Head & Face Medicine

RESEARCH

Open Access

The relationship between endolymphatic hydrops features and hearing loss in Bilateral Meniere's disease

Xu Liu[1,2], Dan Wang[1,2], Yue-Lin Hsieh[1,2], Suming Shi[1,2] and Wuqing Wang[1,2*]

Abstract

Background This study aimed to investigate the relationship between the features of endolymphatic hydrops and hearing loss in patients with Bilateral Meniere's Disease.

Methods A retrospective analysis was conducted on 77 patients diagnosed with Bilateral Meniere's Disease. The features of endolymphatic hydrops in the affected ear were evaluated through gadolinium-enhanced inner ear Magnetic resonance imaging. The Spearman correlation coefficient, paired t-tests, and Wilcoxon signed-rank tests were employed for data analysis.

Results The analysis revealed a significant correlation between the degree of endolymphatic hydrops and hearing loss across all frequencies(0.125–8 kHz), including the cochlear, vestibular, and overall degree of endolymphatic hydrops. The strongest correlation between the overall degree of endolymphatic hydrops and hearing loss was observed at low frequencies ($r = 0.571$, $p < 0.05$), followed by mid-frequencies ($r = 0.508$, $p < 0.05$), and high-frequencies ($r = 0.351$, $p < 0.05$), with a correlation of $r = 0.463$, $p < 0.05$ for the staging of Meniere's disease. Affected Ears with endolymphatic hydrops both in the cochlea and vestibule exhibited more severe hearing loss and Meniere's disease staging compared to those with isolated endolymphatic hydrops within the same patient.

Conclusions The features of endolymphatic hydrops in patients with Bilateral Meniere's Disease were found to correlate with the severity of hearing loss and the staging of Meniere's disease.

Keywords Endolymphatic hydrops, Bilateral Meniere's disease, Hearing loss, Gadolinium-enhanced inner ear MRI

Background

Meniere's Disease (MD) is recognized as a prevalent otological disorder, characterized by episodic vertigo, fluctuating hearing loss, tinnitus, and a sensation of aural fullness. Endolymphatic hydrops(EH) has been long identified as the primary pathological feature of MD, though its pathogenesis remained elusive [1, 2]. The prevailing consensus among researchers is that MD constitutes a syndrome initiated by diverse etiologies, necessitating the subdivision of MD into distinct subtypes. This stratification aims to facilitate the investigation into the clinical manifestations and underlying causes of different subtypes, thereby promoting the development of tailored treatment strategies [3, 4]. Unilateral Meniere's Disease (UMD) encompasses the majority of MD cases, with the prevalence of Bilateral Meniere's Disease (BMD) reported variably across studies, ranging from 2 to 78%, most commonly estimated between 20%-30%. Diagnosing BMD requires individual assessment of each ear; however, once MD advanced in one ear, pinpointing the

*Correspondence:
Wuqing Wang
wwuqing@eent.shmu.edu.cn
[1] Department of Otorhinolaryngology, ENT Institute and, Eye & ENT Hospital, Fudan University, Shanghai 200031, China
[2] NHC Key Laboratory of Hearing Medicine (Fudan University), Shanghai 200031, China

ear responsible for vertigo episodes becomes challenging. It is widely accepted among scholars that the diagnosis of BMD can be established upon the manifestation of contralateral ear symptoms such as hearing loss, tinnitus, and ear fullness [5]. BMD distinguishes itself by a pronounced sense of imbalance due to bilateral vestibular hypofunction and communication challenges stemming from bilateral hearing loss. Additionally, patients with BMD often exhibit a reduced tolerance for vertigo, and then a heightened rate of disability, thus significantly diminishing their quality of life. The management and treatment of BMD necessitate a more meticulous and conservative approach compared to UMD, especially when employing interventions that risk further hearing and/or vestibular damage [6]. Despite the acknowledged significance of precise diagnosis and treatment of BMD, research dedicated to its clinical characteristics remains limited, underlining the need for further investigation to enhance its treatment and management. Currently, the diagnosis of bilateral Ménière's disease remains challenging and requires examinations such as pure-tone audiometry, electrocochleography, vestibular-evoked myogenic potential, and MRI [5]. Furthermore, if a patient presents with bilateral fluctuating sensorineural hearing loss that responds well to diuretic treatment, a high suspicion of bilateral Ménière's disease should be considered.

Endolymphatic hydrops, a cardinal pathological hallmark of Meniere's Disease, was first visualized by a 3 Tesla magnetic resonance imaging (MRI) unit, and three-dimensional fluid-attenuated inversion recovery (3D-FLAIR) imaging was performed in patients by Nakashima et al. [7]. Subsequently, Nakashima described another MRI technology, three-dimensional real inversion recovery (3D-real IR), which demonstrated a higher contrast between the perilymphatic space and endolymphatic vessels compared with 3D-FLAIR imaging [8] Then Naganawa proposed a novel technique called "HYDROPS" to visualize endolymphatic hydrops. [9]. The correlation between the degree of endolymphatic hydrops and hearing loss has been central to academic inquiry, albeit with inconsistent outcomes. One study identified a significant correlation between Pure Tone Audiometry (PTA) thresholds at low to mid-frequency and the degree of endolymphatic hydrops across all cochlear turns, excluding vestibular hydrops, whereas average high-frequency hearing thresholds did not demonstrate a significant correlation with the degree of endolymphatic hydrops either in the vestibule or cochlea [10]. The other study revealed that patients presenting with both vestibular and cochlear hydrops experienced more severe hearing loss and staging of MD than those with isolated cochlear or vestibular hydrops. Nonetheless, no significant correlation was found between vestibular hydrops and hearing loss or disease staging [11]. Conversely, the degree of endolymphatic hydrops both in the vestibule and cochlea were significantly correlated with PTA thresholds [12] [13]. Moreover, disparate cochlear grading methods resulted in varying correlation coefficients between the degree of cochlear hydrops and hearing loss [14].

