Pressing Issues in SIH

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Anschutz Medical Campus

DISCLOSURES

Medical Advisory Board:

Spinal CSF Leak Foundation

Spinal CSF Leak Canada

Consultant:

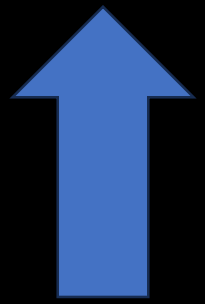
Eli Lilly

Patent:

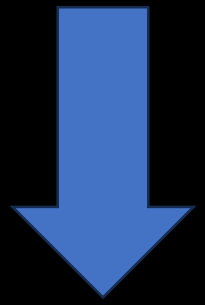
Myelography positioning device

University of Colorado
Anschutz Medical Campus

What could we do better?



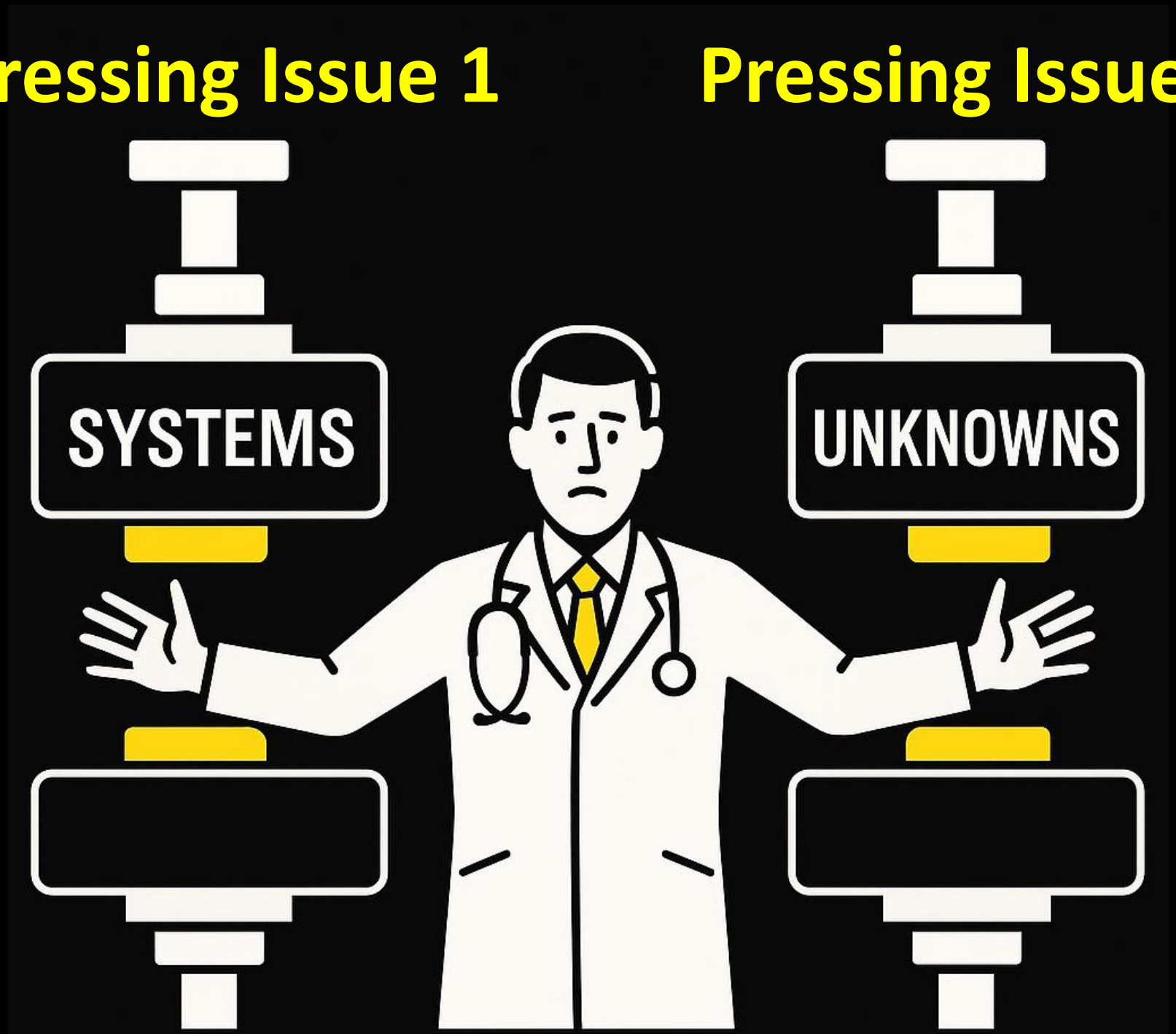
What are *Pressing Issues* in SIH?



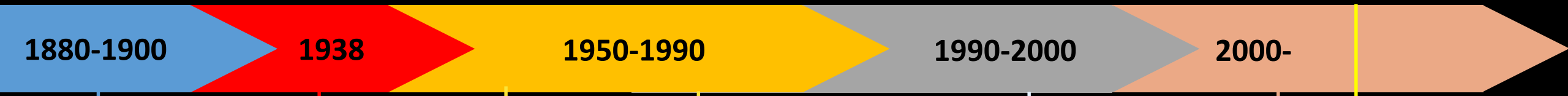
What remains elusive and clinically important?

