

SUSPECTED SPONTANEOUS INTRACRANIAL HYPOTENSION

*A Neurologist's Approach at a
Community Medical Center*



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Disclosures

- ◆ No financial relationships with entities that produce treatment or investigative interventions, devices or therapies.

- ◆ Member of the Medical Advisory Board of the Spinal CSF Leak Foundation

CHALLENGES

The variability in just about every aspect of this disorder is substantial.

It can be difficult to diagnose and treat this syndrome.

-- Bahram Mokri, M.D



CHALLENGES

Sophisticated imaging and experience with the vast clinical presentations of this disorder are keys to the diagnosis.



CASE 1

- ◆ 43 y M
- ◆ Normal weight and body habitus
- ◆ PMHx – several fractures secondary to competitive/extreme mountain biking
- ◆ Chiropractic table drop
 - ◆ Gradual onset of persistent posterior headache, orthostatic – fluctuates, pressure, brain fog
 - ◆ Sharp pain around T7
- ◆ MRI Brain
 - ◆ No abnormalities detected
- ◆ MRI Thoracic Spine
 - ◆ Epidural CSF collection
- ◆ Requested large volume blood patch in thoracic region



LARGE VOLUME THORACIC EPIDURAL BLOOD PATCH

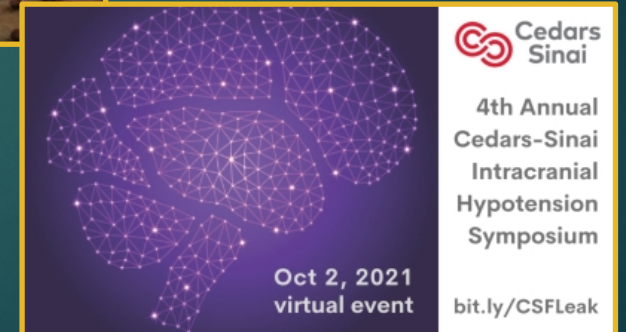
- ◆ **But he hasn't had an LP?!**
- ◆ **Can that even happen?**

CASE 1

- ◆ The interventionalist read about SIH
- ◆ Performed low upper lumbar EBP
- ◆ Patient had temporary improvement
- ◆ Repeat MRI, leak still present
- ◆ Repeat EBP
- ◆ Relief from headache, cognitive fogging, photophobia, fatigue
- ◆ Neck/back pain remains, but dulled and tolerable

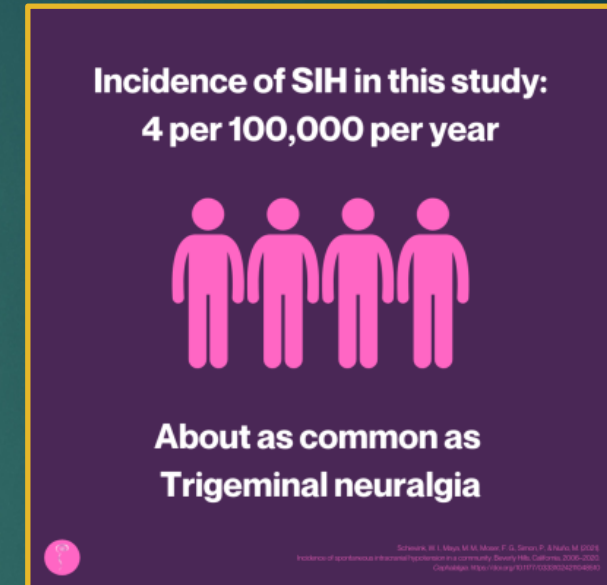
MAKE FRIENDS

- ◆ If possible, try to meet your interventionalists
- ◆ Take food
- ◆ Take coffee
- ◆ Offer to present to the department
- ◆ Suggest great symposiums that they might like to attend



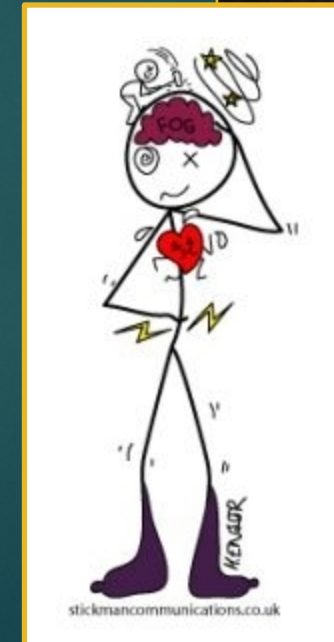
EDUCATE

- ◆ Know the literature
 - ◆ Throw in some numbers and comparisons
 - ◆ trigeminal neuralgia
 - ◆ 50% aneurysmal subarachnoid hemorrhage
 - ◆ idiopathic intracranial hypertension
 - ◆ cluster headache
- ◆ Be prepared with review articles
- ◆ Share
 - ◆ Websites
 - ◆ <https://rarediseases.org/rare-diseases/spontaneous-intracranial-hypotension/>
 - ◆ <https://spinalcspfleak.org/>
 - ◆ Journal Articles
 - ◆ Presentations



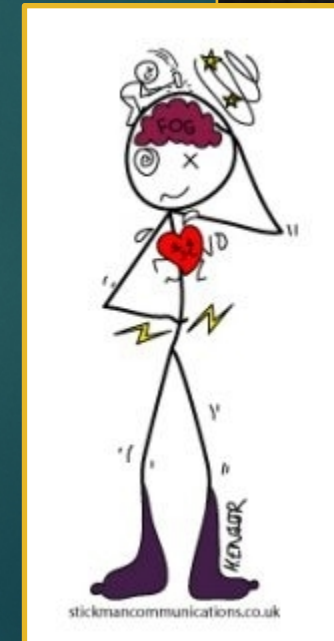
CASE 2

- ◆ 27 y F
- ◆ CM, EDS, POTs, ?MCAS, history of spinal CSF leak
- ◆ Managed with Botox, regular GONB/SONB
- ◆ Worsening HA, cognitive fogging, photophobia
- ◆ No suggestion/evidence of leak on imaging
- ◆ Unable to maintain upright
- ◆ Requested EBP ----->



