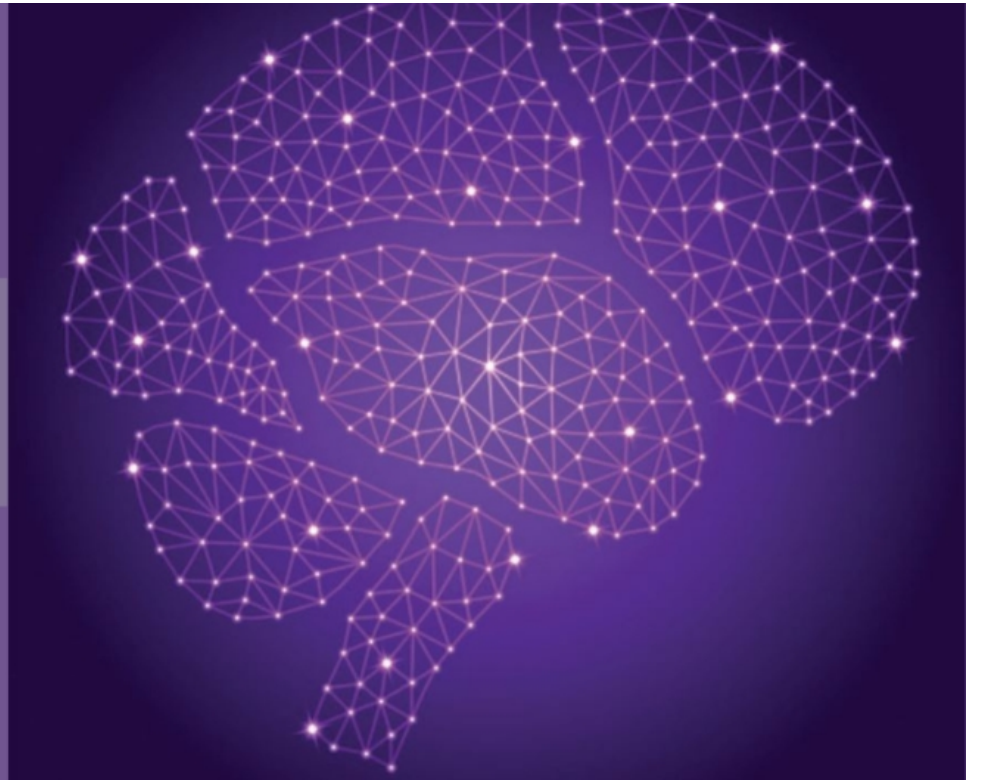


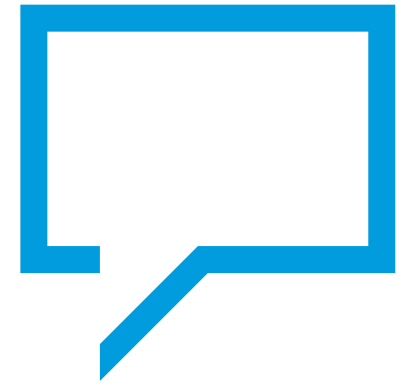
4th Annual Cedars-Sinai
Intracranial Hypotension Symposium





Diagnosis of SIH

The challenging but critical first step



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Cedars-Sinai Intracranial Hypotension Symposium
October 2, 2021

Disclosures

No Relevant Financial Relationships

No Off Label/Investigational Usage

Learning Objectives

Describe the evolution of understanding low CSF pressure syndrome

Recognize limitations of current diagnostic criteria for SIH

Outline a systematic approach to spinal CSF leak diagnosis



Neurosurgery

John Atkinson, MD
Rick Marsh, MD
David Piepgras, MD

Neuro- Ophthalmology

Tariq Bhatti, MD
John Chen, MD, PhD

Anesthesia / Pain Medicine

Neuro- Radiology

John Benson, MD
Waleed Brinjikji, MD
Carrie Carr, MD
Felix Diehn, MD
DK Kim, MD
Greta Liebo, MD
Patrick Luetmer, MD

Neurology



Bahram Mokri, MD
Ivan Garza, MD
Narayan Kissoon, MD
Melissa Payne, RN
Mark Whealy, MD

Ajay Madhavan, MD
Jay Morris, MD
Pearse Morris, MB, BCh
Michael Oien, MD
Courtney Ross, PA-C
Darya Shaplak, MD
Jared Verdoorn, MD

Mokri's landmark description of diffuse pachymeningeal enhancement in low-pressure headache

30th Anniversary

P232. Meningeal Gadolinium Enhancement in Low Pressure Headaches

Babram Mokri, Bruce R. Krueger, Gary M. Miller, and David G. Piepgras, Rochester, MN

Three patients had low pressure headaches and meningeal gadolinium enhancement on magnetic resonance imaging (MRI). The first patient had been treated for a left cavernous sinus meningioma and had a ventriculoperitoneal shunt that had caused low cerebrospinal fluid (CSF) pressure. The second and third patients had history of minor or trivial head trauma without a detectable source of cerebrospinal fluid (CSF) leak. All were asymptomatic when supine, and had headaches when upright. The second and third patients also had nausea in this position. Lumbar CSF pressure was barely

measurable in all. CSF protein was mild to moderately elevated, sugar was normal, cultures were negative, VDRL and FTA-Abs were nonreactive, and there were no blasts or malignant cells in CSF. Gadolinium-enhanced MRI in all three patients showed extensive, diffuse, and thick enhancement of the meninges. Possibility of meningeal carcinomatosis had been raised in all patients. For the first patient, shunt revision was recommended. In the second patient, who also showed small bilateral subdural fluid collections, symptoms and MRI abnormalities have improved. In the third patient, both the symptoms and the MRI abnormalities have resolved completely. Low pressure headaches are another cause of diffuse meningeal enhancement that has not been previously reported and should be included in the imaging differential diagnosis.



