

Orthostatic headache syndromes

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What is orthostatic headache?

Headache which is **consistently** precipitated or significantly worsened by the upright position and resolves or significantly improves on lying flat

Typically associated with **spinal CSF leak** causing **low CSF pressure/volume**

DDx – what is not orthostatic headache?

Migraine / primary NDPH

Cervicogenic headache

Postural tachycardia
syndrome / orthostatic
hypotension

Chiari malformation

DDx – what is not orthostatic headache?

Migraine / primary NDPH

Worsened by **movement** rather than posture

Cervicogenic headache

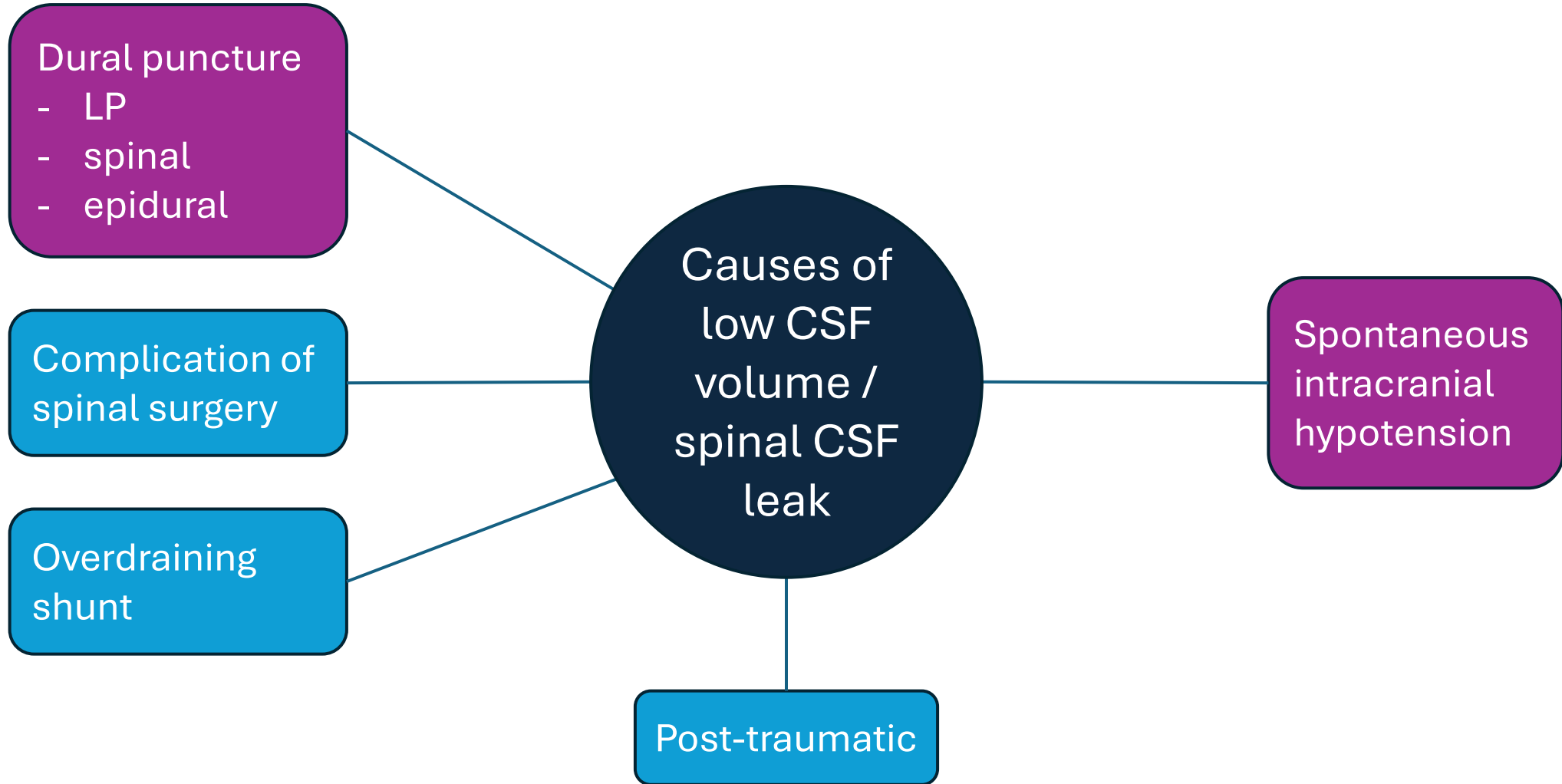
Worsened by **neck position** / movement rather than orthostasis

Postural tachycardia syndrome / orthostatic hypotension

Associated symptoms – e.g. light-headedness and palpitations, headache **less consistently** orthostatic

Chiari malformation

Worsened by Valsalva but not orthostasis, lack of other **MRI** brain features of intracranial hypotension



Post-dural puncture headache

ICHD-3:

“Headache occurring **within 5 days of a lumbar puncture**, caused by cerebrospinal fluid (CSF) leakage through the dural puncture. It is usually accompanied by **neck stiffness** and/or subjective **hearing symptoms**. It **remits spontaneously** within 2 weeks, or after sealing of the leak with autologous **epidural lumbar patch**.”

Incidence:

Diagnostic LP – approx. 5%

Epidural anaesthesia – approx. 0.5%

Spinal – rare (needle type)

Risk factors

Demographic

- Female
- Age 20-50
- Previous PDPH
- Migraine

Procedural

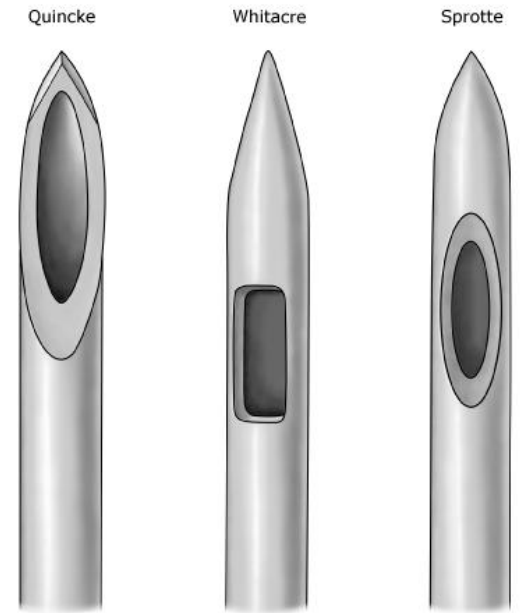
- Multiple attempts / inexperienced operator
- Needle bevel type, size, +/- orientation of entry
- No significant benefit of bed rest post-procedure

