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SIH and/or
POTS?

Disclosures

- No Conflicts of Interest
- **This work is supported by the Considine CSF Leaks Fund**



Stanford
M E D I C I N E

Thank You

- Dr. Schievink, Dr. Maya, and Dr. Louy et al
- Dr. Deline
- Dr. Gray
- The Considine Family

Spinal CSF Leaks have a troubling similarity to POTS

CSF Leak

- Feel worse when upright
- Headache is prominent
- Nausea/vomiting
- Lightheadedness/dizziness
- Brain fog
- Fatigue
- Associated with HDCT
- Understood
- Treatment can be effective

POTS

- Feel worse when upright
- Headache is prominent
- Nausea/vomiting
- Lightheadedness/dizziness
- Brain fog
- Fatigue
- Associated with HDCT
- Mysterious
- Treatment: “learning to cope”

POTS or CSF Leak? One of several patients treated this month...

- She is a 21 Y year old female
- " I have always been hyperflexible...In ballet I used to dislocate my hip all the time. I had to just pop it back in."
- 2014 on distinct day started vomiting every 3 minutes with a 24 hour flu type thing. After that she never really got well.
- Since then she started having head pain and nausea. Initially nausea was the dominating symptom.



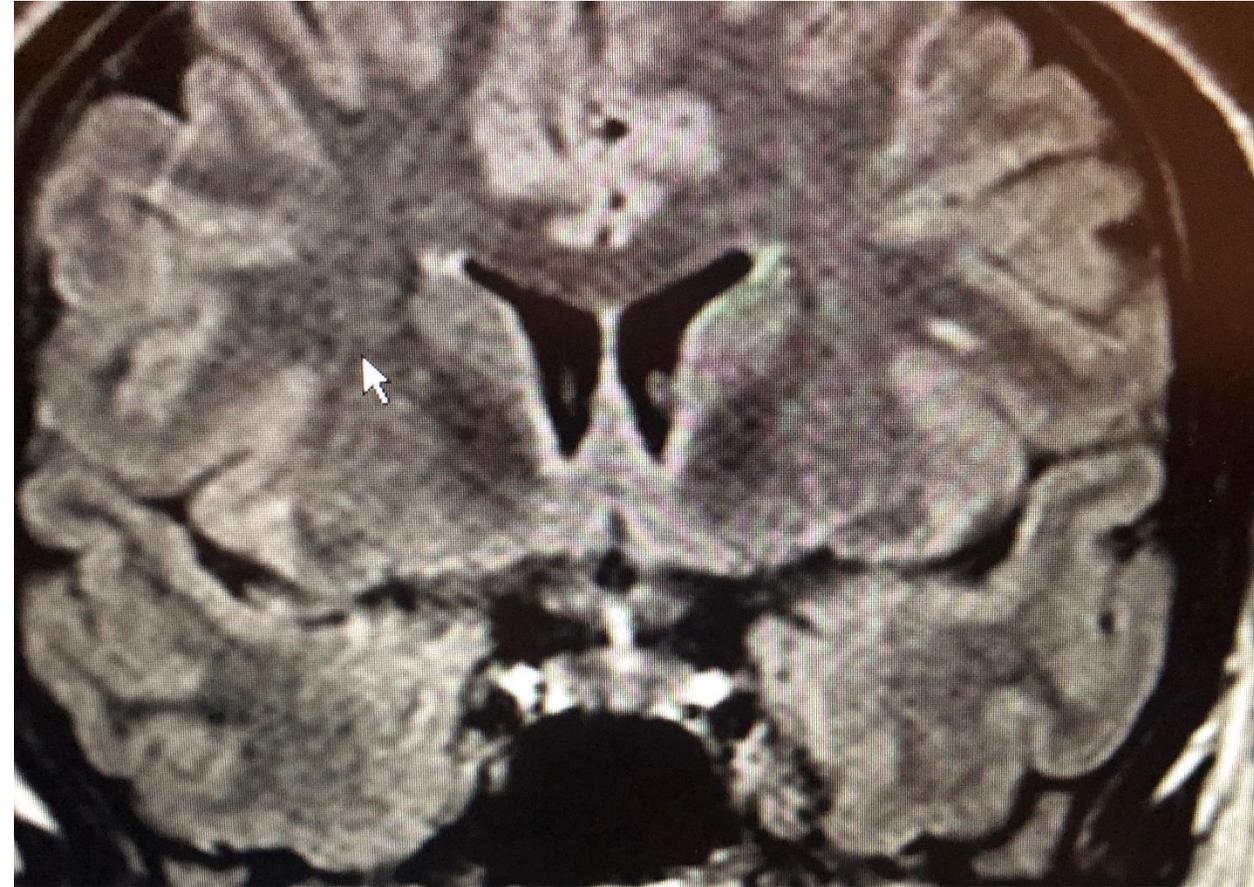
POTS or CSF Leak? Just one patient treated this month...

