



2025 Intracranial Hypotension Conference

What to do: Brain MRI + SIH without detectable spinal CSF leak

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USA



The clinical dilemma

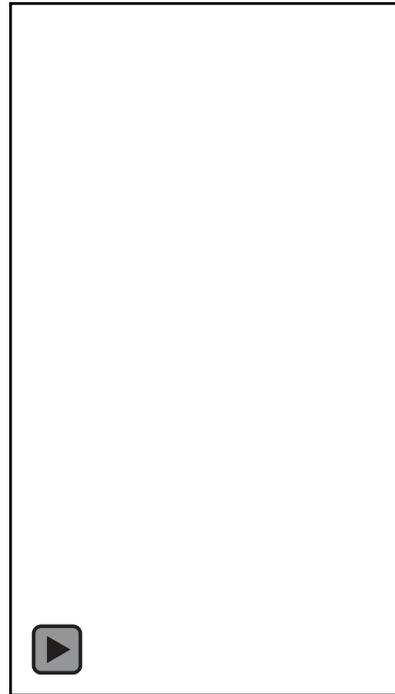
- Patient with brain MRI typical of SIH
- Normal spine MRI/MR-myelogram/conventional CT-myelogram and normal DSM/dynamic CT-myelogram

Reasons for normal spine imaging in CSF deficiency

1. Skull base CSF leak

CSF leaks as a cause of SIH

- **Always spinal!**



Lack of causal association between spontaneous intracranial hypotension and cranial cerebrospinal fluid leaks

Clinical article

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FRANKLIN G. MOSER, M.D., M.M.M.,³ AND TODD D. ROZEN, M.D.⁴**

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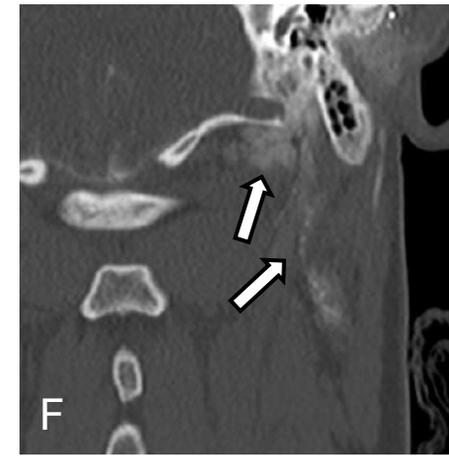
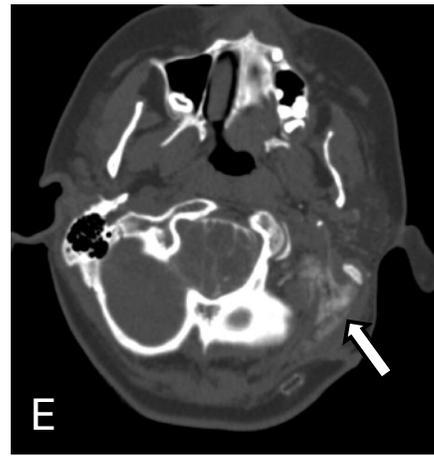
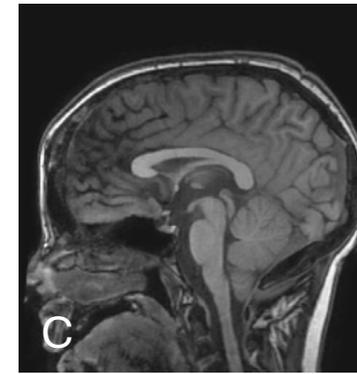
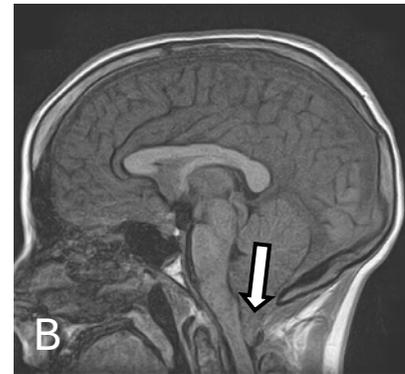
- CSF rhinorrhea?

NEVER!

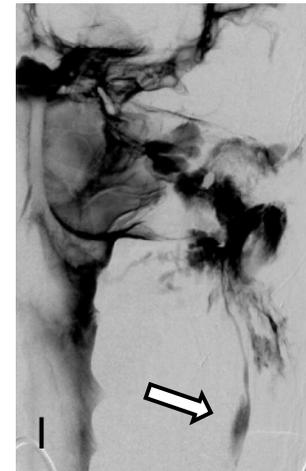
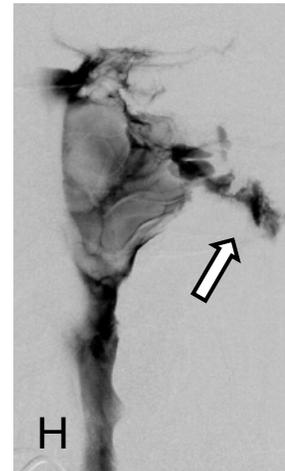
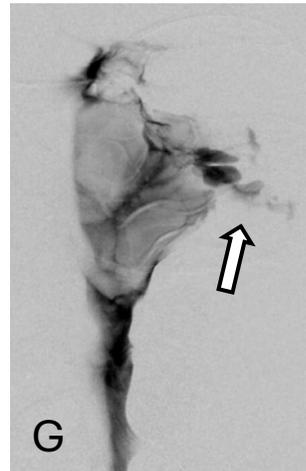
Spontaneous Intracranial Hypotension Due to Skull-Base Cerebrospinal Fluid Leak

Wouter I. Schievink, MD¹, L. Madison Michael II MD,^{2,3,4} Marcel Maya, MD,⁵
Paul Klimo Jr MD,^{2,3,4} and Lucas Elijovich, MD^{2,3,4,6}

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An Official Journal of
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Association and the
Child Neurology Society



13-year-old boy



Spontaneous Intracranial Hypotension Due to Cerebrospinal Fluid Rhinorrhea

Wouter I. Schievink, MD; Marcel M. Maya, MD; William H. Slattery, MD

An 8-year-old girl was seen in our clinic for cerebrospinal fluid (CSF) rhinorrhea, orthostatic headaches, and an acquired Chiari malformation. She had experienced 2 episodes of bacterial meningitis since age 6 years. The first episode of meningitis was complicated by cerebral edema with uncal herniation and a blown pupil requiring a decompressive craniectomy and subsequent autolo-

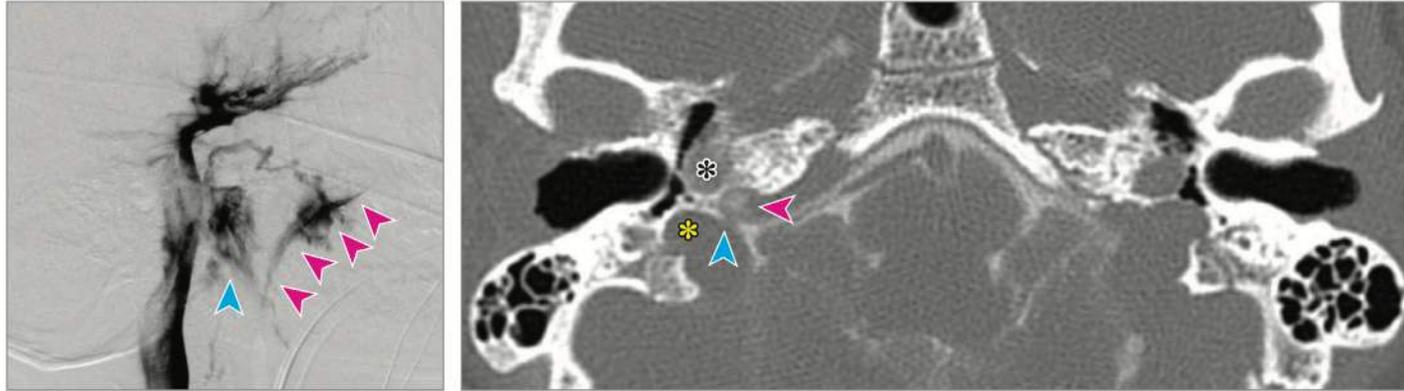
gous cranioplasty. This first episode of meningitis was due to *Streptococcus pneumoniae*, serotype 16F. The second episode of meningitis was due to nontypeable *Haemophilus influenzae*. She was diagnosed with an acquired Chiari malformation and underwent a decompressive craniectomy, cervical-1 laminectomy, and resection of the cerebellar tonsils at age 7 years. An underlying CSF leak was suspected because of the rhinorrhea and acquired Chiari malformation. A computed tomography (CT) cisternogram showed extensive amounts of contrast in the pterygoid muscles, soft palate,

+
Multimedia
+
CME at jamacmelookup.com

Figure 2. Digital Subtraction Myelography Showing Cerebrospinal Fluid Rhinorrhea Originating From the Posterior Cranial Fossa and Computed Tomography (CT) Scan Showing Skull Base Defect

A Lateral digital subtraction myelogram

B Axial postdigital subtraction myelogram CT

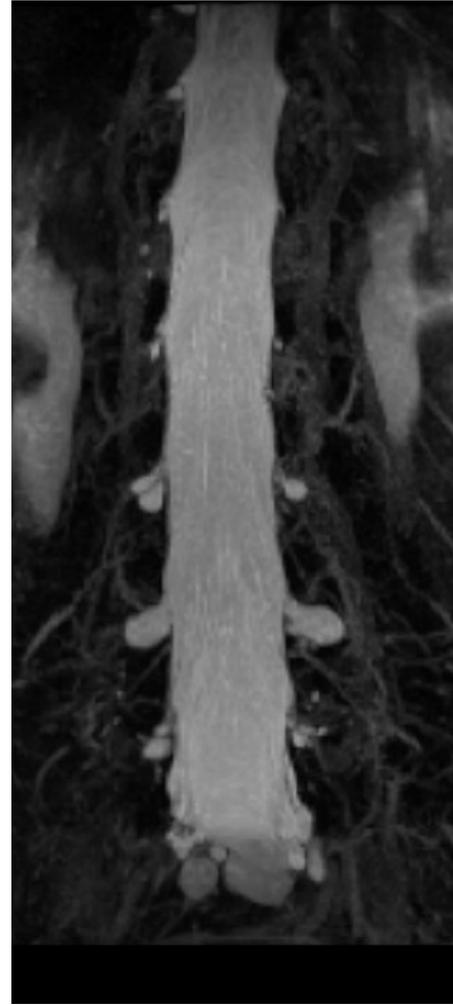
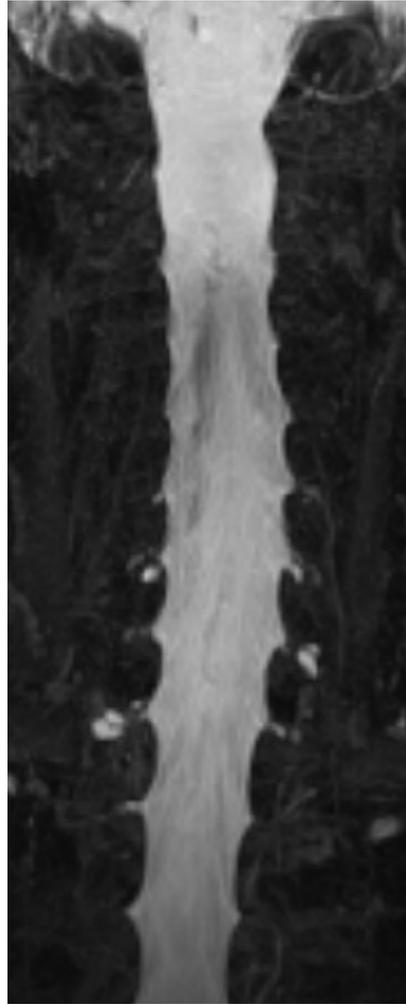
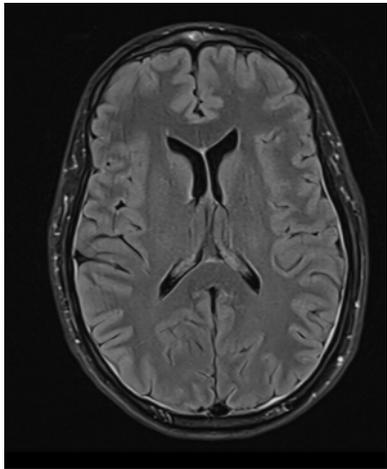
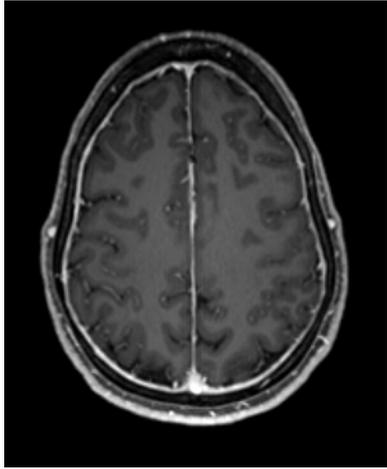


A, Lateral digital subtraction myelogram showing a high-flow cerebrospinal fluid leak from the posterior cranial fossa into the nasopharynx (pink arrowheads) and soft tissues of the neck (blue arrowhead). B, Axial postdigital subtraction myelogram CT scan showing focal sigmoid dehiscence (blue arrowhead) with adjacent focus of contrast accumulation and cerebrospinal fluid leak (pink arrowhead). Note the petrous internal carotid artery (black asterisk) and jugular vein (yellow asterisk).



