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Sensorineural hearing loss in patients with vestibular schwannoma correlates with the presence of utricular hydrops as diagnosed on heavily T2-weighted MRI

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KEYWORDS
Vestibular schwannoma;
Magnetic resonance imaging (MRI);
Endolymphatic hydrops;
Inner ear

Abstract
Purpose: The purpose of this study was to assess whether the volume of the vestibular endolymphatic space correlates with the degree of hearing loss using heavily T2-weighted fast imaging employing steady-state acquisition with cycle phase (FIESTA-C) MRI.
Materials and methods: A total of 23 patients with vestibular schwannoma, as diagnosed on typical image findings, who underwent FIESTA-C MRI were included. There were 13 women and 10 men with a mean age of 63.5 ± 9.3 (SD) years (range: 49—88 years). Two radiologists independently evaluated the volume of the utricle and saccule. Correlation between tumor volume, vestibular endolymphatic space volume and degree of hearing loss — as evaluated with the levels of pure-tone average and speech recognition threshold — were searched for.
Results: The mean saccular, utricular and tumor volumes were 3.17 ± 1.1 (SD) mm^3 (range: 1.45—5.7 mm^3), 14.55 ± 5 (SD) mm^3; (range: 6.6—23.9 mm^3) and 17.4 ± 5.5 (SD) mm^3; (range: 8.3—25.4 mm^3), respectively. There was a moderate correlation between the volume of the utricle and the degree of hearing loss as evaluated with the levels of pure-tone average (rho = 0.5; *P* = 0.015) and speech recognition threshold (rho = 0.58; *P* = 0.004). There were no significant correlations between saccular and tumor volumes and the degree of hearing loss.

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Conclusion: The volume of the utricle in patients with obstructive vestibular schwannoma moderately correlates with the degree of hearing loss.

Abbreviations

VS	vestibular schwannoma
EH	endolymphatic hydrops
PTA	pure-tone average
SRT	speech recognition threshold

Introduction

The origin of cochlear and vestibular symptoms in patients with vestibular schwannoma (VS) is uncertain and various mechanisms related to cochlear dysfunction have been speculated including endolymphatic hydrops (EH) [1—3]. Currently, magnetic resonance imaging (MRI) is the imaging examination of choice for the diagnosis of VS [4,5] and EH using inversion recovery sequences [6]. Previous studies using labyrinthine tap (i.e., invasive platinotomy to obtain perilymphatic fluid sample) revealed that protein levels were 5 to 15 times higher in patients with VS than in healthy subjects [7,8]. Nevertheless, two studies have also reported a precipitate in the endolymphatic space related to the high protein content [3,9].

High-resolution computed tomography and MRI have both a well-established role in the diagnosis of various inner ear diseases [10,11]. Three-dimensional fluid attenuation inversion recovery (3D-FLAIR) sequence allows the visualization of subtle compositional changes of the inner ear fluids in various diseases related to hemorrhage and increased protein levels [12—16]. Kim et al. have demonstrated that the increased cochlear signal on 3D-FLAIR images correlated with the degree of hearing loss, as measured by the pure-tone average (PTA) [16]. Lee et al. were the first to suggest that the high signal seen in the vestibule with 3D-FLAIR sequences in VS patients was related to the well-known high protein concentrations in the perilymph, while the endolymph (utricle and saccule) appeared as dark signal intensity [12]. Based on this condition, Naganawa et al. assessed the rate of EH in patients with VS on non-contrast 3D-FLAIR images and found no correlation between EH and vertigo [17]. However, they did not assess the correlation between EH and the degree of sensorineural hearing loss.

Recently, researchers have reported that fast imaging employing steady-state acquisition with cycle phase (FIESTA-C, General Electrics Healthcare) sequence demonstrated a significant decrease of perilymphatic signal intensity in patients with vestibular VS compared to patients with cerebellopontine angle meningiomas [18,19]. It has also been reported that only the perilymphatic signal decreased on FIESTA-C sequence, due to the elevated perilymphatic protein content caused by the internal auditory canal tumor, allowing recognition of the utricle and saccule in the inner ear [19].

The purpose of this study was to assess whether the volume of the vestibular endolymphatic space correlates with the degree of hearing loss using heavily T2-weighted fast imaging using the FIESTA-C sequence.

Material and methods

Patients

In this retrospective imaging study, 32 patients were initially recruited between December 2015 and May 2017. The study protocol was approved by our institutional review board (IRB E2017-23). All of the patients were recruited when they had typical imaging findings of VS on MRI, including ''ice cream cone'' shape tumor centered on the internal auditory canal on the cochleovestibular nerve, presenting with a relatively high signal on T2-weighted images, a hypo-intense filling defect on steady-state free precession and a homogenous enhancement on post-contrast T1-weighted images [4]. VS was defined as non-obstructive when cerebrospinal fluid was present between the tumor and the internal auditory canal. It was defined as obstructive when the tumor completely obstructed the internal auditory canal without cerebrospinal fluid around even for small tumors of the fundus [19]. Patients with a history of gamma knife radiosurgery or surgery were excluded based on the supplemental risk of post-treatment hearing loss.

Nine patients (28%) were excluded from the analysis: 6 patients had no significant decrease in perilymphatic signal on FIESTA-C, hence not allowing differentiation between the high signal of the endolymph and the low signal of the perilymph (Fig. 1). The other three excluded patients had both perilymphatic and endolymphatic signal decrease, thus the volume of the endolymphatic space could not be performed.

