

“Spinal CSF Leak: Bridging the Gap between Patients and providers”

Spinal CSF leak: clinical diagnosis challenges

Arriving at the diagnosis

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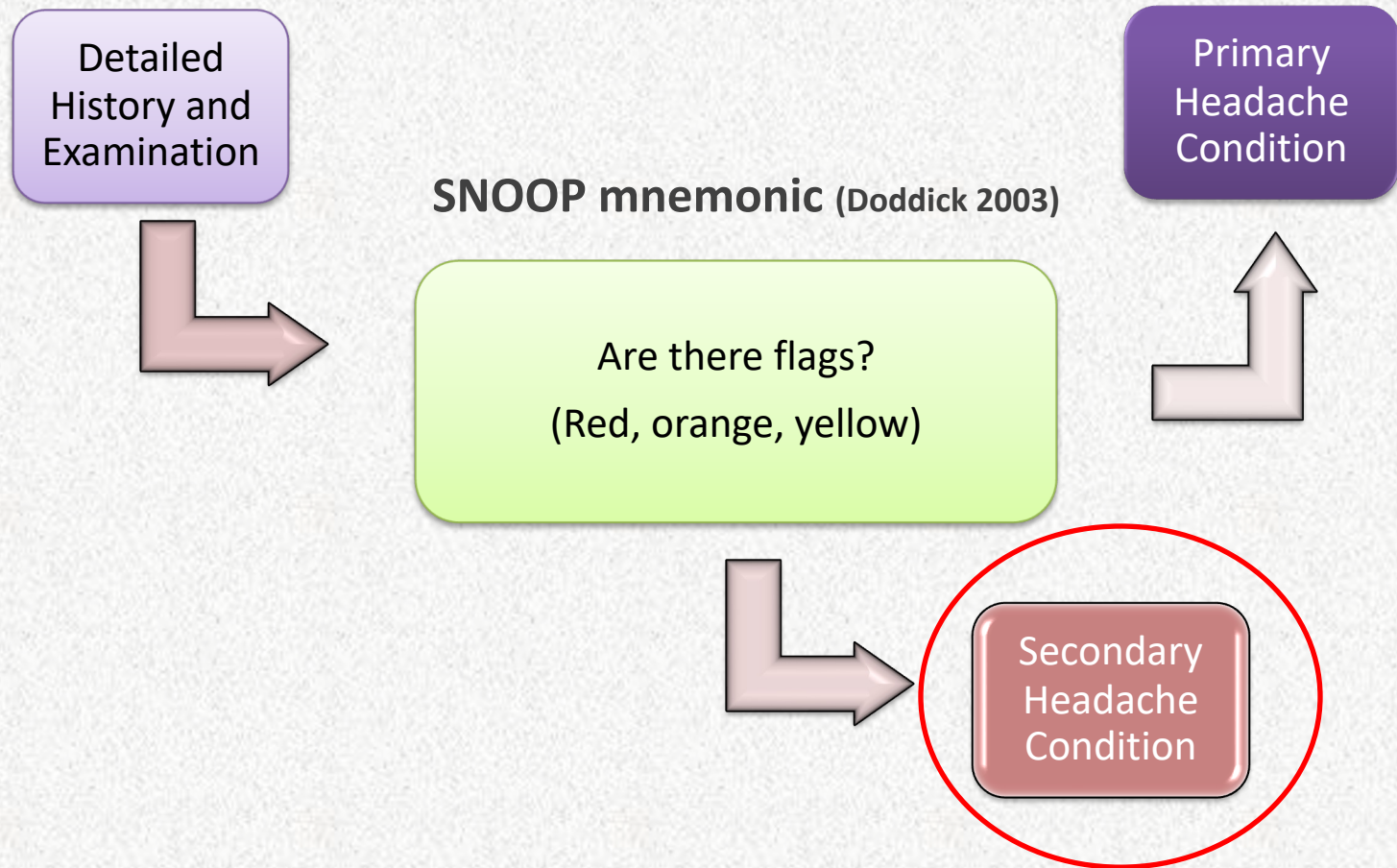


