

Clinical + Imaging Presentation of CVES



Bridging the Gap 2024

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University of Colorado
Anschutz Medical Campus

Disclosures

Medical Advisory Board:
Spinal CSF Leak Foundation
Spinal CSF Leak Canada

Consultant:
Eli Lilly

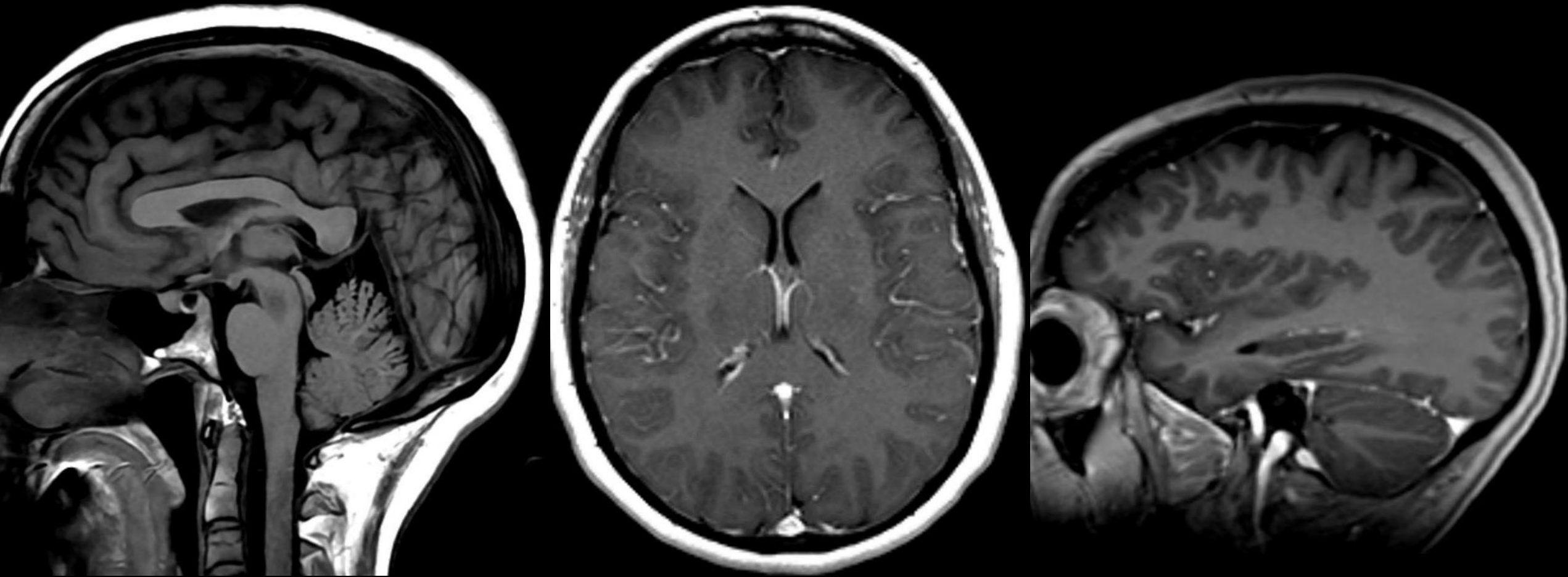


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Case #1 38F

- November 2023: back pain and metallic taste in mouth
- January 2024: Woke up with holocephalic headache, pressure behind eyes and in suboccipital region, worse with cough and bending over. Associated nausea, ear popping, tinnitus, neck stiffness
 - PCP suspects sinus infection → antibiotics → no improvement

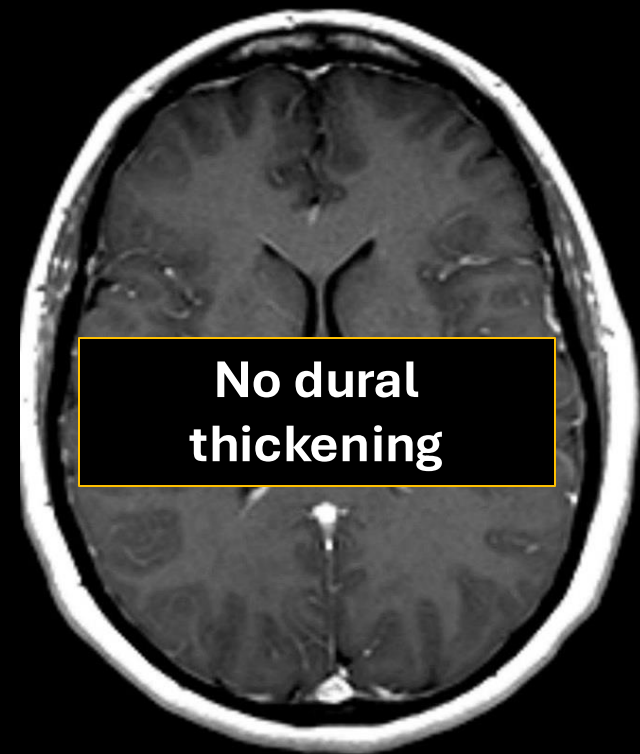
Referred to neurology, orders brain MRI



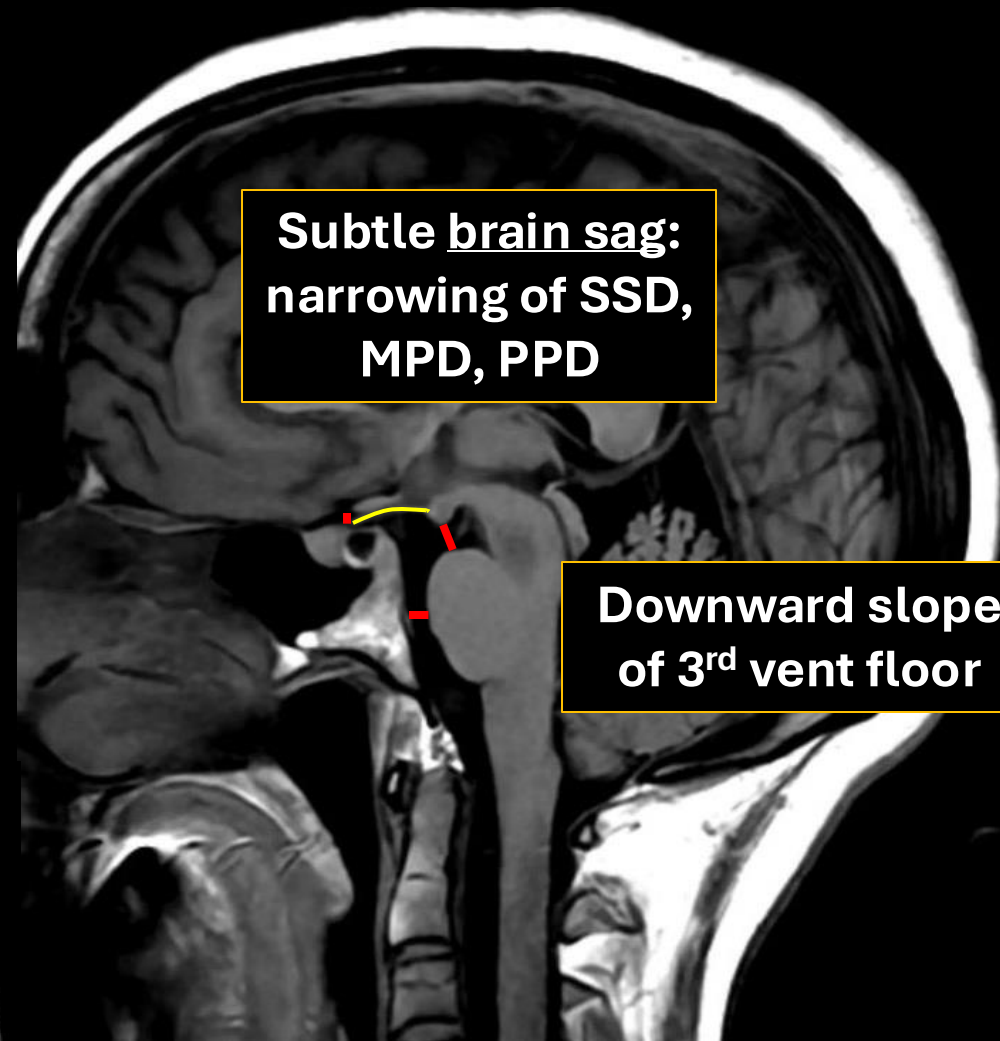
"Normal MRI of the brain"