Describe the evolution of understanding low CSF pressure syndrome



Recognize limitations of current diagnostic criteria for SIH



Outline a systematic approach to spinal CSF leak diagnosis

Historical Considerations

- 1891** Quincke introduces **lumbar puncture**
- 1898** Bier describes **post-dural puncture headaches**
- 1938** Schaltenbrand coins term “**aliquorrhoea**” for clinical syndrome with very low CSF opening pressures
- 1940** Woltman links orthostatic headache with **spontaneous low CSF pressure**, which “may resemble post-puncture headache”
- 1991** Mokri and Sable report association of orthostatic headache with **diffuse pachymeningeal gadolinium enhancement**
- 1993** Fishman and Dillon show **reversible brain sag**



Spontaneous Cerebrospinal Fluid Leaks: From Intracranial Hypotension to Cerebrospinal Fluid Hypovolemia—Evolution of a Concept

BAHRAM MOKRI, MD

Mayo Clin Proc. 1999;74:1113-1123

Clinical-Imaging Syndrome	Headache	Meningeal Enhancement	Low CSF Pressure	Brain Sag
Classic form	+	+	+	+/-
Normal pressure	+	+	-	+/-
Normal meninges	+	-	+	+/-
Acephalgic form	-	+	+	+/-

Contemporary Advances

- 1999** Mokri introduces **low volume** interpretation of spinal CSF leak
- 2003** Luetmer and Mokri describe **hyperdynamic CT myelography** for localizing fast CSF leaks
- 2007** Kumar links extradural fluid collections to **superficial siderosis**
- 2014** Schievink et al. report first cases of **CSF-venous fistula**
- 2019** Dobrocky et al. describe **brain MRI scoring system** for SIH
- 2021** Brinjikji et al. pioneer **transvenous embolization** of CVF

Spontaneous Spinal CSF Leak

Also known as...

- Aliquorrhea, low CSF pressure headache, CSF hypovolemia, CSF volume depletion, spontaneous intracranial hypotension

Distinct from...

- Post-dural puncture headache
- Skull base leak with CSF rhinorrhea/otorrhea or encephalocele

ICD-10 Intracranial Hypotension & CSF Leak Codes

The most important NEW codes, effective October 1, 2020:

G96.811	Intracranial hypotension, spontaneous
G96.819	Other intracranial hypotension
G96.02	Spinal cerebrospinal fluid leak, spontaneous
G96.09	Other spinal cerebrospinal fluid leak
R51.0	Headache with orthostatic component, not elsewhere classified

* There are additional relevant codes

Describe the evolution of understanding low CSF pressure syndrome

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Outline a systematic approach to spinal CSF leak diagnosis

Spontaneous Intracranial Hypotension

2011 Expert Diagnostic Criteria

Orthostatic headache
and ≥ 1 of...

- A. Demonstration of active spinal CSF leakage
- B. Low CSF opening pressure (< 60 mmH₂O)
- C. Cranial MRI showing brain sag or pachymeningeal enhancement
- D. Sustained improvement of symptoms after epidural blood patching

Not all patients have headache

$\geq 50\%$ of patients have normal opening pressure

Sensitivity $\leq 80\%$

Strong placebo effect

Spontaneous Intracranial Hypotension

2018 ICHD-3 Diagnostic Criteria

Not all patients have headache

≥50% of patients have normal opening pressure

Sensitivity ≤80%

Spontaneous Intracranial Hypotension

Schievink Modified Diagnostic Criteria

Sensitivity $\leq 80\%$

$\geq 50\%$ of patients have normal opening pressure



Describe the evolution of understanding low CSF pressure syndrome



Recognize limitations of current diagnostic criteria for SIH



Outline a systematic approach to spinal CSF leak diagnosis

Diagnosis of spinal CSF leak requires sequentially answering 3 questions

1. Is there an active CSF leak?

2. Is the leak fast, slow, or fistula?

3. Where is the leak?

40 y/o ♀

- New daily persistent headache
- Described as intense bifronto-occipital pressure
- Worse when upright or with Valsalva maneuvers
- Accompanied by orthostatic nausea and distorted hearing
- Joint hypermobility (Beighton score 4/9)
- Neuro exam otherwise normal

Does this patient have an active spinal CSF leak?

-
- Common symptoms of SIH
 - Risk factors for SIH
 - Cranial MRI signs of SIH

Common Symptoms of Spinal CSF Leak

Clinical Manifestations	Comments
Orthostatic headache	Orthostatic pattern may be lost over time
Nausea/vomiting	Typically orthostatic
Neck, interscapular > low back pain	Site of pain does not necessarily correspond to site of leak
Diplopia/visual symptoms	Unilateral or bilateral 6 th palsy (often) 3 rd , 4 th , or multiple cranial nerve palsies (less often)
Cochleovestibular symptoms	Tinnitus Muffled, echoed, or distorted hearing Nystagmus Dizziness (vertigo, lightheadedness, wooziness)

Does this patient have an active spinal CSF leak?