CASE 2

- ◆ Went to a different hospital system ER
 - ◆ Temporary benefit ~ 48 hrs
- ◆ Admitted to our hospital system for assistance several weeks later
 - ◆ Despite Neurohospitalist in IR suite encouraging, IR refused to give EBP
- ◆ Called a FRIEND – she was able to get EBP
- ◆ Rebound intracranial hypertension (RIH)
 - ◆ acetazolamide



TREATMENT DILEMMAS



Be creative.



Be persistent.

Shake it off.



BUT WHAT ABOUT DIAGNOSTIC DILEMMAS

- ◆ CSF Leak Imaging -

- ◆ Make certain your Imaging Requests CLEARLY indicate that its for CSF Leak

- ◆ Optimal protocol?

- ◆ Are the radiologists aware of the more nuanced signs/measurements that might be helpful?

MAKE F.R.I.E.N.D.S

+



- ◆ What additional tests might you consider?

- ◆ Autonomic Testing
- ◆ Research Testing

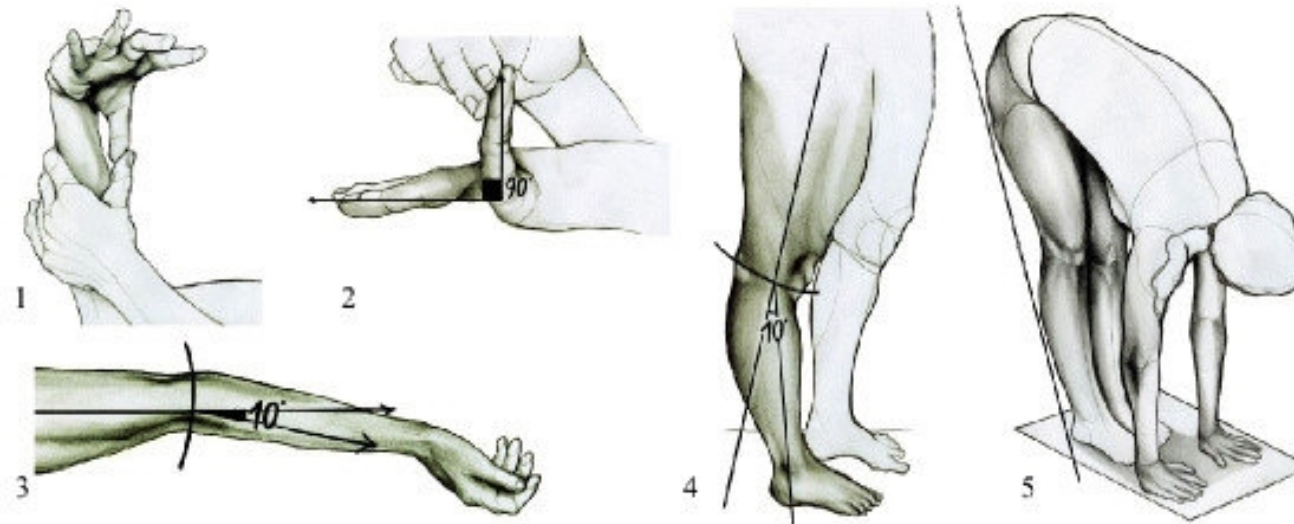
HOW CONVINCED ARE YOU?

- ◆ What are the features that convinces you that your patient has suspected SIH?
 - ◆ Orthostatic headache?
 - ◆ Risk factors – EDS/joint hypermobility, trauma
 - ◆ Signs on Imaging?
 - ◆ Intrascapular pain?
 - ◆ Auricular symptoms? Tinnitus, hyperacusis, muffled hearing, dizziness
 - ◆ Visual changes? Blurriness, diplopia, other
 - ◆ Cognitive complaints
 - ◆ Other – movement disorders, bowel and bladder issues, radicular complaints

BEIGHTON SCORE

Specific joint laxity	YES		NO
1. Passive apposition of thumb to forearm	<input type="checkbox"/> Left	<input type="checkbox"/> Right	<input type="checkbox"/>
2. Passive hyperextension of V-MCP > 90°	<input type="checkbox"/> Left	<input type="checkbox"/> Right	<input type="checkbox"/>
3. Active hyperextension of elbow >10°	<input type="checkbox"/> Left	<input type="checkbox"/> Right	<input type="checkbox"/>
4. Active hyperextension of knee >10°	<input type="checkbox"/> Left	<input type="checkbox"/> Right	<input type="checkbox"/>
5. Ability to flex spine placing palms to floor without bending knees	<input type="checkbox"/>		<input type="checkbox"/>

*Each "YES" is 1 point. A score ≥ 4 out of 9 is generally considered an indication of JH. (MCP: metacarpophalangeal).