The inconsistency among study findings may be attributed to variations in the methods employed for visualizing endolymphatic hydrops via inner ear Magnetic resonance imaging (MRI), differences in criteria for assessing hydrops severity, individual variances among study populations, and potentially due to the lack of strict classification of Meniere's disease subtypes in previous research. Such factors might lead to conclusions that do not accurately reflect the specific context. Ménière's disease is a clinically heterogeneous disorder [15]. In clinical practice, only approximately 20% of patients present with bilateral involvement. Follow-up studies have shown that only a small percentage of patients progress from unilateral to bilateral disease. Consequently, we propose that bilateral Ménière's disease and unilateral Ménière's disease represent distinct subtypes of the condition.Current investigations into the correlation between the degree of endolymphatic hydrops and hearing loss have not differentiated between bilateral and unilateral Meniere's Disease, possibly failing to capture the clinical nuances of BMD accurately. Thus, this study aims to explore the relationship between endolymphatic hydrops features and hearing loss in Bilateral Meniere's Disease, endeavoring to elucidate further the clinical features of BMD and provide a foundation for its diagnosis and treatment.

Materials and methods

Patient cohort and clinical examination protocol

Participants

Inclusion Criteria Individuals diagnosed with bilateral Meniere's Disease by our team from March 1st, 2016, to March 1st, 2023, following the diagnostic criteria [16] for MD were enrolled. Each ear of the patient was assessed individually. A diagnosis of bilateral Meniere's disease is made if gadolinium-enhanced MRI confirms bilateral endolymphatic hydrops and both ears present with symptoms such as hearing loss, tinnitus, and ear fullness [6]. All participants underwent gadolinium-enhanced inner ear magnetic resonance imaging (Gd-enhanced MRI) and Pure Tone Audiometry (PTA). And PTA tests were completed within three days before the MRI examination,

Exclusion Criteria Patients whose MRI images were of insufficient quality for assessing the degree of endolymphatic hydrops, as well as those whose hearing loss

was too severe to yield responses to the PTA test, were excluded from the study.

Ethics

The ethical committee of the Eye and ENT Hospital at Fudan University granted approval for this study (#2,023,128). Given its retrospective nature and the absence of adverse effects on de-identified subjects, the requirement for patient consent forms was waived.

MRI protocol

Participants received an intravenous injection of a double dose (0.4 ml/kg body weight) of Gd-HP-DO3A. Four hours following the injection, MRI scans were conducted using a 32-channel phased-array coil (Verio; Siemens Healthcare, Erlangen, Germany) exclusively for reception. Imaging utilized T2 SPACE and three-dimensional real inversion recovery (3D-real-IR) sequences, with parameters for the 3D-real-IR sequence set as follows: voxel size = 0.2 × 0.2 × 0.6 mm; scan duration = 15 min and 20 s; repetition time = 6000 ms; echo time = 181 ms; inversion time = 1850 ms; slice thickness = 0.6 mm; field of view = 160 × 160 mm; matrix size = 768 × 768.

PTA protocol

Pure Tone Audiometry (PTA) thresholds across all frequencies (0.125–8 kHz) were assessed for participants. Affected ears demonstrated varying degrees of sensorineural hearing loss, with the mean pure tone thresholds at 0.5, 1, 2, and 4 kHz reflecting the average level of hearing loss.

Data

Data collection

Air conduction hearing thresholds, at 0.125–8 kHz, were recorded for participants. The average thresholds at 125, 250, and 500 Hz were considered as the low-frequency hearing level (LH), at 1 kHz and 2 kHz as the mid-frequency hearing level (MH), and at 4 kHz and 8 kHz as the high-frequency hearing level (HH). The overall hearing level (PA) was calculated from the average thresholds at 500 Hz, 1 kHz, 2 kHz, and 4 kHz. Additionally, participants' 3D-real-IR sequences of inner ear gadolinium-enhanced MRI were collected.

Data evaluation

Grading of endolymphatic hydrops Based on the 3D-real-IR sequence of inner ear gadolinium-enhanced MRI, cochlear hydrops (CL) was classified into four grades according to the criteria described by Gürkov et al. [17], as shown in Fig. 1, Grade 0: No expansion of the endolymphatic dark areas(1A); Grade I: The endolymphatic dark areas are round(1B); Grade II: The endolymphatic dark areas are semicircular(1C); Grade III: The endolymphatic dark areas

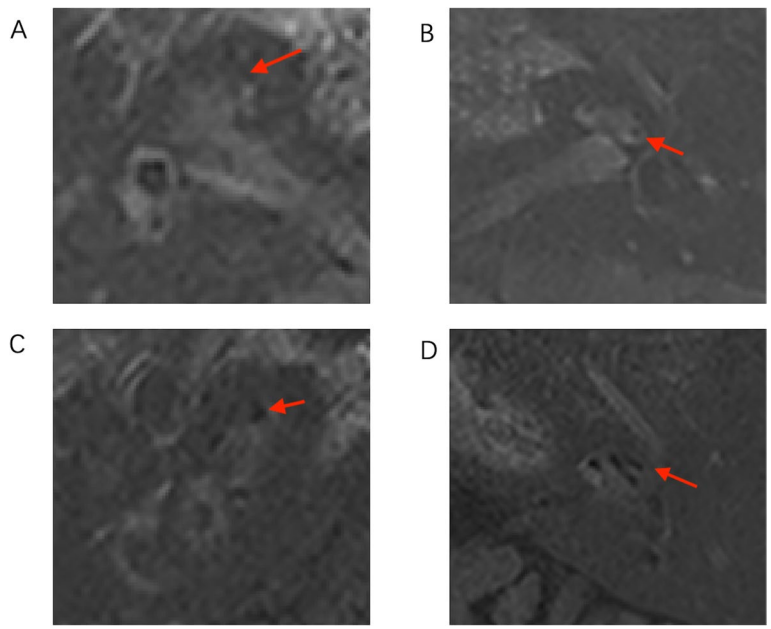

Fig. 1 Illustrations of the Classification method of Cochlear Hydrops degree,1A: Grade 0—No visible cochlear hydrops, indicating normal cochlear condition.1B: Grade I—Mild cochlear hydrops, showing slight expansion of the endolymphatic space.1C: Grade II—Moderate cochlear hydrops, with significant enlargement of the endolymphatic compartment.1D: Grade III—Severe cochlear hydrops, characterized by extensive dilation of the endolymphatic space, indicative of advanced pathological changes