- Never has headache in the morning. They get progressively worse as the day goes on.
- Notes cognitive problems late in the day. This impacted late day test scores.
- Taking a nap at noon helps.



POTS or CSF Leak? Just one patient treated this month...

- She was noted shortly after this to also have a high resting heart rate
- **Has tilt-table test which is positive and she is diagnosed with POTS**



Doesn't the abnormal
tilt table test mean
POTS in the cause?

Does abnormal autonomic testing rule out CSF leak?

- As of June 2018 pubmed search yields

**no
publications**



("Tilt-Table Test"[Mesh]) AND "Intracranial Hypotension"[Mesh] and/or ("Hereditary Sensory and Autonomic Neuropathies"[Mesh]) AND "Intracranial Hypotension"[Mesh]

The Worlds Literature on Tachycardia in CSF Leaks

- **Tachycardia-** ("Tachycardia"[Mesh]) AND "Intracranial Hypotension"[Mesh] pubmed search yields one paper:

[\(Mokri and Low 2003\)](#)

As of June 2018 no papers have looked at the presence or absence of orthostatic tachycardia in CSF leaks.

Tachycardia

- **4 cases with orthostatic headache referred for suspected CSF leak. All thought to have had CSF leak ruled out, but**
 - 3 cases had no spine MRI
 - 2 cases had no CT myelogram
 - None had digital subtraction myelogram
 - 2 had documented temporary benefit from epidural blood patching.
- **All had orthostatic tachycardia so were diagnosed with POTS not CSF leak!**

POTS or CSF Leak? Just one patient treated this month...

- She did a 24 hour flat test and had no headache at all that day which is highly unusual for her. "I have a headache every day".
- Previous headache interventions tried include: verapamil, Topamax, amitriptyline, propranolol; maxalt and sumatriptan
- None of the headache medications were helpful. Sumatriptan gave her tachycardia up to 200 bpm.





Next Steps:

- **Structured Questionnaire**
- “Neuroimaging results, lab results, opening pressure, and clinical symptoms and history reviewed in conference with Dr. HindeyehDr. Hindeyeh and Dr. Carroll both voted to move ahead with 1 epidural patch at T5-6 and L5-S1. If no results then we will get a CT myelogram”

POTS or CSF Leak? Just one patient treated this month...

- **2/18- epidural blood patch with catheter at left paramedian T6-7 with catheter threaded to right ventral T5-6 and L5-S1:**
 - **Was much better for 10 days. For the first 2 days she had some high pressure symptoms. Then had recurrence of symptoms. There has been some persistent benefit as well with increased ability to walk (eg 6-7 miles per day at disneyland)**
- **4/18- bilateral L5-S1 paramedian with catheter to L4-5 epidural blood patch- no benefit.**

- **5/24/18- CT myelogram-** L2-3 right paramedian approach; csf total protein nl at 42; Dilated opacified left nerve root at C5-6 series 301 im 189. At C7-T1 there is a left dorsal osteophyte contacting the dorsal dura without displacing it. T7-8 left facet osteophyte with slight prominence of proximal nerve root (ic1). Left T12 nerve root is lightly broad and has minor opacification (ic2); Left t10-11 foraminal osteophyte; Left L4-5 ventral hazy epidural area may have some contrast in it. (ic3)