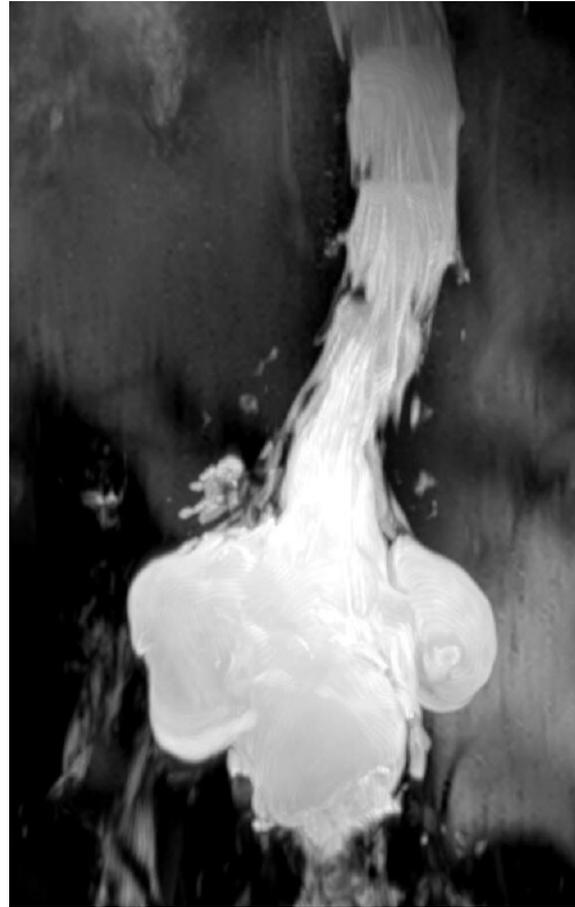
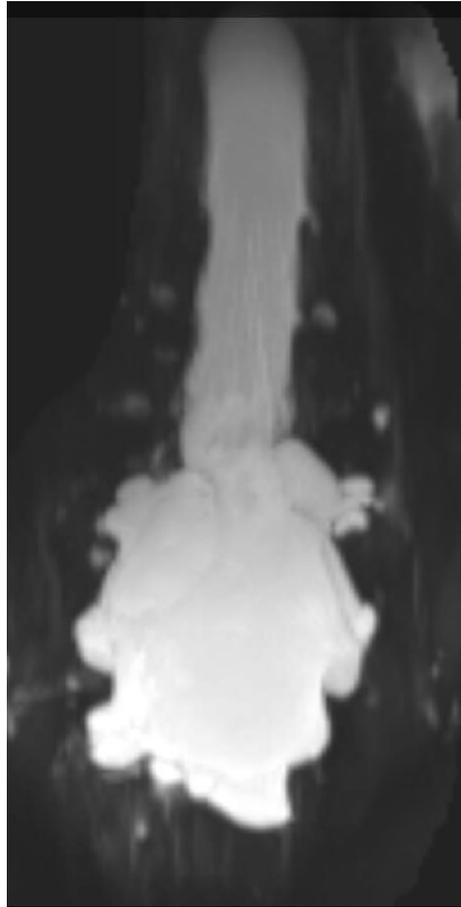
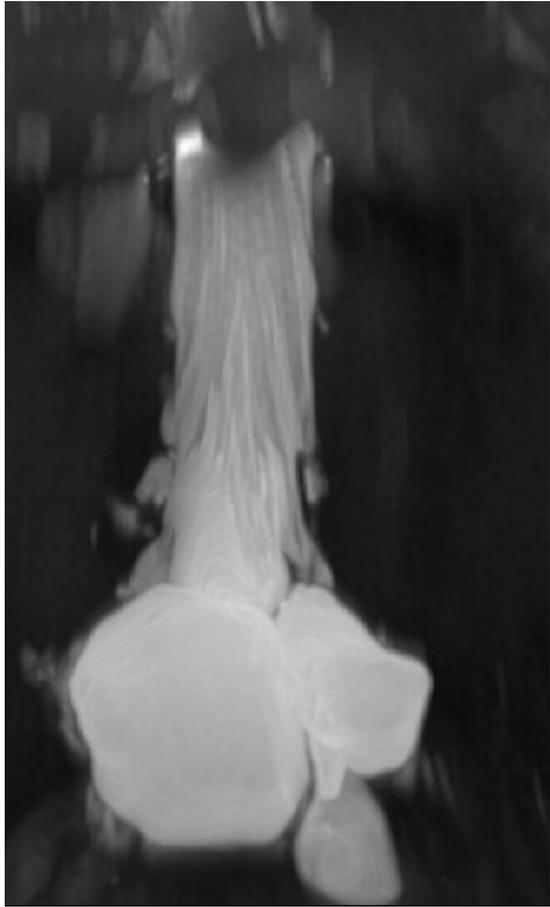
Reasons for normal spine imaging in CSF deficiency

1. Skull base CSF leak
2. “Pooling” of CSF in multiple diverticula/dural ectasia



DSMx6 and
PCCT all normal

Dural ectasia in Marfan and Loeys-Dietz syndrome

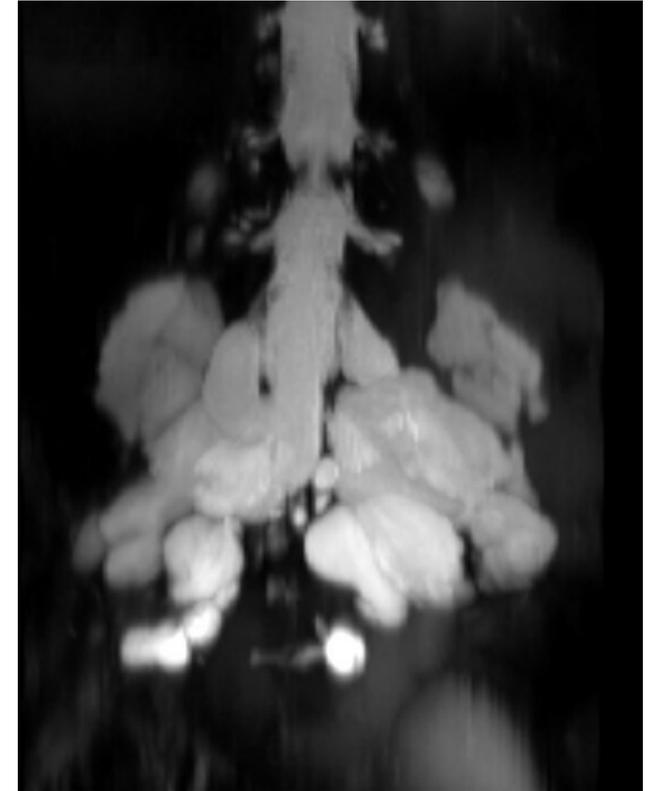


Dural ectasia in patients with tall stature

7'2" (218.5 cm) man



6'6" (198 cm) woman



Intracranial Hypotension Symposium 2025

Amsterdam, the Netherlands, June 28-29



WHAT TO DO?

Persistent headache after CSF leak resolution

1:30PM - 1:50PM

Katharina Wolf, MD

Brain MRI + SIH without detectable CSF leak

1:50PM - 2:10PM

Wouter I. Schievink, MD

Dural ectasia and orthostatic headaches

2:10PM - 2:30PM

Marcus Stoodley, MD, PhD



Reasons for normal spine imaging in CSF deficiency

1. Skull base CSF leak
2. “Pooling” of CSF in multiple diverticula/dural ectasia
3. Small lateral dural CSF collections

The importance of small lateral dural CSF collections in spontaneous intracranial hypotension: A radiologic-anatomic study

Wouter I. Schievink, M.D.,¹ Marcel M. Maya, M.D.,² Andre E. Boyke, M.D.,¹

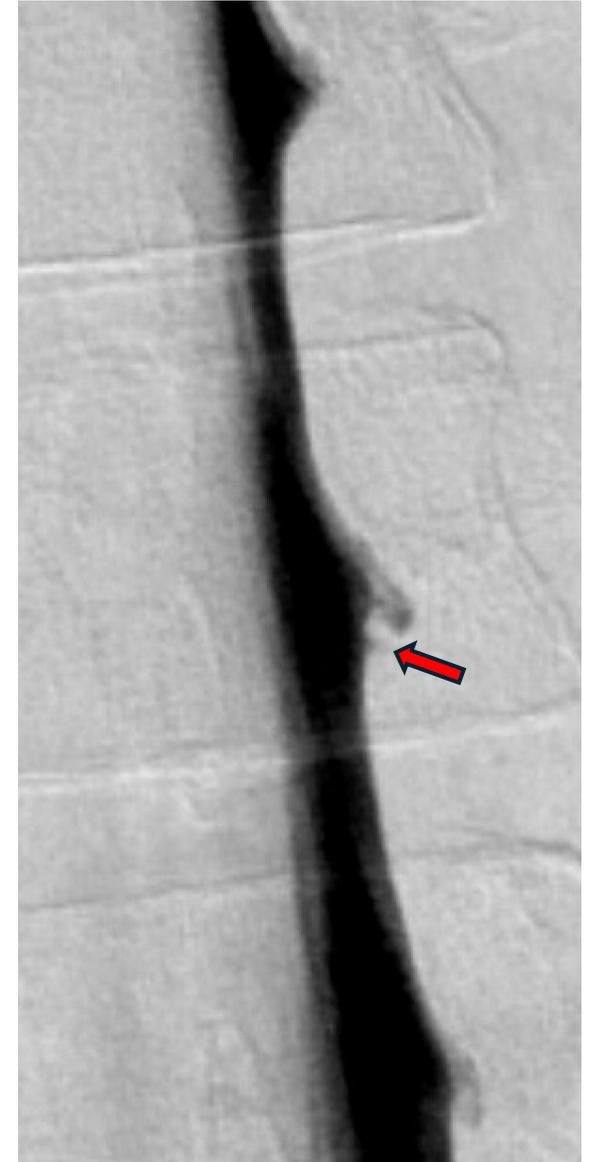
Franklin G. Moser, M.D.,² Ravi S. Prasad, M.D.,² Vikram Wadhwa, M.D.,²

and Xuemo Fan, M.D.³



The importance of small lateral dural CSF collections in spontaneous intracranial hypotension: A radiologic-anatomic study