are expanded and flattened, with the disappearance of the vestibular scale(1D). Vestibular hydrops (VL), as the criteria proposed by Bernaerts et al. [18], was also classified into four grades as Fig. 2 indicated: Grade 0: The saccule and utricle are normal, their combined area is less than half of the vestibular region(2A); Grade I: The saccule is expanded with the saccule area ≥ utricle area, and the saccule/utricle ratio is inverted(2B); Grade II: The endolymphatic areas of the saccule and utricle are expanded, with their delineation becoming blurred or disappearing, yet the peripheral perilymphatic high signal areas remain visible(2C); Grade III: The peripheral perilymphatic high signal areas are no longer visible, leaving only the endolymphatic dark areas(2D). The overall degree of c in the affected ear was represented by summing the grades of cochlear and vestibular hydrops.

Staging of Meniere's disease The Meniere's disease staging (MDS) for affected earswas determined based on the average hearing threshold at 0.5, 1.0, 2.0, and 4.0 kHz, as follows: Stage I: Average hearing threshold ≤ 25 dBHL; Stage II: Average hearing threshold 26–40 dBHL; Stage III: Average hearing threshold 41–70 dBHL; Stage IV: Average hearing threshold > 70 dBHL.

Statistical methods

The Spearman correlation coefficient was employed to analyze the relationship between the degree of endolymphatic hydrops and hearing loss or the staging of Meniere's disease. The differences in hearing loss between the affected ear with both vestibular and cochlear hydrops and that with isolated cochlear or vestibular hydrops within the same patient was analyzed by paired t-tests, and that of Meniere's disease staging was analyzed by Wilcoxon signed-rank tests.

Results

Baseline

As shown in Table 1, 77 patients diagnosed with Bilateral Meniere's Disease, encompassing 154 ears, were included in the study. The cohort had an average age of 50.9 years, comprising 44 males and 33 females. In terms of vestibular hydrops (VL), 27 ears were classified as Grade 0, 29 as Grade I, 85 as Grade II, and 13 as Grade III. For cochlear hydrops (CL), classifications were as follows: 13 ears at Grade 0, 66 at Grade I, 67 at Grade II, and 8 at Grade III. The overall degree of endolymphatic hydrops (EHL) was determined to be Grade I in 25 ears, Grade II in 27 ears, Grade III in 45 ears, Grade IV in 39 ears, Grade V in 16 ears, and Grade VI in 2 ears.

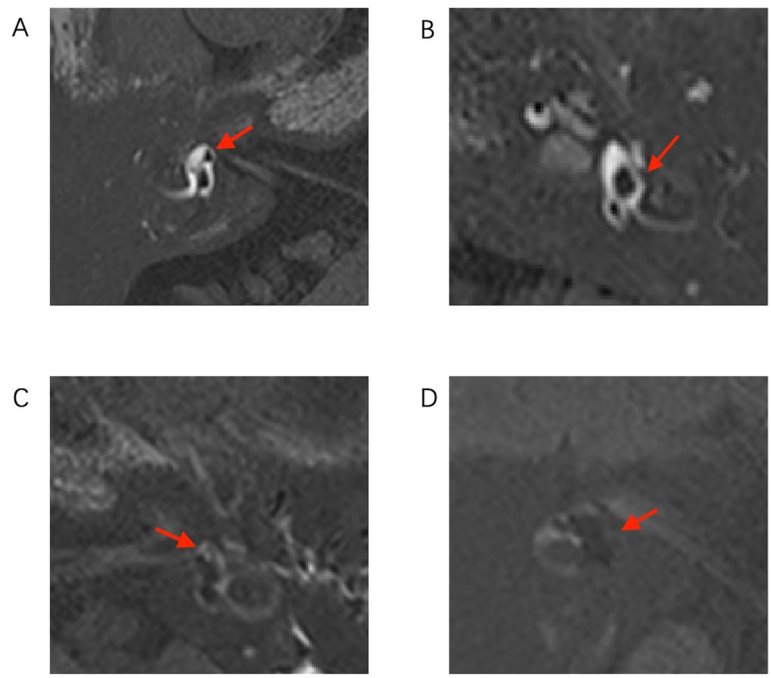

Fig. 2 Illustrations of the Classification method of Vestibular Hydrops degree,2A: Grade 0—No evidence of vestibular hydrops, representing a normal vestibular appearance 2B: Grade I—Mild vestibular hydrops, with slight distension of the saccule.2C: Grade II: The endolymphatic areas of the saccule and utricle are expanded, with their delineation becoming blurred or disappearing, yet the peripheral perilymphatic high signal areas remain visible. 2D: Grade III: The peripheral perilymphatic high signal areas are no longer visible, leaving only the endolymphatic dark areas

Table 1 This table summarizes the demographic and clinical characteristics of the study cohort

Male/Female	44/33
Age	51
VL(0/1/2/3)	27/29/85/13
CL(0/1/2/3	13/66/67/8
EHL(1/2/3/4/5/6)	25/27/45/39/16/2
PA	46.4 dB(95%CI[43.5–49.3])

Table 1 details the distribution of male and female patients (44 males and 33 females), the average age (51 years), the classification of vestibular hydrops (VL) and cochlear hydrops (CL) into grades 0 to 3, and the overall degree of endolymphatic hydrops (EHL) categorized into grades 1 to 6. Additionally, the overall hearing level (PA) is presented with a 95% confidence interval (46.4 dB [95% CI: 43.5–49.3]).

