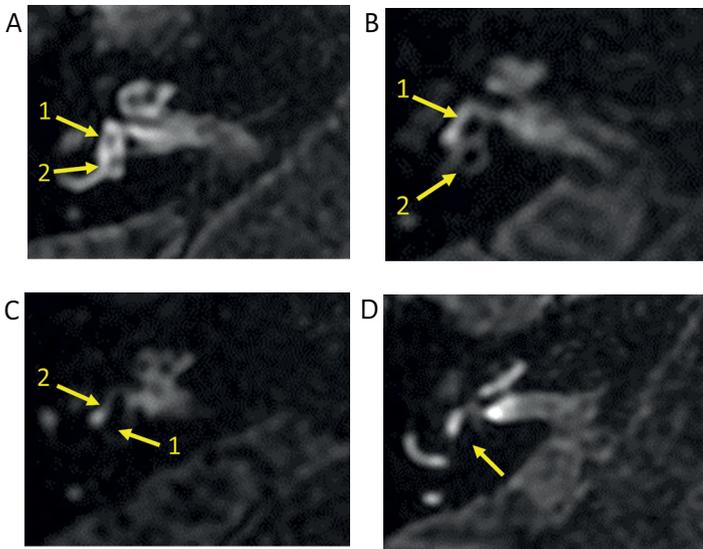


Fig. 2. MRI of the inner ear – a 3D-FLAIR T2-weighted sequence performed 4 hours after intravenous administration of a double dose of the contrast agent. Assessment of the vestibular hydrops grade according to Barath et al. [22] and including modification by Bernaerts et al. [23]. (A) normal vestibule – arrow #1 points to non-enlarged saccule and arrow #2 points to the utricle; the border between the two is maintained and visible; (B) vestibular hydrops of the lowest grade I according to the Bernaerts modification (within normal limits according to Barath) – the vestibule presents with enlarged saccule marked with arrow #1 and utricle marked with arrow #2; the boundary between the two is maintained, and marginal contrast enhancement of the vestibule can be seen; (C) vestibular hydrops of Barath grade I (Bernaerts grade II) – dilatation and overlapping of the saccule and the utricle is marked with arrow #1, peripheral contrast enhancement of the vestibule is marked with arrow #2; (D) vestibular hydrops of Barath grade II (Bernaerts grade III) – the significant enlargement and overlapping of the saccule and utricle is marked with the arrow; no contrast enhancement within the vestibule.

In 2019, Kahn et al. [24] proposed a classification based on the labyrinth's anatomical division where endolymphatic spaces of the saccule, utricle, and ampullae of semicircular canals were assessed separately. Saccular hydrops was detected in 91% of symptomatic ears; in 97% (i.e., all but one) cases in this group, saccular hydrops was accompanied by cochlear hydrops. Utricular hydrops was observed in 45% of symptomatic ears and always co-existed with the involvement of the saccule and the cochlea while the enlargement of semicircular canal ampullae was observed in as few as 8.5% of ears presenting with the symptoms of Ménière's disease; all these cases were associated with significant grades of cochlear, saccular, and utricular hydrops. The above results' analysis confirms the theory suggesting that hydrops develops initially within the cochlear duct to extend sequentially into the saccule, utricle, and semicircular canals [25].

Accurate assessment of the endolymphatic spaces in patients presenting with Ménière's disease symptoms can be achieved using volumetric methods. Gurkov et al. [26] assessed the percentage volumes of endolymphatic spaces within the cochlea and the vestibule of 16 patients. They observed a significant correlation between the enlargement of the cochlear tube and the degree of hearing impairment. The comparison of endolymphatic space volumes in patients with Ménière's disease and healthy volunteers carried out by Ito et al. [27] has proved important. It facilitated the outcomes being referred to the standard. The authors evaluated the total volume of spaces filled with endolymph and