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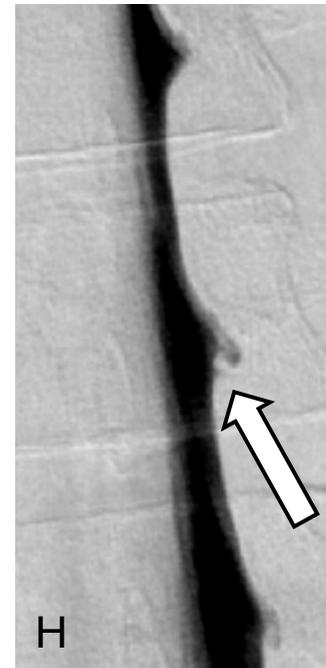
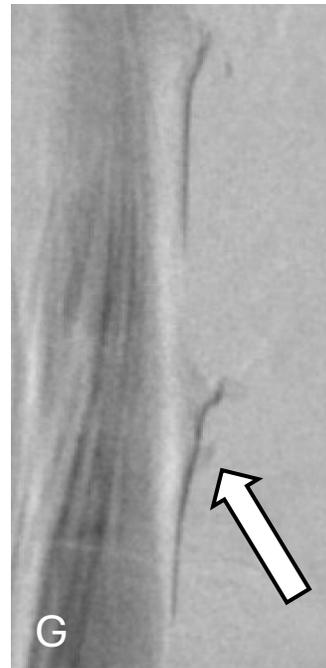
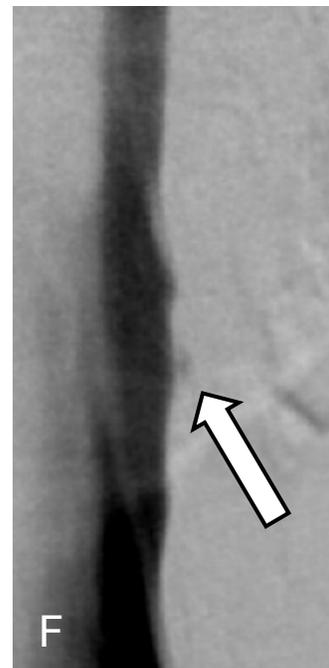
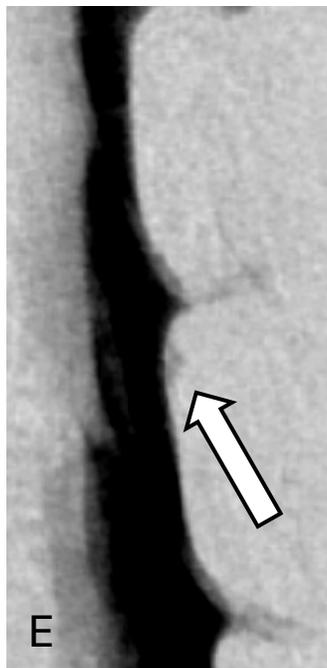
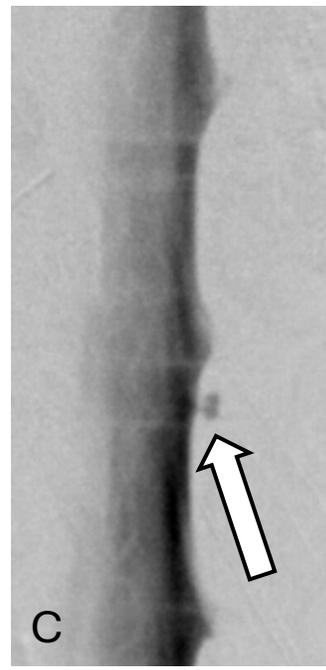
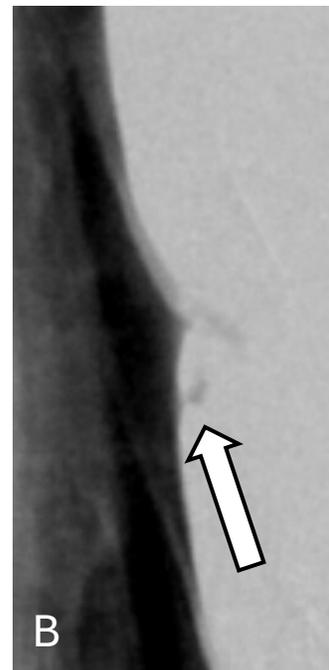
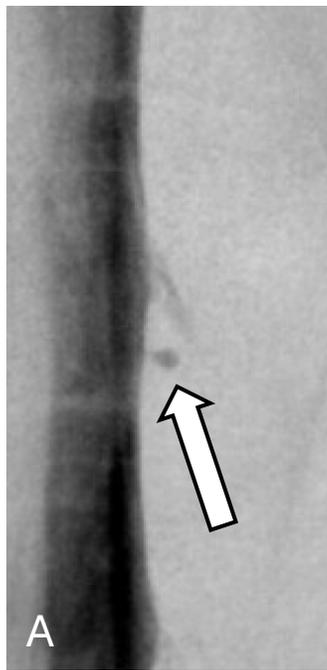
31 patients with small lateral dural CSF collections on DSM

4 patients did not undergo surgical exploration

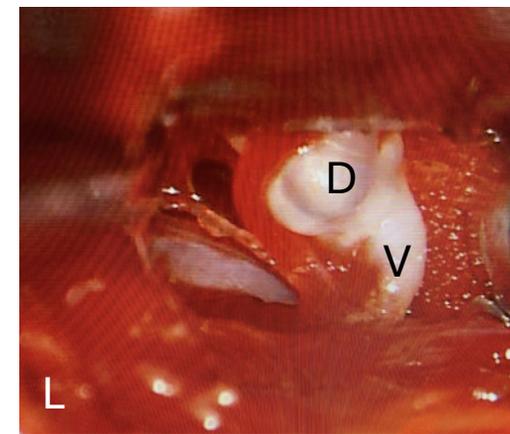
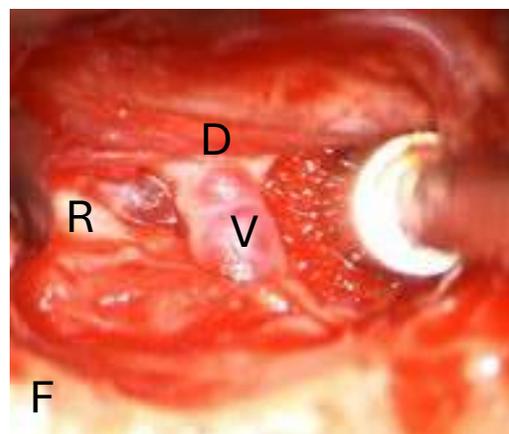
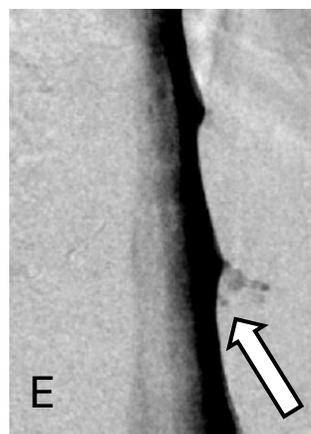
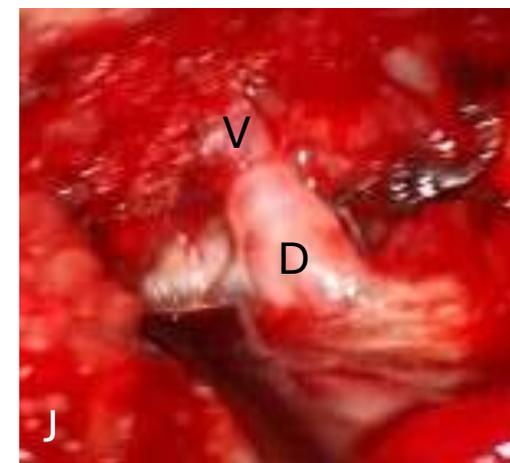
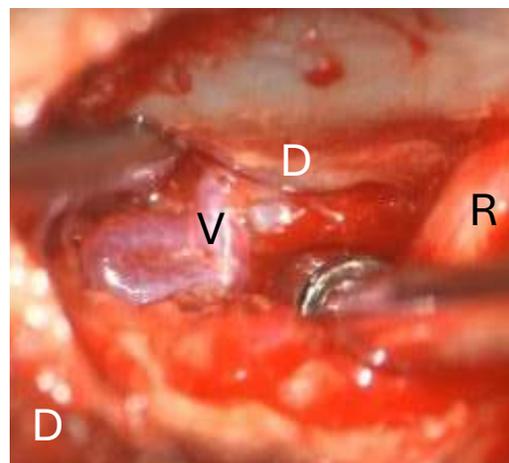
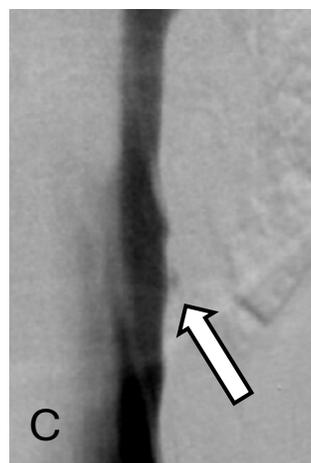
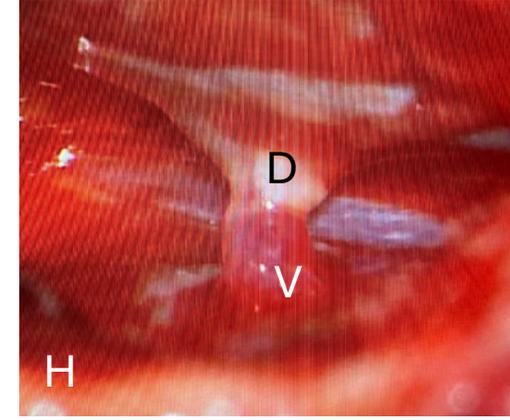
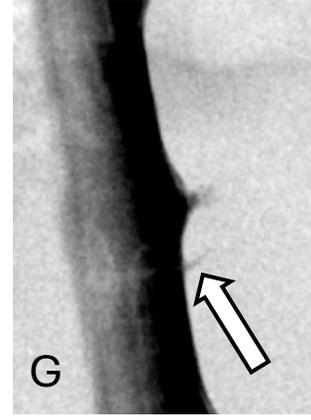
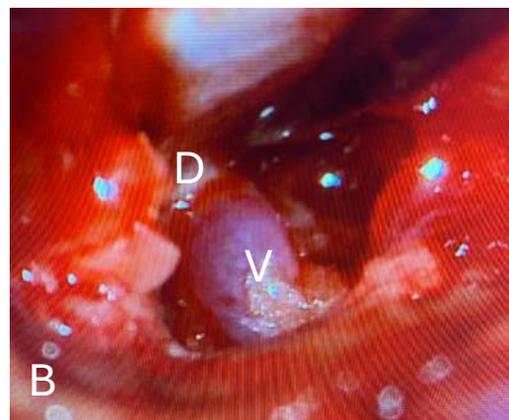
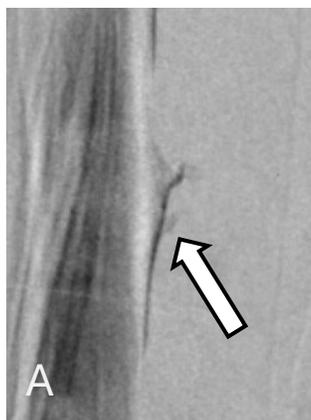
27 patients underwent surgical exploration

27 CSF-venous fistulas
In 23 patients

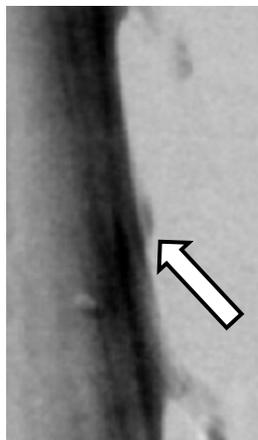
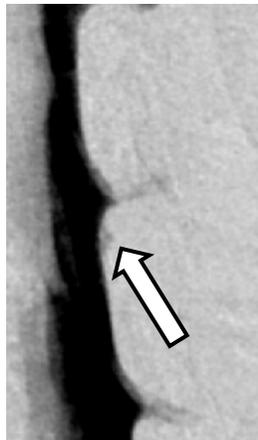
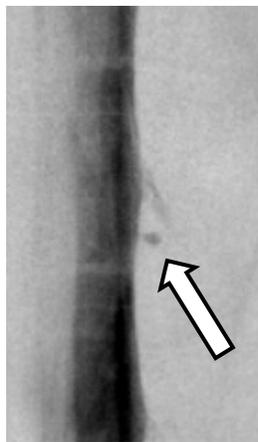
4 pedicular lateral CSF leaks in 4 patients

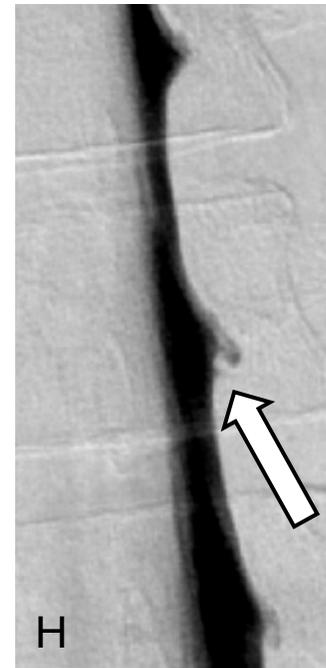
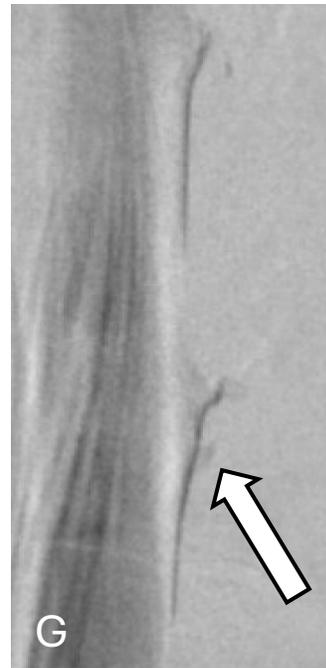
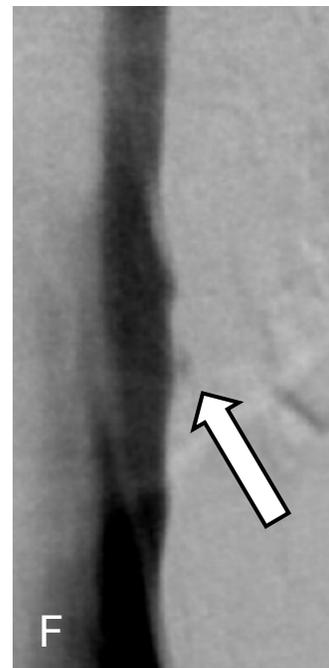
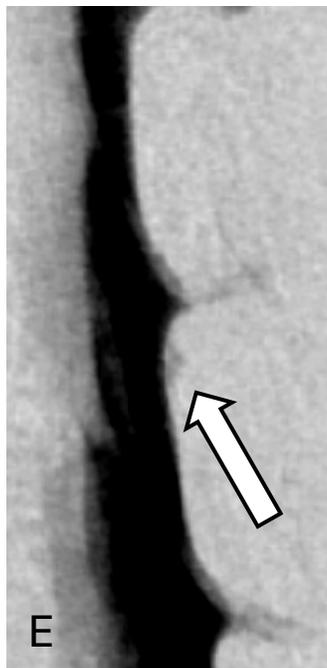
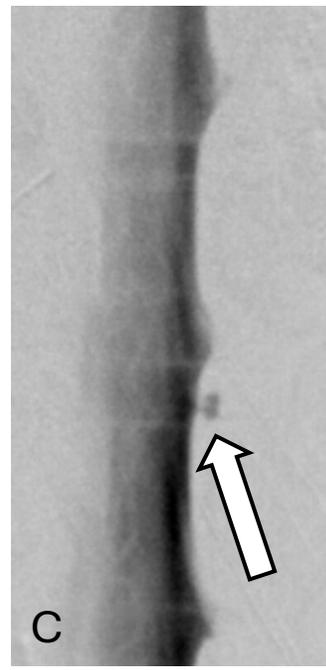
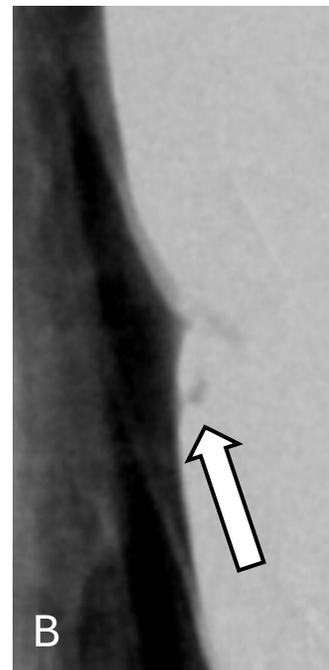
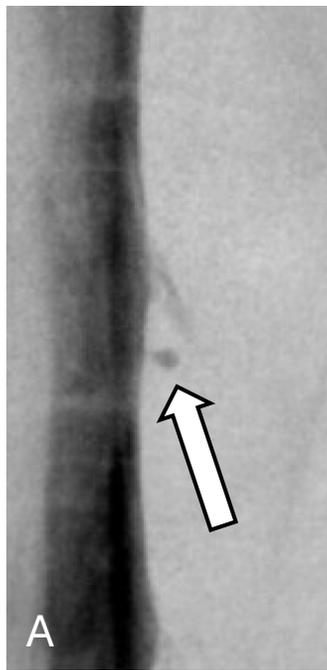


