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Indian Academy of Neurology

Advances in Headache Disorders

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CHAPTER 12

Treatment of Spontaneous Intracranial Hypotension

Ruby Chopra, Neelav Sarma, Ashish Kumar Duggal, Debashish Chowdhury

ABSTRACT

Spontaneous intracranial hypotension (SIH) is a rare and often underdiagnosed condition primarily caused by cerebrospinal fluid (CSF) leaks, typically near spinal nerve roots. Characterized by orthostatic headaches that improve upon lying flat, SIH presents diagnostic challenges due to its variable clinical manifestations and overlapping features with other headache disorders. Diagnosis typically involves a magnetic resonance imaging (MRI) of the brain and spine, with an emphasis on identifying CSF leaks through advanced imaging techniques such as dynamic computed tomography (CT) myelography and digital subtraction myelography. Conservative management, including bed rest, hydration, and analgesics, is initially recommended, with nontargeted and targeted epidural blood patches used for more persistent cases. Surgical intervention may be necessary for patients who do not respond to conservative or invasive treatments. Complications of SIH include subdural hematoma (SDH), cerebral venous sinus thrombosis, and other severe sequelae. While conservative measures are often a first step, most patients will need invasive therapies such as epidural blood patching (targeted or nontargeted) or surgery. Future research is required to refine treatment protocols and improve outcomes for SIH patients.

Keywords: Cerebrospinal fluid (CSF) leak, Orthostatic headache, Myelography, Epidural blood patch.

INTRODUCTION

Spontaneous intracranial hypotension (SIH) is an uncommon, rather underdiagnosed and very debilitating cause of headache. SIH is classically characterized by orthostatic headaches (OHs), most commonly secondary to a spontaneous cerebrospinal fluid (CSF) leak near spinal nerve roots. SIH was first described by Schaltenbrand in 1938.[1] However, the term SIH is a misnomer because the leaks are not spontaneous but are often triggered by trivial trauma in the setting of a weak thecal sac. Moreover, SIH is not a low-pressure state, but a low-volume state as described by Schaltenbrand who coined the term—"aliquorrhea" for the condition. The annual incidence is approximately 5 per 100,000, which is probably an underestimation.[2] There is an unmet need to diagnose and treat SIH optimally.

CLINICAL FEATURES

The SIH can present at any age, but the peak incidence is in middle age (35–55 years).[3]

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Females are affected twice to fifth as often as men.[4-6] Although headache is the most common presenting symptom of SIH, SIH can have several acephalgic presentations as well. Classically, OH (headache that starts or worsens on becoming upright and improves soon after lying flat) is seen in 92% of cases.[7] The headache is typically throbbing, predominantly bilaterally in the occipital region, posterior neck, shoulder, and interscapular areas. OH is characterized by mild pain (1–3/10 on the verbal rating scale) upon waking or after a prolonged supine position. The pain worsens or begins within 2 hours of becoming upright. After lying flat again, the headache should have a good improvement in severity (>50% on the verbal rating scale) within 2 hours. Notably the timing of headache onset and offset is consistent in a particular patient.[8,9]

However, patients with SIH may not always have OH and several different phenotypes can be seen in patients with SIH **(Table 1)**.[7,10] It is essential to determine the nature of the headache at the onset, as the orthostatic component of the headache may diminish over time, leading to transformed OH. The Trendelenburg test is a useful test to determine the orthostatic component of headache in patients suspected to have SIH. The patient is made to lie in the head down position (10–20°) for 5 minutes. A complete pain freedom or substantial pain relief in baseline headaches highly suggests OHs. There may be several associated symptoms in SIH, such as nausea, vomiting, vertigo, impaired hearing, tinnitus, neck and back pain, and cognitive impairment.[11] Patients may present with atypical manifestations such as cognitive decline, coma, peripheral

TABLE 1: Headache phenotypes seen in spontaneous intracranial hypotension.

Headache phenotype	Comments
Second half of day headache	Headaches that gradually worsen over the course of the day resulting in increasing pain in the afternoon and evening hours
	Occur presumably in slow CSF leaks or residual symptoms in a patient who has been partially treated
Wake up headache	Headaches that are present even before getting out of bed in the morning may be seen in a minority of patients
Transformed OH	Orthostatic headache that transforms into a nonpositional headache. This can occur in: • Later in the course of disease • Subdural hematoma • Cerebral venous thrombosis
Paradoxical headache	Headache worsens on lying down
Nonorthostatic daily headaches from start	SIH is an important diagnostic consideration in new daily persistent headache
Exertional and Valsalva-induced headaches	Valsalva maneuver exacerbate the CSF leak through spinal dural defects
Intermittent headaches	Often associated with intermittent leaks
Acute thunderclap headache	Acute thunderclap headache is followed by orthostatic headaches
Neck or interscapular pain preceding OH	It is essential to ask for the positional nature of headache at the onset of headache or neck pain

(CSF: cerebrospinal fluid; OH: orthostatic headache; SIH: spontaneous intracranial hypotension)

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TABLE 2: Atypical manifestations of spontaneous intracranial hypotension.