Twenty-three patients were thus ultimately included. There were 13 women and 10 men with a mean age of 63.5 ± 9.3 (SD) years (range: 49—88 years). The patients had unilateral schwannoma, which was confirmed by MRI: 13 patients had a tumor on the right side and 10 on the left side.

Audiometric tests

The PTA hearing levels of bone conduction were calculated as the mean of the hearing levels measured at 500,

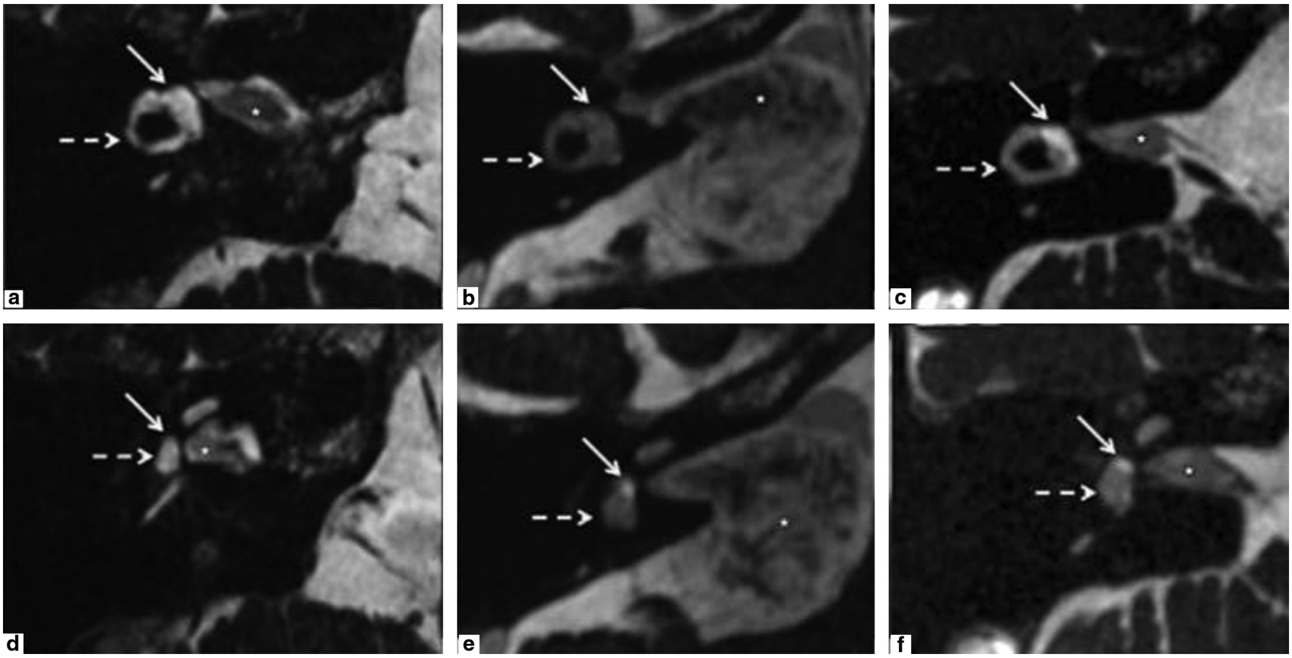

Figure 1. Figures show MR images of three patients with vestibular schwannoma. A and D. 77-year-old woman with a right non-obstructive vestibular schwannoma with a volume of 0.04 cm^3. Fast imaging employing steady-state acquisition with cycle phase (FIESTA-C) image shows a right intracanalicular non-obstructive schwannoma (asterisk) at the level of the utricle (A, arrow) and the saccule (D, arrow). The schwannoma is surrounded by cerebrospinal fluid. The utricle and the saccule remain as a high signal (arrow) as the perilymph from the vestibule and the lateral semicircular canal (dotted arrow). B and E. 73-year-old man with a right obstructive vestibular schwannoma with a volume of 4.5 cm^3. FIESTA-C sequence shows a right intracanalicular obstructive schwannoma (asterisk) at the level of the utricle (B, arrow) and the saccule (E, arrow). The saccule remains displays a high signal (E, arrow) while the perilymph from the vestibule and the lateral semicircular canal (dotted arrow) and the utricle (B, arrow) display a low signal. C and F. 62-year-old male with a right obstructive vestibular schwannoma with a volume of 0.1 cm^3. FIESTA-C image shows a right intracanalicular obstructive schwannoma (asterisk) at the level of the utricle (C, arrow) and the saccule (F, arrow). The utricle and the saccule (arrow) display high signal contrasting with the low signal of the perilymph from the vestibule (dotted arrow).

1000, 2000, and 4000 Hz. Patients' hearing was classified as mild (26—40 dB HL), moderate (41—60 dB HL), severe (61—80 dB HL) and profound (over 81 dB HL). Speech recognition threshold (SRT) was defined as the minimum hearing level for speech at which an individual can recognize 50% of the speech material [20]. For the purpose of data analysis, complete absence of response was assigned a value of 120 dB for PTA and for speech discrimination.

Imaging

All patients had 3T MRI examination before and after intravenous administration of gadolinium-based contrast material. MRI examinations were carried out on a General Electric Discovery MRI 750® (General Electric Healthcare) with a 16-channel head—neck—spine coil. We performed FIESTA-C sequences before contrast administration. Axial T1-weighted spin-echo images were acquired after a single intravenous administration of Gadobutrol (Gadovist®, Bayer Healthcare) at a dose of 0.1 mmol/kg [11].