-
- Common symptoms of SIH
 - Risk factors for SIH
 - Cranial MRI signs of SIH

Risk Factors

- Joint hypermobility
- Connective tissue disorders
 - Marfan syndrome
 - Ehlers-Danlos syndromes
- Disc osteophytes
- Intracranial hypertension
- Bariatric surgery
- Trauma, often minor

Beighton Hypermobility Score

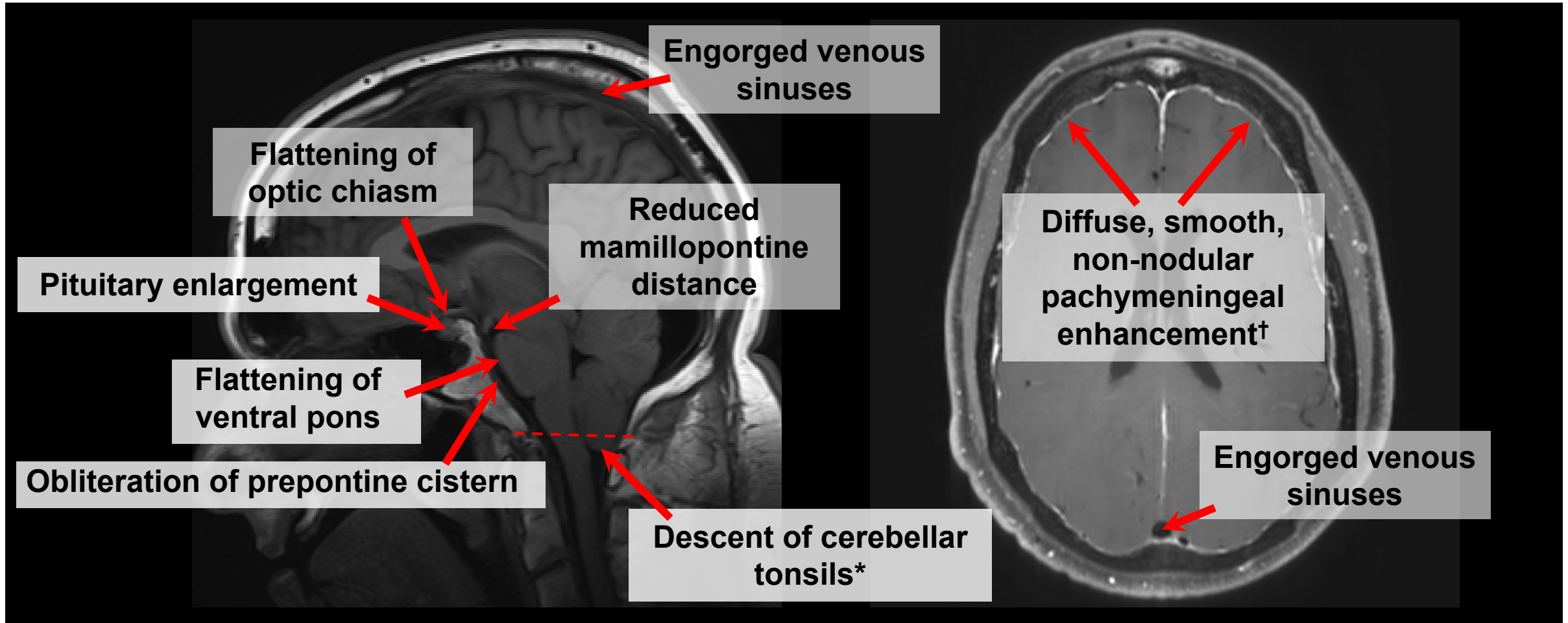


≥5/9 hypermobility syndrome

Does this patient have an active spinal CSF leak?