Case #1 38F

- Neurologist suspects IIH: performs LP → OP 13 cm H₂O, starts Topamax and Rizatriptan for migraines
- Topamax causes depression and lethargy, doesn't help with symptoms, Rizatriptan works once partially for head pain, switched to Ajoovy, does nothing for symptoms
 - July 2024: Referred to CU CSF

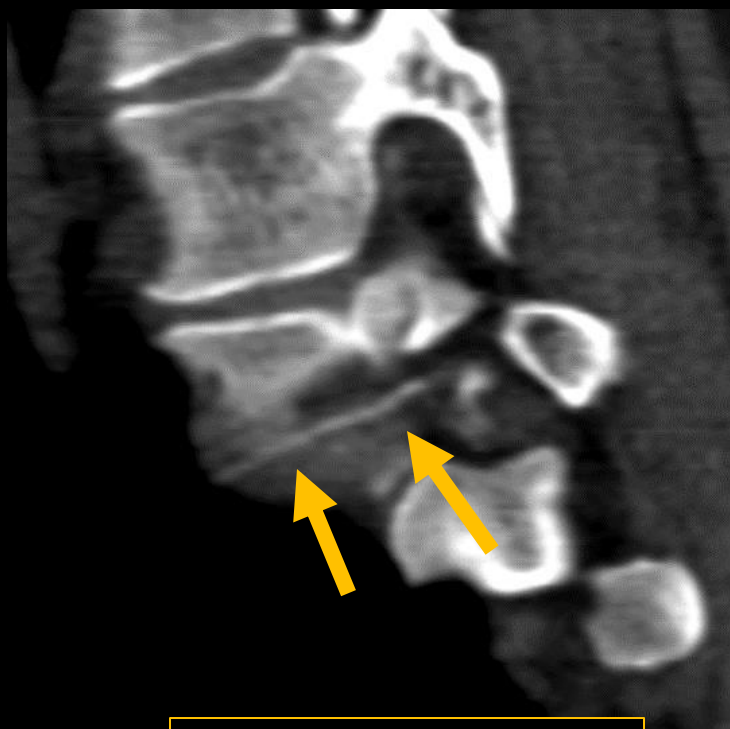


No dural thickening

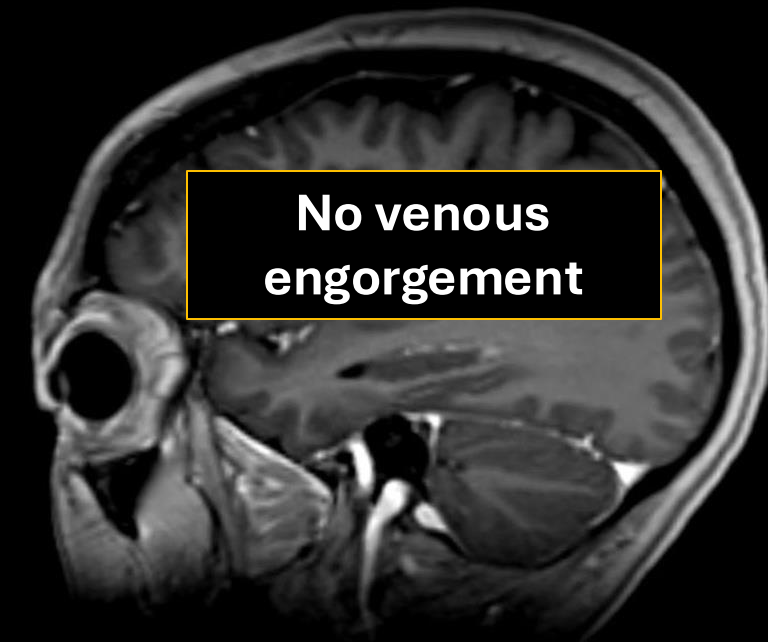


Subtle brain sag:
narrowing of SSD,
MPD, PPD

Downward slope
of 3rd vent floor



Right T8 CSF
Venous Fistula



No venous
engorgement

Bern = 4

Pachymeningeal Enhancement	Venous Engorgement	Suprasellar Effacement (≤ 4 mm)	Subdural Collection	Prepontine Effacement (≤ 5 mm)	Mamilopontine Effacement (≤ 6.5 mm)
2 points	2 points	2 points	1 point	1 point	1 point
2 points or less: LOW PROBABILITY		3-4 points: MODERATE PROBABILITY		5 or more points: HIGH PROBABILITY	

What is a “Normal MRI of the Brain in SIH”?

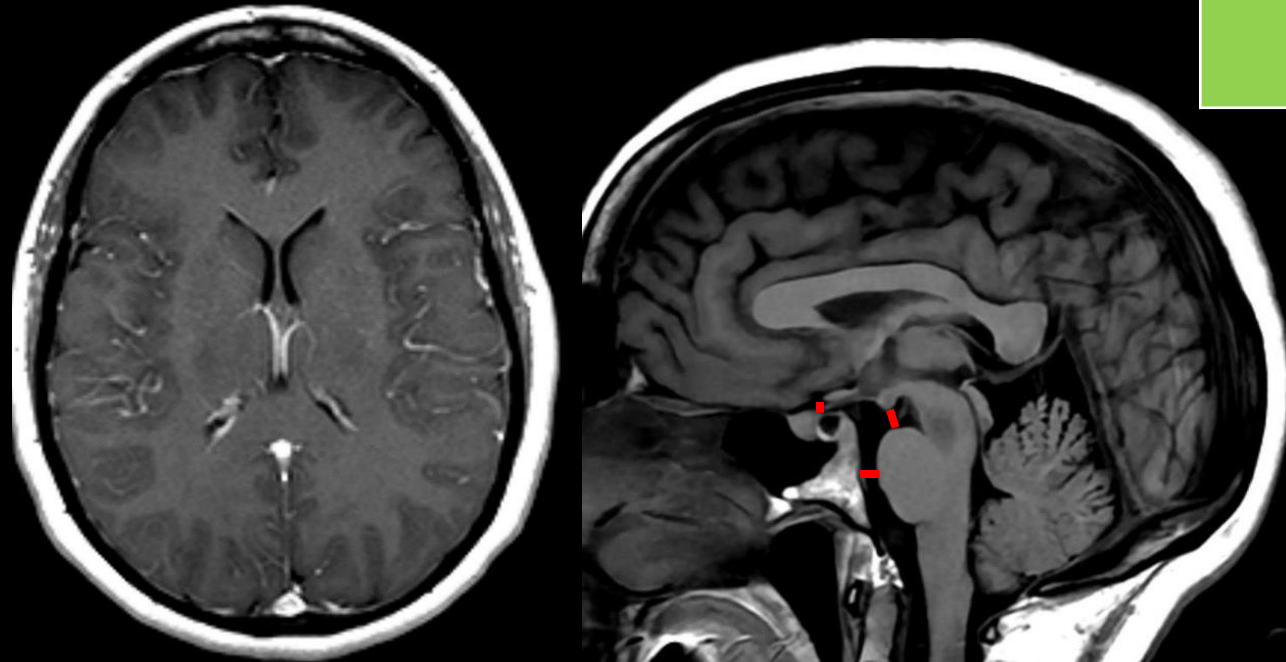
Bern < 2?

Bern = 0?

A normal MRI of the brain is one which is reported as normal.

What % of radiologists would call this suspicious for SIH?