FIESTA-C is a refocused steady-state gradient echo sequence that provides high signals from tissues with elevated T2/T1 ratios and an excellent spatial resolution [21]. The FIESTA-C sequence was performed in the plane of the lateral semicircular canal with the following parameters: TR, 7 ms; TE, 2.8 ms; number of excitations (NEX), 1; matrix size, 484 × 484; flip angle, 60°; bandwidth, 83.3 kHz; and 0.3 mm slice thickness covering the labyrinth with a 20 cm field of view. We employed the autocalibrating reconstruction for cartesian (ARC) parallel imaging technique with an acceleration factor of 2 and a scan time of 4 min and 40 sec. The T1-weighted spin-echo sequence was performed in the axial plane with the following parameters: TR, 580 ms; TE, 19 ms; NEX, 2; spacing between slices, 1.7 mm; matrix size, 416 × 288; and 1.5 mm slice thickness covering the labyrinth with a 16.5 cm field of view.

Visualization of the vestibular endolymphatic space

The presence of the saccule and utricle was verified on the FIESTA-C sequence by two radiologists specialized in head and neck imaging (M. E., G. P.) with 6- and 2-years of experience in hydrops imaging respectively (Fig. 2). The saccule appeared as a high signal area located on the medial and anterior wall of the vestibule, underneath the level of the lateral semicircular canal [22,23]. The utricle appeared as a high signal elliptical zone that occupies the superior part of the vestibule, at the level of the lateral semicircular canal [22,23].

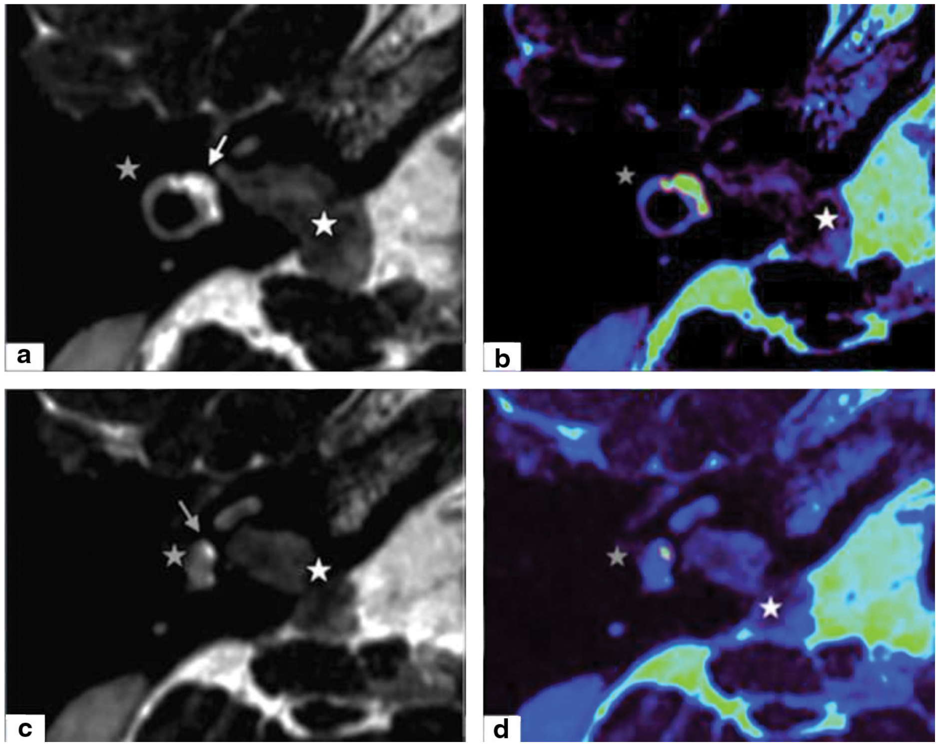

Figure 2. 70-year-old man with right obstructive vestibular schwannoma with a volume of 1.5 cm^3. A—B. Fast imaging employing steady-state acquisition with cycle phase (FIESTA-C) images in the axial plane at the level of the utricle. A. FIESTA-C image shows right intracanalicular schwannoma (white asterisk) and the utricle as a high signal (arrow) while the perilymph from the vestibule and the lateral semicircular canal displays low signal (gray asterisk). B. FIESTA-C image with the rainbow-scale on color map shows the utricle as a green signal and the perilymph (gray asterisk) as a blue signal and demonstrates the manual segmentation for the utricle in order to measure the volume. C—D. FIESTA-C images in the axial plane at the level of the saccule. C. FIESTA-C image shows the saccule (gray arrow) as a high signal contrasting with the low signal of the perilymph from the vestibule (gray asterisk). D. FIESTA-C image with the rainbow-scale on color map showing the saccule as a green signal and the perilymph (gray asterisk) as a blue signal and demonstrating the manual segmentation for the saccule in order to measure the volume.

Tumor volume and vestibular endolymphatic space volume measurements

MR images of each patient were evaluated independently by the same two radiologists, blinded to clinical data. The imaging data of inner ears were analyzed using a workstation (ADW® 4.6, General Electric Healthcare). The two radiologists used the region of interest method with manual segmentation, drawing the surface of the utricle and saccule on axial FIESTA-C images using the color map with the rainbow-scale providing a high-contrast between the endolymph, the perilymph and the surrounding bone. The software then calculated the volume of the utricule, the saccule and of the entire vestibular endolymphatic spaces. The same method was used to measure the schwannoma volume. They also recorded whether each schwannoma obstructed the fundus of the internal auditory canal. Volumetric analysis was expressed in cubic centimeters (cm^3) for the schwannoma and in cubic millimeters (mm^3) for the vestibular endolymphatic space.