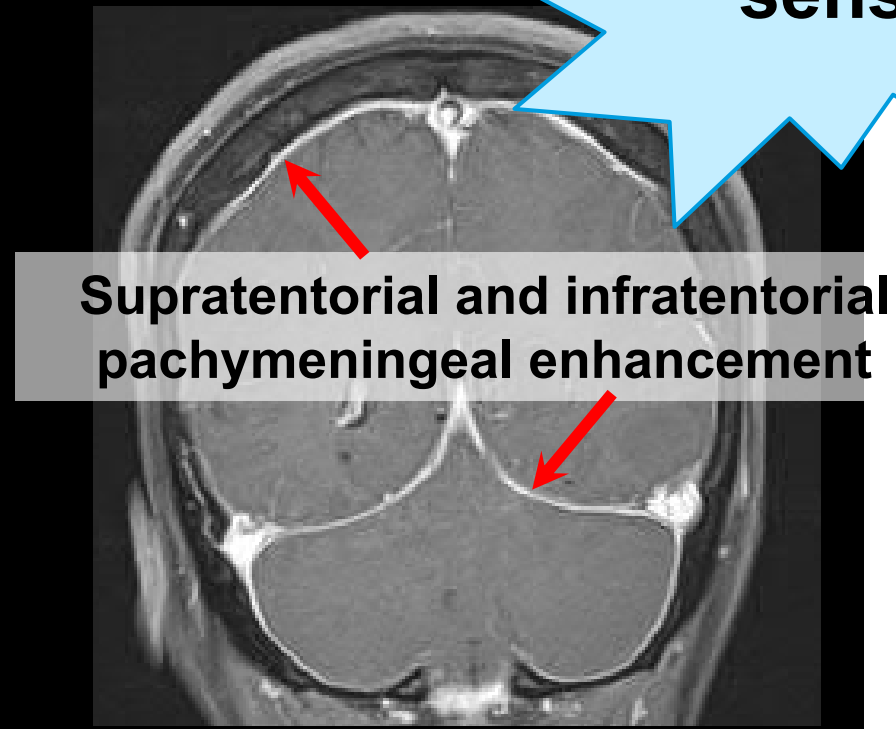
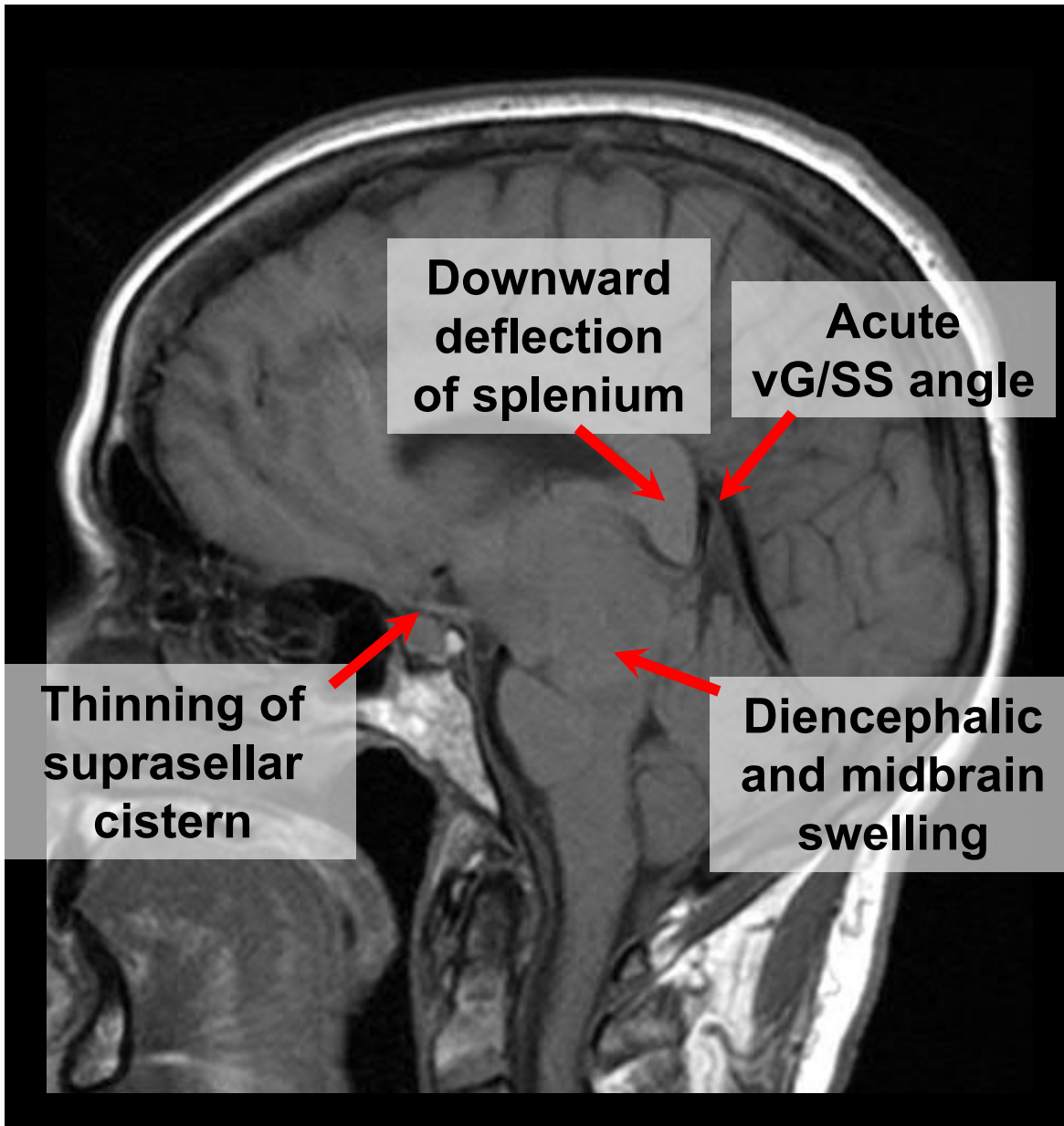
-
- Common symptoms of SIH
 - Risk factors for SIH
 - Cranial MRI signs of SIH

MRI Head with Gadolinium Contrast



* Low lying cerebellar tonsils is least specific sign, also seen in idiopathic intracranial hypertension and Chiari I

† Pachymeningeal enhancement may resolve spontaneously despite persistent CSF leak



**MRI head
≤80%
sensitive**

Does this patient have an active spinal CSF leak?

-
- Common symptoms of SIH
 - Risk factors for SIH
 - Cranial MRI signs of SIH

Assessing Spinal Cerebrospinal Fluid Leaks in Spontaneous Intracranial Hypotension With a Scoring System Based on Brain Magnetic Resonance Imaging Findings

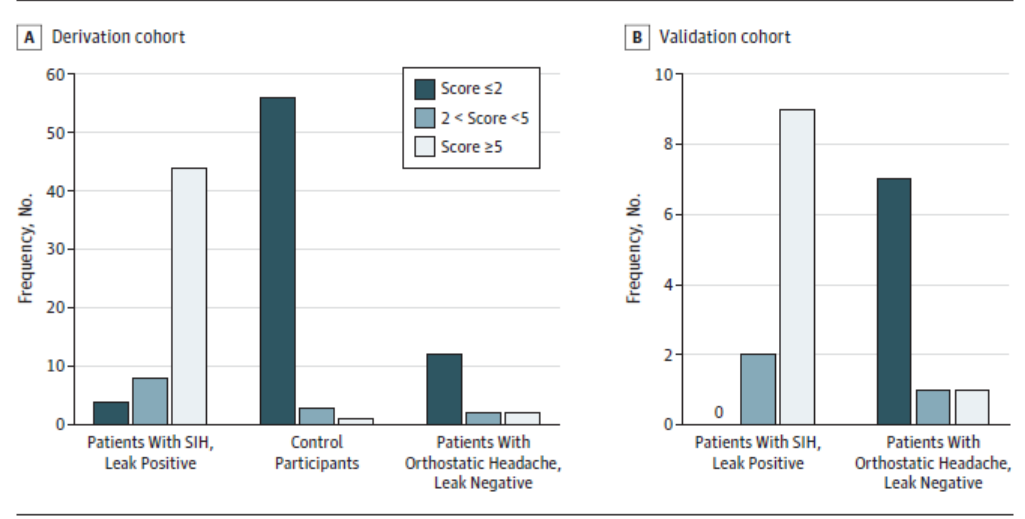
Tomas Dobrocky, MD; Lorenz Grunder, MD; Philippe S. Breiding, MD; Mattia Branca, MSc; Andreas Limacher, PhD; Pascal J. Mosimann, MD; Pasquale Mordasini, MSc; Felix Zibold, MD; Levin Haeni, MD; Christopher M. Jesse, MD; Christian Fung, MD; Andreas Raabe, MD; Christian T. Ulrich, MD; Jan Gralla, MSc; Jürgen Beck, MD; Eike I. Piechowiak, MD