perilymph and determined the percentage share of endolymphatic space within the inner ear. In symptomatic ears, endolymph was found to occupy a significantly larger space than that in the ears of healthy volunteers and the asymptomatic ears of patients with Ménière's disease. Homann et al. [28] compared the volumetric method with the widespread semi-quantitative scale proposed by Nakashima et al. [29]. The authors also took note of the time needed to perform the analysis using both methods, stating that for the semi-quantitative assessment, a single evaluation lasted an average of 2.2 minutes, while 14.5 minutes were required for volumetric analysis. In this study, the volume of endolymphatic space in the group of 11 patients with Ménière's disease correlated with the degree of hearing loss; it was also significantly higher in patients with a longer history of the disease. The volumetric analysis results are consistent with the grade of endolymphatic hydrops as determined using the semi-quantitative scale, making it possible to consider the Nakashima and Naganawa method a reliable and rapid tool for the assessment of endolymphatic hydrops.

CORRELATION BETWEEN THE MR IMAGES OF ENDOLYMPHATIC HYDROPS AND THE CLINICAL PRESENTATION AND AUDIOLOGICAL AND OTONEUROLOGICAL EXAMINATION RESULTS

Ménière's disease's natural history is characterized by exacerbation periods and spontaneous remissions, the frequency of which varies from one case to another. The disease's fluctuating course is reflected in the inner ear imaging results showing variable grades of endolymphatic hydrops in repeated scans acquired in the same patient [29, 30].

Most of the studies carried out in recent years found no link between the endolymphatic hydrops grade and the disease's duration [24, 31, 32]. At the same time, only a few reported the existence of statistically significant correlations [23].

Numerous research papers describe the correlation between the pure-tone audiometry thresholds and the endolymphatic hydrops grade assessed from MR scans [24, 32–35]. This correlation may be crucial for a better understanding of processes developing within the inner ear and demonstrates the impact of endolymphatic space enlargement on hearing impairment.

The relationship between the endolymphatic hydrops grade and electrocochleography examination results was also analyzed. Seo et al. [34] divided their patients' population with established Ménière's disease into groups presenting with endolymphatic hydrops visualized in MRI scans vs. unremarkable MR images of the cochlea to observe a statistically significant difference in the summing to action potential ratios (SP/AP). Yamamoto et al. [36] carried out a similar study and reported the differences in the SP/AP values between groups of patients with different grades of endolymphatic hydrops. However, these differences were only statistically significant when patients with advanced cochlear hydrops were compared to patients with moderate hydrops or unremarkable MRI scans.

Examinations of the balance system and organs were analyzed in correlation with the images of corresponding structures. Caloric test responses obtained in the videonystagmographic (VNG) examination of the labyrinth was assessed in correlation with hydrops grades determined from MRI scans. Cho et al. [3] described the correlations between the reduction in labyrinthine excitability and endolymphatic space enlargement. They also observed that the caloric deficit was significantly higher in endolymphatic hydrops encompassing the horizontal semicircular canal's ampulla. Other studies revealed no significant correlation between the grade of endolymphatic hydrops and VNG results [32, 38]. Cervical and ocular vestibular evoked myogenic potentials (oVEMP and cVEMP) are used to assess the saccule's and the utricle's function, respectively [39–41]. Hence, the results of these examinations were examined for correlations with otolithic organ enlargement. Kahn et al. [24] and Seo et al. [34] failed to identify any significant correlation between the grade of endolymphatic hydrops within the saccule and the utricle and the oVEMP/cVEMP results. Gurkov et al. [33] reported a significant correlation between the grade of vestibular hydrops and reduced response amplitude in the cVEMP study.

MAGNETIC RESONANCE IN THE DIAGNOSTICS OF ATYPICAL COURSE OF MÉNIÈRE'S DISEASE

In addition to patients with the classical clinical presentation of Ménière disease, many patients with isolated symptoms such as periodic tinnitus, feeling of congestion within the ear, or recurring low-frequency sensorineural hearing loss with no vertigo [42]. Such a clinical presentation had been previously referred to as the cochlear form of the disease; however, the term has not been included in the current AAO-HNS guidelines [1, 3, 43], which state that at least two episodes of vertigo are required for the diagnosis of Ménière's disease.