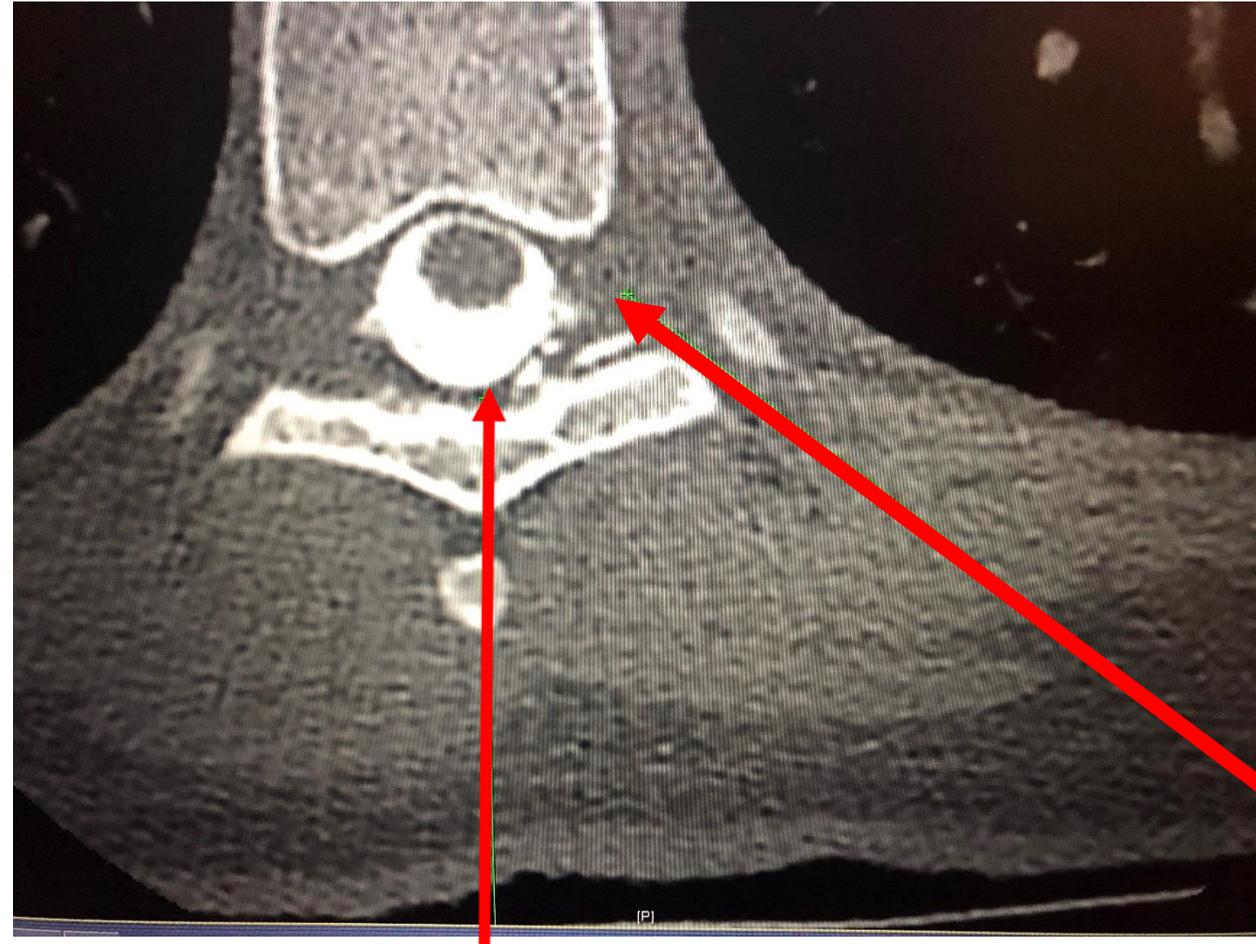
- Left Paramedian facet osteophyte T7-8
- Looks **TOTALLY** benign