Atypical manifestations	Mechanism and comments
Spine pain	• Neck or interscapular pain, may be lower back pain • Can be orthostatic pain • The site of pain may not correlate with the site of leak
Nausea/emesis	• Nausea is much more common than vomiting • Both nausea and vomiting can be orthostatic
Vestibular manifestations	Dizziness, can have a Meniere-like presentation
Cochlear manifestations	• Nonpulsatile tinnitus • Muffled, distant, echoed, and distorted hearing
Diplopia	• Occurs because of 3rd, 4th, and/or 6th nerve palsies • Bilateral 6th nerve palsies more common than 3rd or 4th nerve palsy
Visual symptoms	• Visual blurring and field defects • Photophobia
Movement disorders	• Chorea • Tremor • Parkinsonism • Occurs due to pressure on basal ganglia or their connections
Bibrachial amyotrophy	Weakness and wasting of upper limbs may mimic ALS or Hirayama disease often seen in association with ventral longitudinal epidural spinal fluid collection
Gait unsteadiness	• Regardless of presence or absence of abnormal head sensation • Occurs probably because of spinal cord congestion, cord distortion or resultant deformity
Cognitive/behavioral symptoms	Frontotemporal dementia type of symptomatology occurs due to brain sagging
Other uncommon presentations	• Radiculopathy symptoms in upper limb • Facial numbness • Encephalopathy, stupor, and coma may occur because of brainstem descent or subdural hematoma

(ALS: amyotrophic lateral sclerosis)

and cranial neuropathies, venous sinus thrombosis, etc., usually in chronic cases **(Table 2)**.[12,13]

RISK FACTOR ASSESSMENT

While evaluating patients with suspected SIH, history and examination should focus on the potential risk factors for SIH, such as the history of connective tissue disorders and disc diseases such as herniation and spurs. Iatrogenic factors such as lumbar puncture, spinal anesthesia, bariatric surgery, etc., can also cause low-pressure headaches, but in these cases, the site of the CSF leak is known, and these are defined as postdural puncture headache or CSF fistula headache.[14] Management of these cases is different from SIH since the site of the leak is known.

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DIAGNOSIS

The SIH should be suspected in all cases with OH, new daily persisting headache (NDPH) and lingering chronic daily headache, or cervical/interscapular pain preceding OH by weeks. The presence of postural headache in different headache disorders such as SIH, cervicogenic headache, cough headache, postural orthostatic tachycardia syndrome, and motion sensitivity in migraine may lead to a diagnostic dilemma, thereby necessitating the use of objective criteria of the International Classification of Headache Disorders 3rd edition (ICHD 3) to establish the diagnosis of SIH **(Box 1)**. The ICHD requires demonstrating low CSF pressure (<6 cm CSF) and/or evidence of CSF leakage on imaging.[15] However, routine lumbar puncture is not necessary for diagnosis of SIH. CSF opening pressure (CSFop) may be normal in at least two-thirds of cases and >5 cm CSF in 80%. CSFop in SIH is directly proportional to disease duration and may occasionally be >20 cm CSF. Lumbar puncture may worsen the patient's headache. So, an MRI of the brain with gadolinium is the investigation of choice for all patients suspected of having SIH. MRI is abnormal in 80% of cases with SIH, so a normal MRI does not exclude SIH. **Figures 1A to F** shows the common MRI brain findings in patients with SIH. **Table 3** describes the findings seen in patients with SIH.[16-19] Various quantitative measures have also been described in patients with SIH **(Table 3)**. These can be useful in cases where the MRI appears normal. Bern Index is a score that classifies patients into low, intermediate, or high risk for SIH and can help guide further imaging and management.

