

# Differential Diagnosis of Orthostatic Headache

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# Disclosures

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# Differential Diagnosis of Orthostatic Headache

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## Outline

- Terms: What is orthostasis, orthostatic hypotension, orthostatic headache
- What can gravity do to the head and neck
- What is dural traction and why does it hurt
- Non-SIH causes of orthostatic HA
- Diagnostic definitions of SIH – what matters; lingering questions about diagnosis
- Conclusions

# Terms

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Orthostasis – Maintenance of an upright standing posture

Orthostatic hypotension (postural hypotension) – a drop by at least 20 mm Hg or diastolic blood pressure of at least 10 mm Hg when a person assumes a standing position.

Why does OSH happen? Either significant hypovolemia OR defective autonomic reflexes to control **venous pooling** and reduced cardiac return upon standing.

# Terms

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Orthostatic headache (aka postural headache)—headache that occurs or is made much worse by standing and remaining standing.

In PDPHA the HA occurs within 20 sec of standing and generally resolves within 20 sec of assuming recumbent position\*

\*Vilming ST, Kloster R. Post-lumbar puncture headache: Clinical features and suggestions for diagnostic criteria. *Cephalalgia*. 1997;17:778-784.

# Why might orthostatic headache occur?

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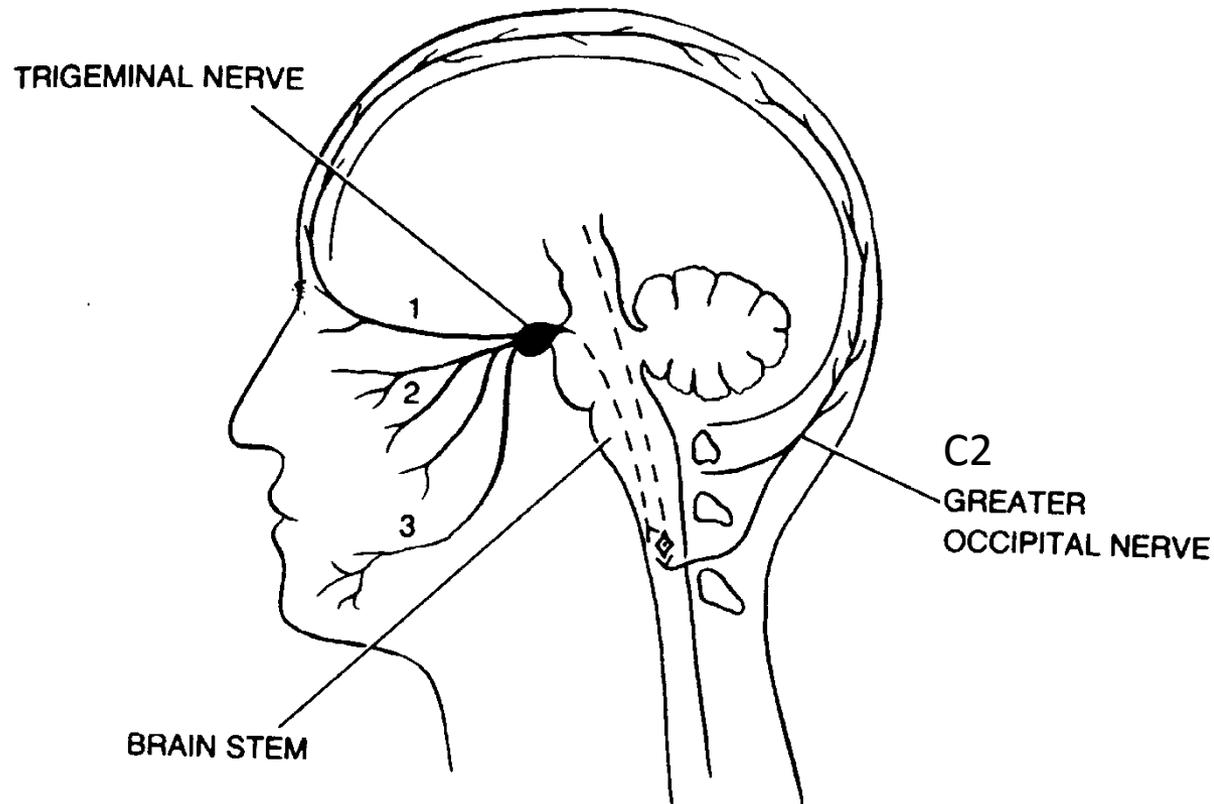
- Sudden decrease in already low intracranial CSF volume → reduced brain buoyancy → skull contents shift downward, stretching the dura
- Sudden dilation of cerebral arteries in an attempt to maintain intracranial volume constant (Monro-Kellie doctrine)
- A large enough intracranial lesion that a sudden shift in position might lead to brain contents shift (mass, hematoma)
- A sinus, orbital, pericranial or cervical lesion which stimulates nociceptors on movement might lead to referred HA