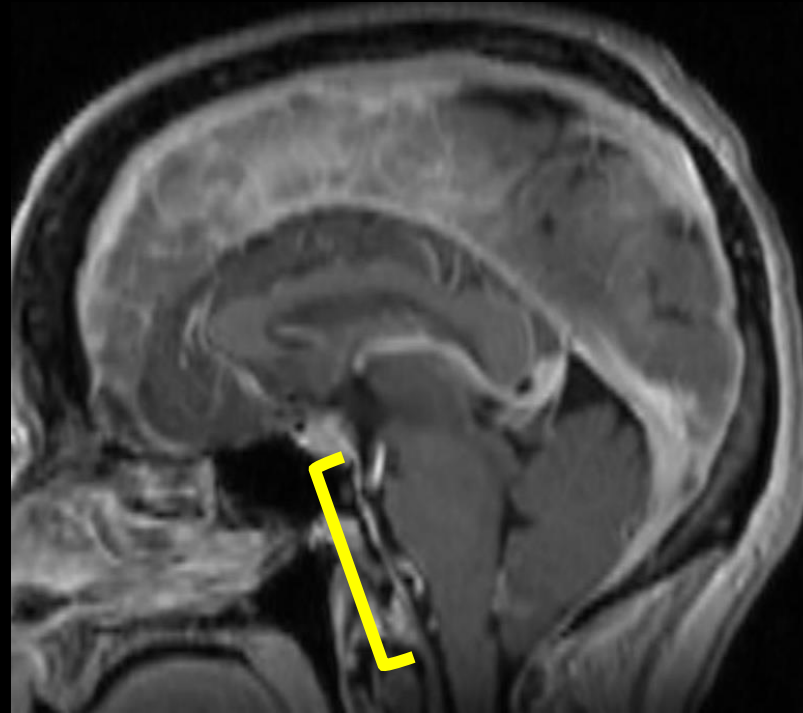
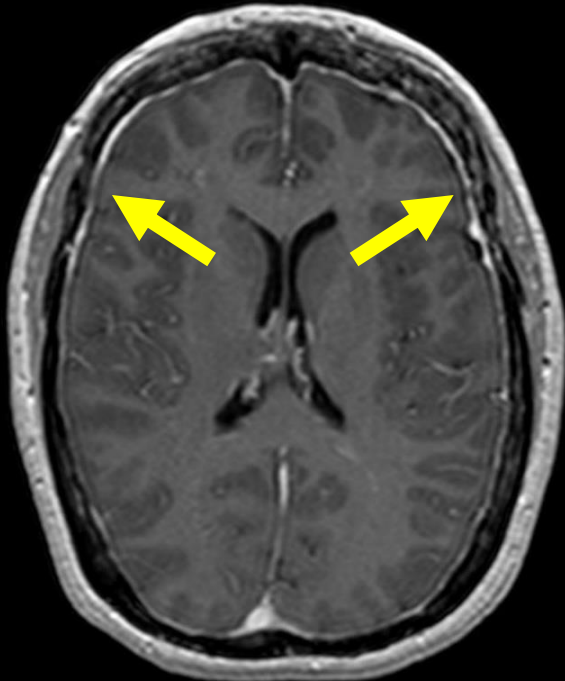
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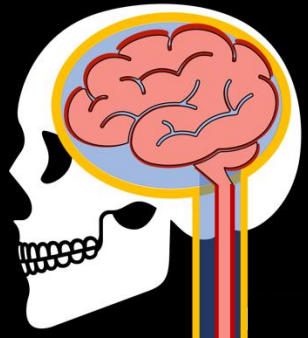


~~“Normal MRI of the Brain”~~
~~“No evidence of intracranial hypotension”~~
“Bern = 4”

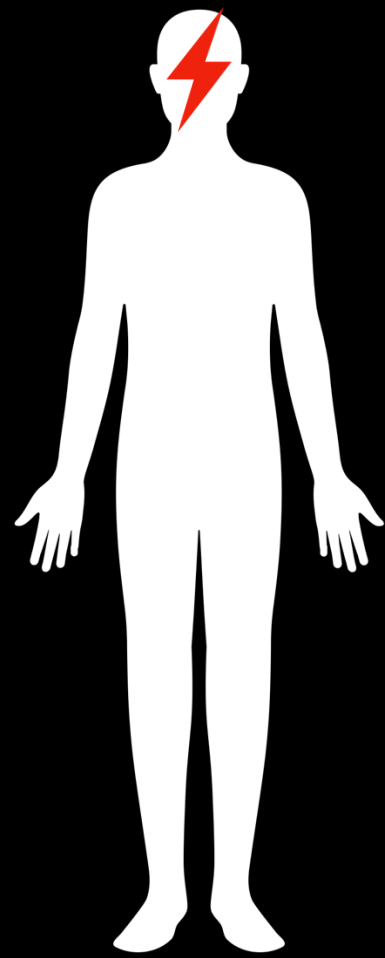
Case #2 61F

- Accompanied by brother. Patient denies any symptoms, brother endorses rapid personality change and memory loss.
- Further questioning: mild left ear hearing loss and tinnitus.
 - No headache, neck pain, nausea, dizziness.

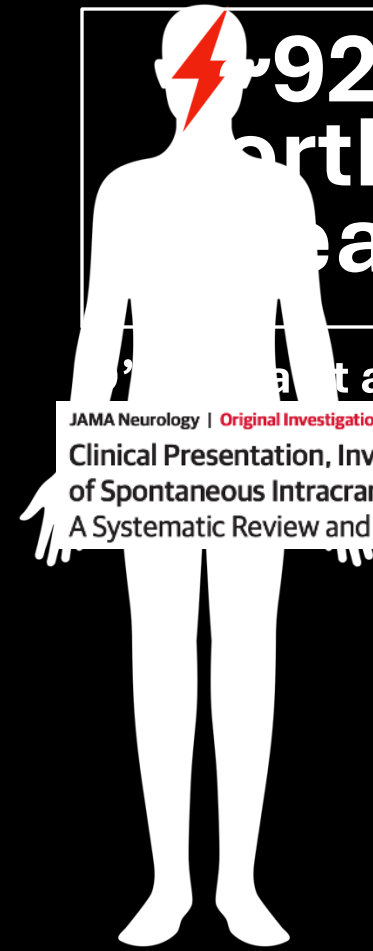




worse when upright



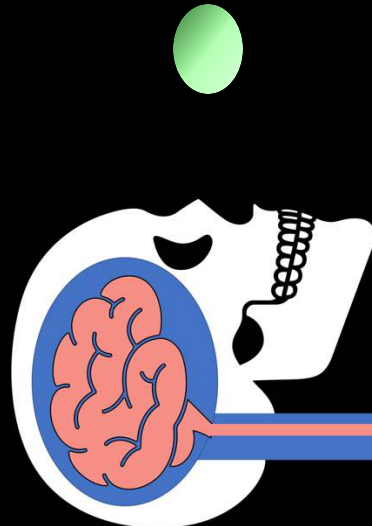
92 % with orthostatic headache'

