



High resolution MRI shows presence of endolymphatic hydrops in patients still symptomatic after endolymphatic shunt surgery



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Abstract

Objective: Endolymphatic hydrops has been well described in patient's with Meniere's disease; however, causation has not been established. Decompression of the endolymphatic sac has been proposed as a means to relieve hydrops and improve vertigo symptoms, but the efficacy of the surgery is debated. Until recently, there have been few objective measures of efficacy other than patients' subjective symptoms. Recent archival human temporal bone studies have shown that patients continue to have hydrops after shunt surgery. We propose using high resolution MRI to determine the efficacy of endolymphatic shunt surgery (ELS) in patients who continue to experience vertigo.

Setting: Academic tertiary care referral center

Patients: Four patients presented with continued vertigo after unilateral endolymphatic shunt surgery. Mean age was 56 years old. Surgery was performed at two different institutions.

Interventions: Magnetic resonance imaging sequences included "cisternographic" three-dimensional T2, and delayed intravenous-enhanced three-dimensional fluid-attenuation inversion recovery (DIVE-3D-FLAIR) sequences, performed with 2350 ms (bright perilymph) and 2050 ms (bright endolymph) inversion times. The bright endolymph images were subtracted from bright perilymph images to create a composite image with bright perilymph, dark endolymph, and intermediate bone signals.

Main outcome measures: MRI finding of endolymphatic hydrops

Results: In all four patients who continued to experience severe vertigo, hydrops was found on high resolution MRI on the operated ear. The appearance on MRI was no different than in patients with EH who have not had surgery.

Conclusions: The present study demonstrates the persistence of endolymphatic hydrops in patients who have failed endolymphatic shunt surgery. Future studies evaluating for the presence or absence of endolymphatic hydrops in patients who claim to obtain relief from endolymphatic shunt surgery.

Introduction

Since its initial description by Prosper Meniere in 1861, Meniere's Disease (MD) has presented a unique therapeutic challenge for the otologist¹. Many medical and surgical therapies have been tried, based on different proposed mechanisms. Endolymphatic hydrops (EH) is an anatomic finding in which the structures bounding the endolymphatic space are distended by enlargement of the endolymphatic volume². This finding was first correlated with Meniere's disease (MD) in the 1930s³⁻⁴, leading researchers to believe that EH is causative of Meniere's. Therefore, several procedures were proposed to decrease the endolymphatic volume in order to treat MD.

Georges Portmann first described surgery to decompress the endolymphatic sac in 1927⁵. Since then, endolymphatic sac decompression and endolymphatic shunt surgery have become commonplace, with a recent systematic review reporting success in controlling vertigo in over 75% of patients⁶. However, recent research has shown that the relationship between EH and MD is not a straightforward causation. In fact, it appears a multitude of insults to the inner ear results in hydrops, and the presence of hydrops alone does not always cause MD symptoms⁷⁻⁸.

Additionally, research in the effectiveness of treatment for MD is limited by reliance on subjective symptoms and high placebo response rates. In fact, in one of the only randomized controlled trials of endolymphatic sac surgery, no difference was seen between decompression and sham surgery, with patients in both arms reporting significant improvement postoperatively⁹. Most studies of treatment for MD report high success rates, even with some interventions such as tympanostomy tubes that have no pathophysiological explanation for effectiveness¹⁰.

A recent examination of temporal bones of patients who had undergone endolymphatic sac surgery showed that out of 15 temporal bones examined, all continued to exhibit hydrops¹¹. In fact, many of the specimens demonstrated that the surgery failed to access the endolymphatic sac at all. Despite this, a majority of the patients experienced symptomatic improvement after surgery.

The advent of new imaging techniques, allowing for the differentiation between perilymph and endolymph in the inner ear, have allowed for evaluation for EH in live patients¹²⁻¹³. This presents an objective measure of the degree of hydrops in patients with MD. Here we present a series of four patients who previously underwent endolymphatic shunt surgery, but continued to be severely symptomatic, and investigate the status of the endolymphatic sac with new MR techniques.