# Head Pain – referral patterns

Intracran supratentorial and anterior head – V

Infratentorial and posterior head, cervicocranial – C2-3

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Head Pain – referral patterns explain non cranial causes of head pain

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Sinus disease

Spinal disease

Ocular disease

All of which can be triggered mechanically

# Differential diagnosis of postural headaches

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- Decreased CSF pressure
- Normal ICP with another cause of movement induced perceived head pain

# Differential diagnosis of postural headaches

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## Decreased CSF / intracranial pressure

Traumatic CSF leak - trauma, diagnostic LP, accidental dural puncture during spinal anesthesia, spinal or cranial surgery

Dural tear from spondylosis or disc herniation (rare)

Weakness of dural sac (meningeal diverticula, abnormalities of connective tissue (with tear or fluid collection triggered by trivial trauma?))

CSF-venous fistulas

CSF shunt overdrainage

Hypovolemia

# 7.2 Headache attributed to low cerebrospinal fluid pressure

- 7.2.1 Post-dural puncture headache
- 7.2.2 CSF fistula headache (leak)
- 7.2.3 Headache attributed to spontaneous intracranial hypotension

## 7.2 Headache attributed to low cerebrospinal fluid pressure

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### ***Description:***

Orthostatic HA in the presence of low CSF pressure (either spontaneous or secondary), or CSF leakage, usually accompanied by neck pain, tinnitus, changes in hearing, photophobia and/or nausea. It remits after normalization of CSF pressure or successful sealing of the CSF leak.

### ***Diagnostic criteria:***

- A. Any headache fulfilling criterion C
- B. Low CSF pressure (<60 mm CSF) and/or 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-III diagnosis.

## 7.2 Headache attributed to low cerebrospinal fluid pressure

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### ***Comment:***

*7.2 Headache attributed to low cerebrospinal fluid pressure* is usually but not invariably orthostatic. Headache that significantly worsens soon after sitting upright or standing and/or improves after lying horizontally is likely to be caused by low CSF pressure, but this cannot be relied upon as a diagnostic criterion. Evidence of causation may depend upon onset in temporal relation to the presumed cause together with exclusion of other diagnoses.

## 7.2.1 Post-dural puncture headache

### Post-lumbar puncture headache.

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***Description:***

Headache occurring within 5 days of a LP, caused by CSF leakage through the dural puncture. It is usually accompanied by neck stiffness and/or subjective hearing symptoms. It remits spontaneously within two weeks, or after sealing of the leak with epidural blood patch.

***Diagnostic criteria:***

- A. Any headache fulfilling criterion C
- B. Dural puncture has been performed
- C. Headache has developed within 5 days of the dural puncture
- D. Not better accounted for by another ICHD-III diagnosis.

## 7.2.1 Post-dural puncture headache

### Post-lumbar puncture headache.

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***Comment:***

Independent risk factors for 7.2.1 *Post-dural puncture headache* have recently been demonstrated: female gender, age between 31 and 50 years, a previous history of 7.2.1 *Post-dural puncture headache* and orientation of the needle bevel perpendicular to the long axis of the spinal column at the time of the dural puncture.

## 7.2.2 CSF fistula headache

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Orthostatic HA occurring after a procedure / trauma causing persistent CSF leakage resulting in low ICP. It remits after successful sealing of the leak.

### ***Diagnostic criteria:***

- A. Any headache fulfilling criterion C
- B. Both of the following:
  - 1. a procedure has been performed, or trauma has occurred that can lead to persistent leak
  - 2. low CSF pressure (<60 mm CSF) and/or evidence of low CSF pressure and/or of CSF leakage on MRI, myelography, CT myelography or radionuclide cisternography
- C. Headache has developed in temporal relation to the procedure/trauma
- D. Not better accounted for by another ICHD-III diagnosis.