The first study in which MRI scans were performed as part of the diagnostic protocol of endolymphatic hydrops had been carried out in patients without a history of typical vertigo and was published in 2009 by Teranishi et al. [44]. The authors demonstrated hydrops' presence within the cochlea and the vestibule in all eight patients, with fluctuating sensorineural hearing loss detected within the low-frequency range or across all audiometric frequencies.

Yoshida et al. [31] performed MRI scans in a group of patients with tinnitus as the main complaint. As in the study mentioned above by Teranishi et al. [44], the patients had no history of vertigo. The authors reported enlargement of the cochlear duct in 56% of symptomatic ears, with the grade of endolymphatic hydrops being significantly correlated with the presence of fluctuating tinnitus and the feeling of congestion within the ear.

The method has also found its use in patients with sudden idiopathic low-frequency sensorineural hearing loss. In a study by Shimono et al. [45], cochlear hydrops was detected in 92% of symptomatic ears following intratympanic administration of gadolinium contrast. It is worth noting that the enlargement of

the endolymphatic space among people affected by the sudden low-frequency hearing loss was also observed within the vestibule, with vestibular hydrops developing in 88% of cases.

The above literature data show that the inner ear's MRI examinations facilitate diagnostics of patients with incomplete clinical presentation, including the predominance of audiological symptoms such as tinnitus or the feeling of congestion within the ear. The natural history of the Ménière disease should be considered, as these symptoms often precede the emergence of vertigo and evolution into a fully symptomatic form of the disease [46].

Cochlear and vestibular hydrops in patients with sudden low-frequency hearing loss are suggestive of the common pathogenesis of isolated sudden low-frequency hearing loss and fully symptomatic Ménière's disease MRI scans of the inner ear are likely to provide an opportunity for quicker diagnosis of endolymphatic hydrops in patients with early symptoms of the Ménière's disease.

A small percentage of patients with Ménière's disease experience transient improvement in hearing during vertigo episodes. Such a clinical presentation is referred to as the Lermoyez syndrome and is observed in about 0.2% of patients with Ménière's disease, although some researchers consider it a separate disease [47]. Zhou et al. [48] reported endolymphatic hydrops in all nine patients with Lermoyez syndrome in their study, which suggests that endolymphatic hydrops is involved in the pathogenesis of this clinical form of the disease. Importantly, compared with the results of patients with classic Ménière's disease, the grade of vestibular and cochlear hydrops was similar and significantly lower, respectively, in patients with Lermoyez syndrome [35]. It suggests that the variability of clinical presentations between individual subtypes of Ménière's disease manifests differences in endolymphatic hydrops' grades within individual inner ear structures.

THE ROLE OF MAGNETIC RESONANCE IN THE EVALUATION OF MÉNIÈRE'S DISEASE TREATMENT EFFICACY

The current guidelines for the management of Ménière's disease suggest an escalation approach. The treatment starts with dietary recommendations, lifestyle modification, and pharmacotherapy, while the next step consists of transtympanic treatment, i.e., steroids of aminoglycoside antibiotics being administered into the tympanic cavity. Upon no improvement, surgical methods are recommended in patients with persistent vertigo, including endolymphatic sac surgery and vestibular nerve section [1, 3].

Evaluation of the efficacy of the treatment of Ménière's disease is carried out mainly to reduce the incidence and severity of vertigo, which is considered the most burdensome disease symptom. However, the progressive nature of the disorder should be taken into account. It may lead to permanent damage to the organ of hearing and balance due to a continued increase in the endolymphatic volume. The use of MRI offers the possibility of assessing the hydrops' dynamics and thus control the pathological process within the inner ear.

Suga et al. analyzed the changes in the endolymphatic hydrops grades following conservative treatment [49]. They acquired MRI scans from a group of 12 patients with suspected Ménière's disease and then repeated the scans after 10 to 76 months. In the group of patients who reported subjective improvement, the hydrops grade was reduced in two out of three ears. At the same time, hydrops remission was observed in only one out of 17 ears, for which no resolution of symptoms was reported. In addition, Gurkov et al. [50] acquired MRI scans of six patients who had received beta-histidine at a dose of 2×24 mg for 3–7 months. The authors reported no changes in the grade of endolymphatic hydrops as a result of the treatment.

