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Review

Update on spontaneous intracranial hypotension

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Abstract

Background
Spontaneous intracranial hypotension (SIH) is a secondary headache disorder caused by spontaneous cerebrospinal fluid (CSF) leakage, typically from dural tears, meningeal diverticula, or CSF venous fistulas, in the absence of preceding trauma or procedures. Although increasingly recognized, SIH remains underdiagnosed and frequently mistaken for primary headache syndromes.

Methods
This narrative review synthesizes current evidence on the epidemiology, pathophysiology, clinical manifestations, neuroimaging features, complications, and management of SIH. Recent literature and expert consensus were analyzed to integrate advances in imaging-based diagnosis and leak type specific therapeutic strategies.

Results
SIH arises from low CSF volume rather than true hypotension, leading to brain descent, venous engorgement, and the characteristic MRI features summarized by the SEEPS mnemonic: subdural collections, pachymeningeal enhancement, venous engorgement, pituitary enlargement, and brain sagging. Orthostatic headache is the hallmark symptom, though up to 15% of patients lack clear positional features. Associated symptoms include vestibulocochlear disturbances, neck or interscapular pain, cranial nerve palsies, and cognitive or motor dysfunction. Brain and whole spine MRI are first line diagnostic tools. MR myelography distinguishes SLEC positive (types 1 and 2) from SLEC negative (type 3) leaks, guiding targeted myelography with dynamic CT or digital subtraction techniques for leak localization. Treatment should be tailored to leak type and site: epidural blood or fibrin patching for dural and diverticular leaks, and transvenous embolization for CSF venous fistulas. Surgical repair is reserved for refractory or anatomically complex cases. Reported complications include subdural hematoma, cerebral venous thrombosis, superficial siderosis, and bibrachial amyotrophy.

Conclusions
SIH represents a heterogeneous spinal disorder with intracranial manifestations. Early recognition, standardized imaging algorithms, and multidisciplinary management are crucial to improving outcomes. Ongoing research into imaging biomarkers, predictors of response, and consensus based care pathways will help advance precision medicine in SIH.

Keywords:
Spontaneous intracranial hypotension
Cerebrospinal fluid leak
Orthostatic headache
CT myelography
Epidural blood patch

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Introduction

Spontaneous intracranial hypotension (SIH) is a syndrome resulting from a spontaneous spinal cerebrospinal fluid (CSF) leak along the spine, typically caused by a dural tear or a CSF–venous fistula (CVF), in the absence of precipitating factors such as major trauma, spinal surgery, lumbar puncture, or spinal anaesthesia (1). The estimated incidence is around 5 cases per 100,000 persons per year, and the condition is more common in middle-aged women, with a female-to-male ratio of approximately 3:2 (2). First described in 1938, SIH remains a diagnostic challenge (3). Although the term "hypotension" is used, only about one-third of patients demonstrate a CSF opening pressure below 6 cm H_2O, because the underlying mechanism primarily involves reduced CSF volume rather than isolated low pressure (4,5).

Orthostatic headache is the most frequent symptom, followed by nausea, neck pain and stiffness, auditory symptoms (such as tinnitus or hearing loss), and dizziness (1). Patients with SIH may also develop acute complications such as subdural haematoma or cerebral venous thrombosis, as well as more delayed complications including bibrachial amyotrophy and superficial siderosis (2,3). Neuroimaging is central to the diagnostic evaluation of SIH, with brain MRI demonstrating the typical intracranial manifestations, spinal MR myelography helping to distinguish dural tears from CSF–venous fistulas, and advanced myelographic modalities such as dynamic CT or digital subtraction myelography (DSM) being used to localize the precise site of CSF leakage.

Management should be tailored to the type and site of the leak and may include epidural blood or fibrin patching, surgical repair, or transvenous embolization. Because SIH can be misdiagnosed or remain unrecognized, it should be systematically considered in patients with new-onset daily headache or chronic daily headache of unclear cause (6). Although orthostatic headache remains the hallmark, some patients present with atypical or weakly positional patterns, further complicating diagnosis (7–9). Clinician awareness and a high index of suspicion are therefore essential to ensure timely recognition and appropriate management, as SIH generally has a favorable prognosis when treated adequately.

Clinical presentation

Orthostatic headache occurs in up to 98% of patients with SIH and is defined as a headache that worsens in the upright position and improves when supine (7). No strict temporal criteria exist regarding the latency between positional change and symptom variation. Earlier diagnostic criteria (ICHD-2) required headache worsening within 15 minutes of standing, but this specification was removed in the ICHD-3, reflecting the variability seen in clinical practice (10).

