

# Endolymphatic Hydrops, Evaluated Using 3T MRI in Patients with Clinically Confirmed Meniere Disease

## Wodniak śródchłonki potwierdzony przy użyciu 3-teslowego skanera MR u pacjentów z obrazem klinicznym choroby Ménière'a

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### ABSTRACT:

Ménière's disease (MD) is characterised by intermittent episodes of vertigo with fluctuating sensorineural hearing loss, tinnitus and aural fullness. Despite numerous studies, the etiology of this disorder remains poorly understood and thus diagnostic criteria are mainly clinical. Development and progress in magnetic resonance imaging (MRI) techniques, in 2007, has enabled visualisation of endolymphatic hydrops, which was the milestone achievement. We present two cases of patients with clinically confirmed MD with a set of audiological, otoneurological examinations and MRI performed. Structures of the inner ear were visualized using contrast-enhanced MR imaging after a 4-hour delayed intravenous gadolinium administration. The cochlea and vestibule were evaluated separately and classified using Barath grading scale. Endolymphatic hydrops in the clinically affected ears were confirmed using MRI. Due to disabling and intractable MD, a decision about vestibular neurectomy was made.

### KEYWORDS:

endolymphatic hydrops, inner ear magnetic resonance, Ménière's disease

### STRESZCZENIE:

Choroba Ménière'a charakteryzuje się występowaniem napadowych zawrotów głowy z towarzyszącym fluktuacyjnym niedosłuchem odbiorczym, szumami usznyimi oraz uczuciem pełności w uchu. Pomimo wielu badań, etiologia choroby pozostaje nieustalona, a jej diagnoza opiera się w głównej mierze na obrazie klinicznym, który charakteryzuje się dużą zmiennością osobniczą i fluktuacyjnym przebiegiem. Możliwość obrazowania ucha wewnętrznego metodą rezonansu magnetycznego po raz pierwszy została opisana w 2007 roku i okazała się znacznym przełomem w diagnostyce wodniaka endolimfatycznego. Prezentujemy opis dwóch przypadków pacjentów z obrazem klinicznym sugerującym chorobę Ménière'a, u których został wykonany pakiet badań audiologicznych, otoneurologicznych, a także rezonans magnetyczny ucha wewnętrznego. W badaniu obrazowym, struktury przedsionka oraz ślimak zostały ocenione po czterech godzinach od podania podwójnej dawki kontrastu gadolinowego dożylnie za pomocą skali zaproponowanej przez Baratha. U opisywanych chorych potwierdzono obecność wodniaka śródchłonki po stronie objawów klinicznych w badaniu MR. Z uwagi na znaczne zaawansowanie kliniczne choroby, opisywani pacjenci zostali zakwalifikowani do chirurgicznego leczenia poprzez przecięcie nerwu przedsionkowego z dostępu przez dół środkowy czaszki.

**SŁOWA KLUCZOWE:** choroba Ménière'a, rezonans magnetyczny ucha wewnętrznego, wodniak endolimfatyczny

### ABBREVIATIONS

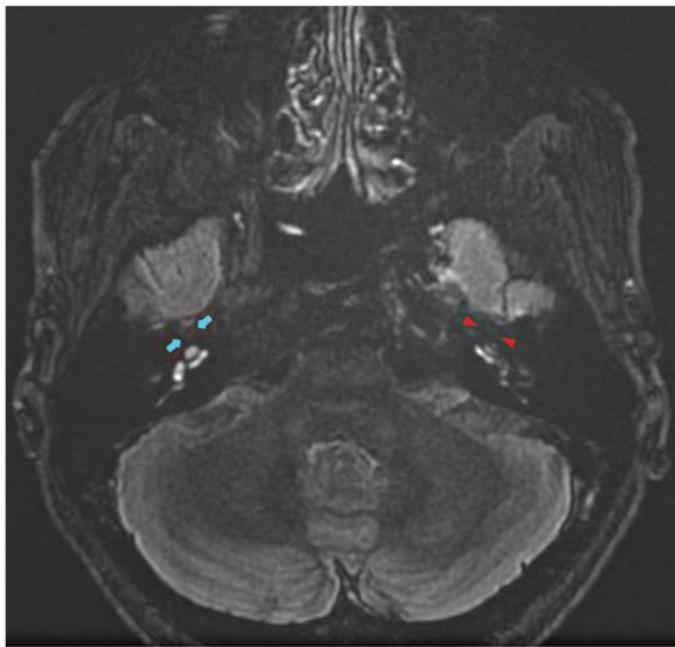
**AAO-HNS** – Committee on Hearing and Balance of the American Academy of Otolaryngology – Head and Neck Surgery

