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Original research

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

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ABSTRACT

Background We aimed to create a multidisciplinary consensus clinical guideline for best practice in the diagnosis, investigation and management of spontaneous intracranial hypotension (SIH) due to cerebrospinal fluid leak based on current evidence and consensus from a multidisciplinary specialist interest group (SIG).

Methods A 29-member SIG was established, with members from neurology, neuroradiology, anaesthetics, neurosurgery and patient representatives. The scope and purpose of the guideline were agreed by the SIG by consensus. The SIG then developed guideline statements for a series of question topics using a modified Delphi process. This process was supported by a systematic literature review, surveys of patients and healthcare professionals and review by several international experts on SIH.

Results SIH and its differential diagnoses should be considered in any patient presenting with orthostatic headache. First-line imaging should be MRI of the brain with contrast and the whole spine. First-line treatment is non-targeted epidural blood patch (EBP), which should be performed as early as possible. We provide criteria for performing myelography depending on the spine MRI result and response to EBP, and we outline principles of treatments. Recommendations for conservative management, symptomatic treatment of headache and management of complications of SIH are also provided.

Conclusions This multidisciplinary consensus clinical guideline has the potential to increase awareness of SIH among healthcare professionals, produce greater consistency in care, improve diagnostic accuracy, promote effective investigations and treatments and reduce disability attributable to SIH.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ There is currently a lack of consistency and established treatment pathways for the investigation and management of patients with suspected spontaneous intracranial hypotension (SIH) due to cerebrospinal fluid (CSF) leak, and most of the published literature on SIH is from single centres with a dedicated service for patients with CSF leak and may be biased by local factors.

WHAT THIS STUDY ADDS

⇒ To the best of our knowledge this is the first multidisciplinary consensus-based guideline for SIH. It covers all aspects of the patient pathway from point of first presentation with suspected SIH to follow-up after treatment.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ This will directly influence clinical practice, produce greater consistency in care, improve diagnostic accuracy, promote effective investigations and treatments and thereby reduce disability related to SIH.

INTRODUCTION

Background

Spontaneous intracranial hypotension (SIH) is a highly disabling syndrome secondary to spinal cerebrospinal fluid (CSF) leak caused by a dural tear, leaking meningeal diverticulum, or CSF-venous fistula (CVF).[1] The estimated annual incidence of SIH is 3.7 per 100 000.[2] The symptoms of SIH resemble intracranial hypotension from other causes such as postdural puncture, postsurgical and post-traumatic CSF leaks, but in SIH the leak occurs spontaneously in the spine at a site which is unknown at the time of presentation. SIH is typically characterised by orthostatic headache and a variety of other neurological symptoms, and in approximately 80% of cases there are MRI features of intracranial hypotension.[3–5]

SIH can present in a variety of settings and to a variety of healthcare professionals and requires coordinated care between multiple medical specialties. Recent evidence suggests that the majority of patients with SIH respond to treatment with

non-targeted epidural blood patches (EBPs), and in the majority of patients with persistent symptoms, the leak can be localised with myelography in order to plan targeted patching, transvenous embolisation or surgery.[3] Despite this, several misconceptions exist in the investigation and management of SIH,[6] and SIH is often misdiagnosed or diagnosed and treated late prolonging a potentially treatable condition.[7 8]

Scope and purpose of the guideline

We aimed to create a multidisciplinary consensus clinical guideline describing best practice in the diagnosis, investigation and management of SIH due to spinal CSF leak, based on current evidence and consensus from a multidisciplinary specialist interest group (SIG), with representation from patients.

This document is intended to increase awareness of SIH among healthcare professionals, produce greater consistency in care, improve diagnostic accuracy, promote effective investigation and treatment and reduce disability related to SIH.

The guideline aims to address all aspects of the usual patient pathway from initial presentation with suspected SIH to follow-up after treatment, as well as several specific situations. The guideline does not apply to cranial CSF leaks, postdural puncture headache, post-traumatic or postsurgical spinal CSF leaks.

The intended target audience includes general practitioners, neurologists, radiologists, neurosurgeons, anaesthetists, pain specialists, emergency medicine specialists, physicians and other healthcare professionals who are involved in the care of patients with SIH.

METHODS

The guideline was developed and written in accordance with the international Appraisal of Guidelines, Research and Evaluation II instrument.[9] Figure 1 summarises the overall guideline development process. The guideline development process was initiated on the recommendation of the chief medical officer for England.

