Case Based Approach to the Diagnosis and Management of Spontaneous Intracranial Hypotension (SIH) from CSF leak and CSF-Venous Fistula

Ochsner Neuroscience Symposium

Paul Gulotta, MD

May 16th, 2024

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DISCLOSURES

- Consultant: Boston Scientific

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Outline

- Algorithmic Multimodality and Multidisciplinary Approach
- Imaging Techniques
- Imaging Findings
- Diagnosis and Management

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Clinical Perspective | Neuroradiology/Head and Neck Imaging | March 9, 2022

Algorithmic Multimodality Approach to Diagnosis and Treatment of Spinal CSF Leak and Venous Fistula in Patients With Spontaneous Intracranial Hypotension

Authors: Andrew L. Callen, MD, Vincent M. Timpone, MD, Adam Schwertner, MD, David Zander, MD, Fabio Grassia, MD, Peter Lennarson, MD, Joshua Seinfeld, MD, Kevin O. Lillehei, MD, Marius Birlea, MD, and Ashesh A. Thaker, MD | AUTHOR INFO & AFFILIATIONS

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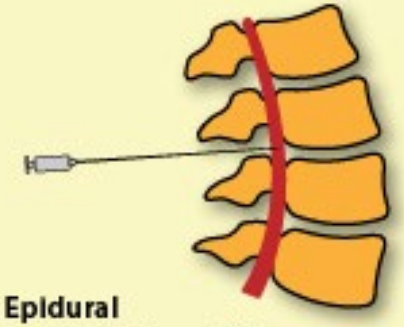
Spinal CSF Leak and Venous Fistula in Spontaneous Intracranial Hypotension

Imaging tests for spinal source

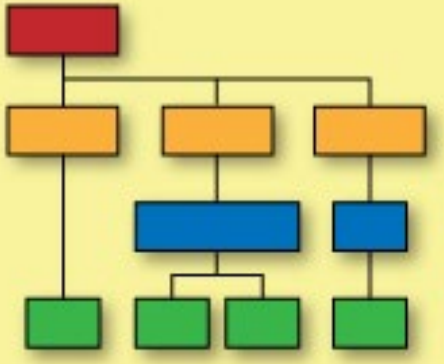
- 3D T2-weighted whole-spine MRI
- Dynamic fluoroscopic myelography
- Dynamic CT myelography
- Delayed MR myelography

Treatments for localized leak



- Epidural blood/fibrin patching
- Percutaneous fibrin glue embolization
- Endovascular embolization
- Surgical ligation

Multimodality algorithmic framework proposed



Callen et al. DOI.10.2214/AJR.22.27485 AJR

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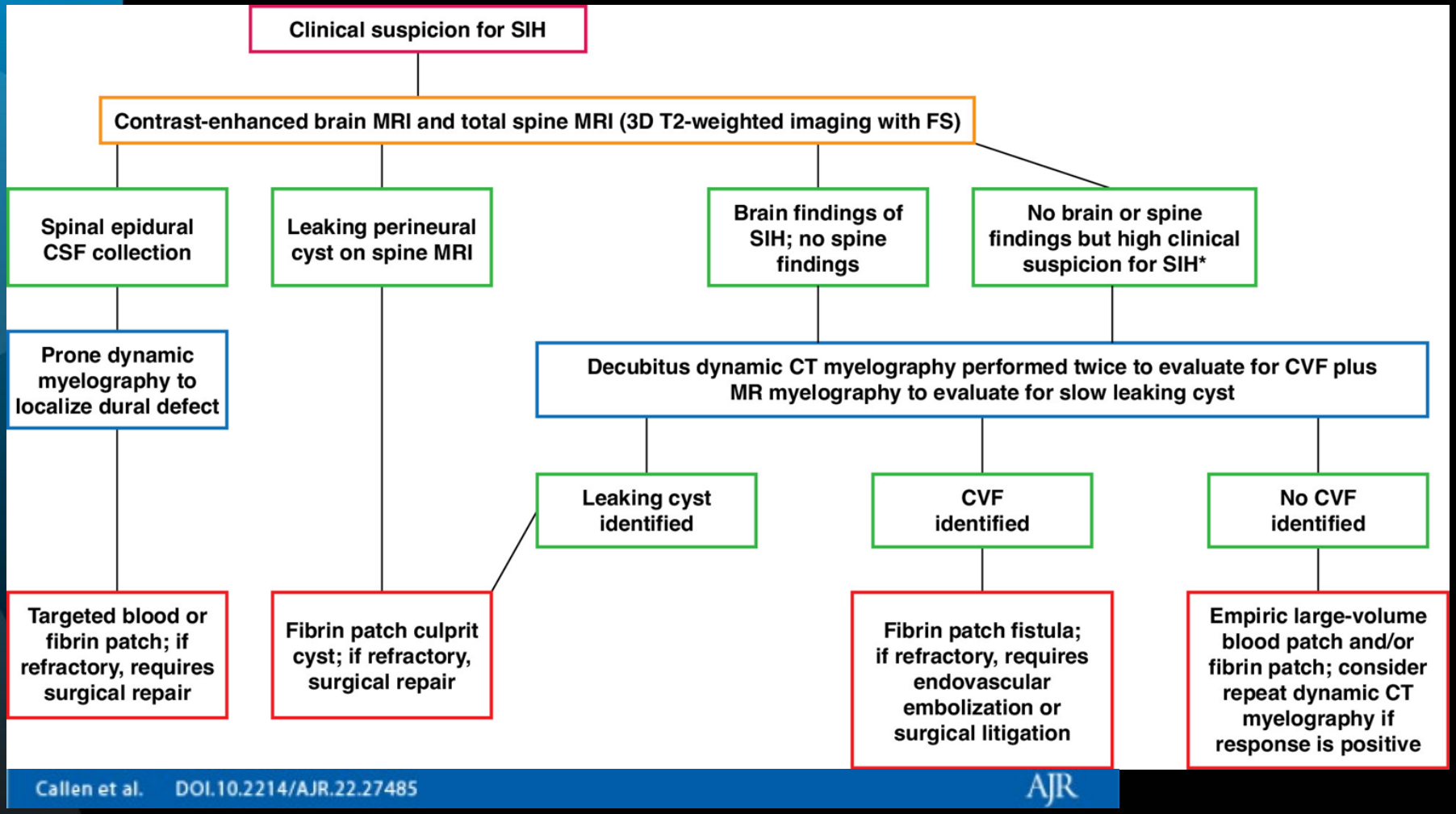

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Imaging Strategy Goals

- Confirm Presence of Intracranial Hypotension
  - Brain MRI can be normal in up to 20% of cases
- Confirm Presence of CSF Leak
- Identify Specific Location and Cause of CSF Leak
- Localize and Potentially Treat Underlying Cause
- May Require Multiple Studies and Modalities

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Imaging Modalities

- Contrast Enhanced MRI Brain
- 3D T2 Weighted MRI of the Entire Spine
- Prone Dynamic Myelography
- Bilateral Decubitus CT myelography
- Digital subtraction myelography
- MR Myelography
- Nuclear Medicine Myelography

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Imaging Findings in SIH

- Qualitative Signs:
  - Pachymeningeal Enhancement
  - Engorgement of Dural Venous Sinuses
  - Pituitary Gland Enlargement
  - Subdural Effusions and Hematomas
  - Reduced CSF Volume: Sagging Brain
  - Other: Layer Cake Skull, Superficial Siderosis

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Imaging Findings

- Quantitative Signs:
  - Mamillopontine distance <6.5mm
  - Pontomesencephalic Angle < 50
  - Interpeduncular angle <40.5

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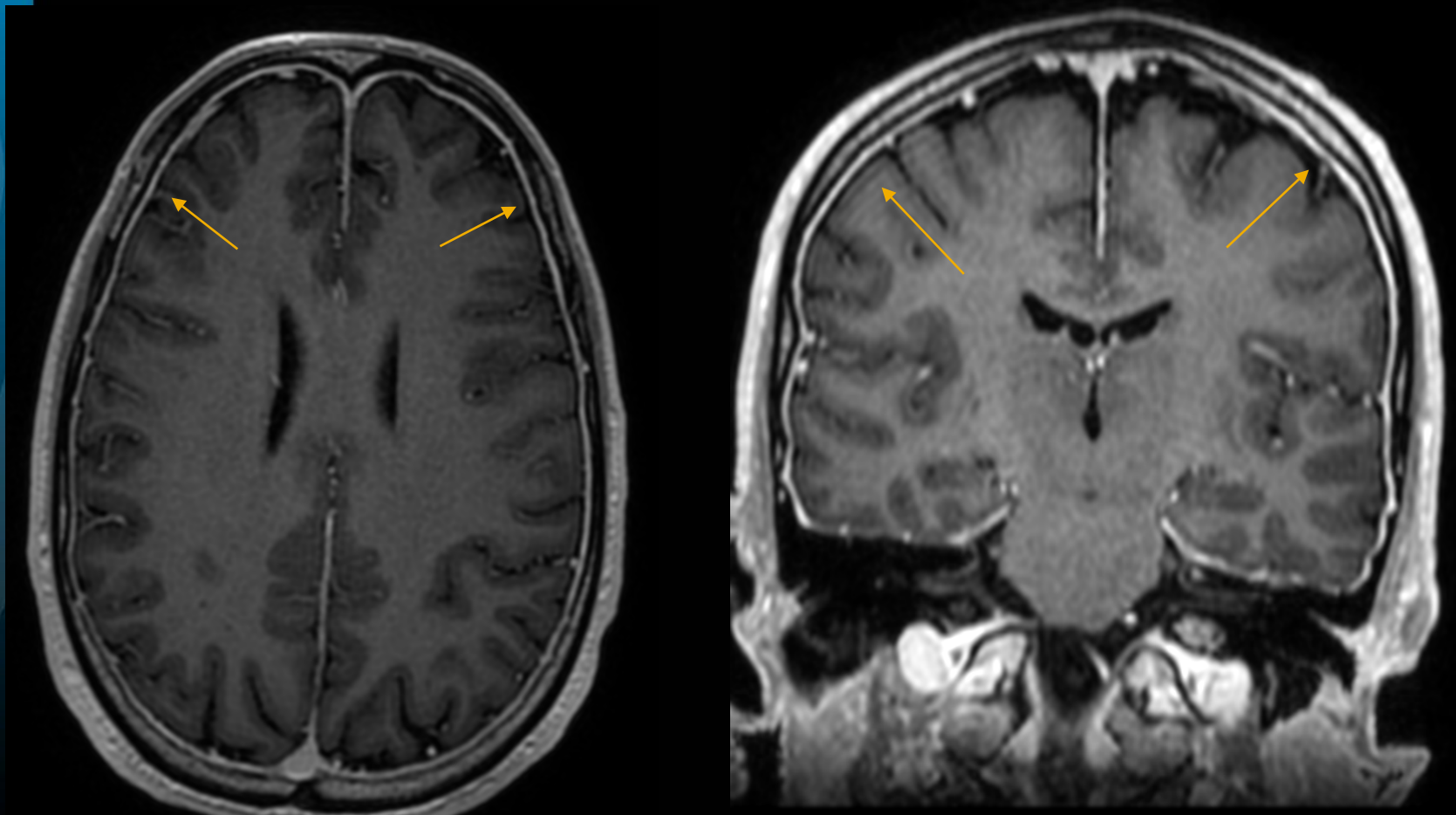
Pachymeningeal Enhancement



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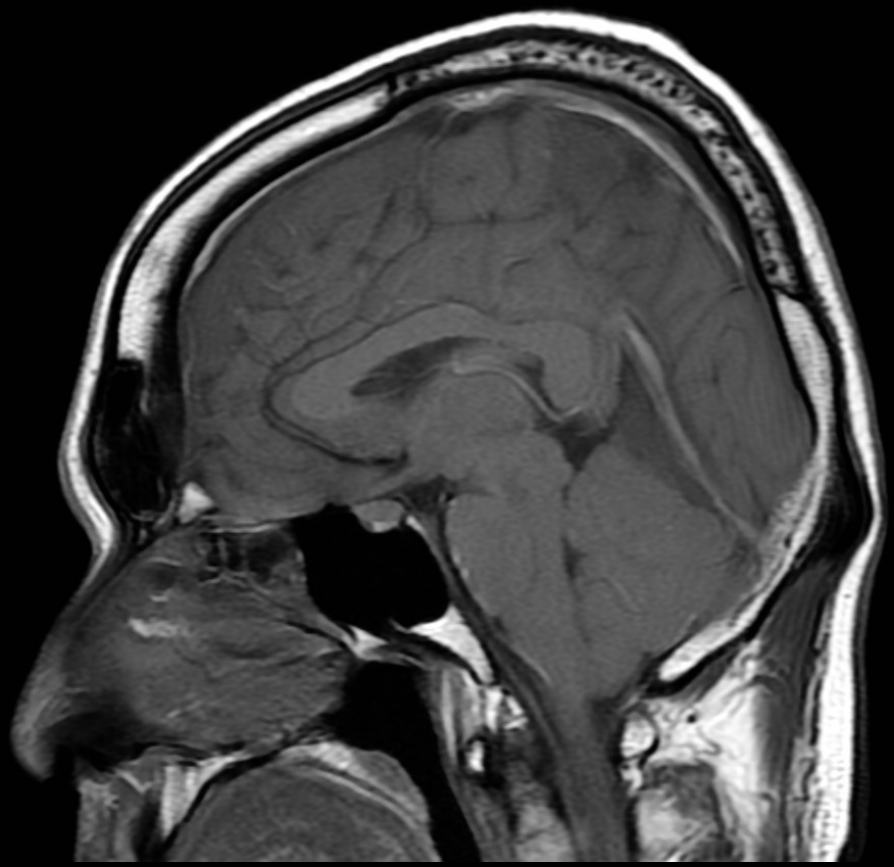
Sagging Brain