**MRI** – magnetic resonance imaging

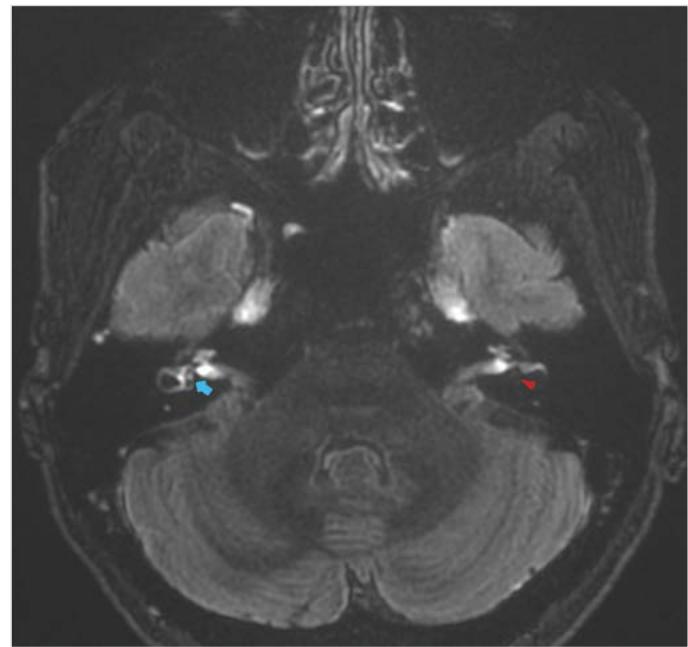
### INTRODUCTION

Prosper Ménière, in his work published in the 'Gazette Médicale de Paris' In 1861, was the first to associate the occurrence of dizziness

with the pathology of the inner ear [1]. By doing that he stood in opposition to the then researchers who associated similar complaints solely with the central nervous system. The triad of symptoms he described, i.e. paroxysmal vortex dizziness, low-frequency hearing loss, and tinnitus or a feeling of fullness in the ear, is referred to as Ménière's disease. The morphological basis of these symptoms was confirmed almost 80 years later by Cairns, Hallpike and Yamakawa, who analysed the temporal bone preparations of patients with a clinical picture corresponding to Ménière's disease and found the presence of distended endolymphatic spaces, subsequently called endolymphatic hydrops [2, 3]. Due to the posthumous nature of the



**Fig. 1.** A 65-year-old woman with suspected Ménière's disease on the left side. Axial cross-section at cochlear level in 3D FLAIR sequence, 4 hours following intravenous administration of a double dose of contrast agent. On the left side, a widened, non-reinforcing cochlear duct is visible, which significantly narrows the vestibular duct. The image corresponds to the grade 2 of cochlear hydrops according to Barath classification (red arrows). On the right side, a picture of properly strengthening cochlear perilymphatic structures with no extension of endolymphatic structures (blue arrows).



**Fig. 2.** A 65-year-old woman with suspected Ménière's disease on the left side. Axial cross-section at vestibular level in 3D FLAIR sequence, 4 hours following intravenous administration of a double dose of contrast agent. On the left side, enlarged utricle and saccule are visible, there is no peripheral atrium reinforcement. The image corresponds to grade 2 of vestibular hydrops according to Barath classification (red arrow). On the right side, the picture of the correct strengthening of the vestibule and the non-reinforcing utricle and saccule (blue arrow).

histopathological examination, which was the only way to confirm endolymphatic hydrops, the identification of Ménière's disease remained a clinical diagnosis based on a typical set of symptoms and documented hearing loss [4]. The fluctuating, individually variable and unpredictable course of the disease impedes the diagnostic process and often delays diagnosis, sometimes by several years. A significant breakthrough turned out to be the possibility of imaging endolymphatic hydrops *in vivo* by employing magnetic resonance imaging (MRI), described in 2007 by Nakashima et al. [5]. They administered gadolinium contrast agent to the tympanic cavity and observed that in the FLAIR (Fluid-attenuated inversion recovery) sequence, the agent migrates to perilymphatic structures while endolymphatic structures remain non-contrasted. In 2014, Naganawa et al. found that the same effect can be obtained by administering a double dose of intravenous contrast agent [6].

The protocols used currently are based on the method proposed by Naganawa, i.e. the assessment is performed in the FLAIR 3D sequence, 4 hours after the administration of a double dose of gadolinium contrast agent intravenously. Ménière's disease is suspected in patients with enlarged endolymphatic structures that do not undergo contrast enhancement, and in this sequence are seen as contrast defects against the background of reinforcing perilymphatic structures. The severity of endolymphatic structures hydrops is assessed using the classification proposed by Barath et al. [7]. In this classification, the severity of endolymphatic structures hydrops of the cochlea and the vestibule is assessed separately. For cochlea, grade 0 means no extension of the cochlear duct, grade 1 entails partially widened cochlear duct with visible narrowing of the vestibular duct, while grade 2 entails a complete extension of the cochlear duct, so that

