



Spontaneous Intracranial Hypotension: A Review of Neuroimaging and Current Concepts

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Abstract

Spontaneous intracranial hypotension (SICH) is the emerging cause of orthostatic headache as it has been better recognized in recent years. SICH happens due to spinal cerebrospinal fluid (CSF) leak; however, the manifestations are predominantly cranial and hence imaging in SICH includes brain and spine. There are few characteristic brain imaging features to be concerned about to diagnose SICH in patients with vague symptoms or low clinical suspicion. Spine screening is recommended in these patients to assess spinal CSF leaks. While neuroradiologists play a significant role from the time of diagnosis to treatment of SICH, there is a need for all the general radiologists to be aware of the condition. Computed tomography myelogram and digital subtraction myelogram are performed for diagnostic and therapeutic management of SICH. There is a known risk for SICH recurrence in patients with sagittal longitudinal epidural collection and hence, targeted blood patch should be used instead of blind patch. Most importantly, slow mobilization is recommended following the patch to avoid recurrence.

Keywords

- ▶ spontaneous intracranial hypotension
- ▶ CT myelography
- ▶ orthostatic headache

Introduction

Spontaneous intracranial hypotension (SICH) is a clinical condition resulting from reduced intracranial pressure that causes imbalances between the blood, cerebrospinal fluid (CSF), and the brain parenchyma. This occurs more commonly secondary to spinal CSF leak into the epidural space or to adjacent venous channels. Even though the name suggests intracranial hypotension, hypotension is not always demonstrated on lumbar puncture and the CSF leak at the skull base rarely results in intracranial hypotension and hence it should be renamed as spontaneous spinal CSF leak syndrome. SICH can also be termed as CSF hypovolemia,

CSF volume depletion, and low CSF volume headache. Spinal CSF leaks are increasingly recognized in recent times due to improved imaging techniques and detection with increasing awareness.

The pathophysiology is understood by Monro-Kellie doctrine where the volume of CSF, blood, and brain parenchyma remains constant in dynamic equilibrium. For instance, in patients with CSF leak, there is reduction in the CSF volume, which results in compensatory dilatation or engorgement of the intracranial and epidural venous structures to maintain the intracranial pressure causing clinical symptoms accordingly (**Chart 1**). Buoyancy of the brain is also reduced in leak patients.¹ Reduced CSF in the brain causes stretching of the

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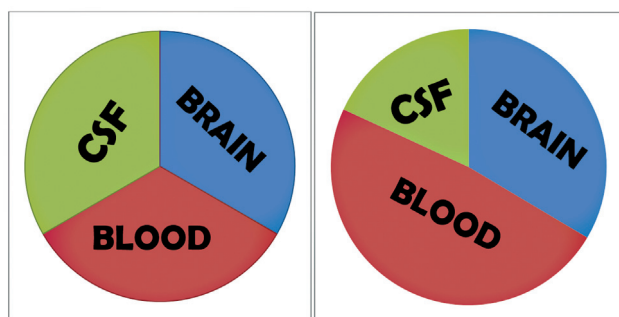


Chart 1 Monro-Kellie hypothesis. CSF, cerebrospinal fluid.

pain sensitive meningeal structures and subsequent venous engorgement results in headache.

The knowledge of physiology of normal intracranial pressure is necessary to understand the mechanism of hypotension in SICH.

patients. There are two points of neuroaxis that play major roles in maintaining the equilibrium. These include hydrostatic indifference point (HIP) and zero CSF pressure in sitting (ZPS). HIP is a point located in the lower cervical or upper thoracic region spinal region, corresponding with the venous pressure at atmospheric level. At this level, the pressure recorded in sitting and lying down position is same. ZPS is located between occipital protuberance and spinous process of seventh cervical vertebra. CSF pressure is negative above ZPS level and positive below this level.

The physiology of orthostatic headache in SICH is that when the person is upright, the CSF pressure is more than the atmospheric pressure. In patients with spinal CSF leaks, pressure point moves downwards, resulting in negative intracranial pressure compared to the spine. However, on lying down, headache resolves on lying down as the pressure attains equilibrium with the point coming back to the normal position.^{2,3} This is the same reason why skull base leaks do not result in symptoms of SICH.

Clinical Features

SICH is the one of the most important and under diagnosed causes of daily disabling headache with an incidence of 1 in 50000. Underdiagnosis of SICH has led to chronic neuro-disabilities despite being a treatable condition. Females are more commonly affected than men. Headache in SICH worsen over the second half of the day and may also worsen during Valsalva maneuver. Predisposing factors include connective tissue disorders causing meningeal disruption, discal spur, and marginal osteophyte.

SICH is an elusive disorder and can mimic many other conditions. The commonest clinical presentation is orthostatic headache, which aggravates against sitting or standing and relieved on lying down. In rare and severe cases, patient may present with dementia, symptoms of brain and/ or spinal cord herniation.^{4,5} ▶Table 1 summarizes the symptoms or presentations of SICH.

Differential diagnoses include orthostatic hypotension, cervicogenic headache, postural tachycardia syndrome,

Table 1 Clinical presentations in SICH patients

Back/neck pain	Paresthesia/ facial numbness
Vomiting/nausea	Hemifacial spasm
Hearing loss	Personality changes
Tinnitus, vertigo	Gait ataxia
Muffled hearing	Frontotemporal dementia
Photophobia	Encephalopathy
Diplopia/blurred vision	Parkinsonian symptoms like tremors/ chorea
Isolated cranial nerve palsies	PRES/CVT/SDH
Amyotrophic symptoms like amyotrophic lateral sclerosis	Lethargy and coma

Abbreviations: CVT, cortical vein thrombosis; PRES, posterior reversible encephalopathy syndrome; SDH, subdural hematoma; SICH, spontaneous intracranial hypotension.

Table 2 Criteria for diagnosing SICH

1.	Brain and spine imaging signs of CSF leak
2.	CSF pressure less than 6 cm H ₂ O
3.	Characteristic orthostatic headache
4.	Symptoms of headache developing in conjunction with low CSF pressure of CSF leak detection
5.	No causative mechanism that could be explained by another diagnosis
6.	Improvement following epidural blood patch

Abbreviations: CSF, cerebrospinal fluid; SICH, spontaneous intracranial hypotension.

Table 3 Types of spinal CSF leaks (▶Fig. 1)

Type 1	Dural tear: SLEC positive 1A—Ventral tear 1B—Lateral tear
Type 2A	SLEC positive—proximal nerve root sleeve tear/ meningeal diverticular/ dural ectasia
Type 2B	SLEC negative—distal nerve root sleeve tear
Type 3	CSF venous fistula (CVF)

Abbreviations: CSF, cerebrospinal fluid; CVF, CSF venous fistula; SLEC, spinal longitudinal epidural collection.

and migraine.⁵ However, typical history and clinical examination would rule out other causes.