Prevention better than cure

- Multiple studies showing benefit of atraumatic needles for lumbar puncture¹ and spinal anesthesia²
- For diagnostic LP incidence of PDPH reduced from 11% to 4%¹
- Odds ratio of needing EBP approx. 0.5¹
- No difference in LP success rate¹
- Size also important - for spinal anesthesia the 26-G atraumatic needle most likely to enable successful insertion while avoiding PDPH²

¹Nath et al. Lancet. 2018;391(10126):1197-1204

²Maranhao et al. Anaesthesia. 2021;76(8):1098-1110



17	Red-Purple	1.4
18	Pink	1.2
19	Cream	1.1
20	Yellow	0.9
21	Dark Green	0.8
22	Black	0.7
23	Deep Blue	0.6
24	Purple	0.55
25	Orange	0.5
26	Brown	0.45
27	Med-Grey	0.4

Symptoms

- Orthostatic headache
- Neck stiffness, nausea, photophobia
- Audiovestibular symptoms – tinnitus, muffled hearing, dizziness

Post-partum headache has other causes:

Primary – migraine, TTH

Secondary – CVST, PRES/eclampsia

Acute management

- “Conservative” management (many resolve spontaneously – estimated in 2/3)
 - offer analgesia, maintain hydration
 - bed rest (but probably does not affect prognosis)
 - small RCT level evidence for caffeine and GON injection
 - negative trial of SPG block
- EBP is gold standard – at level of puncture
- MRI imaging desirable as baseline but not essential

Persistent post-dural puncture headache

MR brain and spine imaging usually negative

Re-review phenotype and treat any primary headache disorder and comorbidities

Options:

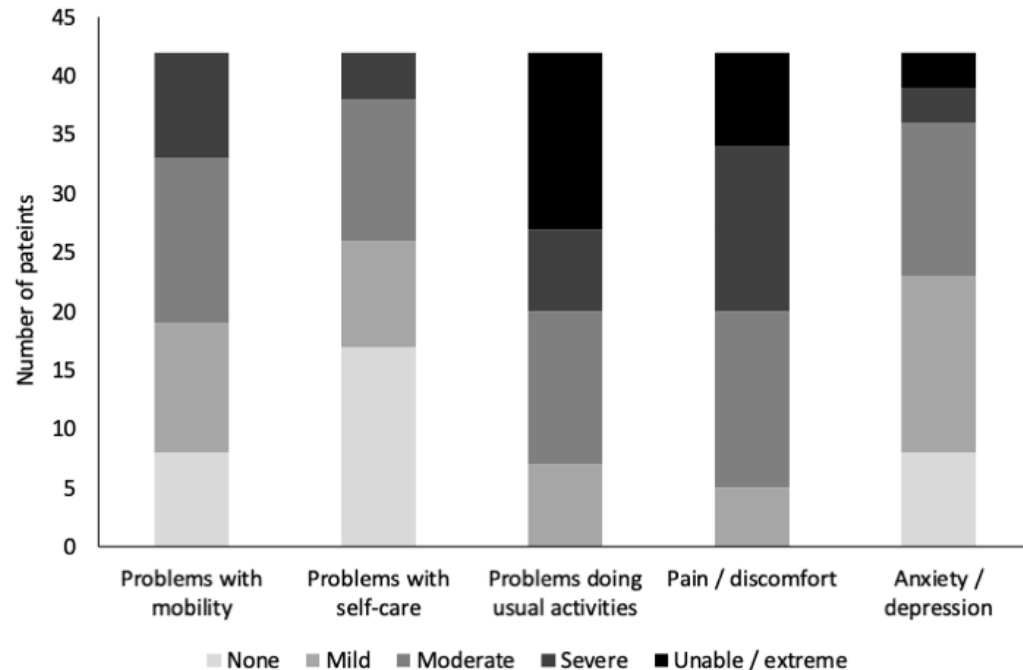
- treat symptomatically – inconsistently effective
- investigate with myelography – usually negative
- repeat EBP at original dural puncture site
- fibrin glue patch at dural puncture site
- exploratory surgery at dural puncture site

Spontaneous intracranial hypotension

= spontaneous spinal CSF leak

Disability

Mean EQ-5D VAS score = 36.4/100



Possible complications

- Subdural haematoma
- Venous sinus thrombosis
- Superficial siderosis
- Bibrachial amyotrophy
- Behavioural variant FTD-like syndrome
- Coma

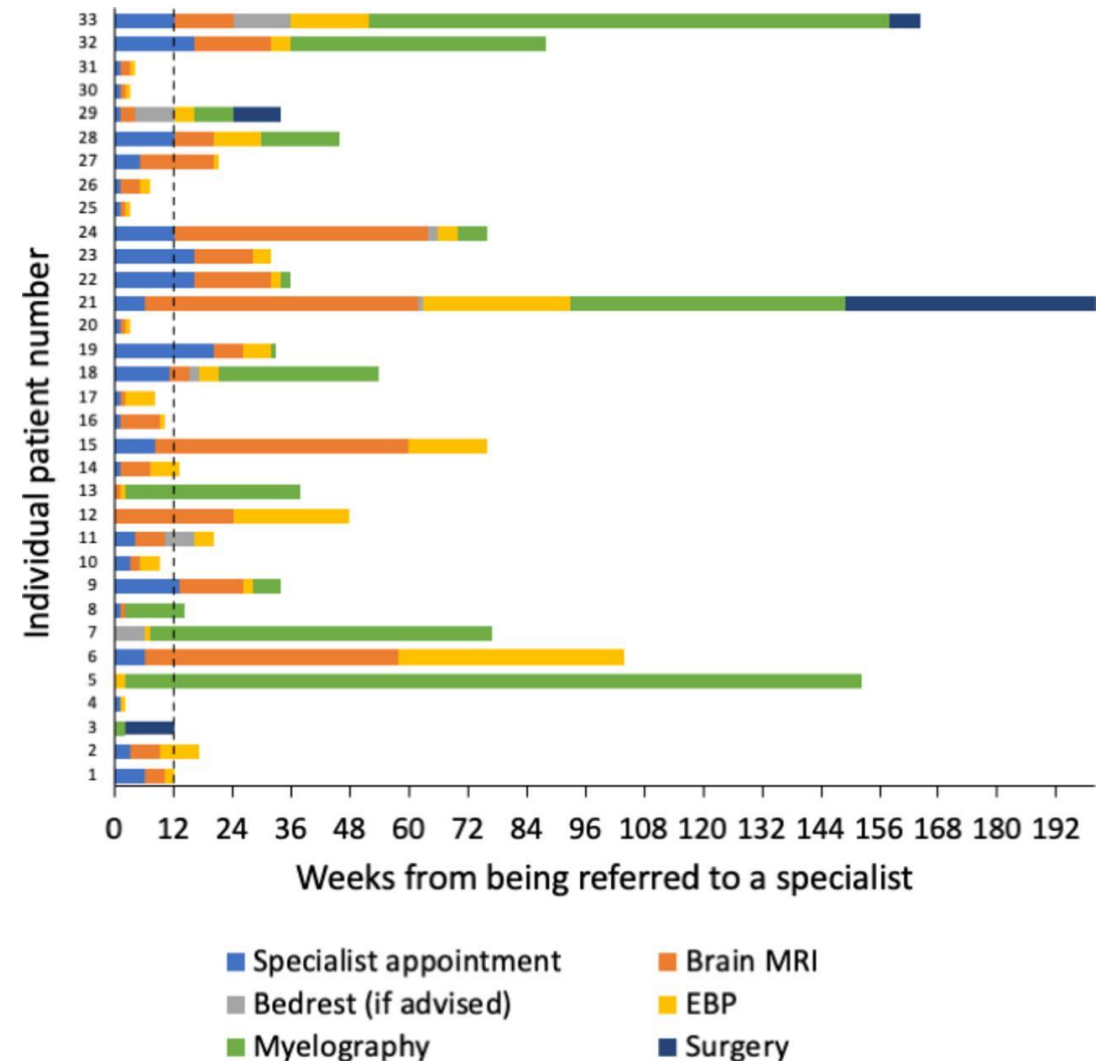
2003 study (US)