11/11/2023

No disclosures

Diagnostic algorithm

HEADACHE



“P” red flags

- | | |
|-----------|---|
| 6 | Pattern change or recent onset of headache |
| 7 | Positional headache |
| 8 | Precipitated by sneezing, coughing, or exercise |
| 9 | Papilledema |
| 10 | Progressive headache and atypical presentations |
| 11 | Pregnancy or puerperium |
| 12 | Painful eye with autonomic features |
| 13 | Posttraumatic onset of headache |
| 14 | Pathology of the immune system such as HIV |
| 15 | Painkiller overuse or new drug at onset of headache |

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ICHD 3 definition

Headache attributed to low CSF pressure	Headache attributed to SIH
A. Any headache fulfilling criterion C.	A. Headache fulfilling criteria for <i>Headache attributed to low CSF pressure</i> , and criterion C below.
B. Either or both of the following: 1. Low CSF pressure (<60 mm CSF) 2. Evidence of CSF leakage on imaging	B. Absence of a procedure or trauma known to be able to cause CSF leakage.
C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage or led to its discovery.	C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage or has led to its discovery.
D. Not better accounted for by another ICHD-3 diagnosis.	D. Not better accounted for by another ICHD-3 diagnosis.

Spontaneous intracranial hypotension (SIH)

some statistics

Characteristics	
Age	42.5 years (2-88)
Gender	Women 63%
Time to diagnosis	32 days (1 day to 19.7 years)
Headache present	97%
Low spinal fluid pressure	67%*

D'Antona et al, JAMA Neurology 2021

* 34% or less in other reports (Kranz et al, Headache 2018)

Annual incidence: 3.8-5/100,000

(Schievink et al, J Headache Pain, 2007, Pradeep et al, Interventional Neurorad 2023)

Clinical Presentation, Investigation Findings, and Treatment Outcomes of Spontaneous Intracranial Hypotension Syndrome

A Systematic Review and Meta-analysis

Linda D'Antona, MD, MBBS; Melida Andrea Jaime Merchan, MD; Anna Vassiliou, iBSc; Laurence Dale Watkins, MD; Indran Davagnanam, MD; Ahmed Kassem Toma, MD; Manjit Singh Matharu, MD, PhD







OPEN ACCESS

Original research


Multidisciplinary consensus guideline for the diagnosis and management of spontaneous intracranial hypotension

J Neurol Neurosurg Psychiatry 2023

Sanjay Cheema ^{1,2}, Jane Anderson,³ Heather Angus-Leppan ⁴, Paul Armstrong,⁵ David Butteriss,⁶ Lalani Carlton Jones,^{7,8} David Choi,^{1,9} Amar Chotai,⁶ Linda D'Antona ^{1,9}, Indran Davagnanam,^{1,10} Brendan Davies,¹¹ Paul J Dorman,¹² Callum Duncan,¹³ Simon Ellis,¹¹ Valeria Iodice,^{1,14} Clare Joy,¹⁵ Susie Lagrata,² Sarah Mead,¹⁵ Danny Morland,¹⁶ Justin Nissen,¹⁷ Jenny Pople,¹⁵ Nancy Redfern,¹⁶ Parag P Sayal,⁹ Daniel Scoffings,¹⁸ Russell Secker,¹⁵ Ahmed K Toma,^{1,9} Tamsin Trevarthen,¹⁵ James Walkden,¹⁹ Jürgen Beck,²⁰ Peter George Kranz,²¹ Wouter Schievink,²² Shuu-Jiun Wang,^{23,24} Manjit Singh Matharu ^{1,2}

Patient experience of spontaneous intracranial hypotension (SIH): qualitative interviews for concept elicitation

J of Patient-Reported Outcomes 2023

Timothy J. Amrhein^{1*} , Molly McFatrigh², Kate Ehle², Michael D. Malinzak¹, Linda Gray¹, Peter G. Kranz¹, E. Hope Weant¹ and Christina K. Zigler²