Audiometric findings relative to hydrops severity

From Table 2, it was observed that hearing thresholds across all frequencies progressively elevated with the advancement of vestibular and cochlear hydrops in the affected ears. Specifically, in cases where the vestibular hydrops in the affected ear was graded from 0 to 3, the average PA(the mean pure tone thresholds at 0.5, 1, 2, and 4 kHz) values for affected ears were 37 dB, 40 dB, 53 dB, and 61 dB, respectively. Correspondingly, for cochlear hydrops levels from 0 to 3, the average PA values for affected ears were 38 dB, 43 dB, 52 dB, and 68 dB, respectively. Based on Fig. 3, it is evident that as the severity of hydrops increases, there is a significant decline in hearing sensitivity, highlighting the progressive impact of endolymphatic hydrops on auditory function.

Table 2 presents the hearing thresholds at different frequencies (125 Hz, 250 Hz, 500 Hz, 1 kHz, 2 kHz, 4 kHz, and 8 kHz) according to the grades of vestibular and cochlear hydrops. The low-frequency hearing level (LH), the mid-frequency hearing level (MH), the high-frequency hearing level (HH), and the overall hearing level (PA) for vestibular hydrops grades (VL = 0 to VL = 3) and cochlear hydrops grades (CL = 0 to CL = 3).

Impact of endolymphatic hydrops site on hearing loss

The analysis included 30 patients with varying numbers of endolymphatic hydropic sites in bilateral ears. Among these, 30 ears exhibited both vestibular and cochlear hydrops, 9 ears exhibited isolated vestibular hydrops, and 21 ears exhibited isolated cochlear hydrops. Hearing thresholds of bilateral ears within the same patient were compared using a paired t-test. As shown in Table 3, ears with hydrops in both the cochlea and vestibule exhibited more severe hearing loss compared to those with isolated hydrops (cochlear or vestibular hydrops) in the same patient. It was found that ears exhibiting both vestibular and cochlear hydrops demonstrated an average low-frequency hearing threshold difference of 17 dB(95%CI 8-26 dB), a mid-frequency difference of 13 dB(95%CI 4-22 dB), a high-frequency difference of 10 dB(95%CI 1-20 dB), and a PA threshold difference of 14 dB(95%CI 5-23 dB), with all p-values less than 0.05. Additionally, the Meniere's Disease (MD) staging of bilateral ears was compared using a paired Wilcoxon test, revealing a statistically significant difference ($p < 0.05$, $z = 2.973$), indicating more severe MD staging in ears with both vestibular and cochlear hydrops compared to ears with isolated hydrops.

Table 3 compares the low-frequency hearing level (LH), the mid-frequency hearing level (MH), the high-frequency hearing level (HH), the overall hearing level (PA), and the staging of Meniere's disease (MDS), between one ear with both vestibular and cochlear hydrops versus the other with isolated hydrops in the same patient. The differences (d), *p*-values (p), and 95% confidence intervals (95%CI) are provided, demonstrating statistically significant differences in the severity of hearing loss and MDS between the two groups.

Table 2 The hearing thresholds of different grades of vestibular and cochlear hydrops

	125	250	500	1 k	2 k	4 k	8 k	LH	MH	HH	PA
VL = 0	33	34	35	35	36	43	51	34	35	47	37
VL = 1	32	35	36	36	39	48	59	34	38	53	40
VL = 2	44	48	51	51	51	57	67	48	51	62	53
VL = 3	51	55	60	63	58	62	73	56	60	67	61
CL = 0	34	36	36	38	35	40	47	35	37	43	38
CL = 1	34	37	39	41	43	51	61	37	42	56	43
CL = 2	45	48	51	51	50	57	68	48	51	63	52
CL = 3	66	70	73	69	64	64	69	70	67	67	68

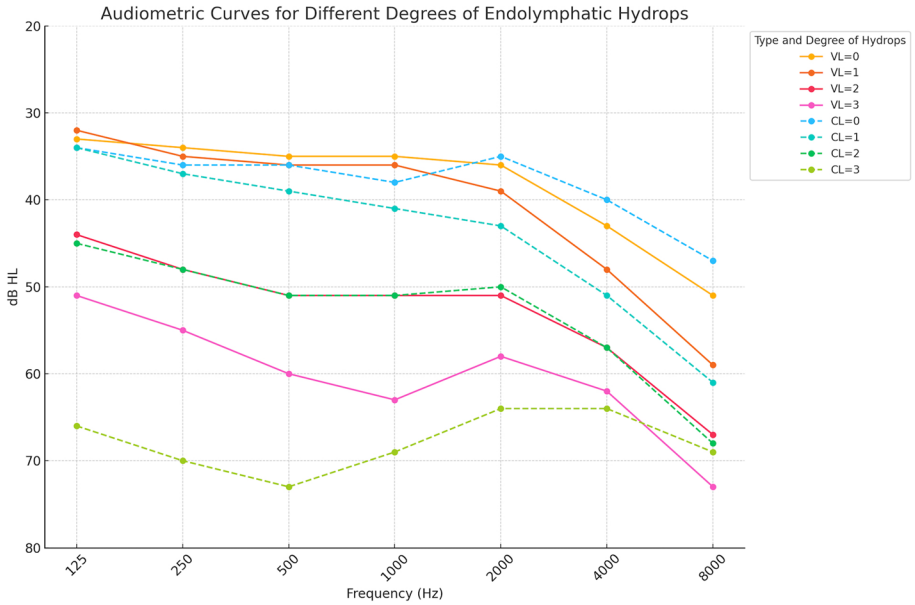

Fig. 3 Audiometric Curves for Different Degrees of Endolymphatic Hydrops

Figure 3 displays average hearing thresholds (dB HL) across frequencies (125–8000 Hz) for varying degrees of endolymphatic hydrops. The curves represent vestibular (VL) and cochlear (CL) hydrops from degrees 0 to 3. As hydrops severity increases, there is a notable decline in hearing sensitivity. These patterns emphasize the progressive impact of endolymphatic hydrops on auditory function