- 94% of patients initially received an incorrect diagnosis
- diagnostic delay ranged from 4 days to 13 years (mean 13 months)

2022 study (UK)

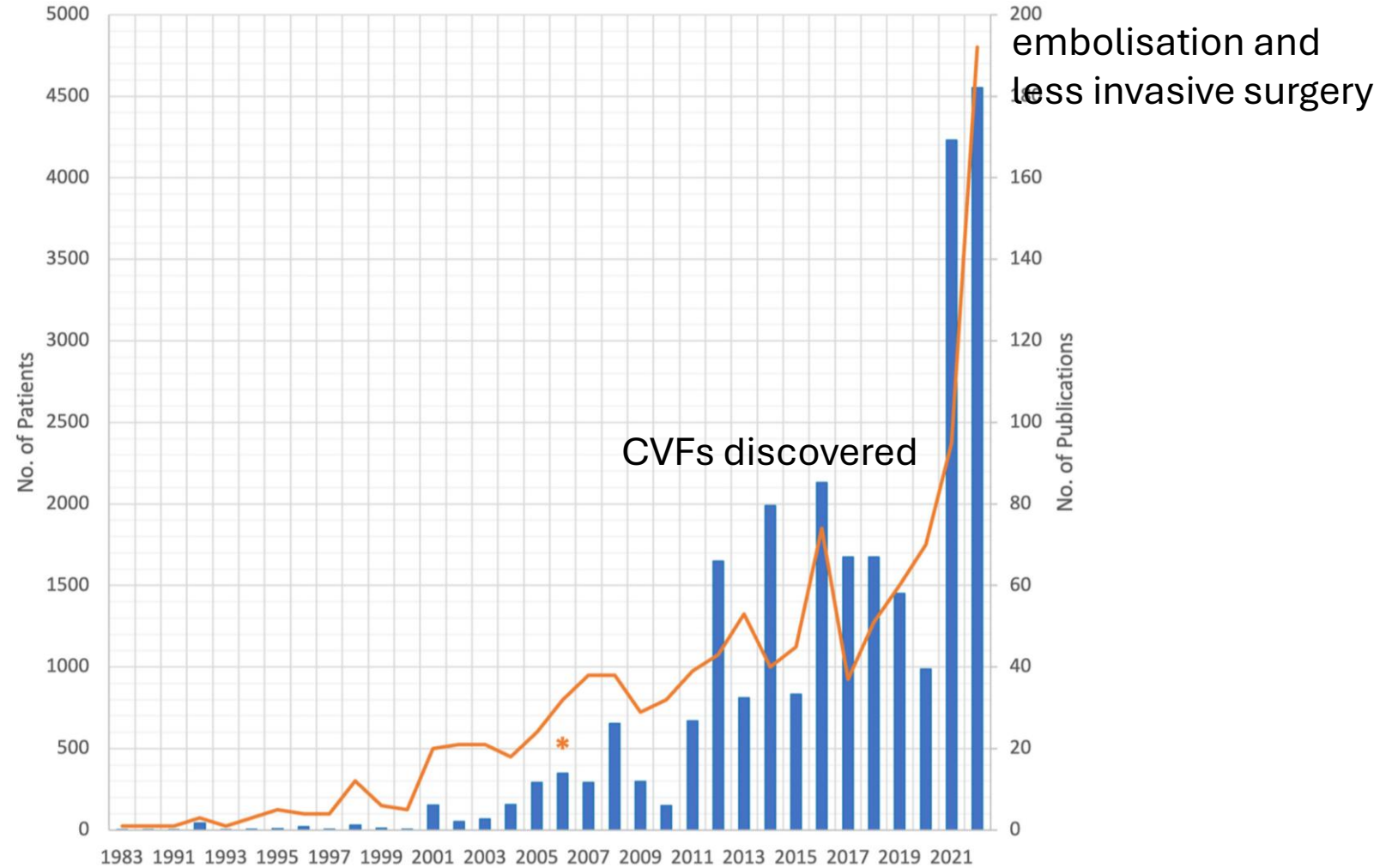
- median of 3 GP visits before referral
- diagnosis made by first specialist in only half
- long delays to investigation and treatment – only 1/3 patients treated within 12 weeks of seeing a specialist

Schievink. Arch Neurol. 2003; 60(12):1713-8.