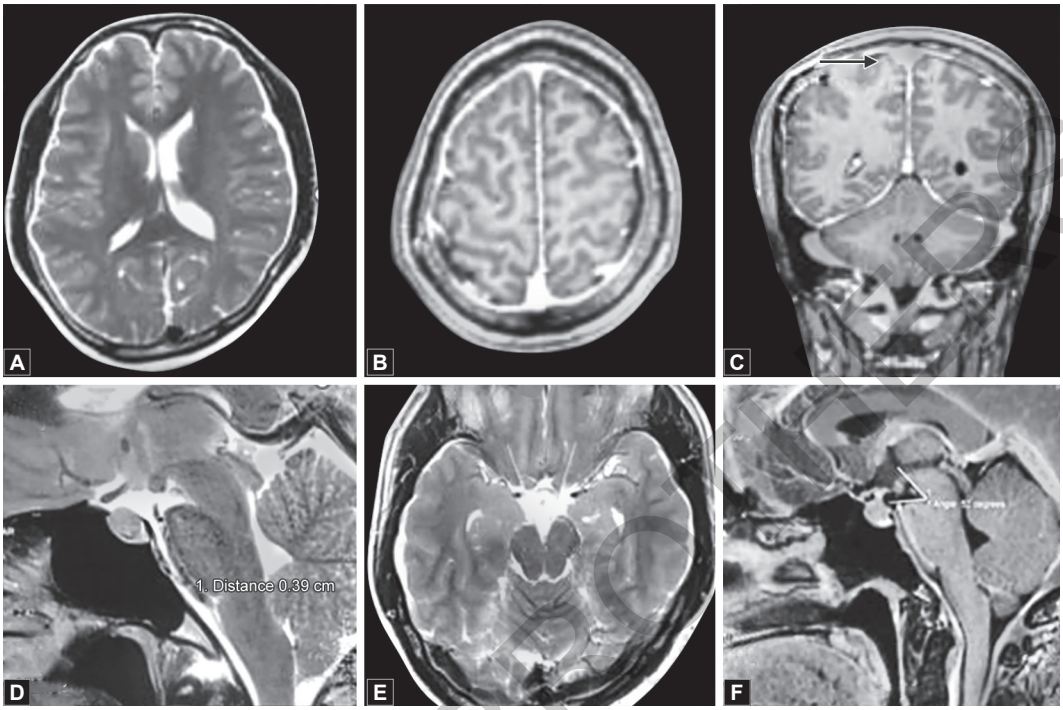
BOX 1: ICHD diagnostic criteria for spontaneous intracranial hypotension.

- Headache fulfilling criteria for 7.2 headache attributed to low cerebrospinal fluid (CSF) pressure, and criterion C below
- Absence of a procedure or trauma known to be able to cause CSF leakage
- Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage or has led to its discovery
- Not better accounted for by another ICHD-3 diagnosis

(ICHD-3: International Classification of Headache Disorders, 3rd edition)

In patients with imaging evidence of SIH or those with high suspicion of having SIH with normal MRI, the next step is to obtain a spinal MRI with heavily T2-weighted images and fat suppression. This noninvasive method can identify the presence of high-flow leaks (but not necessarily their location). Since an MRI of the brain may appear normal in 20% of patients with SIH, those with suspected SIH and a normal MRI should undergo a heavily T2-weighted MRI. Different types of leaks have been described in patients with SIH. CSF leaks can broadly be classified into spinal longitudinal extradural collection (SLEC) as positive or negative **(Flowchart 1)**. The spinal extradural longitudinal collection is an extradural collection of CSF visible on sagittal and axial scans, which indicates the presence of a CSF leak but cannot identify the site of the leak. Identification of SLEC has therapeutic and diagnostic significance. Patients with SLEC on spinal MRI are more likely to respond to nontargeted epidural blood patches. At the same time, SLEC indicates a high-flow CSF leak, most likely due to a dural tear caused by a discogenic microspur. This requires hyperfast CT myelography or digital subtraction myelography (DSM) to evaluate the site of the leak in case a targeted epidural blood patch is needed. On the other hand, in SLEC-negative patients, a CSF venous fistula is the most likely cause, and such patients require a lateral decubitus CT myelogram or lateral decubitus digital subtraction myelogram (on both sides). Dynamic CT myelography and DSM are most effectively conducted by skilled neuroradiologists in

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FIGS. 1A TO F: MRI findings in a patient with SIH showing subdural hygroma: (A and B) Diffuse pachymeningeal enhancement; (C) Engorged superior sagittal sinus and transverse sinuses (black arrow); (C) Reduced mamillopontine distance of 3.9 mm; (D) Reduced interpeduncular angle of 26°; (E) Reduced pontomesencephalic angle of 42°. (F) MRI spine has lower efficiency in localizing CSF leaks but may demonstrate the extent of ventral CSF leak, extradural CSF collection, dilated epidural and intradural veins, dural enhancement, and meningeal diverticulae.[20,21]

(CSF: cerebrospinal fluid; MRI: magnetic resonance imaging; SIH: spontaneous intracranial hypotension)

TABLE 3: Imaging findings in spontaneous intracranial hypotension.