Table 3 The hearing level of the ears with different endolymphatic hydrops site

	d	p	95%CI
LH	17 dB	0.002	(8 -26)
MH	13 dB	0.003	(4–22)
HH	10 dB	0.02	(1–20)
PA	14 dB	0.002	(5–23)
MDS	/	0.003	/

Correlations between endolymphatic hydrops severity and audiological outcomes

Cochlear endolymphatic hydrops and hearing loss

As indicated in Table 4. A significant correlation was identified between the severity of cochlear hydrops and hearing threshold levels across all frequencies, with all p-values less than 0.05. The correlation was most pronounced with low-frequency average hearing levels (0.15–0.50 kHz) ($r = 0.462$, $p < 0.05$), and least pronounced with high-frequency average hearing levels (4-8 kHz) ($r = 0.261$, $p < 0.05$). Significant correlations were also observed with the PA (0.5-4 kHz) ($r = 0.383$) and Meniere's disease staging (MDS) ($r = 0.307$).

Table 4 outlines the Spearman correlation coefficients (r) and p-values (p) assessing the relationship between the degree of cochlear hydrops(CL) and hearing loss across various frequencies (125 Hz to 8 kHz), also including low-frequency (LH), mid-frequency (MH), high-frequency (HH) hearing levels, the overall hearing level (PA), and he degree of CL and Meniere's disease staging (MDS). Significant correlations indicate a strong relationship between CL and hearing impairment across all tested frequencies and MDS.

Vestibular endolymphatic hydrops and hearing loss

As indicated in Table 5, significant correlation was noted between the severity of vestibular hydrops and hearing threshold levels across all frequencies, with all p-values less than 0.05. The strongest correlation was observed with low-frequency average hearing levels (0.15–0.50 kHz) ($r = 0.468$, $p < 0.05$), and the weakest

Table 4 The relationship between the degree of cochlear hydrops and hearing loss

	125	250	500	1 k	2 k	4 k	8 k	LH	MH	HH	PA	MDS
r	0.433	0.453	0.443	0.379	0.316	0.242	0.252	**0.462**	0.368	0.261	0.383	0.307
p	0.000	0.000	0.000	0.000	0.000	0.002	0.001	0.000	0.000	0.001	0.000	0.000

Table 5 The relationship between the degree of vestibular hydrops and hearing loss

	125	250	500	1 k	2 k	4 k	8 k	LH	MH	HH	PA	MDS
R	0.393	0.453	0.495	0.482	0.368	0.304	0.277	**0.468**	0.455	0.314	0.464	0.436
p	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000

with high-frequency average hearing levels (4-8 kHz) ($r = 0.314$, $p < 0.05$). Significant correlations were further noted with PA (0.5-4 kHz) ($r = 0.464$) and Meniere's disease staging (MDS) ($r = 0.436$).

Table 5 details the Spearman correlation coefficients (r) and p-values (p) evaluating the association between the degree of vestibular hydrops(VL) and hearing loss at different frequencies (125 Hz to 8 kHz), also encompassing including low-frequency (LH), mid-frequency (MH), high-frequency (HH) hearing levels, the overall hearing level (PA). And the association between the CL and Meniere's disease staging (MDS). Significant correlations indicate strong relationships between VL and hearing impairment and MDS.

The overall endolymphatic hydrops and hearing loss

As shown in Table 6, significant correlation was established between the comprehensive degree of endolymphatic hydrops and hearing threshold levels across all frequencies, with all p-values less than 0.05. The most substantial correlation was with low-frequency average hearing levels (0.15–0.50 kHz) ($r = 0.571$, $p < 0.05$), and the least substantial with high-frequency average hearing levels (4-8 kHz) ($r = 0.351$, $p < 0.05$). Significant correlations were also seen with PA (0.5-4 kHz) ($r = 0.522$) and Meniere's disease staging (MDS) ($r = 0.463$).

Table 6 presents the Spearman correlation coefficients (r) and p-values (p) illustrating the correlation between the overall degree of endolymphatic hydrops (EHL) and hearing loss at various frequencies (125 Hz to 8 kHz), also including assessments of low-frequency (LH), mid-frequency (MH), high-frequency (HH) hearing levels, the overall hearing level (PA). And the association between the EHL and Meniere's disease staging (MDS). Statistically significant correlations underscore the profound impact of EHL on hearing across and MDS.

Discussion

This study was undertaken to elucidate the association between the features of endolymphatic hydrops and hearing loss in patients diagnosed with Bilateral Meniere's Disease (BMD), a disorder that significantly undermines the quality of life through bilateral vestibular dysfunction and hearing loss. The findings from our investigation revealed a robust correlation between the severity of the overall endolymphatic hydrops, cochlear hydrops, and vestibular hydrops in bilateral Meniere's disease with hearing loss across all frequencies, particularly pronounced in the low to mid-frequency ranges. Existing literature on the correlation between the severity of endolymphatic hydrops and hearing loss has yielded inconsistent results. While a consensus exists among several studies that the severity of cochlear and vestibular hydrops is correlated with hearing loss [12, 13, 19–22], this concurs with our findings. However, certain studies posit that the severity of vestibular hydrops bears no relation to the extent of hearing loss [10, 11], a conclusion at odds with our findings. Such disparities may be attributable to the limited sample sizes in various studies, individual variances among patients, and the disparities in MRI imaging techniques and grading methodologies for vestibular and cochlear hydrops across different clinical settings [22]. For example, disparities in cochlear hydrops grading methods were noted to produce varied correlation coefficient with hearing loss [14].Moreover, other studies did not distinguish between unilateral and bilateral Ménière's disease in their included patients. Therefore, in future research, we will use the same methodology to evaluate the relationship between endolymphatic hydrops characteristics and hearing loss in unilateral Ménière's disease.