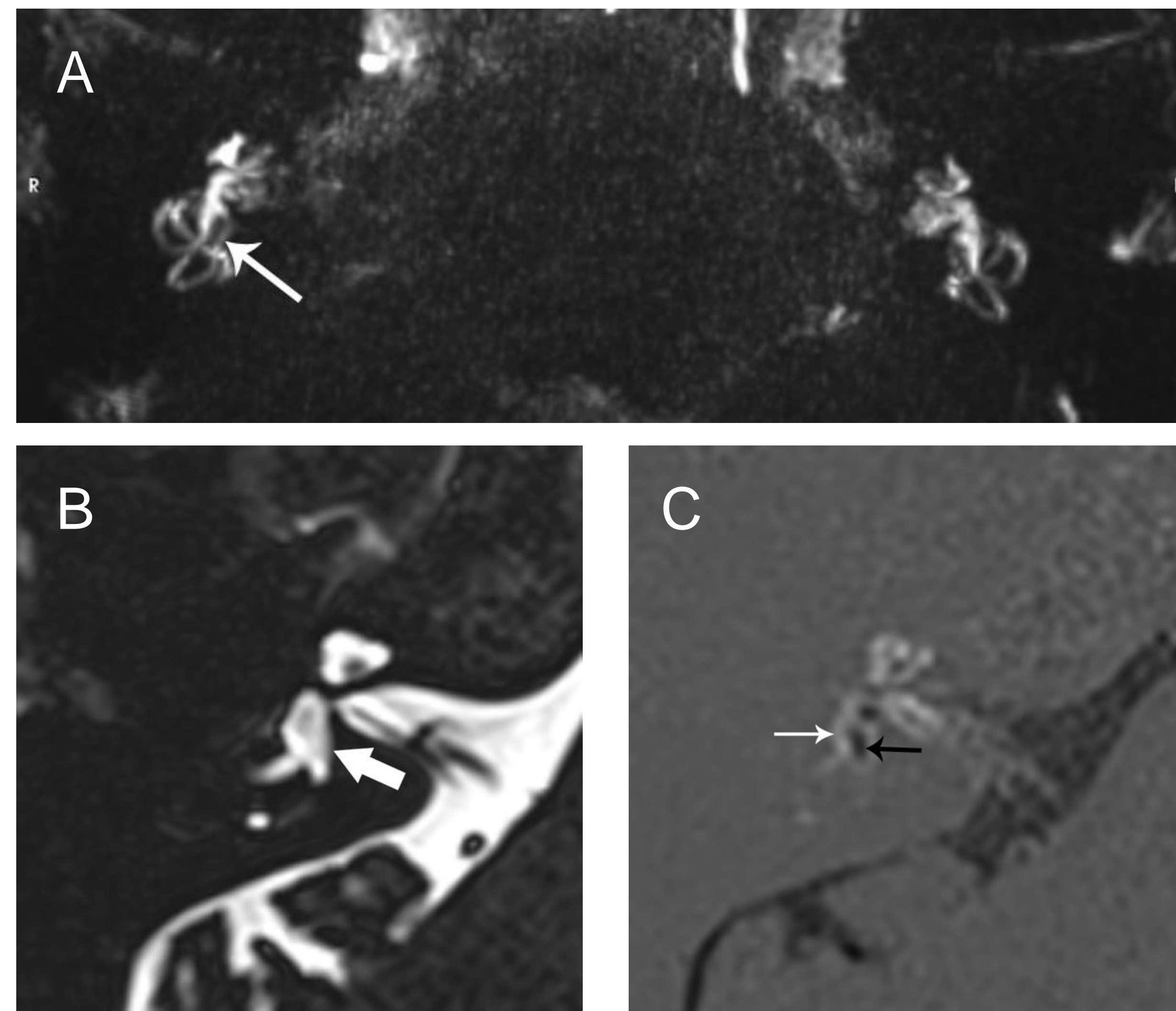


Figure 1. DIVE-3D-FLAIR MR imaging of a normal patient. A. Maximum intensity projection image shows the vestibular endolymph as a small signal void surrounded by bright perilymph (arrow). B. Standard cisternographic T2 image where all the fluid in the inner ear is bright, including both endolymph and perilymph. C. Subtracted image shows black endolymph surrounded by white perilymph.

