

Cardiac diseases occur in 51% of patients of with CAPS. Myocardial infarction, valvular heart disease, and arrhythmias are the most common. Libman-Sacks endocarditis occurs in 6% of patients, involving sterile fibrin-plated thrombi vegetations, which have a propensity for the ventricular side of the mitral valve (3,4).

The mainstay of APS management is heparin for at least 5 days after initial venous thrombosis and long-term warfarin treatment, although the duration and dosage of treatment is debated. Researchers are investigating the efficacy of newer anticoagulants. The most effective combination treatment for CAPS, with a 77.8% recovery rate, includes anticoagulation, corticosteroids, and plasma exchange (1). Physicians should be aware that patients at risk for CAPS after interventional procedures may have an unusual presentation. Early hematology and rheumatology consultations should be sought.

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Truncular Venous Malformations in Ménière Disease Patients: The Key for a New Treatment Perspective?

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Editor:

Until more recently, only the arterial blood components of the cerebrospinal nervous system were assessed, with no attention given to the venous system and its possible role in some cerebrovascular disorders. The initial study by Zamboni et al (1) uncovered the Pandora’s box,

noting the importance of the venous system and identifying the chronic cerebrospinal venous insufficiency syndrome. This syndrome was initially found to cause altered brain venous outflow in patients with multiple sclerosis. Although it has been the subject of controversial debate, this syndrome has opened up the way to assessment of a possible common etiopathogenetic mechanism in patients with other types of diseases.

Ethics board approval was obtained for this study. A 39-year-old woman with frequent objective dizziness associated with highly disabling autonomic phenomena (nausea, vomiting, loss of anal sphincter control), significant and troublesome tinnitus, and auricular fullness was admitted to the Audiology Department of our hospital. She reported a 2-year history of significant and severe progressive hearing loss (especially in the right ear). On examination, she had no spontaneous nystagmus either in the classic positions or after mastoid vibration or hyperventilation test. There were no neurologic deficits, and no dysmetria was demonstrated on finger-to-nose and heel-to-shin testing. Audiometry examination showed right sensorineural hearing loss with pure-tone audiometry thresholds of 50 dB, suggesting cochlear involvement; Weber test lateralized to the right. Based on the clinical criteria proposed by the American Academy of Otolaryngology in 1995 (2), a diagnosis of Ménière disease was made. Cerebral computed tomography and magnetic resonance imaging scans were negative. The patient was treated with betahistine for 60 days with no benefit. Follow-up audiography performed at 6 months showed persistent hearing loss on the right side.

Cerebrospinal outflow color Doppler ultrasound (US) evaluation of the vertebral veins and internal jugular veins (IJV) was performed to assess five hemodynamic criteria of chronic cerebrospinal venous insufficiency. Criteria 1 and 3 were identified: on the left side, a small IJV caliber with bidirectional flow and an immobile septum in segment J1 (Fig 1a), and on the right side, an annulus constriction in segment J2 with bidirectional flow (Fig 1b). Selective venography of both IJVs confirmed the anomalies detected during the color Doppler US examination. In the left IJV, caliber reductions in segment J2 and septum in segment J1 were treated with high-pressure balloon dilation (12-mm-diameter and 40-mm-long balloon for J2 segment and 16-mm-diameter and 40-mm-long balloon for J1 segment) using a MAXI LD dilation catheter (Cordis, Johnson & Johnson, Miami, Florida). In the right IJV, the severe annular constriction (Fig 2a) was treated by 16-mm-diameter and 40-mm-long balloon (MAXI LD) dilation with incomplete expansion at 10 atm because of a resistant annular J2 stenosis. At the end of the procedure, the immediate sensation felt by the patient was the disappearance of auricular fullness and tinnitus. Completion venography demonstrated partial resolution of the annular stenosis (Fig 2b) and improved cerebral outflow confirmed by

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Table E1 is available online at www.jvir.org.

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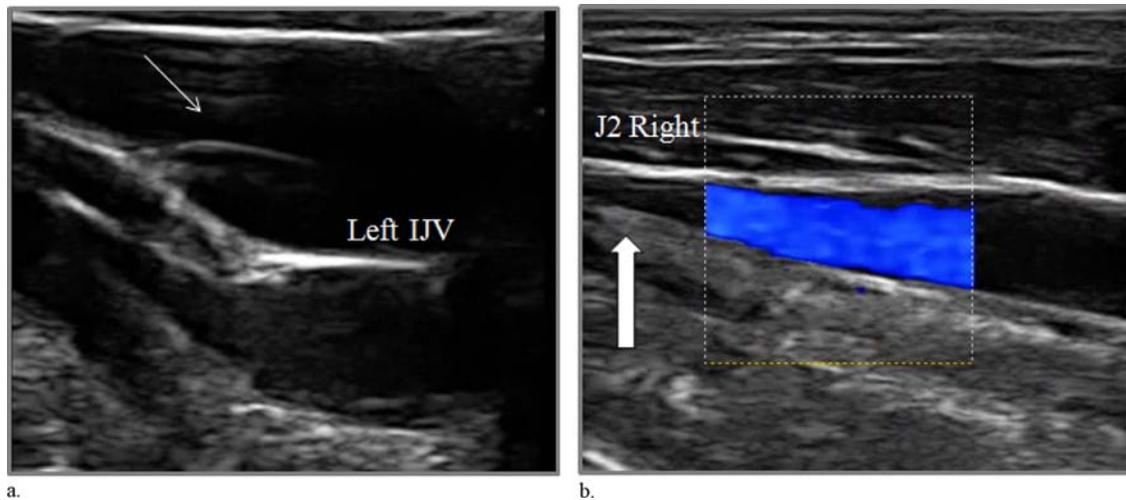