There are few imaging and clinical findings that form the diagnostic criteria for SICH, and these include ▶Table 2.⁶⁻⁹

As SICH is caused by CSF leaks in the spine, three types of leaks are described accordingly depending on the site of leak and whether epidural collection is present or not (▶Table 3).^{8,10,11}

Leaks can be categorized as fast or slow leaks. Fast leaks are seen following ventral dural tear. Slow leaks occur in nerve root sleeve tear and CSF venous fistula (CVF). Tears in the dura can occur ventrally or laterally. An osteophyte or

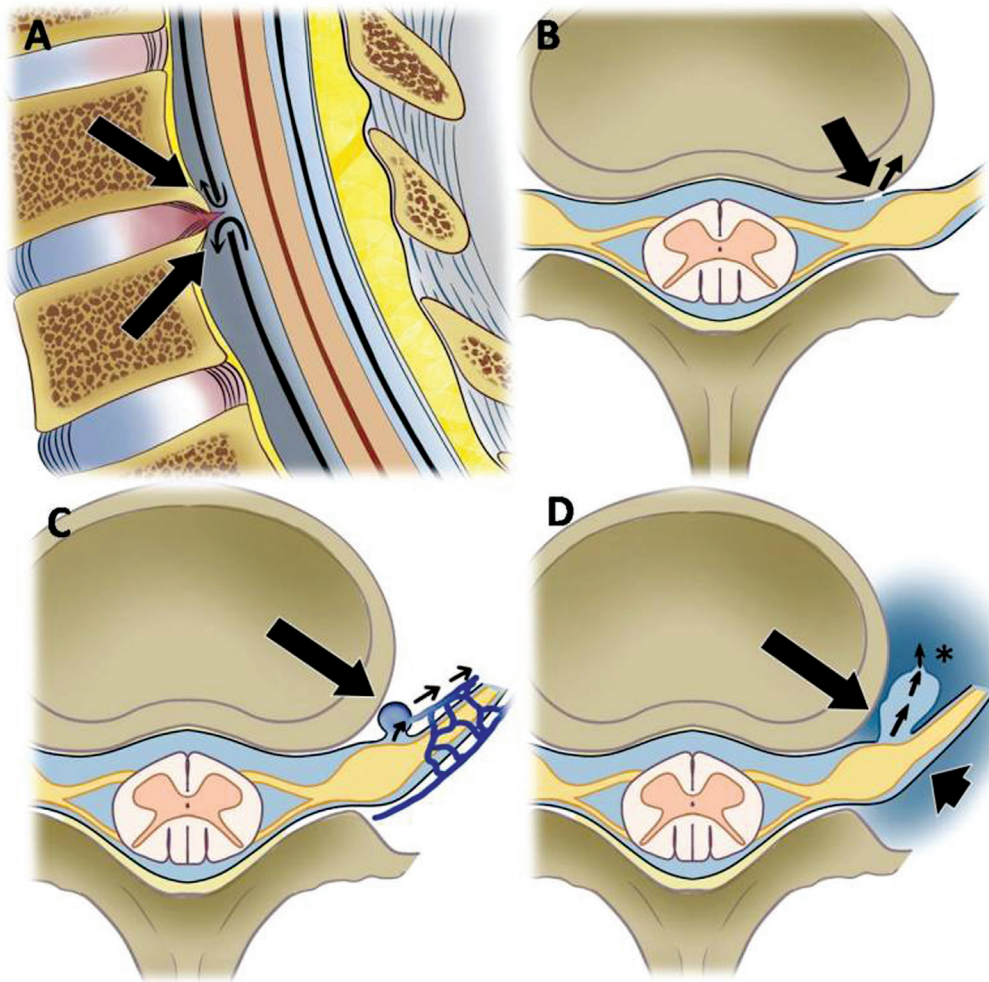


Fig. 1 Types of spontaneous intracranial hypotension (SICH). (A) Type 1 SICH showing spinal ventral dural tear due to calcified discal spur (thick arrows). Curved arrows point to the route of cerebrospinal fluid (CSF) leak into epidural space. (B) Type 2 SICH showing proximal nerve root sleeve tear (thick arrows) points to the meninges, diverticulum with tear. Short thin arrow points to the route of CSF leak. (C) Type 3 SICH showing CSF venous fistula (thick arrow) points to site of CSF venous fistula. (D) Type 2 SICH showing distal nerve root sleeve tear/ meningeal diverticular rupture/ dural ectasia (thick long arrow) leaked CSF (asterisk), and intact posterior dura (thick short arrow).

degenerative disc microspur causes anterior longitudinal tears in the spinal dura and is more commonly seen at upper thoracic levels due to minimal flexibility of the spine along this region, although lower cervical and lumbar dural tears have also been reported. In type 1 spinal leak, there will be epidural collection due to the ventral dural defect that is associated with a discal spur or an osteophyte at the site of leak. It occurs frequently in middle aged females with lesser or normal resting metabolic rate, although the incidence in men is not very uncommon in recent times.^{10,12} Type 2 leaks are classified further as spinal longitudinal epidural collection positive (SLEC-P) which means there is definite spinal longitudinal epidural collection and SLEC negative (SLEC-N) where there is no epidural collection. SLEC-P type 2A leaks occur due to lateral dural tear wherein nerve root sleeve tear occurs proximally with positive SLEC. Other examples are the ruptured meningeal diverticulum and absent nerve root sleeve. These occur between the epidural space and neural foraminal compartment, resulting in definite epidural CSF collection. Meningeal diverticula are frequently seen in

patients with connective tissue disorders like Marfan syndrome, neurofibromatosis, and Ehlers–Danlos syndrome. In type 2B SLEC-N leak, far lateral dural tear or nerve root sleeve tear occurs distally and CSF leaks into the adjacent fascia or connective tissue without any epidural collection. In type 3 leak, the CSF enters directly into the venous channels and hence no epidural collection (SLEC-N).⁹ CVFs are more commonly seen along the lower thoracic spine and in elderly.¹² They may be associated with meningeal diverticulum that is seen as perineural cyst. Such cysts may give rise to CVFs acting as a nidus. CVFs lack dural defect and are seen near nerve root sleeve diverticula close to paraspinal veins.¹³

Other or secondary causes of spinal CSF leaks include injuries to the dura mater following lumbar puncture, spinal anesthesia or surgeries, trauma, and post-ventriculoperitoneal shunt of which post-lumbar puncture intracranial hypotension is more common than other causes.¹⁴ The dural defect is posterior and lumbar in location in these causes as compared to anterior or lateral in SICH, but the pathophysiology is the same regardless of the cause. Post-dural

puncture headache may appear as new daily onset headache after many years of prior dural puncture. There can be an arachnoid bleb formation at the puncture site. Use of atraumatic or pencil point needles over cutting edge needles during lumbar puncture would reduce many complications including post-dural puncture headaches.^{15,16}

Imaging Features

There are few characteristic brain and spine imaging features that suggest intracranial hypotension. Magnetic resonance imaging (MRI) including brain and spine with contrast is the imaging investigation of choice where SICH is suspected clinically due to the clinical symptoms. Computed tomography (CT) brain is not recommended as the temporal resolution is lesser than MRI. However, in few cases, where there is no clinical suspicion of SICH, CT may be the initial imaging modality by the clinicians to look for any cause of headache. CT of the whole spine is important to detect the osteophyte or the discal spur that may be the cause of SLEP-SICH as CT is sensitive for any bone pathology and serves as complementary imaging to MRI study. Brain findings remain the same for all cases of intracranial hypotension regardless the cause.

A description of the MRI protocol is given in ►Table 4, which includes brain and spine sequences.⁹

Below are the distinctive intracranial and spine imaging characteristics.