Pressing Issue 1

Pressing Issue 2



How Far We Have Come



Orthostatic Headaches/Upright Intolerance Noted after Lumbar Punctures

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

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

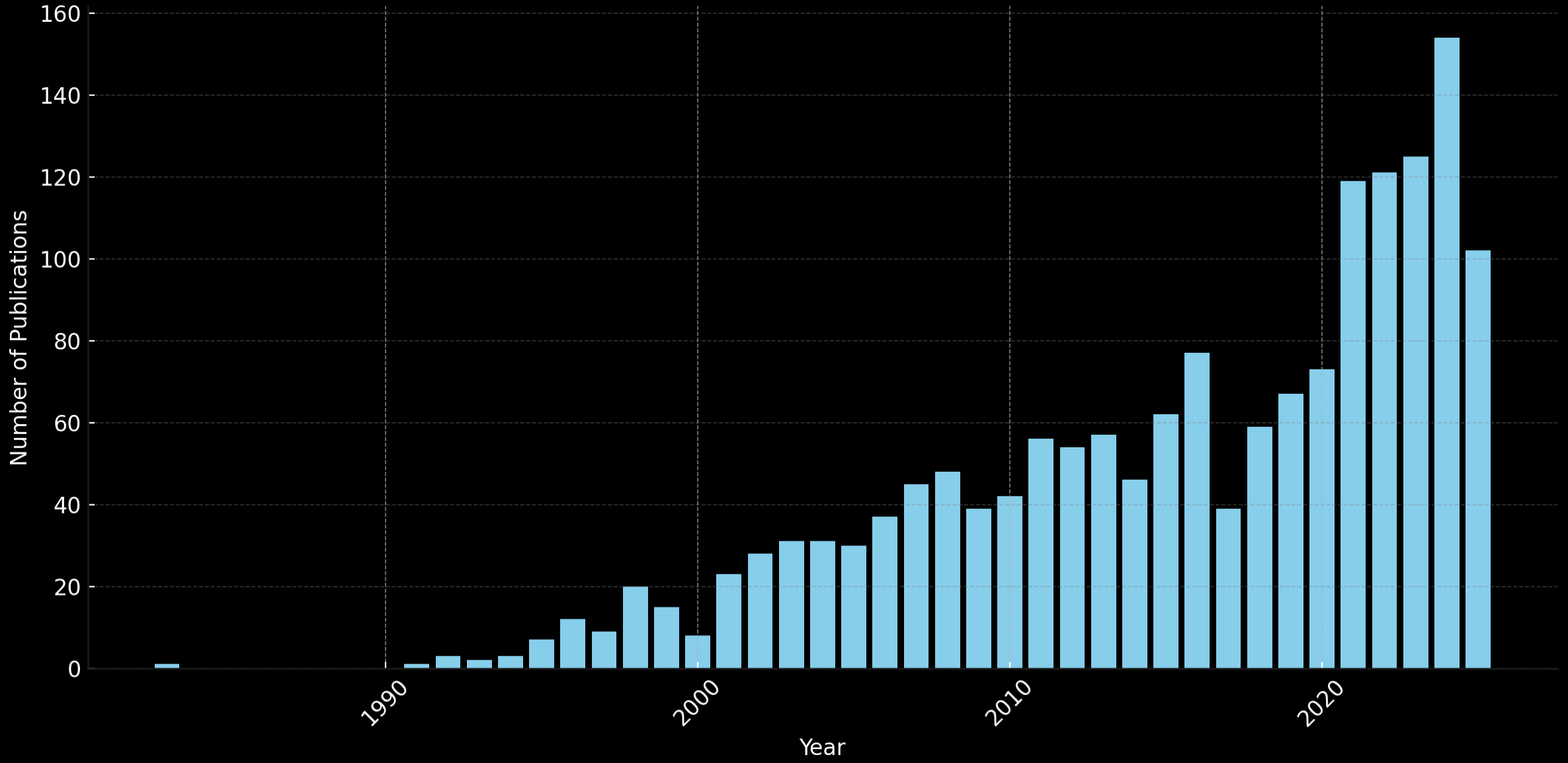
ICHD-1 (1988): no mention of SIH

Dr. Schievink describes spontaneous lateral and ventral dural tears

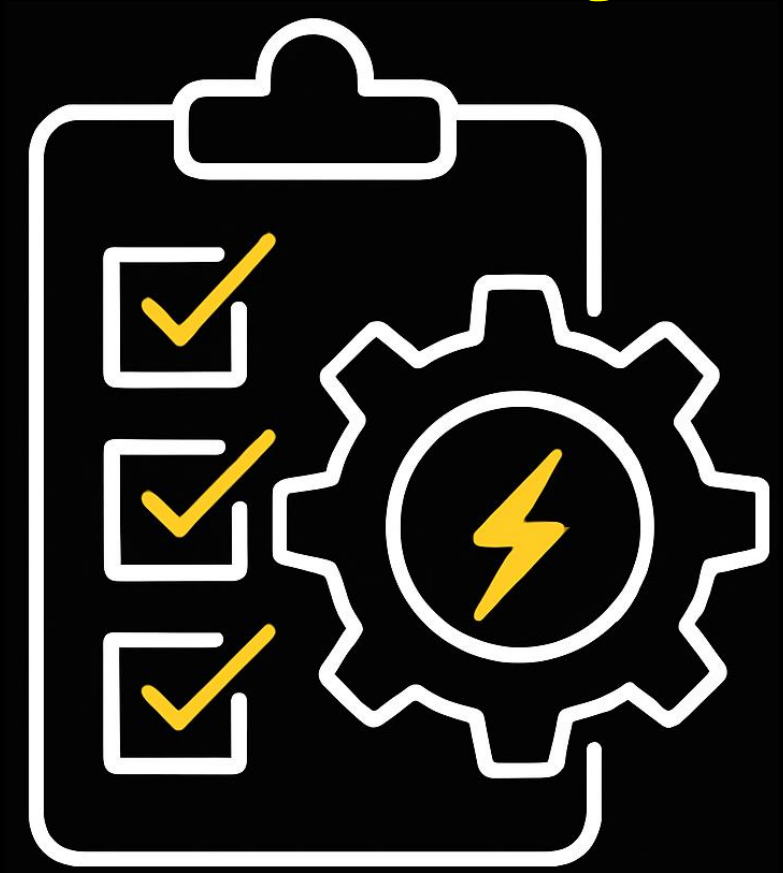
2004: ICHD-2: headache due to low CSF pressure "diffuse/dull headache that worsens within 15 minutes upright and improves within 30 min flat"

CVF DESCRIBED 2014

Pubmed Articles on SIH



Pressing Issues: Systems-Level



- Awareness
- Access
- Education
- Infrastructure/Billing

Underestimation

>90% cases misdiagnosed initially

Mimics migraine, tension type headache, or psychiatric conditions

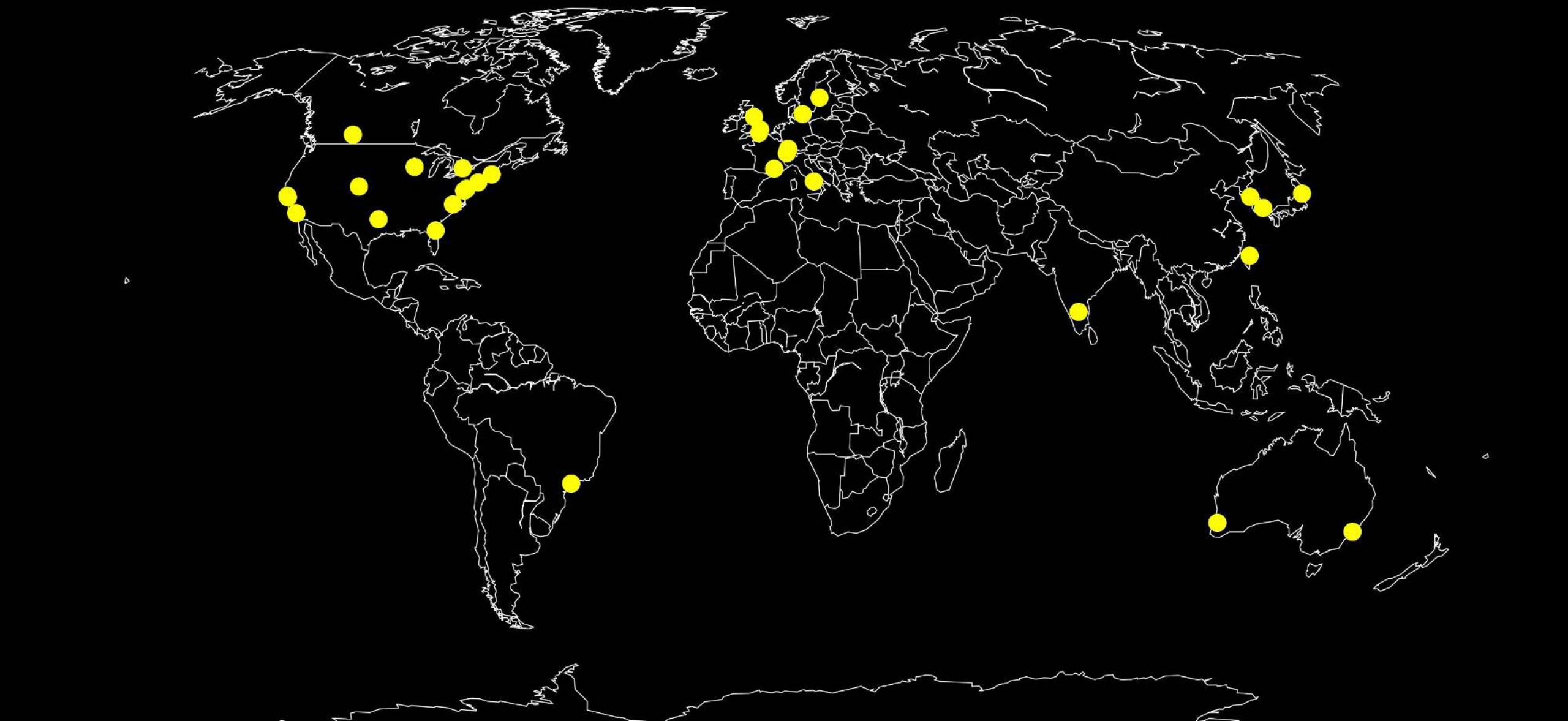
Median delay to diagnosis ~7 months

ICD10 G98.11 (SIH) introduced 2018-19, shifting coding habits takes time/education

Schievink et al 2003
Vukovic-Cvetkovic 2024

Lack of Access

Diagnostic delays, Financial burden, Impact on employment



Access and Disparities

1 month of our CSF leak clinic patients

vs

1 month of ER headache patients who received neuroimaging

Variable	CSF Clinic	ED Headache Pts	Test	*p*-value

PRELIMINARY DATA: Puello Baron, Callen (In Submission)

Are We Underestimating the Impact?

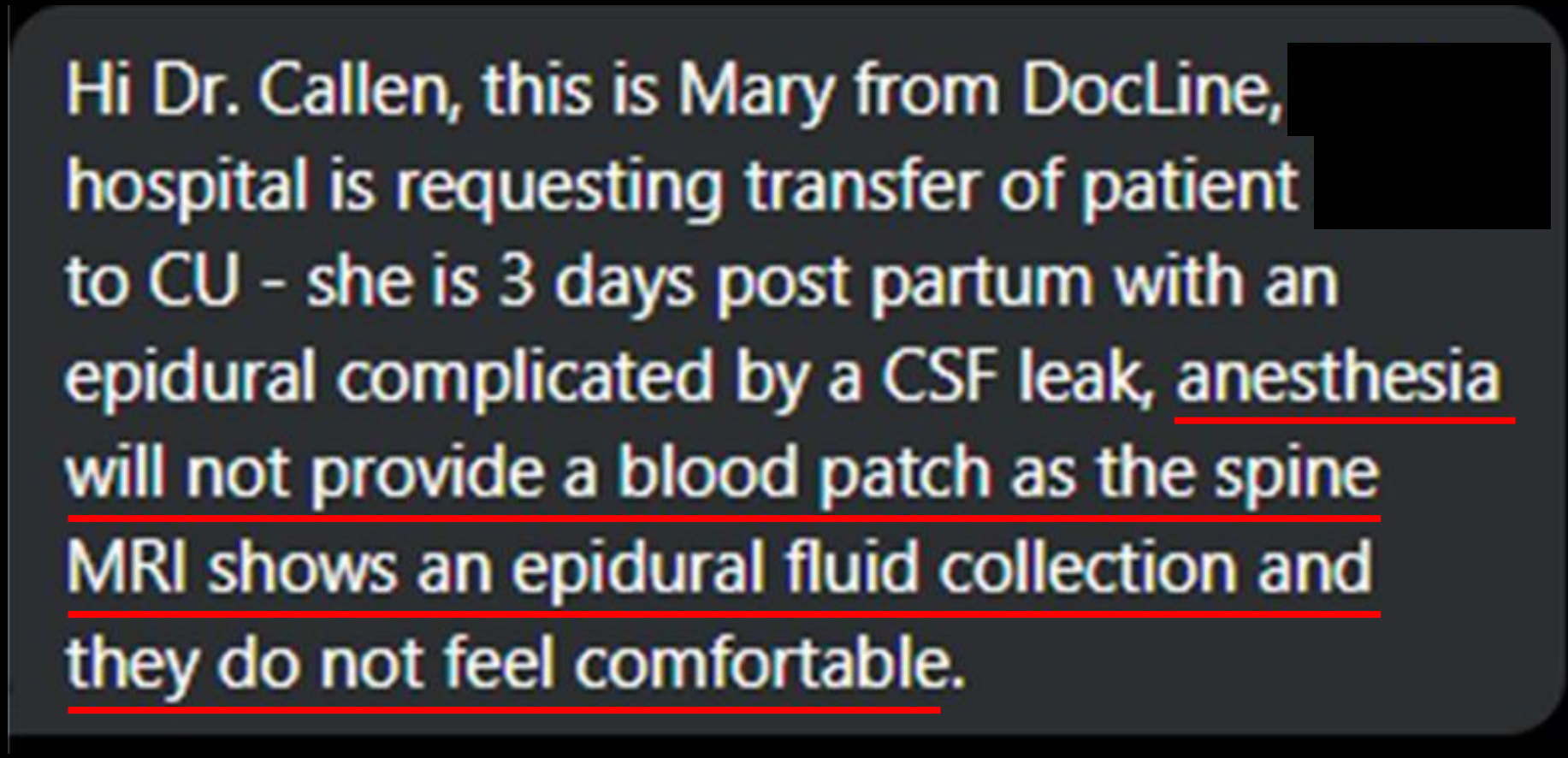
UK Survey: Income loss in 67% of patients; 28% lost jobs (Cheema et al 2022)