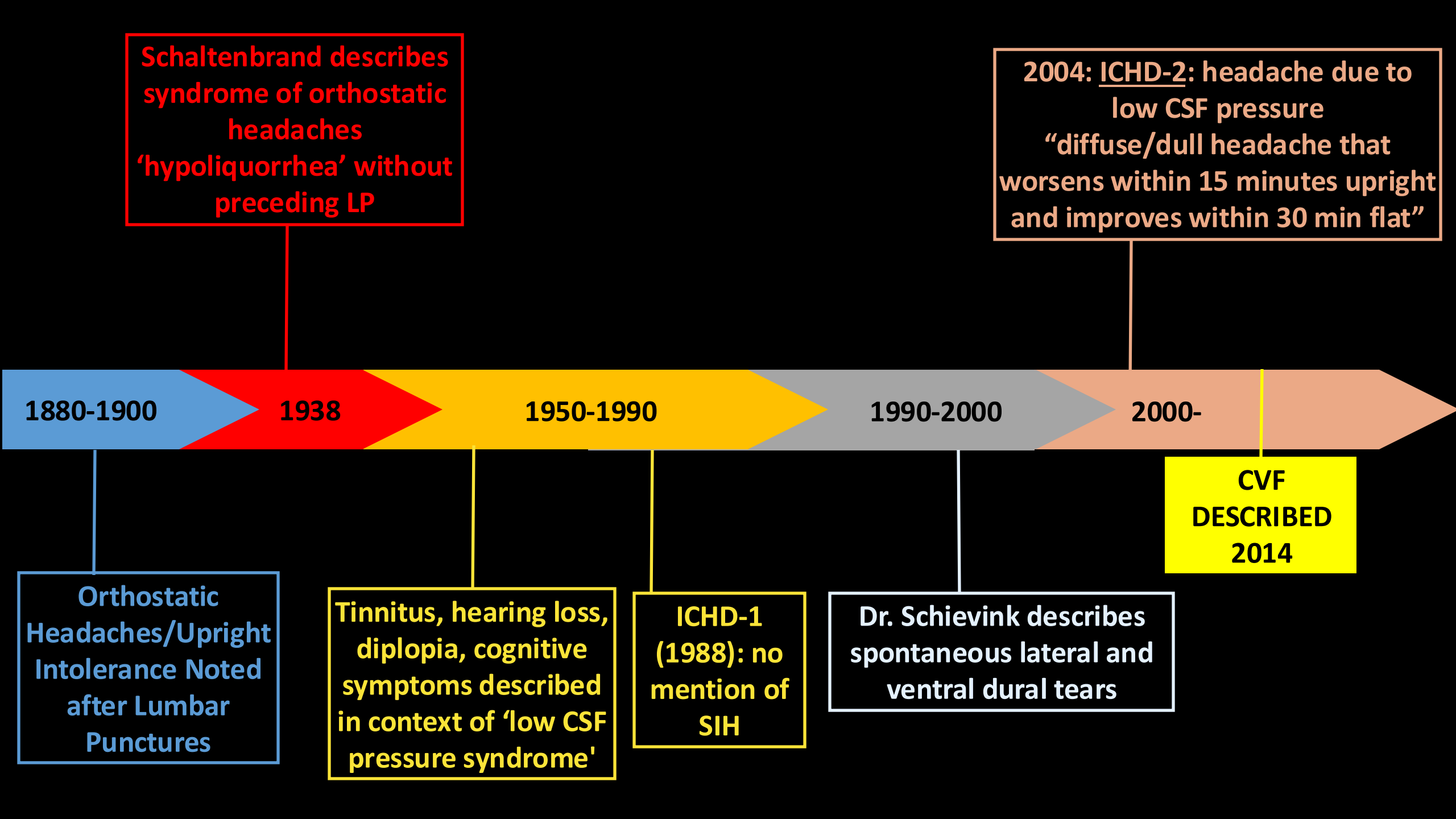


et al. JAMA Neurology 2021

JAMA Neurology | Original Investigation
Clinical Presentation, Investigation Findings, and Treatment Outcomes of Spontaneous Intracranial Hypotension Syndrome
A Systematic Review and Meta-analysis

better when flat





Schaltenbrand describes syndrome of orthostatic headaches 'hypoliquorrhea' without preceding LP

2004: ICHD-2: headache due to low CSF pressure "diffuse/dull headache that worsens within 15 minutes upright and improves within 30 min flat"

1880-1900

1938

1950-1990

1990-2000

2000-

Orthostatic Headaches/Upright Intolerance Noted after Lumbar Punctures

Tinnitus, hearing loss, diplopia, cognitive symptoms described in context of 'low CSF pressure syndrome'

ICHD-1 (1988): no mention of SIH

Dr. Schievink describes spontaneous lateral and ventral dural tears

CVF DESCRIBED 2014

2018:



IHS Classification ICHD-3

7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure

Description:

Orthostatic headache caused by low cerebrospinal fluid (CSF) pressure (either spontaneous or secondary), or CSF leakage, usually accompanied by neck pain, tinnitus, changes in hearing, photophobia and/or nausea. It remits after normalization of CSF pressure or successful sealing of the CSF leak.

Diagnostic criteria:

- A. Any headache¹ fulfilling criterion C
- B. Either or both of the following:
 - 1. low cerebrospinal fluid (CSF) pressure (<60 mm CSF)
 - 2. evidence of CSF leakage on imaging²

**66-96% proven leaks
have normal OP
(particularly CVF)**

Kranz et al. Headache 2018
Callen et al. AJNR 2023

- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or led to its discovery³
- D. Not better accounted for by another ICHD-3 diagnosis.



Comments:

Spontaneous cerebrospinal fluid (CSF) leak has been associated with heritable connective tissue disorders. Patients with CSF leaks should be screened for connective tissue and vascular abnormalities.

While there is a clear postural component in most cases of 7.2.3 Headache attributed to spontaneous intracranial hypotension, it may not be as dramatic or immediate as in 7.2.1 Post-dural puncture headache. Thus, 7.2.3 Headache attributed to spontaneous intracranial hypotension may occur immediately or within seconds of assuming an upright position and resolve quickly (within 1 minute) after lying horizontally, resembling 7.2.1 Post-dural puncture headache, or it may show delayed response to postural change, worsening after minutes or hours of being upright and improving, but not necessarily resolving, after minutes or hours of being horizontal. The orthostatic nature of the headache at its onset should be sought when eliciting a history, as this feature may become much less obvious over time.

In patients with typical orthostatic headache and no apparent cause, and after exclusion of postural orthostatic tachycardia syndrome (POTS), it is reasonable in clinical practice to provide autologous lumbar epidural blood patch (EBP). While EBPs are frequently effective in sealing CSF leaks, the response to a single EBP may not be permanent, and complete relief of symptoms may not be achieved until two or more EBPs have been performed. However, some degree of sustained improvement, beyond a few days, is generally expected. In some cases, sustained improvement cannot be achieved with targeted (to the site of the leak) and/or non-targeted lumbar EBPs, and surgical intervention may be required.

It is not clear that all patients with 7.2.3 *Headache attributed to spontaneous intracranial hypotension* have an active CSF leak, despite a compelling history or brain imaging signs compatible with CSF leakage. The underlying disorder may be low CSF volume. A history of a trivial increase in intracranial pressure (eg, on vigorous coughing) is sometimes elicited.

Postural headache has been reported after coitus: such headache should be coded as 7.2.3 *Headache attributed to spontaneous intracranial hypotension* because it is most probably due to CSF leakage.

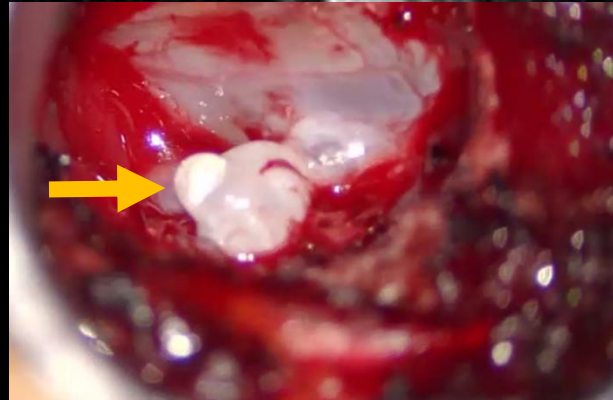
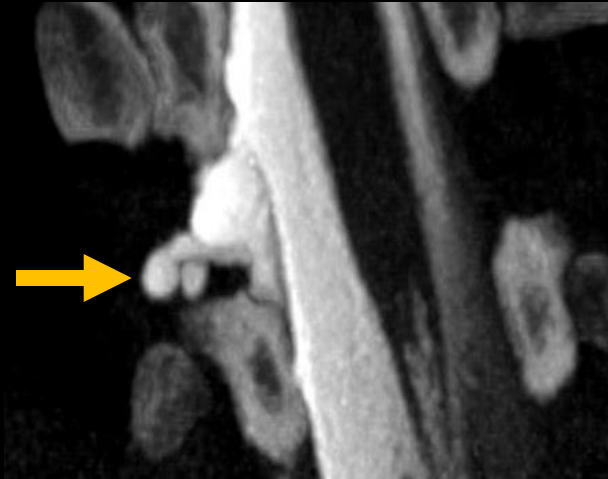
Spontaneous Leak Types and Frequency

(Our Experience)

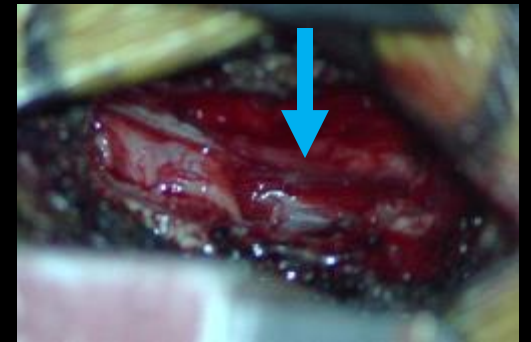
Ventral Defect
28%



Lateral Defect
(17%)



CSF-Venous
Fistula (55%)



Patterns of clinical and imaging presentations in patients with spontaneous intracranial hypotension due to spinal cerebrospinal fluid venous fistula: A single-center retrospective cross-sectional study

48 consecutive patients with CVF localized at CU

- All reported individual clinical symptoms analyzed
- Brain MRI analyzed for each Bern Score component

Orthostatic Headache?