SEEPS AND COPS

Schievnik WI. JAMA. 2006;295:2286-2296

Subdural fluid collection

Enhancement of pachymeninges

Engorgement of veins

Pituitary hyperemia

Sagging of brain

Chiari

Optic chiasm

Pontine flattening *

Sinking iter**



In patients with headache and one of these imaging features, always consider SIH, regardless of whether headache is orthostatic

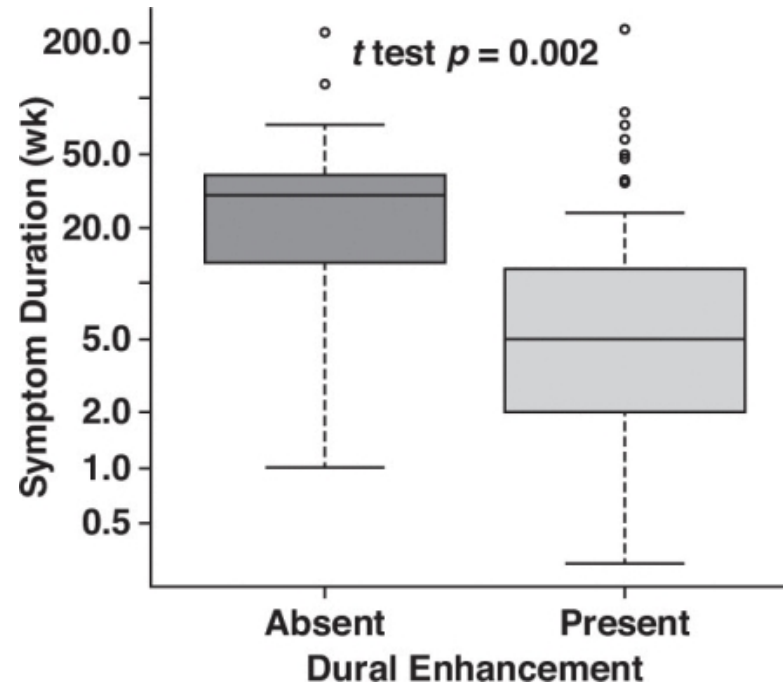
*crowding, distortion of post fossa, obliteration of basal cisterns (peripontine, perichiasmatic)

** descent brainstem, mesencephalon, diencephalon

SIGNS OF CSF LEAK MAY BE TRANSIENT

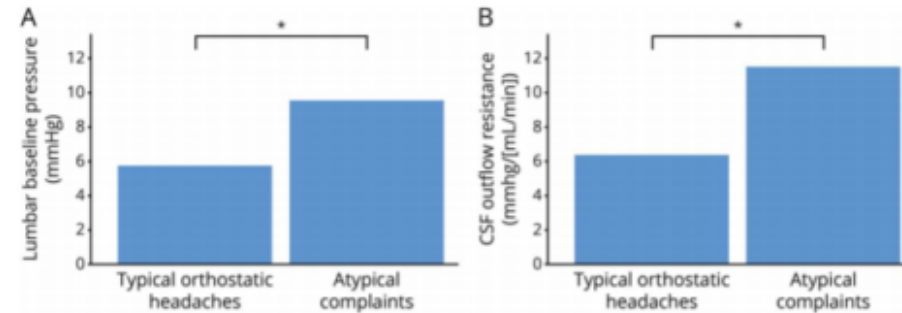
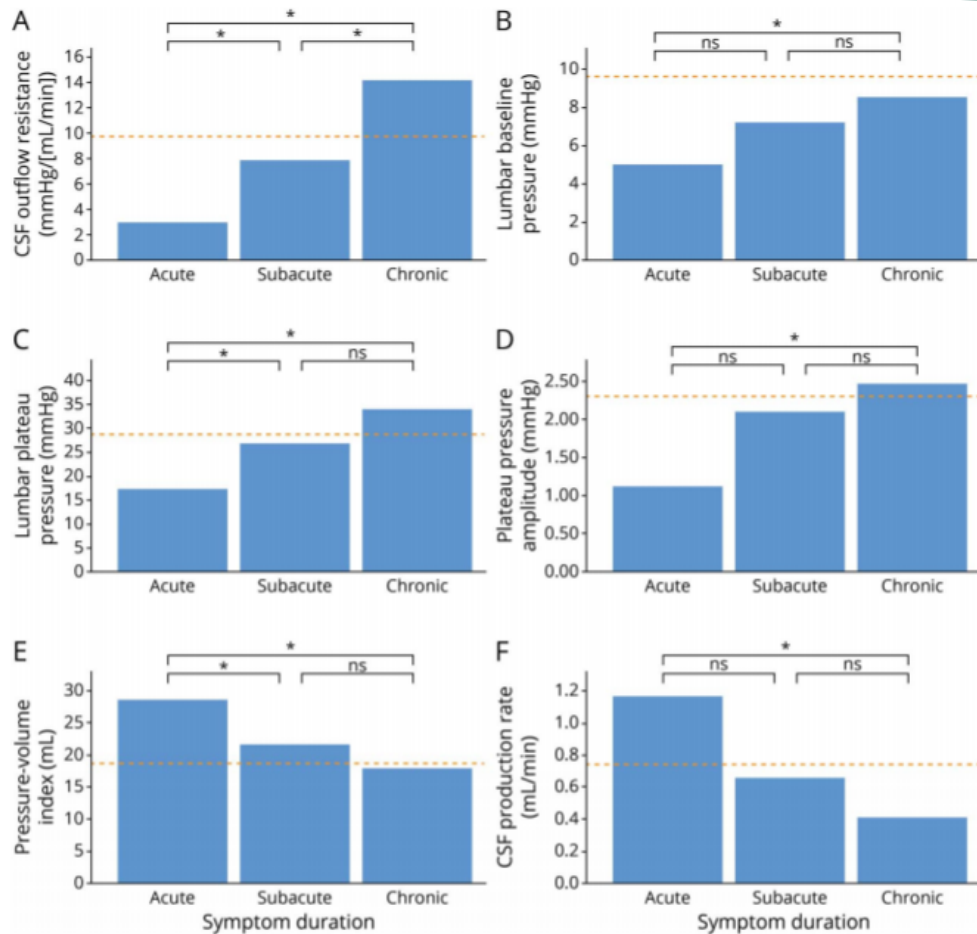
specifically dural enhancement

The mean duration of headache symptoms for subjects with dural enhancement present was 15.1 weeks compared with 45.3 weeks for those with no dural enhancement.