Imaging finding	Comments
Smooth diffuse pachymeningeal enhancement	• Overall sensitivity = 83% • Not seen if MRI done too early (<7 days) or >45 weeks • Diffuse smooth enhancement of dura is almost pathognomic of low CSF pressure syndrome • Other conditions that involve the dura have a pattern that is either not smooth (nodular or plaque-like as in sarcoidosis or infections) or not diffuse (i.e., is localized or unilateral)
Venous distension sign	• High sensitivity (93.3%) and specificity (93%) • Look for the inferior border of transverse sinus in the parasagittal plane on T1 image • Normally, the inferior border is concave but in SIH the inferior border of the transverse sinus becomes convex

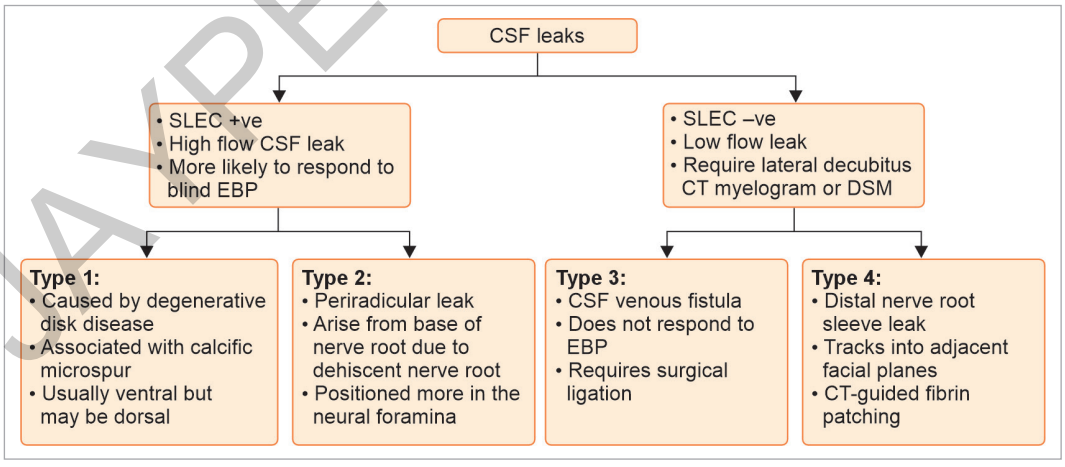
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Imaging finding	Comments
Brain sagging	• Seen in 61% of patients • Can be confused with Chiari malformation which is also associated with cerebellar tonsillar descent • Can be differentiated by measuring the mammilopontine distance
Subdural collection	• Seen in 20% of cases • SIH should be suspected in a patients with spontaneous bilateral subdural collections
Mammilopontine distance	• Assessed on midsagittal images between the inferior border of the mammillary body and the upper surface of the pons • <6.5 mm is characteristic of SIH
Interpeduncular angle	• Measured in the axial plane at or immediately below the level of the mammillary bodies • <40.5°
Pontomesencephalic angle	• It is the angle between two lines drawn along the anterior margin of the midbrain and anterosuperior margin of the pons. • <50° is suggestive of SIH.
Prepontine cistern	• Assessed on midsagittal images measuring the distance between the anterior aspect of the midportion of the pons and the clivus. • ≤5 mm is suggestive of SIH.
Suprasellar cistern	• Assessed on coronal postgadolinium T1-weighted MRI by measuring the distance between the inferior surface of the optic chiasm and the superior surface of the pituitary gland. • ≤4 mm is suggestive of SIH.

(CSF: cerebrospinal fluid; MRI: magnetic resonance imaging; SIH: spontaneous intracranial hypotension)



FLOWCHART 1: Classification of CSF leaks.

(CSF: cerebrospinal fluid; CT: computed tomography; DSM: digital subtraction myelography; EBP: epidural blood patch; SLEC: spinal longitudinal extradural collection)

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a specialized center with a high caseload. This presents a significant challenge in the localization of CSF leaks.

The site of CSF leak can be determined by myelography studies, namely MRI or CT myelography. Specialized spinal imaging in the form of DSM, dynamic CT myelography may be useful in detecting CSF fistula even in the absence of an evident epidural CSF collection. They have also been found to be useful in suspected SIH patients with normal MRI brain findings.[22] DSM is most successful at locating leaks (most commonly found in the thoracic spine—41%), or at multiple levels (24%).[23]

TREATMENT OF SPONTANEOUS INTRACRANIAL HYPOTENSION

Although conservative treatments can be tried, they usually do not succeed. A minority of patients often resolve symptoms spontaneously in 2 weeks, but even in these patients, attempts should be made to identify the site of the leak. Failure to fix the leak may result in complications that follow such as subdural hemorrhage, bibrachial amyotrophy, and superficial siderosis. The treatment should not only focus on alleviating symptoms such as headache, dizziness, tinnitus, and vertigo, but also on repairing any identified spinal CSF leaks. **Flowchart 2** shows a suggested algorithm for management of patients with SIH.