Cheema et al. BMJ Open. 2022; 12(1):e057438

Understanding of SIH

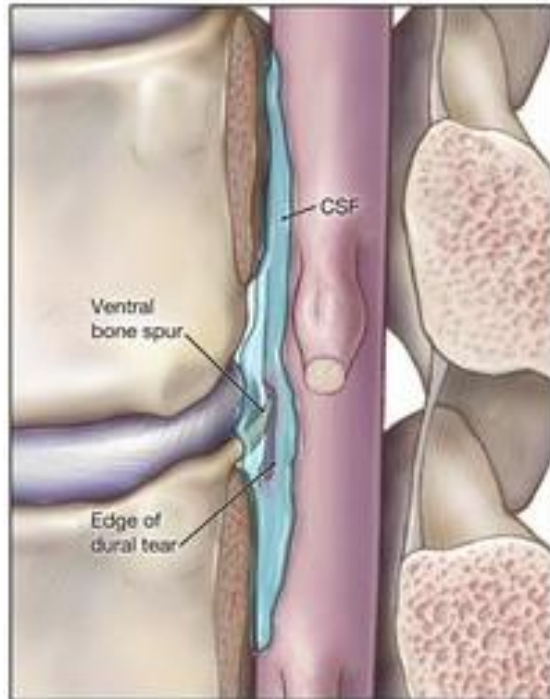


Jesse et al. SIH. A bibliometric analysis. Brain and Spine. 2024; 30:4:103324

Spontaneous intracranial hypotension

Type 1

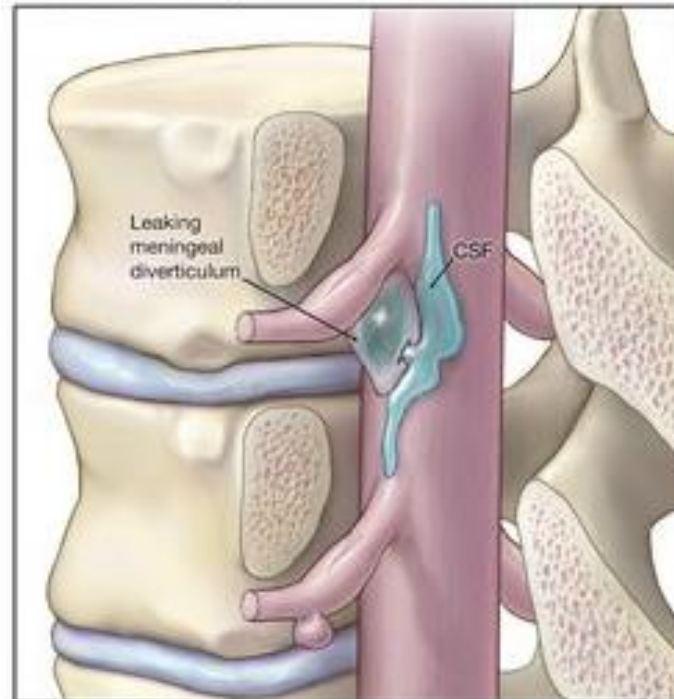
Lateral view of left side



ventral dural tear

Type 2

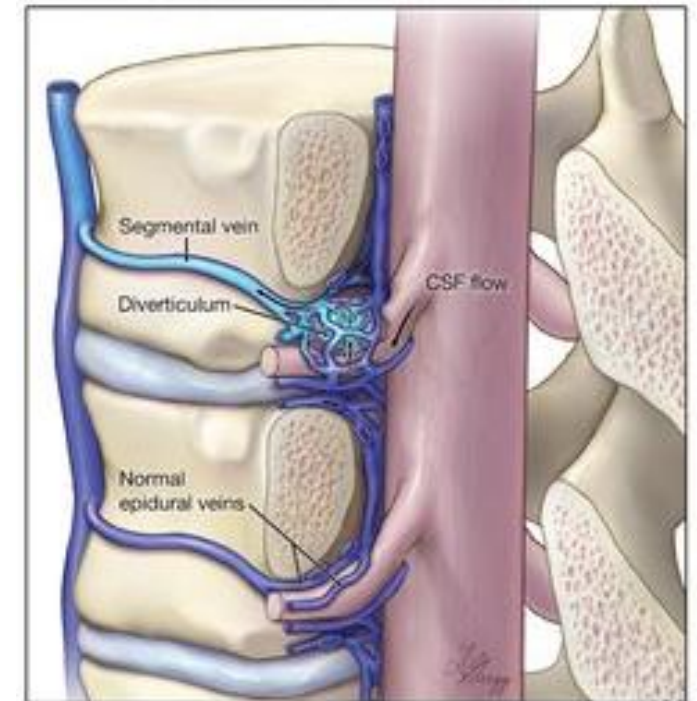
Posterior-oblique view of left side



meningeal diverticula

Type 3

Posterior-oblique view of left side



CSF-venous fistula

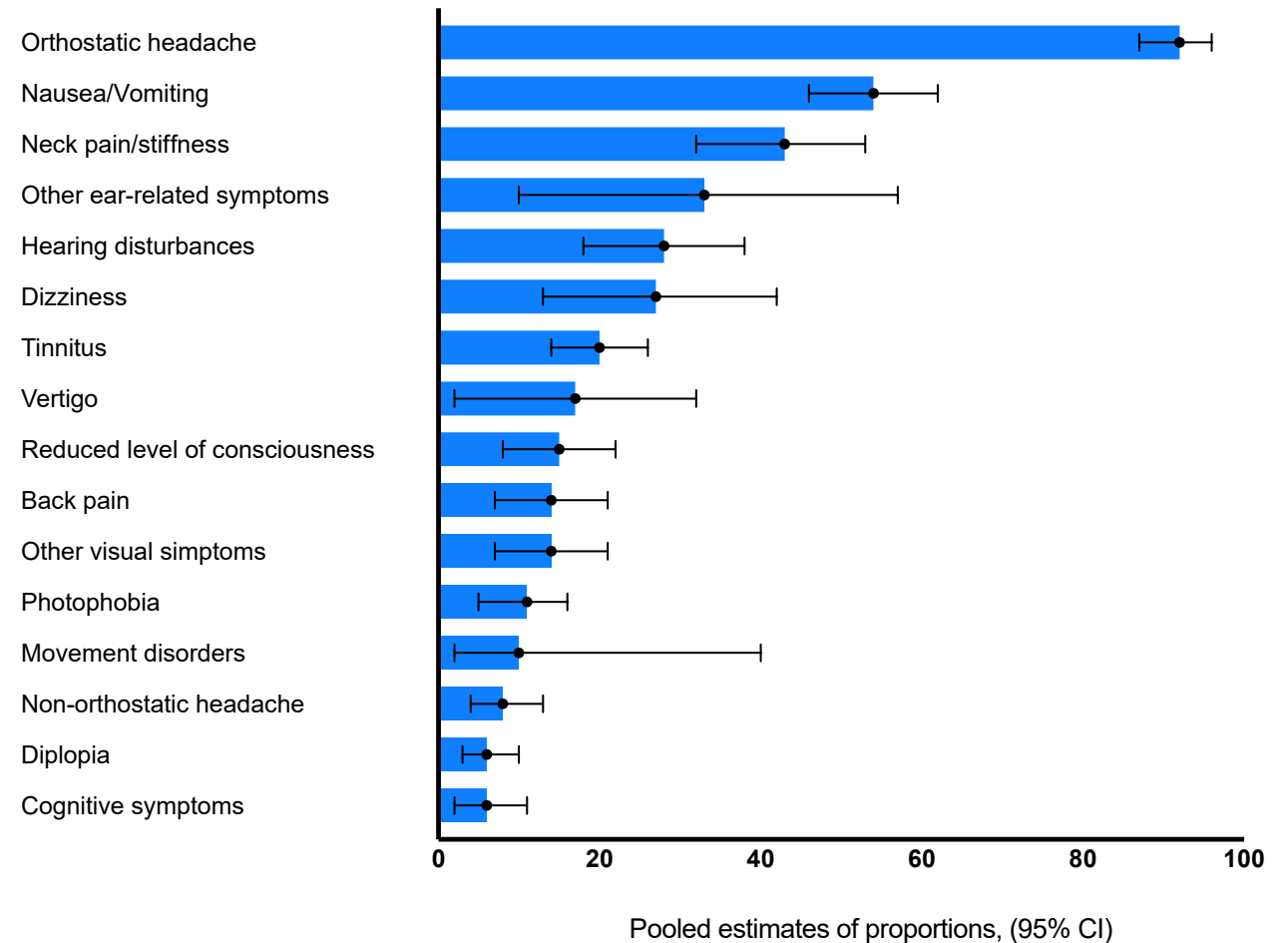
Stages of diagnosis and management of SIH