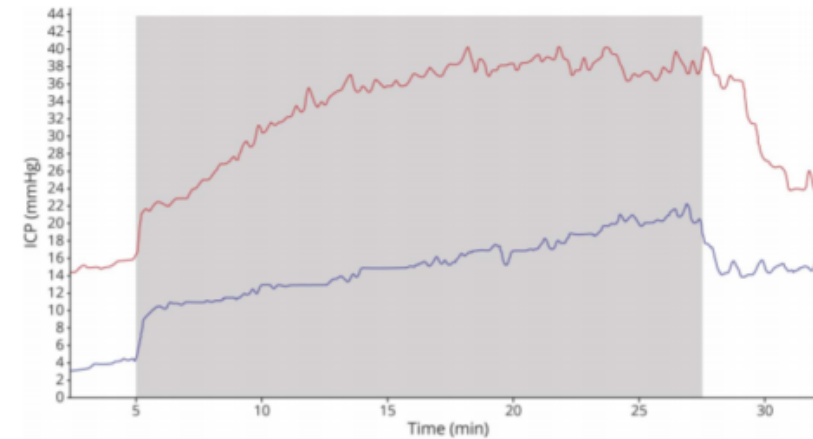


CSF Pressure Dynamics Change Over Time

Häni L, *et al.* (2020)

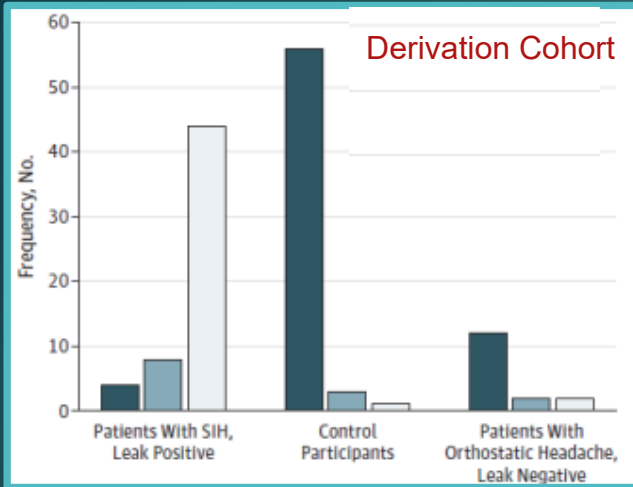


Comparison of lumbar baseline pressure (A) and resistance to CSF outflow (B) between patients with typical orthostatic headache and those with atypical complaints, irrespective of symptom duration. *Statistically significant.



The red curve describes a curve from a patient in the chronic phase of the disease, which is similar to a normal curve. In contrast, the blue curve, describing a patient in the acute phase, starts at a lower baseline pressure, rises more slowly during infusion, and reaches the plateau later, if at all, and at a lower plateau pressure level. ICP = intracranial pressure.

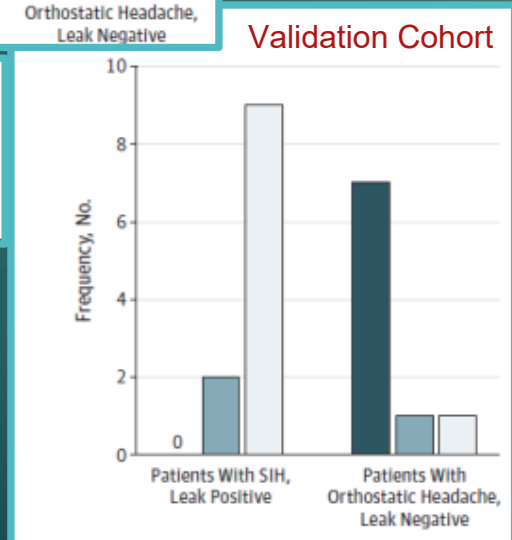
Assessing CSF Leak with a Scoring System Based on Brain MRI



Score ≤ 2
 92.9% sensitivity
 93.3% specificity
Score ≥ 5
 78.6% sensitivity
 98.3% specificity

■ Score ≤ 2
 ■ $2 < \text{Score} < 5$
 ■ Score ≥ 5

Score ≤ 2
 100% sensitivity
 77.8% specificity
Score ≥ 5
 81.8% sensitivity
 88.9% specificity