In a study by Fiorino et al. [51], the impact of transtympanic gentamycin on the degree of endolymphatic space enlargement was assessed in 8 patients. In follow-up MRI scans acquired 3–12 months after the baseline, no inner ear presentation changes were observed in four patients, whereas the other four patients presented with hydrops progression.

Literature reports are also available on MRI in the postoperative assessment of patients having undergone interventional procedures within the endolymphatic hydrops. Higashi-Shingai et al. [52] carried out an MRI study in 21 patients with Ménière's disease, who were subsequently qualified for endolymphatic sac drainage. A follow-up study performed two years after the procedure showed a significant reduction in vestibular hydrops' grade with no significant changes observed within the cochlea. Furthermore, the authors concluded that the regression of hydrops was not correlated with clinical improvement regarding the prevalence of

vertigo, reducing the hearing threshold, or electrocochleographic examination results. Ito et al. [53] carried out a similar study in which they compared the hydrops' grade in 20 patients undergoing endolymphatic sac drainage procedures with intraoperative steroid administration. Two years after the procedure, the follow-up study revealed a link between the decrease in the hydrops' grade as assessed using the MRI and the degree of vertigo resolution.

The available literature presents only the preliminary results from studies conducted on small groups of patients with diverse disease durations and symptoms severities. In most of the above mentioned studies, the period between the baseline examination and the follow-up was as short as few months. Given the progressive, often multiannual natural history of Ménière's disease, long-term follow-up and scheduling of follow-up MRI scans in patients subjected to treatment may be of crucial importance.

SUMMARY

Despite many studies, the etiology of Ménière's disease remains unknown. The MRI examination of the inner ear offers the possibility of performing in vivo evaluation of the grade of endolymphatic hydrops, which is considered the disease's underlying cause. The analysis of correlations between the MRI results, clinical manifestations, and the audiologic and otoneurologic findings may be crucial to understanding the pathogenesis of the disease. In the treatment course, follow-up MRI scans enable the assessment of individual treatment modalities' efficacy in terms of the severity of lesions within the inner ear.