Headache usually begins abruptly, although gradual-onset or even thunderclap-like presentations can occur. It is usually bilateral and posterior, though it may also be unilateral or diffuse, occurring in occipital, frontal, fronto-occipital, or temporal regions, and is commonly described as pressing, tightening, or pulsatile. When pulsatile headache is accompanied by photophobia, phonophobia, and nausea, SIH is frequently mistaken for migraine. In a proportion of patients, the orthostatic component diminishes or disappears over time, which may lead to diagnostic delay (11).

A recent cross-sectional study proposed a standardized definition for orthostatic headache in SIH: baseline pain intensity ≤3 (on a 0–10 verbal rating scale) upon awakening in the supine position; onset within ≤4.5 hours after standing; time to peak intensity ≤7.5 hours; and resolution or return to baseline within 1.5 hours after lying down. Nonetheless, up to 15% of patients may lack a clear orthostatic component (12).

Vestibulocochlear symptoms (e.g. dizziness, vertigo, imbalance, tinnitus, or distorted hearing) occur in about half of patients, and cervical or interscapular pain is reported in roughly one-third (7). Cranial nerve palsies—most often abducens (sixth nerve) and, less commonly, oculomotor or trochlear—may result in horizontal diplopia. Other manifestations include blurred vision, photophobia, visual field defects, facial numbness, galactorrhoea, labyrinthine hydrops, radicular symptoms, altered consciousness, parkinsonism, gait instability, bibrachial amyotrophy, chorea, and cognitive decline, including a phenotype mimicking frontotemporal dementia, termed Frontotemporal brain sagging syndrome (13).

Complications

Although CSF hypotension is commonly regarded as a benign condition, several clinically relevant complications may arise, posing diagnostic and therapeutic challenges. The most frequent complication is the development of subdural fluid collections, which may prompt neurosurgical consultation. Importantly, headache in these patients is often incorrectly attributed solely to the presence of subdural collections. Such collections may range from hygromas—reported in up to half of patients with CSF hypotension—to subdural haematomas, with or without mass effect. Clinicians should maintain a high index of suspicion for SIH in patients presenting with bilateral subdural haematomas in the absence of typical risk factors. In most cases, treatment of the underlying CSF hypotension leads to resolution of these collections, and surgical evacuation should be reserved for those with significant mass effect and associated neurological deterioration (1,14). A less common but potentially more serious complication is cerebral venous thrombosis (CVT), which occurred in <1% of patients in a 202-patient series (1). When present, full-dose anticoagulation is required, posing an additional therapeutic dilemma, particularly in patients undergoing invasive interventions such as epidural blood patch or surgery.

Delayed complications may also occur, including bibrachial amyotrophy and superficial siderosis. Superficial siderosis has been reported in approximately 3% of cases and is characterized by the classic triad of progressive hearing loss, ataxia, and myelopathy. Neuroimaging typically demonstrates posterior fossa superficial siderosis, which helps distinguish it from convexity siderosis associated with prior subarachnoid haemorrhage, traumatic brain injury, or cerebral amyloid angiopathy (15).

Bibrachial amyotrophy, reported in ~1.5% of cases, may clinically mimic Hirayama disease, presenting as a segmental lower motor neuron syndrome affecting the brachial plexus distribution. The risk of bibrachial amyotrophy and superficial siderosis appears to be particularly elevated in patients with prolonged disease duration, reaching nearly 60% at 16 years of follow-up (16).

Pathophysiology: a spinal disease with intracranial manifestations

Spontaneous intracranial hypotension results from three main mechanisms of spinal CSF leak, classified as types 1, 2, and 3 (17). Type 1 leaks are dural tears, most often ventral and often related to calcified disc protrusions or osteophytes that puncture the dura, leading to rapid CSF flow and typically producing a spinal longitudinal extradural collection (SLEC-positive). Type 2 leaks arise from lateral dural tears, where thin-walled arachnoid outpouchings through a dural defect allow CSF to escape; these have been reported to be more common in patients with heritable connective-tissue disorders. Type 3 leaks correspond to CSF–venous fistulas, in which a direct communication between the subarachnoid space and a paraspinal vein permits unregulated shunting of CSF into the venous system, usually without an extradural collection (SLEC-negative). CVFs often occur near nerve-root sleeves and may coexist with diverticula, supporting the role of local meningeal weakness or abnormal venous anatomy.