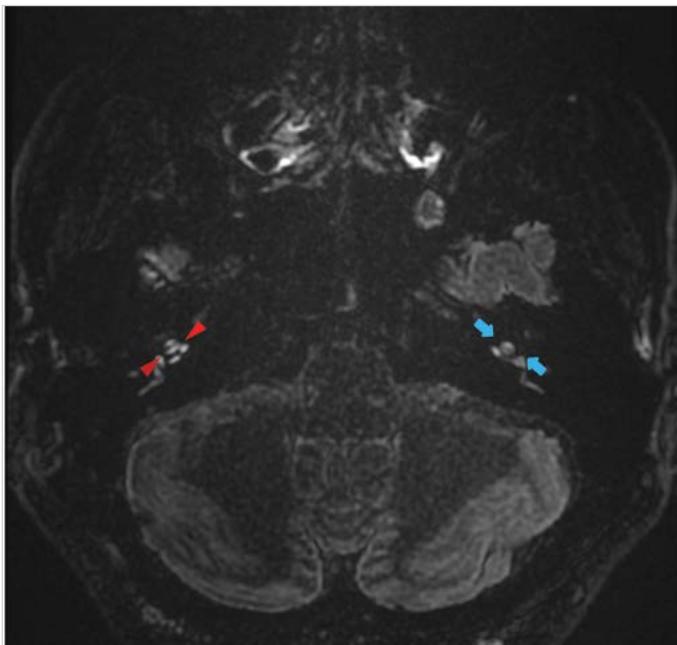
the vestibular duct becomes concealed. Similarly, for the vestibule grade 0 entails lack of dilatation of the utricle and saccule, grade 1 refers to enlargement of the saccule and utricle so that the border between them is not visible and the marginal reinforcement of the vestibule can be observed, and grade 2 entails the extension of the utricle and saccule so significant that the vestibule is not visible.

## CASE DESCRIPTION

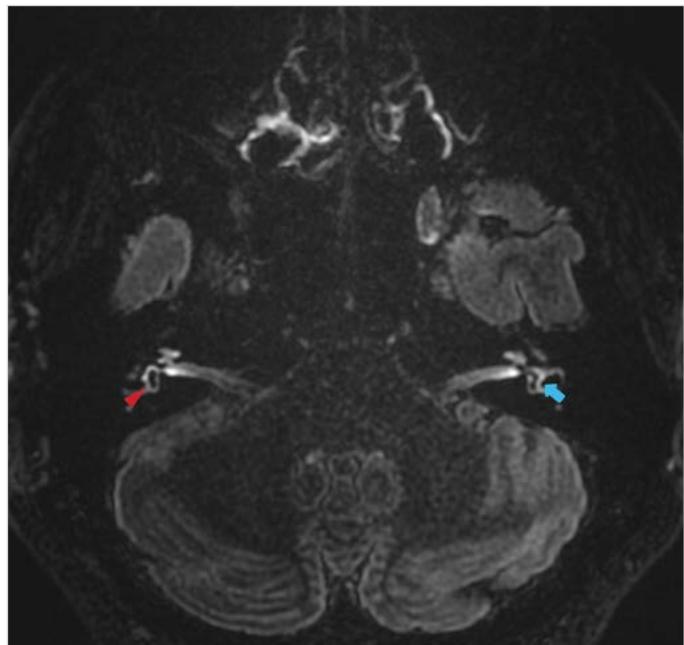
A description of two patients with suspected Ménière's disease was presented; the patients were admitted to the Clinic of Otorhinolaryngology – Head and Neck Surgery at the Medical University of Warsaw for audiological and otoneurological diagnostics. The diagnostic process included: thorough subjective and objective examination, tonal, verbal and impedance audiometry, examination of otoacoustic emissions, brainstem evoked potentials, electrocochleography, vidonistigmography and internal ear imaging using a 3-tesla MRI scanner (GE, SIGNA Architect, Milwaukee, USA). Vestibule and cochlea structures were assessed 4 hours after intravenous administration of a double dose of gadolinium contrast agent (gadobutrol), for which the consent of the bioethics committee and patients was obtained. For the radiological evaluation of the inner ear structures, the classification proposed by Barath was used.

### Case 1

A 65-year-old female patient in whom the first typical attack of dizziness, accompanied by a feeling of fullness in the ear and deterioration of hearing on the left side, occurred 8 years ago. The



**Fig. 3.** A 59-year-old man with suspected Ménière's disease on the right side. Axial cross-section at cochlear level in 3D FLAIR sequence, 4 hours following intravenous administration of a double dose of contrast agent. On the right, a widened, non-reinforcing cochlear duct is visible with a moderate narrowing of the vestibular duct. The image corresponds to grade 1/2 of cochlear hydrops according to Barath classification (red arrows). On the left side, a picture of properly strengthening cochlear perilymphatic structures and no widening of endolymphatic structures (blue arrows).