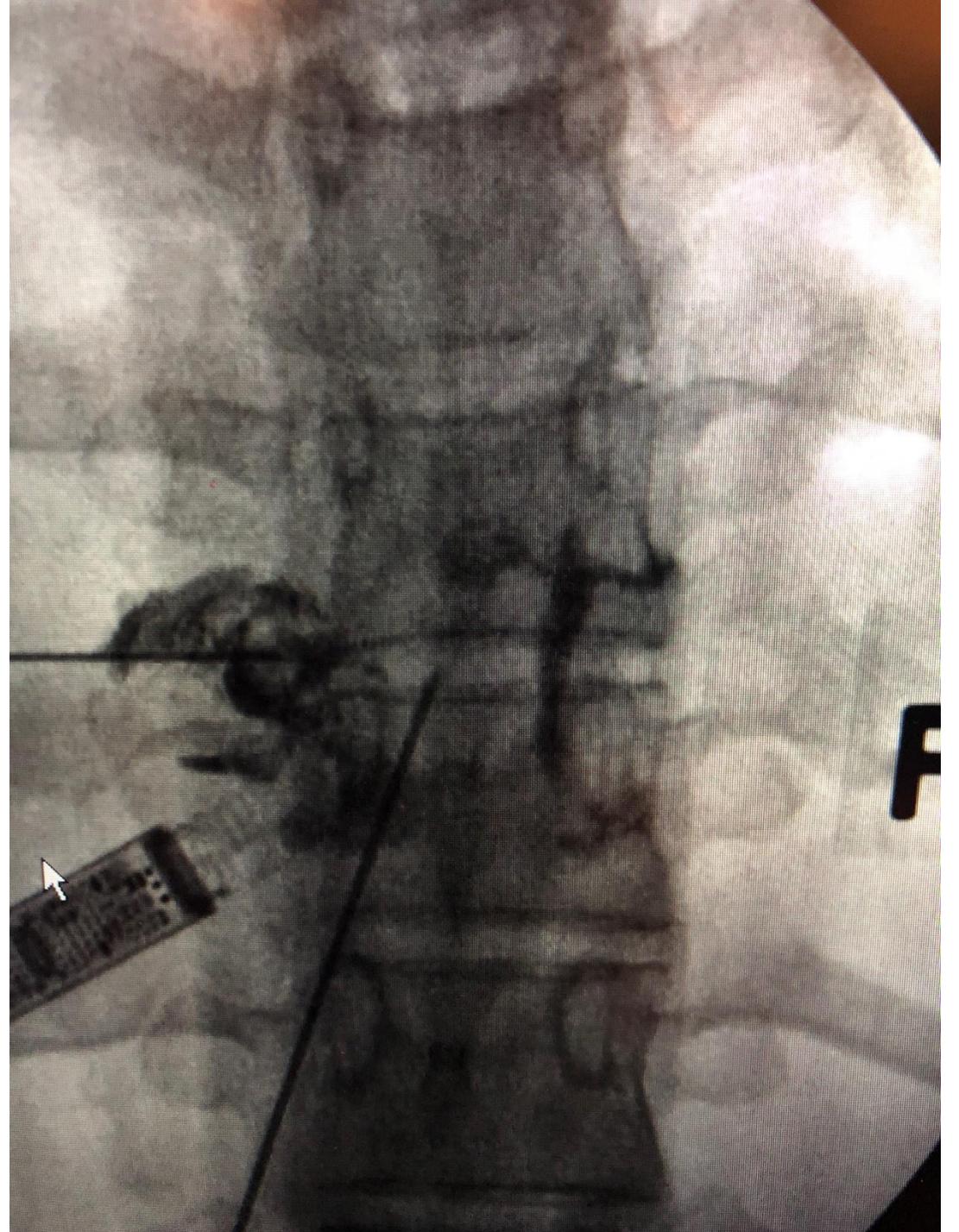


- 5/18- T7-8 left paramedian; L2-3 right paramedian; Left T12-L1 paramedian; Left L4-5 transforaminal epidural fibrin patch:
 - 3 weeks of relief with some rebound intracranial hypertension
- 7/18- T7-8 left paramedian; Left T12-L1 paramedian epidural fibrin patch:
 - developed rebound hypertension for about 10 days then her leak symptoms returned. Despite return of symptoms cognitively, and from fatigue standpoint she is much better than before the patches. However she continues to have head pain and other csf leak symptoms.