Intracranial Findings (SEEPS)

- Subdural collection (bilateral)
- Enhancement of the pachymeninges
- Engorgement of dural venous sinuses
- Paucity of bilateral perioptic CSF
- Pituitary hyperemia
- Sagging of the brain
- Superficial siderosis

The initial imaging finding is the venous sinus engorgement or distension that is easily detected in the transverse and straight sinuses. These sinuses have flattened or concave borders normally. The concave border of the transverse sinus

is seen on sagittal images. In patients with intracranial hypotension, the venous sinus borders appear bulged and convex.¹⁷ Pachymeningeal thickening with enhancement is the next imaging feature to appear. The thickening is smooth/non-nodular and requires contrast imaging to be appreciated. This is due to the noninflammatory fibrocollagenous proliferation of the meninges because of persistent vascular engorgement and transudation of intravascular fluid into the adjacent subdural space. On further progression/persistence of the CSF leak, to maintain the volume within the intracranial space, subdural collection occurs secondary to passive transudation from the intravascular space into the subdural space. The collection may be hemorrhagic in nature but always bilateral and more commonly seen along the frontoparietal convexities.¹⁴ Among these signs, venous distension is very specific to diagnose low intracranial pressure. Pituitary engorgement is another important imaging sign where it enlarges up to 8 to 11 mm in height. However, while recovering, it reverses sooner than pachymeningeal enhancement. Another important finding is sagging of the brain secondary to loss of buoyancy of the brain. This is seen on imaging as down sloping of the floor of third ventricle, mamillary body descent, and effaced basal cisternal spaces. Reduced perioptic CSF fluid is another imaging finding that supports the diagnosis of intracranial hypotension. All these findings reverse back to normal following treatment (►Fig. 2A–H). ►Fig. 3 illustrates other miscellaneous imaging features. All the findings are detected on MRI. CT is not very sensitive to detect these findings other than subdural collection. CT may also show pseudosubarachnoid hemorrhage when there is brain sagging and effaced cisternal spaces.

The probability of CSF leak is assessed using Bern scoring system as low, intermediate, and high with scores of 2 or less, 3 to 4, 5 and more, respectively. This system includes only intracranial findings and does not consider SLEP. Bern scoring system is as follows⁹ (►Table 5).

Spine Findings

- Epidural collection (anterior / posterior)
- Dural defect

Table 4 MRI brain and spine sequences recommended in SICH patients

Brain sequences (4–5 mm thickness or 3D sequences if available)	Spine sequences (3–4 mm thickness)
T2W axial 3D—Look for subdural collections/venous sinuses, look for sagging of brainstem& SICH scoring	T2W sagittal (with and without fat suppression)Whole spine screening - to look for epidural collection
Flair axial/3D sagittal—Look for collection/ venous sinuses	T2W axial—in the region of epidural collection
SWI—To look for hemorrhage including CVST and its complications if any	3D heavily T2W (FIESTA/CISS)—To look for meningeal diverticula, dural tear and extent of epidural collection
T1W pre- and post-contrast—To look for pachymeningeal thickening and enhancement	

Abbreviations: 3D, three-dimensional; CISS, constructive interference in steady state; CVST, cerebral venous sinus thrombosis; FIESTA, fast imaging employing steady state acquisition; MRI, magnetic resonance imaging; SICH, spontaneous intracranial hypotension; SWI, sagittal-weighted imaging; T1W, T1-weighted.

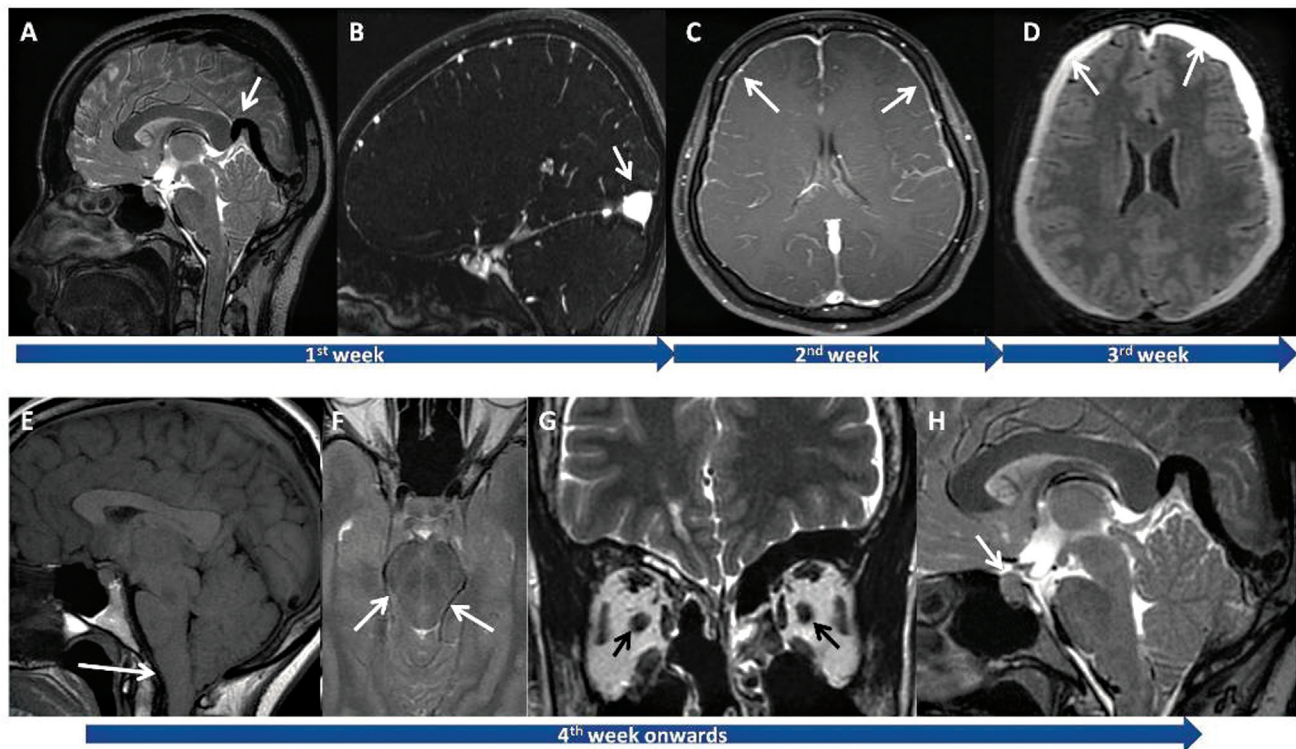


Fig. 2 Sequence of magnetic resonance imaging findings in spontaneous intracranial hypotension. (A) Sagittal T2 image through the brain showing enlarged tortuous straight sinus (white arrow). (B) Sagittal T1 post-contrast image showing rounded venous sinuses with convex inferior borders (white arrow). (C) Axial T1-weighted post-contrast image showing non-nodular, diffuse supratentorial pachymeningeal thickening with enhancement (white arrows). (D) Axial fluid-attenuated inversion recovery image showing bilateral cerebral convexity subdural collections (white arrows). (E) Sagittal T1 image of the brain showing sagging of the parenchyma into the foramen magnum (white arrow). (F) Axial T2 image showing increased anteroposterior diameter of the midbrain with squeezing (white arrow). (G) Coronal T2-weighted image showing reduced cerebrospinal fluid around the bilateral optic nerves (black arrows). (H) Sagittal T2 image showing pituitary engorgement seen as an enlarged dome like pituitary (white arrow).