Not a diagnostic score, rather a predictor of subsequent spinal myelography findings

Table 2. Six Imaging Signs in the Final Diagnostic

Characteristic	Were Included	Score Points
Engorgement venous sinus	1	2
Pachymeningeal enhancement	1	2
Subdural fluid collection	1	1
Suprasellar cistern ^a	1	2
Prepontine cistern ^b	1	1
Mamillopontine distance ^c	1	1

^a ≤4 mm.
^b ≤5 mm.
^c ≤6.5 mm.



Mayo cohort (n=62) who underwent DSM

- Bern score ≥5: leak found in 67%
- Bern score 3-4: 45%
- Bern score ≤2: no leaks found

41 y/o ♀

Monthly migraines became daily orthostatic headaches

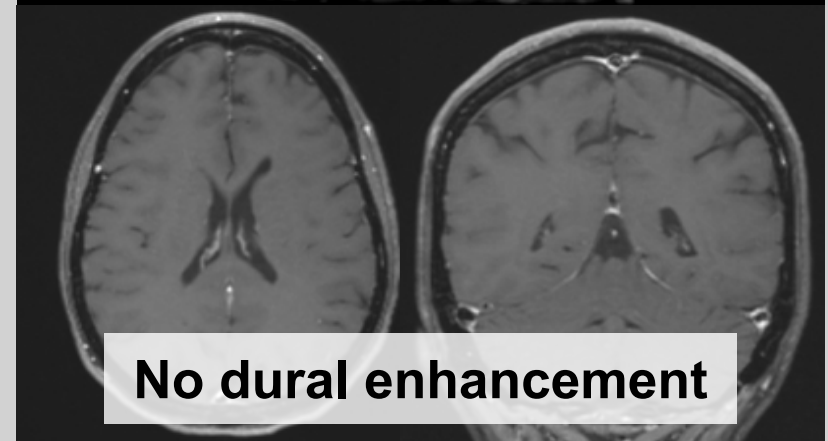
Hearing echoes, seeing black spots

Worse with bending forward, Valsalva, late in day

Better with supine rest of 1-2 hr even without sleep



No brain sag



No dural enhancement

Does this patient have an active spinal CSF leak?

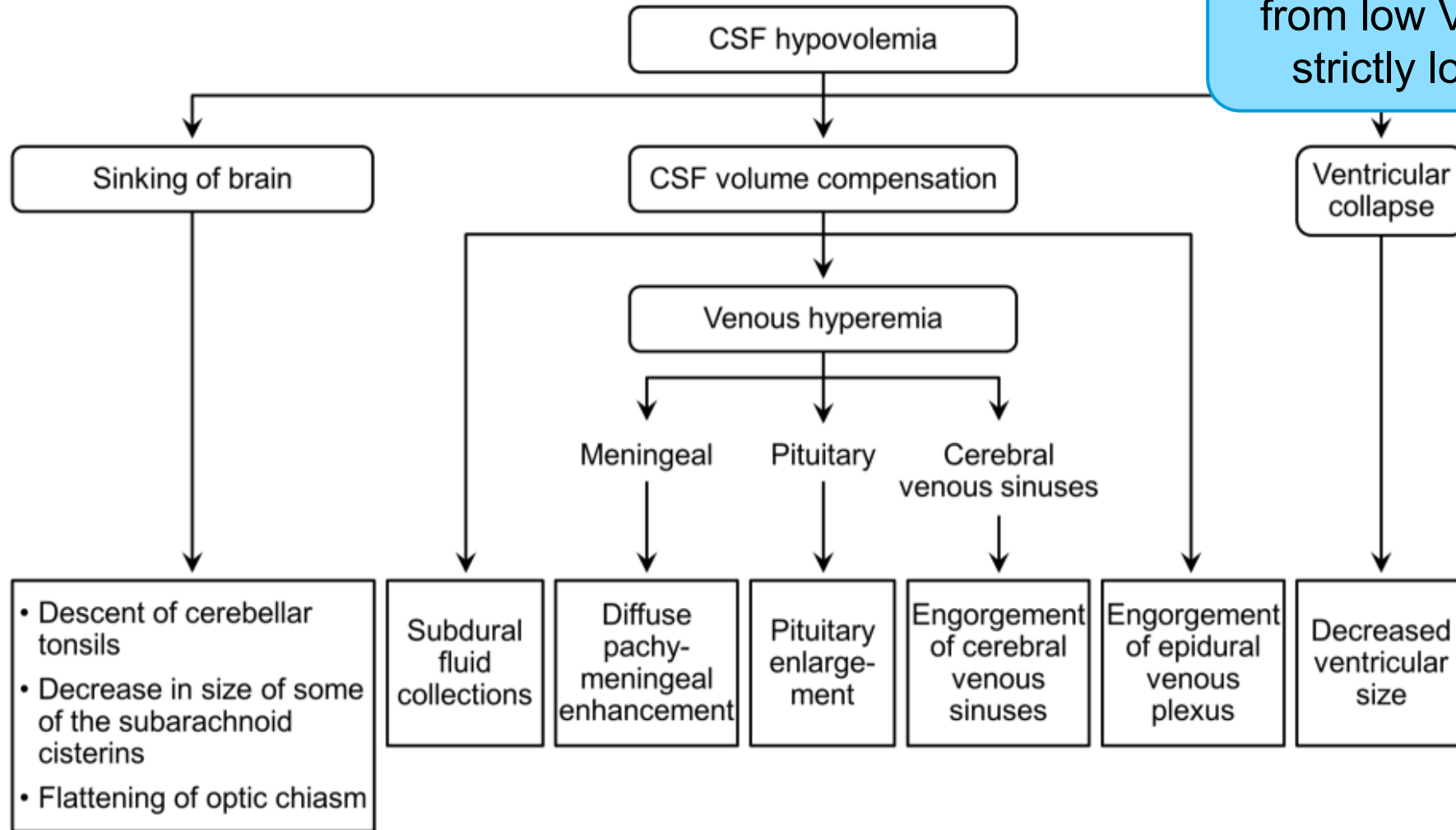
-
- Common symptoms of SIH
 - Risk factors for SIH
 - Cranial MRI signs of SIH