REFERENCES

1. Committee on Hearing and Equilibrium guidelines for the diagnosis and evaluation of therapy in Ménière's disease. American Academy of Otolaryngology-Head and Neck Foundation, Inc. *Otolaryngol-Head Neck Surg.*, 1995; 113(3): 181–185.
2. Goebel J.A.: 2015 Equilibrium Committee Amendment to the 1995 AAO-HNS Guidelines for the Definition of Meniere's Disease. *Otolaryngol Head Neck Surg.*, 2016; 154(3): 403–404.
3. Basura G.J., Adams M.E., Monfared A. et al.: Clinical Practice Guideline: Meniere's Disease. *Otolaryngol Head Neck Surg.*, 2020; 162(2_suppl): S1–S55.
4. Sajjadi H., Paparella M.M.: Meniere's disease. *Lancet.*, 2008; 372(9636): 406–414.
5. Paparella M.M., Djaliliani H.R.: Etiology, pathophysiology of symptoms, and pathogenesis of Meniere's disease. *Otolaryngol Clin North Am.*, 2002; 35(3): 529–545, vi.
6. Noij K.S., Herrmann B.S., Guinan J.J., Jr., Rauch S.D.: Predicting Development of Bilateral Meniere's Disease Based on cVEMP Threshold and Tuning. *Otol Neurotol.*, 2019; 40(10): 1346–1352.
7. Suh M.J., Jeong J., Kim H.J., Jung J., Kim S.H.: Clinical Characteristics of Bilateral Meniere's Disease in a Single Asian Ethnic Group. *Laryngoscope.*, 2019; 129(5): 1191–1196.
8. Huang C.H., Young Y.H.: Bilateral Meniere's disease assessed by an inner ear test battery. *Acta Otolaryngol.*, 2015; 135(3): 233–238.
9. Frejo L., Soto-Varela A., Santos-Perez S. et al.: Clinical Subgroups in Bilateral Meniere Disease. *Front Neurol.*, 2016; 7: 182.
10. Gurkov R., Pyyko I., Zou J., Kentala E.: What is Meniere's disease? A contemporary re-evaluation of endolymphatic hydrops. *J Neurol.*, 2016; 263(Suppl 1): S71–81.
11. Weinreich H.M., Agrawal Y.: The link between allergy and Meniere's disease. *Curr Opin Otolaryngol Head Neck Surg.*, 2014; 22(3): 227–230.
12. Greco A., Gallo A., Fusconi M. et al.: Meniere's disease might be an autoimmune condition? *Autoimmun Rev.*, 2012; 11(10): 731–738.
13. Requena T., Cabrera S., Martin-Sierra C. et al.: Identification of two novel mutations in FAM136A and DTNA genes in autosomal-dominant familial Meniere's disease. *Hum Mol Genet.*, 2015; 24(4): 1119–1126.
14. Ménière P.: Sur une forme de surdité grave dépendant d'une lésion de l'oreille interne. *Gaz Med Paris.*, 1861; 16: 239–240.
15. Hallpike C.S., Cairns H.: Observations on the pathology of Meniere's syndrome. *Proc R Soc Med.*, 1938; 31: 1317–1336.
16. Yamakawa K.: Über pathologische Veränderungen bei einem Menière Kranken. *Otolaryngl Soc Jap.*, 1938; 4: 2310–2312.
17. Nakashima T., Naganawa S., Sugiura M. et al.: Visualization of endolymphatic hydrops in patients with Meniere's disease. *Laryngoscope.*, 2007; 117(3): 415–420.
18. Naganawa S., Nakashima T.: Visualization of endolymphatic hydrops with MR imaging in patients with Meniere's disease and related pathologies: current status of its methods and clinical significance. *Jpn J Radiol.*, 2014; 32(4): 191–204.
19. Nakashima T., Naganawa S., Pyykko I. et al.: Grading of endolymphatic hydrops using magnetic resonance imaging. *Acta Otolaryngol Suppl.*, 2009(560): 5–8.
20. Conte G., Lo Russo F.M., Calloni S.F. et al.: MR imaging of endolymphatic hydrops in Meniere's disease: not all that glitters is gold. *Acta Otorhinolaryngol Ital.*, 2018; 38(4): 369–376.
21. Attye A., Eliezer M., Boudiaf N. et al.: MRI of endolymphatic hydrops in patients with Meniere's disease: a case-controlled study with a simplified classification based on saccular morphology. *Eur Radiol.*, 2017; 27(8): 3138–3146.
22. Barath K., Schuknecht B., Naldi A.M. et al.: Detection and grading of endolymphatic hydrops in Meniere disease using MR imaging. *AJNR Am J Neuroradiol.*, 2014; 35(7): 1387–1392.