- Meningeal diverticulum/ Perineural cyst
- Engorged vertebral venous plexus/epidural veins
- Spinal cord signal intensity changes

Out of all the sequences, T2 is the most useful sequence to rule out SLEC and if present is termed as SLEC-P SICH. SICH can also present without SLEC and is termed as SLEC-N SICH. These SLEC-P SICH are fast leakers that are seen in type 1 and type 2 leaks. Type 3 that includes CVF is a slow leaker without SLEC. The resolution of three-dimensional heavily T2-weighted imaging may help in detecting the site of the dural tear in fast leakers. However, MRI spine has limited outcome in patients with CVF as the fistula is not seen in this modality. In a patient with high clinical suspicion of intracranial hypotension without any brain or spine imaging features, CVF needs to be ruled out. Further investigation of choice is digital subtraction myelography (DSM). Plain CT scan of the whole spine is to be very useful in detecting the discal spur or osteophyte responsible for the spinal leak (► Figs. 4 and 5).

Patients with secondary causes of hypotension have similar intracranial imaging findings and spine imaging findings include spinal epidural collection at the site of defect depending on the respective causes.

Chronic SICH

Chronic complications/ findings in untreated patients of SICH include (► Fig. 6):

- Superficial siderosis is commonly seen in posterior fossa involving superior cerebellar foliae and occurs due to bleeding from the friable epidural veins at the site of dural tear.⁹
- Bibrachial amyotrophy. This feature is seen in chronic SLEC-P patients as the collection compresses the anterior horn cells of the spinal cord and stretches the cervical nerve roots resulting in atrophy.^{18,19}
- The longitudinal spinal collection appears more loculated with rounded margins and with or without thin septations like pseudomeningocele.
- Calvarial thickening with/without prominent transosseous venous collaterals. Thickening of the calvarium in SICH is a compensatory mechanism for the depleted CSF volume. Typically, the thickening occurs along the inner table, more so involving the frontal bone, giving rise to an appearance of layer cake skull.²⁰

Uncommon Findings in SICH²¹

- Ischemia is secondary to brain herniation.
- Venous sinus thrombosis due to venous engorgement and stasis.

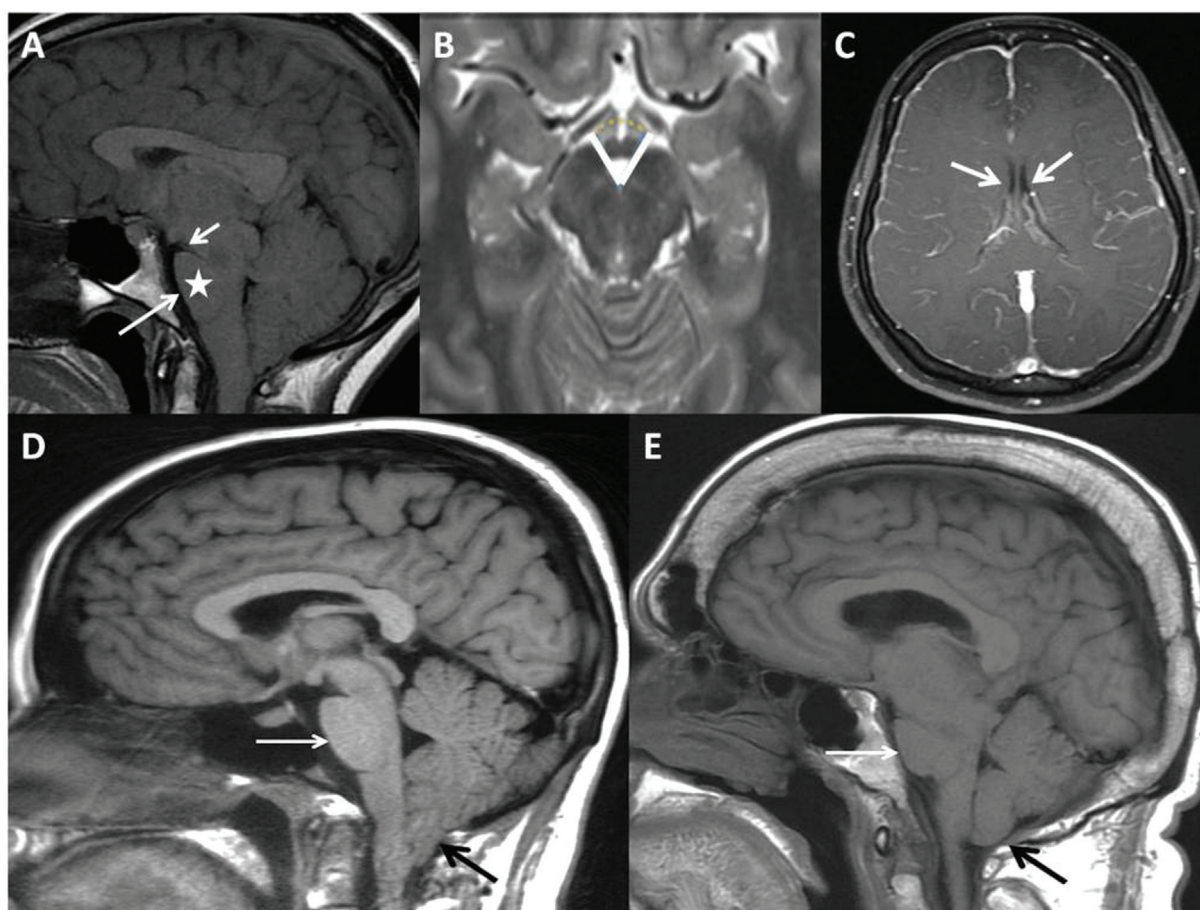


Fig. 3 Miscellaneous imaging findings to diagnose spontaneous intracranial hypotension (SICH). (A) Sagittal noncontrast T1 image through the brain showing flattened ventral pons (asterisk), narrowing of prepontine cistern (long arrow), and reduced mamillopontine distance (short arrow). (B) Axial T2 image showing narrowing of interpeduncular cistern (white lines). (C) Axial contrast T1 image showing slit-like ventricles (white arrows). (D) Patient with Arnold Chiari malformation type 1 reveals sagittal T1 non-contrast image showing normal iter, tonsillar herniation with peg like tonsil (black arrow), and normal pons and prepontine cistern (white arrow). (E) Patient with SICH showing iter below the incisura, tonsillar herniation with normal shaped tonsil (black arrow), flattened pons with reduced prepontine distance (white arrow).

Table 5 Major and minor criteria of Bern/SICH scoring system

Major criteria (2 points each)	Minor criteria (1 point each)
Venous sinus engorgement	Subdural collection
Pachymeningeal enhancement	Prepontine cistern of 5 mm or less
Suprasellar cistern of 4 mm or less	Mamillopontine distance of 6.5 mm or less

Abbreviation: SICH, spontaneous intracranial hypotension.

- Spinal subarachnoid hemorrhage and hemosiderosis.
- Dural calcifications may occur sequelae to chronic blood product deposition secondary to repeated hemorrhage from the epidural venous plexus.

Mimics of SICH²²

- Arnold Chiari 1 malformation
- Other causes of subdural collection
- Other causes of pachymeningeal thickening

- Postural orthostatic tachycardia syndrome (POTS)
- Migraine

Arnold Chiari 1 is a very close mimicker of SICH. Few characteristic imaging findings distinguish between the two and the differences are summarized in **Table 6**.