CSF-venous
fistulas



Pedicular lateral tears



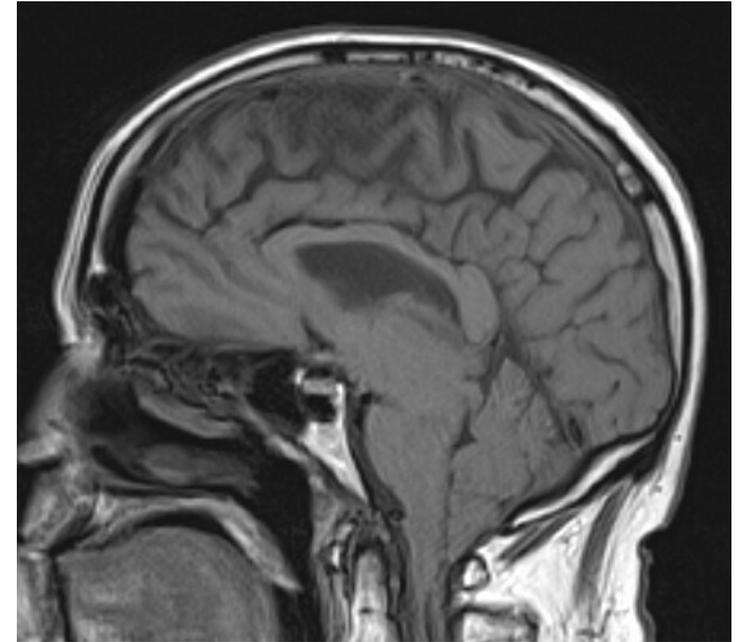
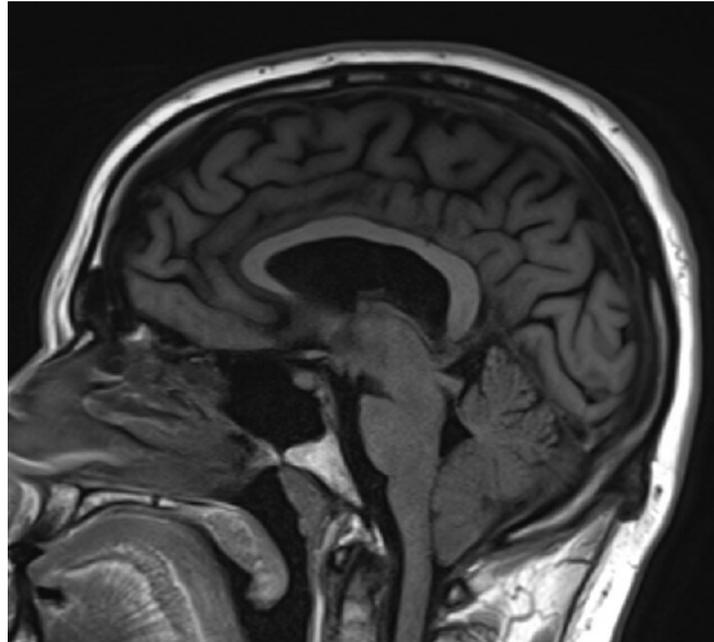
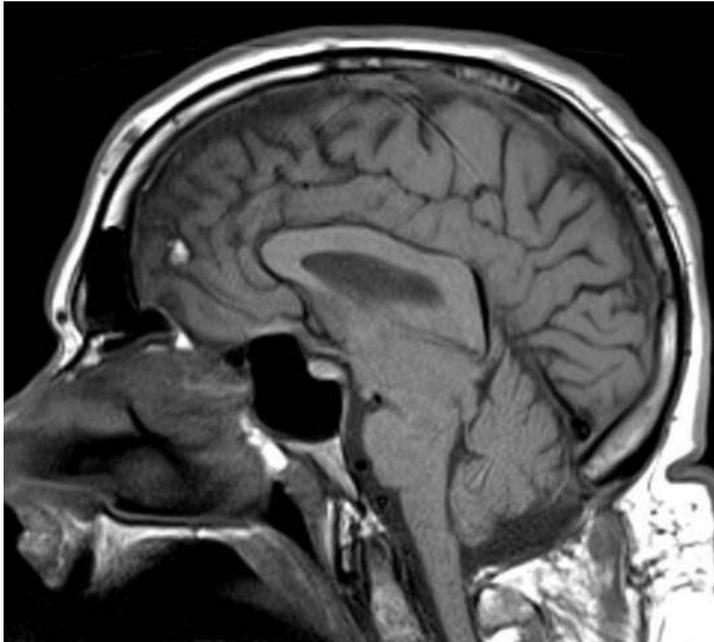


Reasons for normal spine imaging in CSF deficiency

1. Skull base CSF leak
2. “Pooling” of CSF in multiple diverticula/dural ectasia
3. Small lateral dural CSF collections
4. Azygos vein stenosis

Azygos Vein Stenosis in Frontotemporal Dementia Sagging Brain Syndrome

 Wouter I. Schievink,  Marcel M. Maya, Rola Saouaf, H. Gabriel Lipshutz,  Rachele B. Taché,  Daniel Scoffings, and  Jeremy D. Schmahmann



Behavioral – variant frontotemporal dementia

- Progressive deterioration of personality, social comportment, and cognition
- Caused by frontotemporal lobar degeneration
- Incurable
- Second most common cause of dementia in <60 years
- Genetic basis

Reversible variant due to CSF loss!

DIAGNOSIS

By Lisa Sanders, M.d.

Nov. 2, 2017

“So, do you like working here?” the middle-aged man bellowed to the young physician at the other end of the hospital coffee shop. The woman, the object of this not-very-subtle pickup line, ignored him. The man’s sister cringed. When had her younger brother, who was 49, turned into such a jerk? He had always been so quiet and shy. She was living across the country in Washington State, so she didn’t see him often, but he had certainly changed.

In his 20s, he had a problem with alcohol. But back then, his drinking made him quieter. And even during the worst of his drinking days, he had always been tidy and well kempt, fastidious in everything he did. That morning, she drove from the airport to pick him up on the way to visit their father, who was in the hospital after heart surgery. She had taken a red-eye flight from Seattle to Philadelphia, but her brother looked worse than she felt: tired, disheveled, dirty. He said he had just showered, but she could tell it wasn’t true.



The reversible impairment of behavioral variant frontotemporal brain sagging syndrome: Challenges and opportunities

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Abstract

Introduction: Due to loss of brain buoyancy, spontaneous spinal cerebrospinal fluid (CSF) leaks cause orthostatic headaches but also can cause symptoms indistinguishable from behavioral variant frontotemporal dementia (bvFTD) due to severe brain sagging (including the frontal and temporal lobes), as visualized on brain magnetic resonance imaging. However, the detection of these CSF leaks may require specialized spinal imaging techniques, such as digital subtraction myelography (DSM).

Methods: We performed DSM in the lateral decubitus position under general anesthesia in 21 consecutive patients with frontotemporal dementia brain sagging syndrome (4 women and 17 men; mean age 56.2 years [range: 31–70 years]).

Results: Nine patients (42.8%) were found to have a CSF-venous fistula, a recently discovered type of CSF leak that cannot be detected on conventional spinal imaging. All nine patients underwent uneventful surgical ligation of the fistula. Complete or near-complete and sustained resolution of bvFTD symptoms was obtained by all nine patients, accompanied by reversal of brain sagging, but in only three (25.0%) of the twelve patients in whom no CSF-venous fistula could be detected ($P = 0.0011$), and who were treated with non-targeted therapies.

Discussion: Concerns about a spinal CSF leak should not be dismissed in patients with frontotemporal brain sagging syndrome, even when conventional spinal imaging is normal. However, even with this specialized imaging the source of the loss of spinal CSF remains elusive in more than half of patients.

KEYWORDS

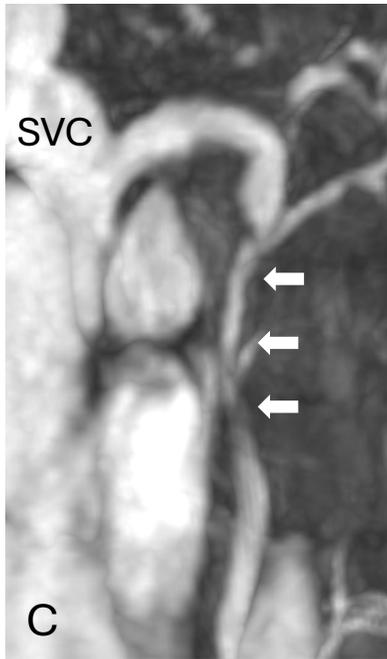
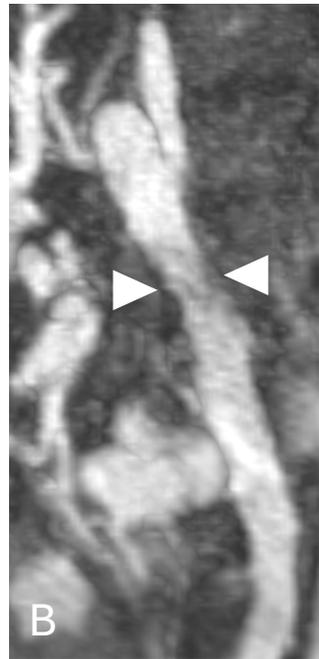
behavioral variant frontotemporal dementia, brain sagging, cerebrospinal fluid leak, frontotemporal dementia, spontaneous intracranial hypotension

TABLE 2 Neurologic manifestations of SIH in 21 patients with behavioral variant frontotemporal brain sagging syndrome

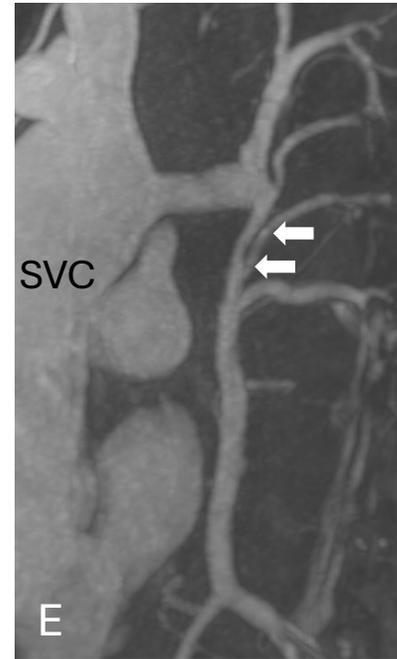
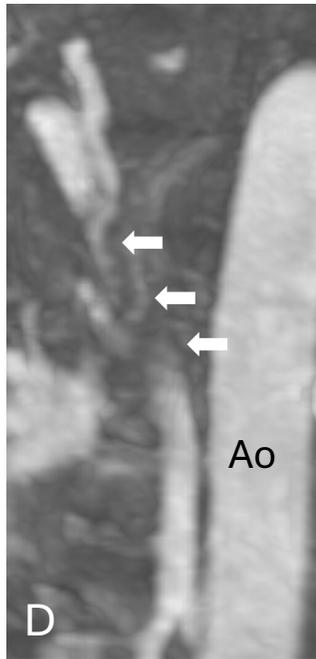
Hypersomnolence	21/21 (100%)
Dysequilibrium/gait dysfunction	19/21 (90.5%)
Headache	17/21 (81%)
Orthostatic	11/17 (64.7%)
Non-positional	3/17 (17.6%)
Reverse orthostatic	3/17 (17.6%)
Dysphagia/dysarthria	14/21 (66.7%)
Tremors	9/21 (42.9%)
Incontinence	7/21 (33.3%)
Orofacial dyskinesia	5/21 (23.8%)
Dysgeusia	4/21 (19.0%)
Hiccupping	4/21 (19.0%)
Hallucinations	3/21 (14.3%)
Impotence/erectile dysfunction	3/21 (14.3%)