Clinic-vs-ED data suggest almost 2x higher private-insurance rate and 3× median travel distance among those who get specialty care

No study yet quantifies missed SIH diagnoses and impact in low-SES or non-English-speaking populations

You Don’t Always Need a “Center”

10/11/2023



Maybe Just Education…

Reimbursement and Sustainability

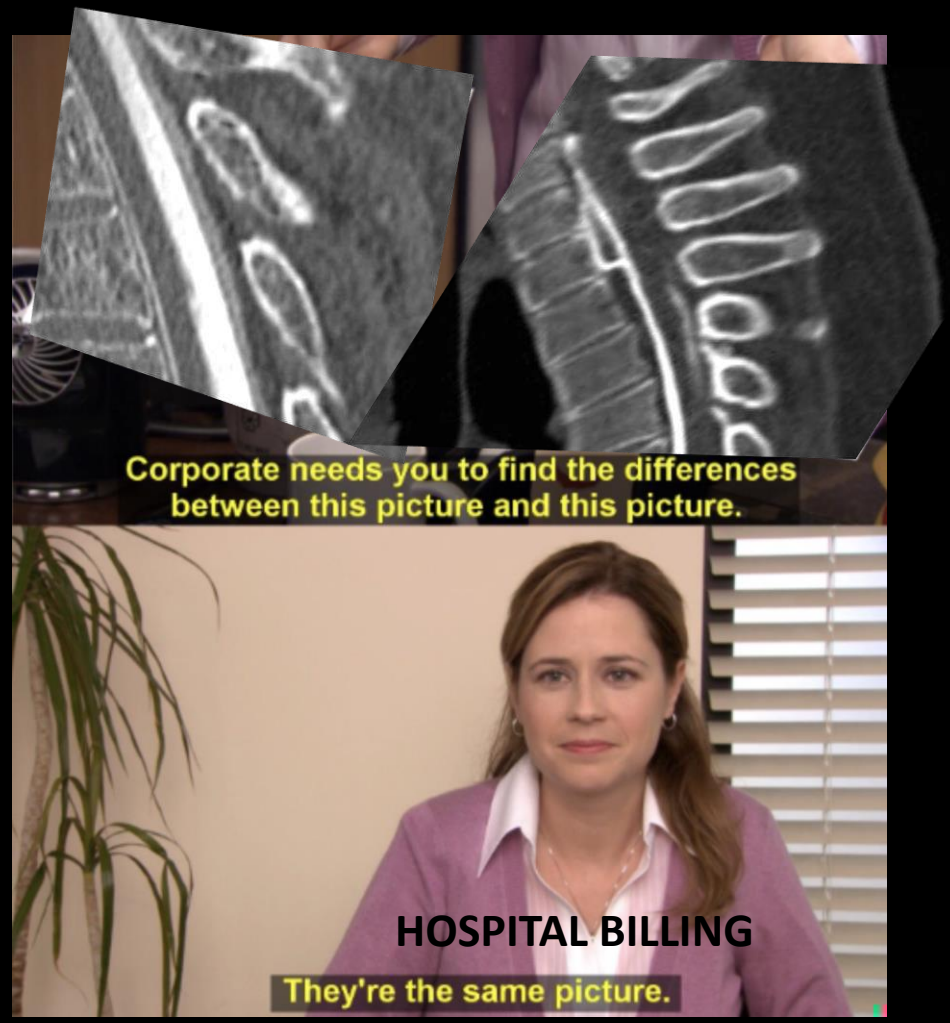
CU estimate: SIH work full time =
~60% RVU of benchmark diagnostic neuroradiologist

Time	Procedure
7:00 AM	CT DYNAMIC MYELOGRAM [IM...
8:00 AM	CT LUMBAR PUNCTURE [IMG22...
9:00 AM	CT INJ EPI BLOOD/CLOT PATCH...
10:00 AM	CT DYNAMIC MYELOGRAM [IM...
11:00 AM	CT FIBRIN GLUE INJECTION LU...
12:00 PM	CT FIBRIN GLUE INJECTION CE...
1:00 PM	CT FIBRIN GLUE INJECTION CE...
2:00 PM	CT INJ EPI BLOOD/CLOT PATCH...
3:00 PM	CT DYNAMIC MYELOGRAM [IM...
4:00 PM	CT INJ EPI BLOOD/CLOT PATCH...

Time	Type
7:00 AM	NEW PATIENT VISIT
8:00 AM	HOME TELEHEALTH
8:30 AM	NEW PATIENT VISIT
9:30 AM	HOME TELEHEALTH
10:00 ...	RETURN PATIENT VISIT
10:30 ...	HOME TELEHEALTH
11:00 ...	NEW PATIENT VISIT
12:00 PM	CT INJ EPI BLOOD/CLOT PATCH
1:00 PM	CT DYNAMIC MYELOGRAM [IM..
2:00 PM	CT INJ EPI BLOOD/CLOT PATCH
4:00 PM	CT INJ CERV/THORA EPIDURAL
5:00 PM	CT INJ EPI BLOOD/CLOT PATCH

Administrative Blind Spots

Lack of CPT/CCSD codes for advanced myelography and targeted patching



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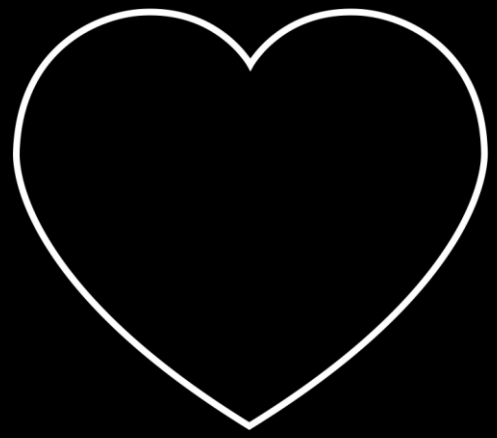
Beyond Systems: Pressing Unknowns

What draws us to this work?

Curiosity:
The Unknown



Altruism:
A Desire to Help

Frustration:
When we can't help, why not?

The Easy Wins

Technical & Clinical Success

61M, YEARS OF ORTHOSTATIC HEADACHE

2016: CTM
"No leak"

2016 Diagnoses:

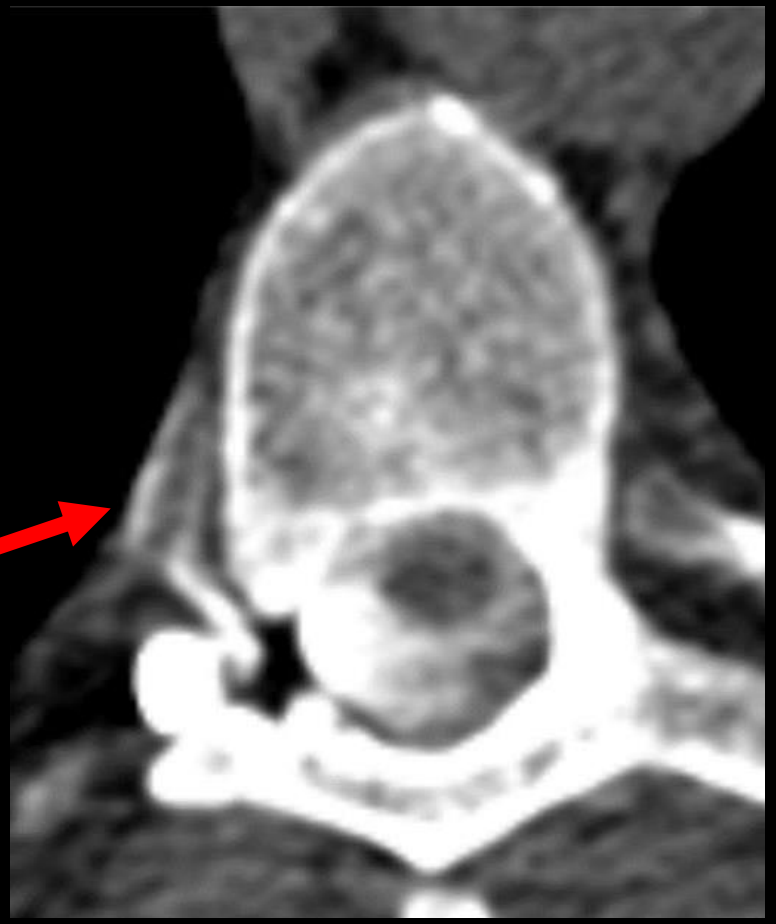
1. Migraine
2. Tension headache
3. Trigeminal Autonomic Cephalalgia
4. TMJ myofascial syndrome
5. Occipital neuralgia
6. Supraorbital neuralgia

61M, YEARS OF ORTHOSTATIC HEADACHE

2016: CTM
"No leak"