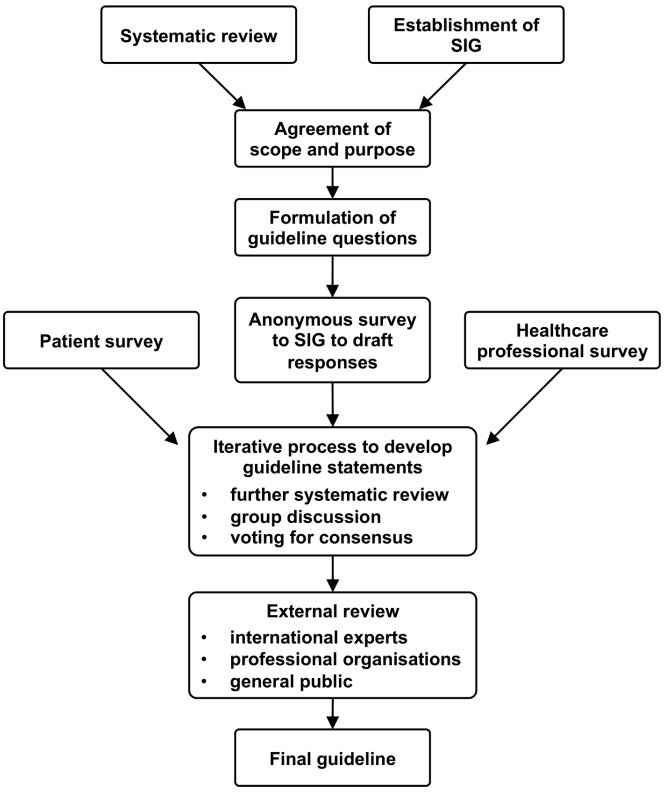

Figure 1 Flow diagram of guideline development process. SIG, specialist interest group.

Table 1 List of guideline question topics formulated by the SIG

Question No	Question
1	What key clinical features should lead to the diagnosis of SIH being considered?
2	What clinical mimics of SIH should be considered and how should the diagnosis be confirmed?
3	What predisposing conditions should be considered?
4	When and where should patients with SIH be referred?
5	What first-line investigations should be performed in patients with suspected SIH?
6	How should patients in whom there is a high clinical suspicion of SIH with normal brain and spine MRI be managed?
7	When should myelography be used in the investigation of SIH?
8	What myelographic strategies should be used in the investigation of SIH?
9	What is the role of intracranial pressure monitoring in the diagnosis of SIH?
10	What are the conservative and pharmacological management strategies that should be considered and for how long?
11	When should non-targeted epidural blood patches (EBP) be performed in the management of SIH?
12	How should non-targeted EBPs be performed?
13	What aftercare is recommended following epidural blood or fibrin sealant patching?
14	When and how should targeted patches be performed?
15	When and how should surgical management of a CSF leak be considered?
16	How should patients with imaging signs of SIH, but who are asymptomatic, be managed?
17	How should complications of SIH be identified and managed?
18	What is the best approach for headache management in SIH?
19	How should post-treatment rebound headache be identified and managed?
20	How should neurological symptoms other than headache in patients with SIH be identified and managed?
21	Is there a role for 'orthostatic rehabilitation' in the long-term management of patients with symptoms of SIH?
22	How should patients be followed up?

CSF, cerebrospinal fluid; SIG, specialist interest group; SIH, spontaneous intracranial hypotension.

The SIG who developed the guideline consisted of nine neurologists, six neuroradiologists, six neurosurgeons, two anaesthetists, one headache nurse specialist and five patient representatives (members of the UK-based CSF Leak Association charity). All medical professionals involved had regular clinical experience in the management of SIH, and all were asked to disclose any relevant conflicts of interest.

The scope and purpose of the guideline, and a series of question topics which the guideline was to address were agreed by the SIG by consensus (see table 1).

A systematic literature review was conducted for each of the questions, according to methods described by D'Antona *et al*,[3] and was updated to include studies published until November 2022, and to include question topics which had not been investigated in the previous publication. Patients were surveyed about their experience of diagnosis and management of SIH in the UK. A survey was also conducted of healthcare professionals outside of the SIG who were expected to be the target audience of the

Table 2 GRADE system for grading recommendations

Strength of the recommendation	Quality of the evidence
1=strongly recommended 2=weakly recommended	A=high quality: RCT(s) B=moderate quality: downgraded RCT(s) or upgraded observational study(s) C=low quality: observational study(s) D=very low quality: downgraded observational study(s)
Factors determining the strength of recommendations: ► Balance between desirable and undesirable effects ► Quality of evidence ► Values and preferences ► Costs of the intervention	Factors that may decrease QoE: ► Study limitations ► Inconsistency of results ► Indirectness of evidence ► Imprecision ► Publication bias Factors that may increase QoE: ► Large magnitude of effect ► Plausible confounding factors would reduce any demonstrated effect ► Dose–response gradient

GRADE, Grading of Recommendations, Assessment, Development and Evaluations; QoE, quality of the evidence; RCT, randomised controlled trial.

Table 3 Commonly associated symptoms and rare presentations of SIH*

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.

guideline. The results of both surveys were used to inform the guideline development process, and the results are published elsewhere.[8 10]

A modified Delphi process was used to develop recommendations for each question topic as follows: SIG members were initially asked to return anonymous draft responses to all guideline questions relevant to their area of expertise with relevant supporting evidence from the literature. Questions were then addressed in a series of five virtual meetings by presenting the anonymous responses, drafting proposed guideline statements based on these, discussion, anonymous voting on any area which did not meet consensus and, finally, voting by the whole SIG on each aspect of the proposed guideline statements. Where statements did not achieve consensus when first presented, they were discussed further among the SIG refined and voted on again. Guideline statements were only accepted for inclusion in the guideline if greater than 70% consensus was reached. The percentage of the SIG who accepted each included statement is shown in online supplemental material 1.