23. Bernaerts A., Vanspauwen R., Blavie C. et al.: The value of four stage vestibular hydrops grading and asymmetric perilymphatic enhancement in the diagnosis of Meniere's disease on MRI. *Neuroradiology*, 2019; 61(4): 421–429.
24. Kahn L., Hautefort C., Guichard J.P. et al.: Relationship between video head impulse test, ocular and cervical vestibular evoked myogenic potentials, and compartmental magnetic resonance imaging classification in meniere's disease. *Laryngoscope*, 2020; 130(7): E444–E52.
25. Pender D.J.: Membrane Stress in the Human Labyrinth and Meniere Disease: A Model Analysis. *Int Arch Otorhinolaryngol*, 2015; 19(4): 336–342.
26. Gurkov R., Berman A., Dietrich O. et al.: MR volumetric assessment of endolymphatic hydrops. *Eur Radiol*, 2015; 25(2): 585–595.
27. Ito T., Inui H., Miyasaka T. et al.: Endolymphatic volume in patients with meniere's disease and healthy controls: Three-dimensional analysis with magnetic resonance imaging. *Laryngoscope Investig Otolaryngol*, 2019; 4(6): 653–658.
28. Homann G., Vieth V., Weiss D. et al.: Semi-quantitative vs. volumetric determination of endolymphatic space in Meniere's disease using endolymphatic hydrops 3T-HR-MRI after intravenous gadolinium injection. *PLoS One*, 2015; 10(3): e0120357.
29. Sone M., Naganawa S., Teranishi M. et al.: Changes in endolymphatic hydrops in a patient with Meniere's disease observed using magnetic resonance imaging. *Auris Nasus Larynx*, 2010; 37(2): 220–222.
30. Wu Q., Dai C., Zhao M., Sha Y.: The correlation between symptoms of definite Meniere's disease and endolymphatic hydrops visualized by magnetic resonance imaging. *Laryngoscope*, 2016; 126(4): 974–979.
31. Yoshida T., Sugimoto S., Teranishi M. et al.: Imaging of the endolymphatic space in patients with Meniere's disease. *Auris Nasus Larynx*, 2018; 45(1): 33–38.
32. Zhang W., Xie J., Hui L., Li S., Zhang B.: The Correlation Between Endolymphatic Hydrops and blood-labyrinth barrier Permeability of Meniere Disease. *Ann Otol Rhinol Laryngol*, 2020:3489420964823.
33. Gurkov R., Flatz W., Louza J., Strupp M., Krause E.: In vivo visualization of endolymphatic hydrops in patients with Meniere's disease: correlation with audiovestibular function. *Eur Arch Otorhinolaryngol*, 2011; 268(12): 1743–1748.
34. Seo Y.J., Kim J., Choi J.Y., Lee W.S.: Visualization of endolymphatic hydrops and correlation with audio-vestibular functional testing in patients with definite Meniere's disease. *Auris Nasus Larynx*, 2013; 40(2): 167–172.
35. Shi S., Guo P., Li W., Wang W.: Clinical Features and Endolymphatic Hydrops in Patients With MRI Evidence of Hydrops. *Ann Otol Rhinol Laryngol*, 2019; 128(4): 286–292.
36. Yamamoto M., Teranishi M., Naganawa S. et al.: Relationship between the degree of endolymphatic hydrops and electrocochleography. *Audiol Neurootol*, 2010; 15(4): 254–260.
37. Cho Y.S., Ahn J.M., Choi J.E. et al.: Usefulness of Intravenous Gadolinium Inner Ear MR Imaging in Diagnosis of Meniere's Disease. *Sci Rep*, 2018; 8(1): 17562.
38. Kato M., Sugiura M., Shimono M. et al.: Endolymphatic hydrops revealed by magnetic resonance imaging in patients with atypical Meniere's disease. *Acta Otolaryngol*, 2013; 133(2): 123–129.
39. Rosengren S.M., Govender S., Colebatch J.G.: Ocular and cervical vestibular evoked myogenic potentials produced by air- and bone-conducted stimuli: comparative properties and effects of age. *Clin Neurophysiol*, 2011; 122(11): 2282–2289.
40. Govender S., Rosengren S.M., Colebatch J.G.: Vestibular neuritis has selective effects on air- and bone-conducted cervical and ocular vestibular evoked myogenic potentials. *Clin Neurophysiol*, 2011; 122(6): 1246–1255.
41. Długaczyk J.: Ocular Vestibular Evoked Myogenic Potentials: Where Are We Now? *Otol Neurotol*, 2017; 38(10): e513–e21.
42. Phillips J.S., Murdin L., Rea P., Sutton L.: Clinical Subtyping of Meniere's Disease. *Otolaryngol Head Neck Surg*, 2018; 159(3): 407–409.
43. Lopez-Escamez J.A., Carey J., Chung W.H. et al.: Diagnostic criteria for Meniere's disease. Consensus document of the Barany Society, the Japan Society for Equilibrium Research, the European Academy of Otolology and Neurotology (EAONO), the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) and the Korean Balance Society]. *Acta Otorrinolaringol Esp*, 2016; 67(1): 1–7.
44. Teranishi M., Naganawa S., Katayama N. et al.: Image evaluation of endolymphatic space in fluctuating hearing loss without vertigo. *Eur Arch Otorhinolaryngol*, 2009; 266(12): 1871–1877.
45. Shimono M., Teranishi M., Yoshida T. et al.: Endolymphatic hydrops revealed by magnetic resonance imaging in patients with acute low-tone sensorineural hearing loss. *Otol Neurotol*, 2013; 34(7): 1241–1246.
46. Kitahara M., Takeda T., Yazawa Y., Matsubara H., Kitano H.: Pathophysiology of Meniere's disease and its subvarieties. *Acta Otolaryngol Suppl*, 1984; 406: 52–55.
47. Shen K.C., Young Y.H.: Lermoyez syndrome revisited: 100-year mystery. *Acta Otolaryngol*, 2018; 138(11): 981–986.
48. Zhou F., Shi S., Wang D., Guo P., Wang W.: MR imaging and clinical characteristics of Lermoyez syndrome. *Acta Otolaryngol*, 2020; 140(7): 528–532.
49. Suga K., Kato M., Yoshida T. et al.: Changes in endolymphatic hydrops in patients with Meniere's disease treated conservatively for more than 1 year. *Acta Otolaryngol*, 2015; 135(9): 866–870.
50. Gurkov R., Flatz W., Keeser D. et al.: Effect of standard-dose Betahistine on endolymphatic hydrops: an MRI pilot study. *Eur Arch Otorhinolaryngol*, 2013; 270(4): 1231–1235.
51. Fiorino F., Pizzini F.B., Barbieri F., Beltramello A.: Magnetic resonance imaging fails to show evidence of reduced endolymphatic hydrops in gentamicin treatment of Meniere's disease. *Otol Neurotol*, 2012; 33(4): 629–633.
52. Higashi-Shingai K., Imai T., Okumura T. et al.: Change in endolymphatic hydrops 2 years after endolymphatic sac surgery evaluated by MRI. *Auris Nasus Larynx*, 2019; 46(3): 335–345.
53. Ito T., Inui H., Miyasaka T. et al.: Three-Dimensional Magnetic Resonance Imaging Reveals the Relationship Between the Control of Vertigo and Decreases in Endolymphatic Hydrops After Endolymphatic Sac Drainage With Steroids for Meniere's Disease. *Front Neurol*, 2019; 10: 46.