These three types comprise the principal mechanisms of SIH and form the basis for diagnostic imaging algorithms and targeted treatment approaches.

The clinical manifestations of SIH arise primarily from low CSF volume rather than isolated low CSF pressure (1,18). Loss of CSF volume causes downward displacement of the brain, exerting traction on pain-sensitive structures such as meninges, bridging veins, and cranial nerves, which leads to orthostatic headache and associated symptoms. According to the Monro–Kellie doctrine, the cranial cavity maintains a fixed total volume; thus, CSF loss is compensated by venous engorgement and dural hyperaemia, explaining imaging findings such as pachymeningeal enhancement and venous-sinus distension. This compensatory response also increases craniospinal elastance, reflecting altered intracranial compliance (19). With prolonged leakage, compensatory mechanisms become less effective, leading to subdural collections, pituitary enlargement, and sometimes structural brain descent, which may account for atypical or non-positional headache presentations.

Imaging and diagnostic work-up

Imaging plays a key role in the diagnosis and management of patients with suspected SIH, both to confirm the syndrome and to define the leak type and site. Whenever possible, all patients should initially undergo brain MRI and whole-spine MRI (20). In many cases, this will be sufficient to confirm the diagnosis of SIH and to suggest the type of spinal CSF leak involved.

Brain MRI is the cornerstone for detecting classic signs of intracranial hypotension (figures 1 and 2), often summarized by the SEEPS mnemonic: subdural collections (S), pachymeningeal enhancement (E), venous engorgement (E), pituitary enlargement (P), and brain sagging (S) (21). Protocols for brain MRI should ideally include post-contrast images, as they are most sensitive for identifying pachymeningeal thickening/enhancement. Where contrast is contraindicated, T2-FLAIR may provide similar sensitivity (22). MRI of the brain is positive in at least 80% of patients with SIH. However, in patients with chronic dural tears (types 1 and 2 leaks), brain findings may normalize, and a negative study should not be interpreted as absence of a leak (23). Moreover, pachymeningeal enhancement and brain sagging may also be absent in the first 1–2 weeks of symptom onset (24).

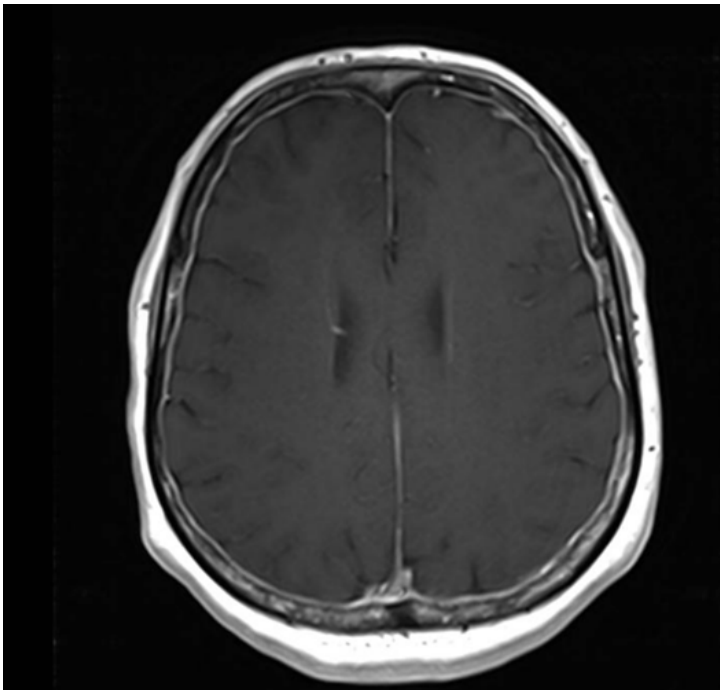

Figure 1. Axial T1-weighted post-contrast MR image of the brain shows diffuse regular pachymeningeal thickening and enhancement.