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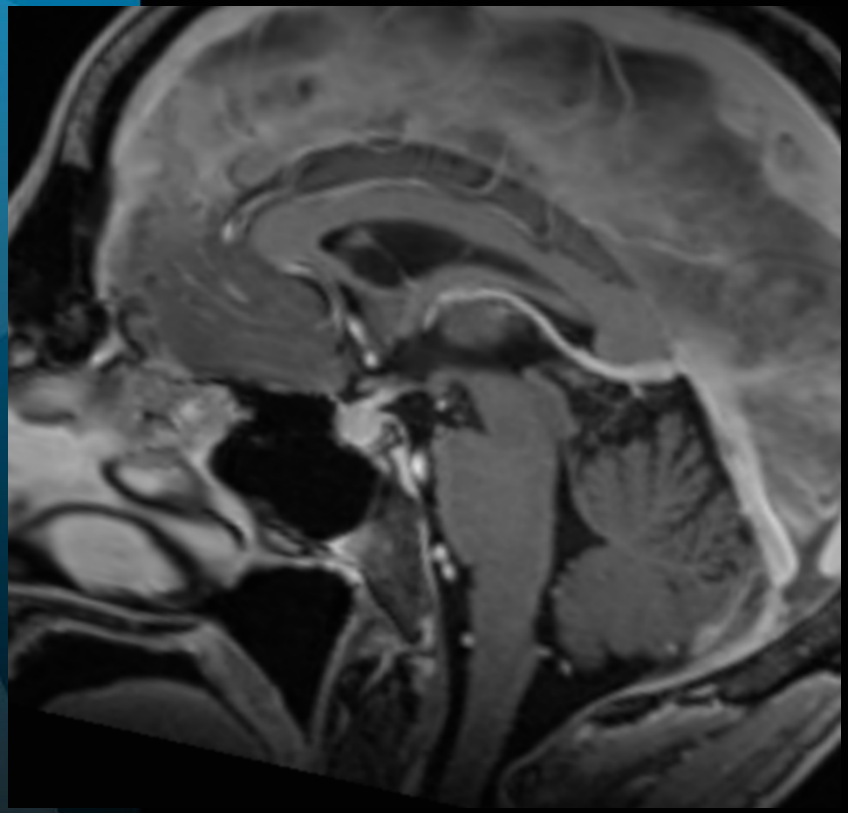
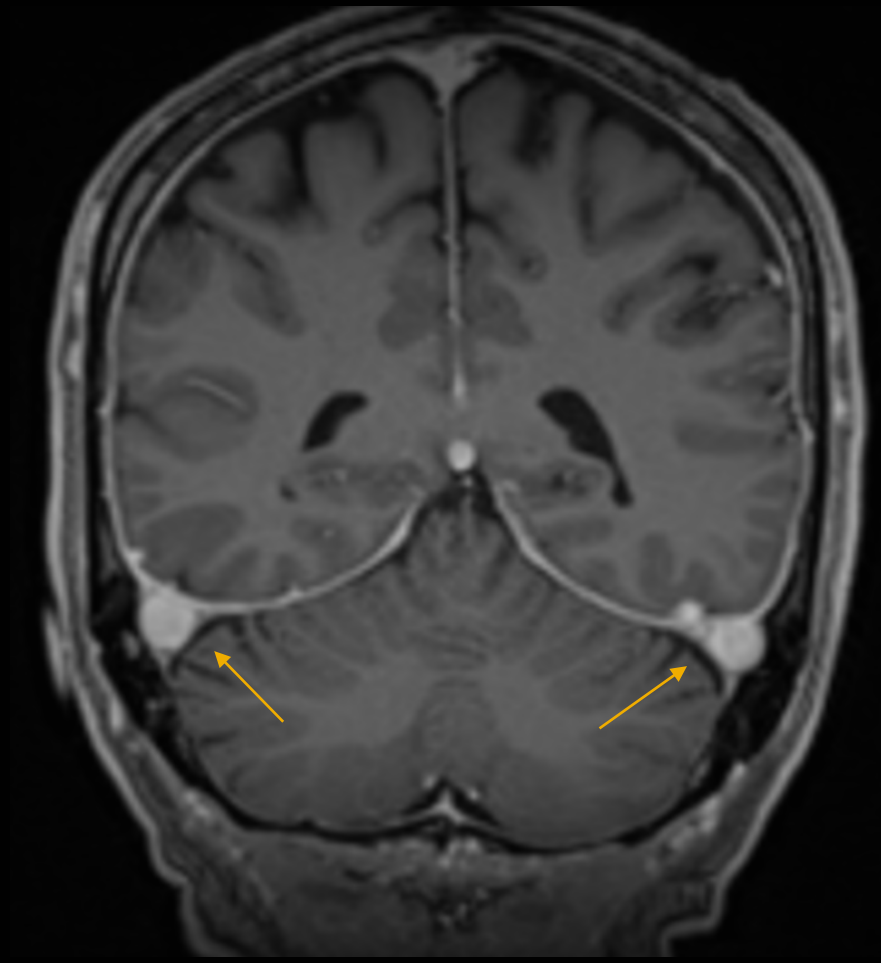
Pituitary Enlargement and Venous Engorgement





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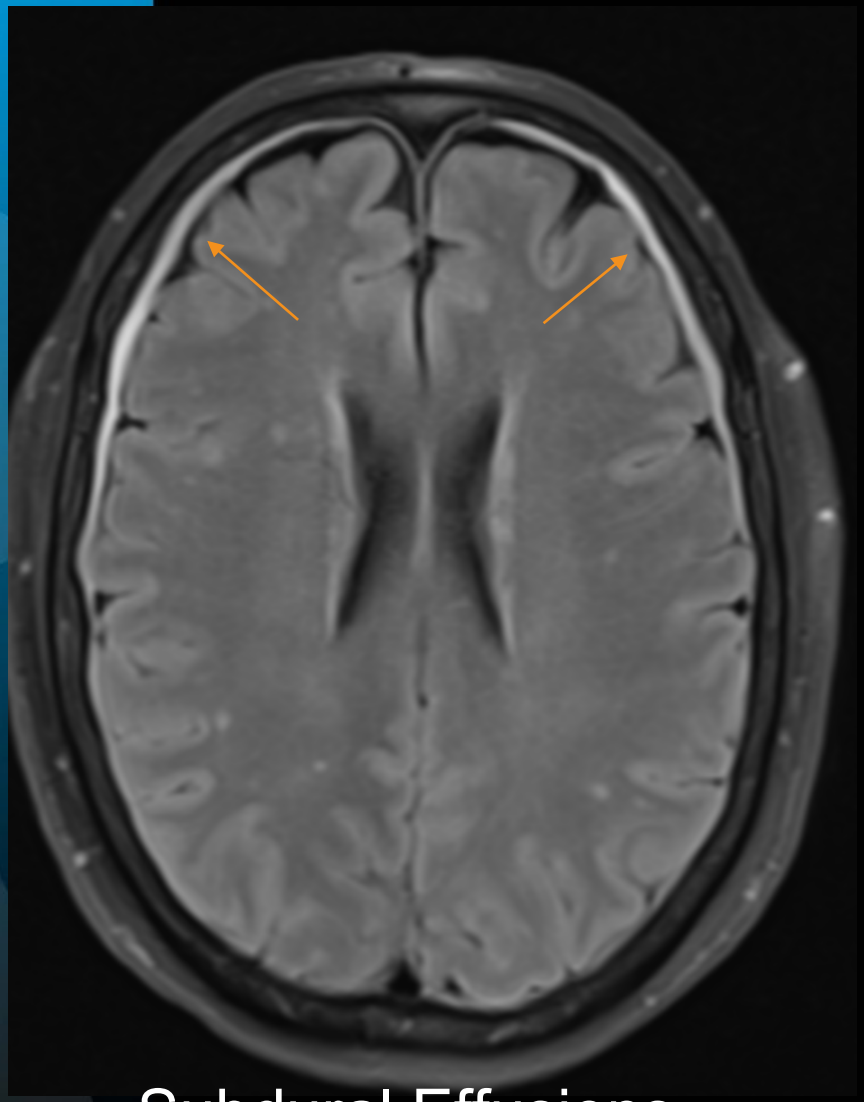
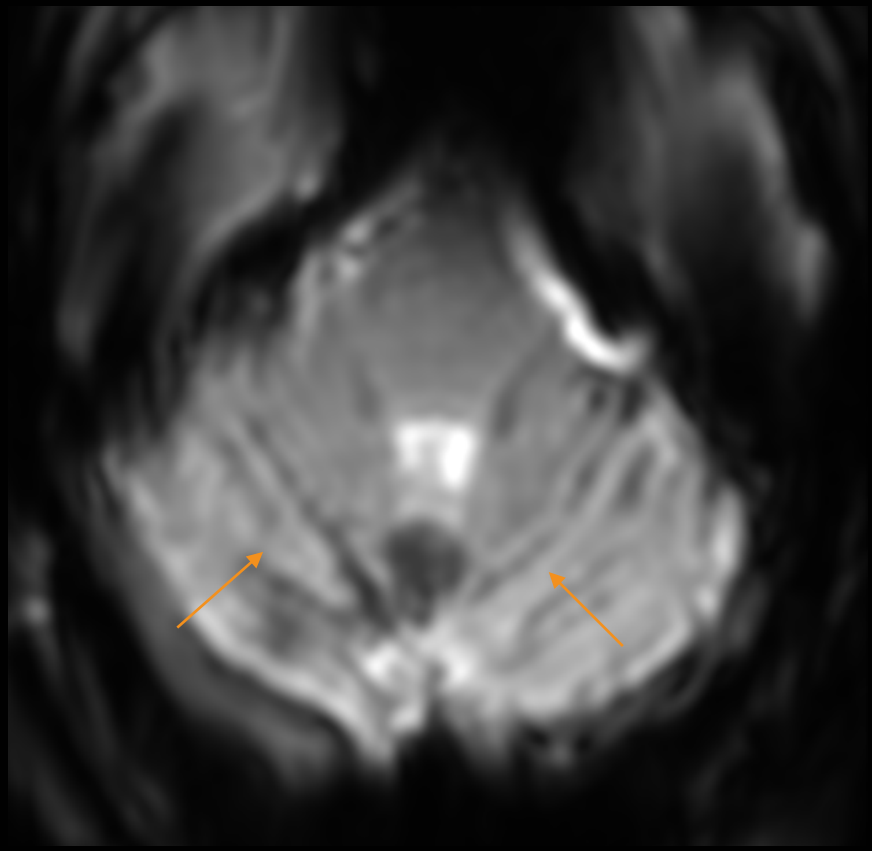


Subdural Effusions



Superficial Siderosis

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CSF Leak Types

- Type 1: Dural Tear (40%)
  - Often Ventral and Thoracic
  - Calcified Disc Osteophytes
- Type 2: Lateral Leak (17%)
  - Meningeal Diverticulum
  - Lower Thoracic
- Type 3: CSF-Venous Fistula (23%)
  - Associated with Meningeal Diverticulum
  - Thoracic Spine
- Type 4: Distal Nerve Root Sleeve Leaks (rare)
  - “Nude Nerve Root”

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Treatment Options

- Type 1: Dural Tear
  - Targeted epidural blood or fibrin glue patch
  - Surgery
- Type 2: Lateral Leak
  - Targeted epidural blood or fibrin glue patch
  - Surgery
- Type 3: CSF-Venous Fistula
  - Targeted fibrin glue patch
  - Transvenous Embolization
  - Surgery

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Case 1

Patient with history of chronic headaches which have changed in character and increased in the past 6 months. Headaches are exacerbated by positional changes, coughing and valsalva.