Other causes of subdural collection may include trauma, bridging vein rupture in elderly following minor trauma. However, these are frequently on one side and rarely bilateral unlike SICH that is always bilateral. Pachymeningeal thickening can also be seen in other conditions like IgG4-related disease, neurosarcoidosis, histiocytosis, other autoimmune conditions, and infections including tuberculosis. In these conditions, other sites, which include brain parenchyma, cranial nerves, and bone may also be affected. In intracranial hypotension, the pachymeningeal thickening is smooth and non-nodular.²⁴

Clinically, few conditions mimic SICH and these are POTS, vestibular migraine, orthostatic hypotension, and cervicogenic headache. POTS patients also develop headache worsening in standing or upright position. Heart rate and blood pressure while sitting and standing would help in diagnosing

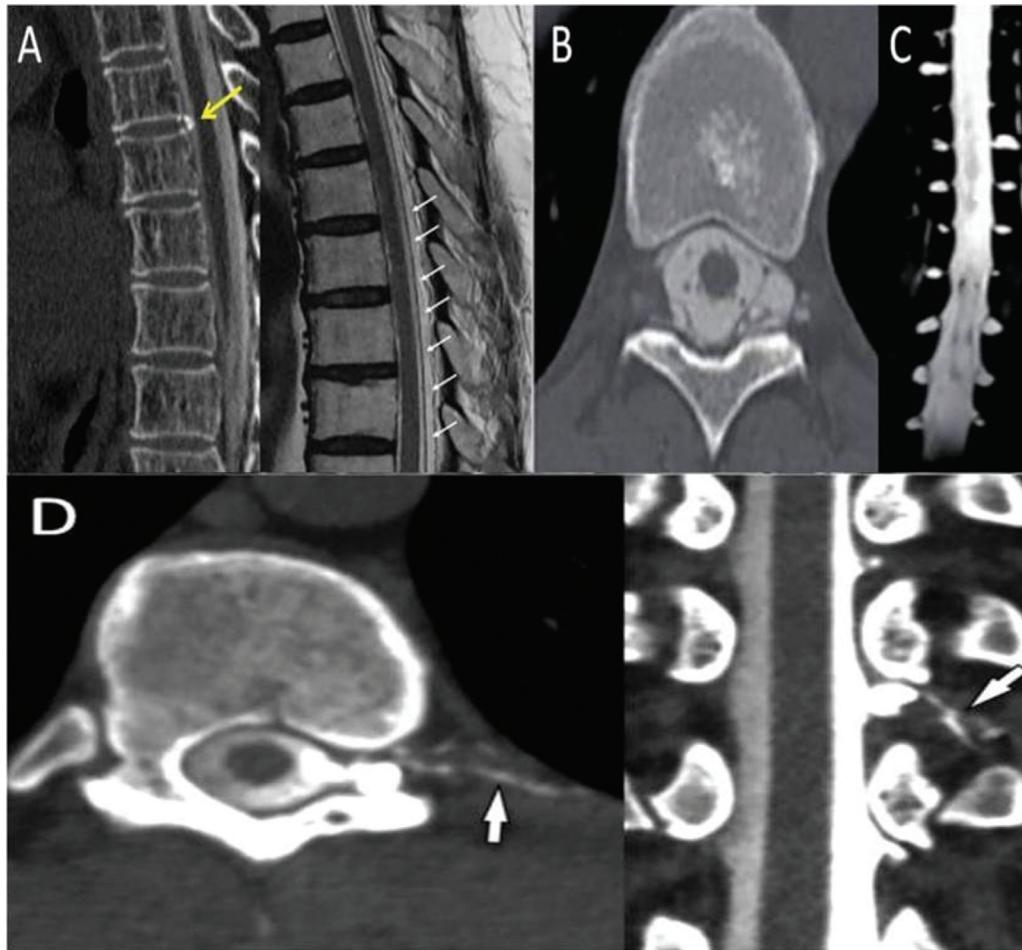


Fig. 4 Types of spontaneous intracranial hypotension (SICH) on cross-sectional imaging. (A) Type 1 SICH with calcified disc spur (arrow). (B and C) Type 2 SICH showing multiple meningeal diverticula on both sides. (D) Type 3 SICH reveals cerebrospinal fluid venous fistula (arrow).

POTS as these patients typically have rapid increase in heart rate on standing with relatively stable blood pressure. Cervicogenic headache is due to osteoarthritic changes of the cervical spine. Vestibular migraine is diagnosed by the presence of associated vertigo.²²

Further Managing SICH

Dynamic CT myelogram (CTM) or DSM performed to confirm spinal leak. Both the techniques involve expertise and radiation exposure. Myelogram confirms the site of dural tear, detection of meningeal diverticulum and CVF. Both the techniques are invasive in nature.

Ultrafast CT Myelography Technique

Patient is positioned in prone Trendelenburg position. By using the lumbar puncture technique, 10 mL of 300mg I/mL iodinated contrast is injected into the thecal sac and real time serial images are taken while injecting the contrast. Back and forth table movement in the CT gantry is accompanied by image acquisition every 10 to 20 seconds. In case of SLEC-N leaks, lateral decubitus position is suitable, although CTM may be negative in CVF and hence DSM is used.²³ In the event of contrast extravasation into

the epidural space, the procedure must be stopped.¹⁴ In fast leakers, the contrast enters the epidural space. If the imaging is quickly performed, the site of leak can be readily detected. In case of CVFs, CTM may show prominent parasagittal vein with adjacent small foraminal radicular veins.

Digital Subtraction Myelogram

It has greater temporal resolution than convention techniques, but limited coverage as DSM makes use of planar images.²⁵ Lateral decubitus position is preferred to demonstrate CVFs. A newer technique has been recently described for better appreciation of CVF. It is a two-day procedure carried out with right lateral decubitus on day 1 and repeating the same in left lateral decubitus on the day 2. To increase the sensitivity of detection of CVF in DSM, few modifications are suggested including respiratory modulation. It is based on the principle of changes in venous pressure secondary to changes in the intrathoracic pressure during different phases of respiration. This technique involves breathing continuously into a 5 mL syringe between the lips and image acquisition during breathing into the syringe and finally during Valsalva maneuver. There is disappearance of the

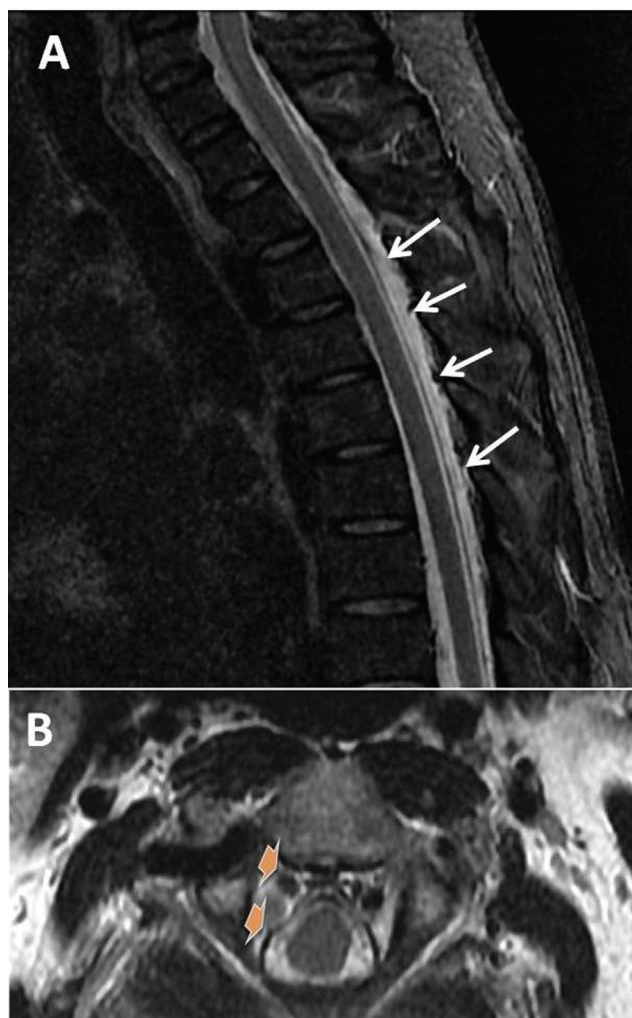


Fig. 5 Spinal imaging findings in spontaneous intracranial hypotension. (A) Sagittal short tau inversion recovery sequence of thoracic spine showing posterior epidural collection (arrows). (B) Axial T2 image of the thoracic spine showing prominent epidural veins (thick arrows).

prominent paraspinous vein and appearance of a prominent external epidural vein during Valsalva.