The strength of recommendations and quality of evidence for interventions were graded according to the Grading of Recommendations, Assessment, Development and Evaluations system (table 2).[11] Good clinical practice statements based on face validity and expert opinion (EO), where there is little available direct evidence but a high level of certainty that the recommendation would do more good than harm, were not graded but marked as EO. The evidence supporting each of the guideline statements and areas of uncertainty are also outlined for each question in online supplemental material 1.

Auditing and monitoring criteria were developed to assess rates of guideline implementation and adherence to recommendations (see online supplemental materials 2 and 3).

The first draft of the guideline was reviewed by several international experts (JB, PGK, WS, S-JW) and several UK-based professional bodies of relevant specialties, and underwent a publication consultation. Following this, further discussion and voting was held by the SIG members about any suggested changes before the final series of guideline statements were finalised. The final guideline was approved by the Association of British Neurologists and endorsed by the Royal College of Physicians.

GUIDELINE STATEMENTS

Q1. What key clinical features should lead to the diagnosis of SIH being considered?

SIH should be considered in any patient presenting with orthostatic headache (other than following iatrogenic dural puncture or major trauma); 'end of the day' or 'second half of the day' headache with improvement of the headache on lying flat (as defined below); thunderclap headache which is followed by orthostatic headache; and new daily persistent headache with an initial orthostatic quality. The presence of associated symptoms (see table 3) should increase the suspicion of SIH.

We recommend a working definition of orthostatic headache as headache which meets the following criteria:

- Absent or only mild (1–3/10 on verbal rating scale (VRS)) on waking or after prolonged lying flat.
- The onset of the headache occurs within 2 hours of becoming upright.
- After lying flat, the headache should have a 'good' improvement in severity (>50% on VRS) within 2 hours.
- The timing of headache onset and offset is consistent.

Q2. What differential diagnoses of SIH should be considered and how should the diagnoses be confirmed?

Differential diagnoses of SIH include postural tachycardia syndrome (PoTS), orthostatic hypotension, cervicogenic headache and migraine.

PoTS and orthostatic hypotension are diagnosed from a detailed autonomic history and haemodynamic autonomic responses to formal standing tests to document objective evidence of postural tachycardia (increase in heart rate by >30 beats per minute) or orthostatic hypotension (fall of >20 mm Hg in systolic blood pressure and/or >10 mm Hg in diastolic blood pressure).[12] A negative standing test does not exclude the diagnosis of PoTS and if clinical suspicion is high consider additional autonomic testing.

Cervicogenic headache (in the presence of cervical pathology) can be diagnosed with a history confirming that the headache is provoked by cervical movement rather than posture, reduced cervical range of motion and associated myofascial tenderness.

Migraine can be diagnosed with a history confirming that the headache is provoked by movement rather than posture, establishing migrainous biology, including history and trajectory

of episodes, presence of aura and vertigo (rather than hearing impairment and tinnitus).

Thunderclap headache presentations are most likely to be related to acute subarachnoid haemorrhage and its wider differential, of which SIH should be considered.

Q3. What predisposing conditions should be considered?

There may be no predisposing conditions to the development of SIH. The evidence identifying possible predisposing conditions is limited but enquiry may be made about connective tissue disorders and joint hypermobility disorders; and spinal pathology including osteophytes, disc herniation and discogenic microspurs in direct relation to the site of the spinal leak.

Q4. When and where should patients with SIH be referred?

Patients with suspected SIH should be referred to their local neurologist. If the patient is able to care for his or her self, the urgency of the referral should be 2–4 weeks, depending on the severity of clinical features including mental health impact. If the patient is not able to care for his or her self but has help, the urgency should be within 48 hours; and if they are not able to care for themselves and do not have help there should be an emergency admission. If the local neurologist does not have access to a practitioner skilled in performing EBPs they should be referred urgently to a regional centre with this expertise.

Patients should have early referral to a specialist centre if the diagnosis is in doubt, first-line treatments fail or there is a rapid clinical deterioration or serious complications such as subdural haematoma with mass effect (urgent referral to a tertiary neuroscience centre). For reasons other than rapid clinical deterioration, the time to assessment in a specialist neuroscience centre with expertise in SIH management should be within 1 month.

A specialist neuroscience centre should have the following services:

- Neuroradiological investigations and expertise including CT myelography (CTM) and/or digital subtraction myelography (DSM).
- Specialist clinical opinion, familiar and skilled in diagnosis and treatment of SIH.
- Practitioners skilled in epidural blood patching.
- Multidisciplinary team (MDT) meeting where patients with SIH are discussed.
- Expertise in performing targeted patching.
- Local guidelines for the use of fibrin sealant.
- Surgical expertise to repair a spinal CSF leak.