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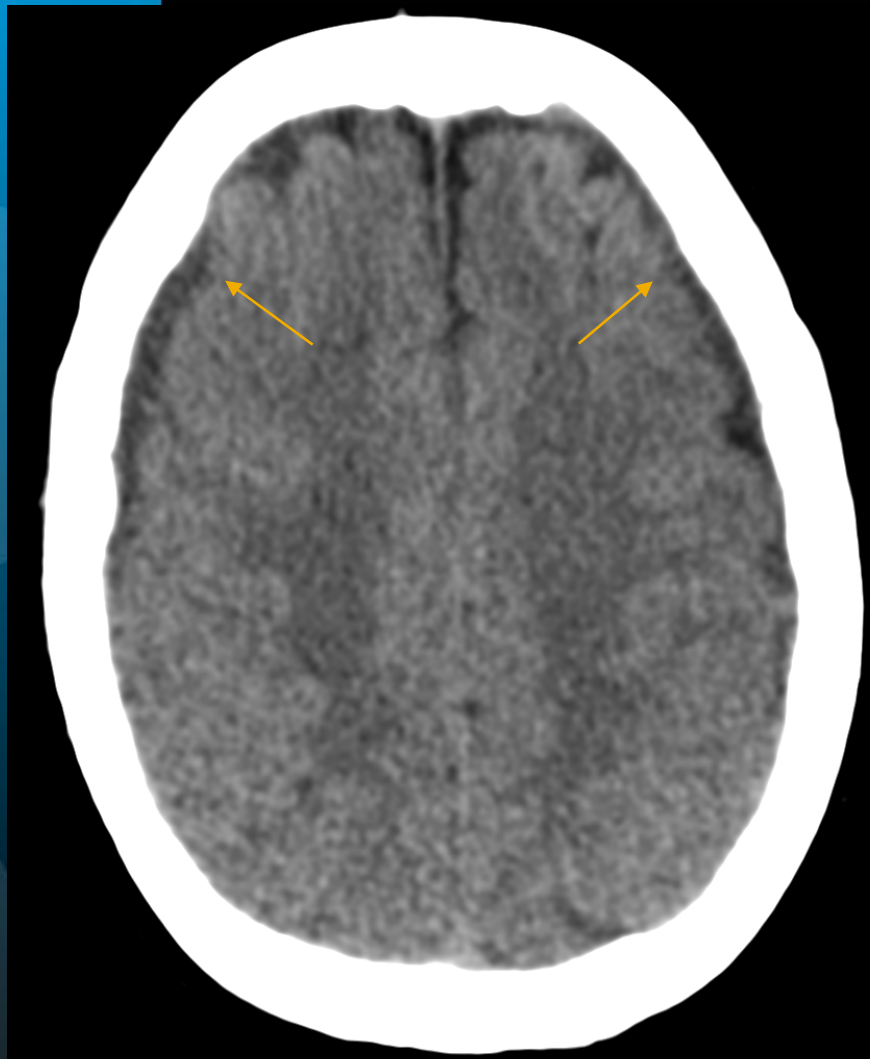

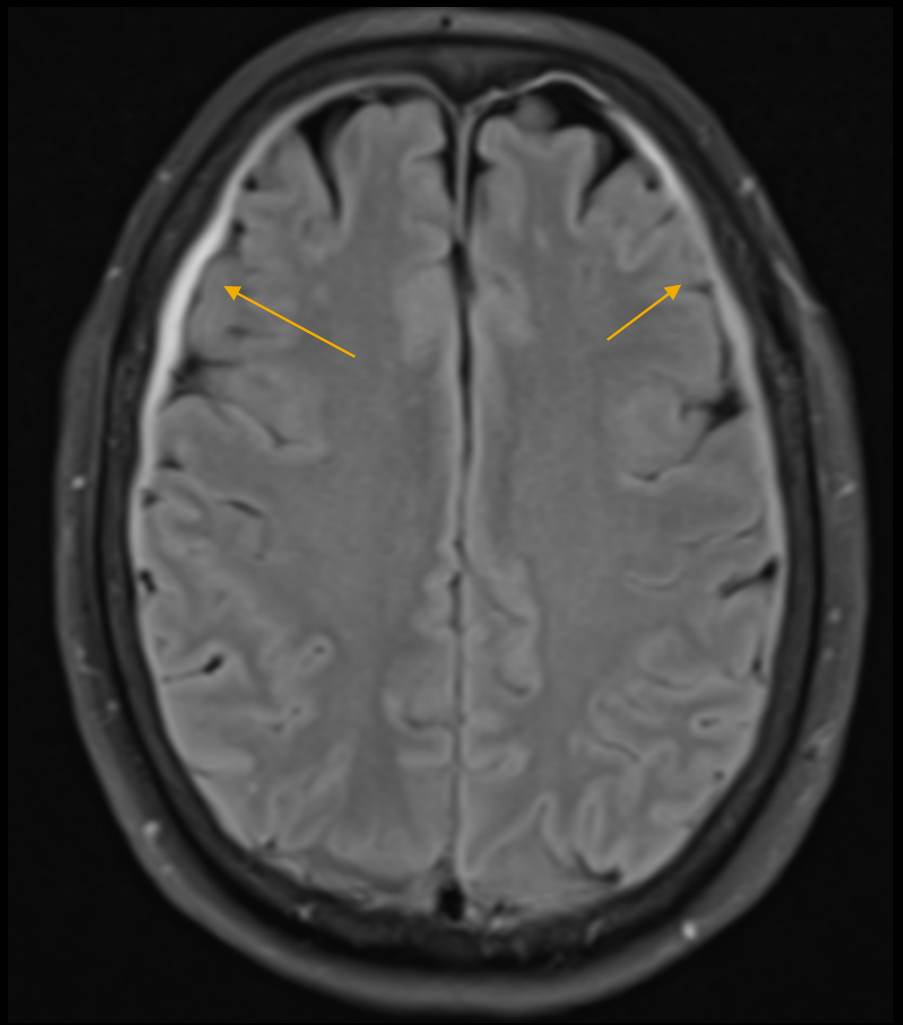

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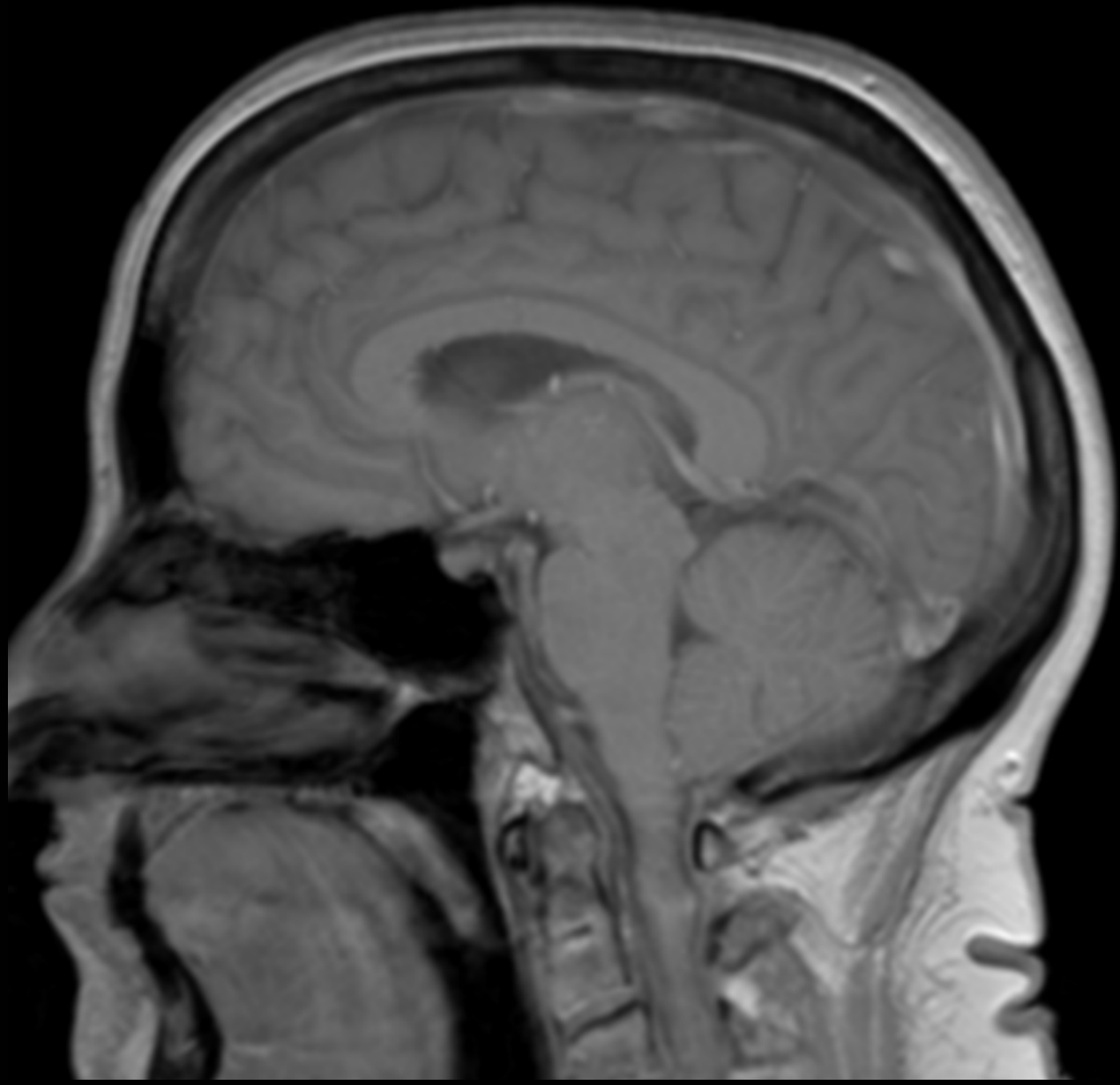

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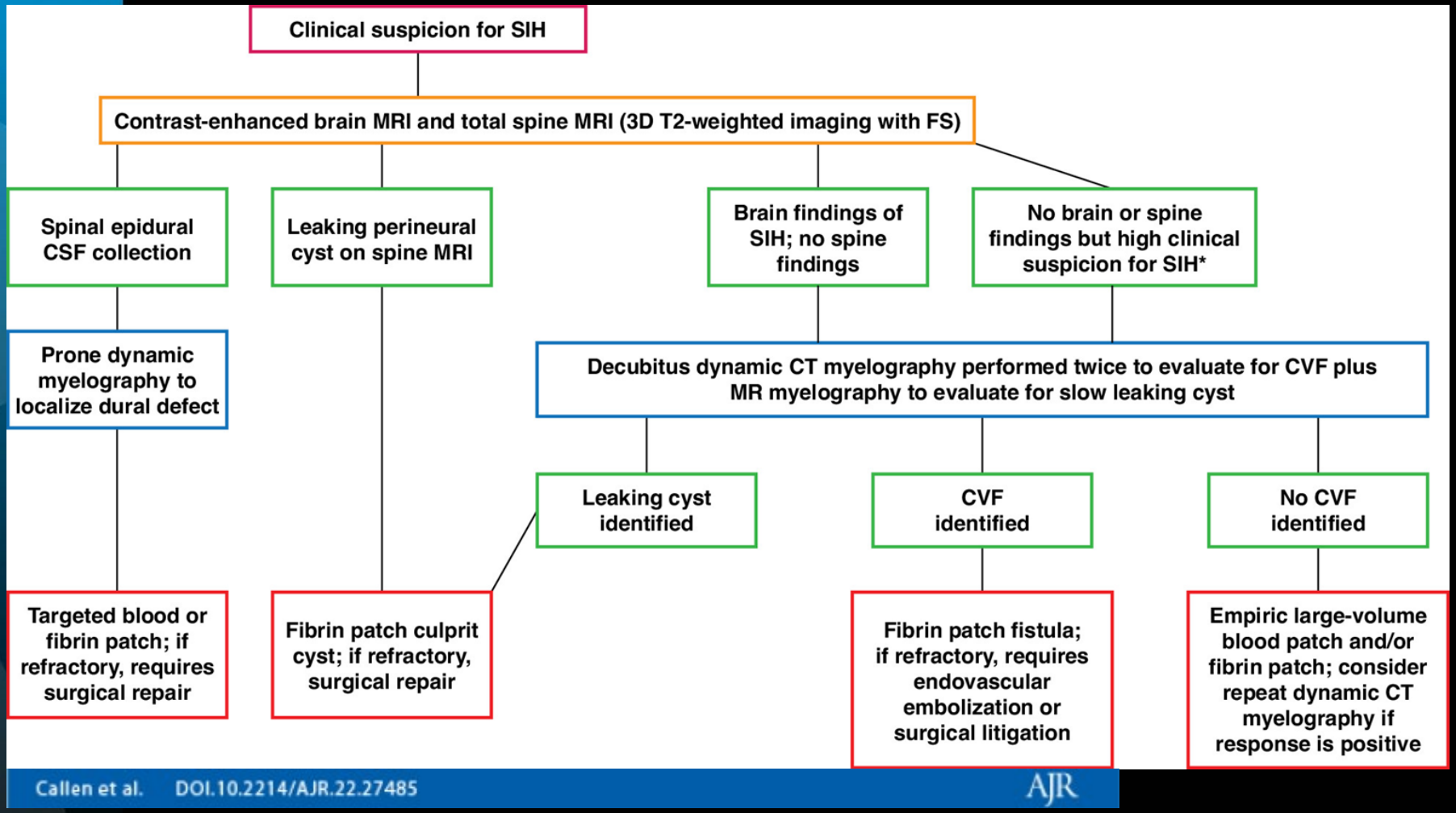

