## **Clinical Mimics of SIH**

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# Disclosures (past year)

Role	Organization	
Advisory Board/Consultant	Allergan/Abbvie, Biohaven Pharmaceuticals/Pfizer, Eli Lilly, Impel, Lundbeck, Optomed, Pfizer, Satsuma, Teva, Theranica, Zosano	
Speaker Bureau	Abbvie, Impel	
Grant support	Spinal CSF Leak Foundation	
Support: Clinical trial site PI	Allergan, Eli Lilly	
Medical Advisory Board	Healthy Women, Spinal CSF Leak Foundation	
Editorial Board	Neurology Reviews, Journal of Neuro-ophthalmology	
Contributing author	Medlink Neurology	
	<b>Bold</b> = relevant to content	

### Post-traumatic Headache

Lifetime prevalence 4.7% in males and 2.4% in females

Causes: TBI, whiplash, craniotomy (headache inversely proportional to extent of trauma)

Relationship of headache onset to trauma is controversial

ICHD-3 specifies 7 days based on consensus, may extend to 3 months ("delayed PTH")

With a pre-existing headache disorder, current headache must change or worsen in close temporal relationship to the injury.

No defined headache frequency or phenotype

30-60% have persistent symptoms 3 months after TBI

Acute: resolves within 3 months

Persistent: present for >3 months after onset (30-50% of cases)

Literature is based primarily on clinic-based studies; 50% do not seek medical care

### ICHD-3 Criteria: Acute Headache Attributed to TBI from Head Injury (5.1)

- Any headache fulfilling and D
- Traumatic injury to the head has occurred
- Headache is report to have developed within 7 days after one of the following:
  - The injury to the head
  - Regaining consciousness following the injury to the head
  - Discontinuation of medication(s) impairing ability to sense or report headache following injury to the head
- Either of the following:
  - Headache has resolved within 3 months after its onset
  - Headache has not yet resolved but 3 months have not yet passed since onset
- Not better accounted for by another ICHD-3 diagnosis
- \* After 3 months, it is considered Persistent Headache Attributed to TBI from Head Injury (5.2)

# Headache Attributed to Mild TBI to the Head (5.1.2, 5.1.4)

- A. Headache fulfilling criteria for 5.1
- B. Injury to the head fulfilling both of the following:
  - 1. Associated with **none** of the following:
    - Loss of consciousness for >30 minutes
    - Glasgow Coma Score <13</li>
    - Post-traumatic amnesia for >24 hours
    - Altered level of awareness for < 24 hours</li>
    - Imaging shows skull fracture, ICH, contusion or other sign of TBI

- 2. Associated with one or more of the following:
  - Transient confusion, disorientation or impaired consciousness
  - Anterograde of retrograde amnesia
- 3. Two or more of the following symptoms suggestive of mild TBI:
  - Nausea
  - Vomiting
  - Visual disturbances
  - Dizziness and/or vertigo
  - Gait and postural imbalance
  - Impaired memory and/or concentration

## Post-Concussion Syndrome

#### Headache

Risk factors:

Female sex

Family and personal history of headache and migraine

History of prior concussion

Mood disorders

Phenotype is migraine-like > Tension-type headache-like >>> Cluster (TAC)-like

Most often bilateral, moderate to severe in intensity

Treat with acute and preventive migraine medications

Cognitive Difficulties

Generally improves all the time but not universally May impede school and job performance

- Sleep Difficulties
- Cervical Injury
- Vestibular and Oculomotor

Vertigo, dizziness

Pursuits, saccades, convergence and accommodation may be affected

Psychological

Depression, anxiety, irritability, emotional lability

Predictive factors of PCS at 3 months: Pre-injury psychiatric history (depression or anxiety), acute post-traumatic stress (~5 days post injury), life stressors, pain, resilience/coping styles

## Whiplash and Whiplash-Associated Disorders

Whiplash: Acceleration-deceleration head movement with flexion and extension of the neck caused by trauma