On CTM, false localizing sign has been described. This sign depicts retrospinal fluid collections at C1-C2 level in patients with intracranial hypotension that occurs as the epidural fluid escapes into the retrospinal soft tissues.^{27,28}

Treatment

Conservative Approach

Hydration and absolute bed rest may be tried but do not respond in many cases. Oral caffeine and abdominal binders may be tried but not at the expense of other definitive management (– Fig. 7).

Epidural blood patch (EBP) can be given empirically or targeted. This is done following no more than 2 weeks of conservative management by performing CTM. Conventional CTM is not useful in fast leakers especially those with type 1 SICH and requires ultrafast dynamic CTM (UFCTM). As part of the radiological intervention for SLEC-P cases, UFCTM is performed to localize the rent followed by 8 to 10 mL of autologous EBP mixed with approximately 1 mL of iodinated contrast into the epidural space. EBP is targeted at either the localized site or empirically if unsuccessful to locate the exact site of leak. However, for all blind patches, lumbar region is used. Image-guided targeted EBP is recommended over blind EBP. Fibrin glue may be used instead of blood, only in failed EBP cases or as the initial treatment if the treating doctor has experience or expertise in using fibrin glue. Post-EBP instructions are to be followed to prevent recurrence. Strict bed rest for at least 8 hours after blood patch. Immediate sitting upright post EBP, long hours of travelling or lifting heavy weights and performing maneuvers which tend to increase the intra-abdominal pressure are to be avoided for few days following the patch as these activities may increase the chance of recurrence. Recommended follow-up after

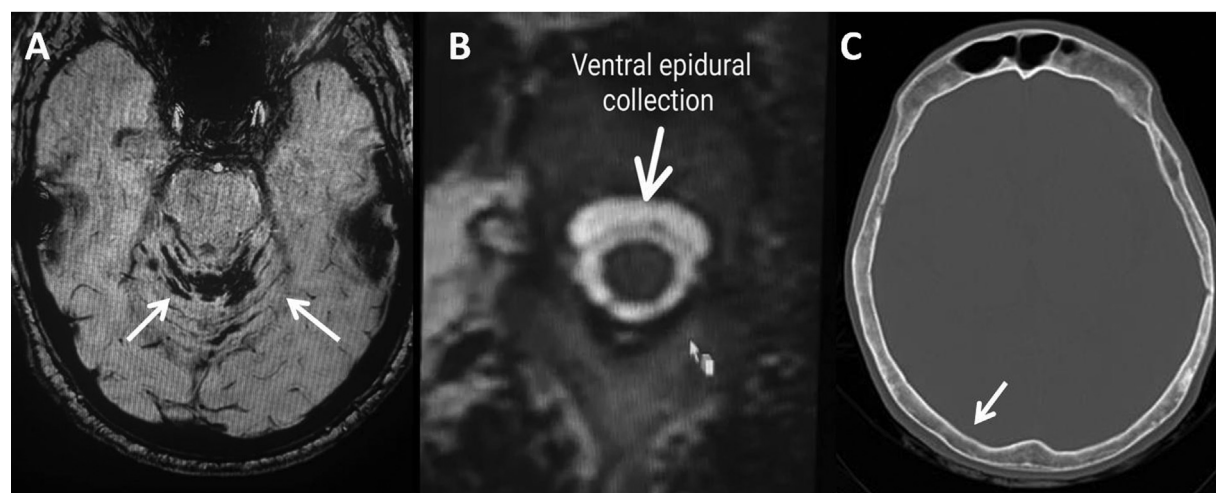


Fig. 6 Imaging findings indicating chronicity of spontaneous intracranial hypotension. (A) Axial susceptibility-weighted image of the brain showing blooming in the region of bilateral cerebellar folia, suggesting superficial siderosis (arrows). (B) Axial T2 weighted image of thoracic spine showing rounded spinal epidural collection in the anterior aspect. (C) Axial plain computed tomography bone window of the head showing thickened cortex (arrow).

Table 6 Differences between Arnold Chiari 1 and SICH

	Arnold Chiari malformation 1	SICH
Tonsils	Peg like	Normal shape
Tonsillar ectopia	More	Less
Midbrain	No descent	Descent
Ister	Normal	Below incisura
Pons	Normal	Effaced prepontine effacement and flattened ventral pons
Corpus callosum	Normal	Drooping of splenium
Contrast	Nil	Pachymeningeal enhancement
SDH	Nil	Common
Syrinx and/or hydrocephalus	Associated	Rare
Third ventricular floor descent	> 15 degrees	< 15 degrees
Pontomesenchymal angle	> 45 degrees	< 45 degrees

Abbreviations: SDH, subdural hematoma; SICH, spontaneous intracranial hypotension.