2021 Diagnosis:

1. SIH due to CVF



2021: LAT DECUBITUS CTM

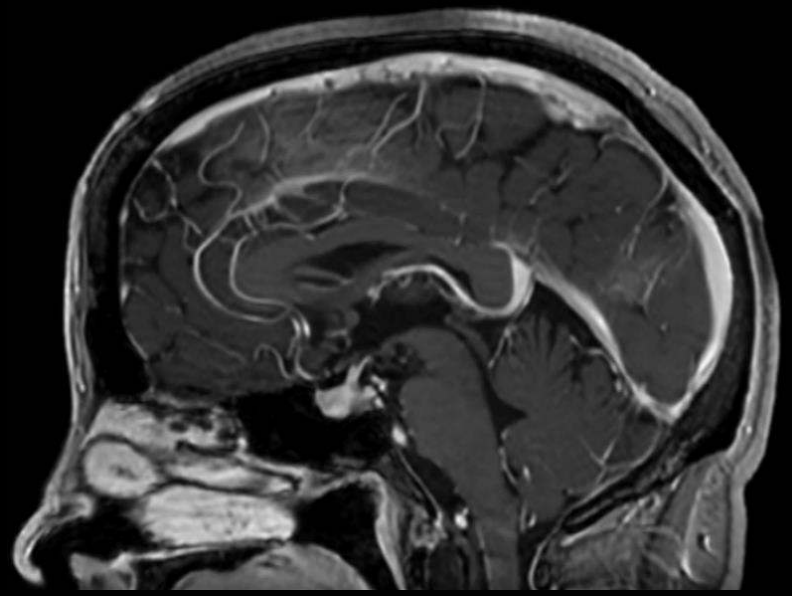
Diagnostic Dead Ends

No leak found, no fix possible

Sounds Like a Leak…

34F sudden onset suboccipital HA

Sounds Like a Leak…



Brain MRI -

Spine MRI -

- Negative bilateral lateral decubitus myelography
- Empiric blood patch with transient improvement <1 week, back to baseline

Over next 3 years, patient undergoes:

- Bilateral styloidectomy
- Occipital-cervical fusion
- Surgery for occult tethered cord

Symptoms unchanged, remains profoundly disabled.

But… we tried, used our tools/understanding, and found no answer

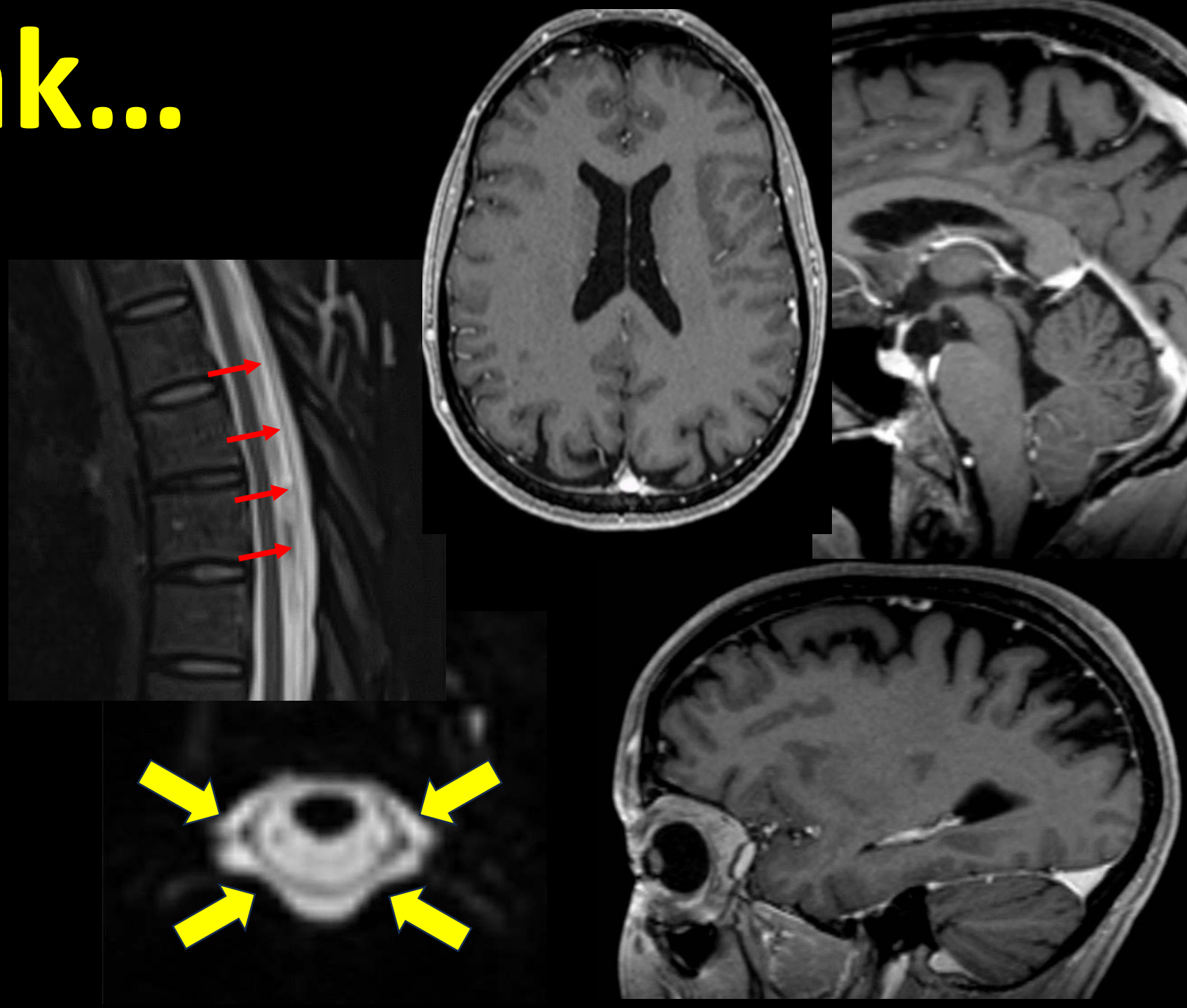
The Fix-Failure Paradox

Technical success, clinical failure

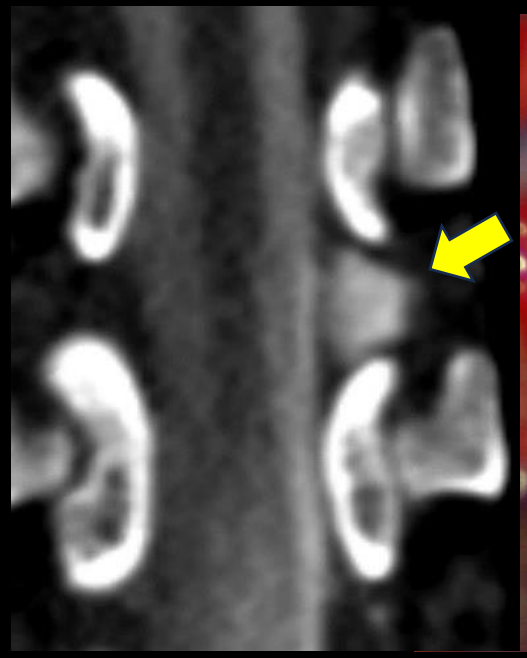
It is a Leak…

Patient A

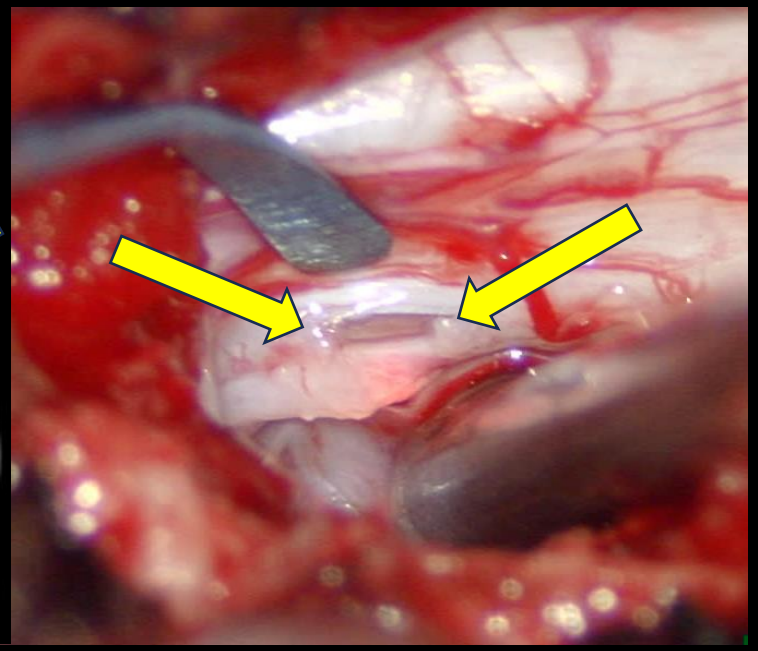
51F, sudden onset suboccipital orthostatic headache 1.5 years ago, associated with severe vertigo and tinnitus