Word count: 4300 Tables: 3 Figures: 2 References: 53

DOI: 10.5604/01.3001.0014.6176 Table of content: <https://otolaryngologypl.com/issue/13708>

Copyright: Some right reserved: Polish Society of Otorhinolaryngologists Head and Neck Surgeons. Published by Index Copernicus Sp. z o.o.

Competing interests: The authors declare that they have no competing interests.



The content of the journal „Polish Society of Otorhinolaryngologists Head and Neck Surgeons” is circulated on the basis of the Open Access which means free and limitless access to scientific data.



This material is available under the Creative Commons – Attribution-NonCommercial 4.0 International (CC BY-NC 4.0). The full terms of this license are available on: <https://creativecommons.org/licenses/by-nc/4.0/legalcode>

Corresponding author: Asst. Prof. Magdalena Lachowska MD, PhD (ORCID: 0000-0003-3631-2551); Department of Otorhinolaryngology, Head and Neck Surgery, Medical University of Warsaw; Banacha street 1a, 02097 Warsaw, Poland; Phone: +48 22599252; E-mail: mlachowska@wum.edu.pl

Cite this article as: Jasinska A., Lachowska M., Wnuk E., Niemczyk K.: Magnetic resonance imaging of the inner ear in the diagnostics of Ménière's disease; Otolaryngol Pol, 2021: 75 (2): 1-8