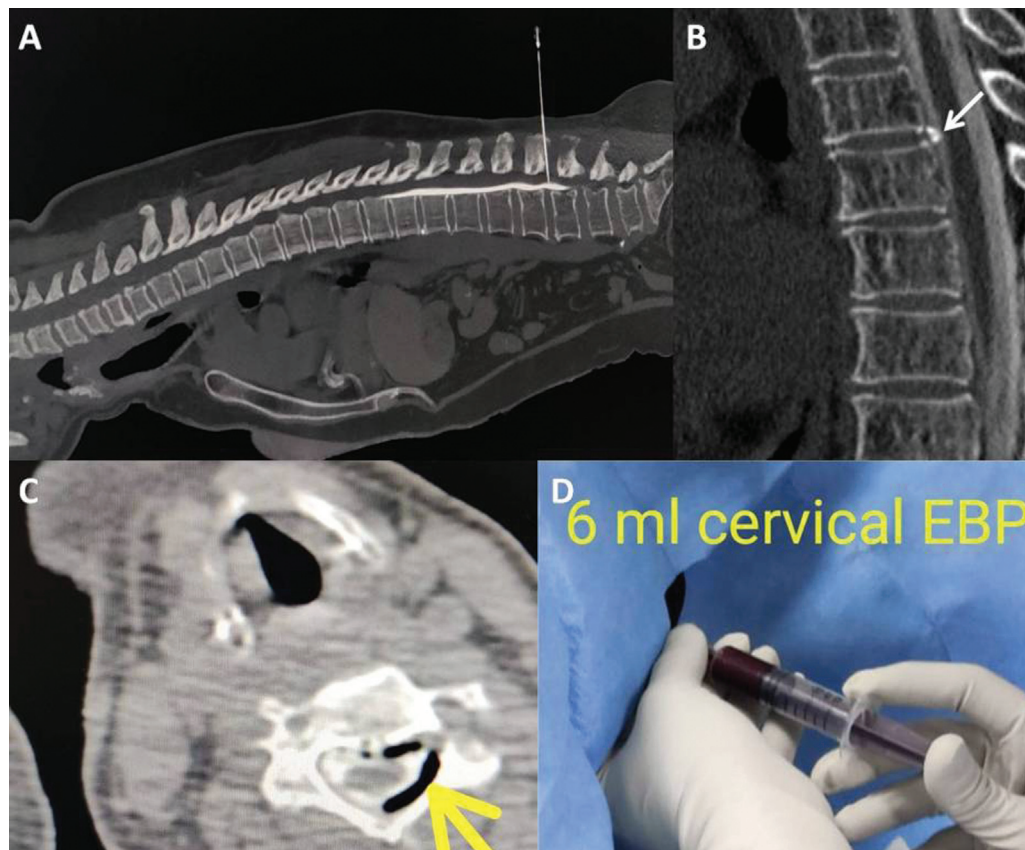


Fig. 7 Computed tomography (CT) myelogram procedure. (A) Injecting CT contrast through lumbar puncture technique in Trendelenburg position. (B) Sagittal CT myelogram showing bony spicule along the posterior aspect of the disc (arrow). (C) Epidural air confirms the needle is in epidural space before injecting autologous blood (yellow arrow). (D) Localized epidural blood patch (EBP) in prone position.

blood patch is between 10 and 14 days and late follow-up is between 3 and 6 months. In case of recurrence in clinical symptoms, multidisciplinary meet is performed, and repeat imaging or intervention may be considered.³

- Repeat EBPs recommended for recurrent cases. Image-guided targeted EBP is recommended over empirical

patches. CT-guided epidural fibrin glue patches may also be tried for failed EBP cases.

- CT-guided fibrin glue injection to meningeal diverticula and CVF.
- CT-guided epidural fibrin glue injection or transvenous endovascular embolization of foraminal veins using liquid embolic agents for CVF.

- Neurosurgical role
 - Subdural collections evacuation for causing significant mass effect or midline shift.
 - Dural repair and excision of the bony spurs or microspecies.
 - Microsurgical repair of the dura and ligation of foraminal vein in cases of CVF and ligation repair of meningeal diverticulum.

Intrathecal Gadolinium MR Myelogram Remains Obsolete
Nuclear medicine cisternography used in patients with image-negative SICH. Lumbar puncture radionuclide tracer is injected. After 24 hours, complete coverage of cerebral convexity is obtained. In case of incomplete coverage after 24 hours, CSF flow dynamics are affected, and these are considered as SICH-positive cases and are subjected to DSM/CTM to assess possible SLEC-N leak.²⁹

Anticoagulation in Case of Cerebral Venous Thrombosis
 At places where there is no expertise to perform the UFCTM or DSM, two level nontargeted autologous blood patch is preferred, one at the cervicothoracic junction and the other at dorsolumbar junction, respectively.

Rebound Intracranial Hypertension
 SICH may result in rebound intracranial hypertension following EBP or surgical repair, where the patient presents with headache worsening on lying down and improves in upright position. This is more commonly seen in patients with chronic SICH and is believed to be caused by increased CSF production and disrupted CSF reabsorption during the

CSF leak period. It is treated by head end elevation and analgesics in mild cases, acetazolamide orally or intravenously in moderate cases, and lumbar puncture to remove the fluid in severely affected cases.³⁰

Post-Treatment Imaging

Imaging findings are temporary and revert to normal. Intracranial features of venous distension, pituitary engorgement, herniation, and pachymeningeal enhancement disappear within a month following treatment. However, bilateral subdural collection may take a few more weeks to resolve.

Conclusion

SICH is an emerging elusive neurological condition, with an increase in incidence in recent years due to improved detection techniques and increasing awareness. The flow-chart (→Fig. 8) summarizes the steps need to be undertaken from the time of suspicion of diagnosis followed by the management. The knowledge of imaging findings of SICH is to be made aware among all the general radiologists that can further improve in detecting and treating this condition. MRI brain with contrast and screening the whole spine would give the radiologist a clue regarding spinal CSF leak. Utilizing appropriate diagnostic techniques is the key to confirming the diagnosis. Targeted EBP is the preferred first line of the treatment at present and use of CT guidance for injecting blood and fibrin glue is on the rise. In case of failure, repeat patches can be given and may consider alternative treatment strategies especially surgical repairs. However, the key success in SICH treatment is targeted

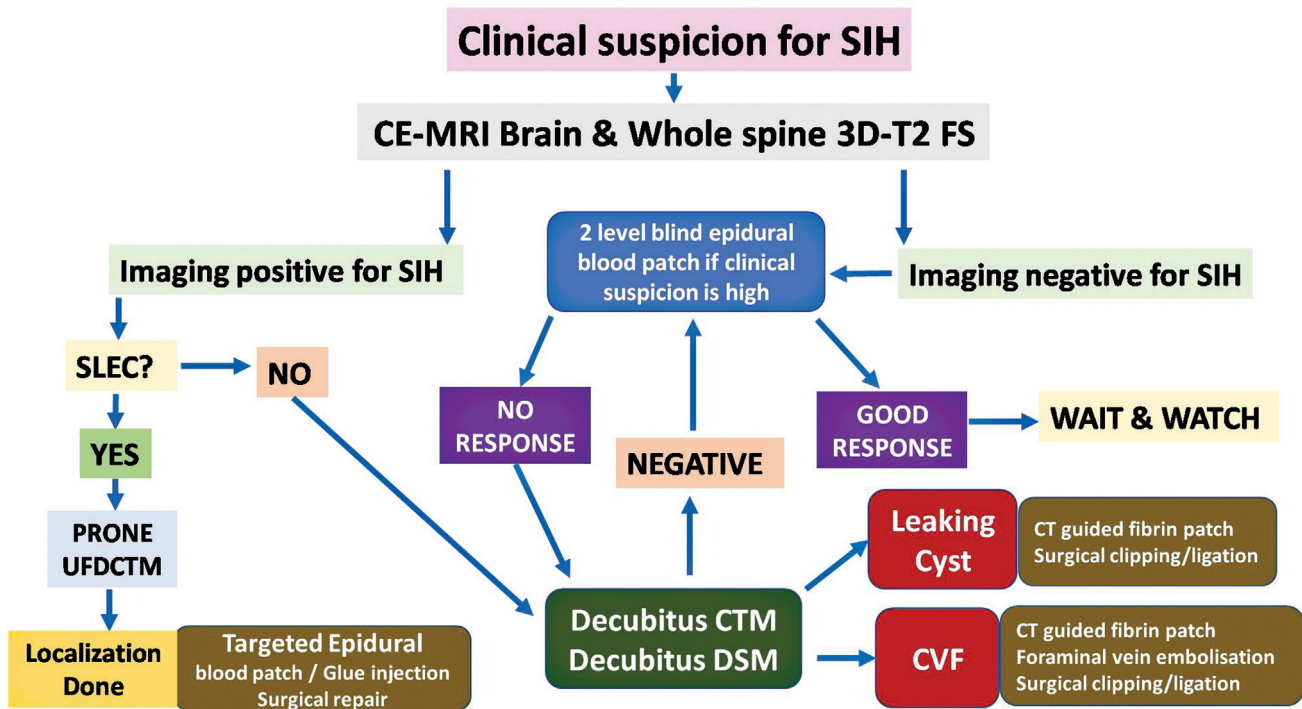


Fig. 8 Flowchart illustrating the steps involved in the management of spontaneous intracranial hypotension. CTM, CT myelogram; CVF, CSF venous fistula; DSM, digital subtraction myelogram; SICH, spontaneous intracranial hypotension; SLEC, sagittal longitudinal epidural collection; UFDCTM, ultrafast dynamic CT myelogram.

image-guided blood patch and following strict post-procedure instructions.