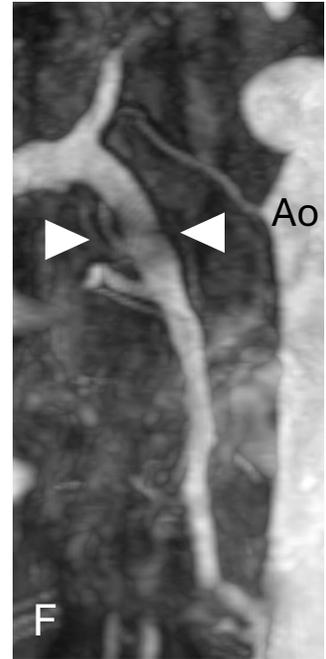
Patient 1

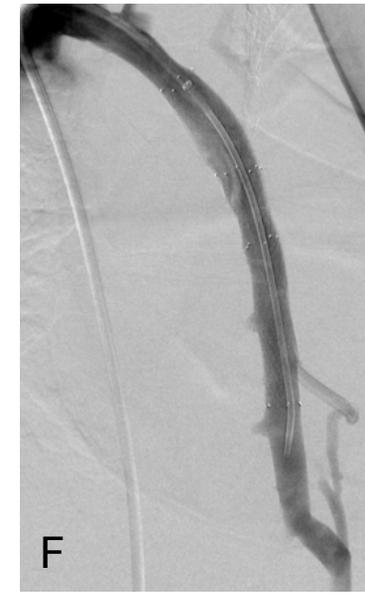
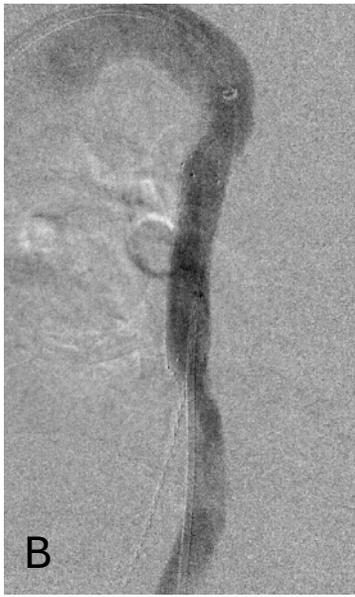
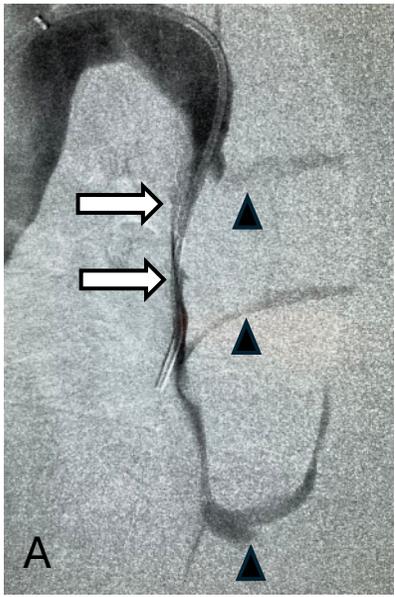


Patient 2



Patient 3



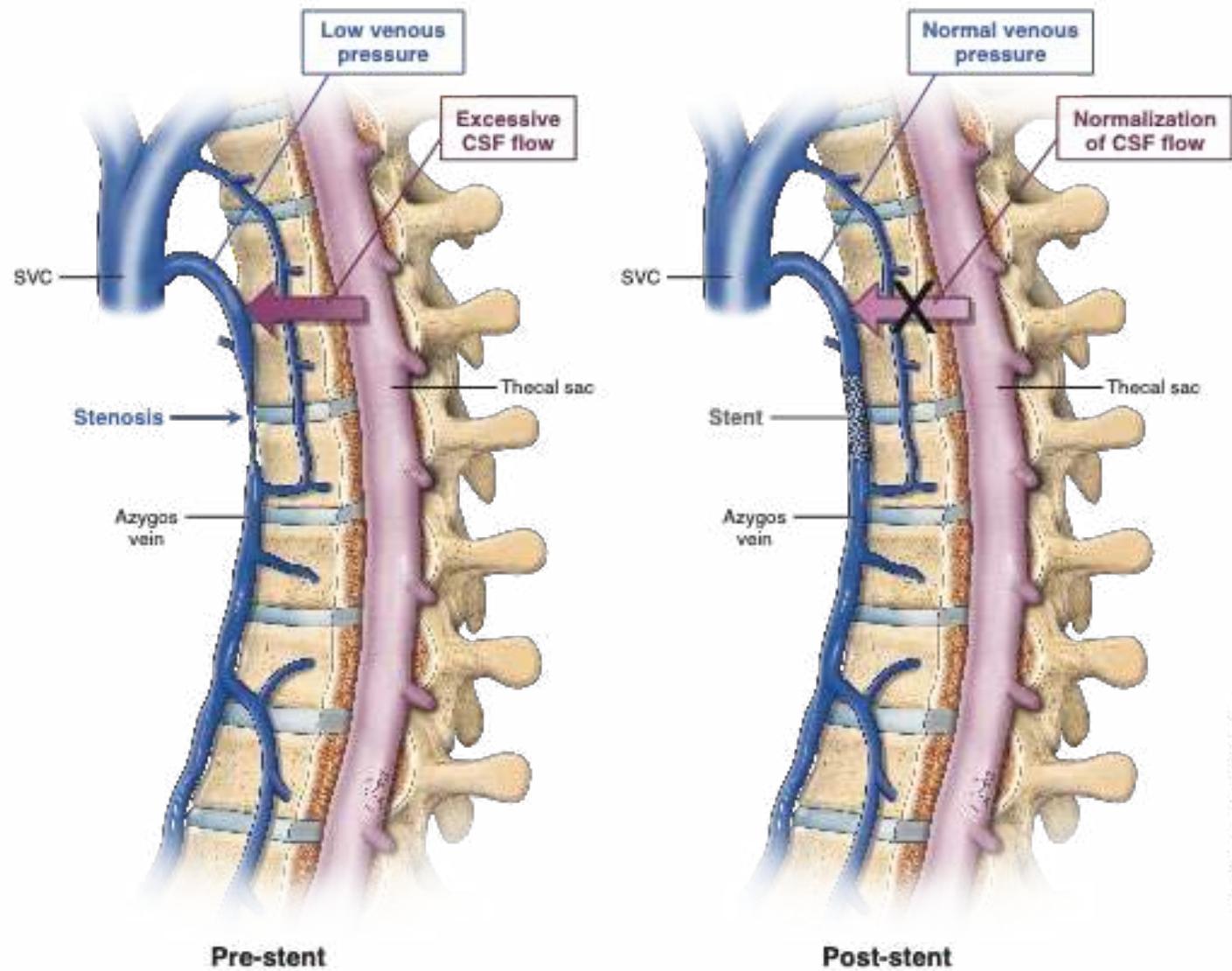


SIHDAS score pre- and post-stenting

Patient 1: 180 → 3

Patient 2: 187 → 0

Patient 3: 70 → 30



approximately 1 cm. Just distal to this site at T7-8, there is an approximately 2 cm long segment of similar-appearing ribbon like narrowing to approximately 2 mm, series 2, image 68, again in-between the nondilated esophagus and spinal column. The azygos vein just distal to this site measures up to 9 mm in diameter, corresponding to an approximately 80% diameter stenosis. The azygos arch is widely

COMPARISON: None

FINDINGS:

Left-sided aortic arch with three-vessel arterial branching pattern. The visualized great vessels arteries are patent and of normal caliber. No evidence of aortic dissection or aortic aneurysm. No evidence of pulmonary emboli. Normal cardiac chamber size. The visualized abdominal aorta is patent and of normal caliber. Patent celiac, superior mesenteric and renal arteries.

The internal jugular, subclavian, axillary, and innominate veins are patent. The SVC and visualized IVC are patent. The azygos arch is patent as visualized on images 93 through 108 of series 5. The azygos and hemiazygos veins below this level are small in nature with likely areas of stenosis near the T10 vertebral body. This is best seen on images 155 through 198 of series 5.

Postembolization changes lower thoracic spine. Simple cyst within the liver. Calcified granuloma left lower lung. Areas of linear fibrosis in both lung bases. The remainder of the upper abdominal contents are negative.

Reasons for normal spine imaging in CSF deficiency

1. Skull base CSF leak
2. “Pooling” of CSF in multiple diverticula/dural ectasia
3. Small lateral dural CSF collections
4. Azygos vein stenosis
5. Skull defect

RESEARCH ARTICLE

Skull defect – Frontotemporal dementia sagging brain syndrome

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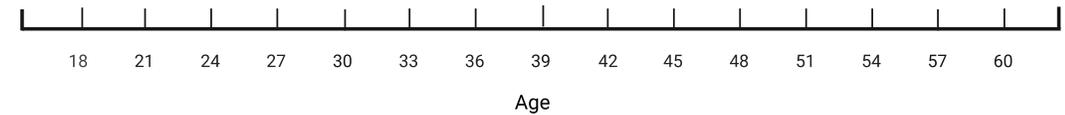
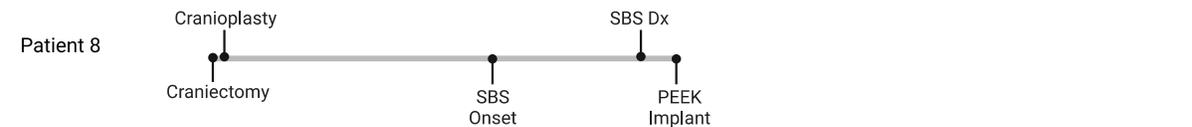
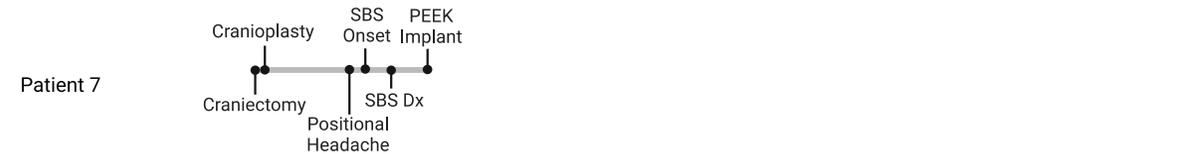
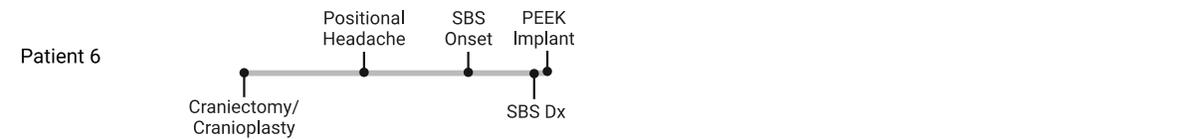
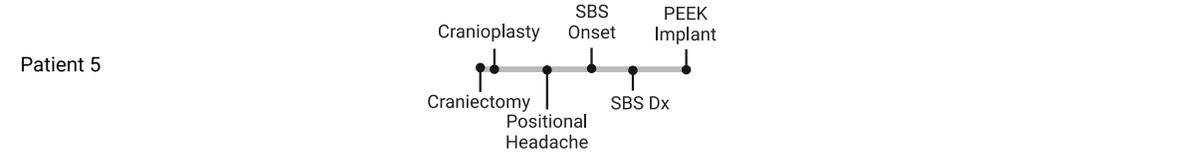
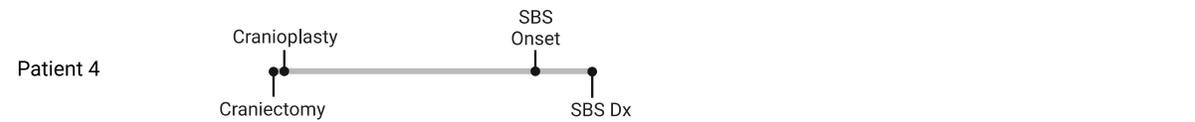
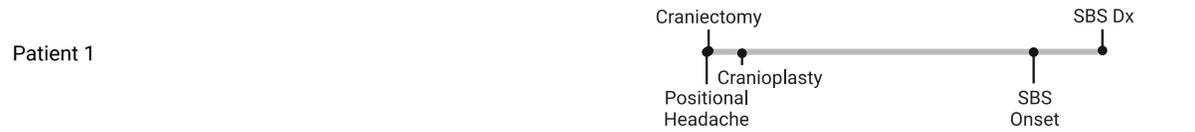
Annals of Clinical and Translational Neurology 2025; 12(1): 226–234