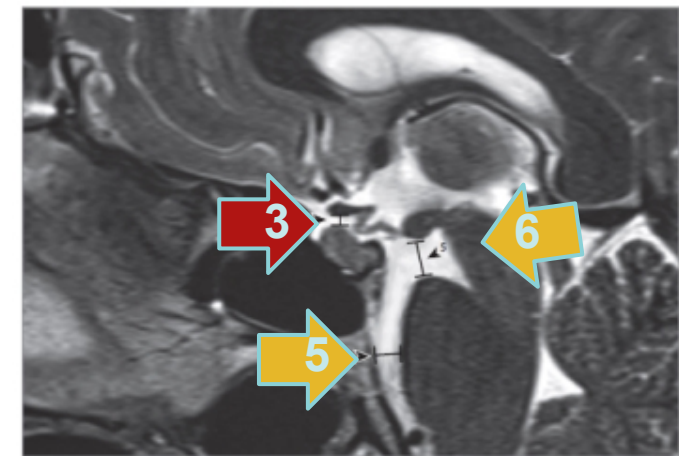
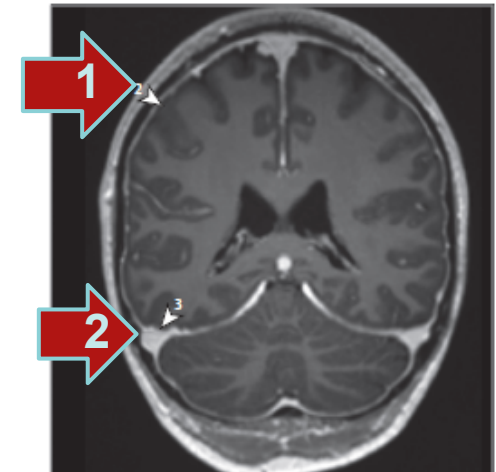
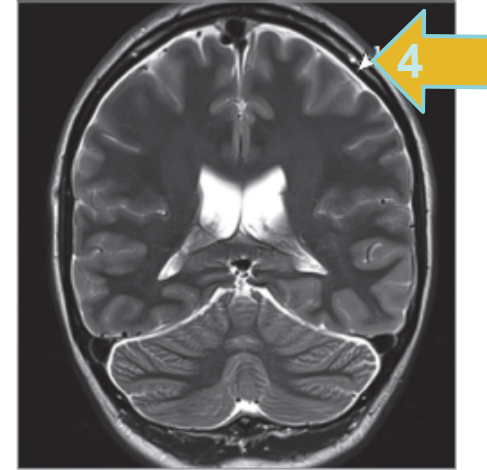


Major (2 points each)

- 1) pachymeningeal enhancement
- 2) engorgement of the venous sinus
- 3) effaced suprasellar cistern (≤ 4 mm)

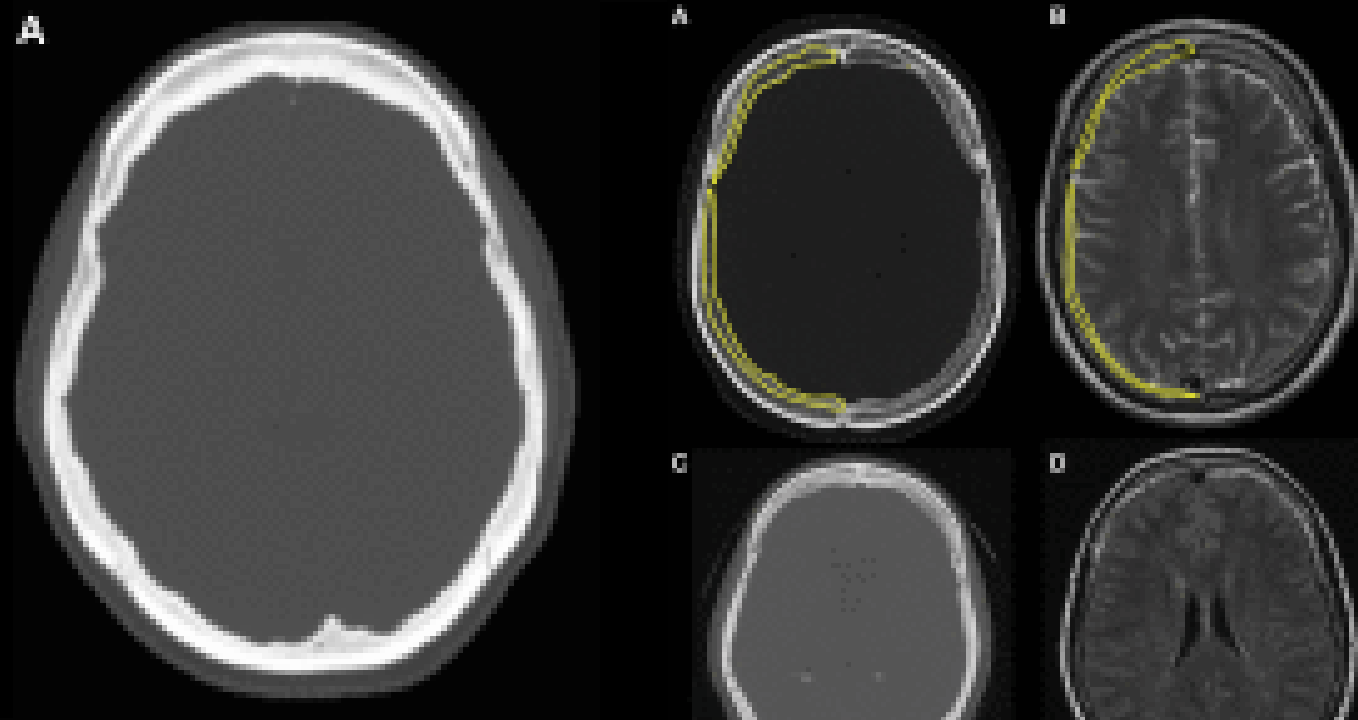
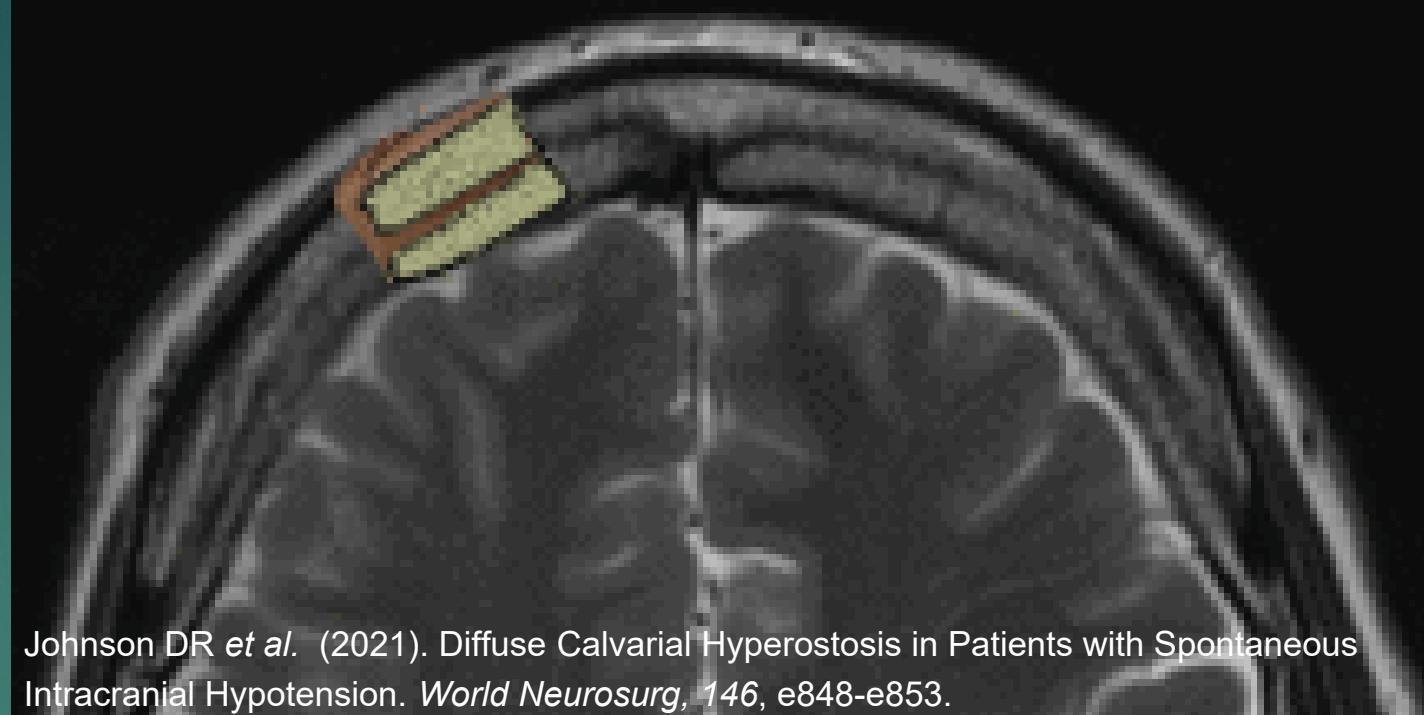
Minor (1 point each)