Monro-Kellie Doctrine

- Assumption: Cranial compartment is incompressible and the volume inside the cranium is fixed
- The cranium contains brain tissue, blood, and CSF...
- In a state of volume equilibrium...
- Any increase in volume of one constituent must be compensated by a decrease in volume of another...
- And therefore, any decrease in volume of one constituent must be compensated by an increase in volume of another

Mechanisms of MRI abnormalities in SIH

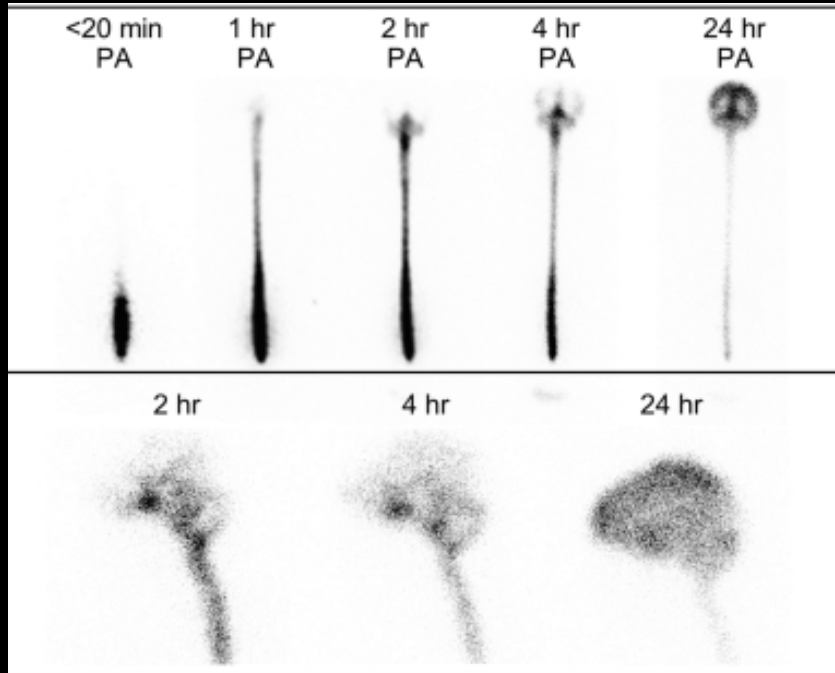
All of these changes result from low VOLUME, not strictly low pressure



Radioisotope Cisternogram

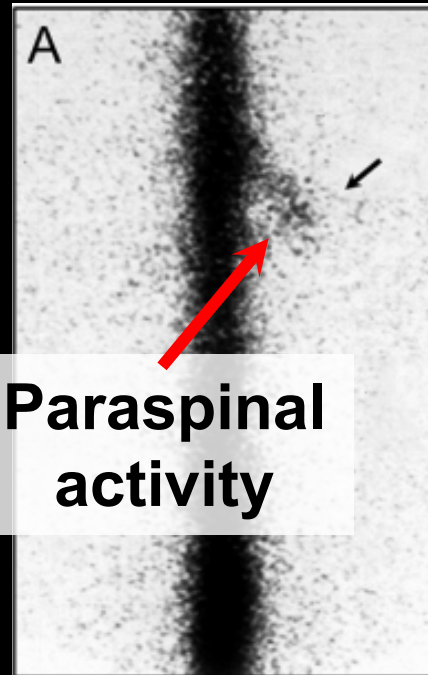
Assess for abnormal CSF FLOW

Cisternogram
~100% sensitive
(for localizable leaks)