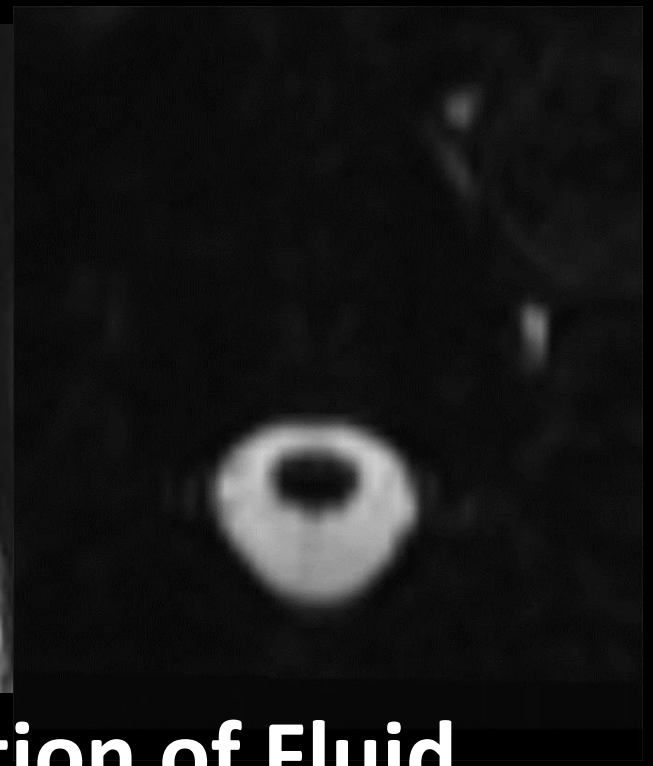
It is a Leak… but



L T9-T10
Lateral Tear



Operative Repair



Resolution of Fluid
Collection

Zero Clinical Improvement

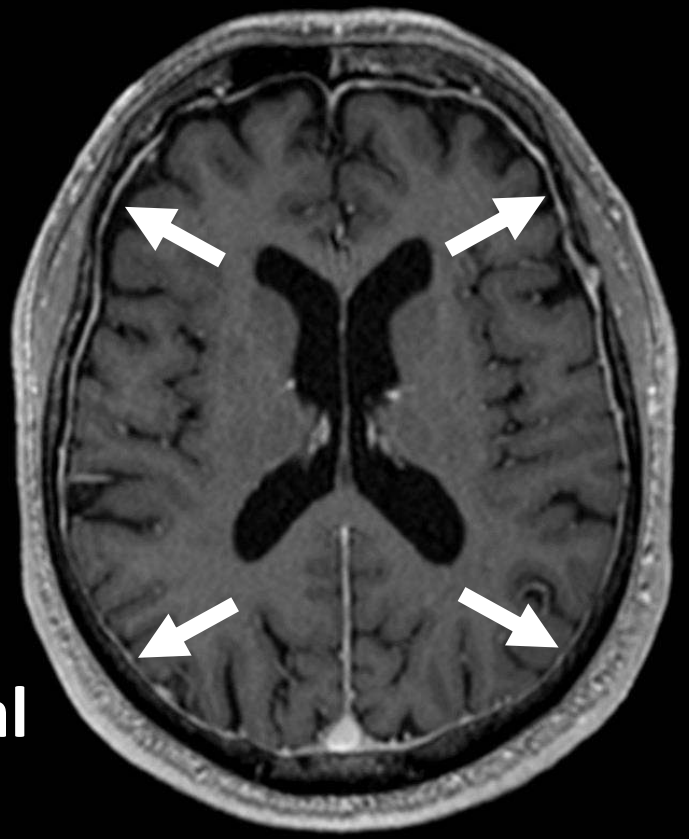
Zero Clinical Improvement

- Repeat myelo: negative
- Patched LP site: 0 benefit
- Diamox trial: 0 benefit

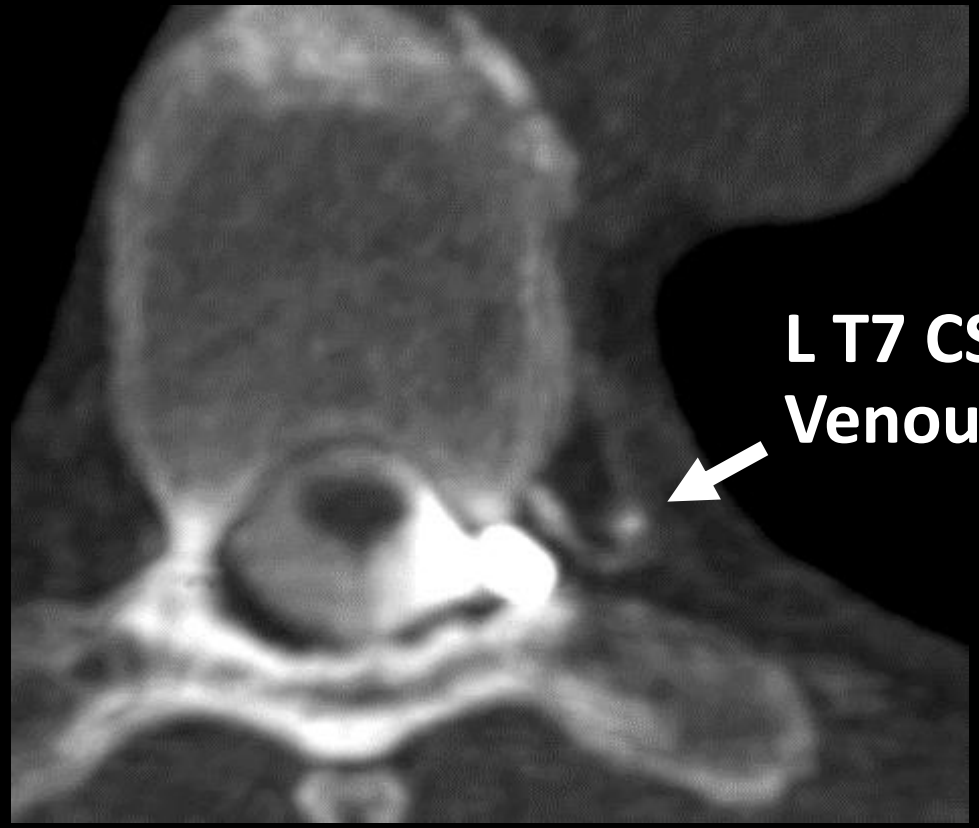
It is a Leak…

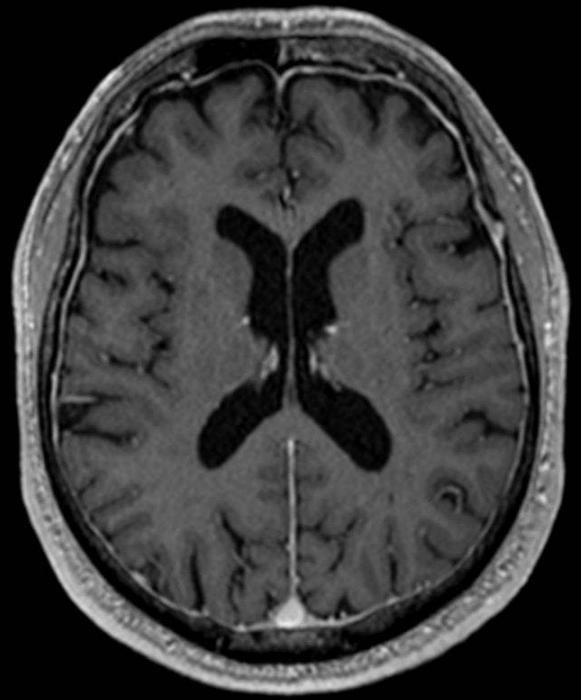
Patient B

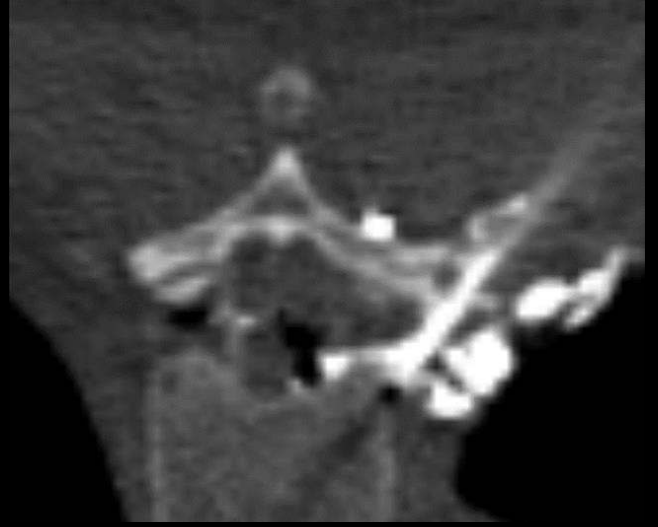
73M, 1 month of left sided HA, worse with cough/bending over, + R sided tinnitus



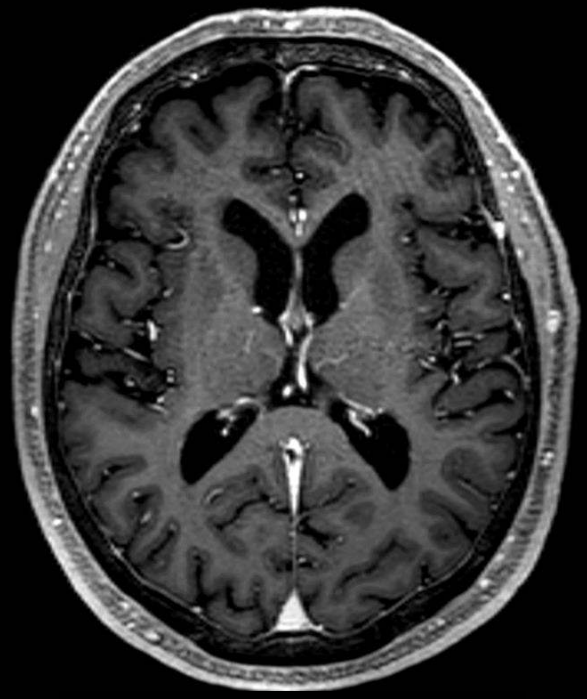
Diffuse Pachymeningeal Thickening







Fibrin x 2:
No symptom improvement



Brain MRI:
Resolved

Transvenous embo:
No symptom improvement

Repeat dCTM
+ DSM
Negative

Surgical Ligation :
No symptom improvement

What is going on???