- 4) Subdural fluid collection
- 5) effacement of the prepontine cistern (≤ 5 mm)
- 6) mamillopontine distance (≤ 6.5 mm)



Diffuse Calvarial Hyperostosis in SIH

- ◆ Retrospective review of CT & MRI of 285 patients who had myelography for SIH
 - ◆ generalized calvarial thickening
 - ◆ secondary layer of bone
 - ◆ typical benign hyperostosis frontalis was excluded
- ◆ 14% - diffuse calvarial hyperostosis
 - ◆ 80.0% - distinct circumferentially layered appearance
 - ◆ 20.0% had generalized calvarial thickening without layering

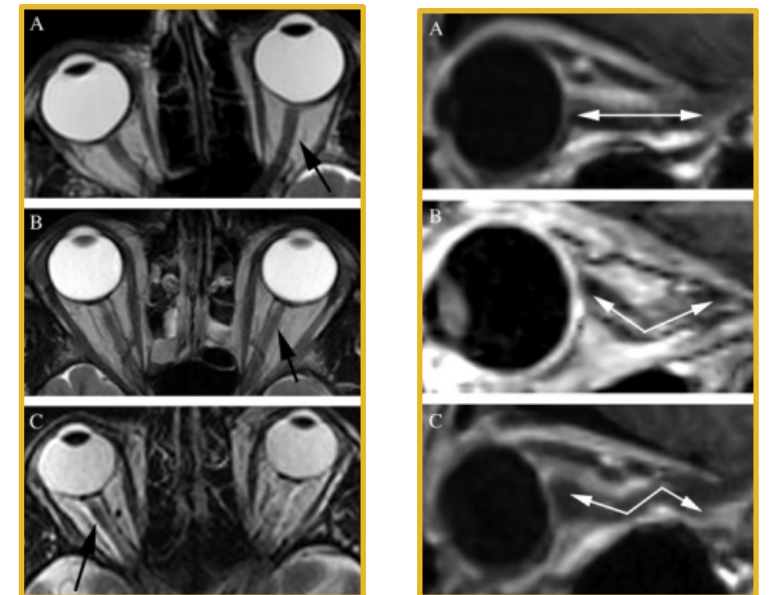
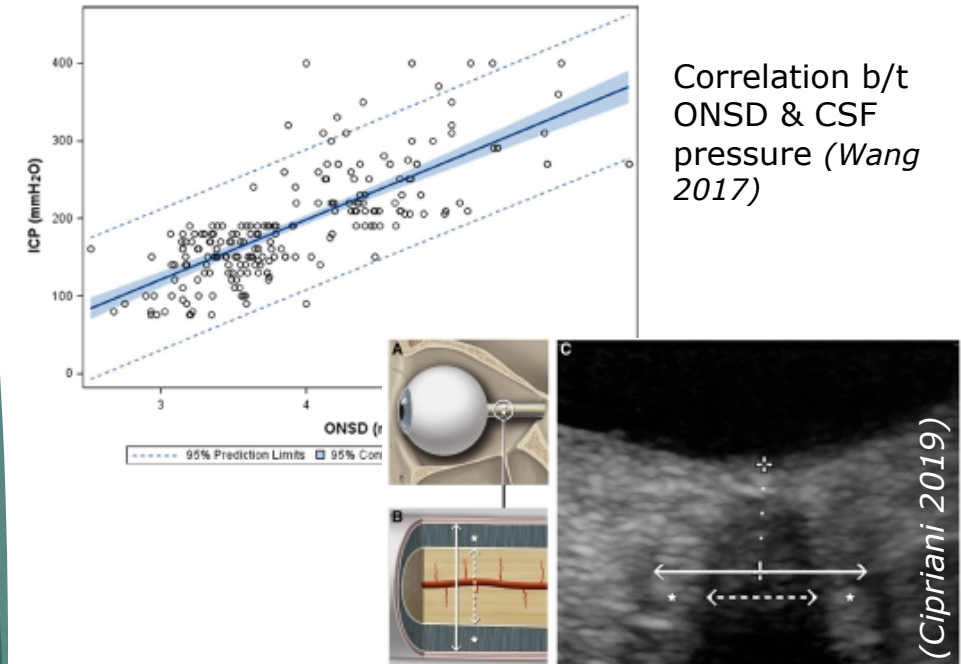