Materials and Methods

Imaging was done on a 3-T scanner (Skyra, Siemens Healthcare, Erlangen, Germany) using a 12-channel head coil, 4 hours after administration of 0.2 mmol/kg gadobutrol intravenous contrast (Gadavist, Bayer HealthCare). Imaging consisted of three sequences: 1) "Cisternographic" 3D turbo spin echo T2; 2) "Perilymph bright, endolymph dark" heavily T2-weighted (hT2w)-3D-FLAIR, obtained with an inversion time of 2350 ms, 3) "Endolymph bright, perilymph dark" hT2w-3D-FLAIR, obtained with inversion time of 2050 ms. All sequences were acquired in the axial plane along the infraorbitomeatal line, as three dimensional volumetric scans. The cisternographic T2 was obtained with 0.3 x 0.3 x 0.3 mm isotropic voxels. The 3D-FLAIR sequences were obtained with 0.8 mm isotropic voxels. The endolymph bright hT2w-3D-FLAIR images were subtracted from the perilymph bright hT2w-3D-FLAIR images, in order to obtain an image with bright perilymph, dark endolymph, and intermediate signal bone. The cisternographic T2 was used to assist with anatomic reference.

Results

A 58-year-old female presented with a 5 year history of episodic vertigo, nausea, vomiting, and tinnitus in the right ear. The episodes were occurring several times a week and lasting 8-9 hours each time. A year prior to presentation, the patient had a shunt placed in the right ear, but she continued to have the same symptoms. MRI demonstrated hydrops on the right side (figure 2). The patient subsequently underwent right retrosigmoid vestibular neurectomy, and 6 months postoperatively reports complete resolution of vertigo and improvement in hearing.

A 44-year-old male presented with a two year history of episodic vertigo. He underwent ELS without improvement in his symptoms. He underwent MRI showing left-sided hydrops. He tried multiple medications and had brief relief of his symptoms with medical management. While his symptoms were controlled, he underwent an MRI, which showed resolution of hydrops. However, his symptoms returned, and he opted for a vestibular neurectomy. One year after a left-sided vestibular neurectomy, the patient is doing well without suffering any further vertigo attacks after surgery.

A 60-year-old male presented for intractable recurrent episodic vertigo who previously underwent ELS. Postoperatively, he noted profound sensorineural hearing loss in that ear. Quantitative vestibular testing demonstrated unilateral canal paresis. MRI demonstrated presence of hydrops on the operated ear. Due to his persistent symptoms, he elected to proceed with transmastoid labyrinthectomy. He has been fully asymptomatic since the surgery without any further vertigo spells.