1

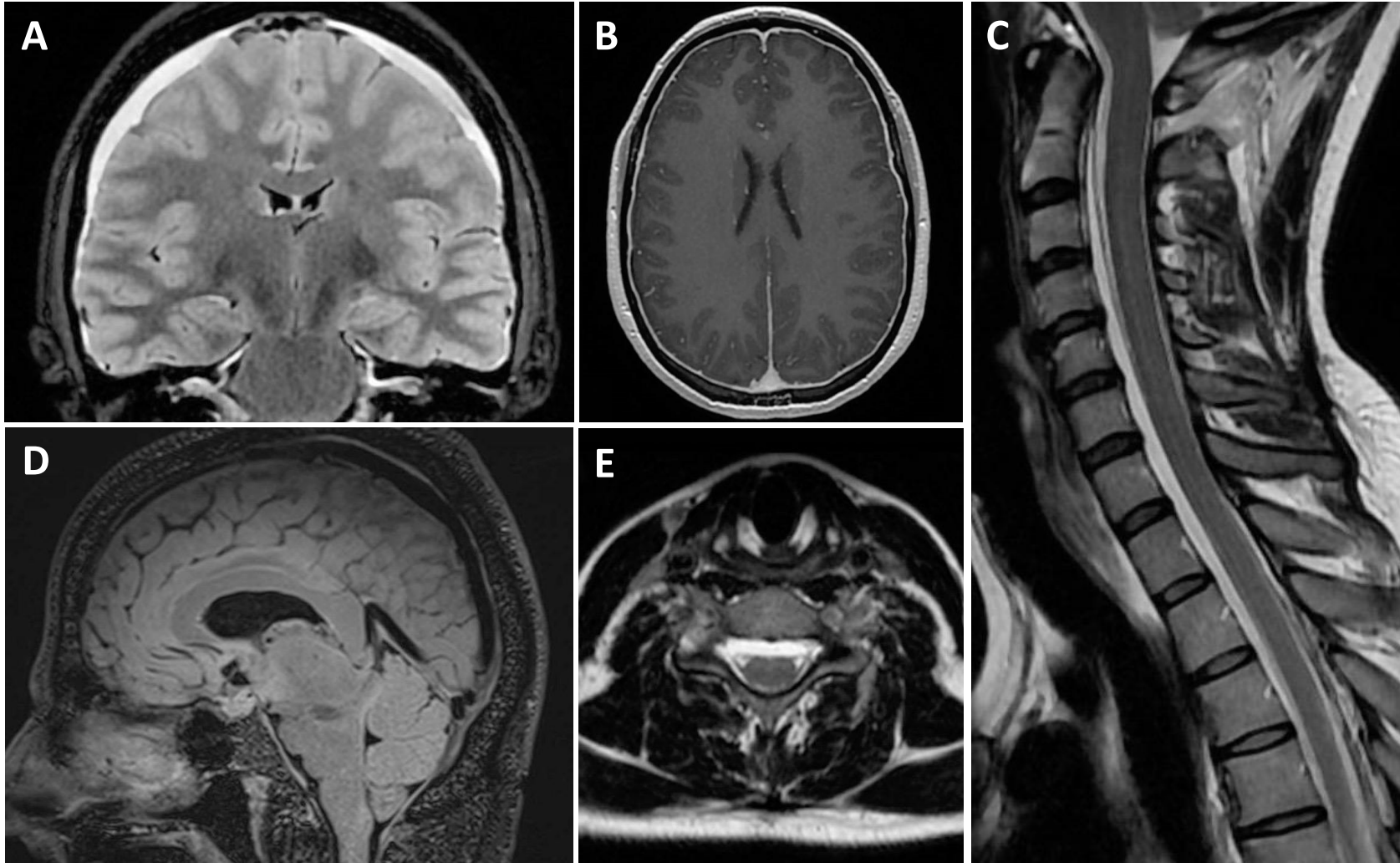
Clinical suspicion

- 92% orthostatic headache
- 5% non-orthostatic headache
- 2% no headache
- Common associated symptoms
- Timing of onset / offset variable
- Orthostatic relationship may attenuate over time
- Consider differential diagnosis



2

Radiological identification of low CSF pressure/volume



Conservative/symptomatic measures

- Bed rest, hydration, abdominal binders, avoidance of Valsalva
- Consider analgesia (but often not effective)
- Oral or intravenous caffeine can be considered but limited evidence in SIH
- Consider measures to reduce deconditioning and DVT risk
- Should not delay investigations or more definite treatment by more than two weeks
- Treat comorbid headache phenotypes e.g. migraine (but caution with topiramate, candesartan, propranolol)

Epidural blood patch

- Offer after no more than two weeks of conservative management¹
- Consider repeating after 2-4 weeks if lack of / transient improvement to first EBP¹
- Volume of blood >20ml most important predictive factor of response
- Followed by bed rest for 2-24h, be aware of red flag features of SCC/CES, and possibility of rebound headache
- **“a single epidural blood patch was “successful” in 64% (95% CI, 56%-72%)”²**

¹ Cheema S, et al. JNNP. 2023; 94:835-843

² D'Antona L, et al. JAMA Neurol. 2021;78(3):329-37

How often does non-targeted EBP really work?