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Findings and Plan

- CT head and MRI Brain Findings Suspicious for SIH
  - Subdural effusions and Sagging Brain
- MRI Spine negative
- Algorithm:
  - Bilateral Decubitus CT myelogram performed on sequential days to evaluate for CSF venous fistula

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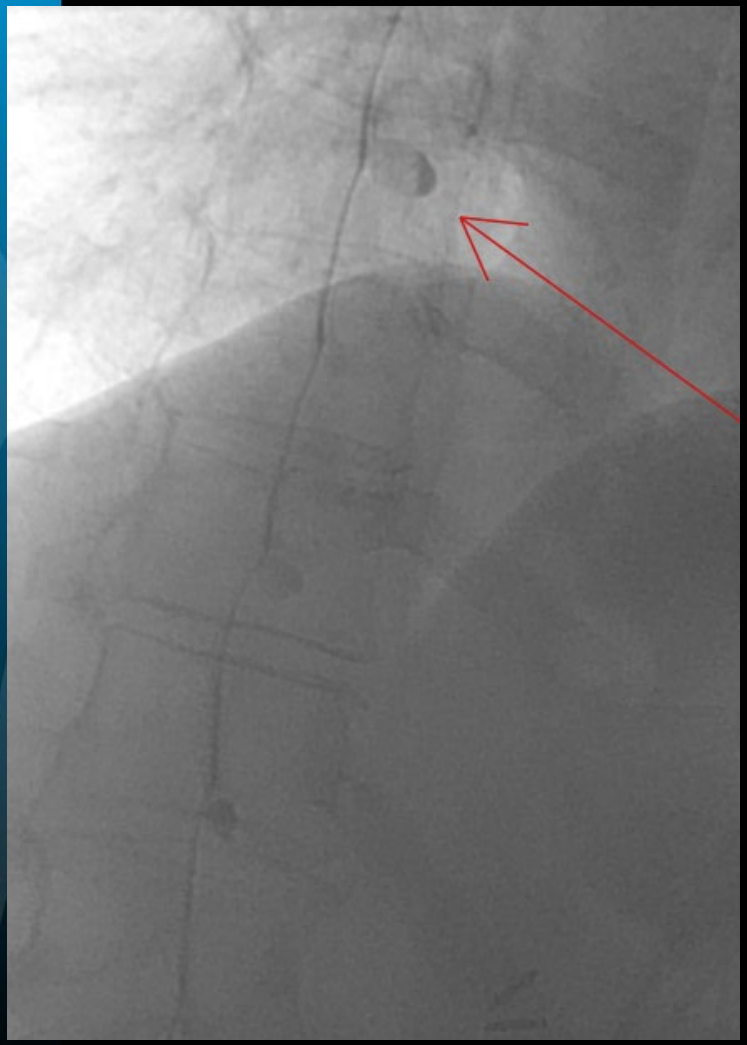

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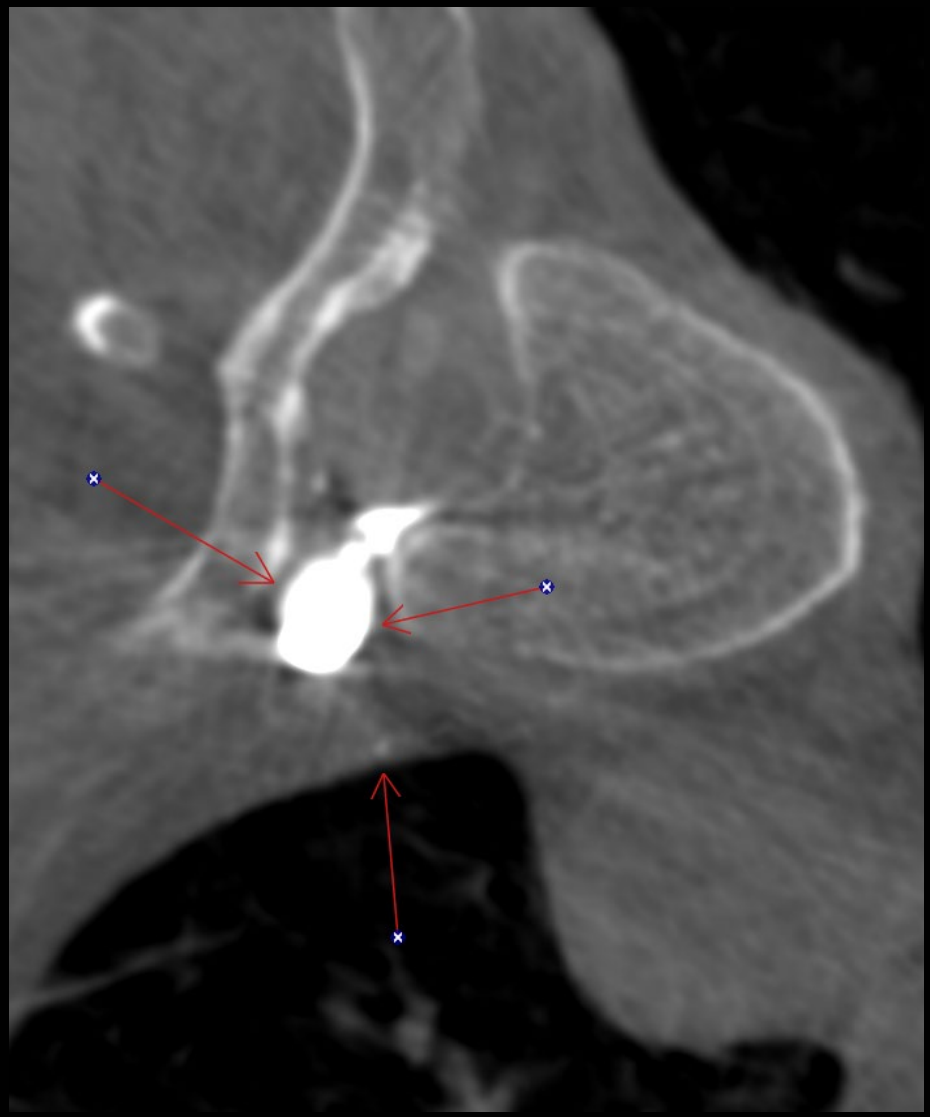

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Myelographic Findings

- Suspected CSF venous fistula arising from the left T9-10 level
- Subsequent right lateral decubitus myelogram negative for leaking diverticulum or fistula
- Plan: Targeted blood patch at T9-10 on the left performed during the second myelogram

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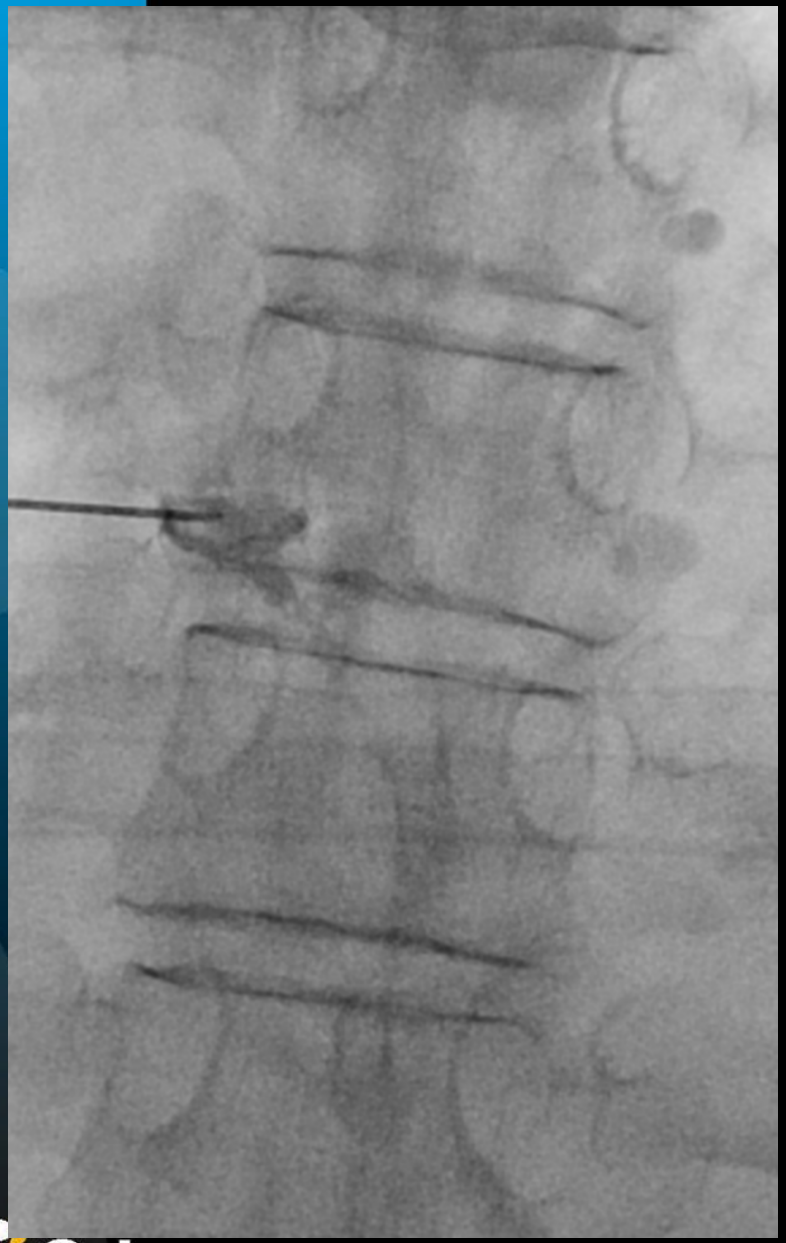

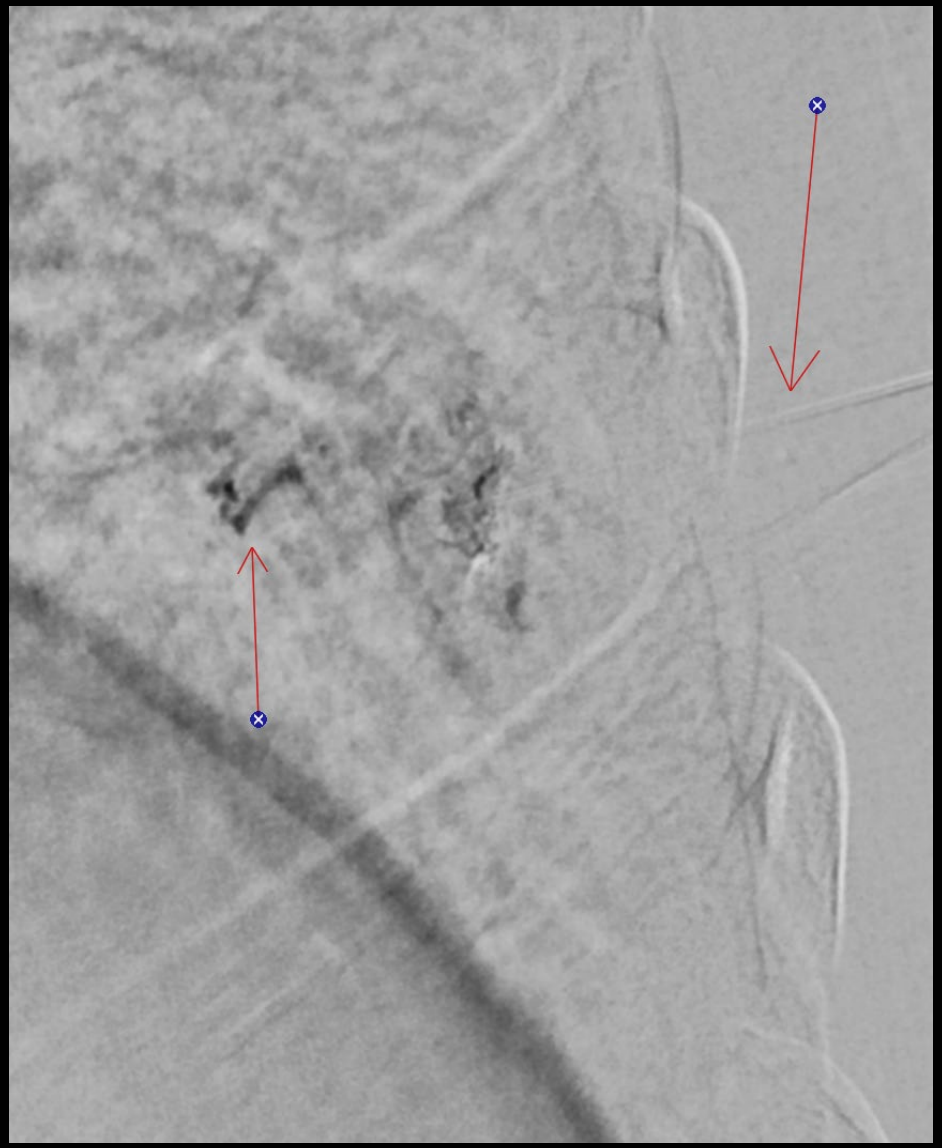

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Case 2

Patient with history of refractory headaches suspicious for intracranial hypotension. Patient underwent empiric large volume blood patch that helped with symptoms for 1 day. Patient also has history of Chiari decompression.