Q5. What first-line investigation(s) should be performed in patients with suspected SIH?

Ideally, MRI of the brain with intravenous contrast and MRI whole spine should be performed as first-line investigations. If not possible to achieve both at the same time, MRI of the brain with contrast should be performed as the first-line investigation.

MRI of the brain with contrast is essential to look for imaging signs that confirm the diagnosis of SIH (see figure 2). MRI of the whole spine is not always necessary for the diagnosis and is unlikely to locate the site of the CSF leak, but it can be helpful to identify the presence of findings that may direct subsequent invasive myelography.

If MRI is unavailable or if it is contraindicated, CT of the brain may show some of the findings supportive of the diagnosis.

Lumbar puncture should not routinely be performed for the sole purpose of confirming the diagnosis of SIH. If lumbar puncture is being performed for other reasons, such as to exclude alternative diagnoses, a CSF opening pressure should be measured at the time.

MRI of the brain protocol should include:

- T2 weighted (any plane) at 4–5 mm thickness or isotropic volume.
- Fluid-attenuated inversion recovery (axial or coronal) at 4–5 mm thickness or isotropic volume.
- T2*-weighted gradient echo (GRE) or susceptibility-weighted imaging (SWI) (axial) at 2–5 mm thickness.
- Precontrast and postcontrast 3D isotropic volumetric T1-weighted acquisitions OR T1-weighted spin echo at 4–5 mm thickness in the sagittal and one other plane.

Spine MRI protocol should include:

- Fat-supressed T2-weighted sequence such as short-tau inversion recovery (STIR) or other similar alternative.
- T2 weighted (sagittal) at 3–4 mm thickness in three parts.
- T2 weighted (axial) at 3–4 mm thickness of select segments of the spine.
- High-resolution steady-state or equivalent heavily T2-weighted 3D sequence (eg, constructive interference in steady state (CISS), fast imaging employing steady-state acquisition (FIESTA), balanced fast field echo (bFFE), Cube, or sampling perfection with application optimized contrast using different flip angle evolution (SPACE)) at a minimum isotropic resolution of 1 mm in three parts to cover the whole spine.

Q6. How should patients in whom there is a high clinical suspicion of SIH with normal brain and spine MRI be managed?

The presence of normal brain and spine MRI does not rule out SIH but is a recognised rare finding in patients with subsequently confirmed SIH. Ensure imaging has been reviewed by

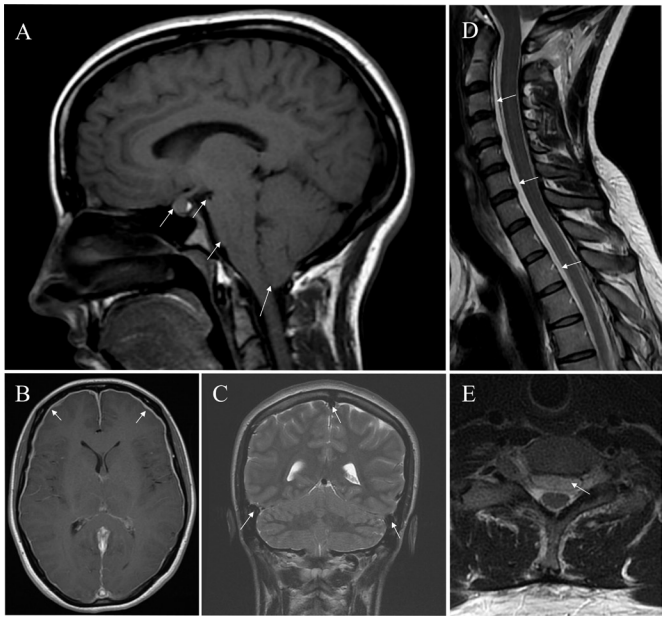

Figure 2 Typical MRI findings of spontaneous intracranial hypotension (SIH). (A) Sagittal T1 image showing enlargement of the pituitary, decreased mamillopontine distance, sagging of the brainstem and cerebellar tonsillar descent. (B) Axial T1 postcontrast image showing diffuse smooth dural thickening and pachymeningeal contrast enhancement. (C) Coronal T2 image showing distension of the dural venous sinuses. (D) Sagittal T2 image showing extensive ventral spinal longitudinal epidural collection (SLEC) extending from the upper cervical to thoracic regions. (E) Axial T2 image showing ventral SLEC.

a neuroradiologist and differential diagnoses have been considered. If a high clinical suspicion remains after consideration of the differential diagnosis and the imaging is confirmed as normal, then the patient should be referred to a specialist centre for MDT discussion and further management. Although there is limited evidence regarding the efficacy of performing empirical EBP in this context, up to two high-volume non-targeted lumbar EBPs could be considered.

Q7. When should myelography be used in the investigation of SIH?

The purpose of myelography in SIH is to locate the site of a spinal CSF leak in order to plan the targeted treatment.