Clinical diagnostic challenges

- Orthostatic nature
- Atypical presentations
- Cerebrospinal fluid pressure
- Predisposing and precipitating factors
- Cognitive bias

Orthostatic Headache (OH)

- **Prevalence:** 92% or less in SIH
- **Time to onset of being upright and time to peak**
 - 2 hours? 1 hour? can function upright at all?
- **Time to improvement after lying down (flat):**
 - may take up to 2 hours, may not need to be complete
- **Second half of the day** headache (Dr. Mokri)
- **Orthostatic feature may disappear over time:**
 - 93% if presented within 10 weeks of symptoms onset vs
 - < 63% after 10 weeks
- **Paradoxical headache:** worse lying down
- **Neck involvement:** cervicogenic?

Atypical presentations: timing

- Chronic Daily Headache (CDH +/-O)
- New Daily Persistent Headache
- Thunderclap headache
- Recurrent
- Other: children, older, pregnancy

Atypical presentations: symptoms other than headache

Commonly associated symptoms	Rare presentations
Dizziness or vertigo (50.5%)	Interscapular pain (10.9%)
Nausea and vomiting (49.0%)	Dysgeusia (7.4%)
Disequilibrium (42.6%)	Hyperacusis (5.9%)
Muffled hearing or aural fullness (37.1%)	Behavioural variant frontotemporal dementia syndrome (2.5%)
Posterior neck pain (34.2%)	Reverse orthostatic headache (2%)
Cognitive impairment† (31.7%)	Bibrachial amyotrophy (1.5%)
Tinnitus (27.7%)	Superficial siderosis (1.5%)
Hypoacusis (26.2%)	Cerebral venous thrombosis (1%)
Fatigue (24.3%)	Abducens nerve palsy (1%)
Photophobia or phonophobia (20.3%)	Spinal cord herniation (1%)
Visual blurring (17.8%)	Coma (0.5%)
Facial numbness, paraesthesia or pressure (15.8%)	Syringomyelia (0.5%)
	Hemifacial spasm (0.5%)

*Adapted from Schievink [4].
†Most commonly non-specific problems with concentration and word finding.⁴
SIH, spontaneous intracranial hypotension.

Seizures; Movement disorders; Functional Neurological Disorder (FND)

Cheema et al, JNNP 2023

Amrhein et al, J of Patient-Reported Outcomes, 2023

Pere-Vega et al, Neurological Sciences 2020

Ozge&Bolay, Curr Pain and Headache Rep 2014

Atypical presentations: prompted and non-prompted symptoms

Table 2 Frequency[^] of symptoms reported by 15 adult participants with SIH.

Symptom as described by the patient (medical term)	Total (n)	Unprompted ¹ (n)
Head pain/headache	15	15
Ringing in ears / muffled hearing (tinnitus)	13	5
Ear fullness / pressure, and/or pain	9	6
Neck pain	9	5
Interscapular pain	9	3
Nausea	9	2
Fatigue	8	2
Cognitive/memory issues	8	1
Dizziness / light headedness / sense of imbalance (disequilibrium)	7	5
Difficulty finding words (anomic aphasia)	6	0
Head pressure	6	6
Sensitivity to sound (hyperacusis)	6	0
Sensitivity to light (photophobia)	6	1
Blurry vision / double vision (diplopia)	3	0
Hearing loss	3	1
Impaired speech (e.g., difficulty forming words, stutter)	2	2
Runny nose or liquid drainage (rhinorrhea)	2	1
Vomiting	2	0
Loss of mobility	1	1
Low back pain	1	1
Difficulty swallowing (dysphagia)	1	1
Eyeball pain (ophthalmalgia)	1	1
Sensitivity to smell (hyperosmia)	1	0

“Patient experience of spontaneous intracranial hypotension (SIH): qualitative interviews for concept elicitation”

Patient experience of spontaneous intracranial hypotension (SIH): qualitative interviews for concept elicitation