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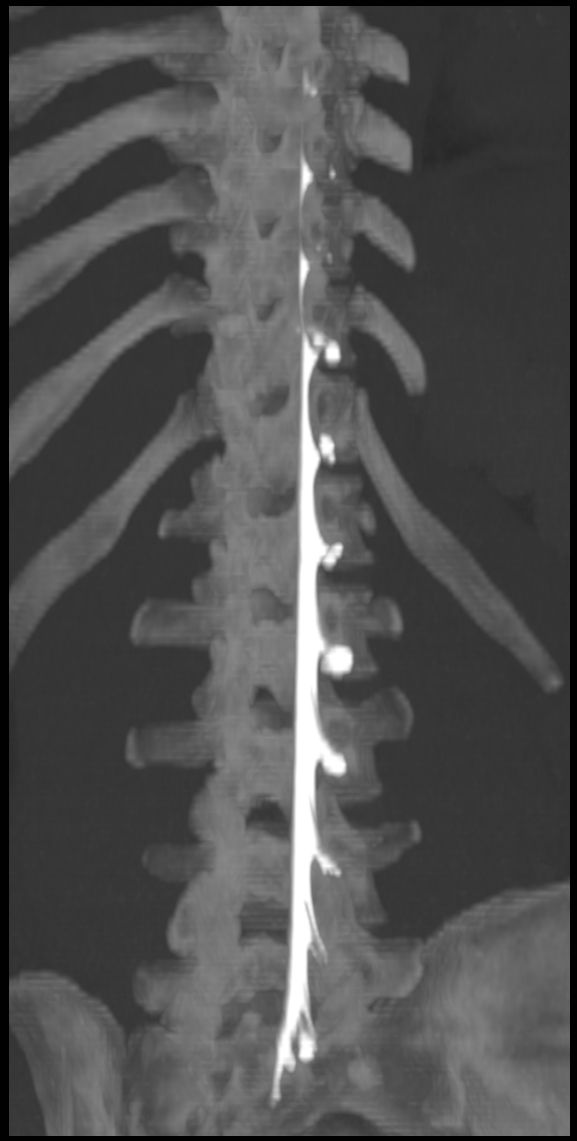

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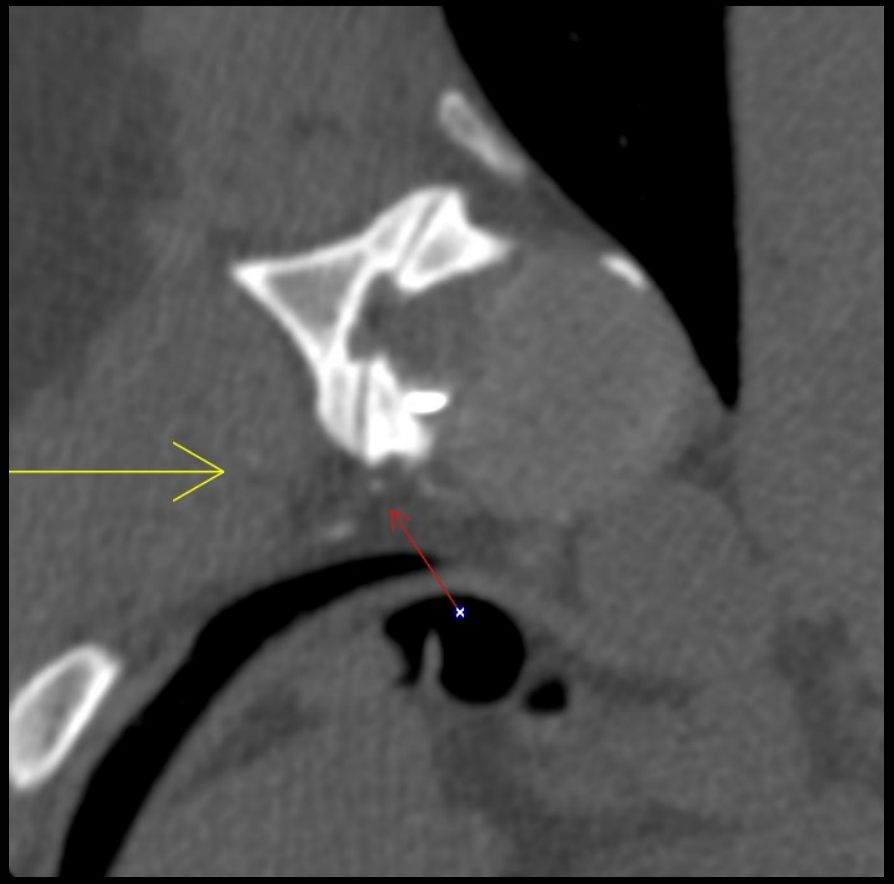

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Myelographic Findings

- Suspected CSF venous fistula arising from the left T10-11level
- Subsequent right lateral decubitus myelogram negative for leaking diverticulum or fistula
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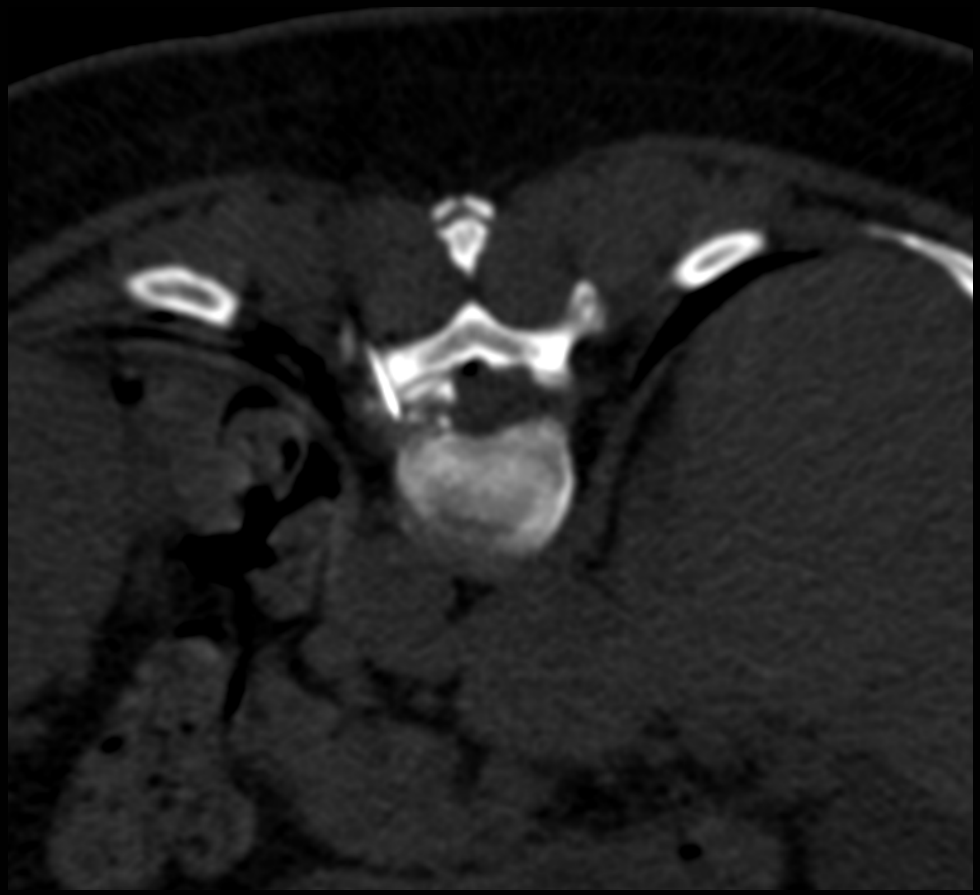
Fibrin Glue



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Conclusion

- Multidisciplinary approach is key!
  - Neurology, Neurosurgery, Radiology
- Multimodality often requiring multiple examinations and procedures
- Important: Up to 20% may have negative imaging
- Other treatment options including transvenous CSF-venous fistula embolization and surgical repair

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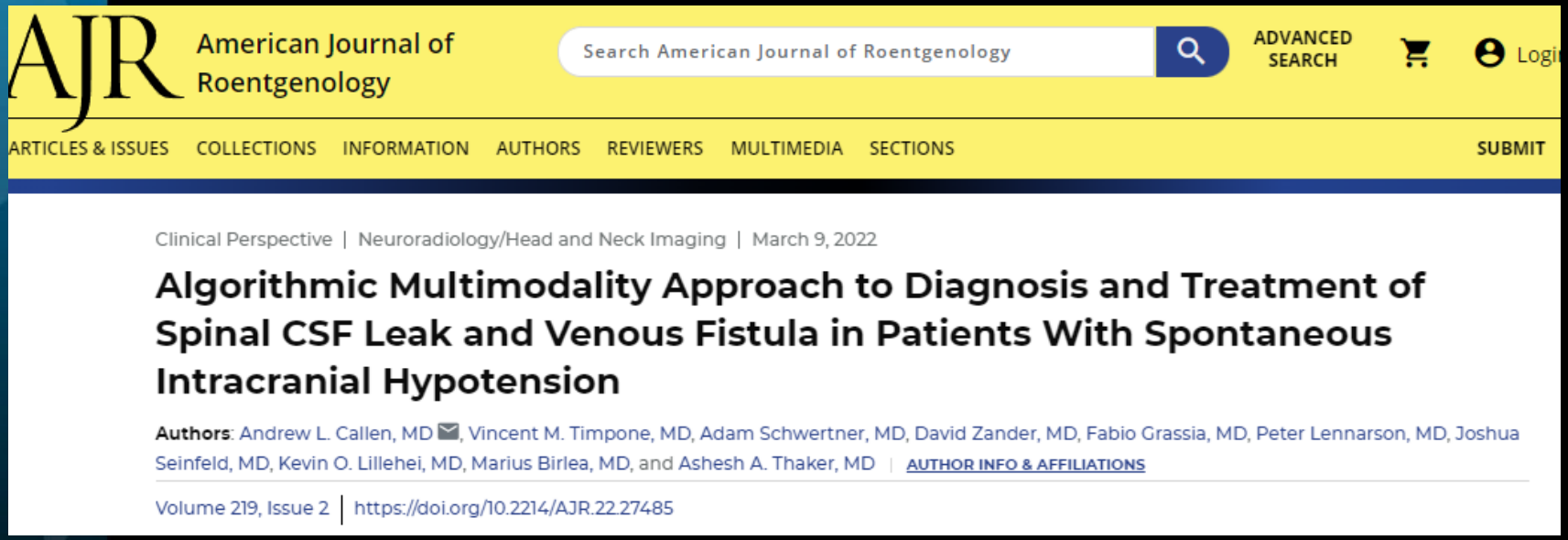
References



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THANK YOU!
QUESTIONS?

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2024 Ochsner Regional Neurosciences Symposium

Breakout Session A

2:20 PM - 3:00 PM	CSF leak and Intracranial Hypotension Panel Discussion Paul Gulotta, MD and Fawad Khan, MD

ICHD 3 definition

Headache attributed to low CSF pressure	Headache attributed to SIH
A. Any headache fulfilling criterion C.	A. Headache fulfilling criteria for *Headache attributed to low CSF pressure*, and criterion C below.
B. Either or both of the following: 1. Low CSF pressure (<60 mm CSF) 2. Evidence of CSF leakage on imaging	B. Absence of a procedure or trauma known to be able to cause CSF leakage.
C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage or led to its discovery.	C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage or has led to its discovery.
D. Not better accounted for by another ICHD-3 diagnosis.	D. Not better accounted for by another ICHD-3 diagnosis.