Statistical analysis

Data were analyzed using SPSS software v22.0 (IBM). The intraclass coefficient correlation (ICC) was used to evaluate the reproducibility of the volume by the two observers. ICC values < 0.5 were considered to indicate of poor reliability, values between 0.5 and 0.75 moderate reliability, values between 0.75 and 0.9 good reliability, and values > 0.90 excellent reliability [24]. The mean absolute deviations between the two radiologists for the saccular, utricular and vestibular endolymphatic volumes were estimated. A range of deviation of 10% between measures was considered as adequate for volume estimations. A paired Student's t-test was used to compare the degree of hearing loss in symptomatic and asymptomatic ears in patients with VS. Pearson test, Spearman test and Brunner—Munzel test were used for categorical data to explore the correlation between the volume of the schwannoma, the volume of the vestibular endolymphatic space and the degree of hearing loss. For the purpose of the study, the volumes evaluated by the most experienced radiologist were taken as reference values in order to explore the correlation with the audiometric results. Significance was set at $P < 0.05$.

Results

MRI data

All data are summarized in Table 1. The mean tumor volume of the 23 included patients was 1.74 ± 2.5 (SD) cm^3 (range: 0.1—11.6 cm^3). All these patients had obstructive VS.

Table 1 Clinical and radiological characteristics in 32 patients with vestibular schwannoma.

Patients	Sex	Age (years)	Tumor side	PTA (dB)	SRT (dB)	Endolymph volume 1 (mm^3)			Endolymph volume 2 (mm^3)			Schwannoma volume (cm^3)
						Utricle	Saccule	Total	Utricle	Saccule	Total	
1	F	58	R	18.75	20	14.1	2.2	16.3	14.6	2.5	17.1	0.52
2	F	57	R	22.5	30	13	3.5	16.5	12.7	3.3	16	1.19
3	F	69	L	68.75	70	11.9	3.2	15.1	13	2.8	15.8	0.23
4	F	88	L	56.25	70	17.6	3	20.6	16.7	2.7	19.4	1.74
5	F	74	R	53.75	45	9.6	2.6	12.2	9.3	2.2	11.5	5.85
6	M	58	L	120	120	23.9	2.2	26.1	23.4	2.4	25.8	1.54
7	F	67	L	73.75	85	17.9	3.8	21.7	18	3.7	21.7	11.64
8	M	63	L	108.75	120	21.6	3.2	24.8	19.8	2.8	22.6	0.78
9	F	73	R	71.25	120	16.3	2.2	18.5	15.1	2.8	17.9	5.12
10	F	51	R	52.5	50	14.9	4.1	19	13.3	3.8	16.8	0.32
11	M	70	R	52.5	30	7.1	1.45	8.5	6.6	1.4	8	1.55
12	M	49	R	22.5	20	11.6	2.5	14.1	12.6	2.5	15.1	0.94
13	F	59	L	53.75	55	9.6	3.3	12.9	10.4	2.4	12.8	1.01
14	M	68	R	48.75	55	6.6	1.7	8.3	9.9	2.7	12.6	0.29
15	M	52	L	48.75	35	21.3	4	25.3	20.3	3.4	23.7	0.69
16	M	65	R	36.25	30	13.2	1.6	14.8	11.8	1.6	13.4	0.30
17	F	54	R	102.5	70	10.7	3.5	14.2	9.8	3.8	11.9	0.40
18	F	59	R	15	25	8.8	2.5	11.3	9.6	2.3	11.9	0.99
19	F	67	L	56.25	35	9.9	2.7	12.6	11.3	1.6	12.9	0.14
20	F	63	R	65	70	19.7	5.7	25.4	18.9	5.4	24.3	0.92
21	M	68	L	53.75	80	12.2	3.8	16	10.8	3.7	14.5	0.16
22	M	79	R	80	—	20.1	1.9	22	21.9	1.2	23.1	3.36
23	M	62	L	66.25	85	20.3	5	25.3	21.5	3.9	25.4	0.10
24	F	83	R	47.5	45	No PL signal decrease						0.009
25	M	73	R	15	15	No PL signal decrease						0.001
26	M	70	L	30	25	No PL signal decrease						0.03
27	M	65	R	25	25	No PL signal decrease						0.2
28	F	77	R	51.25	65	No PL signal decrease						0.04
29	F	70	R	63.75	80	No PL signal decrease						0.57
30	M	73	R	78.75	90	PL and EL signal decrease						4.51
31	F	55	R	27.5	35	PL and EL signal decrease						1.16
32	M	50	R	10	10	PL and EL signal decrease						1.25

VS: vestibular schwannoma; EH: endolymphatic hydrops; PTA: pure-tone average; SRT: speech recognition threshold; M: male; F: female; R: right; L: left; PL: perilymphatic; EL: endolymphatic.

The mean tumor volume of the 6 excluded patients with no perilymphatic signal decrease was 0.14 ± 0.22 (SD) cm^3 (range: 0.001—0.57 cm^3). These 6 patients had non-obstructive internal auditory canal VS. The 3 patients who were excluded due to the endolymphatic signal decrease (added to perilymphatic signal decrease, thus rendering impossible the distinction between endolymph and perilymph fluids [Fig. 1]) had a mean tumor volume of 2.30 ± 1.9 (SD) cm^3 (range: 1.16—4.51 cm^3). These three patients had obstructive VS.