Conservative Treatment

Conservative treatment is the initial treatment offered to all patients with new-onset OHs. Literature suggests that conservative treatment is successful (resolution of symptoms with no further intervention needed) in 28% of patients.[7] Bed rest, hydration, and analgesia are the cornerstones of conservative treatment. Other modalities include use of methylxanthines (caffeine, theophylline), corticosteroids, and abdominal binders. Results of large meta-analysis[7] suggest that bed rest is the most effective conservative measure, beneficial in 88% patients. Hydration follows it, being effective in 83% patients. Use of oral or injectable analgesics, steroids, and caffeine was found effective in 27.4, 4, and 0.3% patients, respectively.

Bed Rest

Strict bed rest, avoidance of upright position, and Valsalva maneuvers such as coughing, straining, etc., are advised for several days to weeks in all cases. Bed rest should continue while awaiting results of investigations for localization of site of CSF leak and/or response to treatment. Recumbent position of the body is favorable for dura mater regeneration, improved CSF fluctuation, and shifting of the center of mass of brain upward.[24,25] Bed-bound patients may, however, develop symptoms of orthostatic intolerance, mild cognitive deficits, decreased reasoning and concentration, visual difficulties, blood pressure fluctuations, etc. This is more likely in patients with preexisting postural orthostatic tachycardia syndrome (POTS) or hypermobility syndromes. Deconditioned skeletal muscles and autonomic postural responses need to be addressed by orthostatic rehabilitation.[26] Deep vein thrombosis prevention with anticoagulants may be needed in patients on prolonged bed rest.[26]

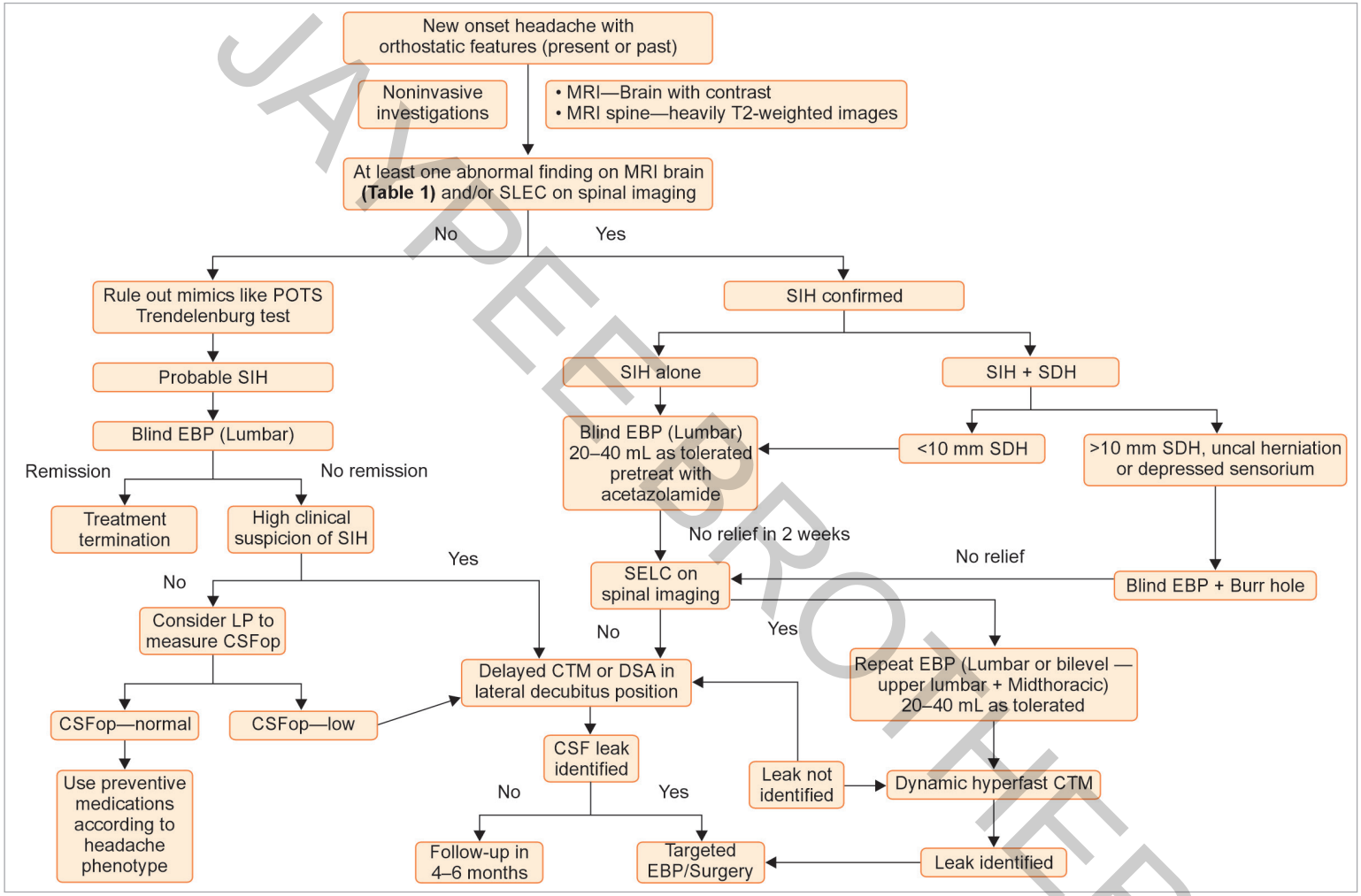
Hydration

Adequate hydration is advocated, at least 2–2.5 liters of fluids by oral route, and if needed, intravenous (IV) fluids.[26]