It should be considered in any of the following scenarios:

- Patients who have at least one brain or spine MRI finding of SIH and have derived no benefit or only temporary benefit from one or more non-targeted EBPs.
- Patients who have normal brain and spine MRI, with meningeal diverticula, in whom the clinical suspicion of finding a CVF is high, and who have derived no benefit or only temporary benefit from one or more non-targeted EBPs.
- Patients who have normal brain and spine MRI, without meningeal diverticula, in whom the clinical suspicion is high and in whom myelography has been recommended after MDT discussion.
- If a patient is already under the care of a specialist MDT where myelography is available, and has not yet had a non-targeted EBP, the MDT may decide based on individual patient factors to proceed directly to myelography.

Q8. What myelographic strategies should be used in the investigation of SIH?

Myelography for spinal CSF leaks should be undertaken by a neuroradiologist with appropriate expertise and working as part of an MDT. The choice of myelographic technique (see table 4) depends on a number of factors, including whether a spinal longitudinal epidural collection (SLEC) is present or not, and the suspected underlying cause of the leak.

In patients with high clinical suspicion but normal brain and spine MRI, a CVF is the most likely cause of SIH. The likelihood of finding a leak in such patients is low, but decubitus CTM or lateral decubitus DSM are the recommended options.

Intrathecal gadolinium MR myelography lacks the temporal resolution of CTM and DSM and is not recommended as a first-line or second-line technique. It may sometimes be useful in cases of a suspected slowly leaking meningeal diverticulum when CTM or DSM has been negative. The use of intrathecal gadolinium is off-label and informed consent should be sought from patients for this.

Radionuclide cisternography has poor spatial and temporal resolutions and is not recommended as a tool for localising leaks.

Table 4 Selection of myelographic technique based on spinal MRI findings

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

CSF, cerebrospinal fluid; CTM, CT myelography; DSM, digital subtraction myelography; SLEC, spinal longitudinal epidural collection; UFCTM, ultrafast CT myelography.

It may rarely have a role in confirming the presence of a CSF leak in patients with normal brain and spine MRI in whom there is a high clinical suspicion of SIH but the above methods have all been negative.

Q9. What is the role of intracranial pressure monitoring in the diagnosis of SIH?

It is unclear whether intraparenchymal intracranial pressure monitoring has a role in SIH and it is not recommended as part of the standard clinical pathway.

Q10. What are the conservative and pharmacological management strategies that should be considered and for how long?

Conservative management should be discussed with all patients with suspected SIH and implemented for up to 2 weeks from symptom onset, while offering non-targeted EBP as soon as possible, if symptoms do not resolve with conservative management alone. Conservative measures recommended should include bed rest and hydration (2.0–2.5 L daily). Other strategies which may be recommended are use of abdominal binders and avoidance of Valsalva manoeuvres.

Measures to reduce the risk of deconditioning and risk of deep vein thrombosis should be advocated during the period of bed rest.

Though evidence for use of medication is sparse, oral caffeine or intravenous caffeine could be considered but this should not delay investigations or definitive treatment.

Q11. When should non-targeted EBPs be performed in the management of SIH?

A non-targeted EBP should be offered in all patients with a clinical and/or imaging diagnosis of SIH, after no more than 2 weeks of conservative management.

If there is no response or a transient response to the first EBP, a second EBP could be considered before proceeding to myelography.

The recommended time interval between EBPs (or following symptom recurrence in those with a transient response) should be 2–4 weeks.

Q12. How should non-targeted EBPs be performed?

Non-targeted EBPs should be performed by an experienced practitioner; under local anaesthetic; with the option of using conscious sedation; and with the option of using fluoroscopic or CT guidance to access the epidural compartment. A full discussion of the rationale for epidural blood patching including potential risks and complications must be held and the patient's informed consent must be documented. The practitioner should consider adjunctive preprocedural and/or periprocedural analgesia. Chlorhexidine skin preparation above 0.5% concentration should not be used.

As much blood as possible should be administered up to 40 mL, ideally at a minimum total volume of 20 mL. The administration of autologous blood should cease when the patient experiences back pain/pressure, headaches or radicular symptoms that they can no longer tolerate.

Q13. What aftercare is recommended following epidural blood or fibrin sealant patching?

Following targeted or non-targeted EBP or fibrin sealant patch, patients should be monitored in a recovery area and undergo basic physiological observations (heart rate, blood pressure and pulse oximetry) as well as spinal observations. A period of 2–24 hours bed rest and observation is recommended.

Following non-targeted blood patches patients should be either in the supine or Trendelenburg position. Following targeted patches patients should be in the supine position with head elevated as comfortable.

Thromboprophylaxis should be considered during immobilisation following EBP, according to local institution venous thromboembolism policy.

The patient should have a clinical review prior to discharge. If not admitted overnight, patients should be contacted the following day to exclude the presence of concerning features.