- 45/51 (88%) had complete or substantial symptom improvement in the short term¹
- 36/51 (71%) patients had a persistent spinal CSF leak on imaging and/or intraoperatively¹

Clinical response	
Category	Frequency (%)
Resolution	17 (23.9)
Improvement	27 (38)
Unchanged	25 (35.2)
Worsening	2 (2.8)
Radiological response	
Category	Frequency (%)
Resolution	4 (5.6)
Improvement	33 (46.5)
Unchanged	34 (47.9)

¹ Piechowiak E, et al. Clin Neuroradiol. 2023;33:211–218

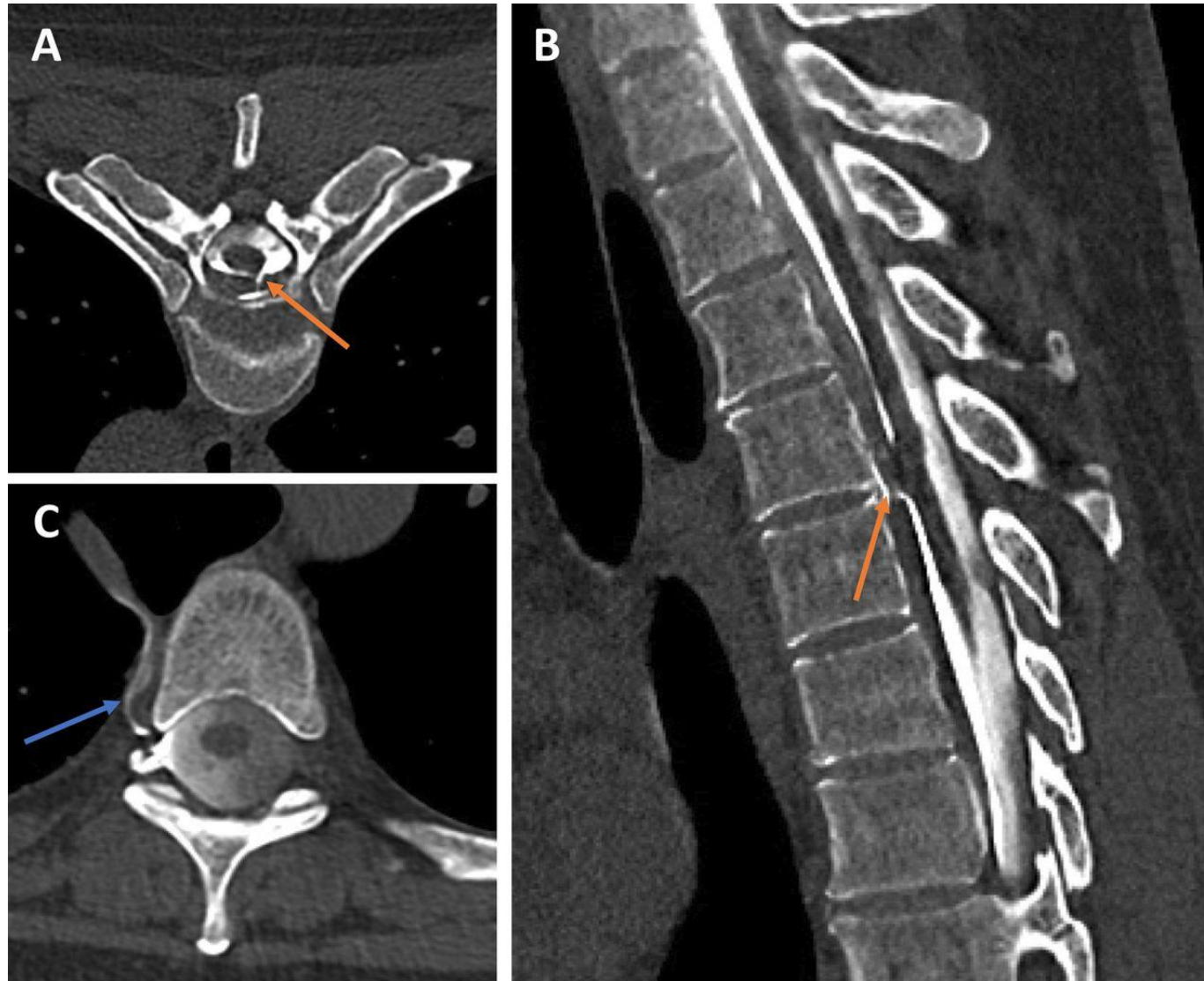
² Mehta D, et al. Unpublished

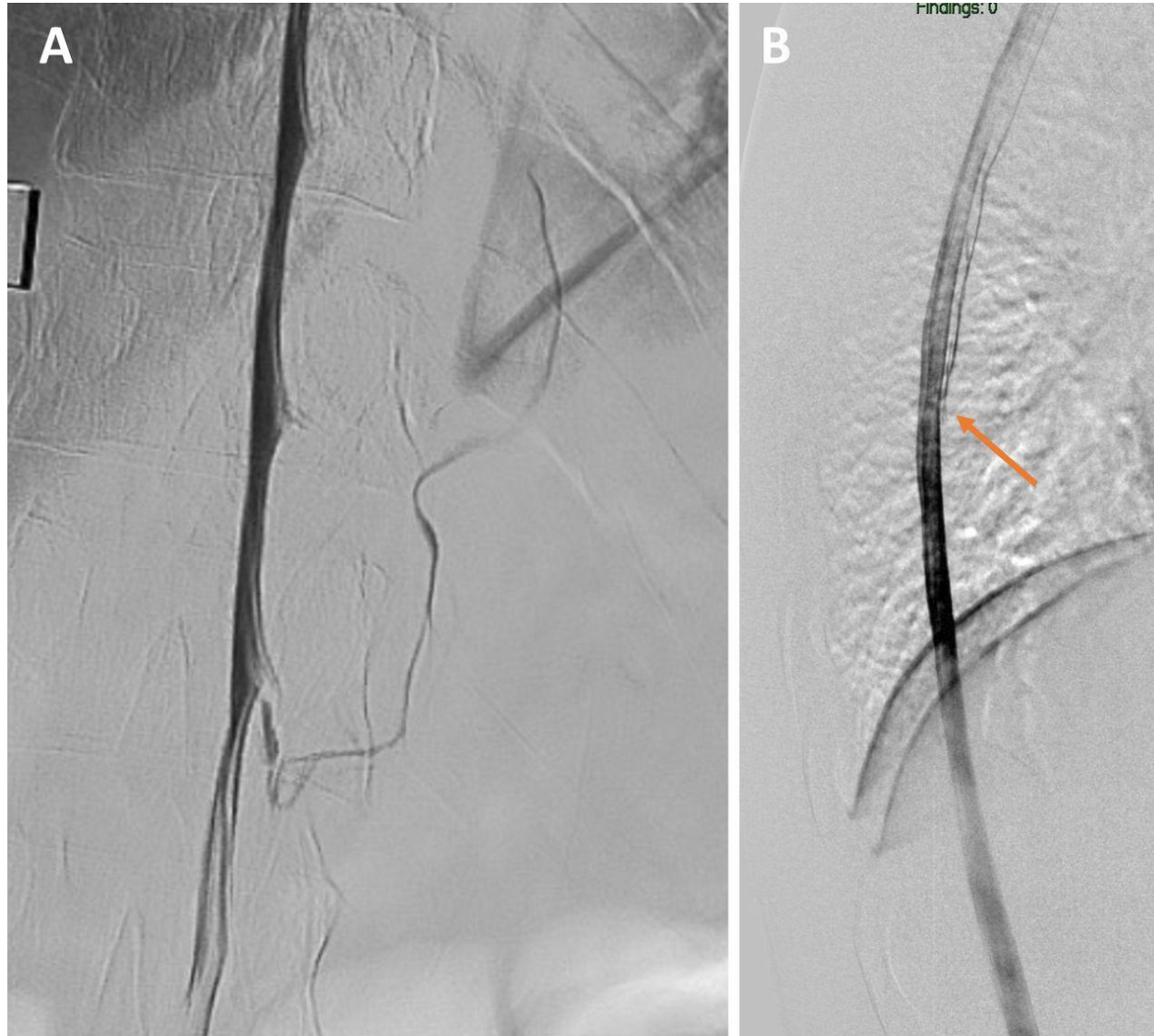