In the 23 included patients, the mean saccular volume was 3.17 ± 1.1 (SD) mm^3 (range: 1.45—5.7 mm^3) and the mean utricular volume was 14.4 ± 5 (SD) mm^3 (range: 6.6—23.9 mm^3). The mean volume of the vestibular endolymphatic space was 17.45 ± 5.5 (SD) mm^3 (range: 8.3—25.4 mm^3). The ICC (for the saccular, utricular and vestibular endolymphatic volumes) were 0.867, 0.966 and 0.967, respectively (Fig. 3). The mean absolute deviations between the two radiologists for the saccular, utricular and vestibular endolymphatic volumes were 0.42 mm^3 (10.5%), 1.10 mm^3 (8%) and 1.03 mm^3 (5.1%), respectively.

Correlation between vestibular endolymphatic space volume and audiometric findings

The mean PTA level on the tumor side (57.9 ± 26.5 [SD] dB HL; range: 15—108.75 dB HL) was higher than that on contralateral side (25.7 ± 13.7 [SD] dB HL; range: 10—71.25 dB HL) ($P < 0.0001$). There were 3 patients with mild sensorineural hearing loss, 9 patients with moderate sensorineural hearing loss, 6 patients with severe sensorineural hearing loss and 3 patients with profound sensorineural hearing loss. Two patients had no hearing loss. SRT was available for all patients except one (22/23; 96%). The mean SRT on the

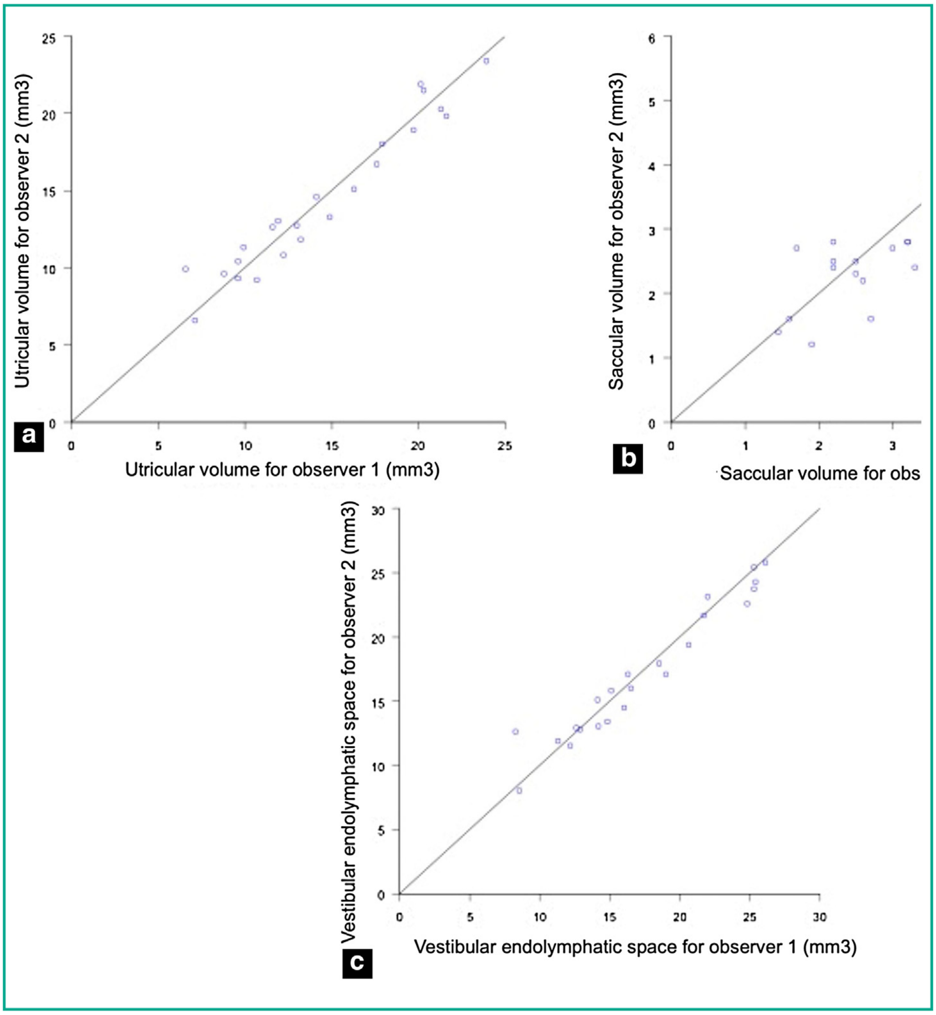

Figure 3. Diagrams show reliability of utricular (A) and saccular (B) and vestibular endolymphatic space (C) volume measurement between two independent radiologists. The intraclass correlation coefficients for the saccular, utricular and vestibular endolymphatic volumes are 0.867, 0.966 and 0.967, respectively.

tumor side (60.2 ± 31 [SD] dB SPL; range: 10—120 dB SPL) was higher than that of contralateral side (23.7 ± 13.3 [SD]; range: 5—65 dB SPL) ($P < 0.0001$).

In the 6 patients who were excluded due to no perilymphatic and endolymphatic signal decrease, the mean PTA and SRT were 23.3 ± 7.6 (SD) dB HL (range: 15—63.75 dB HL) and 42.5 ± 25.6 (SD) dB SPL (range: 15—80 dB SPL), respectively on the symptomatic ear. In the 3 excluded patients that presented both with endolymph and perilymph signal decreased, the mean PTA and SRT on the symptomatic ear were 38.75 ± 35.7 (SD) dB HL (range: 10—78.75 dB HL) and 45 ± 40.9 (SD) dB SPL (range: 10—90 dB SPL), respectively.

