


# Orthostatic Headaches: Differential Diagnosis & Diagnostic Criteria of SIH

David W. Dodick, MD  
Professor  
Department of Neurology  
Mayo Clinic  
Phoenix Arizona



# Orthostatic Headache

- 1. Does the patient have a CSF leak?**
    - a. Clinical features**
    - b. Diagnostic criteria**
  
  - 2. What else could present with OH in the absence of CSF leak?**
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# The Syndrome of Spontaneous Low CSF Pressure (SIH) or Low CSF Volume (CSF Hypovolemia)

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- **Orthostatic headache is invariably due to SIH**
- **SIH almost always a result from spontaneous CSF leak**
- Spontaneous CSF leaks **almost always at the level of spine**



## Brief Communication

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### Diagnostic Criteria for Headache Due to Spontaneous Intracranial Hypotension: A Perspective

Wouter I. Schievink, MD; David W. Dodick, MD; Bahram Mokri, MD; Stephen Silberstein, MD;  
Marie-Germaine Boussier, MD; Peter J. Goadsby, MD, PhD


- A. Orthostatic headache
- B. The presence of **at least one** of the following:
  1. Low opening pressure (60 mm H<sub>2</sub>O)
  2. Sustained improvement of symptoms after epidural blood patching
  3. Demonstration of an active spinal CSF leak
  4. Cranial MRI changes of intracranial hypotension (e.g. brain sagging, dural enhancement)
- C. No recent history of dural puncture
- D. Not attributable to another disorder

## 7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure

**Description:** OH caused by low CSF pressure or CSF leak, usually accompanied by neck pain, tinnitus, changes in hearing, photophobia, nausea. It remits after normalization of CSF pressure or successful sealing of the CSF leak.

### Diagnostic criteria:

- A. Any headache<sup>1</sup> fulfilling criterion C
- B. Either or both of the following:
  1. **low cerebrospinal fluid (CSF) pressure (<60mm CSF)**
  2. **evidence of CSF leakage on imaging**
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.



Imaging diagnostic procedure of choice; LP rarely needed

# Headache attributed to spontaneous intracranial hypotension (ICHD-3:7.2.3)

Orthostatic headache caused by low CSF pressure of spontaneous origin. It is usually accompanied by neck stiffness and subjective hearing symptoms. It remits after normalization of CSF pressure.

Diagnostic criteria:

- A. Headache fulfilling criteria for 7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure , and criterion C below
- B. Absence of a procedure or trauma known to be able to cause CSF leakage
- C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage, or has led to its discovery
- D. D. Not better accounted for by another ICHD-3 diagnosis.

## Notes from ICHD-3

- “Patients with S-CSF leaks should be **screened for connective tissue and vascular abnormalities**”
- “. . . may show delayed response to postural change . . . **orthostatic features may become less obvious over time**”
- “In patients with typical OH and no apparent cause (after exclusion of POTS) lumbar EBP is reasonable”
- “It is not clear that all patients with 7.2.3 Headache attributed to SIH 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”

# OH-CSF leak

## Variability in Clinical Phenotype of Headache

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- **OH** (present in upright relieved in supine position)
- **OH evolving to non-orthostatic** daily headache (NODH)
- **NODH**
- **NODH evolving to OH** (sometimes preceded by **neck or interscapular pain**)
- **Exertional, cough, Valsalva-induced**
- **Thunderclap**
- **“Second-half-of-day”** headaches (often with some orthostatic features)
- **Paradoxical OH** (relieved when upright)
- **Intermittent headaches** from intermittent CSF leak
- **No headache “acephalgic form”**



# Most common symptoms



**Bilateral, occipital**



**Neck pain, stiffness**

**Associated symptoms (mimics migraine)**



Adapted from Mokri B. Neurology 2015;21(4):1086–1108  
Schievnik WI. JAMA. 2006;295:2286-2296  
Schwedt TJ, Dodick DW. Curr Pain Headache Rep 2007 Feb;11(1):56-61.