## 7.2.3 Headache attributed to spontaneous intracranial hypotension

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### ***Description:***

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. Any headache fulfilling criterion C
- B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
- D. Not better accounted for by another ICHD-III diagnosis.

# Postural HA Case 1

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LP performed on a 32 year old obese woman presenting with frequent headaches accompanied by nausea and vomiting, particularly frequent around menses. Fundus exam had revealed papilledema (later found to be benign drusen)

Following LP pt complained of severe holocranial headache worsened with sitting or standing accompanied by severe nausea.

After EBP headache improved.

# Postural HA Case 2

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After micro surgery for removal of a L1 herniated disc, a 56 year old man complained of perirectal pain, incontinence and a new headache, worse with sitting or standing and relieved by sitting.

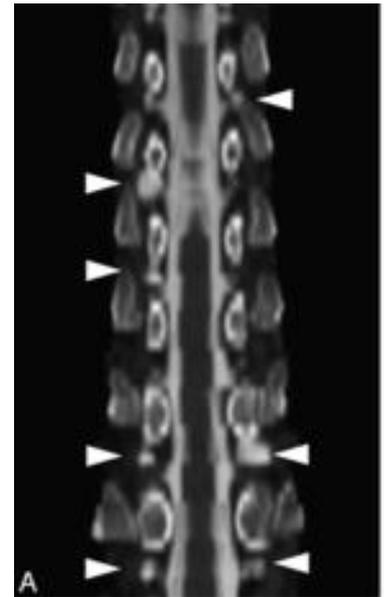
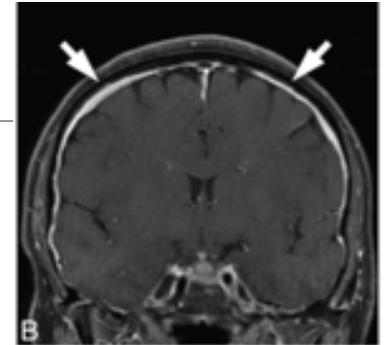
CT myelogram revealed a dural leak at the level of surgery and was repaired with immediate resolution of HA. Perirectal pain, incontinence were felt to be due to conus medullaris trauma and resolved over wks.

# Postural HA Case 3

48 year old tall slender woman with possible EDS complained for several months of frequent headaches generally worse with standing and relieved by laying down flat or at an angle.

Brain MRI was remarkable for 7 mm cerebellar tonsil descent, and diffuse pachymeningeal enhancement. Spinal MRI revealed meningeal diverticulae at multiple levels, but no evident of extrathecal CSF leak.

Multiple EBPs were ineffective.



# Diverticulae cause postural HA?

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Kranz et al Spinal meningeal diverticulae in SIH: Analysis of prevalence myelographic appearance. Am J Neuroradiol 2013 34:1284–89.

“Despite the well-established association between spinal meningeal diverticula and SIH, we found no difference in the prevalence or myelographic appearance of diverticula in patients with SIH compared with controls”

# Differential diagnosis of postural headaches

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## Normal ICP with other cause of movement induced pain

Migraine

Benign exertional headache

Sinusitis induced HA

Meningitis

Pregnancy-induced hypertension (pre-eclampsia)

Cerebral venous thrombosis

Subdural hematoma

Subarachnoid hematoma

Brain tumor

Stroke (ischemic and hemorrhagic)

# ICHD 1.1 Migraine without aura

- A. At least 5 attacks fulfilling criteria B-D
- B. Headache attacks lasting 4-72 h (untreated or unsuccessfully treated)
- C. Headache has  $\geq 2$  of the following characteristics:
  1. unilateral location
  2. pulsating quality
  3. moderate or severe pain intensity
  4. aggravation by or causing avoidance of routine physical activity (eg, walking, climbing stairs)
- D. During headache  $\geq 1$  of the following:
  1. nausea and/or vomiting
  2. photophobia and phonophobia

# ICHD 4.2 Primary exercise HA

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- A. At least 2 headache episodes fulfilling criteria B and C
- B. Brought on by and occurring only during or after strenuous physical exercise
- C. Lasting <48 h
- D. Not better accounted for by another ICHD-3 diagnosis

# Headache due to sinusitis

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“Sinus” headaches are overdiagnosed but can occur in both acute and chronic sinus infections, particularly if sphenoid sinus is involved.