Figure 1. Color Doppler US analysis of the vertebral veins and IJVs, assessing five venous hemodynamic criteria of chronic cerebrospinal venous insufficiency. **(a)** Immobile septum in the J1 segment of the left IJV (arrow). **(b)** Annulus constriction in the J2 segment of the right IJV (arrow).

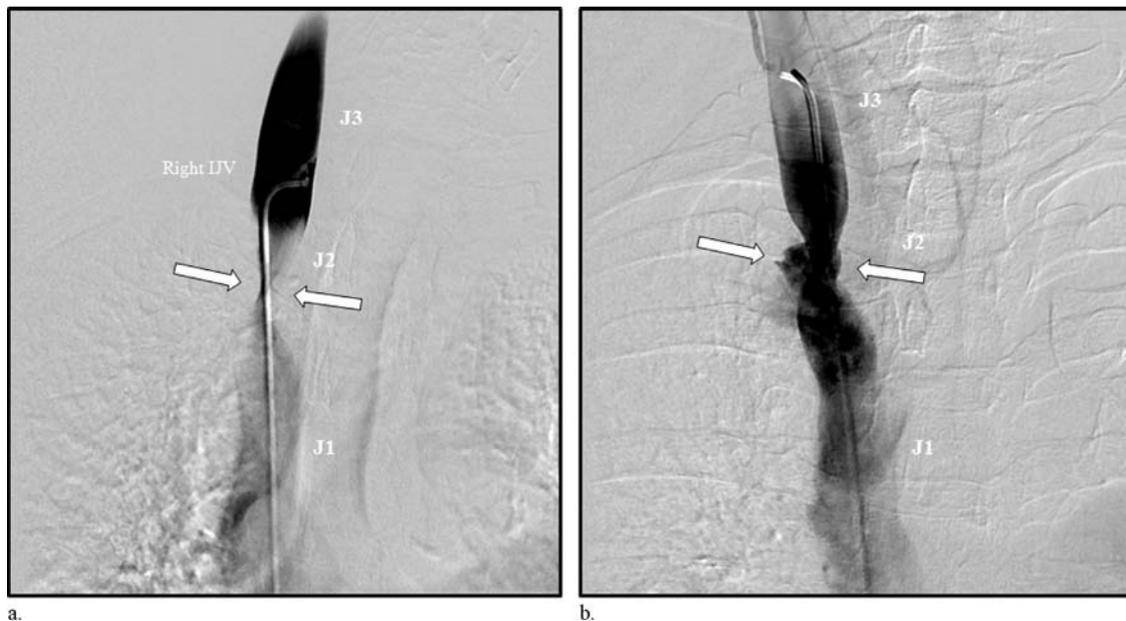


Figure 2. Selective venography of the right IJV. **(a)** Severe annular constriction in the J2 segment of the right IJV (arrows). **(b)** Partial reduction of annular constriction after 16/40 mm balloon dilation (arrows).

final color Doppler US analysis, which was performed again at the 1-month follow-up evaluation. An 8-month follow-up by the Audiology Department highlighted improvement in her quality of life as a result of both a subjective improvement (ie, disappearance of tinnitus, vertigo episodes, and auricular fullness) and an objective improvement on the audiometric examination (**Fig 3**).

Truncular venous malformations are the result of developmental anomalies of the vascular trunk occurring during embryogenesis and related to chronic cerebrospinal venous insufficiency syndrome. This condition has unclear pathophysiologic significance that has led to a controversial debate. However, doubts regarding treatment efficacy are likely related to correct diagnosis and

US analysis: this examination is strictly operator dependent because many practical measures have to be carefully taken into consideration (ie, regarding patient position, breathing, hydration, and neck position) (**Table E1** [available online at www.jvir.org]).