Incidence 300-600/100,000 in N. America and Western Europe

Following whiplash, headache reported by 60% within 7 days, 23% at 3 months, 30% at 6 months, 38% at 12 months

Motor vehicle accidents (rear-ended) are the most common cause

#1 reason for personal injury compensation claims (and malingering) after traffic accidents

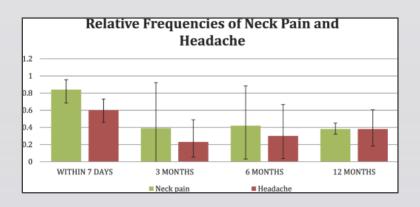
Most common symptoms: Neck pain and headache

Meta analysis:

Neck pain in 84% (95% CI 68-95) within 7 days of injury

Declines to about 40% up to a year out

Headache in 60% (95% CI 45-73) within 7 days of injury



## Whiplash-Associated Disorders

Literature is WEAK and discordant

Headache is more prevalent in those with pre-existing headache

Quebec Task Force classification:

Grade	Neck Symptoms	Physical Signs
0	None	None
I	Pain, stiffness or tenderness	None
II	Pain, stiffness or tenderness	Musculoskeletal signs (decreased ROM, point tenderness
Ш	Pain, stiffness or tenderness	Neurological signs (plus musculoskeletal signs)
IV	Pain, stiffness or tenderness	Fracture or dislocation

## Post-Concussion Syndrome/Mild TBI or Spinal CSF Leak?

	ТВІ	CSF Leak
History of head or neck trauma	Required	Possible
History of surgery	Brain	Spine
Headache	Usually migraine-like	Any phenotype, orthostatic
Neck pain	Common	Common
VIII nerve symptoms	Vestibular	Vestibular, cochlear
Cognitive dysfunction	Yes	Yes
Sleep difficulties	Yes, insomnia	Nocturnal awakening possible
Mood and behavior disorders	Common	Common
Brain MRI	Normal	Usually abnormal

# New Daily Persistent Headache (NDPH)

A **primary** headache disorder

- Distinct and clearly remembered onset, becoming continuous and unremitting within 24 hours
- Present for at least 3 months
- Not attributed to another ICHD-3 diagnosis

May resemble migraine or tension-type headache

Self-limiting and refractory subtypes

SIH is a secondary cause of NPDH

# NDPH or Spinal CSF Leak?

	NDPH	Spinal CSF Leak
Clearly recalled onset	Yes	Often
Thunderclap onset	Possible	Possible
Orthostatic headache	No	Yes
Other neurologic symptoms (vestibular, cognitive, etc.)	No	Yes

## Occipital Neuralgia

Unilateral or bilateral attacks in the distribution of the greater, lesser and/or third occipital nerves

At least 2 of the following:

Recurring paroxysmal attacks lasting seconds to minutes

Severe in intensity

Shooting, stabbing, or sharp in quality

Both of the following:

Dysesthesia and/or allodynia during innocuous stimulation of the scalp or hair

Either of the following:

Tenderness of the affected nerve branches

Trigger points at the emergence of the GON or in the C2 distribution

Pain is eased temporarily by local anesthetic block of the affected nerves

Not accounted for by another ICHD-3 diagnosis

- Pain may sometimes extend to the fronto-orbital area
- Distinguish from referred occipital pain arising from the atlantoaxial or upper zygapophyseal joints or tender trigger points in neck muscles or their insertions

Likely over-diagnosed based on response to anesthetic blockade which is also effective in migraine, cluster headache, cervicogenic headache and post-spinal puncture headache

# Occipital Neuralgia or Spinal CSF Leak?

	Occipital Neuralgia	Spinal CSF Leak
Occipital pain	Yes	Very common
Brief, paroxysmal attacks	Yes	No
Orthostatic pain	No	Yes
Stabbing, shooting, sharp pain	Yes	Infrequent
Neck pain	Possible	Very common
Transient relief with anesthetic block	Yes	Possible
Other neurologic symptoms	No	Yes
Brain imaging	Normal	Usually abnormal
Cervical imaging	Spine abnormal if upper cervical etiology	Spine abnormal if upper cervical etiology