Patients should be advised to seek urgent medical attention should they develop any of the following: new-onset severe back or leg pain, lower limb motor weakness or sensory disturbance, urinary or faecal incontinence, urinary retention, perineal sensory disturbance, nausea and vomiting or fever. Advice regarding the possible symptoms of post-treatment rebound headache should be provided, including a change in the nature and site of headache.

Patients should not drive themselves home. Patients should be advised to lie flat as much as possible for 1–3 days after procedure. Patients should be advised to minimise the following for 4–6 weeks: bending, straining, stretching, twisting, closed-mouth coughing, sneezing, heavy lifting, strenuous exercise and constipation.

Q14. When and how should targeted patches be performed?

Targeted patches should be performed in patients who remain symptomatic following appropriate conservative management and/or non-targeted EBPs, in whom a causative lesion has been identified on DSM or CTM which is safely accessible via an image-guided transcutaneous approach.

The risks and benefits of image-guided patching should be discussed with the patient. Discussion may include risks/benefits of surgical management where appropriate.

Targeted patching should be performed by a consultant radiologist with appropriate training and experience in image-guided spinal interventional techniques in a neurosciences centre with local guidelines for the use of percutaneous fibrin sealant patching (off-label use/new procedure). This will usually be the neuroradiologist who has performed the myelography that demonstrated the spinal CSF leak/CVF. Exact technique will vary according to specific requirements of the leak type/site.

Q15. When and how should surgical management of a CSF leak be considered?

Surgical management of SIH should be considered in patients who remain symptomatic following appropriate conservative management and/or non-targeted EBPs in whom a causative lesion has been identified on DSM or CTM. The decision to offer surgery should consider the response to previous treatments, severity of symptoms, site and type of the leak or CVF, feasibility and risk of surgery and patient preference. The decision to undertake surgery (vs targeted patching) should be made after discussion involving the neurosurgeon, neurologist, neuroradiologist and patient.

Surgery should be performed by a neurosurgeon with expertise in managing spinal CSF leaks. Exact technique will vary according to specific requirements of the leak type/site.

If a CVF is shown on myelography, then endovascular treatment may also be considered as a first-line treatment (along with targeted patching and surgery).

Q16. How should patients with imaging signs of SIH, but who are asymptomatic, be managed?

Asymptomatic patients with radiological evidence of SIH should be referred to a specialist neuroscience centre and discussed in an MDT.

There is emerging evidence of potential significant long-term sequelae (particularly superficial siderosis) from persistent ventral spinal CSF leaks. This information should be discussed with asymptomatic patients.

Clinicians should discuss with patients and offer to investigate and treat asymptomatic spinal CSF leak with SLEC, in light of the potential long-term risks, particularly of superficial siderosis.

Patients who opt for a conservative approach should be offered a clinical review and repeat neuroimaging (MRI of the brain including SWI or GRE sequence and spine MRI) every 1–2 years.

Q17. How should complications of SIH be identified and managed?

Subdural haematoma

MRI of the brain with contrast and whole spine should be performed to investigate the possibility of spinal CSF leak in patients with subdural haematoma/hygromas where there is a high index of suspicion such as supportive history of orthostatic headache, or absence of trauma/coagulopathy/alcohol misuse.

Small or asymptomatic haematomas should be managed conservatively while treating the CSF leak. Symptomatic haematomas with significant mass effect may need burr hole drainage in conjunction with treating the leak.

Cerebral venous thrombosis

CT or MR venography should be considered in any sudden change in headache pattern or neurological examination in the context of SIH.

EBP should be prioritised as initial treatment of SIH with cerebral venous thrombosis. Addition of anticoagulation may be considered balancing the risks of bleeding complications on an individual basis.

Superficial siderosis

Patients with SIH undergoing MRI should have MRI of the brain and spine with blood-sensitive sequences which can detect superficial siderosis. A higher index of suspicion is needed in patients with SIH who develop ataxia, hearing loss or myelopathic features. CSF ferritin levels and xanthochromia may be measured.

Patients with SIH with siderosis should be managed in a specialist centre of expertise for this disorder. Symptomatic patients with superficial siderosis should be offered non-targeted EBP, or targeted treatment of the CSF leak site if detected on imaging. Deferiprone may be considered in symptomatic patients where the underlying CSF leak is unable to be found or treated.

Q18. What is the best approach for headache management in SIH?

Treatment of headache in SIH should focus primarily on management of the CSF leak, in tandem with best symptomatic management. Appropriate pain relief should be given as part of best symptom management. Paracetamol and/or non-steroidal anti-inflammatory drugs can be considered. Opioid medication may be required to provide adequate pain relief, but should be avoided in the routine long-term management of headache in SIH.