Optic Nerve as a Diagnostic Tool

- ◆ Wang *et al.* (2017)
 - ◆ $ICP = -111.92 + 77.36 \times ONSD$
 - ◆ ONSD independent predictor of ICP.
 - ◆ Sex, age, BMI, waist circumference, head circumference, and DBP
- ◆ Gupta and Pachisia (2019)
 - ◆ CSF pressure of >20 cm H₂O ~ ONSD >0.63 cm (n=100 patients, 81% men, $r = 0.715$)

Orbit findings in Intracranial Hypotension are typically the inverse of the findings seen in IIH: Diminished or absent optic nerve sheath CSF, relative straightening of the optic nerve angle. (Holbrook 2017)

Optic nerve sheath CSF: A–C: Axial T2w images through the orbits; A. Decreased CSF in the optic nerve sheath in a patient with IH (arrow); B. Normal optic nerve sheath CSF in a normal control (arrow); C. Increased CSF in the optic nerve sheath in a patient with idiopathic intracranial hypertension (arrow).



Orthostatic change alters ONS diameter in SIH, but not POTs

	POTS + OSH (n = 7)	POTS—OSH (n = 7)	SIH (n = 5)	Control (n = 8)	P value
Optic nerve sheath diameter					0.003**
Supine	4.8 ± 0.1	4.5 ± 0.2	5.3 ± 0.1	4.8 ± 0.2	
Upright	4.8 ± 0.2	4.5 ± 0.2	4.6 ± 0.1	4.6 ± 0.2	
Optic nerve diameter					0.080
Supine	3.7 ± 0.3	3.1 ± 0.2	3.5 ± 0.2	3.8 ± 0.2	
Upright	3.8 ± 0.2	3.3 ± 0.2	3.4 ± 0.2	3.6 ± 0.2	
Perineural space diameter					0.050*
Supine	2.5 ± 0.1	2.6 ± 0.2	3.1 ± 0.1	2.2 ± 0.1	
Upright	2.1 ± 0.2	2.4 ± 0.1	2.4 ± 0.2	2.2 ± 0.1	

Note. Values are reported in mm. Data are reported as mean ± S.E.M. P values refer to the interactions of group and position, derived from analyses of variance for repeated measures.

*P ≤ 0.05;

**P ≤ 0.01

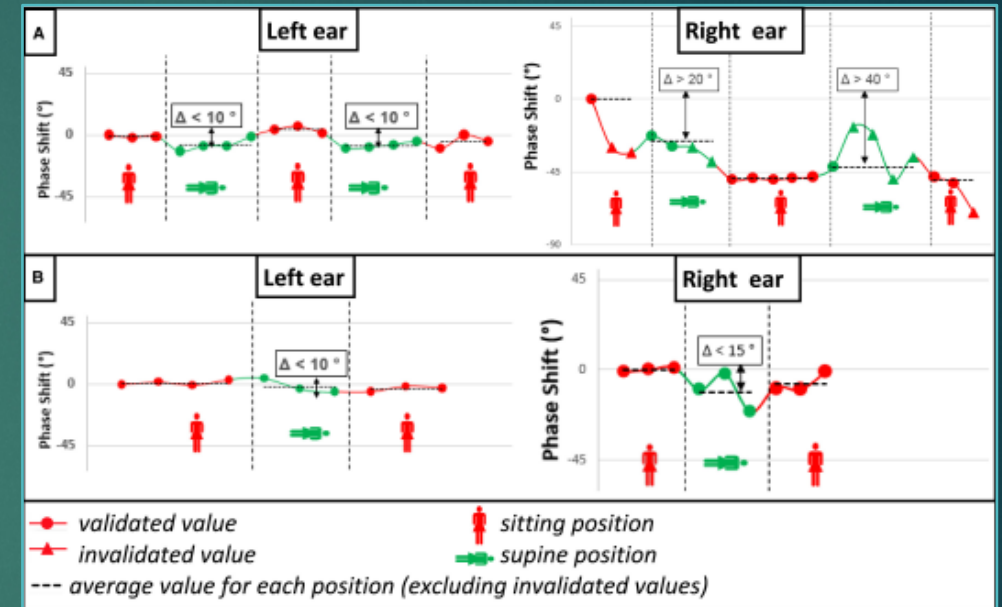
POTS, postural orthostatic tachycardia syndrome; OSH, orthostatic headache; SIH, spontaneous intracranial hypotension

Otoacoustic Emissions (OAE) for Diagnosis of SIH

Redon S, et al. (2019)

- ◆ OAE are low - level sounds generated by the cochlea spontaneously or in response to external stimuli
- ◆ Distortion product OAE (DPOAE) are a subtype of OAE produced in response to 2 pure tone frequencies (f_1 and f_2) in the ear canal
- ◆ 4 patients' DPOAE w/ positional change
 - ◆ (reported norms between 9° - 37°)