Central sensitization?

Persistent undetectable leak?

Rebound intracranial hypertension/CSF disequilibrium?

Question:
Can we predict non-responders?

92 ICHD3+ SIH Patients With Documented Clinical Followup

32 ventral, 14 lateral, 46 CVF

Mean follow-up: 5.3 months

Documentation of patient-reported complete, partial, or no improvement

Bern Score Pre and Post With Subcomponents Recorded

PRELIMINARY DATA: Callen, Montes (In Submission)

	Complete Symptom Resolution (n = 42)	Partial (n = 40) or No (n = 10) Symptom Resolution (total n = 50)	p-value
Symptom Duration	2.1 ± 3	5.7 ± 10.3	0.03
Opening Pressure (n = 42)	11.7 ± 4.9	14.8 ± 4.7	0.02
Pre Treatment Bern Score	6.9 ± 1.9	5.3 ± 2.5	0.001
Female Sex	31 (73.8%)	26 (52.0%)	0.032
Dural Thickening	36 (85.7%)	38 (76.0%)	0.24
Subdural Collection	13 (73.8%)	19 (38.0%)	0.48
Mamillopontine Narrowing	31 (73.8%)	32 (64.0%)	0.31
Prepontine Narrowing	32 (76.2%)	29 (58.0%)	0.07
Suprasellar Narrowing	34 (81.0%)	32 (64.0%)	0.07
Venous Engorgement	38 (90.5%)	23 (46.0%)	<0.001

PRELIMINARY DATA: Callen, Montes (In Submission)

Complete Clinical Resolution Predicted By…

Shorter symptom duration
Lesser opening pressure
Higher pre-treatment Bern Score
Venous engorgement…

PRELIMINARY DATA: Callen, Montes (In Submission)

Adjusted Predictors of Clinical Resolution

	Adjusted Odds Ratio	95% CI	p-value
Female Sex	7.0	1.3-38.8	0.026
Pre Treatment Venous Engorgement	27.8	2.4-318	0.008
Symptom Duration	0.98	0.86-1.12	0.776
Opening Pressure	0.91	0.78-1.05	0.195
Pre-Treatment Bern Score	0.78	0.48-1.28	0.329

PRELIMINARY DATA: Callen, Montes (In Submission)

Adjusted Predictors of Clinical Resolution

	Adjusted Odds Ratio	95% CI	p-value
Female Sex	2.76	0.89-8.60	0.079
Symptom Duration	0.99	0.88-1.12	0.882
Pre-treatment Venous Engorgement	14.68	2.12-101.51	0.006
Bern Score Change	0.94	0.71-1.24	0.640

PRELIMINARY DATA: Callen, Montes (In Submission)

Looking Further…

Patients without venous engorgement had significantly higher OP than those with engorgement

(15.9 ± 4.4 cm H_2O) vs

(12.1 ± 4.8 cm H_2O)

p = 0.004

PRELIMINARY DATA: Callen, Montes (In Submission)

RESEARCH ARTICLE OPEN ACCESS

Relationship of Bern Score, Spinal Elastance, and Opening Pressure in Patients With Spontaneous Intracranial Hypotension

Andrew L. Callen, MD, Jack Pattee, PhD, Ashesh A. Thaker, MD, Vincent M. Timpone, MD, David A. Zander, MD, Ryan Turner, BA, Marius Birlea, MD, Danielle Wilhour, MD, Chantal O'Brien, MD, Jennifer Evan, MD, Fabio Grassia, MD, and Ian R. Carroll, MD, MS

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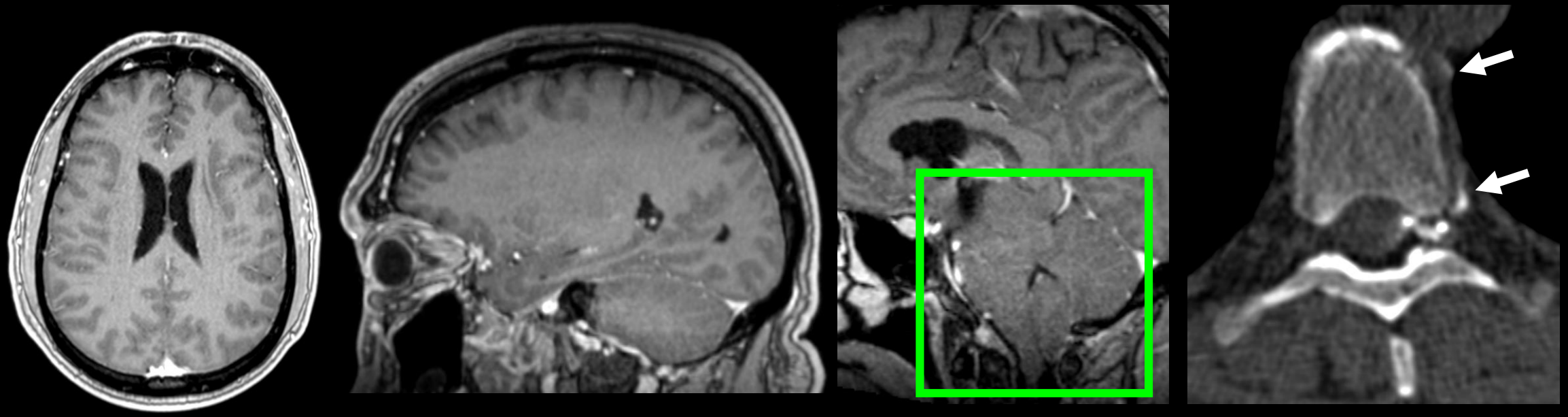
Neurology® 2023;100:e2237-e2246. doi:10.1212/WNL.0000000000207267

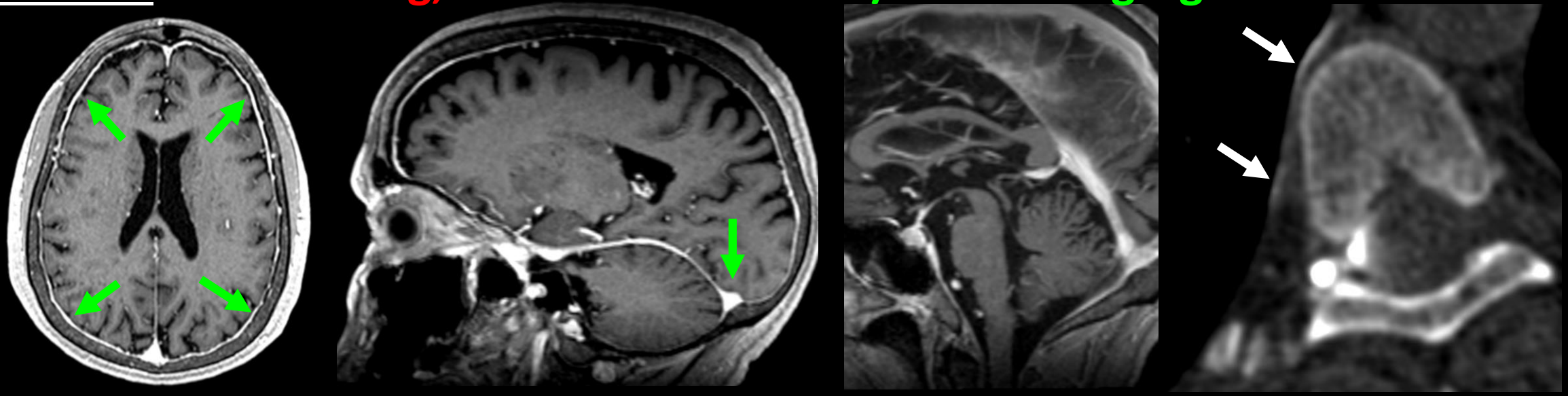
Craniospinal Compliance Related to Venous Engorgement

Beta = 0.269, p < 0.001, 95% CI 0.13-0.41

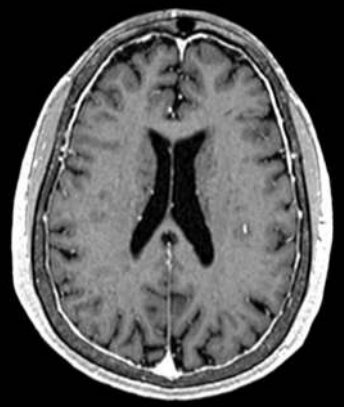
Patient A: Brain Sag Present, No Dural Enhancement/Venous Engorgement

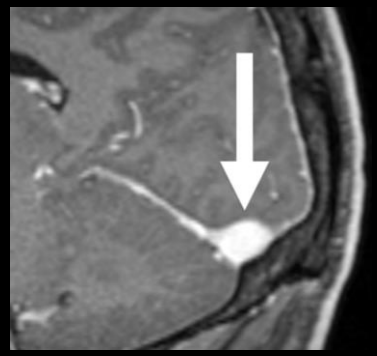


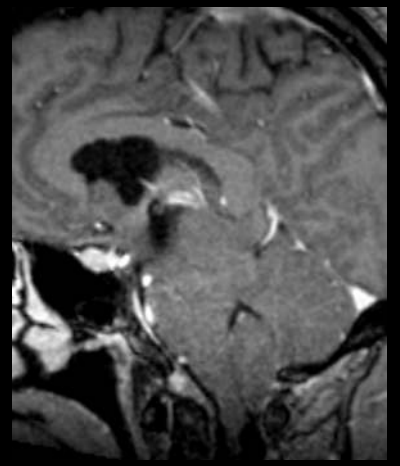
Patient B: No Brain Sag, Dural Enhancement/Venous Engorgement Present



MRI Findings Cluster Together!