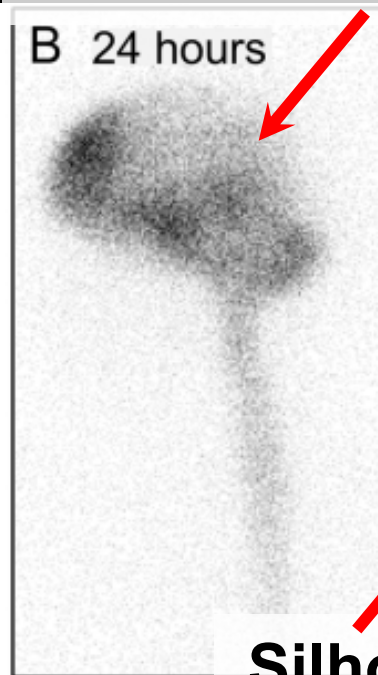


Normal

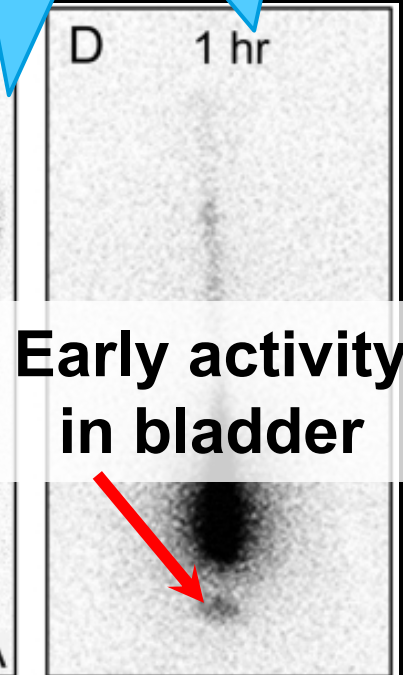
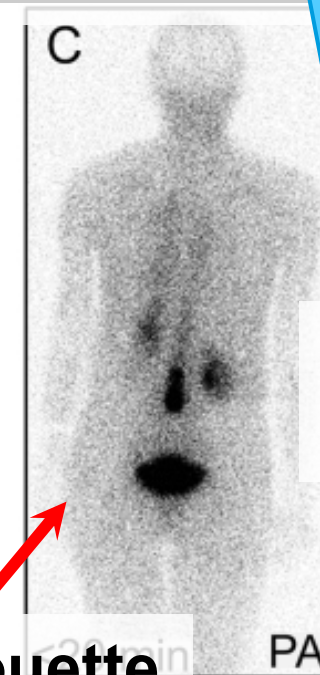
Reduced activity over cerebral convexities



Paraspinal activity



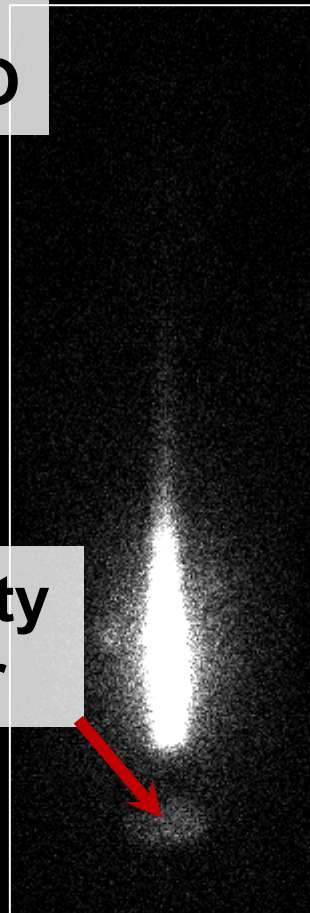
Silhouette sign



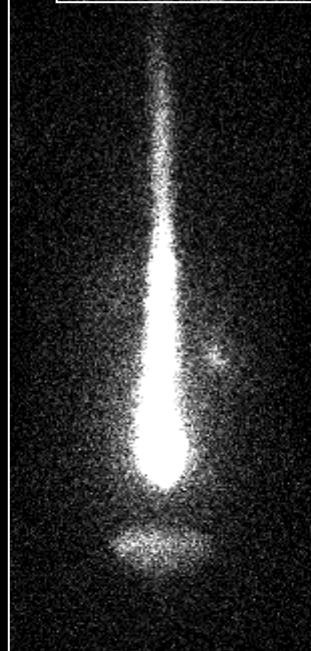
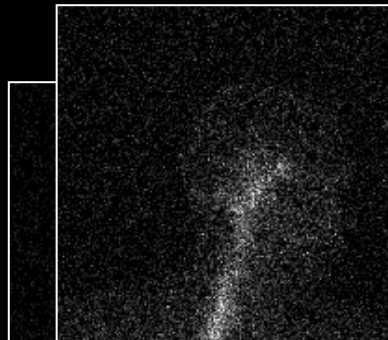
Early activity in bladder

**Opening
pressure
190 mmH₂O**

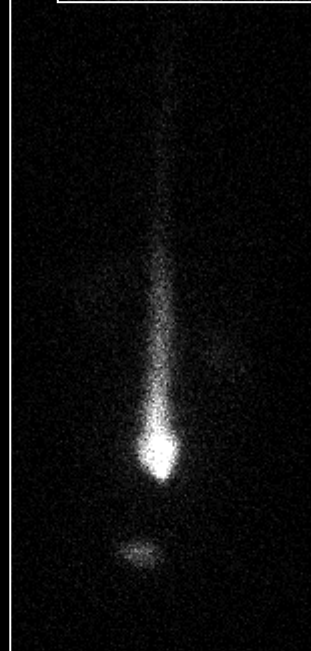
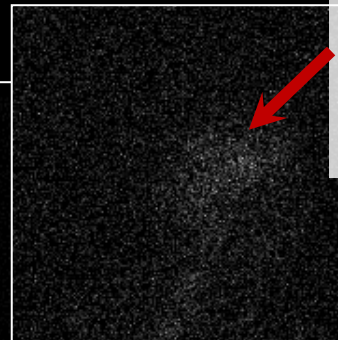
**Early activity
in bladder**



1 hr

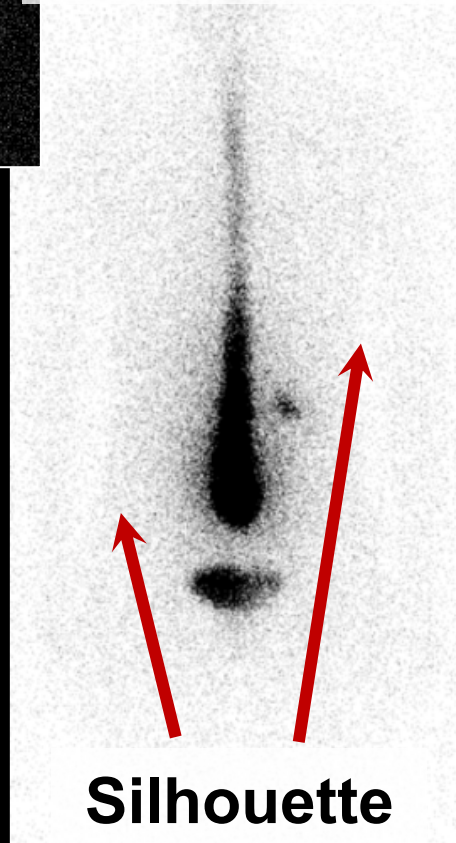


6 hr



24 hr

**Reduced activity over
cerebral convexities
at 24 hr**



**Silhouette
sign**

Does this patient have an active spinal CSF leak?