Mehta et al, Frontiers in Neurology 2023

- 5 per 100 000 individuals every year
- The duration of symptoms at the time of diagnosis and/or treatment was variable ranging from 1 day to 19.7 yrs

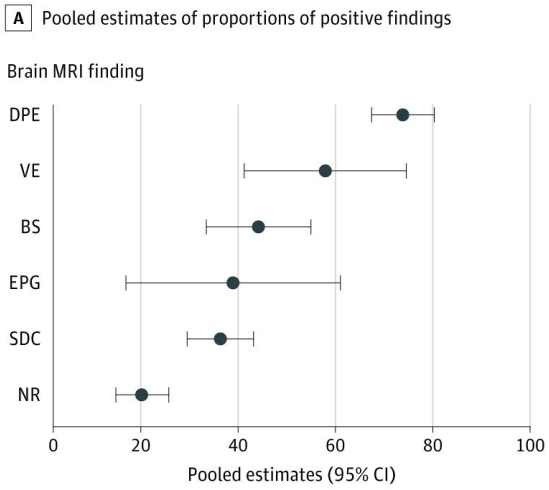
Causes

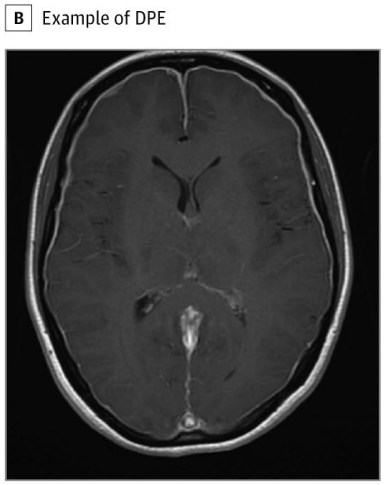
- Weakness of the spinal meninges- development of meningeal diverticula or perineural cysts, and subsequently spontaneous dural dehiscence.
- Calcified degenerative thoracic disc protrusions may cause trivial traumatic events leading to progressive dural tears, which can be either ventral or lateral
- CSF–venous fistula (CVF)
- Congenital absence of the entire nerve root sleeve
- Connective-tissue disorders, particularly the Marfan syndrome, the Ehlers–Danlos syndrome (type II), and autosomal dominant polycystic kidney disease (ADPKD)
- Spinal pathologies (ie, osteophytes, disc prolapse, and discogenic micro spurs)
- Bariatric surgery

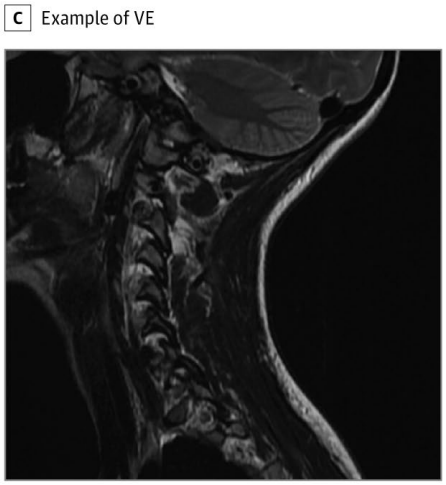
Opening pressure

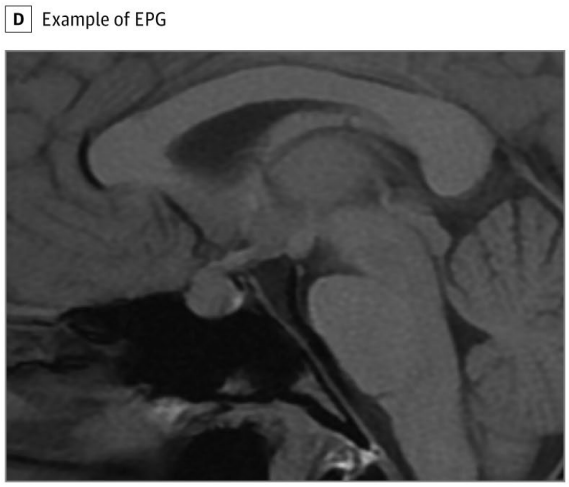
- Up to 80% of patients had low pressure (<60 mm H_2O)
- Up to 44% had normal pressure (60-200 mm H_2O)
- Up to 6% had high pressure (>200 mm H_2O). The highest reported opening pressure was 228 mm H_2O

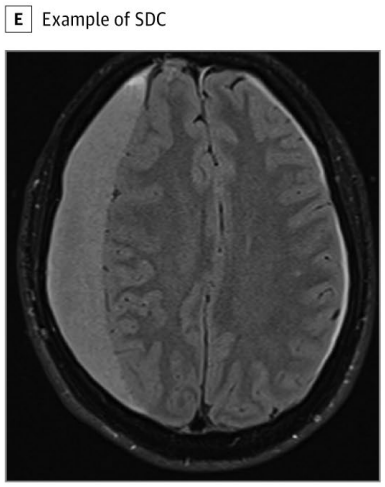
- Tissue compliance determines the physiological relationsh between pressure and volume
- Compliance with regards to the CSF component is highly dynamic and variable between individuals, as it is influenc by posture (upright vs. recumbent), body habitus, and the extent of the spinal epidural venous plexus
- Low volume compartment!

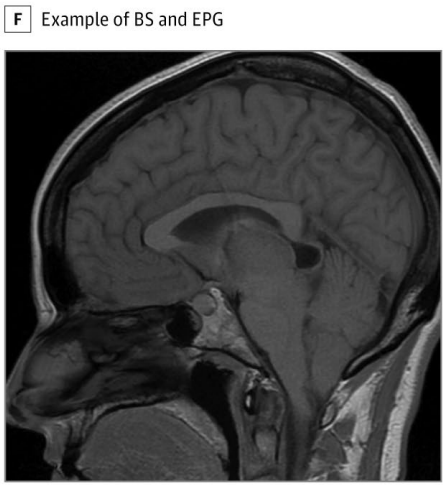












2078 patients

B, Example of diffuse pachymeningeal enhancement (DPE) in T1-weighted axial MRI sequence. C, Example of venous engorgement (VE, transverse sinus venous distension sign) in T2-weighted sagittal MRI sequence. D, Example of enlarged pituitary gland (EPG) in T1-weighted sagittal MRI sequence. E, Example of subdural collection (SDC) in T2-weighted axial MRI sequence. F, Example of brain sagging (BS) and enlarged pituitary gland (EPG) in T1-weighted sagittal MRI sequence. NR indicates normal.

Presence of extradural CSF was detected in 48% to 76%

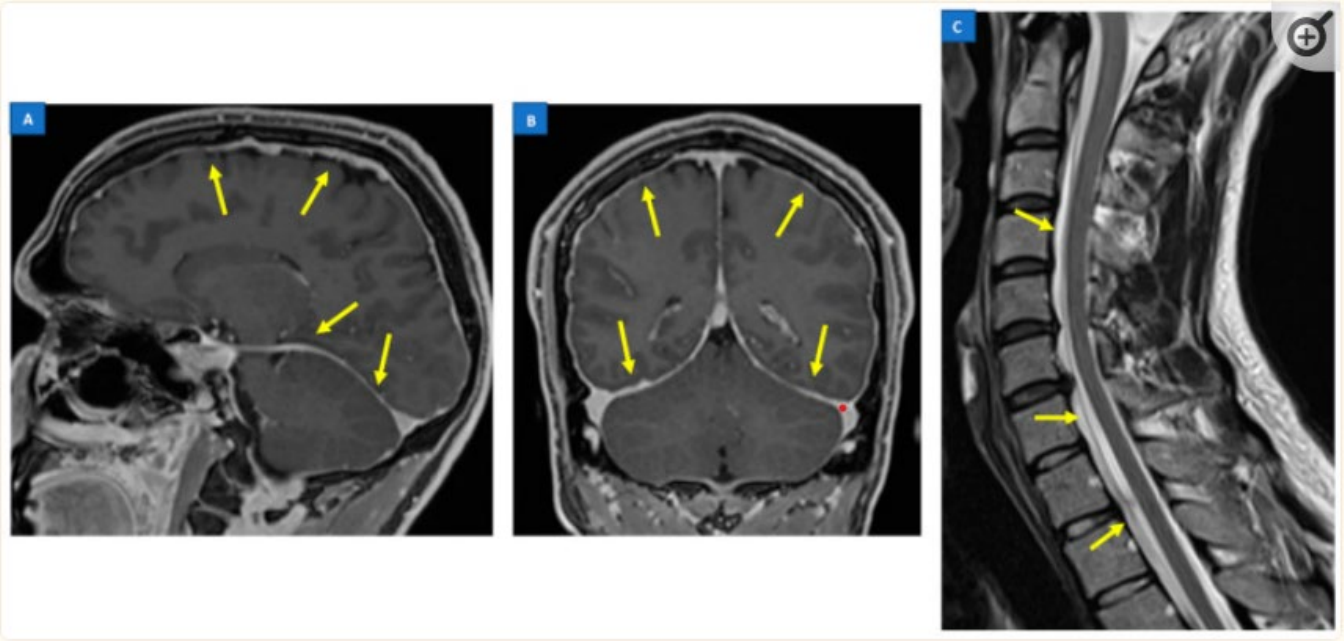


Figure 3

Smooth and diffuse pachymeningeal enhancement (arrows) in a post-gadolinium MRI ((**A**,**B**): sagittal and coronal planes): the most sensitive intracranial sign pointing toward SIH. Cervicodorsal spine MRI (**C**) showing the presence of a spinal longitudinal extradural collection (SLEC) (arrows), which is a specific sign of SIH with dural mechanical tears along the thecal sac.

Other tests

spinal MRI
computed tomography myelograms (CTM)
dynamic CTM
digital subtraction myelography (DSM)
MR myelography
radionuclide cisternography.

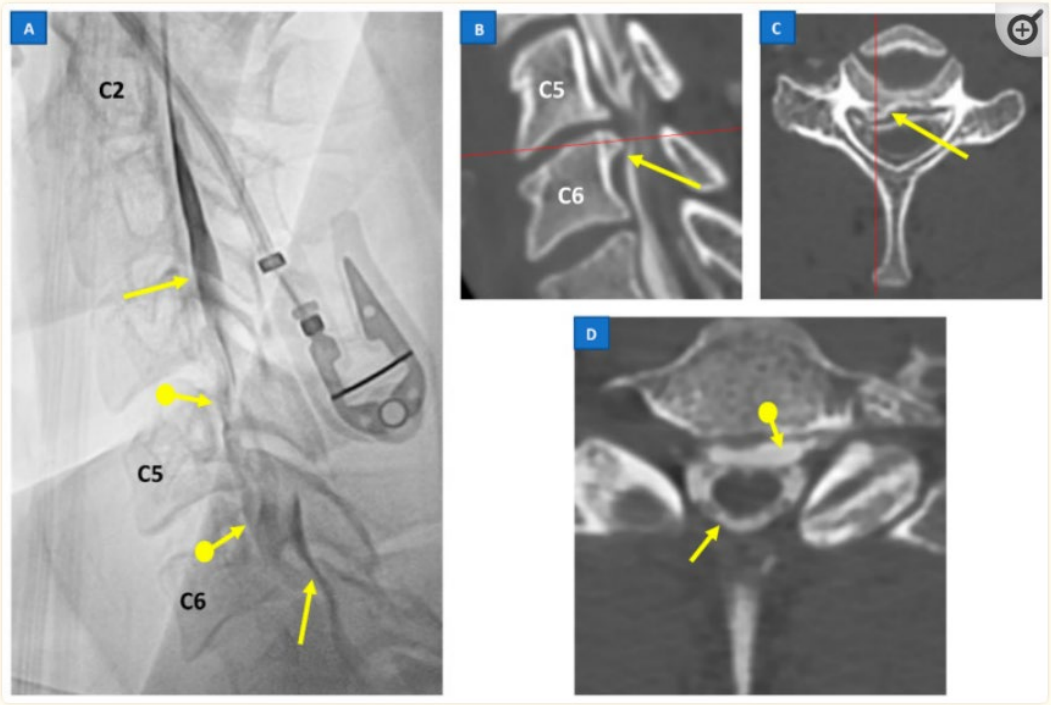


Figure 1

Prone myelography (**A**) showing a regular ventral profile of the dural sac (arrows) with anterior extravasation at C5–C6 into the epidural space (arrows with dot). Myelo-CT ((**B**,**C**): sagittal and axial planes) shows the presence of a C5–C6 disc herniation, with an osteophyte (bone spur) of the upper somatic margin of C6 (arrows), in the right paramedian site, probable cause of the dural tear (the red line indicates in image (**B**) the axial plane of image (**C**) while in image (**C**) the sagittal plane of image (**B**)). Myelo-CT (**D**) also shows the presence of contrast in the epidural space (arrow with dot), anterior to the dural sac (arrow).