Dural Thickening

Prepontine Narrowing

P = 0.007, OR 18.7

No significant associations across groups

p < 0.001, OR 10.1

p = 0.004, OR 7.5

Venous Engorgement

Mamillopontine Narrowing

p < .001 OR 28.9

Suprasellar Narrowing

Callen et al Headache 2024.

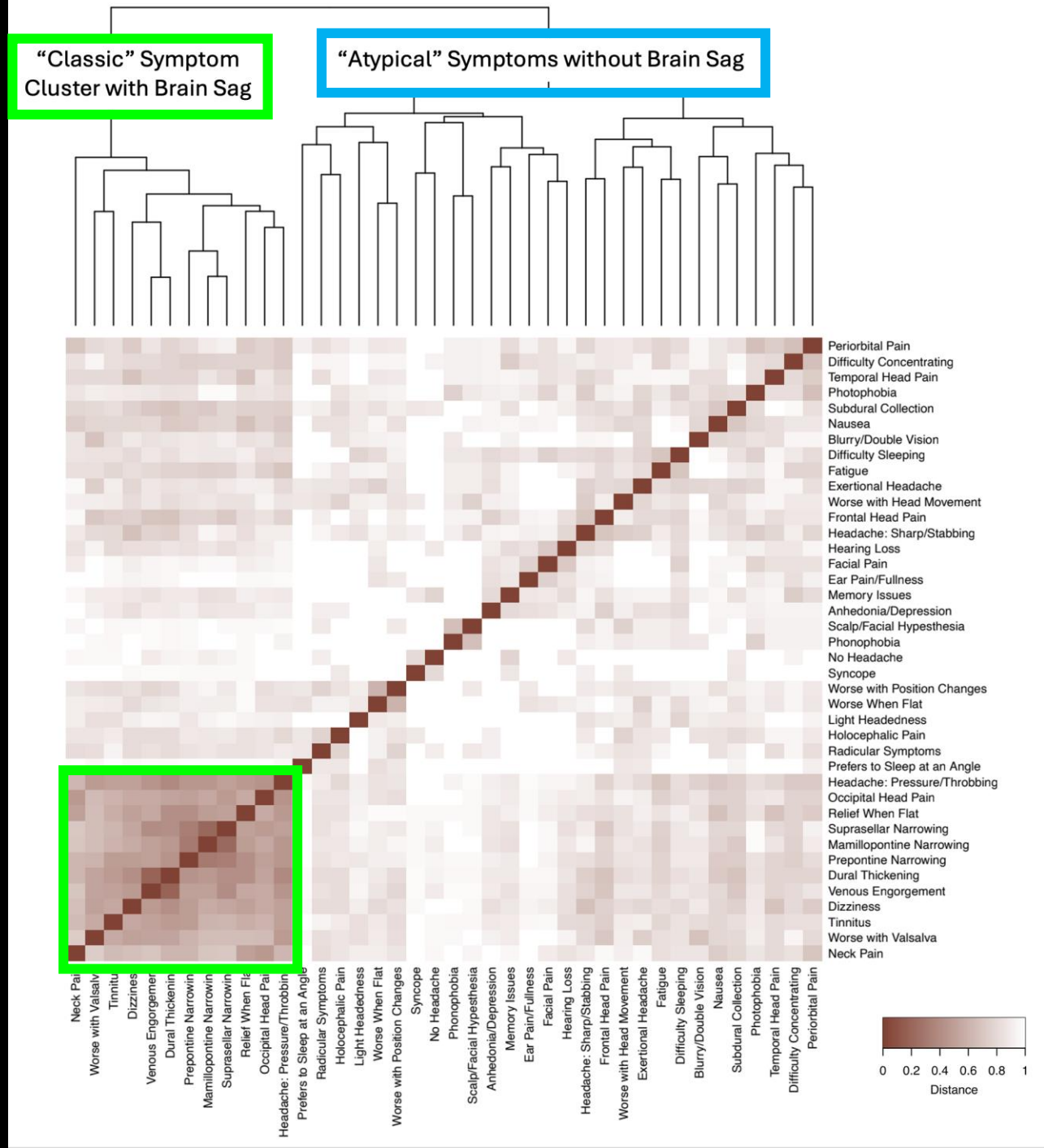
Clinical/Imaging Clusters

‘Classic’ Symptom Cluster

- Quality: Pressure/Throbbing
- Location: Occipital
- Relief when flat
- Comorbid neck pain

‘Atypical’ Symptom Cluster

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



Callen et al Headache 2024.

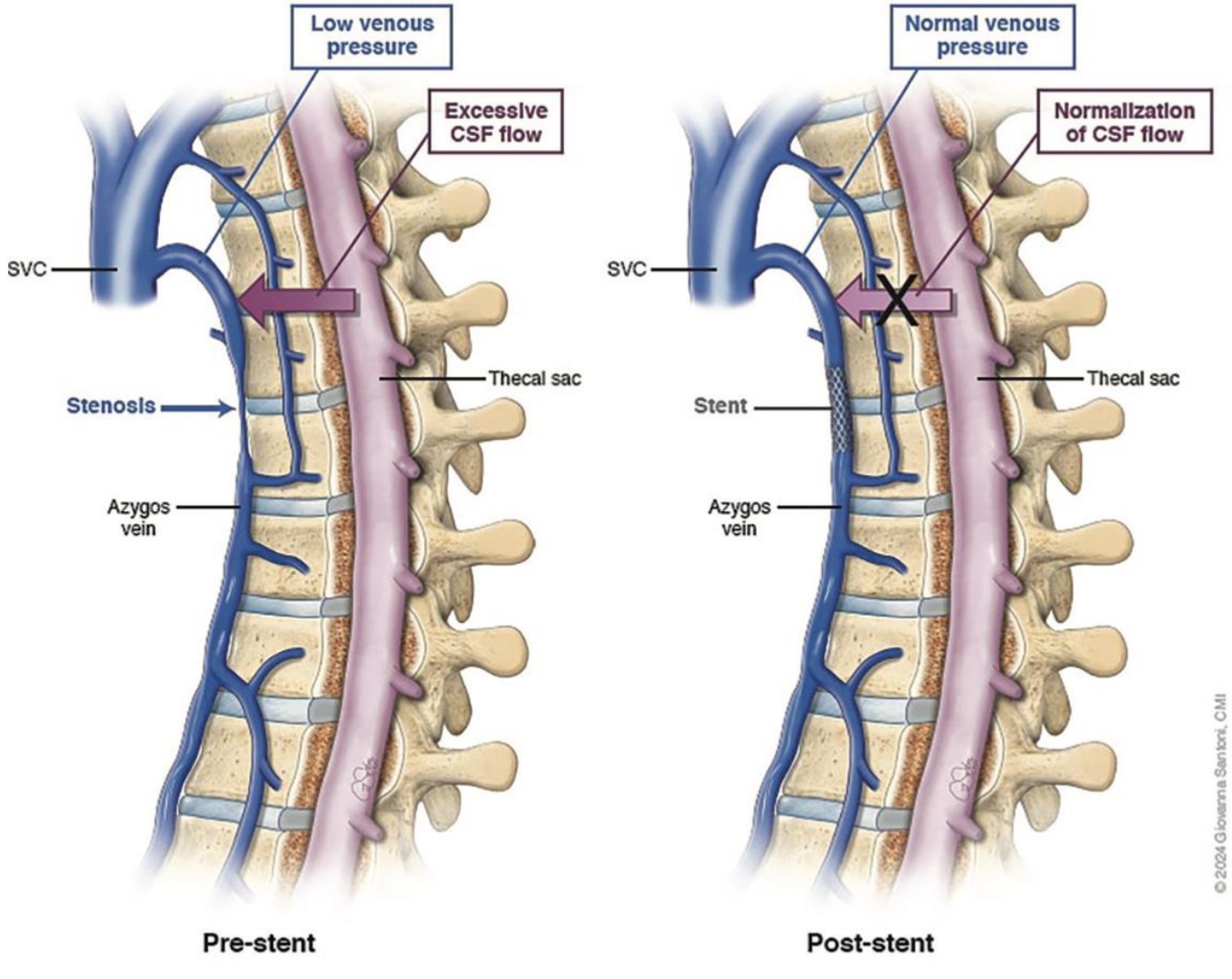
What else have we learned recently about venous pathology and SIH?