Different findings in several studies have led to difficult interpretation of the results. A possible explanation of these discrepancies reflects differences in operator learning curves, methodology, and interpretative skills of color Doppler US assessment of the extracranial and intracranial veins, which is the optimal screening tool for chronic cerebrospinal venous insufficiency. These anomalies have a possible impact on the cerebral vascular system; retrograde venous hypertension in patients with multiple

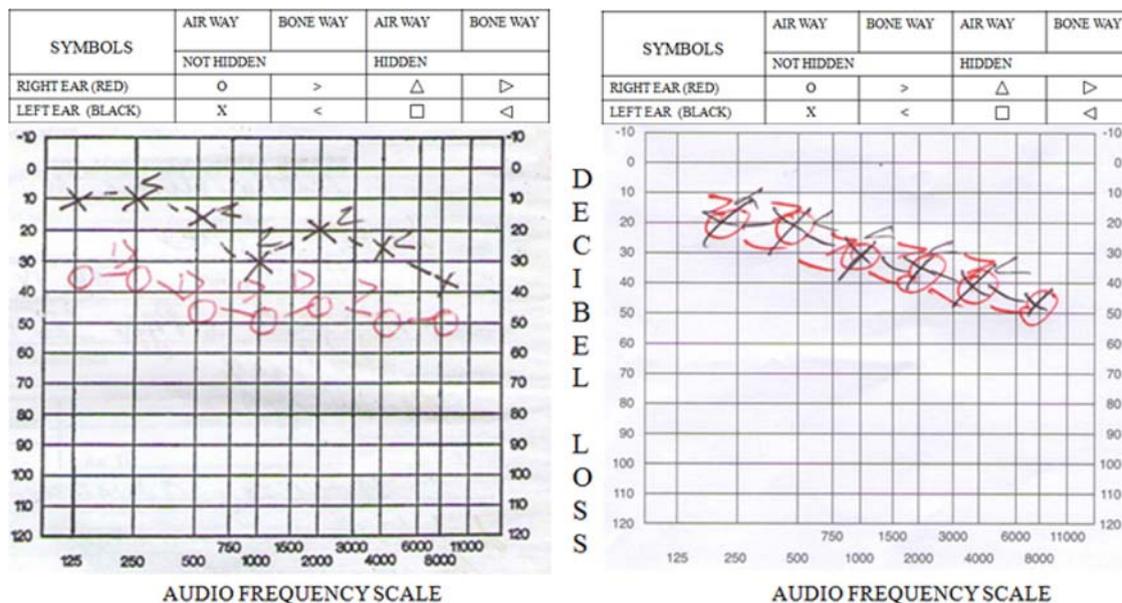


Figure 3. Audiometry examination of the right ear (red signals) and left ear (black signals) via air and bone impulse transmission. In the left panel (before percutaneous transluminal angioplasty), pure-tone audiometry shows a low sensorineural hearing loss on the left side and medium sensorineural hearing loss on the right side. In the right panel (after percutaneous transluminal angioplasty), pure-tone audiometry shows bilateral low sensorineural hearing loss (improvement on the right side).

sclerosis has been suggested to be due to an anomalous cerebral outflow, and increased cerebral blood flow pulsatility has been linked with microstructural white matter damage (3).

These anomalies not only may involve patients with multiple sclerosis but also may involve patients with audiologic symptoms with the same etiopathogenetic mechanism, and the primary mechanism may be an endolymphatic hydrops caused by an excess of endolymphatic volume, secondary to chronic reduced or altered venous drainage of the anterior and posterior vestibular veins or of the cochlear veins into the venous cerebrospinal system (IJVs and vertebral veins) (4). Patients with Ménière disease have poor response to conventional treatment therapies (eg, betahistine, steroids, diuretics, loop diuretics, osmotics, vasoactive drugs), with persistent dizziness, spells of acute vertigo, hypoacusis, ear fullness, and tinnitus resulting in a poor quality of life.

In recent years, percutaneous transluminal angioplasty procedures on IJVs and the azygos vein of patients with truncular venous malformations have become widespread, producing good results and low morbidity. In our experience, this endovascular treatment appears safe and appropriate in patients with Ménière disease when a correct diagnosis is obtained, with a final reassessment of the vascular venous neck and brain circulation and an improved cerebral outflow leading to major benefit in these patients and allowing an improvement in quality of life in subjective terms (ie, disappearance or reduction of tinnitus, vertigo episodes, and auricular fullness) as well as an objective

improvement in audiometric examination. This option offers new treatment possibilities.

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Transhepatic Arterial Approach for Successful Embolization of Hepatic Hilar Pseudoaneurysm Fed by Tortuous Collateral Vessels

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Table E1. Color Doppler US Examination Recommendations

Skill and experience of operator	The operator's training level is a key aspect of the color Doppler US examination. The operator's ability to interpret the images without influencing their acquisition (an adequate gel apposition is required to avoid venous compression during the examination) is important.
Patient position	Both positions (supine and upright) need to be comfortable for the patient avoiding muscle contractions and muscle compression, which may falsify the venous flow.
Head position	The head should be placed in a natural position, looking upward and forward, without neck rotation.
Breath	The patient should breathe normally, preferably through the nose.
Hydration	It is mandatory for the patient to be adequately hydrated before the color Doppler US examination to analyze the correct vessel caliber.