Meaningful patient experience identified via qualitative interviews	Outcome of Interest	Existing patient-reported outcome measures (PROMs) that may be useful in measuring the outcome of interest ¹
Primary Symptom - pain	Headache pain* (intensity/frequency)	Headache Impact Test (HIT-6); PROMIS Pain Intensity; Numeric Rating Scale; Pain, Enjoyment, General Activity (PEG) Scale Assessing Pain Intensity and Interference
Other symptoms - varied	List of common and uncommon symptoms associated with SIH (presence/absence)	Due to unique nature of SIH, the development of an SIH-specific symptom battery could be considered
Other symptom – fatigue	Fatigue (intensity)	PROMIS Fatigue
Other symptoms – cognitive impairment	Cognitive impairment	PROMIS Cognitive Functioning
Functioning	Pain interference*	Pain, Enjoyment, General Activity (PEG) Scale Assessing Pain Intensity and Interference
Functioning	Physical Functioning*: Impact to daily activities and limits to physical activities/exercise	PROMIS Physical Functioning
Functioning	Social Role Functioning/Participation	PROMIS Ability to Participate in Social Roles and Activities
Headache characteristics	Headache pain characteristics (frequency) ² : Improve when lay down, worsen when bent over, worsen when lift heavy objects, worsen as day progresses	The development of SIH-specific items could be considered.

Predisposing and Precipitating factors

Predisposing

- degenerative spine changes
- connective tissue disease (HSD, EDS, Marfan, etc)
- Other: i.e. dramatic weight loss

Precipitating

- Sitting/standing
- exercise
- Valsalva
- “just getting up in the morning”
- “just living”

Cognitive biases

“The diagnostic failure rate is estimated to be 10 to 15%” (Society for the Improvement of Diagnosis in Medicine, Graber et al, JAMA 2012)

Greater than 90% of SIH patients are initially misdiagnosed (Schievink et al, Arch Neurol 2003)

Physician features (i.e. “Chiari malformation”, CM):

Availability: unfamiliarity with CSF hypovolemia presenting as “CM”

Confirmation: a “malformation” is present, symptoms are present → need for decompression

Outcome Bias: this is “someone I can fix”

Zebra Retreat:

- pursuing this alternative diagnosis (SIH) to explain symptoms less desirable
- not worth time and effort
- the need to order costly tests, possibly perceived as wasting resources.

Clinical diagnostic challenges: consequences

- **Complications:**

- subdural hematoma (13-20%, ! if bilateral)

- superficial siderosis (up to 10.3 %)

- cerebral vein thrombosis (1-2%)

- spinal cord herniation (1%)

- coma (0.5 %, reversible!)

- **Under- and overdiagnosis**

- **Suboptimal outcomes**

Quality of Life in Patients with Confirmed and Suspected Spinal CSF Leaks

“Improved identification and treatment of SIH are imperative to improve patients’ QoL”