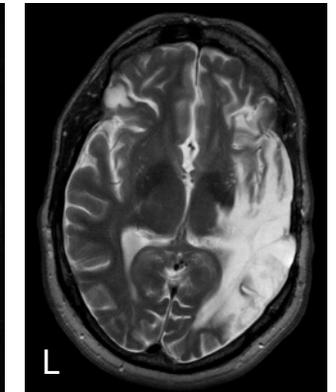
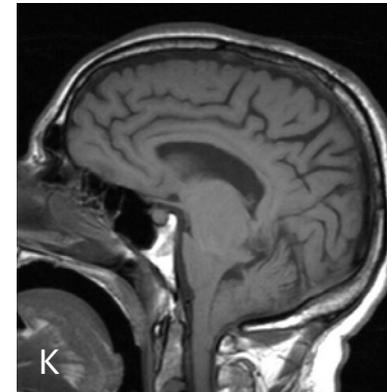
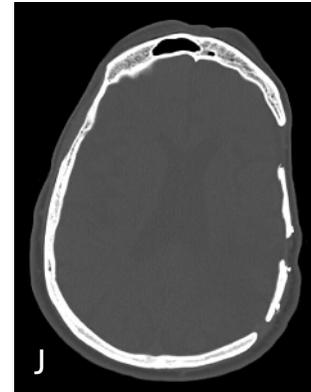
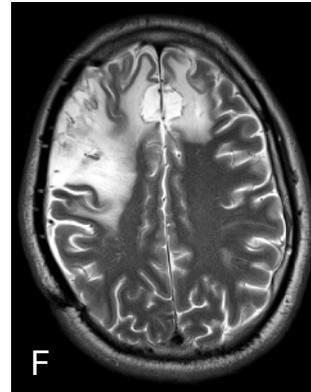
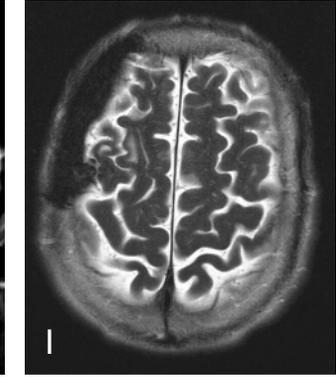
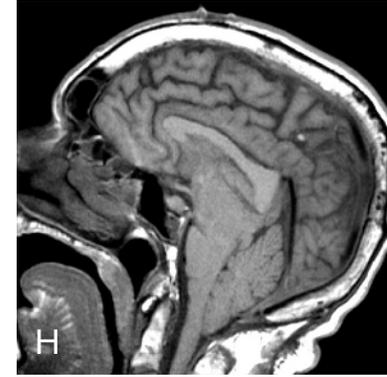
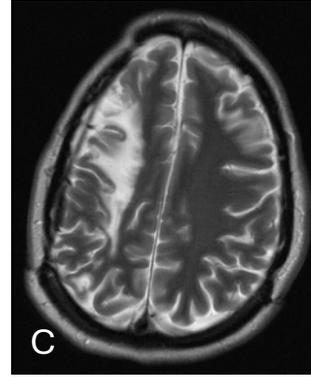
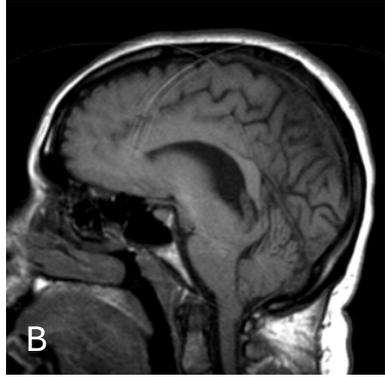
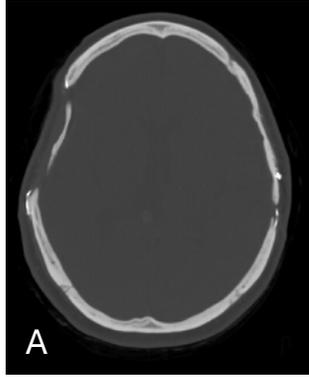
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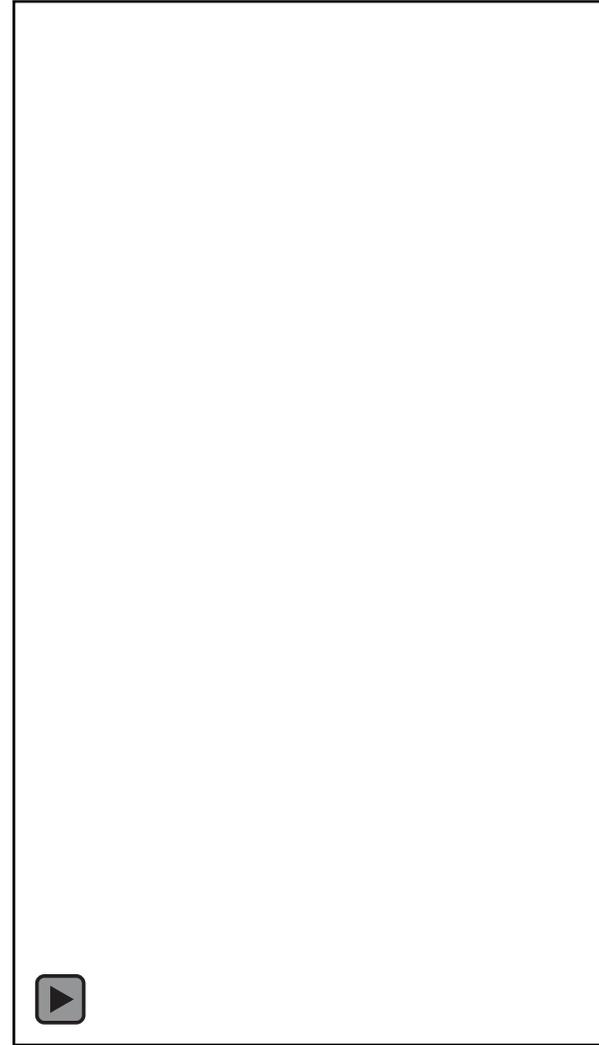
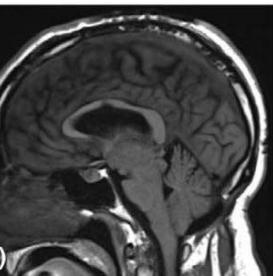
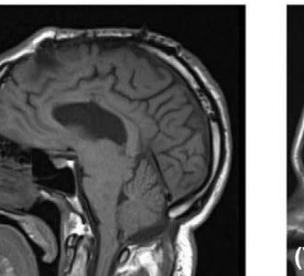
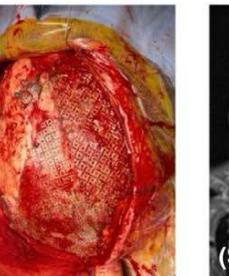
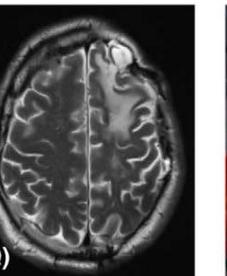
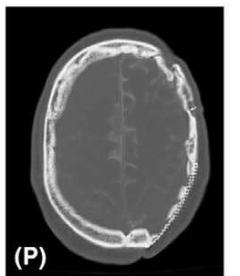
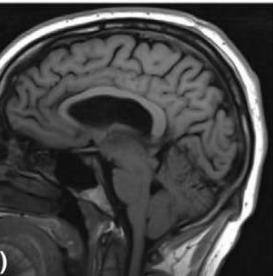
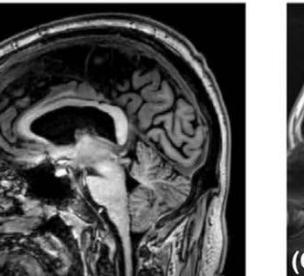
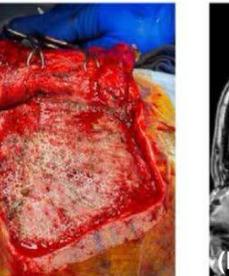
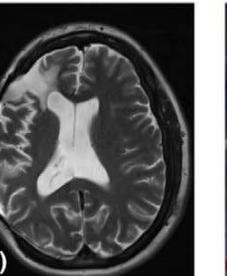
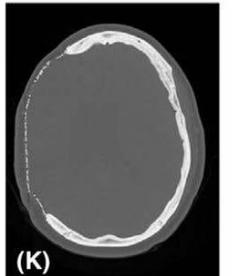
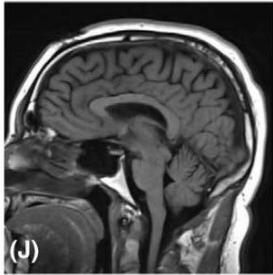
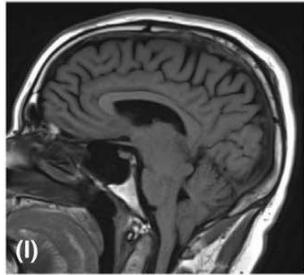
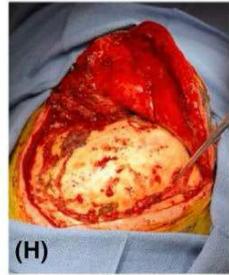
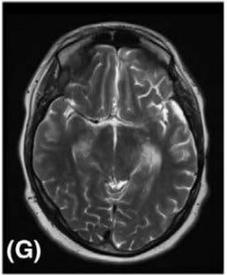
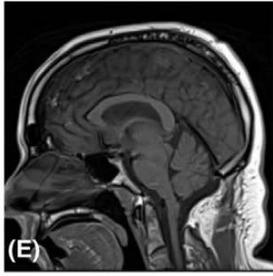
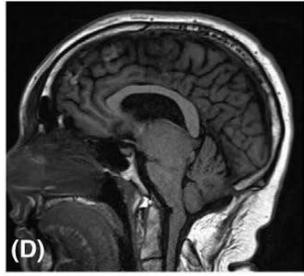
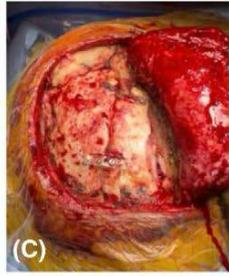
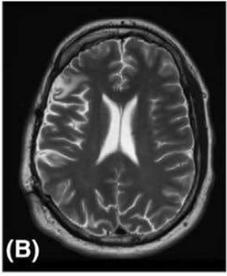
Abstract

Objective: Frontotemporal dementia (FTD) sagging brain syndrome is a disabling condition. An underlying spinal Cerebrospinal fluid leak can be identified in only a minority of patients and the success rate of non-directed treatments is low. Some of these patients have a remote history of craniectomy/cranioplasty and we report a positive response to custom implant cranioplasty revision many years after their initial cranioplasty. **Methods:** We reviewed medical records and imaging studies of 61 consecutive patients with FTD sagging brain syndrome. A SIH Disability Assessment Score (SIHDAS) questionnaire was completed to assess the severity of the symptoms before and after custom implant cranioplasty. Pre- and post-operative brain MRI was obtained to assess degree of brain sagging. **Results:** Eight (13.1%) of the 61 patients had a history of craniectomy/cranioplasty 1.5–13.5 years prior to onset of symptoms of FTD sagging brain syndrome. The mean age of the one woman and seven men at the time of presentation to our medical center was 50 years (range, 26–68 years). None had sinking scalp flap syndrome. Prior treatments included epidural blood patching and dural reduction surgery. Custom cranial implant surgery was performed in four patients and resulted in prompt and remarkable improvement of symptoms in three patients (SIHDAS: very severe disability to no or mild disability) and mild improvement in one patient. Brain MRI showed improvement of brain sagging. **Interpretation:** A disproportionate number of patients with FTD sagging brain syndrome have a remote history of supratentorial craniectomy/cranioplasty and revision cranioplasty should be considered.