48 Patients with CVF

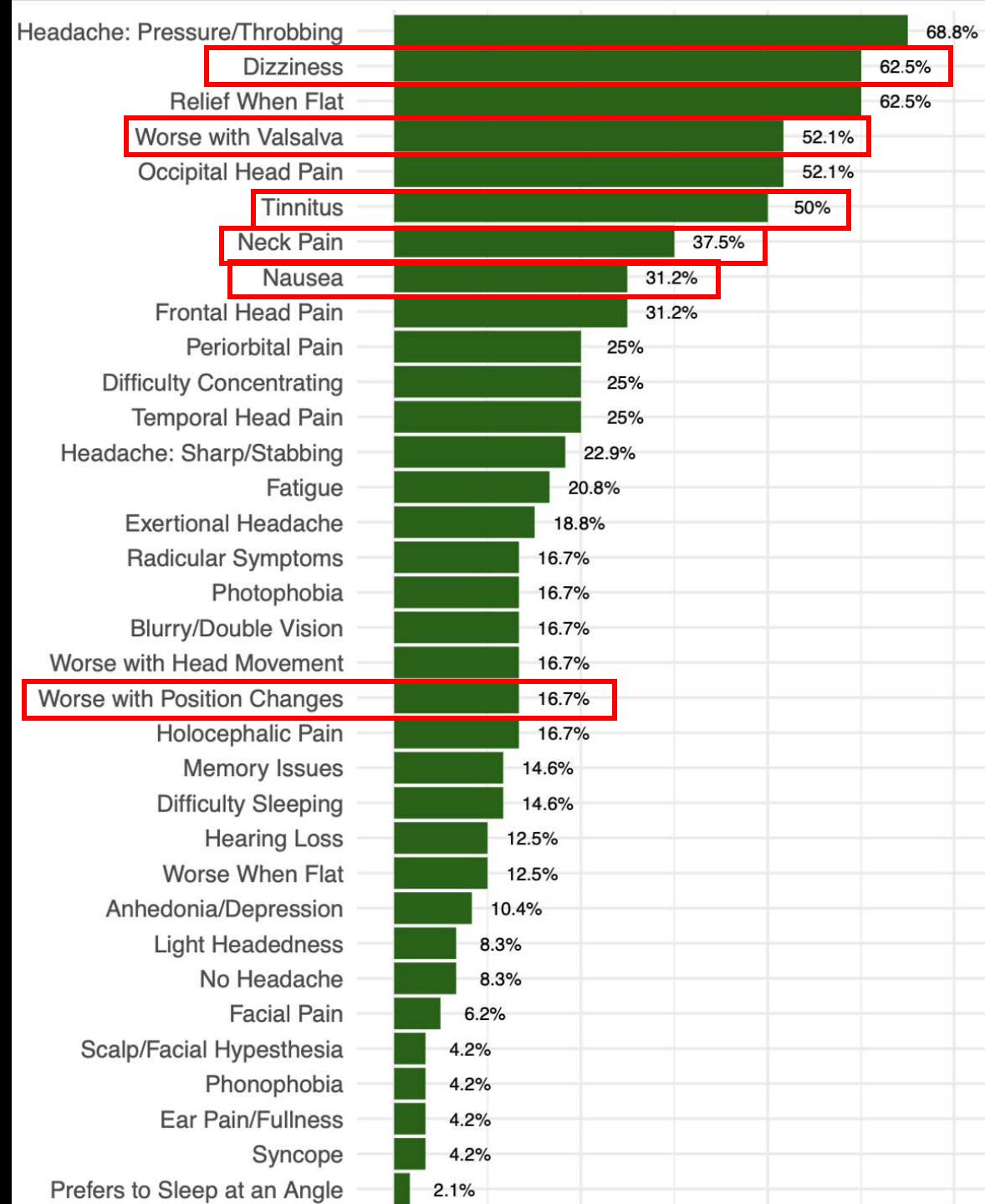
37% - no relief when flat

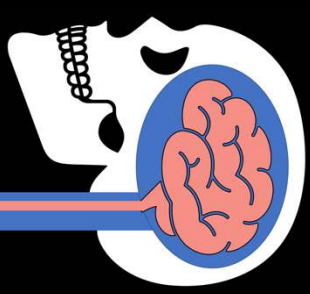
12.5% - **WORSE** when flat

8% - no headache at all

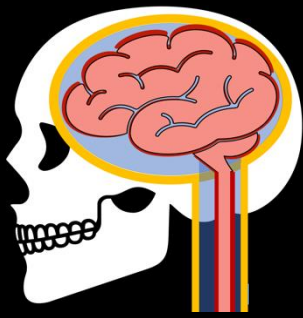
12.5% - no relief when flat

**AND ≥ 1 migrainous symptom
(phono/photophobia, nausea)**





What is an 'orthostatic headache'???

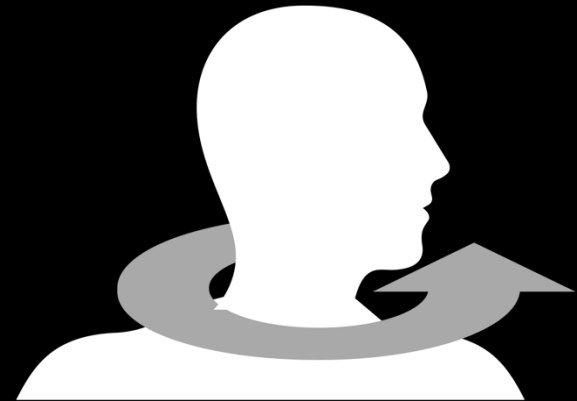
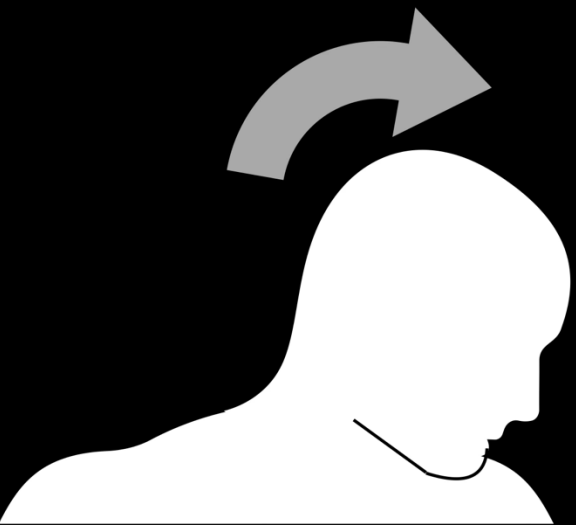


“Do you feel better when you lay down?”