There was a moderate correlation between the utricular volume and the degree of hearing loss (Figs. 4, 5 and 6) as evaluated with the levels of PTA (rho = 0.5; 95% CI: 0.11, 0.76) ($P = 0.015$) and SRT (rho = 0.58; 95% CI: 0.21, 0.8) ($P = 0.004$), but not significant between the saccular volume and the levels of PTA ($P = 0.71$) and SRT ($P = 0.38$). There was also a significant correlation between the volume of the vestibular endolymphatic space and the degree of hearing loss as evaluated with the levels of PTA (rho = 0.42; 95% CI: 0.04, 0.70) ($P = 0.032$) and SRT (rho = 0.55; 95% CI: 0.20, 0.77) ($P = 0.004$).

Correlation between the tumor volume and audiometric findings

No significant correlation was found between tumor volumes and PTA levels (Rho = 0.20; 95% CI: −0.20, 0.54) ($P = 0.33$) and SRT (Rho = 0.22; 95% CI: −0.18, 0.56) ($P = 0.28$).

Discussion

In this study, we demonstrated that the volume of the vestibular endolymphatic space can be assessed with high reliability in patients with obstructive VS and that it moderately correlates with the degree of hearing loss as evaluated with the levels of PTA and SRT. The volume of the utricle correlates with the degree of hearing loss evaluated with the levels of PTA and SRT. On the other hand, the saccule volume does not correlates with the levels of PTA and SRT.

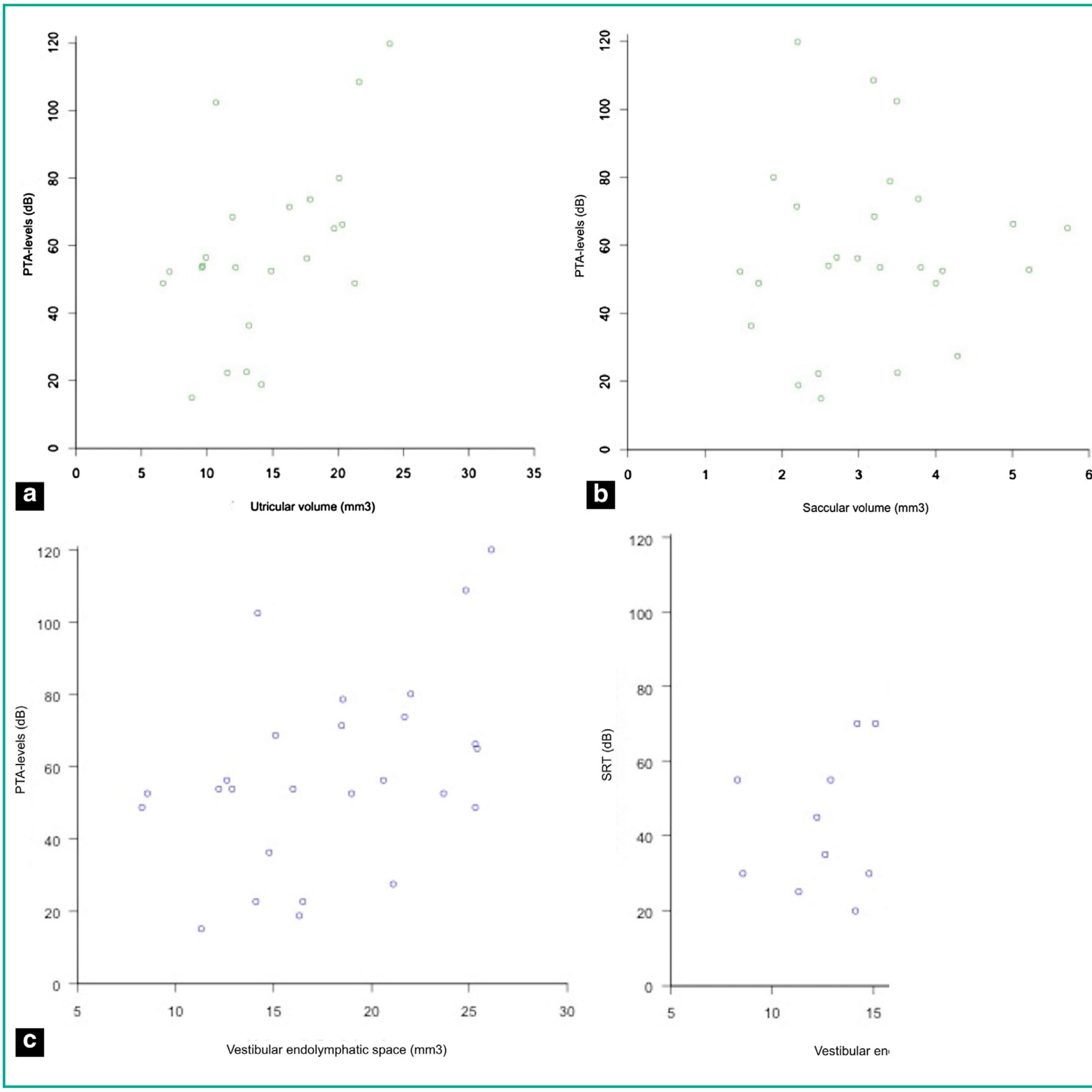

Figure 4. Graphs show correlation between the utricular (A), saccular (B) and vestibular endolymphatic space volumes and the hearing thresholds evaluated by pure-tone average (PTA) and speech recognition threshold (SRT). There is a moderate correlation between the utricular (PTA: rho = 0.5, $P = 0.015$; SRT: rho = 0.58, $P = 0.004$) and vestibular endolymphatic spaces (PTA: rho = 0.42, $P = 0.032$; SRT: rho = 0.55, $P = 0.004$) with the degree of hearing loss but not significant for the saccular volume ($P = 0.71$ for PTA and $P = 0.38$ for SRT).