Plan to Patch just one level at a time to determine which level is responsible for change.

- 9/18- T7-8 left paramedian and transforaminal Fibrin epidural patch.

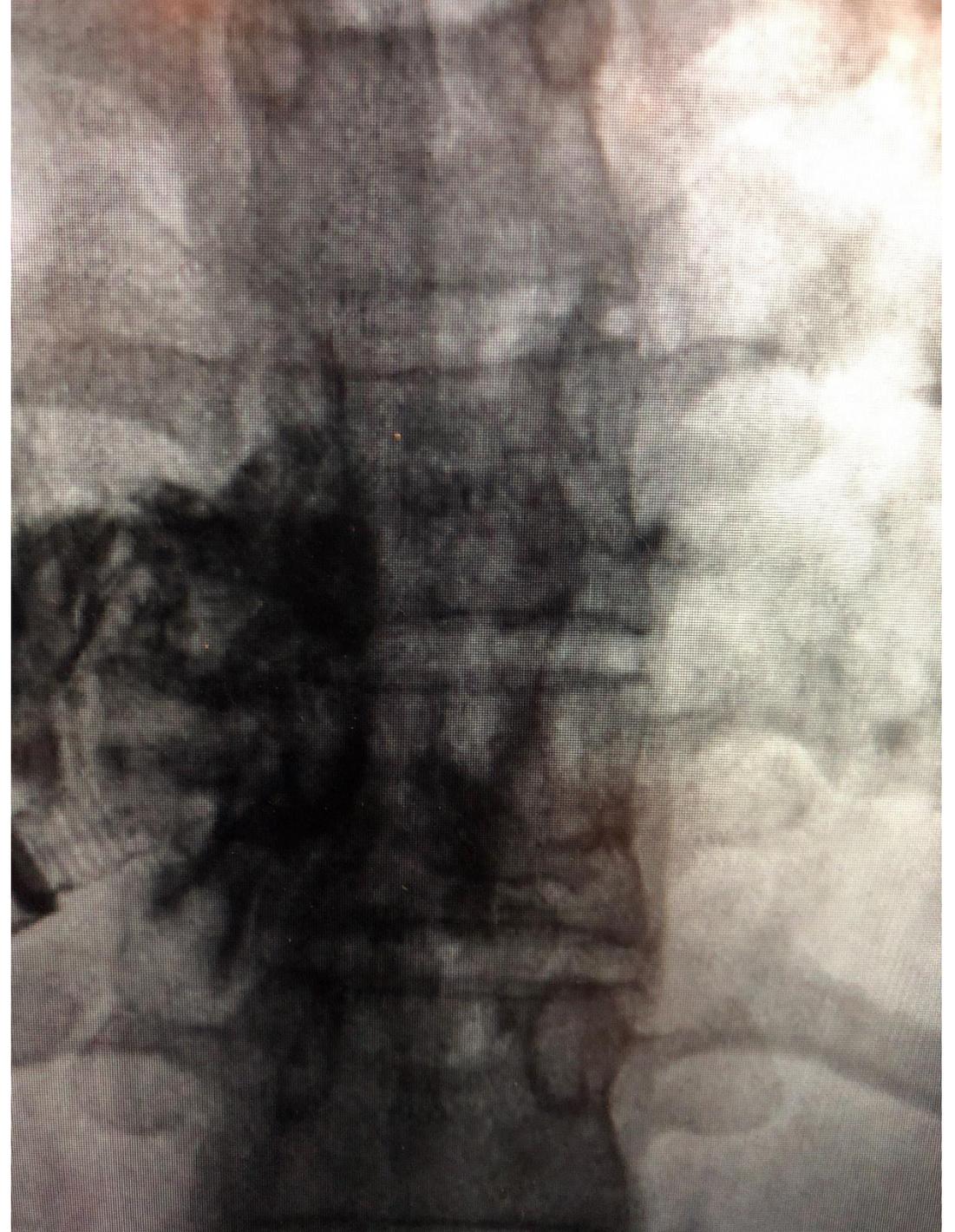




- **DSA of transforaminal T7-8 fibrin injection**



- 9/18- T7-8 left paramedian and transforaminal Fibrin epidural patch.
 - immediate rebound intracranial hypertension TREATED WITH Acetazolamide and Amiloride
 - followed by marked clinical improvement
- Remember:
 - **4/18- bilateral L5-S1 paramedian with catheter to L4-5 epidural blood patch- no benefit.**