Targeted Review of Symptoms

17%- worse with position *transition*, but not after being upright

17%- worse with *head movement*

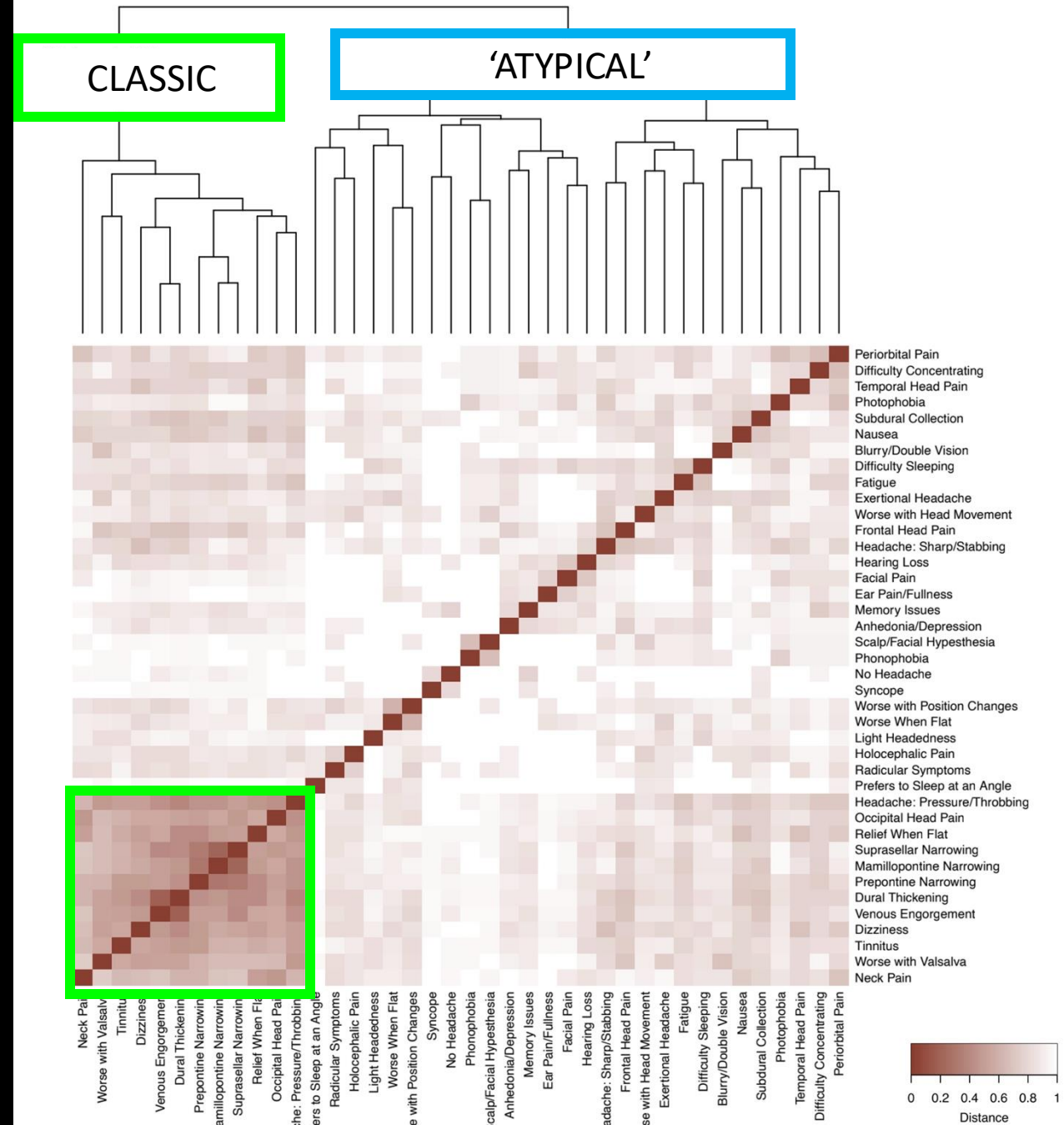


'Classic' Symptom Cluster (48%)

- **Quality: Pressure/Throbbing**
- **Location: Occipital**
- **Relief when flat**
- **Neck pain**

'Atypical' Symptom Cluster (46%)

- **Quality: Sharp/Stabbing**
- **Location: Frontal**
- **Less relief when flat**
- **Less neck pain**



No Headache – Acephalgic Cluster

50% - memory issues

12.5%

syncope

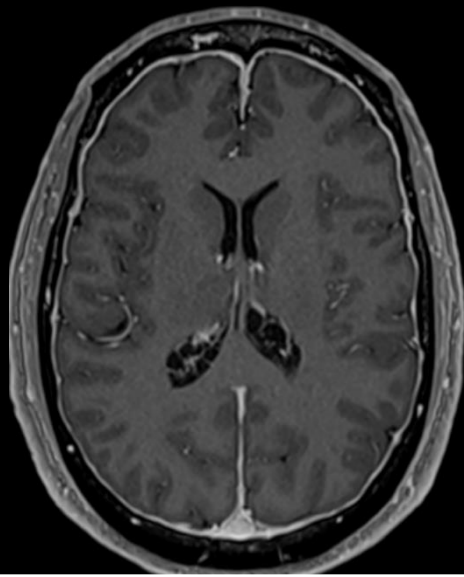
difficulty concentrating

hearing loss

radicular pain/paresthesia

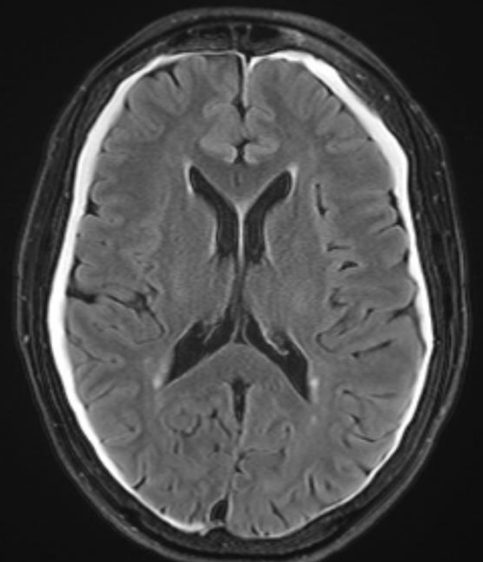
tinnitus

neck pain without headache

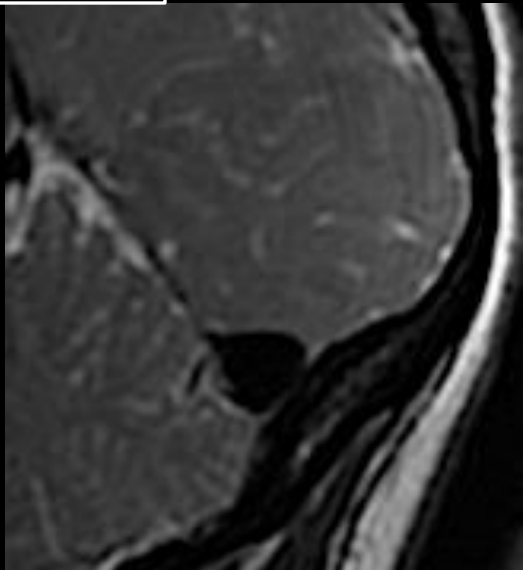


DURAL ENHANCEMENT

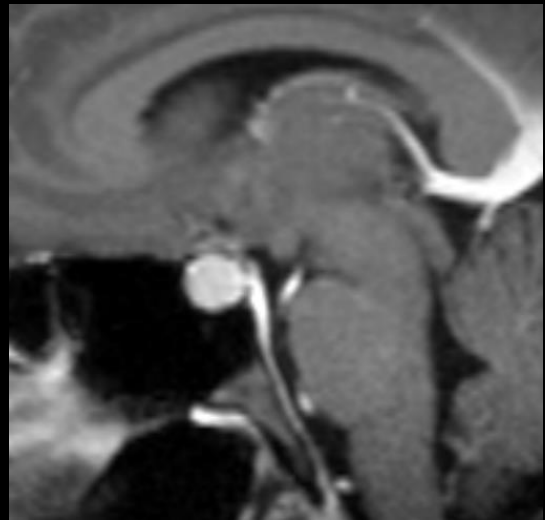
BRAIN SAG



SUBDURAL COLLECTIONS

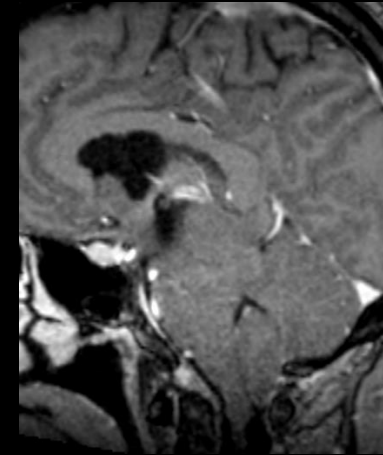
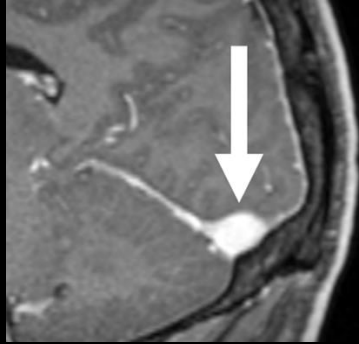
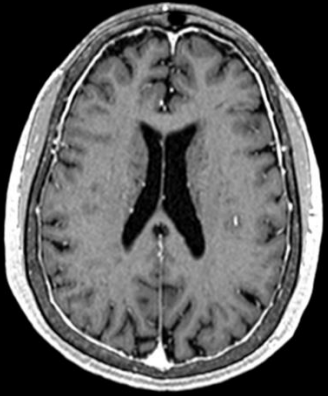


VENOUS ENGORGEMENT



PITUITARY ENGORGEMENT

MRI Findings Cluster Together!