Azygos Vein Stenosis in Frontotemporal Dementia Sagging Brain Syndrome

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

AJNR
AMERICAN JOURNAL OF NEURORADIOLOGY

- 21 patients with bvFTD Sagging Brain Syndrome
- Severe brain sag
- No leak found in any
- 7 patients with venous stenosis
- Treatment of azygos stenosis reversed sag/symptoms in 3



Impaired Cranial Venous Outflow Predicts RIH:

RESEARCH

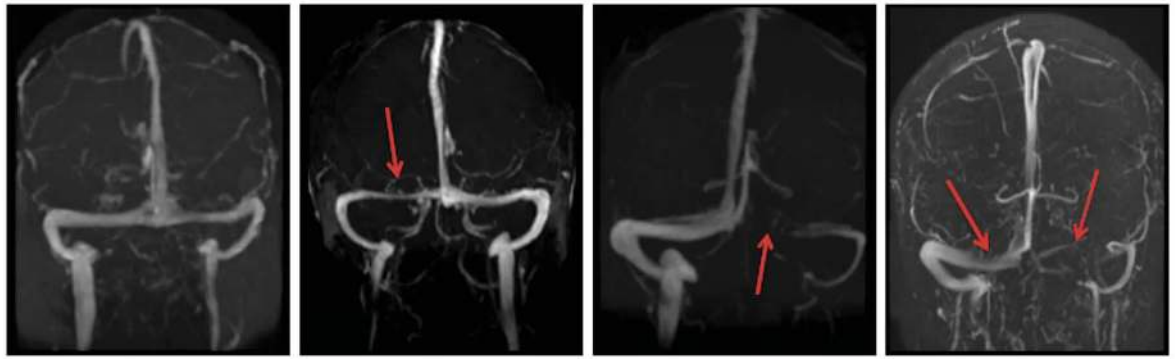
Rebound high-pressure headache after treatment of spontaneous intracranial hypotension

MRV study

Wouter I. Schievink, MD, M. Marcel Maya, MD, Stacey Jean-Pierre, PA-C, Franklin G. Moser, MD, MMM, Miriam Nuño, PhD, and Barry D. Pressman, MD

Correspondence
Dr. Schievink
schievinkw@cshs.org

Figure Magnetic resonance venography (MRV) scoring system (Higgins et al.[25]) for patients with rebound high-pressure headaches following treatment for SIH

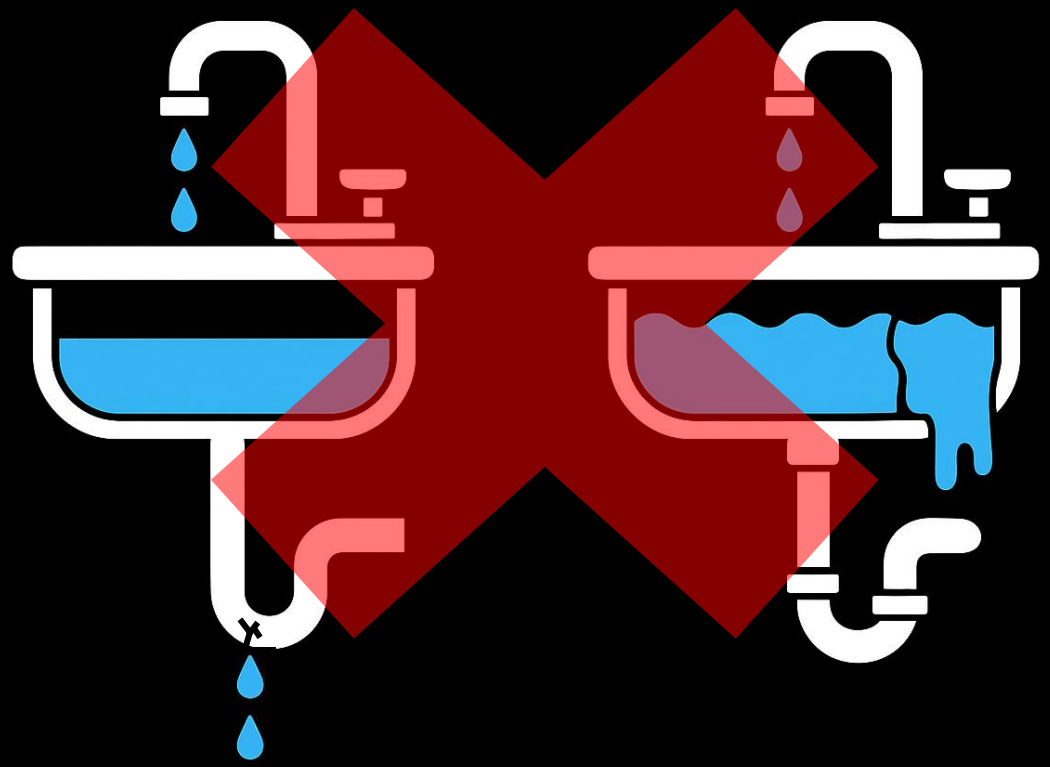


0/0 MRV score: 0 | 1/0 MRV score: 1 | 0/2 MRV score: 2 | 1/2 MRV score: 3

Normal is 0; one or more areas of focal narrowing is 1; one or more signal gaps is 2. The scores are summated to give values ranging from 0 to 4.

RIH means leak is fixed?

No.



RIH reflects a phenomenon of venous dynamics in response to pressure change

Three multicenter studies on each leak type.
No association between RIH and treatment success.

Factors Predictive of Treatment Success in CT-Guided Fibrin Occlusion of CSF-Venous Fistulas: A Multicenter Retrospective Cross-Sectional Study

Andrew L. Callen, Lalani Carlton Jones, Vincent M. Timpone, Jack Pattee, Daniel J. Scoffings, David Butteriss, Thien Huynh, Peter Y. Shen, and Mark D. Mamlouk

Radiology

ORIGINAL RESEARCH • NEURORADIOLOGY

Efficacy of Traditional Epidural Patching versus Patching within Spinal Longitudinal Extradural Collections for Ventral Dural Cerebrospinal Fluid Leaks

Andrew L. Callen, MD[1] • Samantha L. Pisani Petrucci, MD, PhD[1] • Peter Lennarson, MD[2] • Mark F. Sedrak, MD[3] • Adriana Gutierrez, PA[4] • Mark D. Mamlouk, MD[4,5]

Outcomes of CT-Guided Targeted Epidural Patching For Lateral Dural Tears In Spontaneous Intracranial Hypotension: A Multicenter Retrospective Cohort Study

Andrew L. Callen, Daniel Montes, Debayan Bhaumik, Peter Lennarson, Mark D. Mamlouk, Niklas Lützen, Jürgen Beck, Horst Urbach, Daniel Scoffings, David Butteriss, Lalani Carlton Jones

So Should We Be Looking at the IJVs?

Human cerebral venous outflow pathway depends on posture and central venous pressure

J. Gisolf[1], J. J. van Lieshout[2], K. van Heusden[3], F. Pott[4], W. J. Stok[1] and J. M. Karemaker[1]

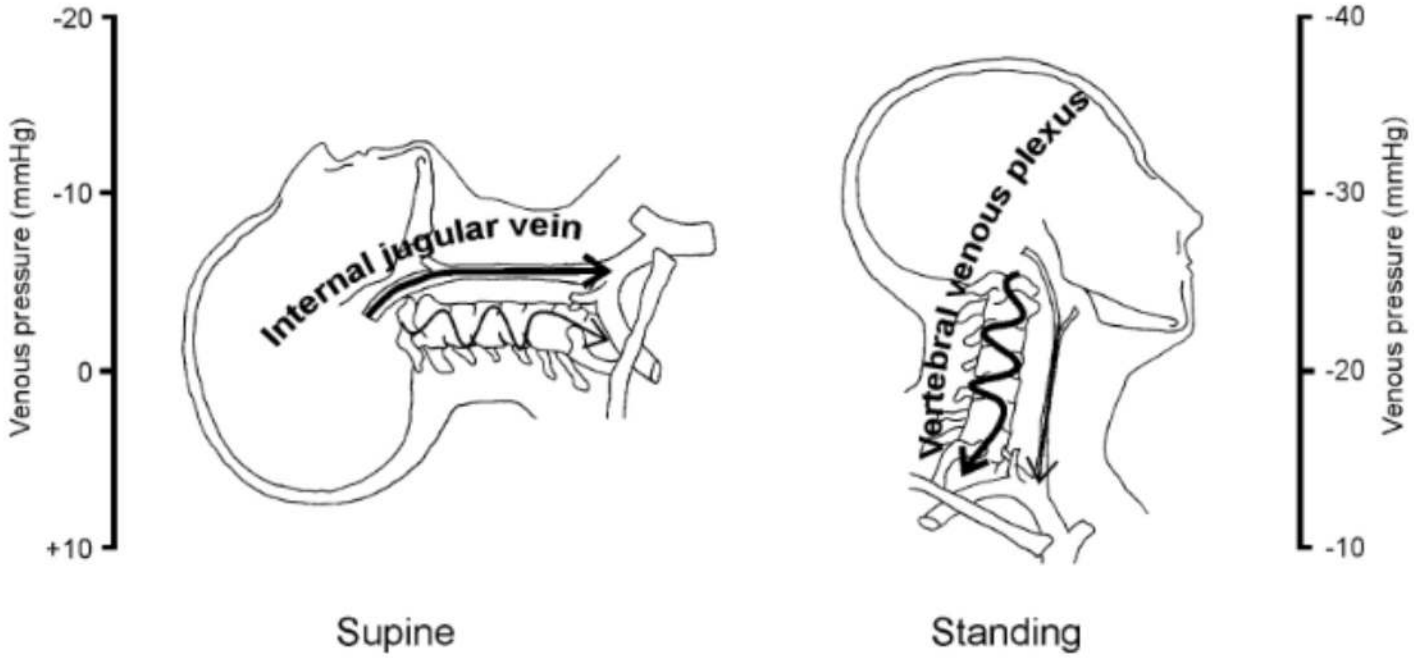


Figure 6. Illustration of the cerebral venous outflow pathways
Schematic representation of cerebral venous outflow pathway primarily via the internal jugular veins in the supine position (left) and the vertebral venous plexus in the upright position (right).

The vertebral venous plexus is responsible for majority of venous drainage in the upright position.

Vertebral venous plexus = next frontier of understanding?

Clinical Outcomes

CSF Dynamics Hi + Low

Venous Plasticity

~~Pressing~~ *(Pressure?)* Issues in SIH

System Pressures

Timely Diagnosis, Access, Billing/Infrastructure

Clinical Pressures

When Cure Isn't Cure
What We Do Not Yet Understand

Thank You!

Maria Puello, MD

CU CSF Leak Team

Peter Lennarson, MD

Debayan Bhaumik, MD

Katie Kerrigan, NP

Andrea Walters

Erika Rivas

Daniel Montes, MD

University of Colorado
Anschutz Medical Campus

University of Colorado
CSF Leak Program