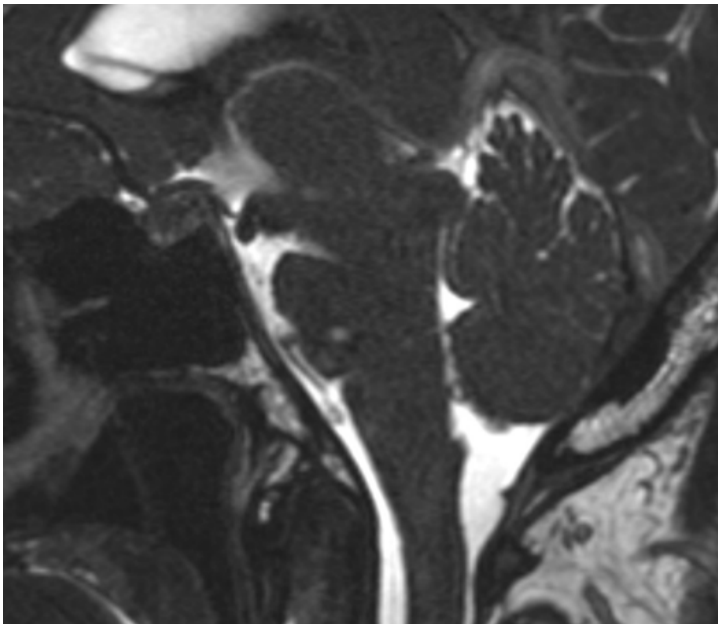

Figure 2. Midline sagittal steady-state MR image of the brain shows brain sagging, with descent of the optic chiasm, floor of the third ventricle and mammillary bodies, with reduction of the cisterns of the posterior fossa.

MR myelography (MRM) is a non-contrast MRI of the whole spine with heavily T2-weighted, fat-saturated sequences (figure 3). Its main purpose is to distinguish patients with SLEC-positive leaks (dural tears, type 1 or 2) from those with SLEC-negative leaks (typically CSF–venous fistulas, type 3), as only the former will exhibit epidural fluid, often called spinal longitudinal epidural collections (SLEC) (25). Identification of SLEC both confirms the diagnosis of SIH in cases where the brain MRI was normal and narrows the aetiology toward a dural tear. In many cases, MRM brings additional information that may help narrow down the type of leak (e.g. lateral/sacral) as well as the probable level. Ventral thoracic SLEC is often associated with ventral leaks, whereas predominantly posterior fluid may point to lateral or sacral leaks. The identification of a pseudomeningocele next to a nerve-root sleeve ("bud-on-branch" sign) has been shown to be sensitive and highly specific for localizing the level of a lateral leak (26). Early data also suggest that ventral leaks may be identified by the flow-void sign or by dedicated high-resolution sequences (0.5-mm isotropic CISS) (27,28).

Once the diagnosis is confirmed and the presumed leak type is known, invasive myelographic modalities are usually necessary to pinpoint the exact site of CSF leakage (figures 4, 5 and 6) (6). This is most often performed using dynamic CT myelography (dCTM) or digital subtraction myelography (DSM). Both involve lumbar puncture, often with the patient in the Trendelenburg position, followed by rapid image acquisition during and after intrathecal injection of iodinated contrast. In dural tears, the diagnostic yield is very high, although a minority of patients may require repeated studies in cases of initially equivocal findings (29). For suspected CSF–venous fistulas, yield is highly dependent on brain MRI findings (30). Patients with brain MRI positive for signs of SIH will have a CVF identified in around 70% of cases, with incremental yield on subsequent studies (31). Patients with normal brain MRI, on the other hand, have much lower rates of CVF identification (~10%). In this scenario, looking for ancillary imaging signs—such as decreased perioptic nerve-sheath diameter on coronal orbital MRI or the presence of multiple thoracic meningeal diverticula—may help select patients for invasive imaging (32,33).

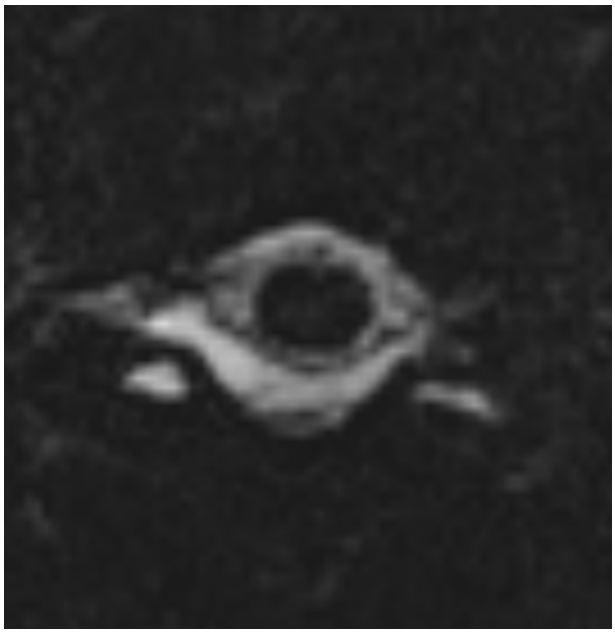

Figure 3. Axial T2-weighted MR myelography image shows circumferential spinal epidural fluid.