Analgesics

Appropriate pain relief should be tried as part of symptomatic treatment. Paracetamol and/or nonsteroidal anti-inflammatory drugs and antiemetics can be considered. Opioid medications are resorted to at times because most simple analgesics fall short of providing

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FLOWCHART 2: Algorithm for management of suspected SIH.

(CSF: cerebrospinal fluid; CSFop: CSF opening pressure; CTM: computed tomography myelography; DSA: digital subtraction angiography; EBP: epidural blood patch; MRI: magnetic resonance imaging; LP: lumbar puncture; POTS: postural orthostatic tachycardia syndrome; SDH: subdural hematoma; SIH: spontaneous intracranial hypotension; SLEC: spinal longitudinal epidural collection)

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adequate pain relief.[26] All analgesics and especially opioids are to be used with proper patient education about the potential of developing medication overuse headache. If a patient is on treatment for preexisting primary headaches, drugs such as topiramate and indomethacin are better avoided, as they tend to lower CSF pressure. β-blockers are to be used with caution, as these may exacerbate the postural symptoms of SIH due to the lowering of blood pressure.[26]

Methylxanthines (Caffeine and Theophylline)

Caffeine is a stimulant of central nervous system, produces cerebral vasoconstriction and reduces cerebral blood flow transiently. It is quickly absorbed after oral administration with a bioavailability of 100%. It freely penetrates the blood brain barrier (BBB). Inhibition of phosphodiesterase and adenosine antagonism are the mechanisms behind antinociceptive effects of caffeine.[27] Caffeine has been found effective in established cases of postdural puncture headache (PDPH); thus, the expected benefits of caffeine in SIH are another headache due to low CSF pressure.[28] The recommended dose is 300–500 mg of oral or IV caffeine preparation once or twice a day. A Cochrane review concluded that these doses were better than a placebo in treating headaches due to CSF leaks.[29] Caffeinated beverages are usually advocated; one cup of coffee contains 50–100 mg of caffeine, a black tea cup has 60–90 mg, and soft drinks contain 35–50 mg of caffeine.[30] The use of caffeine should not delay the diagnosis and treatment of the underlying spinal CSF leak.

Theophylline, another methylxanthine derivate can also be used to increase intracranial pressure (ICP). Venous dilatation is implicated in low CSF pressure headaches, hence explaining the efficacy of vasoconstrictor drugs like theophylline.[31] Recommended daily dose is 200 mg by oral or IV route. There is evidence in SIH patients, where sustained relief in postural headache occurred only with long-term theophylline administration.[32,33] A retrospective review of six studies concluded theophylline to be an effective treatment of headache in PDPH. Since the clinical phenotype of PDPH and SIH are similar, encouraging results are expected in headache relief in SIH as well.[34] A head on trial compared theophylline with combination of paracetamol-caffeine and placebo in the treatment of PDPH. A statistically significant change in pre- and posttreatment mean visual analog scale (VAS) scores was found in theophylline group only.[35] *Despite all the evidence available, theophylline should be used cautiously because it has a narrow therapeutic window.* It causes nausea, vomiting, diarrhea, insomnia, irritability and headache at plasma concentration (pc) >20 μg/mL, while cardiac arrhythmia, hypotension, hypoglycemia, and hypokalemia at (pc) >30 μg/mL. When theophylline exceeds a concentration of 40 μg/mL it can cause epilepsy, brain damage, and death.[36] Moreover, these medications only provide temporary relief and do not address the CSF leak.

Abdominal Binders

A tight abdominal binder raises the intra-abdominal pressure which is transmitted to the epidural space and may relieve the headache. They are not in much use in clinical practice due to abdominal discomfort.