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Identification of precise spinal CSF leak site

- Contrast enhanced myelography (CTM / DSM)
- Usually performed if there is a lack of response to non-targeted EBPs

SLEC	Likely cause of leak	Patient position	Technique
Present	Discogenic microspur Lateral or dorsal dural tear	Depends on distribution of SLEC	CTM, DSM, UFCTM
Absent	CSF-venous fistula	Lateral decubitus	CTM, DSM





5

Targeted treatment

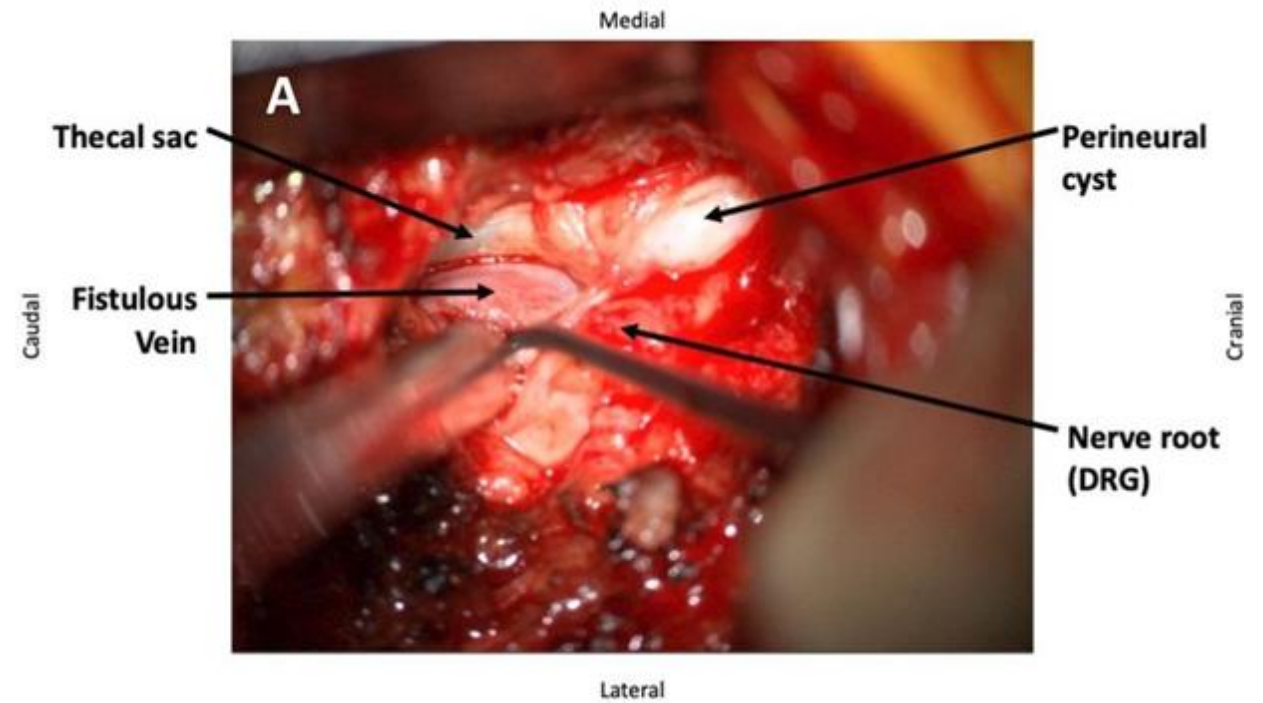
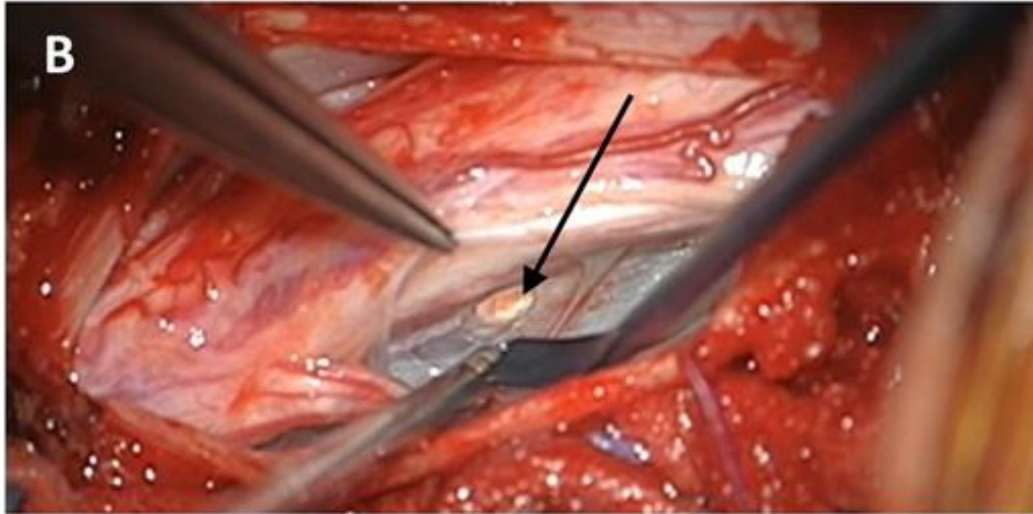
Type 1 / 2 leaks