# Postural Orthostatic Tachycardia Syndrome (POTS)

A cardio-autonomic disorder with no apparent disease

Affects 1-2% of the U.S. population

Typically young, White women

Joint hypermobility syndromes common (EDS)

Gastrointestinal disorders (e.g., irritable bowel)

Other comorbidities (e.g., MCAS, migraine)

At least 30 bpm increase in heart rate in adults or 40 bpm or more in children OR an absolute heart rate of  $\geq$  120 bpm within 10 min of standing

Secondary causes must be excluded (medications, **deconditioning**/post-COVID, dehydration, hyperthyroidism, anemia, orthostatic hypotension, arrhythmias)

### Headache in POTS

Overall prevalence of headache 36.8% (95% CI 2.1-70.7%)

Orthostatic headache ranges from 2.2%-58.3%

Most patients have non-orthostatic headaches that are migraine-like

A case-control study (9 confirmed SIH, 48 POTS) identified:

Shorter disease duration (p<0.05), orthostatic headache (p=0.001) and neck stiffness (p=0.008), older age(p=0.052) more common with SIH

Syncope (p=0.033), worsening with menses (p=0.004) and myofascial pain (p=0.018) more common with POTS

Neck stiffness was only present in patients with SIH (p=0.008)

But...many symptoms were present in both groups

### Other Manifestations of POTS

#### Hyperadrenergic POTS

Headache, dizziness, tremulousness, anxiety, increased urination, cold and diaphoretic extremities

#### **Neuropathic POTS**

Dizziness, chronic fatigue, weakness, insomnia, anhidrosis, acral cyanosis of the feet when standing

#### Hypovolemic POTS

Weakness, brain fog, fatigue and exercise intolerance in the upright posture

#### Adolescents with POTS

Dizziness, weakness, dyspnea, tremulousness, orthostatic syncope

## It's Complicated...

It is difficult to distinguish POTS from SIH clinically

Orthostatic vital signs and tilt table test are warranted but not always conclusive

Not all patients with orthostatic headache develop a headache during a tilt-table test and a headache during upright tilt does not correlate with orthostatic headache during daily activities

SIH patients can also have increased systolic BP with head-up tilt and heart rate variability with deep breathing, identical to POTS

POTS can develop as a result of SIH due to deconditioning, so SIH patients may also have POTS and its associated manifestations

# POTS or Spinal CSF Leak or Both?

	POTS	Spinal CSF Leak
Headache phenotype	Usually migraine	Anything
Headache location	Typically frontal or holocranial	Often posterior but no specific location
Orthostatic headache	2% to 58%	Over 90% at some point
Joint hypermobility	Common	Common
VIII nerve involvement	Dizziness, usually lightheadedness	Vestibular (spinning, lightheadedness), cochlear
Syncope	Common	Uncommon
Tremulousness	Common	Uncommon
Abnormal tilt table test	Yes	Sometimes
Abnormal brain MRI	No	Please!!

### Chiari I Malformation

- 1% prevalence overall and up to 3.6% of pediatric population
- Most cases are sporadic although familial clusters exist
- May resolve or arise spontaneously (children)
- May be associated with A-A instability and connective tissue disorders