Corticosteroids

Treatment decisions in SIH patients managed conservatively are individualized. Evidence for using steroids is not robust[37] and lack large clinical trials. A short-term high-dose IV corticosteroid therapy can be considered in selected immunocompetent SIH patients, refractory to initial conservative management, before trying epidural blood patch (EBP) or surgery.[38] In some case reports, remission

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of persistent headache in SIH patients was seen with high dose IV methylprednisone for 3 days, followed by tapering for 2 weeks. Patient responded significantly in 24 hours and remained asymptomatic at 1 month.[38] Resolution of symptoms has been reported with intramuscular dexamethasone (20 mg/d),[39] oral prednisolone (40–80 mg/d)[40-42] In some cases headaches subsided slowly in 2–4 weeks and radiological improvement occurred over 6–12 months.[43] A rare case of SIH, presenting with frontotemporal dementia, improved clinically and radiologically with oral prednisolone (80 mg/d).[44] Acute psychotic symptoms may develop in patients receiving high doses of corticosteroids.[45] Corticosteroids mediate the establishment of the loosened dural tight junctions, as indicated in vasogenic edema.[46] Improving brain edema, decreasing vascular leakage, and anti-inflammatory properties of steroids have been hypothesized as the possible mechanism of action.[47]

The duration for which conservative treatment has been attempted ranges from 7 to 9 weeks.[7] Since a large majority of patients do not respond to conservative measures alone, delaying EBP treatment beyond 2 weeks results in suboptimal treatment response. Therefore, it is advisable not to wait beyond 10–14 days for patients to respond to conservative treatment.[26] Early initiation of invasive treatments like EBP is recommended for greater likelihood of treatment response and to prevent the emergence of late complications like superficial siderosis.[48]

NONTARGETED EPIDURAL BLOOD PATCH

A nontargeted blood patch should be offered to all patients with a clinical and/or imaging diagnosis of SIH after no more than 2 weeks of conservative management. This procedure involves injecting autologous blood obtained from venipuncture and then injecting it into the lumbar epidural space.[22] Alternatively, blood can also be injected at two levels (lumbar and thoracic).[49] The injected blood, irrespective of the site of introduction, spreads through the entire epidural space and seals the site of the leak. An initial effect of rise in ICP occurs due to mass effect, resulting in a short-lasting initial improvement. Long-term benefits are believed to occur due to coagulation of blood at site of leakage and hence aiding in repair of dural tear.[50] This procedure can be performed by anesthetists or neuroradiologists and is done under local anesthesia or sedation. Blood is injected epidurally either blindly or under fluoroscopic/CT guidance. Following injection, patients are usually observed for around 2 hours for complications, either in the supine or in the Trendelenburg position.[11] Patients are advised to rest and lie flat as much as possible for a period of 24–72 hours and avoid the Valsalva procedure for 4–6 weeks after the procedure.[11] Response to treatment is usually assessed at the end of 2 weeks. A repeat injection is considered in 2–4 weeks if there is no response after the first patch.[11] Adverse effects include back pain, radicular pain, tinnitus, paresthesias, numbness, bradycardia, and dizziness which are usually transient.[7] Rare, more serious side effects include dural puncture, cauda equina syndrome, spinal cord compression, and spinal arachnoiditis.[11] Another potential complication following any form of epidural patching is rebound intracranial hypertension, reported in 7–27.4% of the patients.[7] Some patients need acetazolamide to correct rebound headache and ICP.[7]

A meta-analysis found that single sitting of blood patching is successful in 64% of patients [95% confidence interval (CI) 56–72%].[7] Large volume patches (>22.5 mL) are more effective than smaller ones (<22.5 mL, 67.9% vs. 47%, $p = 0.01$).[51] It is recommended to inject a volume between 20–40 mL as tolerated by the patient.[2] Pretreatment with acetazolamide

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can decrease the CSF volume and allow the injection of a larger blood volume. Important predictors of response include anterior epidural CSF collection length (<8 segments vs. 8 or more segments; 72.5% vs. 37.3%, $p < 0.001$) and midbrain-pons angle (<40° vs. >40°, 71.3% vs. 37.5%, $p < 0.001$).[51]

A recently published prospective cohort study analyzed the efficacy of epidural blood patch in 69 patients with symptomatology suggestive of SIH but had a normal MRI brain and spine. They found clinically significant benefits in headache, neck pain, fatigue, and global physical and mental health without significant adverse effects.[52] They recommend epidural blood patch should be offered to patients with OHs and normal imaging after ruling out other diagnoses such as POTS, cervicogenic headache, etc.[53] At the same time, patients with an MRI (brain or spine) suggestive of SIH without symptoms should be offered an EBP because of the potentially devastating long-term complications.