1. Targeted blood and/or fibrin glue patches
2. Surgery

Type 3 leak (CVF)

1. Surgery
2. Embolisation
3. Targeted fibrin glue patching

MDT
decision





Not recommended routinely

- Lumbar puncture for the sole purpose of confirming the diagnosis of SIH
- Intrathecal gadolinium MR myelography
- Radionuclide cisternography
- Intraparenchymal intracranial pressure monitoring



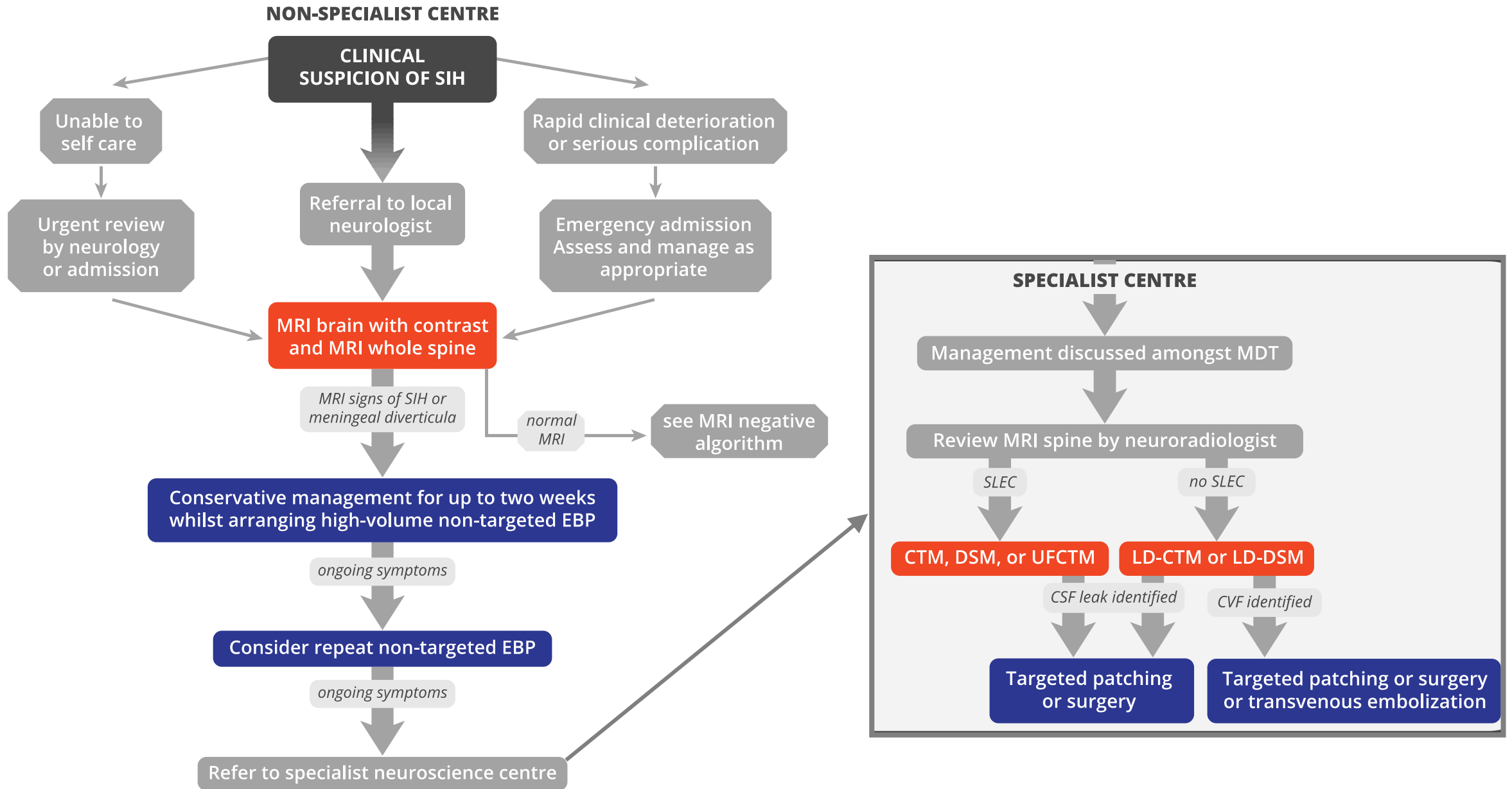
Original research

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

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}

<https://jnnp.bmj.com/content/94/10/835>





High clinical suspicion of SIH with normal MRI brain

- Consider **differential diagnoses**
- Ensure **spine MRI** done (approx. 5% brain negative but spine positive)
- Ensure imaging has been reviewed by a **neuroradiologist**
- Trial **symptomatic treatments**
- If a high clinical suspicion remains refer to a **specialist centre for MDT discussion** and further management
- Although limited evidence, consider up to two non-targeted lumbar EBPs
- Consider lateral decubitus myelography but yield of finding CVF is low (7-10%)

LEARNING POINTS

- When seeing headache patients, enquire about an orthostatic relationship
- SIH is highly disabling and has several serious complications
- Management of SIH should focus on identifying and treating the leak early rather than non-specific symptomatic measures
- High volume (>20ml) non-targeted EBP is first line treatment
- Experienced neuroradiologists with current myelographic techniques can now identify leak site in majority of (MRI positive) patients
- Patients who do not respond to one or two non-targeted EBPs should be referred to a specialist centre for dynamic myelography to localise the site of leak and facilitate targeted treatment
- Targeted treatment highly successful and SIH is curable in the majority of patients