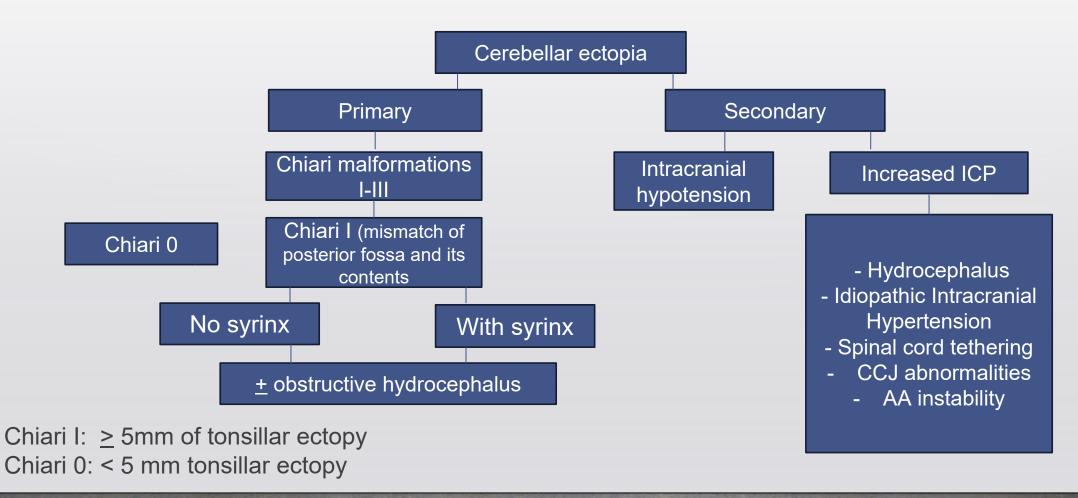
www.jeffreyhatef.med.com

Radsource thinks that I am a Chiari malformation!



https://radsource.us

### Chari I Malformation



# Symptoms of Chiari I

In general, symptoms arise from

**CSF** obstruction

Brainstem or cerebellar compression/dysfunction (cranial nerves VIII-XII, autonomic, nystagmus)

Related to spinal cord or syringomyelia

#### **Pediatric**

Brainstem dysfunction (central sleep apnea, feeding difficulties)

Headache (crying, irritability, failure to thrive, opisthotonos)

#### Adults

Pain or headache in occipital or cervical regions

Produced by Valsalva maneuvers (coughing, sneezing, laughing)

### Orthostatic Intolerance and Chari I?

Fatigue, lightheadedness/dizziness, blurred vision, impaired memory, tremors, palpitations, nausea, vomiting, syncope and non-syncopal orthostatic intolerance described

Case reports of improvement following decompression

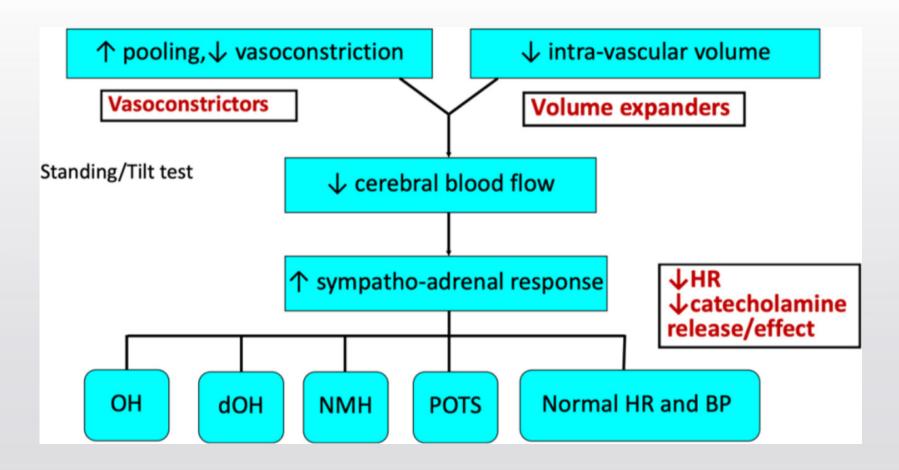
Co-existing POTS and EHS and other connective tissue disorders may be causative

# Chiari I or Spinal CSF Leak?

	Chiari I	Spinal CSF Leak
Occipital headache	Yes	Common
Orthostatic headache	With co-existing POTS	Yes
Neck pain	Yes	Yes
Syncope	Yes	No
Dizziness	Common (lightheaded)	Common (vertigo, imbalance)
MRI	Small, crowded posterior fossa, peg-like tonsils	Tonsillar descent + other abnormalities



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OH=orthostatic hypotension; dOH=delayed OH, NMH = neurally mediated hypotension, POTS = postural orthostatic tachycardia syndrome