2 Cases of Tachycardia following Iatrogenic
CSF leak at Stanford

45 y/o Man with Neuropathic Chest Pain

- 4/30/09- Pulse 86
- 5/8/09- Pulse 81
- 7/30/09 Spinal cord stim placement at T8-9 and T9-10 with 12 ga dural puncture
- 7/31/09- **Pulse 98** post op then Lumbar puncture L3-4 with 22 quincke
- 8/1/2009 – **Pulse 104**: Epidural Blood Patch T8-9 20 CC
- 8/2/09- **Pulse 130-150**
- 8/3/09- **Pulse 121**: repeat L3-4 and T9-10 thoracic epidural patch:
- 8/5/09- per medicine note: "HR better controlled, mainly 80s to 90s, not above 100s."
- 10/1/09- pulse 84

17 y/o with iatrogenic CSF Leak and postural tachycardia

- 2/14/2012- foot surgery and CRPS
- 4/5/2012- Pulse 71
- 4/19/2012- Pulse 82 -Lumbar sympathetic block
- 5/3/2012- Pulse 90- Lumbar sympathetic block
- 5/31/2012- **pulse 120- new onset nausea and tachycardia after second lumbar sympathetic block**
- 7/6/2012- **KNOWN 12 ga dural puncture during scs trial**
- 7/7/2012- pulse 108 Postural Headache
- 7/8/2012- pulse 79 (staying flat)
- 7/13/2012- **7/13/12- Epidural blood patch Lumbar L3/4 14 cc blood**
- 7/15/2012 **pulse 73- while in bed in hospital**
- 7/16/2012 **pulse 116 when upright and discharge to clinic**

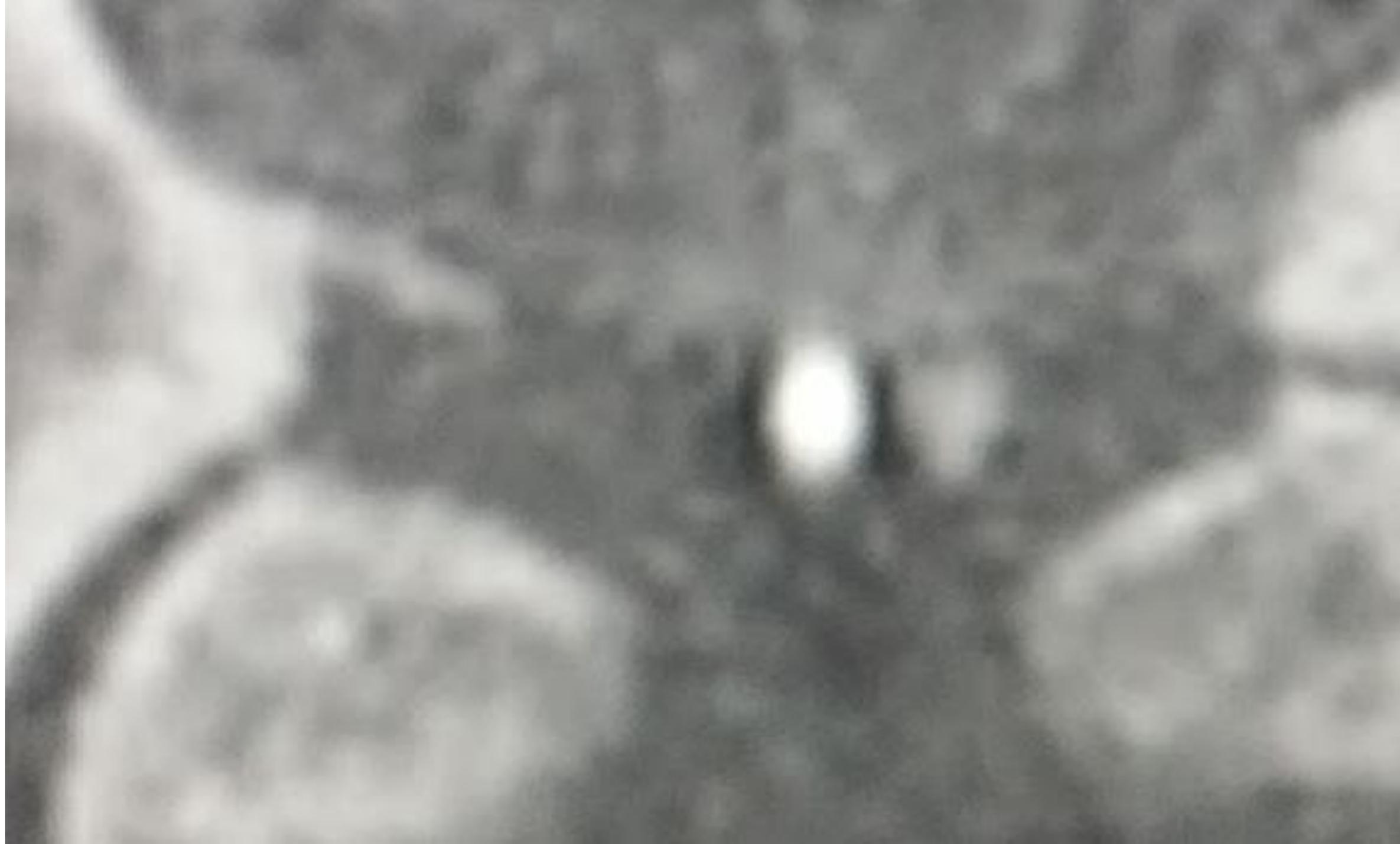
17 y/o with iatrogenic CSF Leak and postural tachycardia