# Other Non-Headache Manifestations of Spontaneous CSF Leaks

Manifestations	Comments
Spine pain	Neck and/or interscapular pain (common), low back pain (much less common). Don't expect the level of the pain to match the site of the leak
Cochleovestibular manifestations	<ul style="list-style-type: none"> <li>• Tinnitus, non-pulsatile</li> <li>• Change in hearing (muffled, distant, echoed, distorted, decreased)</li> <li>• Dizziness (vertigo, lightheaded feeling, wooziness)</li> </ul> Rarely Meniere's-like picture
Diplopia/visual symptoms	Unilateral or bilateral 6th CN palsy more often than 3rd or 4th or multiple CN palsies. Visual blurring, field defects
Cognitive/behavioral changes	<ul style="list-style-type: none"> <li>• Memory difficulties</li> <li>• Personality change, less organized, can't get things done or complete projects, less tidy, inappropriate behavior, FTD-like picture</li> </ul>
Gait unsteadiness	Regardless of presence or absence of abnormal head sensation
Movement disorders	Chorea, Parkinsonism, tremor
Uncommon or rare manifestations	<ul style="list-style-type: none"> <li>• Numbness/paresthesias (face posterior head, upper limbs)</li> <li>• Radicular upper extremity manifestations</li> <li>• Encephalopathy, stupor, coma</li> <li>• Trouble with sphincter control</li> <li>• Galactorrhea</li> <li>• Visual field cut</li> <li>• Bibrachial amyotrophic</li> </ul>

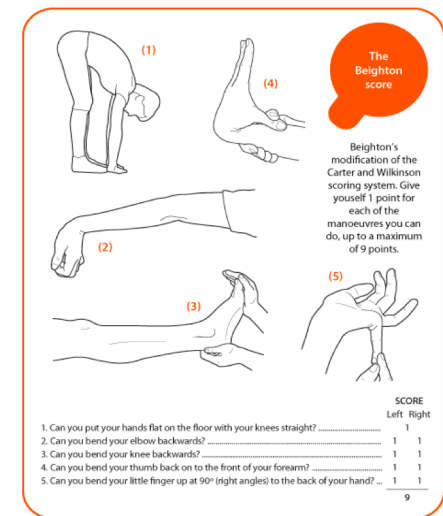
# Etiology of Spontaneous CSF Leaks and IH

## Connective Tissue Disorders

- Marfan syndrome
- Ehlers-Danlos syndrome type II
- Autosomal dominant PCKD
- Spontaneous retinal detachment
- Arterial dissections, aneurysms or nonrheumatic valvular heart disease
- Isolated skeletal features of Marfan syndrome (20%)\*\*
- Joint hypermobility (40%)

## Trauma

- Bony spurs, herniated intervertebral discs
- Injury (MVA, sports injuries, Valsalva)



PCKD=polycystic kidney disease

\*\*Skeletal features of Marfan (tall stature, arachnodactyly, high arched palate, and joint hypermobility)

# Differential diagnosis SIH/Orthostatic Headache

## SIH

- Meningeal diverticula ( $\geq 8\text{mm}$ )
- Dural ectasia (complex diverticula)
- Post-craniotomy (occipital decompression)
- Spinal CSF-leak (DSM)
- CSF leak (“Indeterminate type”) without demonstrated CSF leak

## OH without IH

- Postural orthostatic tachycardia syndrome (POTS)
- Headache with radiographic mimics (Chiari, inflammatory/granulomatous pachymeningitis, subdural hematoma)
- Other primary/secondary headache disorders that worsen with changes in posture/activity

# Complications

Imaging in Headache Medicine

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## Recurrent brainstem infarction caused by spontaneous intracranial hypotension

Benjamin Matosevic, Manuela Prieschl, Gerhard Luef, Michael Knoflach, Christoph Schmidauer, Johann Willeit and Peter Lackner

**JNS** SPINE

CASE REPORT  
J Neurosurg Spine 26:751–753, 2017

## Superficial siderosis of the central nervous system associated with intraspinal hemorrhage from ventral thoracic epidural veins and a ventral spinal CSF leak: case report

Keisuke Takai, MD, PhD,<sup>1</sup> Takashi Komori, MD, PhD,<sup>2</sup> Manabu Niimura, MD, PhD,<sup>1</sup> and Makoto Taniguchi, MD, PhD<sup>1</sup>

Medical Principles  
and Practice

### Case Report

Med Princ Pract 2012;21:392–394  
DOI: [10.1159/000336782](https://doi.org/10.1159/000336782)

Received: August 16, 2011  
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## Cerebral Venous Sinus Thrombosis Associated with Spontaneous Intermittent Cerebrospinal Fluid Rhinorrhea: A Case Report

Uygar Utku<sup>a</sup> Mustafa Gokce<sup>a</sup> Mehmet Senoglu<sup>b</sup>

Departments of <sup>a</sup>Neurology and <sup>b</sup>Neurosurgery, Faculty of Medicine, Kahramanmaraş Sutcu Imam University, Kahramanmaraş, Turkey

Brief Report

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## Diffuse non-aneurysmal SAH in spontaneous intracranial hypotension: Sequela of ventral CSF leak?

Wouter I Schievink<sup>1</sup> and M Marcel Maya<sup>2</sup>


# Spontaneous Intracranial Hypotension Imaging Features



- SEEPS & COPS

- 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