Skull defect – Frontotemporal dementia sagging brain syndrome









CLINICAL/SCIENTIFIC NOTE

Compulsive Repetitive Flexion With Breath-Holding in Sagging Brain Syndrome

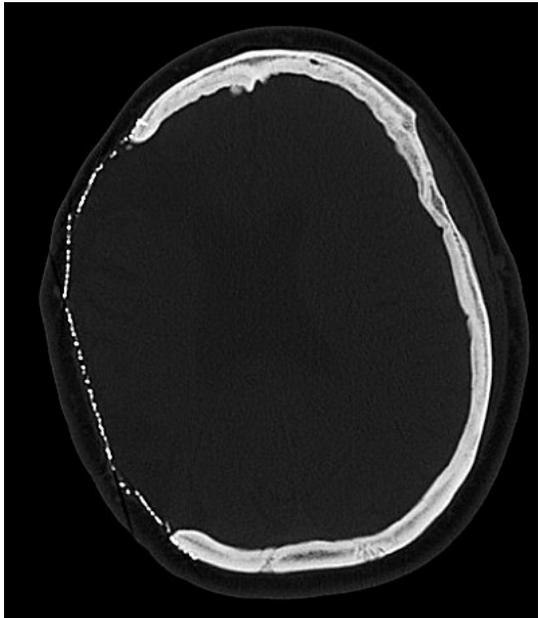
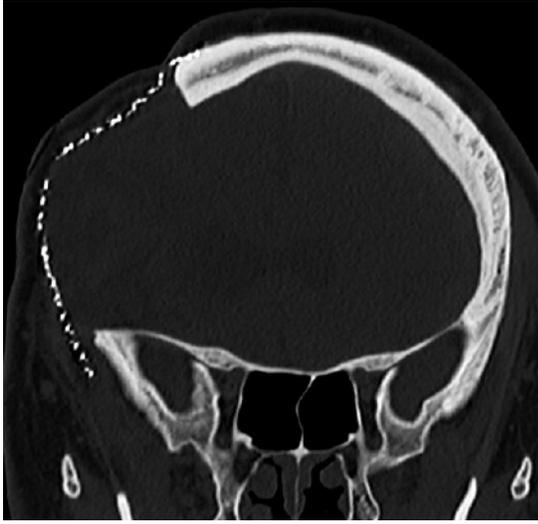
Jeremy D. Schmahmann, MD, and Wouter I. Schievink, MD

Neurology: Clinical Practice 2024;14:e200261. doi:10.1212/CPJ.0000000000200261

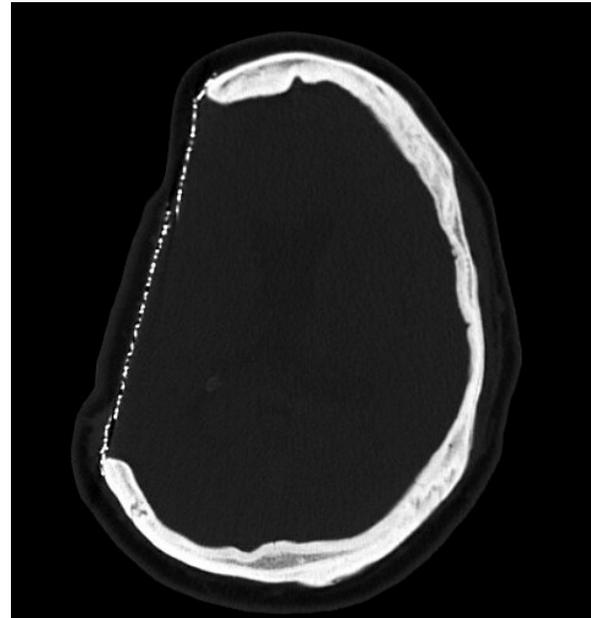
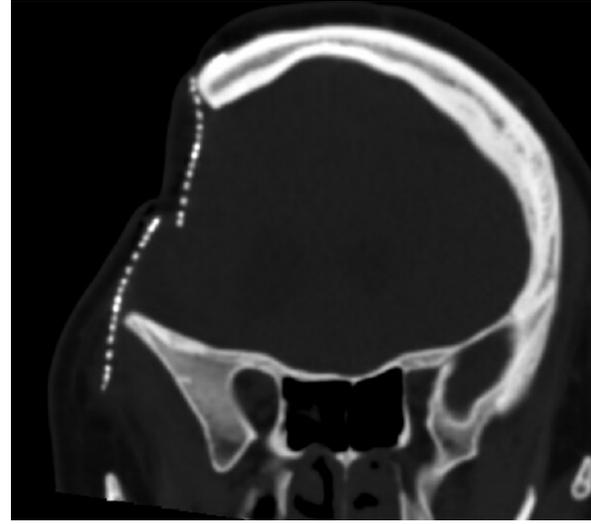
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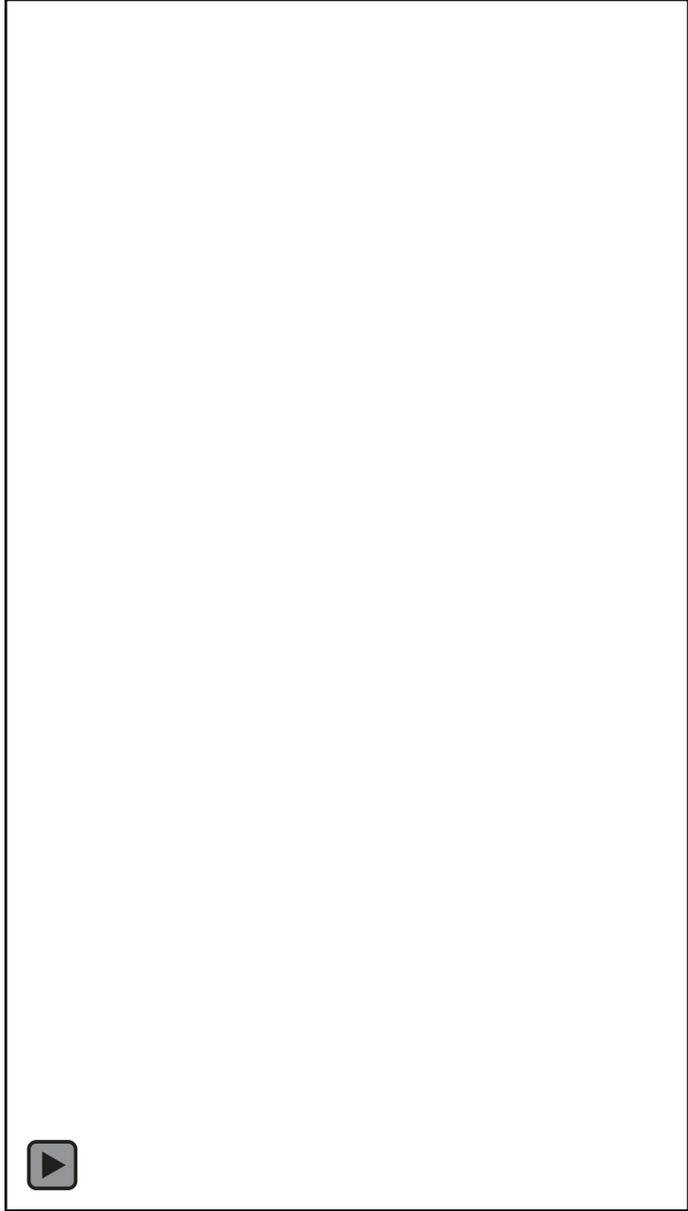
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or Dr. Schmahmann
jschmahmann@partners.org

CORFBIS

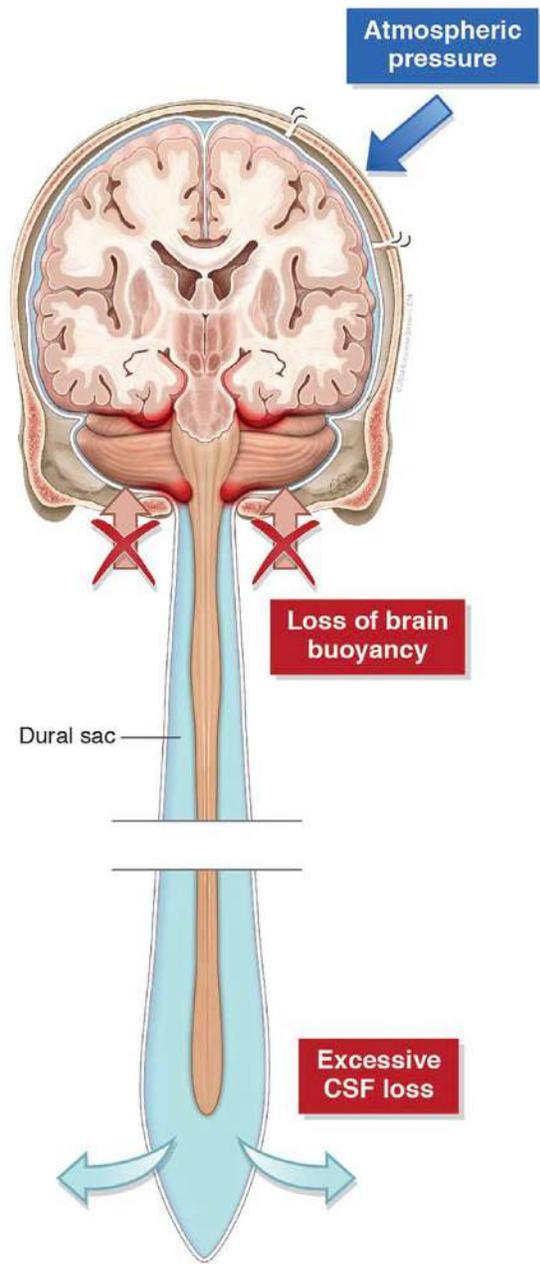


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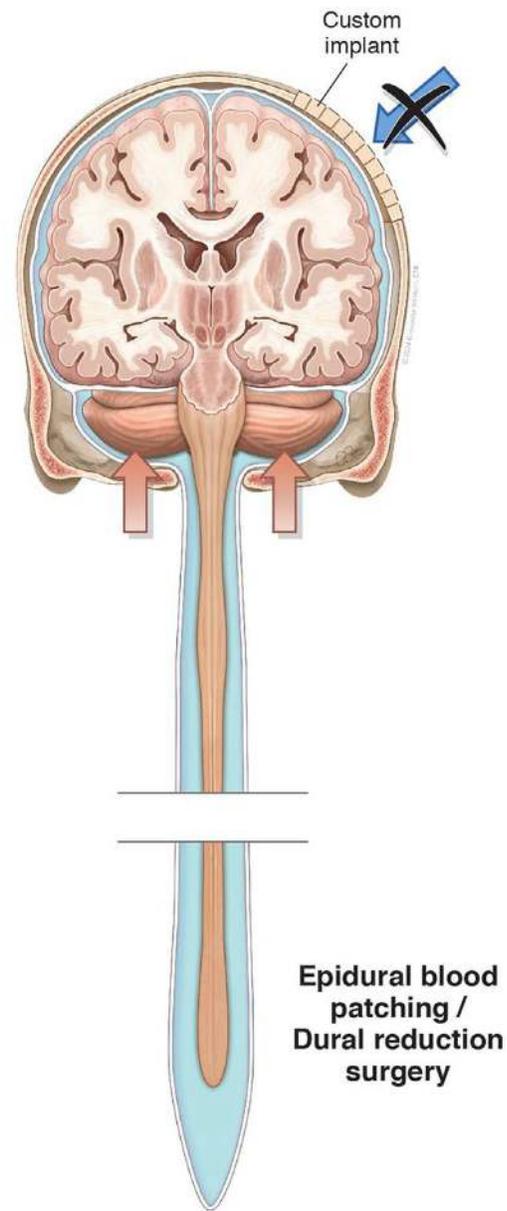