- 8/23/2012 pulse 125
- 9/6/2012 pulse 120
- 9/21/12- pulse 122
- 10/4/12- pulse 123
- 10/18/12- pulse 120
- 11/8/2012- ER visit due to palpitations
- 11/29/2012- 121
- 2/2013 Holter: Pulse 45 flat; 145 upright
- 3/21/13- pulse 132
- 4/2013- **Stim implant with repeat KNOWN 12 ga dural puncture**
- 4/4/2013- caudal epidural with catheter to L3-4 epidural blood patch 20 ml
- 4/18/2013 pulse 103
- 5/9/ 2013- pulse 120
- 6/27/2013- pulse 109
- 7/26/2013- pulse 123
- 8/7/2013- pulse 102
- 8/23/2013- pulse 109
- 12/19/2013- pulse 128
- 6/19/2014- pulse 116
- 8/28/2014- pulse 121
- 1/5/15- pulse 120

17 y/o with iatrogenic CSF Leak and postural tachycardia

- 1/15/15- **caudal epidural blood patch (post procedure immediately hr 81-89)- headache and tachycardia resolves.**
- **nausea, visual disturbance, neck pain, balance problems remain. Headache eventually returns but not tachycardia.**
- 1/22/15- pulse 99
- 2/05/15- pulse 84
- 3/06/2015- pulse 85
- 4/09/2015- pulse 92
- 5/4/2015- pulse 90
- 6/21/16- Ct myelogram: Tiny contrast filling outpouching within the left paramedian dorsal thecal sac at the upper L3 level.
- 6/30/16- Lumbar laminectomy (L1-2) for closure of presumed dural tear. "As expected based on her CT myelogram, a 3-4 mm arachnoid bleb/dural opening was seen just to the left of the lead at the level of the L2 pars"
- 10/12/18- pulse 91





Can Imaging distinguish CSF leak from POTS?

Sensitivity of MRI of the spine compared with CT myelography in orthostatic headache with CSF leak

■ Ref 2

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ABSTRACT

Objective: To investigate the sensitivity of MRI of the spine compared with CT myelography (CTM) in detecting CSF leaks.

Methods: Between July 1998 and October 2010, 12 patients with orthostatic headache and a CTM-confirmed spinal CSF leak underwent an MRI of the spine with and without contrast. Using CTM as the gold standard, we retrospectively investigated the sensitivity of spinal MRI in detecting a CSF leak.