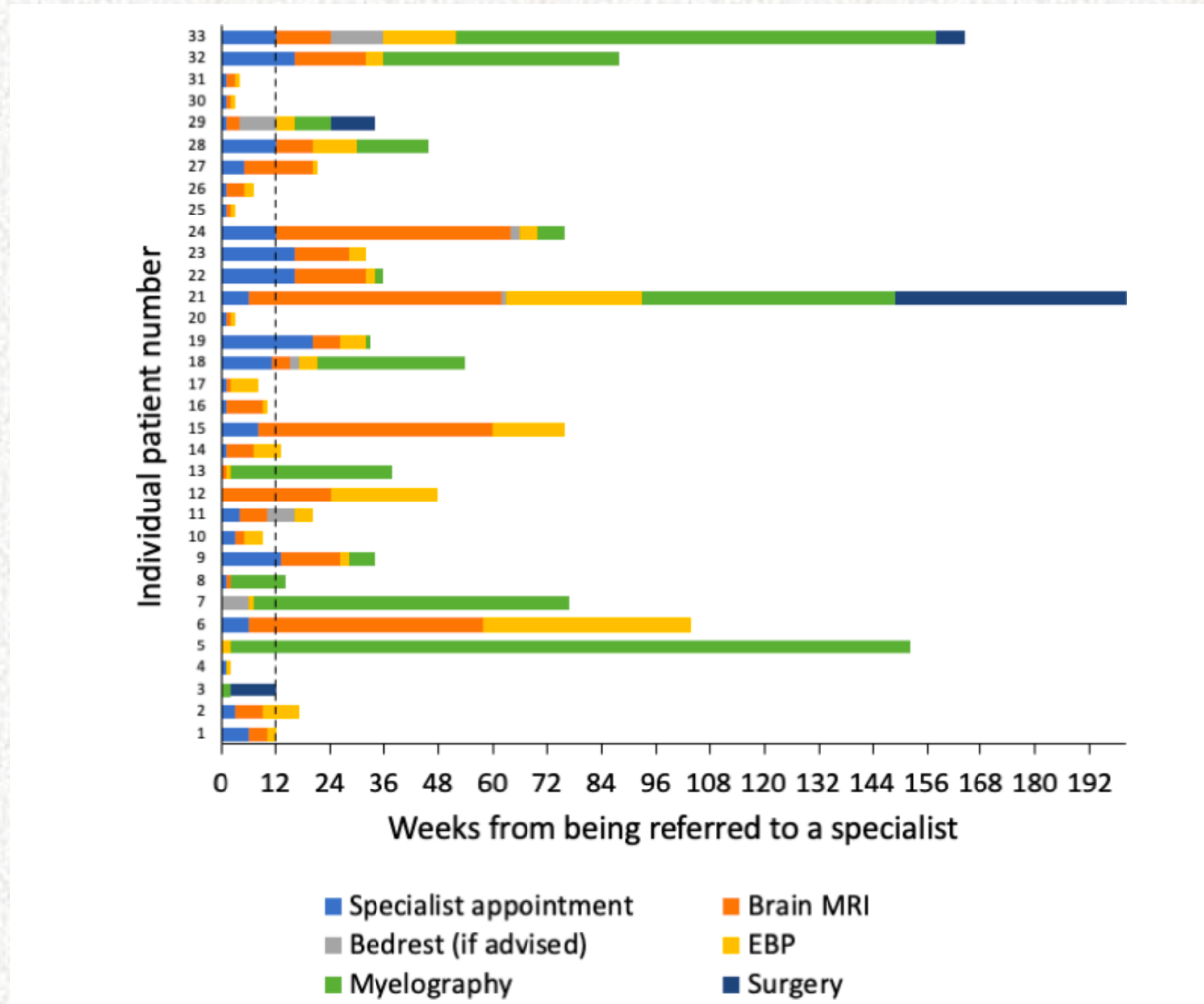
**THANK YOU VERY MUCH
FOR YOUR ATTENTION!**

Other causes

TABLE 5 Secondary causes of intracranial hypotension.

Lumbar puncture
Spinal anesthetic (e.g., labor/cesarean section)
Spinal surgery
Other surgeries (e.g., bariatric)
Lumboperitoneal or ventriculoperitoneal shunt (leading to over-drainage)
Spinal manipulation (e.g., chiropractic)
Traumatic

Delays from referral to a specialist to investigation and treatment.



Who (mis)diagnoses SIH?

Table 5. Specialty of Physician Misdiagnosing Spontaneous Intracranial Hypotension

Specialty	No. (%) of Patients
Emergency medicine	17 (34)
Neurology	15 (30)
Family practice	7 (14)
Neurosurgery	5 (10)
Internal medicine	4 (8)
Dentistry	1 (2)
Orthopedic surgery	1 (2)

Greater than 90% of patients are initially misdiagnosed

Urgency of referral to neurologist

- 2–4 weeks: if the patient is able to care for his or her self, the urgency of the referral should be based on the severity of clinical features including mental health impact.
- 48 hours: if If the patient is not able to care for his or her self but has help
- Emergency admission: if patients are not able to care for themselves and do not have help