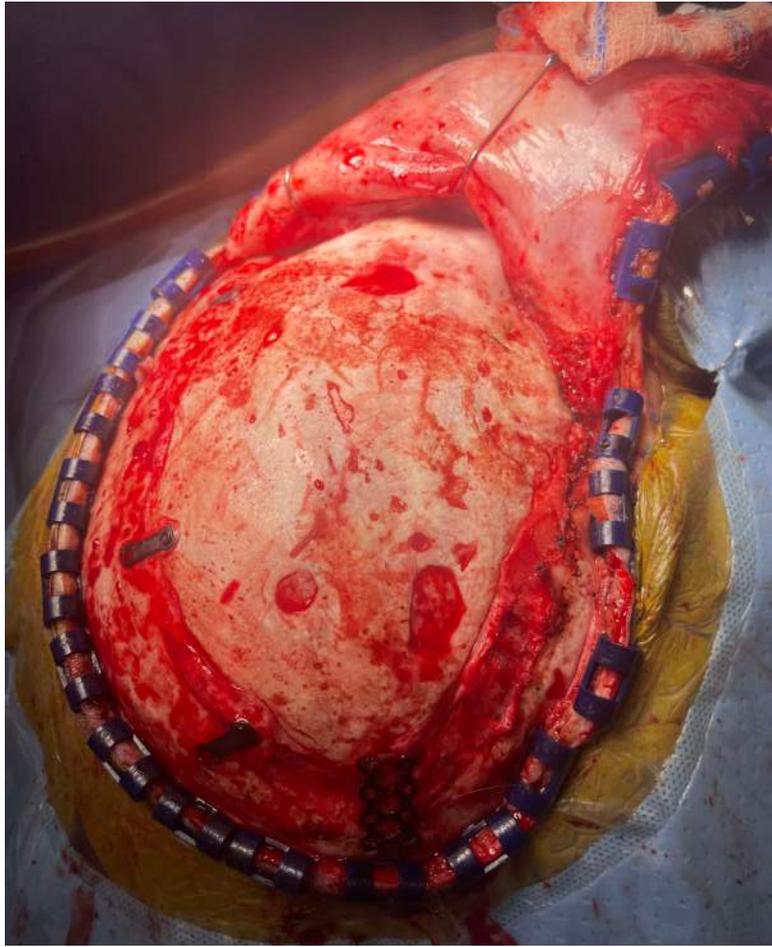
PEEK implant

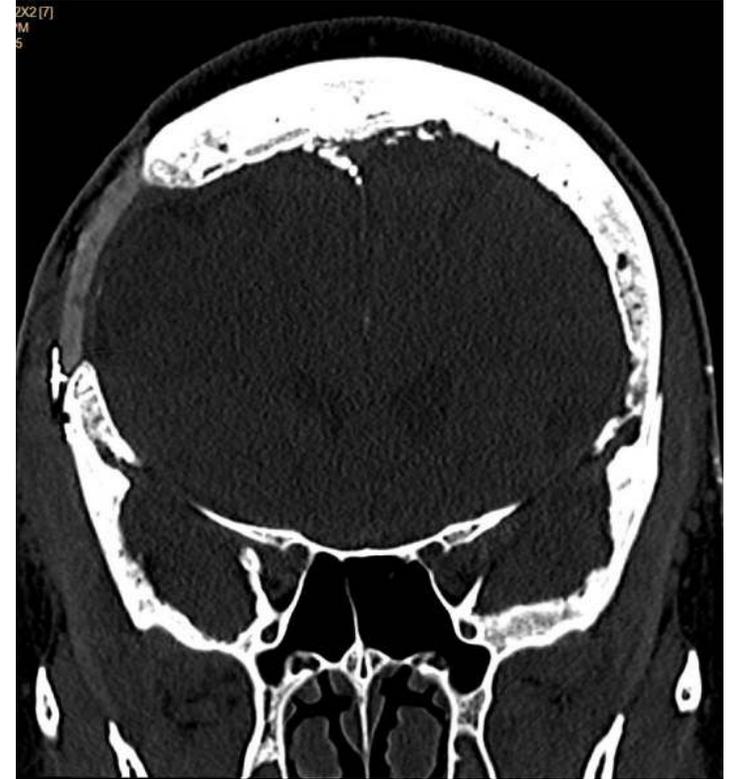
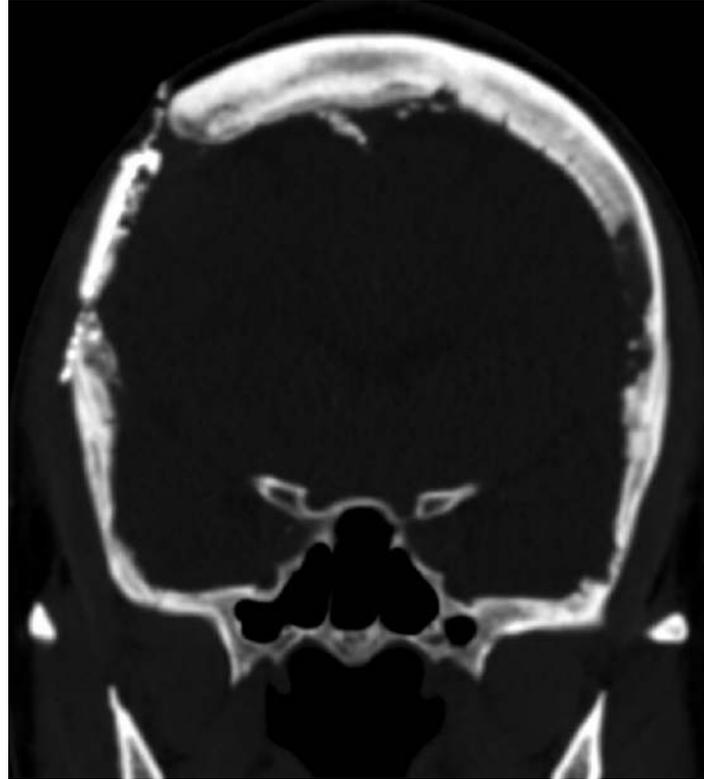
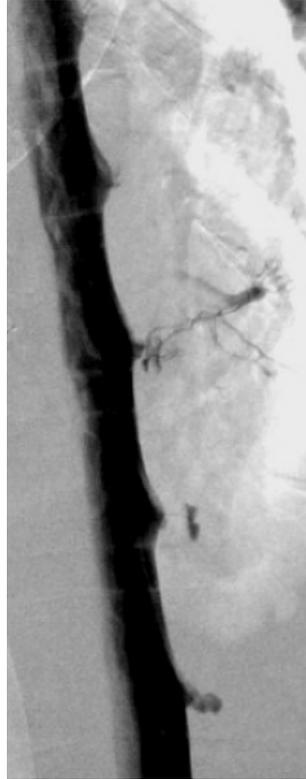


BEFORE Cranioplasty

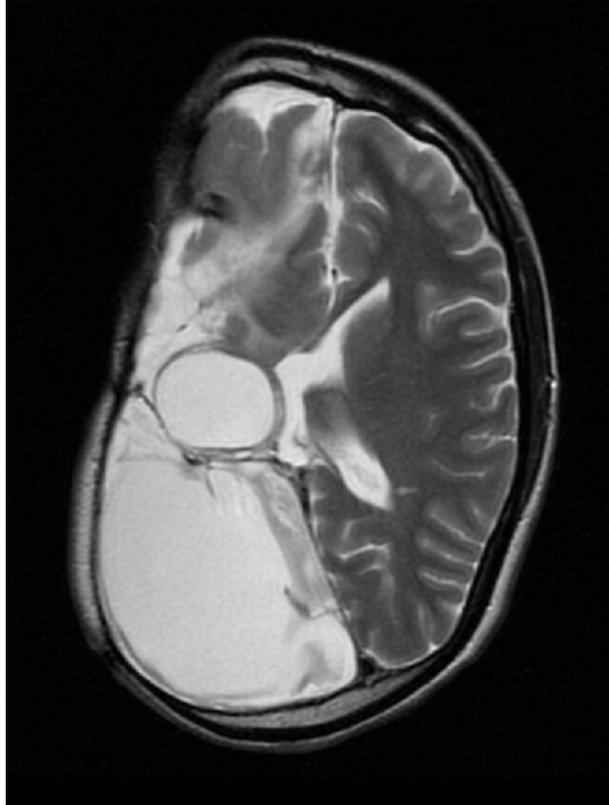


AFTER Cranioplasty

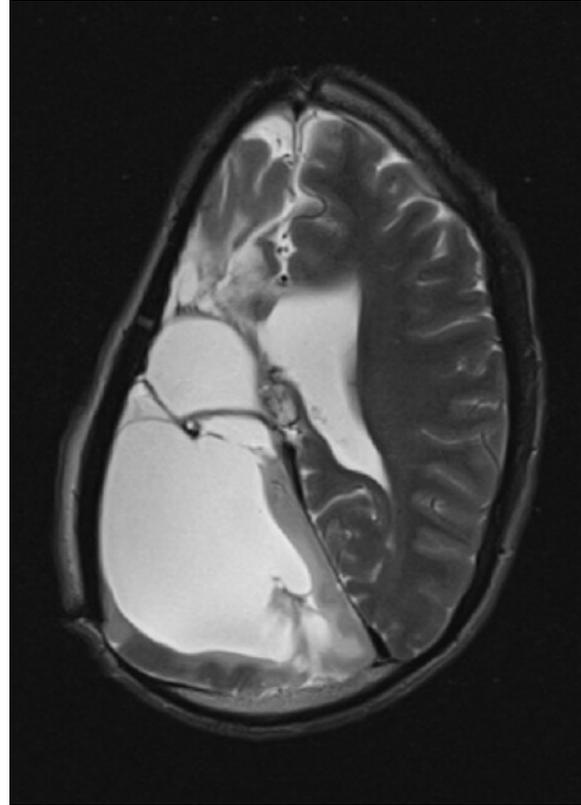




Resolution of orthostatic headaches



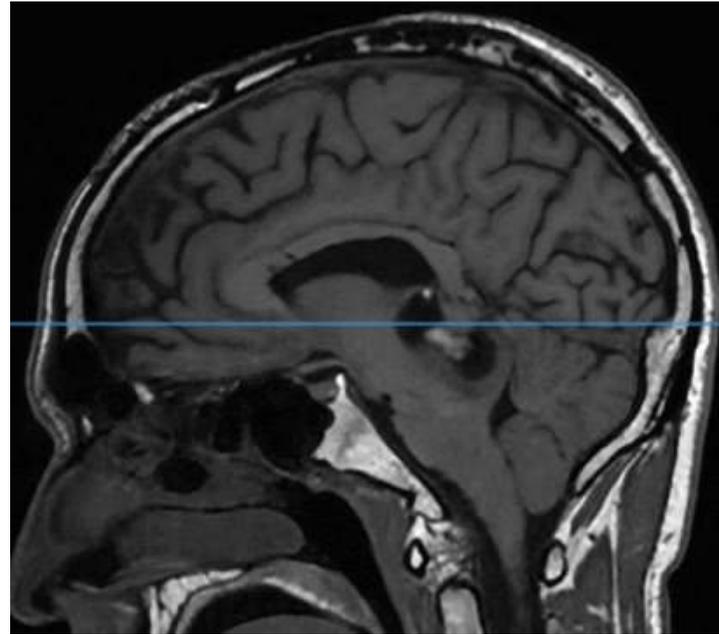
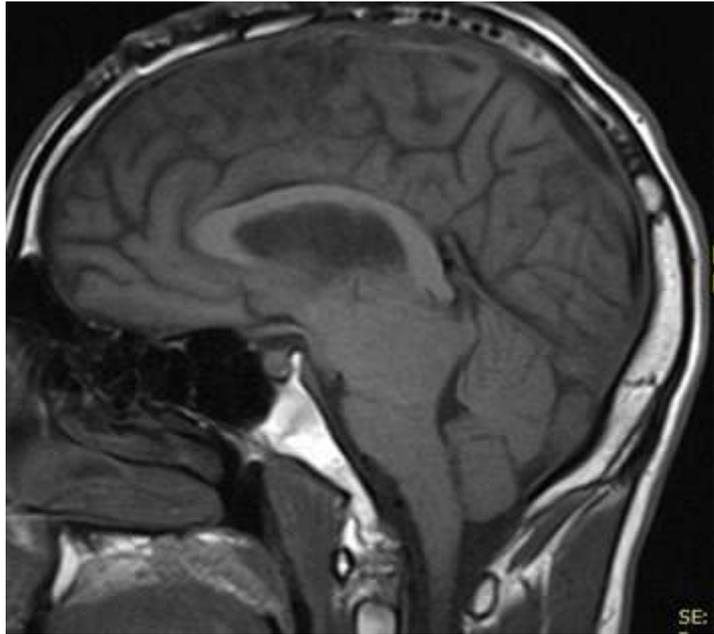
Pre-op



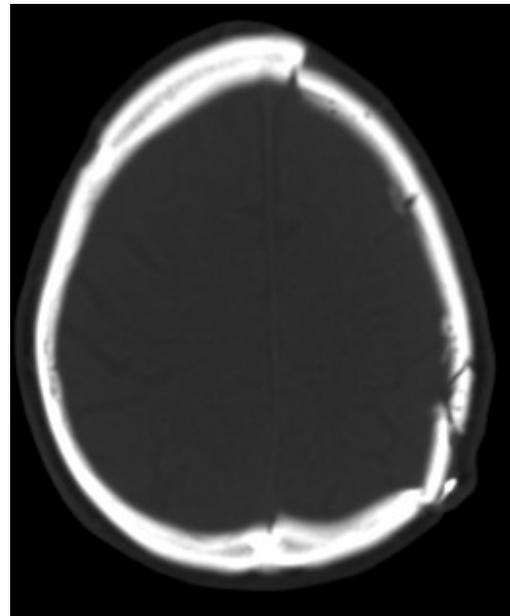
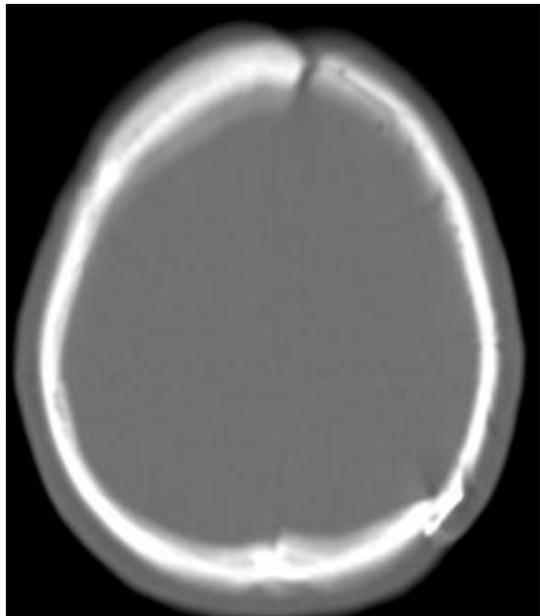
Post-op



Resolution of orthostatic headaches



Young man
Severe TBI many years prior
Sagging brain syndrome x 3 years



Awaiting PEEK implant

Brain + for SIH but no spinal CSF leak

- Maybe not in the spine?
- Pooling of CSF?
- Are there any small innocuous looking lateral dural CSF collections?
- MRV or CTV to look for azygos vein stenosis?
- Is there a history of craniectomy/(inadequate)cranioplasty?

Thank you



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