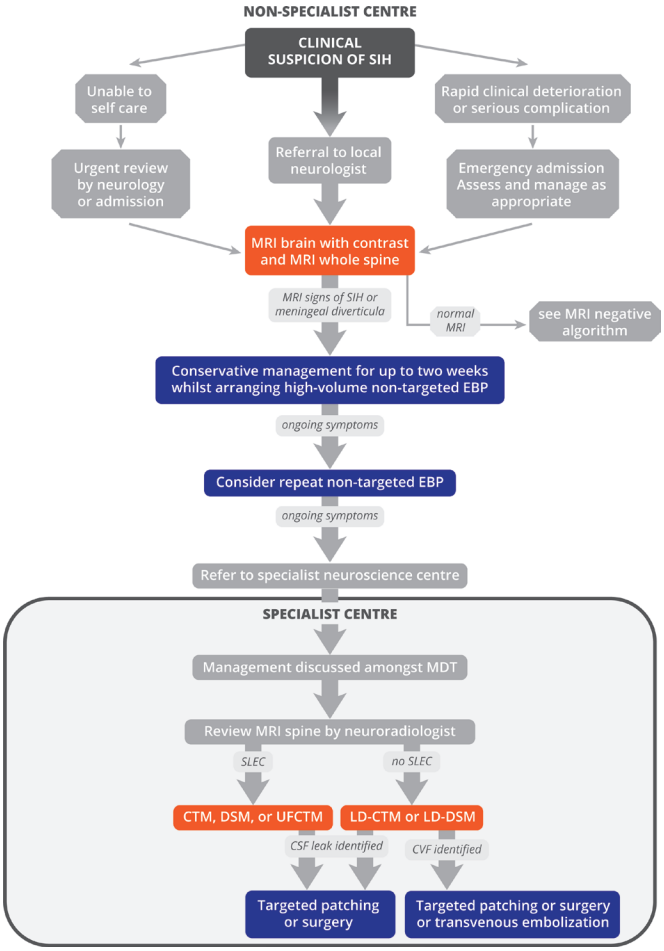

Figure 3 Algorithm for MRI-positive patients. This algorithm is designed to show the recommended pathway for most patients rather than capture every single possible situation which may occur in the management of a patient with spontaneous intracranial hypotension (SIH). CSF, cerebrospinal fluid; CTM, CT myelography; CVF, CSF-venous fistula; DSM, digital subtraction myelography; EBP, epidural blood patch; LD-CTM, lateral decubitus CT myelography; LD-DSM, lateral decubitus digital subtraction myelography; MDT, multidisciplinary team; SLEC, spinal longitudinal epidural collection; UFCTM, ultrafast CT myelography.

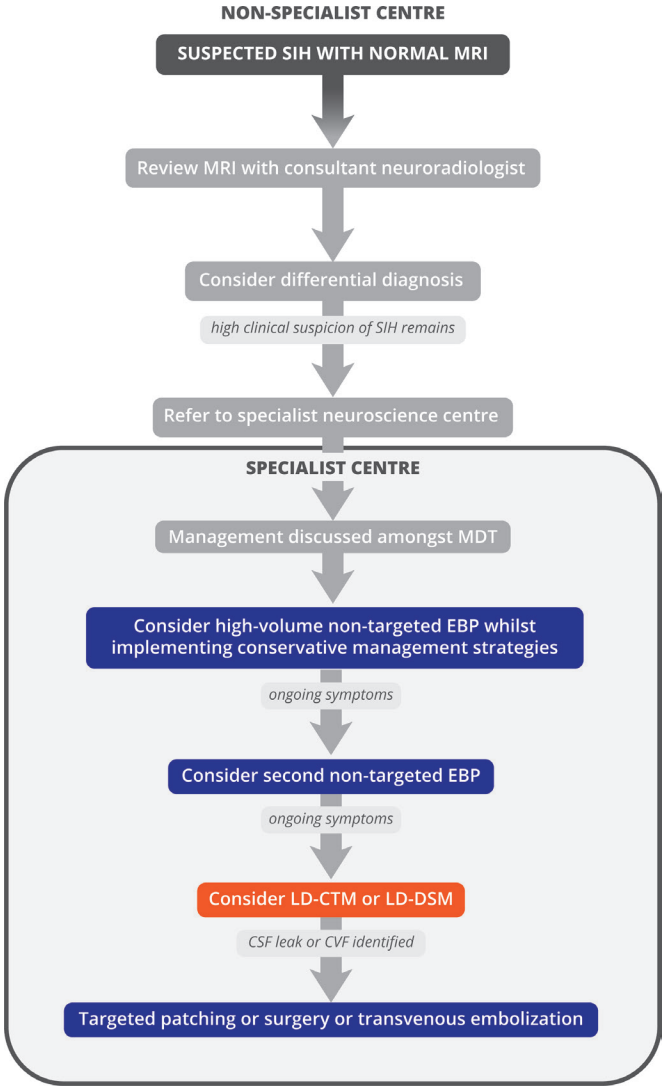

Figure 4 Algorithm for MRI-negative patients. CSF, cerebrospinal fluid; CVF, CSF-venous fistula; EBP, epidural blood patch; LD-CTM, lateral decubitus CT myelography; LD-DSM, lateral decubitus digital subtraction myelography; MDT, multidisciplinary team; SIH, spontaneous intracranial hypotension.