On FIESTA-C imaging the contrast is based on T2/T1 ratios [11]. Thus, it is conceivable that the elevated protein content in the perilymphatic space results in a decreased perilymphatic signal by shortening both the T1 and T2 of the perilymphatic fluid. Venkatasamy et al. showed significant decrease in perilymphatic signal on FIESTA-C sequence, depending on the degree of obstruction of the internal auditory canal, potentially responsible for the failure of perilymphatic drainage [19]. In this study, there was no perilymphatic signal decrease in patients with non-obstructive schwannomas (17% of patients) [19]. Our results also support this theory, since 6 patients with non-obstructive schwannomas were excluded in our study due to the absence of a significant perilymphatic signal decreased on FIESTA-C. They had lower PTA and SRT levels than patients with obstructive schwannomas. Here, we demonstrate that FIESTA-C sequences enable the assessment of the utricle and the saccule in 72% of patients with obstructive VS and with a high spatial resolution and a reasonable acquisition time (4 minutes 40 seconds).

In our study, we excluded 3 patients with obstructive VS because they presented with both endolymphatic and perilymphatic signal decrease. We raise the hypothesis that in these patients the endolymphatic signal decrease was related to the proteinaceous precipitates involving the endolymphatic space, as it has been observed in only two histopathological studies that found a high rate of endolymphatic space involvement (8/12 ears and 11/32 ears) [3,9]. Interestingly, these 3 patients presented with lower PTA and SRT levels than patients with obstructive schwannomas with isolated perilymphatic signal decrease.

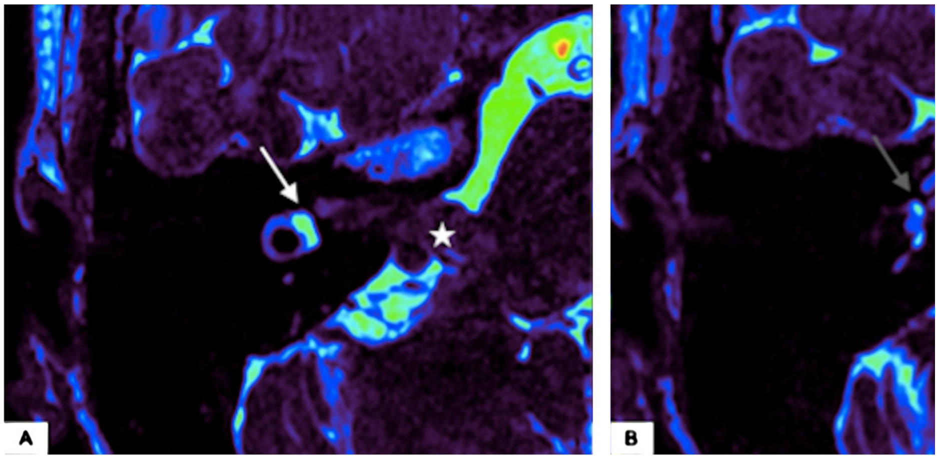

Figure 5. 57-year-old woman with a right vestibular schwannoma and conserved pure-tone average (22.5 dB HL) and speech recognition threshold (30 dB SPL). Fast imaging employing steady-state acquisition with cycle phase (FIESTA-C) image in the axial plane with the rainbow-scale on color map at the level of the utricle (A, white arrow) and the saccule (B, gray arrow). MR image shows schwannoma (asterisk) with a volume of 1.19 cm^3. The volume of the vestibular endolymphatic space is 16.5 mm^3.

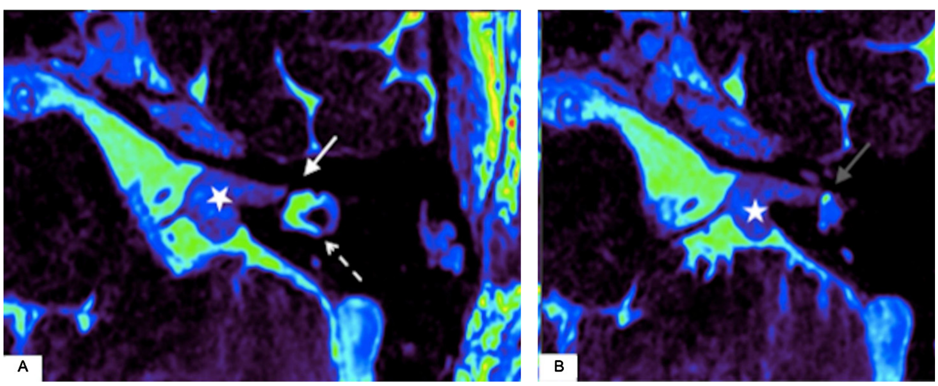

Figure 6. 63-year-old man with a left vestibular schwannoma and profound hearing loss (pure-tone average = 108.75 dB HL and speech recognition threshold = 120 dB SPL). Fast imaging employing steady-state acquisition with cycle phase (FIESTA-C) image in the axial plane with the rainbow-scale on color map at the level of the utricle (A, white arrow) and the saccule (B, gray arrow) shows left vestibular schwannoma (asterisk) with a volume of 0.76 cm^3. The utricle is dilated (21.6 mm^3), protruding in the lateral semicircular canal (dotted arrow) and the volume of the vestibular endolymphatic space is 26.1 mm^3.