This study found that the severity of bilateral Meniere's Disease (MD) endolymphatic hydrops and MD staging are correlated, which partially diverges from the

Table 6 The relationship between the degree of overall endolymphatic hydrops and hearing loss

	125	250	500	1 k	2 k	4 k	8 k	LH	MH	HH	PA	MDS
R	0.503	0.555	0.579	0.532	0.418	0.333	0.324	**0.571**	0.508	0.351	0.522	0.463
P	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000

conclusions of the study [23]. That study posited that traditional MD staging does not correlate with the degree of endolymphatic hydrops (EH) visible on MRI, although advanced staging of MD may indicate increased severity of cochlear and vestibular EH.

Furthermore, it was observed that from Table 3, within the same patient, ears afflicted with both vestibular and cochlear hydrops manifested more severe hearing loss and MD staging compared to those with isolated cochlear hydrops or vestibular hydrops, particularly in LH, suggesting that isolated vestibular or cochlear hydrops may represent an earlier stage of the disease. Previous study [24] has suggested that isolated hydrops in the affected ear results in milder hearing loss. Our study, by comparing different affected ears within the same patient, further substantiates this finding. Early management of the disease at the stage of isolated hydrops may improve patient outcomes. This underscores the importance of continuous follow-up in clinical practice for such patients to monitor disease progression and identify influencing factors.

The novelty of our study lies in its focus on BMD to enhance the extant body of evidence on the correlation between the severity of endolymphatic hydrops and hearing loss in bilateral Meniere's disease, which provided BMD data to the existing literature. Moreover, a distinguishing feature of our study is the employment of novel methodologies for determining the grades of cochlear and vestibular hydrops. Utilizing the latest grading method proposed by Gürkov et al. [17] for cochlear hydrops, which classifies the condition into grades 0 through 3, which demonstrated the highest correlation with the degree of hearing loss in this study compared other grades methods [14]. For vestibular grading, following the criteria outlined by BERNAERTS et al. [18], vestibular hydrops was categorized into grades 0 through 3, with grade 1 identified as saccular expansion surpassing the utricle's area, thereby facilitating the early diagnosis of vestibular hydrops [25]. The findings from our study underscored a strong correlation between the severity of vestibular hydrops and hearing loss, thereby validating the scientific robustness of the grading method mentioned above. Finally, by amalgamating the grades of vestibular and cochlear hydrops, the overall severity of endolymphatic hydrops in the affected ear was represented. It was concluded that the aggregate severity of hydrops, encompassing both endolymphatic and cochlear hydrops, exhibited a stronger correlation with hearing loss. This suggests that such an approach enhances the accuracy in assessing the correlation between endolymphatic hydrops and hearing loss, considering the impact of both vestibular and cochlear involvement on hearing.

The limitations of this study include its reliance on specific imaging techniques (3D-real-IR [8]) for assessing endolymphatic hydrops, which may not be universally accessible or standardized across healthcare facilities. The 3D-FLAIR [7] and HYDROPS [9] MRI imaging techniques are also methods for visualizing endolymphatic hydrops. The integration of radiomic features extracted from conventional T2-weighted MRI scans has shown promise in the diagnosis of Menière's disease through a multi-layer perceptron classification model [26] Therefore, MRI can aid in the differential diagnosis of symptoms similar to those of Meniere's syndrome. Future research should endeavor to standardize imaging protocols for the diagnosis of endolymphatic hydrops and conduct prospective studies to gain a more comprehensive understanding of the progression of BMD and the factors influencing hearing loss. Such endeavors aim to refine treatment modalities for patients afflicted with this condition. In this study, PTA tests were completed within three days before the MRI examination, and no diuretic treatment was administered before the above examinations. This ensures the reliability of our study results. However, since this is a retrospective study, information regarding the patients' previous use of diuretics was not considered, which could potentially affect the study outcomes. Therefore, future prospective studies may consider analyzing patients at their initial visit to enhance the credibility of the research. Prospective cohort studies are still needed to explore whether the hearing levels in the same patient change with variations in the degree of endolymphatic hydrops. Additionally, future studies should examine the relationship between the summation potential (SP) from electrocochleography and the degree of endolymphatic hydrops.

Future research should evaluate the functional performance of patients using the matrix sentence test, especially for those with bilateral Ménière's disease. This test is essential for assessing hearing performance in noisy environments. Patients with bilateral hearing loss may face greater difficulties, exacerbated by different audiometric curve morphologies. Additionally, considering the use of hearing aids in these patients might improve listening performance in challenging acoustic environments [27]. This approach could provide valuable insights for the management of bilateral Ménière's disease.

In conclusion, the findings of this research into the correlation between the characteristics of endolymphatic hydrops and hearing loss in BMD contribute to an enhanced comprehension of the syndrome, particularly by focusing on the distinct clinical features of BMD. This lays the foundation for subsequent research directed at improving the quality of life for those affected by this debilitating condition.

Conclusion

The degree of endolymphatic hydrops in affected ears of patients with Bilateral Meniere's Disease was found to correlate with the severity of hearing loss and the staging of Meniere's disease. The degree of endolymphatic hydrops positively correlated with hearing loss across all frequencies, with the strongest correlation observed in low-frequency hearing loss and the weakest in high-frequency hearing loss, and the overall degree of endolymphatic hydrops and hearing loss has the strongest correlation between hearing loss. Patients with both vestibular and cochlear hydrops exhibited more severe hearing loss and staging of MD than those with isolated hydrops.