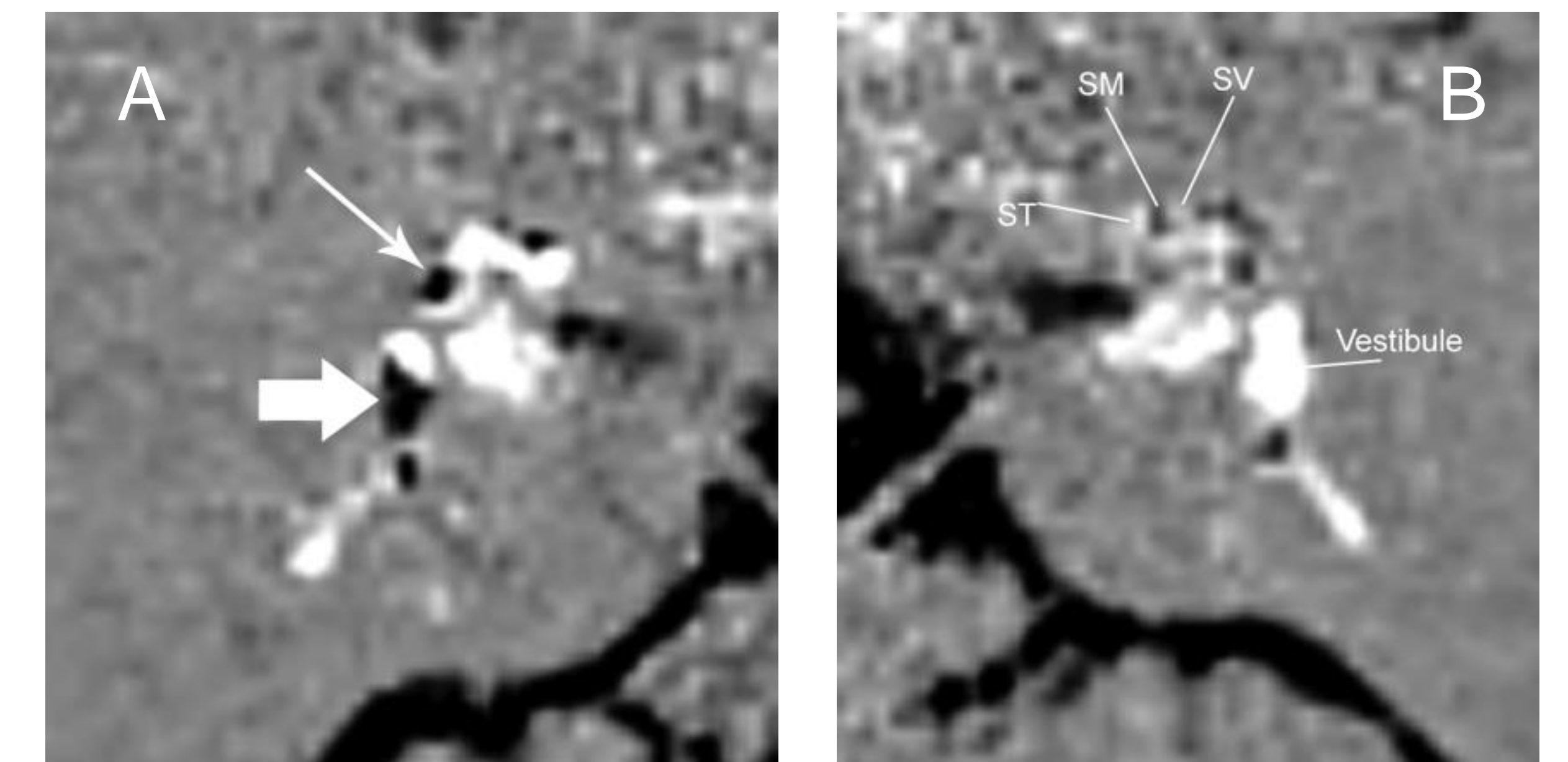


Figure 2. DIVE-3D-FLAIR MR imaging of one affected patient. Both images represent the subtracted images showing black endolymph surrounded by white perilymph. A. Affected right inner ear showing dilated endolymphatic space (black regions with arrows pointing to them). B. Normal left inner ear. ST – scala tympani. SM – scala media. SV – scala vestibuli.

Results

A 62-year-old female underwent ELS for recurrent episodic vertigo spells. Despite the surgery, she continued to experience recurrent episodic vertigo spells. She was placed on Diamox but she demonstrated no improvement. MRI showed the presence of endolymphatic hydrops on the left ear. Patient subsequently underwent retrosigmoid vestibular neurectomy and has been asymptomatic for several years.

Discussion

- Endolymphatic sac surgery has been shown to have high success rates in relieving symptoms of patients with MD.
- Most interventions for Meniere's Disease have shown high success rates, possibly due to placebo effect.
- A recent temporal bone study showed that all patients continued to have hydrops after endolymphatic sac surgery, even though the majority of them experienced symptomatic relief; additionally, successful access to the endolymphatic sac did not correlate with symptom relief.
- New MRI techniques are allowing an ability to objectively diagnose hydrops in live patients.
- This series of 4 patients showed that patients who continued to be symptomatic after shunt surgery continued to have hydrops.
- One explanation could be that the endolymphatic sac is difficult to access, and even with good surgical technique, the actual sac may not be truly accessed.
- Though the number is few, the patients in this series improved after vestibular nerve section or labyrinthectomy.
- Further studies are needed to compare these interventions.

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