EBP



- ◆ 4 OSHA, 3 negative MRI brain/spine, 2 tinnitus, 1 dizziness
- ◆ All abnormal postural phase shift DPOAE pre-EBP
- ◆ All normalized post-EBP

PREPARE YOUR PATIENTS *and yourself...*

- ◆ May be a long process
- ◆ Risks/adverse reactions
 - ◆ EBP
 - ◆ Invasive testing
 - ◆ Out of state referrals
- ◆ Insurance
- ◆ Support



Post-SIH-treatment Rebound Pressure-HA

N = 113 (59% F)

OSHA: 109

non-positional HA: 3

reverse OSHA: 1

Rebound high-pressure HA

31 (27.4%)

RHP-HA reported

MRV score 0: 14%

MRV Score 1: 24%

MRV Score 2: 44%

MRV Score 3: 67%

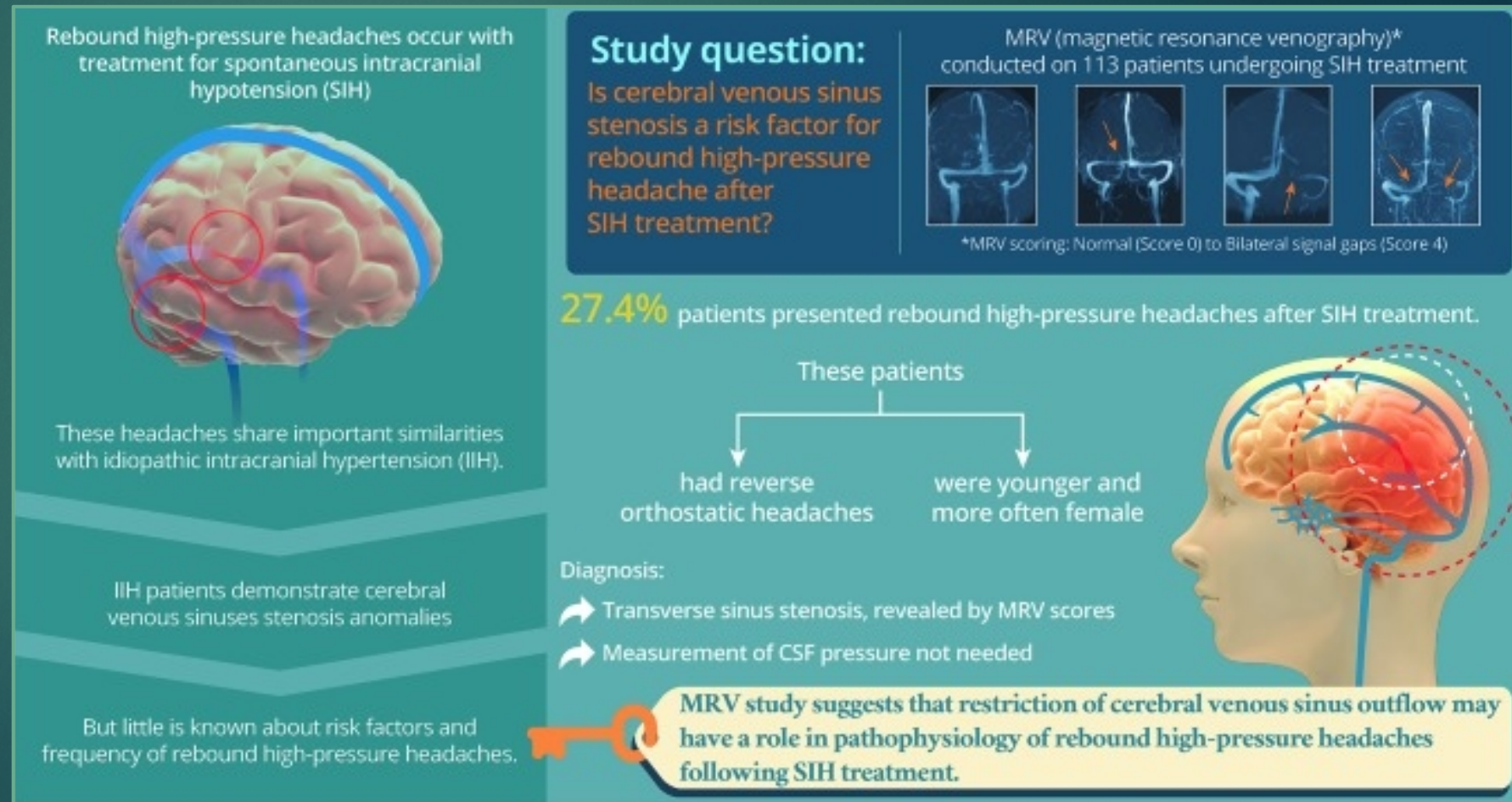
Funduscopy: 25

Papilledema: 2

No difference in BMI ~ 25

Schievink WI, et al. (2019)

MRV studies were performed prior to treatment of SIH.



SUMMARY

SUMMARY



Be creative.



Be confident in your own diagnostic suspicion.



Be persistent.



Be prepared.



because your dura matters®

Thank you.

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Review Articles

- ◆ Bond, K. M., et al. (2020). "Spontaneous Intracranial Hypotension: Atypical Radiologic Appearances, Imaging Mimickers, and Clinical Look-Alikes." AJNR Am J Neuroradiol **41**(8): 1339-1347.
- ◆ Ferrante, E., et al. (2020). "Spontaneous intracranial hypotension: review and expert opinion." Acta Neurol Belg **120**(1): 9-18.
- ◆ Martineau, P., et al. (2020). "Imaging of the Spontaneous Low Cerebrospinal Fluid Pressure Headache: A Review." Can Assoc Radiol J **71**(2): 174-185.
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