Abbreviations

MD	Meniere's Disease
EH	Endolymphatic hydrops
UMD	Unilateral Meniere's Disease
BMD	Bilateral Meniere's Disease
PTA	Pure Tone Audiometry
MRI	Magnetic resonance imaging
Gd-enhanced MRI	Gadolinium-enhanced inner ear magnetic resonance imaging
3D-real-IR	Three-dimensional real inversion recovery sequences
LH	Low-frequency hearing level
MH	Mid-frequency hearing level
HH	High-frequency hearing level
PA	Overall hearing level
CL	Cochlear hydrops
VL	Vestibular hydrops
EHL	Endolymphatic hydrops
MDS	Meniere's disease staging

Authors' contribution

XL contributed to the conceptualization, methodology, and supervision of the study. DW was responsible for data curation, formal analysis, and writing the original draft. Yue-Lin Hsieh conducted the investigation, validation, and visualization of the results. SS managed resources, project administration, and contributed to the writing, review, and editing of the manuscript. WW provided software support, secured funding for the study, and contributed to the writing, review, and editing of the manuscript.

Funding

The authors declare no financial interests in the present study and have no potential conflicts of interest to disclose.This study received support from the National Natural Science Foundation of China (Nos. 82101222) and the Natural Science Foundation of Shanghai (No. 20ZR1409600).

Availability of data and materials

The data in this study are clinical in nature and therefore cannot be publicly disclosed.

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

The ethical committee of the Eye and ENT Hospital at Fudan University approved this study (#2023128).

Consent for publication

I confirm that I have obtained consent from all participants for the publication of the content of this paper.

Competing interests

The authors declare no competing interests.

Received: 1 July 2024 Accepted: 5 September 2024
Published online: 16 September 2024

References

1. Rizk HG, Mehta NK, Qureshi U, Yuen E, Zhang K, Nkrumah Y, et al. Pathogenesis and etiology of Ménière disease: a scoping review of a century of evidence. JAMA Otolaryngol Head Neck Surg. 2022;148(4):360–8. https://doi.org/10.1001/jamaoto.2021.4282. (PubMed PMID: 35142800).
2. Gürkov R, Pyykö I, Zou J, Kentala E. What is Menière's disease? A contemporary re-evaluation of endolymphatic hydrops. J Neurol. 2016;263 Suppl 1:S71–81.Epub 20160415. https://doi.org/10.1007/s00415-015-7930-1. PubMed PMID: 27083887; PubMed Central PMCID: PMC4833790.
3. Hoskin JL. Ménière's disease: new guidelines, subtypes, imaging, and more. Curr Opin Neurol. 2022;35(1):90–7. https://doi.org/10.1097/wco.0000000000001021. PubMed PMID: 34864755.
4. Phillips JS, Murdin L, Rea P, Sutton L. Clinical Subtyping of Ménière's Disease. Otolaryngol Head Neck Surg. 2018;159(3):407–9. https://doi.org/10.1177/0194599818773077. PubMed PMID: 29688822. Epub 20180424.
5. Nabi S, Parnes LS. Bilateral Ménière's disease. Curr Opin Otolaryngol Head Neck Surg. 2009;17(5):356–62. https://doi.org/10.1097/MOO.0b013e3283304cb3. (PubMed PMID: 19617826).
6. Pérez-Carbonell T, Orts-Alborch M, Pla-Gil I, Pérez-Guilén V, Tenías-Burrillo JM, Marco-Algarra J, et al. Bilateral Ménière's disease according to its form of debut: synchronous and metachronous disease. J Laryngol Otol. 2023;137(7):782–8. https://doi.org/10.1017/s0022215122002262. PubMed PMID: 36200516. Epub 20221006.
7. Nakashima T, Naganawa S, Sugiura M, Teranishi M, Sone M, Hayashi H, et al. Visualization of endolymphatic hydrops in patients with Meniere's disease. Laryngoscope. 2007;117(3):415–20. https://doi.org/10.1097/MLG.0b013e31802c300c. PubMed PMID: 17279053.
8. Naganawa S, Satake H, Kawamura M, Fukatsu H, Sone M, Nakashima T. Separate visualization of endolymphatic space, perilymphatic space and bone by a single pulse sequence; 3D-inversion recovery imaging utilizing real reconstruction after intratympanic Gd-DTPA administration at 3 Tesla. Eur Radiol. 2008;18(5):920–4. Epub 20080307. https://doi.org/10.1007/s00330-008-0854-8. PubMed PMID: 18324405.
9. Naganawa S, Yamazaki M, Kawai H, Bokura K, Sone M, Nakashima T. Imaging of Ménière's disease after intravenous administration of single-dose gadodiamide: utility of multiplication of MR cisternography and HYDROPS image. Magn Reson Med Sci. 2013;12(1):63–8. Epub 20130311. https://doi.org/10.2463/mrms.2012-0027 . PubMed PMID: 23474961.
10. 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):974–9. https://doi.org/10.1002/lary.25576. PubMed PMID: 26333096. Epub 20150829.
11. Huang Y, Zhao P, Han Z, Xie J, Liu Y, Gong S, et al. Evaluation of the relationship between endolymphatic hydrops and hearing loss in Meniere's disease based on three-dimensional real inversion recovery sequence. Braz J Otorhinolaryngol. 2023;89(5):101314. Epub 20230828. https://doi.org/10.1016/j.bjorl.2023.101314. PubMed PMID: 37688911; PubMed Central PMCID: PMC10504479.
12. Yang S, Zhu H, Zhu B, Wang H, Chen Z, Wu Y, et al. Correlations between the degree of endolymphatic hydrops and symptoms and audiological test results in patients With Menière's disease: a eeevaluation. Otol Neurotol. 2018;39(3):351–6. https://doi.org/10.1097/mao.0000000000001675. PubMed PMID: 29287037.
13. 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):286–92. Epub 20181217. https://doi.org/10.1177/0003489418819551. PubMed PMID: 30556402.
14. Han Z, Qiu X, Huang Y, Sun Q, Ding H, Xie J, et al. Correlation between grading methods of the cochlear endolymphatic hydrops and hearing loss in meniere's disease using three-dimensional real inversion recovery sequences. Acta Otolaryngol. 2023;143(5):370-5. Epub 20230517. https://doi.org/10.1080/00016489.2023.2209134. PubMed PMID: 37195094.