**Fig. 4.** A 59-year-old man with suspected Ménière's disease on the right side. Axial cross-section at vestibular level in 3D FLAIR sequence, 4 hours following intravenous administration of a double dose of contrast agent. On the right side, the enlargement of saccule and utricle is slightly visible, marginal vestibular strengthening is visible. The image corresponds to grade 0/1 of vestibular hydrops according to Barath classification (red arrow). On the left side a picture of the correct strengthening of the vestibule and the non-reinforcing saccule and utricle (blue arrow).

patient reported low-frequency tinnitus, whose average severity over the past 6 months was 4.75 on a scale from 0 to 6 as proposed by Arenberg, as well as fluctuating, worsening over time deterioration of hearing on the left side. Tonal audiometry indicated a significant degree of pantoneal hearing loss in the left ear. Analysis of the results of the brainstem evoked potential showed a cochlear-like receiving damage in the left side. In the MRI of the inner ear a widened cochlear duct was observed, resulting in complete obliteration of the vestibular waterway (grade 2 cochlear hydrops according to Barath) and a significant degree of enlargement of the utricle and saccule, with non-visible marginal vestibule reinforcement (grade 2 vestibular hydrops) (Fig. 1.–2.). On the right side the image of the inner ear in the imaging examination was normal. The described patient was receiving chronic betahistine treatment at a dose of 24 mg, 2 times a day; attempts were also made to treat her with diuretics and steroids administered transtympanally. In a subjective assessment, the patient determined the severity of symptoms and the impact of ailments on daily functioning to be at grade 4 on a six-point scale proposed by the Committee on Hearing and Balance of the American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNS). Due to the clinical advancement of the disease and the ineffectiveness of previous treatment, based on the analysis of the results of audiological, otoneurological and imaging tests, the patient was qualified for vestibular neurectomy with access through the middle cranial fossa. The perioperative period was without any complications. A follow-up audiological and otoneurological examination is planned, as well as MRI examination of the patient's inner ear after 6 months.

## Case 2

A 59-year-old man with a one-year history suggesting Ménière's disease. The first symptom was a typical attack of dizziness accompanied by a feeling of fullness in the right ear, tinnitus, and subjective hearing impairment on this side. During the first 6 months from the onset of the disease, the patient experienced an average of 6.5 typical seizures per month, followed by over 4 months of spontaneous remission, interrupted by 2 seizures in the last month. On the functional scale, the subjective average severity of fullness in the ear during the 12 months of illness was graded 4 on the Arenberg scale. Tinnitus during this period was rated at 3.45 on a corresponding scale. In the audiological study performed during the asymptomatic period, hearing loss of medium and high tones was found. Records of transtympanic electrocochleography indicated distinct features of the labyrinthine hydrops on the right side.

A partial dilatation of the cochlear duct (grade 1/2) as well as a small enlargement of the utricle and saccule against the background of the strengthening vestibule was observed in an MRI examination (Fig. 3.–4.). Due to the aggressive, rapidly progressing clinical course of the disease and significant impairment of the patient's daily functioning (grade 5 on a 6-point scale), he was qualified for surgical treatment and is currently awaiting vestibular nerve surgery.

## DISCUSSION

Despite many studies conducted to date, the etiology of Ménière's disease remains undetermined and the diagnosis is mainly based

on the clinical picture. It is known, however, that the symptoms are caused by disturbances in hydrostatic and osmotic balance between endothelium and epithelium, which results in the formation of endolymphatic hydrops [8, 9]. The possibility of imaging the widened endolymphatic spaces of the inner ear with the help of 3T resonance imaging is an extremely important, minimally invasive method of confirming the presence of hydrops. Numerous studies confirmed the correlation between the clinical picture and the severity of the disease in the imaging study [10, 11]. There have been reports of patients in whom the features of widening endolymphatic spaces in a clinically healthy ear were detected by magnetic resonance imaging, which suggests the possibility of imaging hydrops before the onset of clinical symptoms [12]. MRI of the inner ear is of particular importance in a group of patients

who do not present a complete clinical picture of Ménière's disease. This is indicated by research confirming the presence of endolymphatic hydrops in an imaging examination in patients with atypical disease [13].

An aggressive course of the disease, with frequent and persistent attacks of dizziness, in the absence of improvement despite conservative treatment and administration of drugs to the tympanic cavity, is an indication for surgical treatment. When qualifying the patient for an effective yet invasive procedure of cutting the vestibular nerve, it may be helpful to confirm the presence of endolymphatic hydrops to be the cause of the symptoms. Inner ear imaging using MRI may also be part of the assessment of the effectiveness of treatment in patients with symptoms of Ménière's disease [14].

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