Decubitus CT Myelography

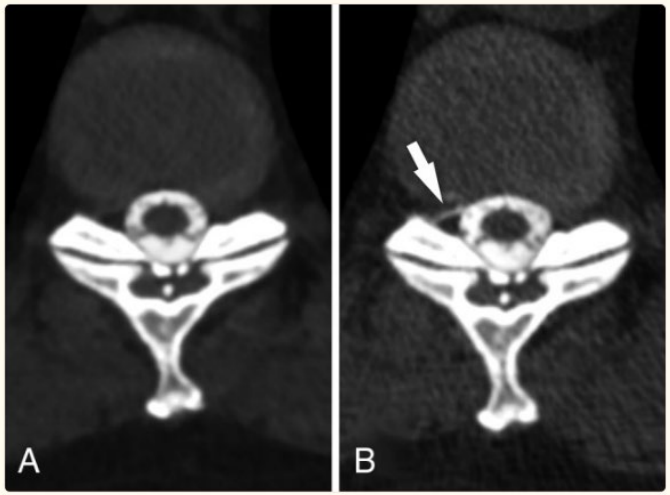


Fig 5.

Low-flow CSF leak seen only on decubitus myelography. Axial prone CTM image (*A*) shows no CSF leak at the T10–11 level. The patient was turned into the left lateral decubitus position and re-scanned, again with the scan showing no leak (not shown). The patient was then turned to the right lateral decubitus position. An axial CTM image from the decubitus myelogram (*B*) obtained 6 minutes later shows a low-flow CSF leak not seen on prone myelogram (*arrow*).

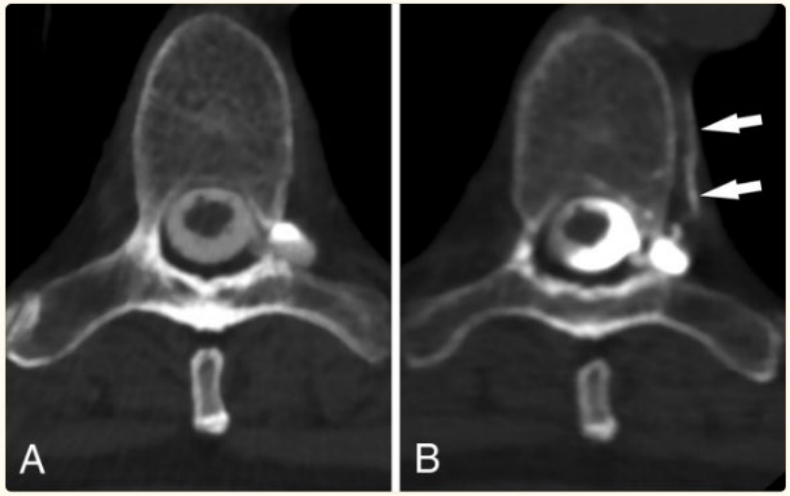


Fig 4.

CSF venous fistula visualized best on decubitus CTM. Axial prone CTM image (*A*) shows a perineural diverticulum, but no clear leak. Axial image (*B*) from a subsequent CTM obtained with the patient in the decubitus position after dynamic myelography shows clear filling of a segmental vein (*arrows*), confirming a CVF.

Chiari vs SIH

- Chiari malformation type I (CM1) is classified as a rare disease and is historically defined as cerebellar tonsillar position greater than or equal to 5 mm below the level of the foramen magnum
- SIH shares some MRI features with Chiari malformation type I (CM1), especially low-lying cerebellar tonsils.
- Both can cause chronic headaches
  - CM1 generally does not cause an abrupt orthostatic chronic daily headache, but rather causes headaches in the neck/occipital area, often as nonprogressive short attacks of 'cough headaches'
  - SIH is associated with orthostatic headache within thirty minutes of standing and should be alleviated within 15–30 min after lying in a recumbent position, especially early in the disease course
  - SIH can also cause a 'cough headache', but it is usually progressive in nature and can be associated with an underlying daily headache
- CM1 treatment involves suboccipital craniectomy, while treatment for SIH involves invasive interventional procedures or spinal surgery.
- A misdiagnosis can lead to delays in care and incorrect treatments, such as unnecessary surgery or procedures, with a needless risk of surgical or procedural complications.
- Imaging features:
- Measures of midbrain sagging, including cut-off values for slope of the third ventricular floor and pontomesencephalic angle, may help discriminate CM1 from SIH
- Low-lying cerebellar tonsils from SIH should maintain the normal tonsillar rounded shape
- Pachymeningeal enhancement and other MRI features, including pituitary hyperemia, subdural collections, and venous engorgement, may be a useful means of distinguishing SIH from CM1. [these features are not always present]
- Features associated with CM1, including syringomyelia and tethered cord, are not commonly observed in SIH [these features are not always present].

Postural Orthostatic Tachycardia Syndrome (POTS) vs SIH

- A cardio-autonomic disorder
- Affects 1-2% of the U.S. population
- Typically young, White women
- Joint hypermobility syndromes common (EDS)
- Gastrointestinal disorders (e.g., irritable bowel)
- Other comorbidities (e.g., MCAS, migraine)
- Def: At least 30 bpm increase in heart rate in adults or 40 bpm or more in children OR an absolute heart rate of > 120 bpm within 10 min of standing
- Secondary causes must be excluded (medications, deconditioning/post-COVID, dehydration, hyperthyroidism, anemia, orthostatic hypotension, arrhythmias)

- Headache in POTS
- > 1/3 experience headaches (orthostatic headache can occur)
- Most patients have non-orthostatic headaches that are migraine-like
- Syncope, worsening with menses, and myofascial pain more common with POTS

Treatment

- **Conservative treatment:** Authors reported a successful conservative treatment (resolution of symptoms with no further intervention needed) in up to 37%
  - Bed rest, Hydration, Caffeine, Abd binder, Amantadine
- **Epidural blood patches:**
- First EBP was reported to be successful 56%-72%
- [similar proportions of successful results for the targeted and nontargeted techniques]
- Large EBPs (>20 mL) had a higher success rate than small EBPs
- No serious adverse events were reported after EBP treatments.
- The minor transient adverse events included back pain, radicular pain, tinnitus, paraesthesia, numbness, bradycardia, and dizziness
- <u>Post procedure HA:</u>
- The incidence of rebound intracranial hypertension after treatment (EBP, percutaneous, or microsurgical treatment) has been reported to be between 7% and 27.4%.
- Spread of autologous blood in the subarachnoid space has been reported as a complication of EBP occurring in 8.5% of procedures. transient (mostly intraprocedural) symptoms: palpitation, nausea, and headache

Surgical treatment

- Surgical repair of dural defects, meningeal diverticula, or CSF-venous fistulas

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Review Article

Spontaneous Intracranial Hypotension: 10 Myths and Misperceptions

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1. Currently, there is no single diagnostic test that excludes SIH with a high level of sensitivity. When SIH is suspected, a combination of brain imaging, spine imaging, and CSF pressure measurement may be necessary for a complete evaluation.
2. Orthostatic headache is the most common, but not the only, clinical presentation of SIH. The lack of orthostatic headache should not exclude the diagnosis when there is objective imaging evidence of SIH.
3. Orthostatic headache can be caused by other conditions such as POTS, cervicogenic headache, and craniocervical instability.
4. Diffuse, smooth dural enhancement on brain MRI is a highly specific finding for SIH.
5. Cerebellar tonsillar ectopia can be seen with both SIH and Chiari I malformation. Other imaging signs of brain sagging can help differentiate these entities.
6. Spinal imaging is often useful in detecting CSF leaks, which can help to confirm the diagnosis of SIH and direct treatment efforts.
7. Causes of CSF leaks include meningeal diverticula, ventral dural tears caused by disk protrusions or osteophytes, and CSF-venous fistulas.
8. SIH is not typically caused by skull base CSF leaks.
9. Recovery from spontaneous spinal CSF leaks differs from lumbar puncture related headaches, typically taking longer.
10. Recovery from SIH can be complicated by RIH. Awareness of this entity is important for appropriate management after treatment for a spinal CSF leak.

Prognosis

- Mixed
- Subdural hematoma
- Cerebral vein thrombosis
- Spinal cord herniation
- Superficial siderosis

Case discussion

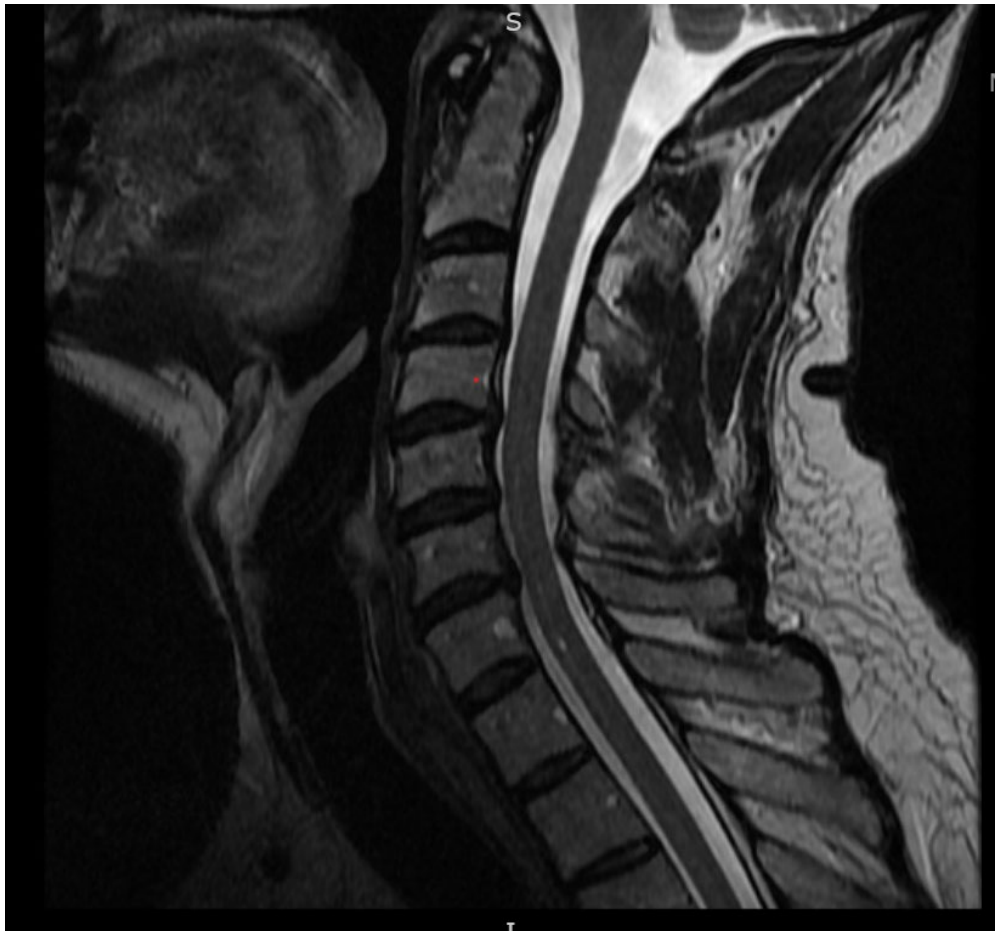
- **55 yr Male**
- In 2015 had fall at home - onset of LBP - dull ache
- MRI - unrevealing , PT - no help
- 2016 facet inj L5/S1 - not help , PT – no help
- Jan 2017 throbbing orthostatic HA onset
- Autoimmune work up - neg
- Inpatient for severe HA exacerbation, on dilaudid
- MRI spine - unremarkable
- Blood patch July - not help
- Repeat admission - dilaudid 2 mg Q6,
  - ativan 8 mg/day, pred 20 mg/day
- Aug 8 Mayo - imaging - unremarkable
- cisternogram - pressure 23, no thinning noted
- LP - CSF pressure 59
- Rheum work up – neg

Intracranial BOLT
Provocative tests

Activity	Pressure	Pain	
Sitting	-2 to - 4	3.5	
Lying	4 to 5	3.25	
Symptomatic episode while sitting	+3 to -11	3.75	
Trendelenburg	18 to 21	3.25	
Trendelenburg with pillow	13 to 15	3.25	
Standing	-3 to -9	3.5	
Walking	**-3 to -9**	**3.75**	
Hyperventilation	**-9 to -13**	**4.0**	
Squatting	-3 to -5	3.5	
Standing up quickly	-7	3.5	
Valsalva	17 to 20	3.75	