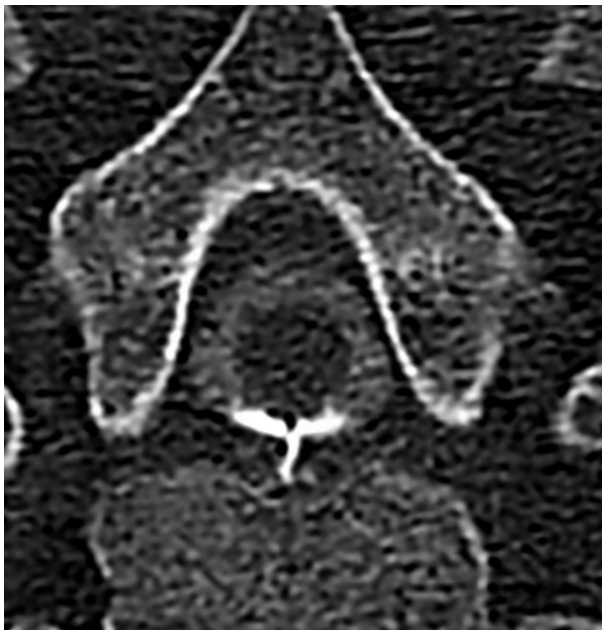

Figure 4. Axial dynamic CT myelography image in the prone position showing a ventral spinal CSF leak at the T2-T3 level, with active contrast leakage to the epidural space.

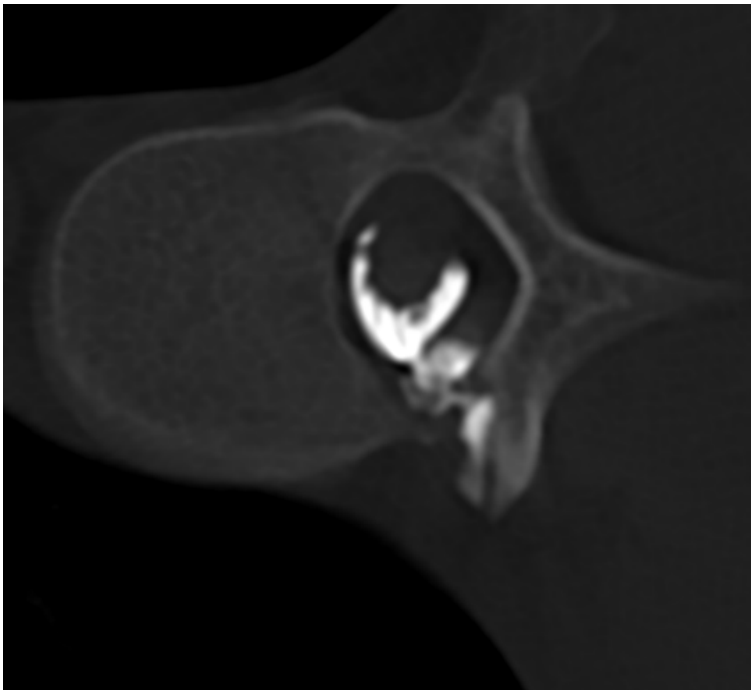

Figure 5. Axial dynamic CT myelography image in right lateral decubitus showing a lateral spinal CSF leak at the T11-T12 level, with active contrast leakage to the epidural space.