Features included diffuse pain, but particularly anterior, worsened with Valsalva and bending forward, and may be exacerbated by other changes in position.

Hearing changes including tinnitus may occur

Nausea and vertigo may occur

# ICHD 7.4.1 Headache attributed to intracranial neoplasm

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- Space-occupying intracranial neoplasm demonstrated
- $\geq 2$  of:
  - 1.headache has developed in temporal relation to development of the neoplasm, or led to its discovery
  - 2.either or both of: a) headache has significantly worsened in parallel with worsening of the neoplasm; b) headache has significantly improved in temporal relation to successful treatment of the neoplasm
  - 3.headache has  $\geq 1$  of the following 3 characteristics:
    - a)progressive; b) worse in the morning or after daytime napping; c) **aggravated by Valsalva-like manœuvres**

# Headaches due to colloid cyst of the third ventricle

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- Recurrent relatively sudden headaches triggered by a change in position (generally relieved by lying down).
- Sometimes accompanied by reduced level or loss of consciousness
- Headache is the most common presenting symptom

Young, WB and Silberstein, SD. Paroxysmal headache caused by colloid cyst of the third ventricle: case report and review of the literature. *Headache* 1997, 37:15-20

# Chronic Subdural hematoma

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Known to cause intermittent head pain, worse with position change

May produce cognitive decline without clear focal deficits

Traumatic cause may have been trivial

# Postural HA Case 4

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21 year old college student presented with severe HA for 1 day with mild nausea. Seen in the ER crying due to pain unable to sit or stand due to pain intensification. She cannot answer questions or describe the headache or how it started other than to say “Make it stop!”

Exam revealed only meningismus. CT scan was normal. LP revealed Protein of 80, WBC count of 250, 90% lymphocytes, glucose 60. Morphine IV helped to reduce the pain.

# Postural HA Case 5

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34 year old woman transferred from another hospital due to “screaming headaches for the past 1 week”. Only opioid medications have been helpful. On arrival she resisted any movement and remained in the fetal position.

CT of the head was normal by report and LP was likewise normal without any change in her pain.

She denies recent trauma, history of headaches in the past, and any other new symptoms. She restarted OCP 5 weeks ago.

# Clinical features of intracranial hypotension

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- Postural headaches, also worsened by activity and/or Valsalva
- Pain intensity is generally severe but not invariably
- Pain character: burning dull or throbbing, variable locations
- History of a procedure – but pain may start after a lag >2 days (33%)
- Valsalva may worsen pain

# Clinical features of intracranial hypotension

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- Vertigo, tinnitus, decreased hearing may occur (due to perilymph fluid changes)
- Cranial nerve palsies especially diplopia (traction on cranial nerves due to crowding of brainstem)
- Nausea, photophobia, phonophobia
- HA may abate with pressure on abdomen<sup>1</sup>, or when putting pt in Trendellenberg<sup>2</sup>

1. Gutsche BB. Lumbar Epidural analgesia in obstetrics: Taps and patches. In: Reynolds F, ed. *Epidural & Spinal Blockade in Obstetrics*. Oxford: Bailliere Tindall; 1984:95-106.

2. Rozen T, Swidan S, Hamel R, Saper J. Trendelenburg position: A tool to screen for the presence of low CSF pressure syndrome in daily HA pts. *Headache*. 2008;48:1366-1371.

# Radiological features of Intracranial hypotension and their mimics

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**Brain sag** – also seen in Chiari malformations and read incorrectly in normal cases

**Pachymeningeal enhancement** – Also seen in meningitis, meningeal carcinomatosis, post LP, and venous sinus thrombosis

**Bilateral subdural collections** – may be bilateral chronic subdural hematomata

**Enlarged pituitary** – may be a diffusely enhancing tumor or normal for the patient (e.g. postpartum)

# Intracranial Hypotension Ddx

## *Conclusions*

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Strong clues are

- Orthostatic HA (at least initially) with resolution on recumbency
- Accompanying symptoms of hearing change and vertigo as well as possible cranial nerve palsies
- No signs of systemic illness
- No meningismus
- Response to EBP
- Typical brain MRI features

Suspect it after epidural anesth and spine surgery

Be skeptical of the diagnosis in pregnancy and delivery

Obtain a careful head MRI with and without contrast