Results: Eleven of 12 patients with a CSF leak documented by CTM also had extradural fluid collections on spinal MRI (sensitivity 91.7%). Six patients with extradural fluid collections on spinal MRI also had spinal dural enhancement.

Conclusion: When compared with the gold standard of CTM, MRI of the spine appears to be a sensitive and less invasive imaging modality for detecting a spinal CSF leak, suggesting that MRI of the spine should be the imaging modality of first choice for the detection of spinal CSF leaks.

Neurology® 2013;81:1789-1792

Methods: Between July 1998 and October 2010, 12 patients with orthostatic headache and a CTM-confirmed spinal CSF leak underwent an MRI of the spine with and without contrast. Using CTM as the gold standard, we retrospectively investigated the sensitivity of spinal MRI in detecting a CSF leak.

Results: Eleven of 12 patients with a CSF leak documented by CTM also had extradural fluid collections on spinal MRI (sensitivity 91.7%). Six patients with extradural fluid collections on spinal MRI also had spinal dural enhancement.

Correlation vs sensitivity

(Hosoya et al. 2013)
2008-2011, 11 hospitals

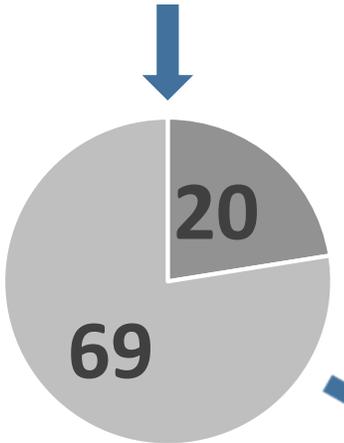
100 Prospectively gathered
orthostatic headaches

89 Brain MRI

89 Radionuclide
cisternogram

70 axial T2
Spine MRI

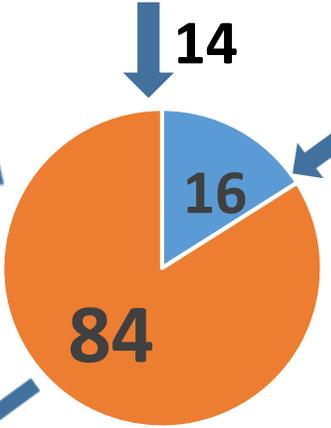
86 MR
Myelogram



■ Positive ■ Negative

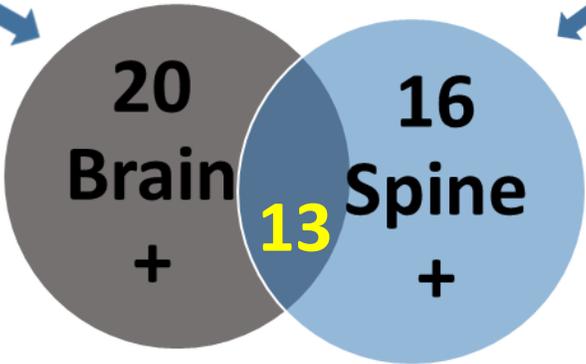
How many of 69 were false
negative results?

?



How many of 84
were false negative?

■ Positive ■ Negative



**7/20 (35%) of MRI proven
SIH missed on multiple
spine imaging!**

The First Stanford Hypothesis

- The **specificity** of all existing neuroimaging for CSF leak is **high**, but the **sensitivity is low**.
- **le Test-/True+** may be 80% with imaging missing most leaks

The Second Stanford Hypothesis

- Patients with imaging-negative CSF Leaks/SIH will be given stereotyped alternative diagnoses.

We have seen CSF Leaks diagnosed as

- Chronic Migraine, Chronic Daily headache, New Daily Persistent headache
 - Chiari Malformation
 - POTS
 - Chronic Fatigue Syndrome
 - Traumatic Brain Injury
 - Post-Concussion Syndrome
-
- Except POTS none of these should improve with prolonged flatness

What to do in the face of ambiguity?

- “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).”
- “...Complete relief of symptoms may not be achieved until two or more EBPs have been performed.”



IHS CLASSIFICATION ICHD-3





A Call to Service