15. Frejo L, Soto-Varela A, Santos-Perez S, Aran I, Batuecas-Caletrio A, Perez-Guillen V, et al. Clinical subgroups in bilateral meniere disease. Front Neurol. 2016;7:182. https://doi.org/10.3389/fneur.2016.00182. PubMed PMID: 27822199; PubMed Central PMCID: PMC5075646. Epub 20161024.
16. Lopez-Escamez JA, Carey J, Chung WH, Goebel JA, Magnusson M, Mandalà M, et al. Diagnostic criteria for Menière's disease. J Vestib Res. 2015;25(1):1–7. https://doi.org/10.3233/ves-150549. PubMed PMID: 25882471.
17. Gürkov R, Flatz W, Louza J, Strupp M, Krause E. In vivo visualization of endolyphatic hydrops in patients with Meniere's disease: correlation with audiovestibular function. Eur Arch Otorhinolaryngol. 2011;268(12):1743–8. https://doi.org/10.1007/s00405-011-1573-3 . PubMed PMID: 21431434. Epub 20110323
18. Bernaerts A, Vanspauwen R, Blaivie C, van Dinther J, Zarowski A, Wuyts FL, et al. The value of four stage vestibular hydrops grading and asymmetric perilymphatic enhancement in the diagnosis of Menière's disease on MRI. Neuroradiology. 2019;61(4):421–9. https://doi.org/10.1007/s00234-019-02155-7. PubMed PMID: 30719545; PubMed Central PMCID: PMC6431299. Epub 20190205.
19. Hu Y, Zhang Y, Zhao X, Li J. Endolymphatic hydrops imaging and correlation with clinical characteristics, audiovestibular function and mental impairment in patients with Meniere's disease. Eur Arch Otorhinolaryngol. 2023;280(9):4027–36. https://doi.org/10.1007/s00405-023-07899-w. PubMed PMID: 36849561; PubMed Central PMCID: PMC10382354. Epub 20230227.
20. Kahn L, Hautefort C, Guichard JP, Toupet M, Jourdaine C, Vitaux H, et al. Relationship between video head impulse test, ocular and cervical vestibular evoked myogenic potentials, and compartmental magnetic resonance imaging classification in menière's disease. Laryngoscope. 2020;130(7):E444–e52. Epub 20191119. https://doi.org/10.1002/lary.28362. PubMed PMID: 31742710.
21. Zhang W, Xie J, Li S, Zhang B. Correlation between quantitative value of endolymphatic hydrops and hearing threshold using magnetic resonance imaging. Ann Otol Rhinol Laryngol. 2023;132(10):1149–55. https://doi.org/10.1177/00034894221134729. PubMed PMID: 36412145. Epub 20221122.
22. de Pont LMH, van Steekelenburg JM, Verhagen TO, Houben M, Goeman JJ, Verbist BM, et al. Hydropic ear disease: correlation between audiovestibular symptoms, endolymphatic hydrops and blood-labyrinth barrier impairment. Front Surg. 2021;8:758947. https://doi.org/10.3389/fsurg.2021.758947. PubMed PMID: 34805261; PubMed Central PMCID: PMC8601159. Epub 20211104.
23. Yan HY, Young YH. Role of conventional MD staging in modern era of hydrops MR imaging. Laryngoscope Investig Otolaryngol. 2024;9(1):e1213. Epub 20240115. https://doi.org/10.1002/lio2.1213. PubMed PMID: 38362181; PubMed Central PMCID: PMC10866591.
24. Niu Y, Chen W, Lin M, Sha Y. Development and characteristics of hearing loss with the progression of endolymphatic hydrops. Ear Nose Throat J. 2022:1455613221101088. Epub 20220720. https://doi.org/10.1177/01455613221101088. PubMed PMID: 35856637.
25. Jasińska A, Lachowska M, Wnuk E, Pierchała K, Rowiński O, Niemczyk K. Correlation between magnetic resonance imaging classification of endolymphatic hydrops and clinical manifestations and audiovestibular test results in patients with definite Ménière's disease. Auris Nasus Larynx. 2022;49(1):34–45. https://doi.org/10.1016/j.anl.2021.03.027. Epub 20210415. PubMed PMID: 33865653.
26. van der Lubbe M, Vaidyanathan A, de Wit M, van den Burg EL, Postma AA, Bruintjes TD, et al. A non-invasive, automated diagnosis of Menière's disease using radiomics and machine learning on conventional magnetic resonance imaging: A multicentric, case-controlled feasibility study. Radiol Med. 2022;127(1):72–82. Epub 20211125. https://doi.org/10.1007/s11547-021-01425-w. PubMed PMID: 34822101; PubMed Central PMCID: PMC8795017.
27. Portelli D, Loteta S, Ciodaro F, Salvago P, Galletti C, Freni L, et al. Functional outcomes for speech-in-noise intelligibility of NAL-NL2 and DSL v.5 prescriptive fitting rules in hearing aid users. Eur Arch Otorhinolaryngol. 2024;281(6):3227–35. Epub 20240328. https://doi.org/10.1007/s00405-024-08587-z. PubMed PMID: 38546852.

Publisher's Note