Patients with VS mostly complain of progressive sensorineural hearing loss, low speech discrimination, tinnitus and balance disorders [25]. The neural degeneration induced by compression of the nerve is not sufficient to explain sensorineural hearing loss. Nadol et al. reported that hearing loss level does not correlate with the size of the tumor and that a subset of patients with VS have a relative preservation of speech discrimination with pure-tone sensorineural hearing loss [26]. Various additional mechanisms have been suggested including proteinaceous precipitates in the perilymph and EH [1,3]. It has been demonstrated that the elevated perilymphatic protein content is caused by obstruction of the cochlear aperture rather than by blockage of the neuroaxonal transport, breakdown of the blood-perilymph barrier and immune response to the tumor [9,19].

Patients with VS presented EH on histopathological examination involving the saccule in 50% of ears and the utricle in 40% of ears. However, Mahmud et al. have never reported EH in non-operated VS when speech discrimination was greater than 50% [1,9]. Schindler demonstrated that in patients with Menière's disease the endolymphatic sac presents changes similar to those in patients with schwannoma, suggesting that sac degeneration is the result of the disease potentially leading to EH [27]. Lee et al. suggested that the high signal observed in the vestibule with 3D-FLAIR sequences in patients with schwannoma could be related to the high protein concentrations in the perilymph, while the endolymph appears as dark signal intensity [12]. Due to the high signal of the perilymph causing by the increased protein content, Naganawa et al. assessed the rate of EH in patients with VS on non-contrast 3D-FLAIR images [17]. However, no correlation between the presence of EH and the hearing threshold was searched for.

To our knowledge, our study is the first to measure the volume of the saccule and utricle in vivo without contrast media injection in patients with schwannoma. Morita et al. measured the volume of each part of the membranous labyrinth on 31 normal temporal bones, with mean volumes and upper normal volume limits of the saccule and the utricle of 2.42 and 3.68 mm^3 and 10.65 and 16.45 mm^3, respectively [28]. The mean volumes of the saccule and utricle in our study were 3.17 and 14.4 mm^3 respectively, which is greater than those found by Morita et al. and close to their upper normal volume limits, suggesting EH in some patients with schwannoma.

Our study has several therapeutic implications. It has been reported that 75% of VS show no growth, leading to a ''wait and scan'' approach. In growing tumors, the mean increase in size varies between 2- and 4-mm per year [29]. The decision of surgery (destructive or conservative) or gamma knife therapy is based on the experience of the surgical team, the degree of hearing loss, the growth rate and the patient's morbidities [30]. Gamma knife radiosurgery is the first-line treatment in patients with small- to medium-size tumors and functional hearing while surgery is usually considered for large-size tumors [31]. Imaging can reveal EH associated with schwannoma in patients on conservative treatment to preserve hearing thus allowing the implementation of therapies such as diuretics to reduce EH. Indeed, it has been demonstrated on MRI that some patients with Menière's disease show improvement of symptoms and resolution of EH after having been treated with acetazolamide [32]. Recently, patients with schwannoma and disabling vertigo were treated with intratympanic gentamicin, which is presumed to reduce the production of endolymph and consequently EH [33].

Our study has several limitations. The vestibular endolymphatic space could not be evaluated in 28% of patients with VS on non-contrast FIESTA-C sequences either because of a lack of significant decrease in perilymphatic signal in cases with non-obstructive schwannoma or the decreased endolymphatic signal related to the high endolymphatic protein content [17]. The volume of the utricle and the saccule could not be evaluated on the contralateral side of the schwannoma since there was no perilymphatic signal decreased, thus we could not compare the volume of the endolymphatic space between both sides. Regarding the correlation between the utricular volume and hearing loss, the rho coefficient indicates a moderate correlation. However, we believe that the origin of hearing loss could be multifactorial, also implying perilymphatic protein content variation, as it has been previously speculated [13,31]. Interestingly, in our study the patients with non-obstructive schwannomas and no perilymphatic signal decreased presented lower PTA and SRT than patients with obstructive schwannomas. The main problem we encountered was the impossibility of confirming the tumor type and the volume of the vestibular endolymphatic space with pathological analysis in vivo. Another limitation is to what extent our results can be extrapolated to other T2-weighted echo-gradient sequences.

In conclusion, FIESTA-C sequence enables the assessment of the vestibular endolymphatic space in 72% of patients with a high reliability and its size is moderately correlated with the degree of hearing loss as evaluated with the levels of PTA and SRT. As most patients with schwannoma benefit from a ''wait and scan'' approach or a conservative treatment, based on the MRI results, therapies to reduce EH in order to improve hearing could be administrate.

Authors' contribution

Michael Eliezer and Arnaud Attyé: conceptualization, data curation, formal analysis, funding acquisition, investigation, methodology, project administration, resources, software, supervision, validation, visualization, writing.

Guillaume Poillon: data curation, investigation, methodology, resources, supervision, validation, visualization, writing.

Charles Maquet: funding acquisition, investigation, project administration, resources.

André Gillibert: data curation, formal analysis, software, validation.

Julien Horion: conceptualization, data curation, resources, software, visualization.

Jean-Paul Marie: funding acquisition, investigation, project administration, resources.

Jean-Pierre Guichard: validation, visualization, writing.

Nicolas Magne: formal analysis, funding acquisition, investigation, project administration, resources.

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Disclosure of interest

The authors declare that they have no competing interest.

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