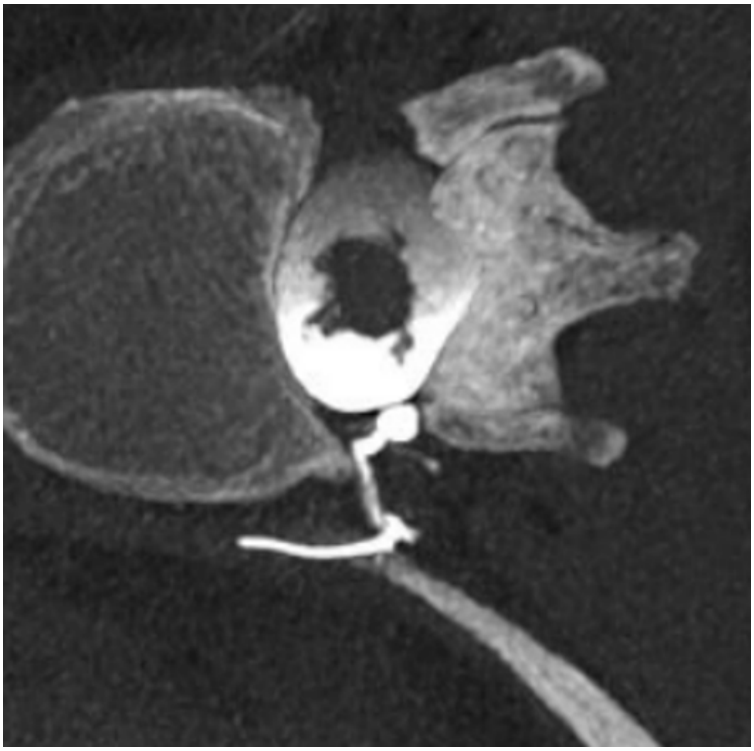

Figure 6. Axial maximum intensity projection (MIP) image of a decubitus CT myelography in the right lateral decubitus position showing a CSF-venous fistula on the right T10-T11 level, with opacification of intervertebral and paraspinal veins.