**Dural
Enhancement**

P = 0.007,
OR 18.7

**Venous
Engorgement**

No
significant
associations
across
groups

Prepontine Narrowing

p < 0.001, OR 10.1

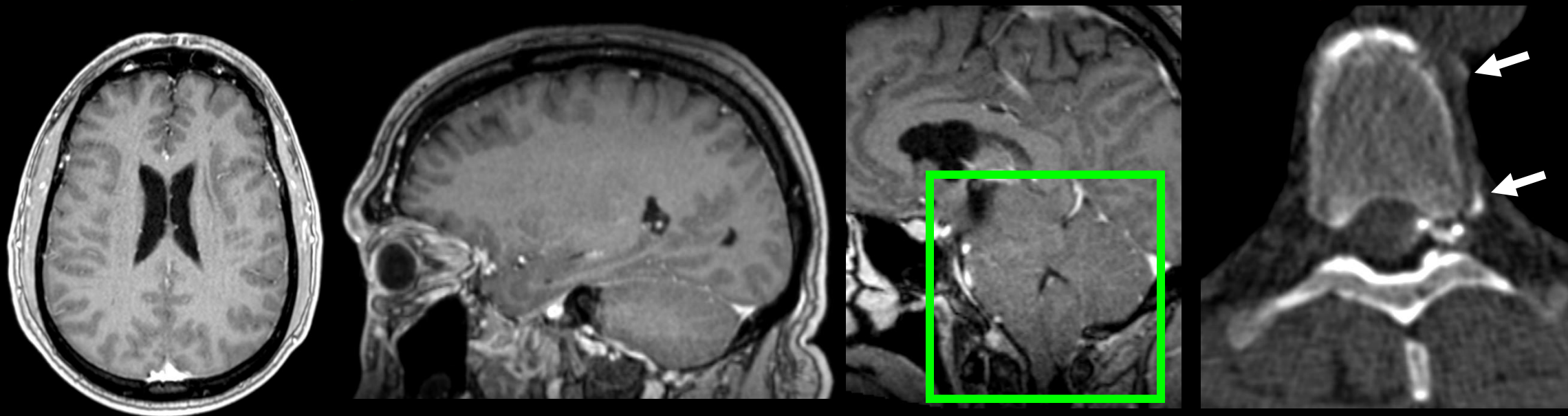
p = 0.004, OR 7.5

**Mamillopontine
Narrowing**

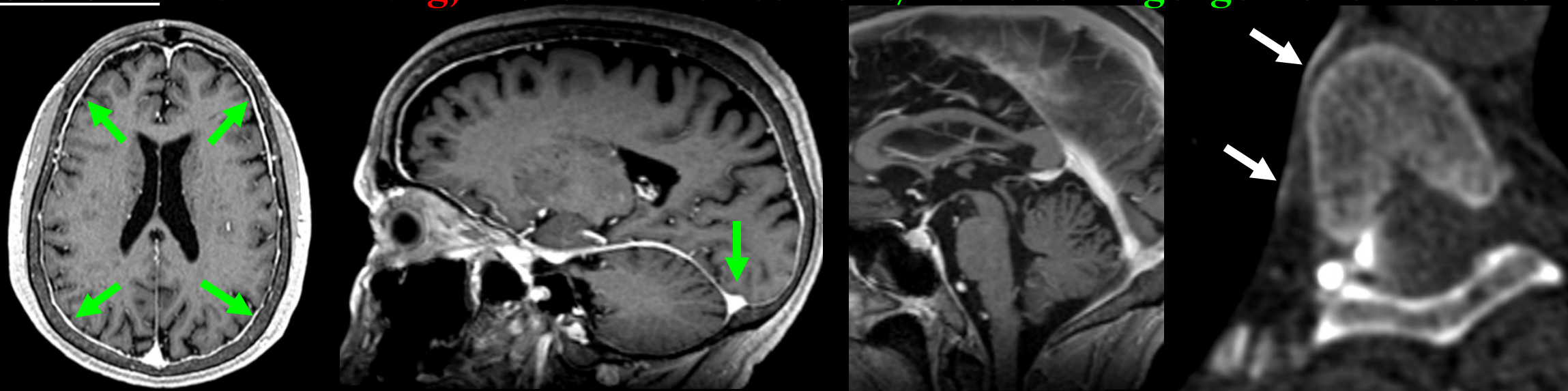
p < .001
OR 28.9

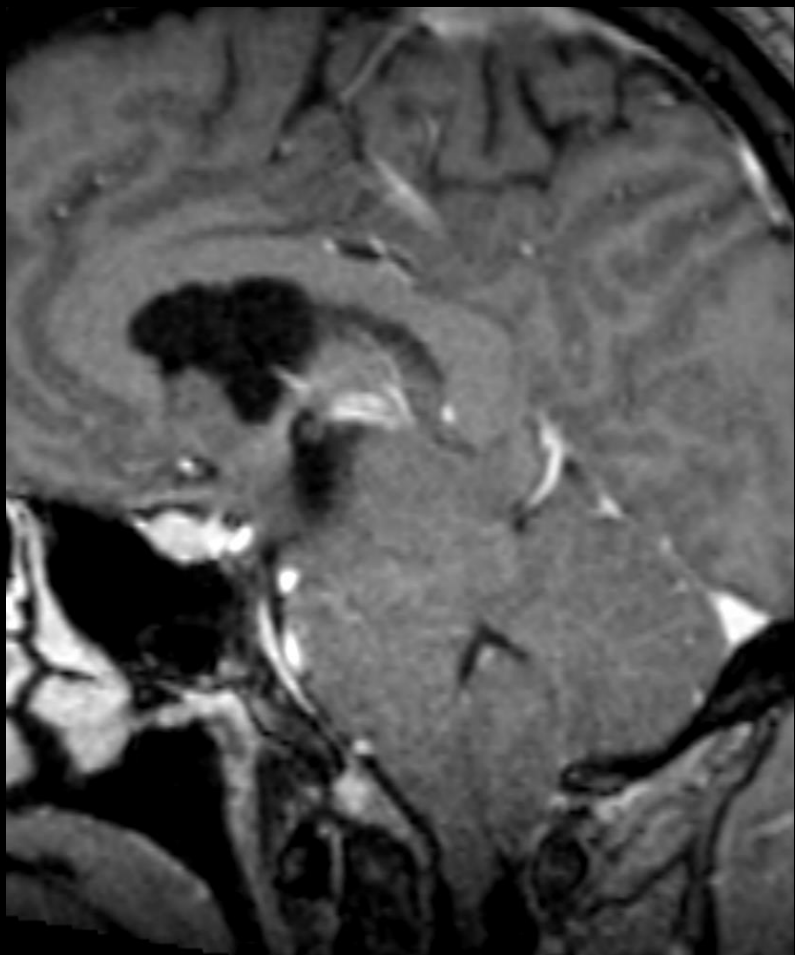
**Suprasellar
Narrowing**

Patient A: Brain Sag Present, No Dural Enhancement/Venous Engorgement



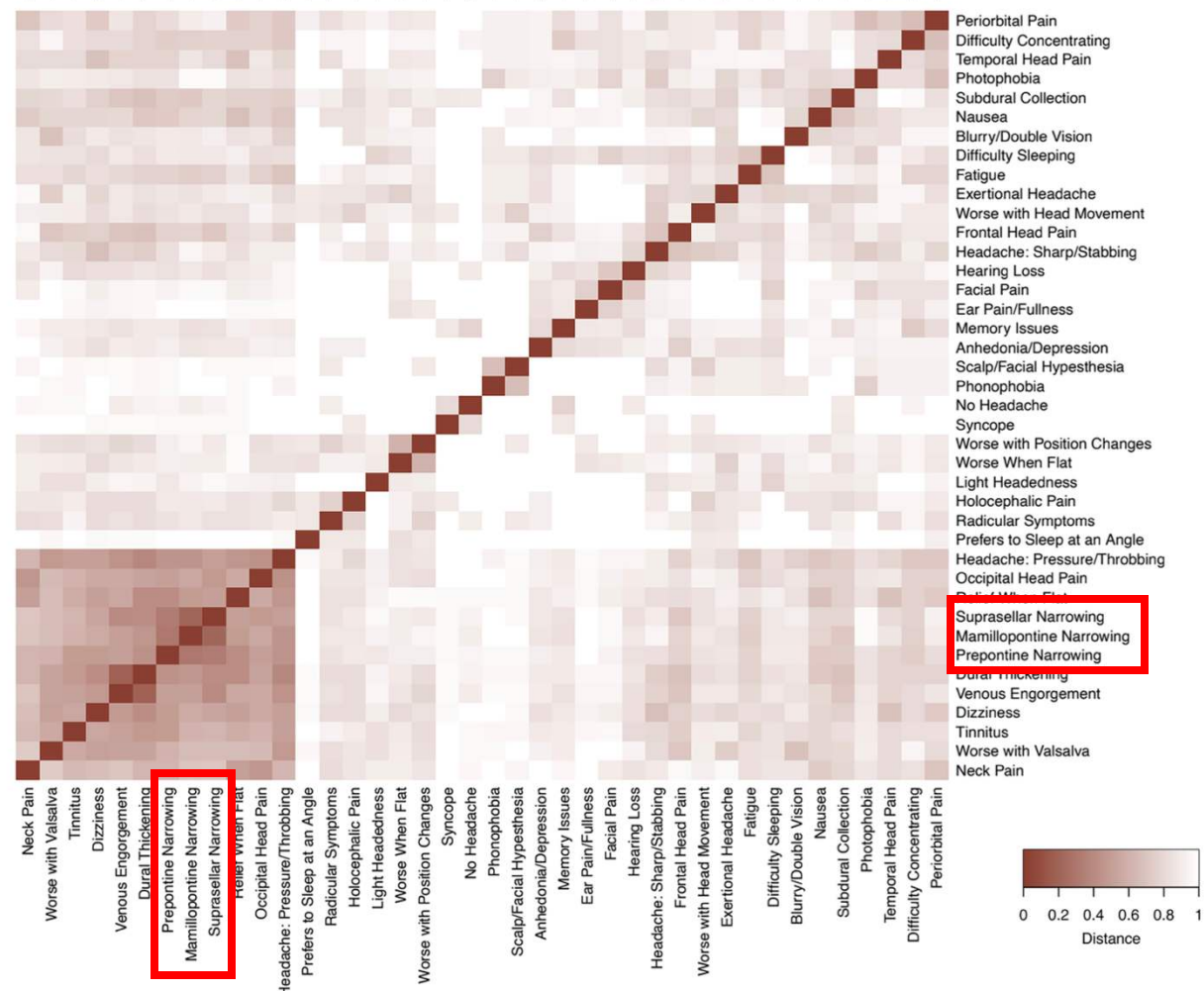
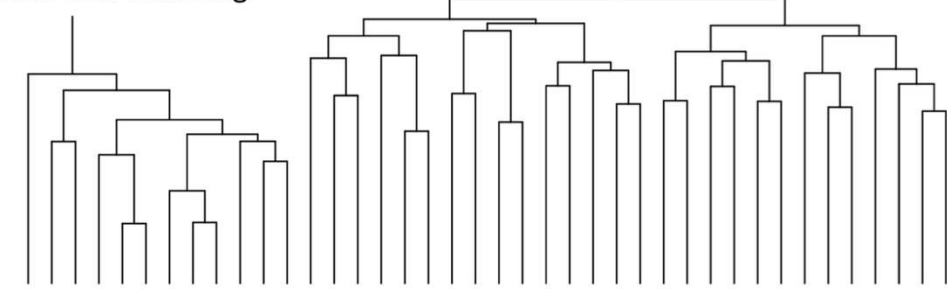
Patient B: No Brain Sag, Dural Enhancement/Venous Engorgement Present





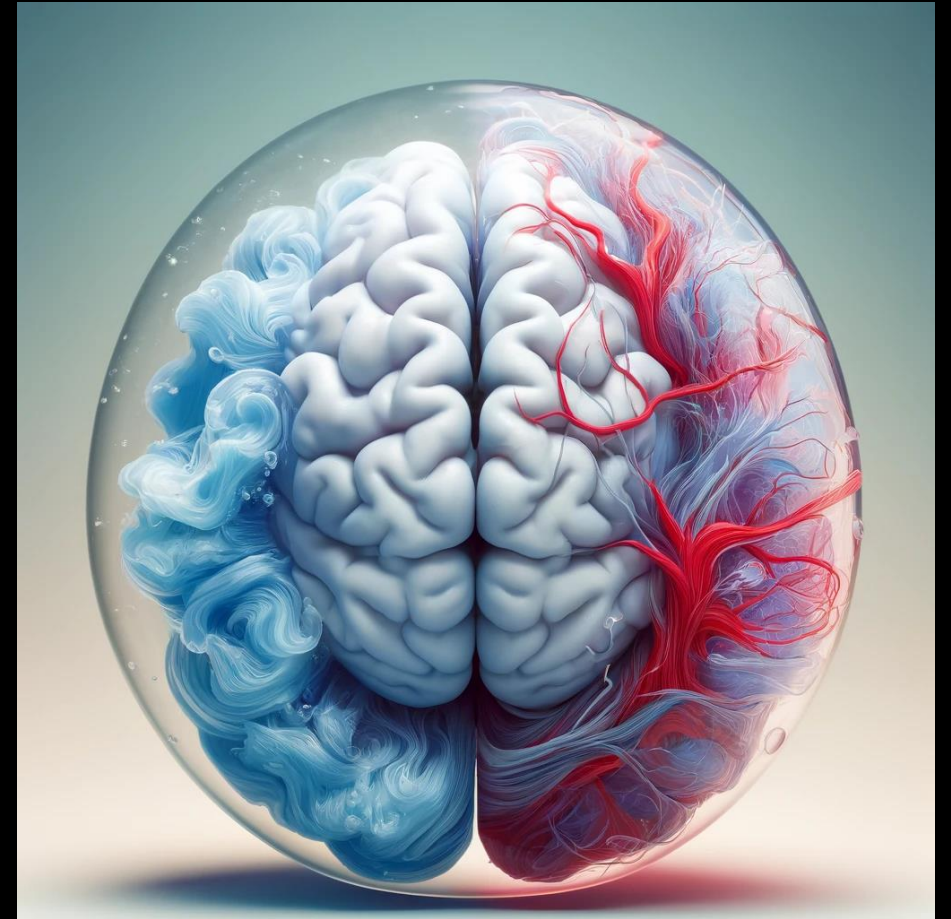
“Classic” Symptom Cluster with Brain Sag

“Atypical” Symptoms without Brain Sag





***Loss of Buoyancy
Sag → Classic Symptoms?***



***Venous Engorgement
(Monro Kellie)***

Take Home Points

Brain MRI findings of a leak may be incredibly subtle. Relief when flat: classic leak symptom, but Radiology report of "normal" deserves a second look. A patient with a leak can lack dural thickening and have absence of headache, absence of positional component, and/or presence of migrainous symptoms. Dizziness, tinnitus, nausea, cough headache, symptom should not preclude leak workup. Clinical and imaging findings may cluster together. neck pain are common.

What is the significance of this for our patients?

Thank you!



Peter Lennarson MD



Dave Bhaumik, MD



Samantha Petrucci, MD PhD



Nadya Andonov, NP