**MRI head
≤80%
sensitive**

- Common symptoms of SIH
- Risk factors for SIH
- Cranial MRI signs of SIH
- Low opening pressure
- Cisternographic signs of SIH

**Cisternogram
~100% sensitive**

Diagnosis of SIH

DIRECT EVIDENCE

- Observation of CSF escaping thecal sac at surgery
- Observation of contrast escaping thecal sac on myelogram
- Extradural CSF on spinal MRI

INDIRECT EVIDENCE

- Dural enhancement, venous engorgement, brain sag, subdural fluid collections on brain MRI
- LP opening pressure <60 mm H₂O
- LP OP reduced from prior measurement
- Hyperdense paraspinal vein on myelogram
- Reduced tracer over cerebral convexities at 24 hr on cisternography
- Tracer activity in bladder before 4 hr on cisternography
- Post-myelogram renal contrast
- Response to epidural patching

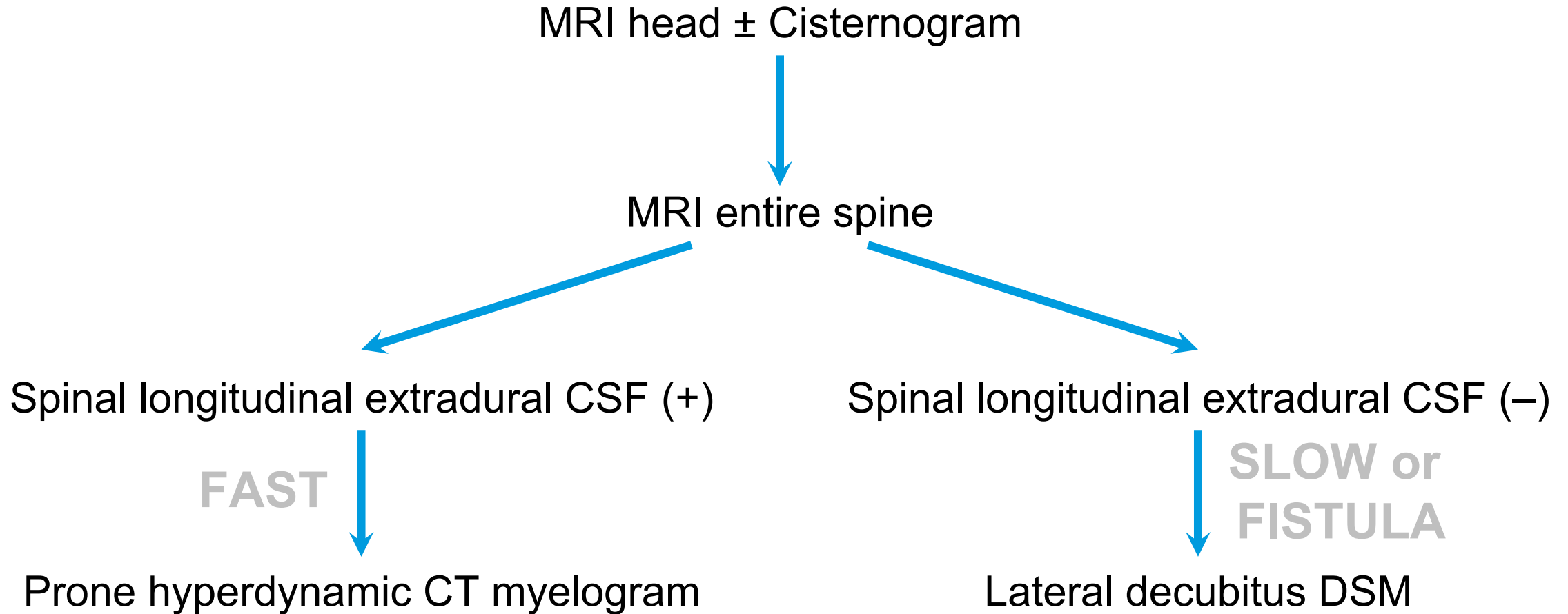
Diagnosis of spinal CSF leak requires sequentially answering 3 questions

1. Is there an active CSF leak?








2. Is the leak fast, slow, or fistula?

3. Where is the leak?

Mayo Clinic Diagnostic Algorithm for Suspected Spinal CSF Leaks



Spontaneous Intracranial Hypotension: A Systematic Imaging Approach for CSF Leak Localization and Management Based on MRI and Digital Subtraction Myelography

 R.I. Farb,  P.J. Nicholson,  P.W. Peng,  E.M. Massicotte,  C. Lay,  T. Krings, and  K.G. terBrugge

Spinal longitudinal extradural CSF (+)

Spinal longitudinal extradural CSF (-)

↓
Prone DSM

↓
If (-)

↓
Lateral decubitus DSM





Describe the evolution of understanding low CSF pressure syndrome



Recognize limitations of current diagnostic criteria for SIH



Outline a systematic approach to spinal CSF leak diagnosis



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