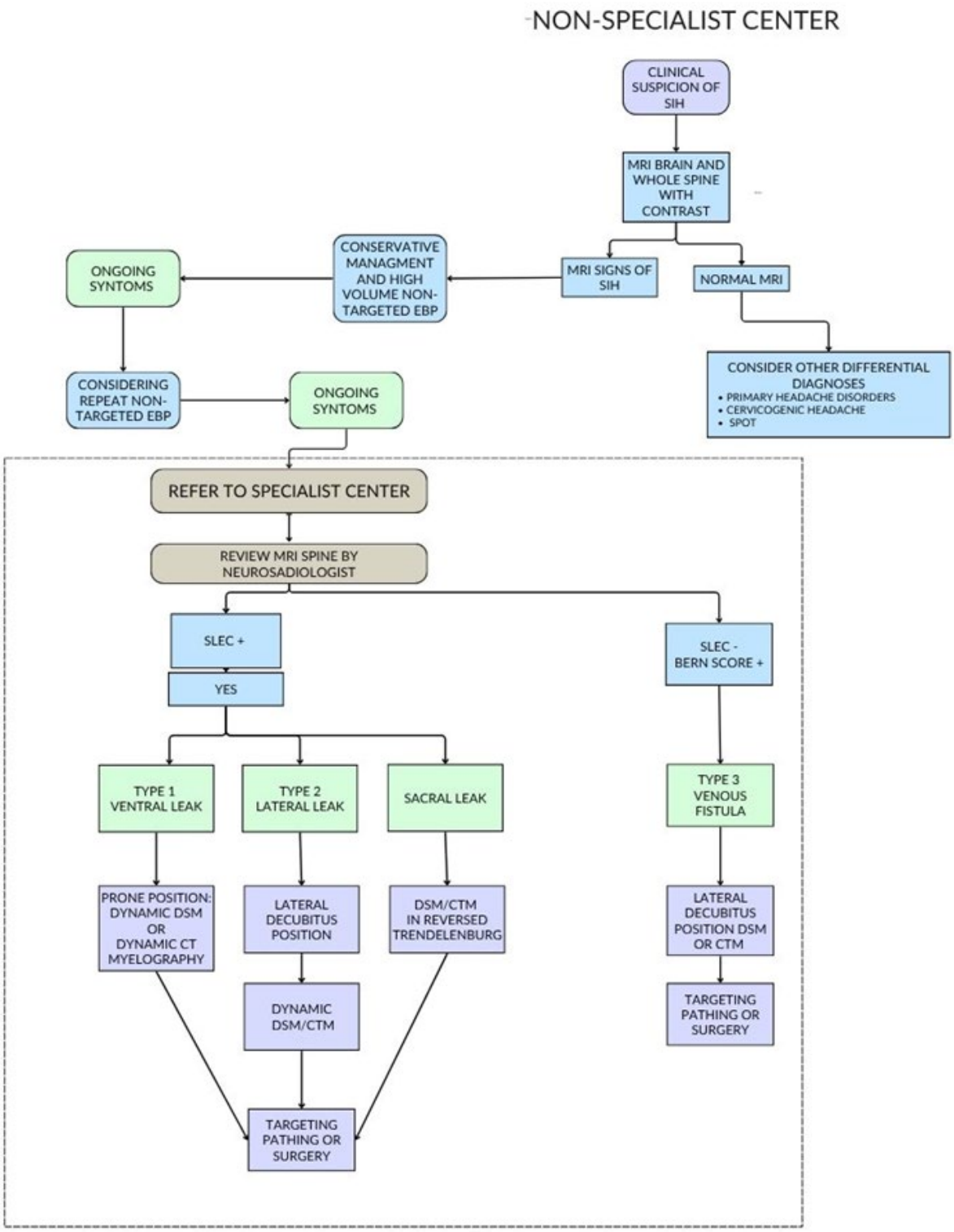

Figure 7. Algorithm for neuroimaging investigation in suspected spontaneous intracranial hypotension (SIH), illustrating a stepwise approach from initial brain MRI findings to advanced spinal imaging modalities. SLEC: spinal longitudinal extradural collection, DSM: Digital Subtraction Myelography, POTS: Postural Orthostatic Tachicardia Syndrome CTM: CT Myelography. Adapted from: Cheema et al.(34)

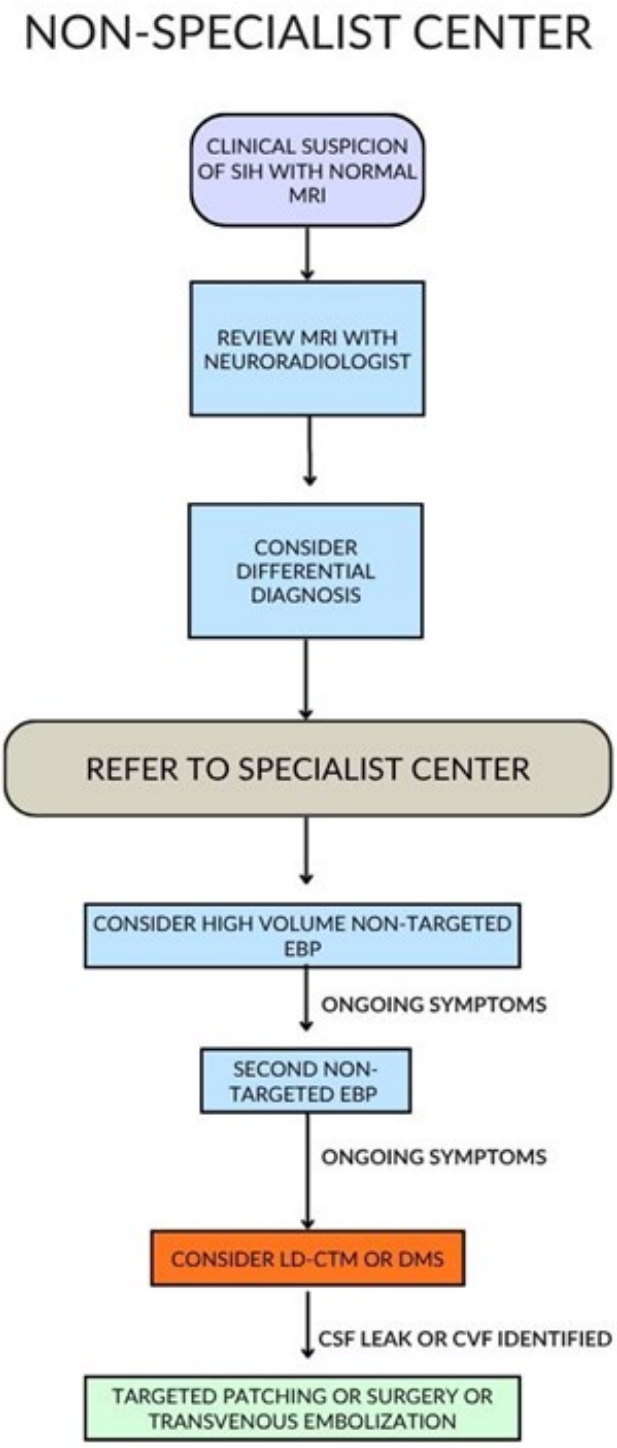

Figure 8. Algorithm for MRI-negative patients. EBP, epidural blood patch; LD-CTM, lateral decubitus CT myelography; LD-DSM, lateral decubitus digital subtraction myelography; MDT, multidisciplinary team; SIH, spontaneous intracranial hypotension. Adapted from: Cheema et al.(34)

Treatment options

Treatment for spinal CSF leaks is determined by the type and location of the defect identified on diagnostic studies such as myelography or MRI. The main therapeutic options include CT-guided epidural blood or fibrin patching, surgical repair, and transvenous embolization. When a large longitudinal epidural collection is present, the most likely cause is a type 1 or 2 leak (dural tear), and initial treatment usually consists of an epidural blood or fibrin injection (20,27). However, if an osteophyte is identified in association with a ventral tear, and EBP is not effective, surgical repair is often required for definitive treatment (20). In the absence of an extensive epidural collection, the mechanism is most likely a type 3 leak (CSF–venous fistula). For type 3 leaks, epidural blood patching is generally ineffective, and the treatment of choice in most cases is transvenous embolization (20,35,36). Surgical repair may be considered when embolization fails or is not feasible.