Case discussion

- 2018-2022: medications that provide some temporary, and moderate, pain relief: Diazepama (30 me daily), Prednisone
- Failed : Galcanezumab, Botos for migrates (two sets of injections), amitriptyline, CBD Oil, Hydroxyzine, nortriptyline, DHE, acetazolamide , gabapentin, duloxetine, baclofen, ketamine infusions
- CT MYELOGRAM 1/12/22 IMPRESSION:
  - 1. Negative for CSF leak or CSF to venous fistula. There is congenital absence of the right C5 cervical pedicle causing retrolisthesis of the facet on the right at the C5 level. There is also spina bifida occulta involving the C5 vertebral body. Significant degenerative changes are noted at the C5-6 level with disc osteophyte present.
  - 2. Significant facet arthropathy at the L5-S1 level bilaterally.
- Working Diagnosis: cervicogenic headaches. cervicogenic pain at C5-6 and recommended fusion.
- He underwent a lidocaine injection near the C5 root which resulted in 45 minutes of pain freedom. He then underwent an injection with steroids in this same region which had no effect.
- Second opinion: patient undergo MBB w/ RFA.
- Failed MBB C4,C5
- Failed ESI c5-6
- Back in New Orleans he had a repeat lidocaine injection near the C5 root at total hospital. He again experienced a 45 minute pain relief.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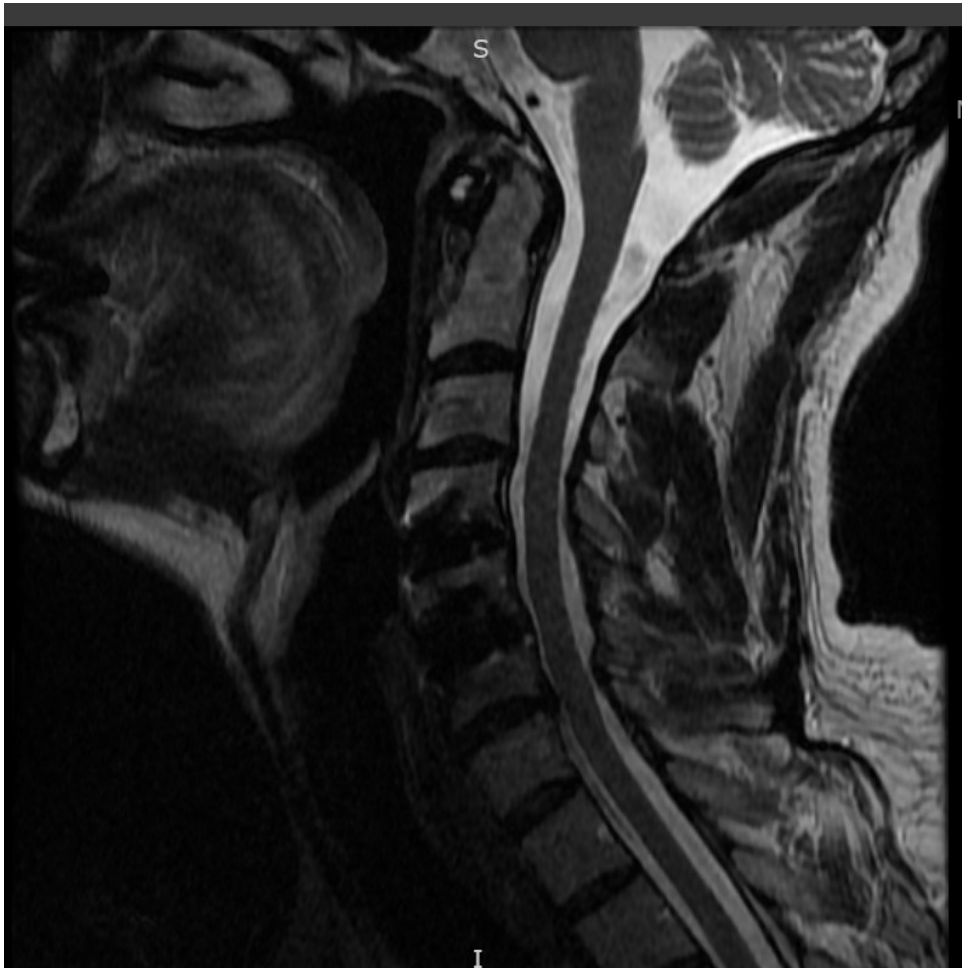
Case discussion

- Cervical fusion surgery in April 2022.
- Feeling much better in July and August (being upright for 8-10 hours/day without significant pain), he started going downhill in early September.
- Sept 2022-current: chronic headache – PT and pregabalin
  - Patient notes primary difficulty is upright standing tolerance before HA becomes limitation. HA begin at back of head and progress to top of head. This limits his tolerance to household chores (cooking end of day) and comfort /c travel.
  - Symptoms interfere with daily activity, sleeping and work.
  - Exacerbating factors: Sitting, Standing and Getting out of bed/chair.
  - Mitigating factors laying down and rest. States that he lays down up to 21 hours a day.



MRI Jan 2024

- At C2-3, there is mild facet joint arthropathy. No spinal canal stenosis or significant neural foraminal narrowing.
- At C3-4, there is broad-based posterior disc osteophyte complex with uncovertebral spurring and facet joint arthropathy, contributing to moderate left and mild right-sided neural foraminal narrowing. No spinal canal stenosis.
- C4 through C6 is fused
- At C6-7, there is broad-based posterior disc osteophyte complex with ligamentum flavum hypertrophy, contributing to moderate spinal canal stenosis and moderate bilateral neural foraminal narrowing.
- At C7-T1, there is mild facet joint arthropathy. No spinal canal stenosis or significant foraminal narrowing.

Second opinion pending

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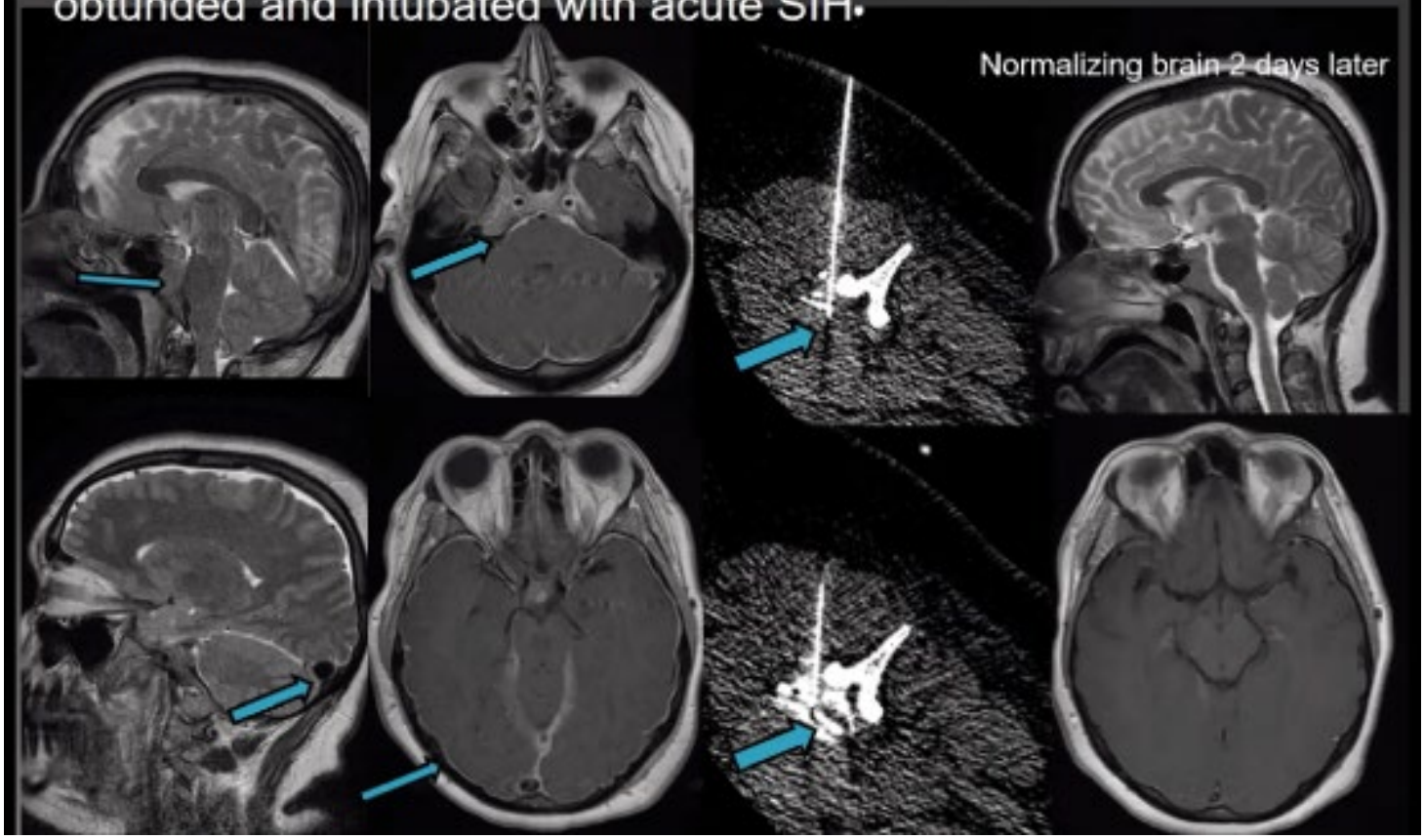
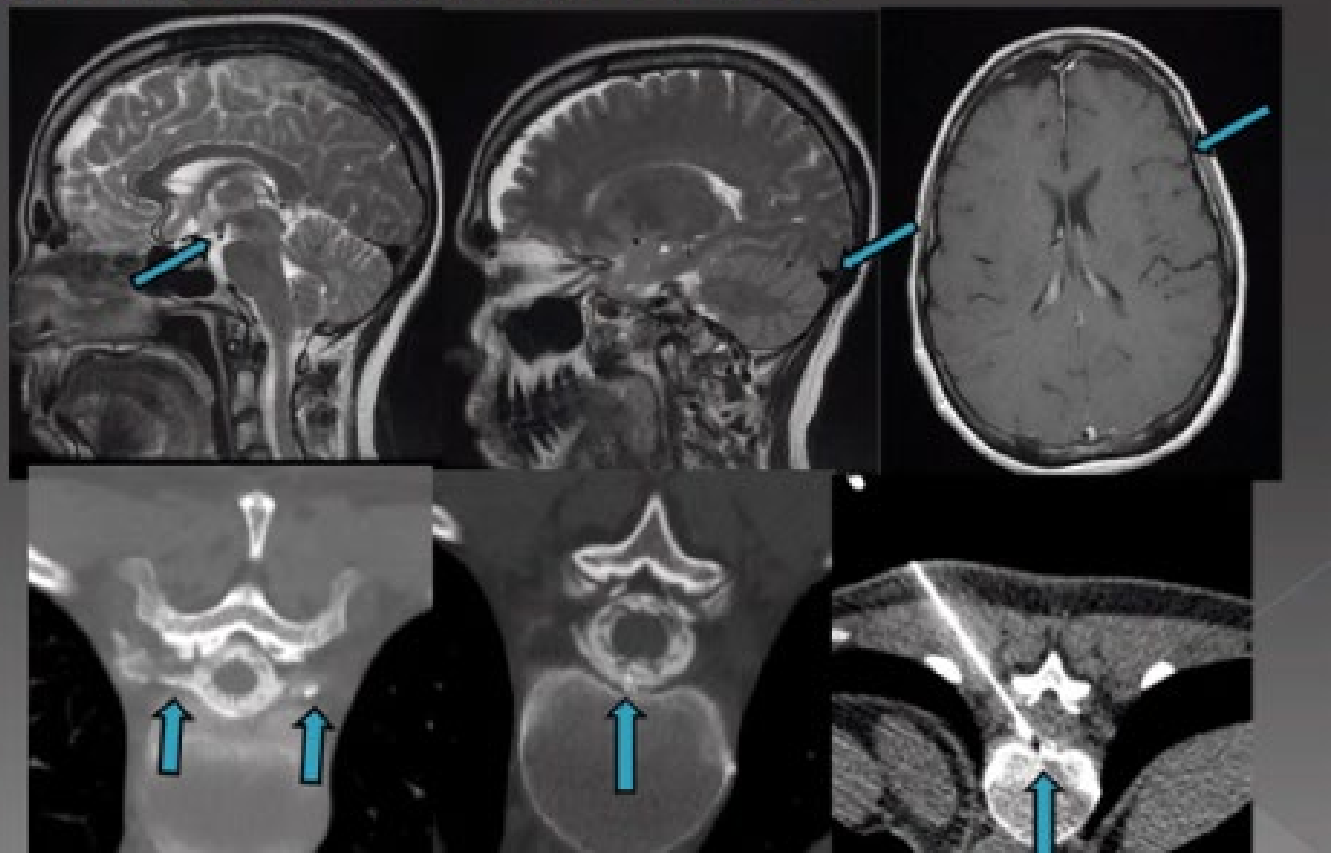
DUKE cases

Targeted Patches----Circumferential Patch:

21 yo s/p 4 attempts at lumbar epidural for delivery presents obtunded and intubated with acute SIH.



56 yo rocking on her porch in the mountains of NC suddenly develops a 10/10 HA sitting or standing and 0/10 when supine, despite NL Brain MRI, thoracic CSF leak