Nontargeted patching may be repeated at 2–4 weeks with albeit lower responsive rates. A patient who has not responded to two sittings of nontargeted patching should be considered for imaging for localizing site of leak and subsequent targeted therapies.[54]

TARGETED PATCHING

This procedure is recommended for patients who do not respond to conservative treatment or nontargeted blood patches. This is possible after accurate localization of the site of the CSF leak by dynamic CT myelography or lateral decubitus DSM, depending on the presence or absence of SLEC. Targeted patching can be performed with blood, fibrin glue, or their combination.[54] An open label study compared 31 patients of targeted blood patching with 25 patients who received blind epidural patching, and found significantly better efficacy in the former group (87.1% vs. 52%, $p < 0.05$).[55] However, another open label study which compared 28 patients of blind epidural patching with 88 patients of targeted patching found similar level of efficacy in the two groups.[56] The requirement of subsequent blood patching was also similar in the two groups.[56] Epidural fibrin glue injection can be considered in patients who do not respond to epidural blood patch therapy.[4] It has been used successfully as targeted therapy in patients with dural CSF leaks and with CSF-venous fistula.[57,58] Potential adverse effects of targeted patching akin to nontargeted therapy include spinal cord compression and chemical meningitis.[54] Fibrin glue injections can be associated with aseptic meningitis, arachnoiditis, and risk of embolization when used in treatment of CSF-venous fistula.[54]

SURGERY

Surgical treatment is recommended in patients who do not respond to conservative treatment, fail to improve after at least two epidural patches, and have an identifiable site of CSF leak on myelogram.[54] In certain scenarios, such as in CSF venous fistula, surgical modalities may be considered at an earlier stage due to rather ineffectiveness of nontargeted epidural patching in these groups of patients. Surgical modalities include ligation of diverticula; repair of leak by placing absorbable gelatin sponge, fat, or muscle; aneurysm clipping of fistula; electrocautery to ligate fistula.[59] A prospective cross-sectional study of 20 patients with SIH due to CSF fistula who underwent surgery found 83% patients to have major improvement in headache severity.[60] All patients reported clinically significant levels of satisfaction with treatment, while 90% reported highest level of satisfaction.

Another prospective unblinded case series of 69 surgically treated SIH patients found 94.2% to have partial improvement

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and 52.2% to have complete resolution of symptoms.[48] The authors found duration of symptoms to be an important predictor of outcome with a cutoff of 12 weeks, procedures done before this time interval had better results.[48] Counterintuitively, more intense preoperative headache patients had a better outcome, which has been attributed to a more aggressive presentation resulting in earlier diagnosis.[30]

Statistically nearly two-thirds of patients with SIH shall require invasive treatment at some point of time. Lack of randomized controlled trials makes it difficult to formulate an optimum, universally applicable pathway. Although targeted blood patching looks promising scientifically, it is yet to be proven superior to blind patching in a robust way and the response depends on the type and site of leak. Surgical therapy will be effective in most of the cases; however, it shall have a higher rate of success when done at an earlier stage (<12 weeks duration). This emphasizes the importance of not only quickly diagnosing SIH but also giving equal weightage to localizing the cause of a leak in nonresponding patients.

TREATMENT OF COMPLICATIONS OF SPONTANEOUS INTRACRANIAL HYPOTENSION

Known complications include SDH, cerebral venous sinus thrombosis, superficial siderosis, syringomyelia, and frontotemporal dementia like syndrome.[26]

- *Subdural hematoma:* Asymptomatic or small hematomas are managed by addressing the primary condition. Significantly large hematomas and mass effects may require drainage.[11] It is essential to note that in patients with SDH, nontargeted epidural patch should precede the drainage because of the potential for worsening of SIH after SDH drainage.
- *Cerebral venous sinus thrombosis:* EBP should be considered in these group of patients of SIH.[11] Patient will also require anticoagulation which should be started after assessing potential risks of bleeding.[11]

CONCLUSION

Treatment of SIH requires a multidisciplinary approach. Whereas around 28% of patients respond to conservative treatment alone, the majority of others require invasive therapy. Around two-thirds of these patients will improve with a single epidural blood patching. Surgical repair is needed for the rest of the patients, who have a higher rate of success when done at an earlier stage. This requires early diagnosis and localization of pathologic site. Future randomized trials will help in determining optimal duration of conservative treatment while balancing against the potential risk of delaying interventional therapy resulting in less favorable outcomes.

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Advances in Headache Disorders

Advances in Headache Disorders is one of the publications released by the Indian Academy of Neurology (IAN) during the IANCON 2024 in Visakhapatnam. This specialized text focuses on the current treatment guidelines and algorithms for various headache disorders. Written by a team of stalwarts in neurology and headache medicine, the book includes contributions from leading practitioners and researchers, ensuring that the content is authoritative and relevant. This book is a practical guide for clinicians seeking to enhance their approach to diagnosing and treating headache disorders, leveraging the most recent advancements in medical research and clinical practice. It offers evidence-based recommendations for pharmacological and nonpharmacological interventions. The book features flowcharts, tables, and diagrams that simplify complex treatment protocols and facilitate quick reference during clinical practice. This book is intended for healthcare professionals involved in diagnosing and treating headache disorders, including neurologists, headache specialists, and general practitioners. It also serves as an essential resource for medical students and researchers focused on innovative treatment methodologies.

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