Epidural patches can be performed with autologous blood, and high-volume injections are preferred, as they allow dissemination to multiple spinal levels. Fibrin patches are reserved for focal leaks, leaks refractory to EBP, or for patients with contraindications to autologous blood. Empirical (non-targeted) patches have variable success rates (37–80%), but their efficacy is much lower in type 3 leaks (~14%) (7,35,37).

Given the considerable difference in prognosis after empirical treatment, a minimal diagnostic work-up with brain and spinal MRI should ideally be performed in all patients, whenever available. For those with positive brain MRI findings and a positive SLEC, the likelihood of a type 1 or type 2 leak is higher; therefore, an epidural blood patch may be performed even without precise leak localization. To improve efficacy, a minimum volume of 22.5 mL of autologous blood has been recommended (37). The most common adverse effects of patches include pain at the puncture site, radicular pain, and rebound intracranial hypertension. Patients who are refractory to non-targeted EBP should undergo CT myelography or DSM to identify the exact site of the leak, followed by a targeted EBP—with or without fibrin patch—or surgical repair, as indicated. After the procedure, the patient should remain supine for 2–24 hours, depending on institutional protocol. Activities that increase intrathoracic or intracranial pressure (e.g. Valsalva manoeuvres, heavy lifting) should be avoided for 4–6 weeks to minimize the risk of treatment failure (20).

In patients with positive brain MRI findings but negative SLEC, a type 3 fistula is the most likely mechanism, and further evaluation with CT myelography or DSM is strongly recommended to confirm the diagnosis. Transvenous embolization is the treatment of choice for most type 3 fistulas, with reported success rates of 84–95% (36,38). This procedure consists of endovascular occlusion of the involved vein by injecting a permanent embolic agent, yielding results comparable to surgery but with less morbidity and shorter recovery.

Surgical repair has been described for all three SIH mechanisms, with effectiveness in approximately 70–96% of cases (39,40). Microsurgical approaches are preferred because they provide adequate exposure with fewer complications. Reported complications include urinary retention, paraesthesia, haematoma, and lower-back pain.

Conservative management (bed rest, hydration, analgesia, caffeine, sometimes theophylline) should be offered to all patients, particularly in the acute or mildly symptomatic phase, but it should not delay or replace definitive treatment. Bed rest is only useful in the very acute phase (≤2 weeks); in chronic cases, it is counterproductive, as it promotes deconditioning (20,41). Oral hydration (2–2.5 L/day) is safe and may partially improve symptoms. SIH headaches often respond poorly to standard analgesics. Although NSAIDs and caffeine may provide partial relief in severe exacerbations, evidence for sustained benefit is limited (20). Peripheral nerve blocks can be considered as adjunctive symptomatic therapy, but they do not replace treatment of the underlying leak (42).

Conclusion and future perspectives

Spontaneous intracranial hypotension should be recognized as a syndrome encompassing multiple underlying pathologies rather than a single disease entity. This perspective captures the diversity of CSF-leak aetiologies, including dural tears, meningeal diverticula, and CSF–venous fistulas, each requiring a distinct diagnostic and therapeutic approach. Understanding this heterogeneity is crucial to improving outcomes.

The future direction of SIH management lies in personalized, imaging-driven strategies, integrating the clinical phenotype, imaging characteristics, and local expertise. Treatment should be guided by the suspected leak type and tailored to the individual. A multidisciplinary team—neurologists, neuroradiologists, anaesthesiologists/pain specialists, and neurosurgeons—is essential to achieve durable results.

Despite growing awareness, delayed recognition and diagnostic uncertainty are still common, often resulting in prolonged disability and impaired quality of life. The implementation of standardized diagnostic may help shorten diagnostic delays and improve prognosis. International registries and collaborative research networks will be crucial to align diagnostic criteria, collect real-world data, and evaluate long-term outcomes. Future studies should focus on validating imaging biomarkers, defining predictors of treatment response, and developing consensus-based clinical pathways.

Ultimately, embracing the syndromic nature of SIH and applying an individualized, multidisciplinary approach will pave the way toward more precise and timely care, with better quality of life for affected patients.

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