In patients not responding to initial management of SIH, it is important to look for comorbid primary headache and treat as per phenotype, and important to consider and warn patients about the risk of medication overuse headache. For management of associated primary headache, drugs that potentially lower CSF pressure such as topiramate and indomethacin, and migraine preventives that can reduce blood pressure such as candesartan and beta blockers should be used with caution, as they may exacerbate the postural symptoms of SIH.

Q19. How should post-treatment rebound headache be identified and managed?

Before an EBP, fibrin sealant patch or surgical repair of spinal CSF leak, patients should be informed about the entity of post-treatment rebound headache.

When rebound headache after treatment of SIH occurs, patients need to be evaluated for secondary intracranial hypertension. If very severe or worsening continues after 1–2 weeks further clinical review may be indicated. The development of rebound headache after treatment for SIH may indicate postprocedural intracranial hypertension which is self-limiting in most individuals and can often be managed without medical treatment.

There is anecdotal use of acetazolamide, topiramate and diuretics for rebound intracranial hypertension but these agents are not well tolerated and recommended treatment duration is not well defined in SIH treatment-related rebound headache.

Q20. How should neurological symptoms other than headache in patients with SIH be identified and managed?

Treatment of non-headache symptoms in SIH should focus primarily on management of the CSF leak, in tandem with best symptomatic management, for example, antiemetics for nausea and vomiting and encouragement of adequate hydration. Symptomatic management and advice on ways of coping with symptoms should be discussed with patients, while attempting treatment for CSF leak, but the evidence base for their use is lacking.

Q21. Is there a role for 'orthostatic rehabilitation' in the long-term management of orthostatic intolerance in patients with SIH?

Orthostatic rehabilitation should be considered for patients who have been bedbound, in particular those who have developed symptoms of orthostatic intolerance and patients with pre-existing PoTS and/or hypermobility syndromes. The rehabilitation programme should address both deconditioning affecting skeletal muscle and deconditioning affecting autonomic postural responses.

Q22. How should patients be followed up?

All patients (all types of blood patch, surgery, any person who has had therapeutic intervention) should be followed up clinically and should be given contact details for their responsible clinical team. We recommend follow-up at the following intervals:

- Early review for complications (following any intervention): 24–48 hours.
- Intermediate follow-up after EBP: 10–14 days.
- Intermediate follow-up after surgery: 3–6 weeks.
- Late follow-up (after any intervention): 3–6 months.

We recommend assessing for the following during follow-up:

- Peak headache severity on 0–10 scale.
- Time to severe headache onset after becoming upright.
- Severity of other symptoms, for example, audiovestibular/cognitive.
- Time able to spend upright before needing to lie down.
- Cumulative hours able to spend upright per day.
- Headache disability and quality of life outcome scores may be used; however, they are not validated for SIH.

In cases where there is no clinical improvement, or initial improvement with subsequent relapse following any intervention, it is recommended the patient is referred back to the MDT/specialist for discussion. Further imaging or intervention may be required.

In cases where there is a sustained long-term improvement, no further specialist/MDT involvement may be necessary. Further follow-up imaging to act as a baseline for any further imaging/treatment is at the discretion of the specialist who performed the procedure.

Repeat invasive imaging techniques should not be performed for the purpose of determining a baseline in patients who are asymptomatic or significantly improved.

DISCUSSION

We hope that this multidisciplinary consensus clinical guideline will lead to improved and more uniform pathways in the investigation and management of SIH in the UK, and potentially internationally, stimulating interest in the topic and highlighting future research questions. The guideline recommendations are supported by algorithms (figures 3 and 4) summarising the recommended pathway suitable for most patients. The guideline is intended to guide non-experts on the principles of management rather than serve as mandatory recommendations. Suggested auditing and monitoring criteria to aid implementation and adherence to the guideline are included as online supplemental materials 2 and 3.

To our knowledge a multidisciplinary consensus-based guideline for SIH has not previously been produced. Previously published algorithms for management of SIH are from single centres which may be biased by local factors, or do not cover the whole patient pathway.[13–16] We have also included aspects of SIH which were identified as especially important to patients including differential diagnosis, identification of comorbidities and symptom management.

Potential barriers to implementation of this guidance include the lack of provision for non-targeted EBPs, advanced myelographic techniques and targeted patching. However, we anticipate that the publication of this guidance will stimulate training and establishment of more widespread local services for these procedures. Non-targeted EBPs are commonly performed by obstetric anaesthetists for postdural puncture headache using the same technique which is employed in SIH. Myelography and targeted patching are limited to the smaller subset of patients who do not respond to first-line treatments and are provided by a small number of clinicians in specialist centres.

We recognise the limited evidence base for some of the recommendations. Hence, a modified Delphi method was used to develop the consensus guideline statements, and the guideline was reviewed by several international experts and professional bodies. We also recognise the recently expanding volume of SIH publications in the literature. Therefore, we plan to update the guideline regularly, with the next revision planned in 3 years' time.

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