Learning Points

1. Headache with postural nature is an important symptom.
2. Imaging in SICHD should include both brain and spine.
4. Dynamic myelogram is preferred over conventional myelogram.
5. Multimodality treatment is available at present.

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Conflict of Interest

None declared.

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References

- 1 Mokri B. The Monro-Kellie hypothesis: applications in CSF volume depletion. *Neurology* 2001;56(12):1746–1748
- 2 Robblee J, Secora KA, Alhilali LM, Knievel KL. Spontaneous Intracranial Hypotension Requires a High Index of Suspicion Because of Potentially Normal Diagnostic Test Results and Refractory Symptoms. *Practical Neurology* 2020
- 3 Sun-Edelstein C, Lay CL. Spontaneous intracranial hypotension: pathophysiology, clinical features, and diagnosis. *UpToDate* 2022
- 4 Headache classification committee of the international headache society (IHS) the international classification of headache disorders, 3rd edition. *Cephalalgia* 2018;38(01):1–211
- 5 Cheema S, Anderson J, Angus-Leppan H, et al. Multidisciplinary consensus guideline for the diagnosis and management of spontaneous intracranial hypotension. *J Neurol Neurosurg Psychiatry* 2023
- 6 Urbach H, Fung C, Dovi-Akue P, Lützen N, Beck J. Spontaneous intracranial hypotension. *Dtsch Arztebl Int* 2020;117(27-28):480–487
- 7 Kim D, Small JE. Intracranial Hypotension. In Small JE, Noujaim DL, Ginat DT, Kelly HR, Schaefer PW (Ed), *Neuroradiology*. Elsevier 2019:158–162. Doi: <https://doi.org/10.1016/B978-0-323-44549-8.00017-1>
- 8 Kranz PG, Gray L, Malinzak MD, Amrhein TJ. Spontaneous intracranial hypotension: pathogenesis, diagnosis, and treatment. *Neuroimaging Clin N Am* 2019;29(04):581–594
- 9 Carlton Jones L, Butteriss D, Scoffings D. Spontaneous intracranial hypotension: the role of radiology in diagnosis and management. *Clin Radiol* 2022;77(03):e181–e194
- 10 Farb RI, Nicholson PJ, Peng PW, et al. Spontaneous intracranial hypotension: a systematic imaging approach for CSF leak localization and management based on MRI and digital subtraction myelography. *Am J Neuroradiol* 2019;40(04):745–753
- 11 Luetzen N, Dovi-Akue P, Fung C, Beck J, Urbach H. Spontaneous intracranial hypotension: diagnostic and therapeutic workup. *Neuroradiology* 2021;63(11):1765–1772
- 12 Mamlouk MD, Shen PY, Jun P, Sedrak MF. Spontaneous spinal CSF leaks stratified by age, body mass index, and spinal level. *Am J Neuroradiol* 2022;43(07):1068–1072
- 13 Callen AL, Timpone VM, Schwertner A, et al. Algorithmic multimodality approach to diagnosis and treatment of spinal CSF leak and venous fistula in patients with spontaneous intracranial hypotension. *Am J Roentgenol* 2022;219(02):292–301
- 14 Michali-Stolarska M, Bladowska J, Stolarski M, Szaśiadek MJ. Diagnostic imaging and clinical features of intracranial hypotension – review of literature. *Pol J Radiol* 2017;82:842–849
- 15 Callen AL, Lennarson P, Carroll IR. A causative role for remote dural puncture and resultant arachnoid bleb in new daily persistent headache: a case report. *Headache* 2023;63(07):981–983
- 16 Nath S, Koziarz A, Badhiwala JH, et al. Atraumatic versus conventional lumbar puncture needles: a systematic review and meta-analysis. *Lancet* 2018;391(10126):1197–1204
- 17 Kim SC, Ryoo I, Sun HY, Park SW. MRI findings of spontaneous intracranial hypotension: usefulness of straight sinus distention. *Am J Roentgenol* 2019;212(05):1129–1135
- 18 Lützen N, Zeitlberger A, Beck J, Urbach H. Teaching neuroimages: dynamic digital subtraction myelography discloses a ventral CSF leak in a patient with upper limb amyotrophy. *Clin Neuroradiol* 2023;33(01):245–246
- 19 Girão MMV, Sousa RMP, Ribeiro MC, Cardoso TAMO, França Júnior MC, Reis F. Spontaneous intracranial hypotension and its complications. *Arq Neuropsiquiatr* 2018;76(08):507–511
- 20 Benson JC, Madhavan AA, Cutsforth-Gregory JK, Johnson DR, Carr CM. The Monro-Kellie doctrine: a review and call for revision. *Am J Neuroradiol* 2023;44(01):2–6
- 21 Callen AL, Dillon WP, Shah VN. Correction to: Unusual neuroimaging findings in spontaneous intracranial hypotension. *Neuroradiology* 2023;65(05):883
- 22 Bond KM, Benson JC, Cutsforth-Gregory JK, Kim DK, Diehn FE, Carr CM. Spontaneous intracranial hypotension: atypical radiologic appearances, imaging mimickers, and clinical look-alikes. *Am J Neuroradiol* 2020;41(08):1339–1347
- 23 Houk JL, Amrhein TJ, Gray L, Malinzak MD, Kranz PG. Differentiation of Chiari malformation type 1 and spontaneous intracranial hypotension using objective measurements of midbrain sagging. *J Neurosurg* 2021;136(06):1796–1803
- 24 Dobrocky T, Mosimann PJ, Zibold F, et al. Cryptogenic cerebrospinal fluid leaks in spontaneous intracranial hypotension: Role of dynamic CT myelography. *Radiology* 2018;289(03):766–772
- 25 Kranz PG, Luetmer PH, Diehn FE, Amrhein TJ, Tanpitukpongse TP, Gray L. Myelographic techniques for the detection of spinal CSF leaks in spontaneous intracranial hypotension. *Am J Roentgenol* 2016;206(01):8–19
- 26 Kranz PG, Malinzak MD, Gray L, Willhite J, Amrhein TJ. Resisted inspiration improves visualization of CSF-venous fistulas in spontaneous intracranial hypotension. *Am J Neuroradiol* 2023;44(08):994–998
- 27 Schievink WI, Maya MM, Chu RM, Moser FG. False localizing sign of cervico-thoracic CSF leak in spontaneous intracranial hypotension. *Neurology* 2015;84(24):2445–2448
- 28 Medina JH, Abrams K, Falcone S, Bhatia RG. Spinal imaging findings in spontaneous intracranial hypotension. *Am J Roentgenol* 2010;195(02):459–464
- 29 Farnsworth PJ, Madhavan AA, Verdoorn JT, et al. Spontaneous intracranial hypotension: updates from diagnosis to treatment. *Neuroradiology* 2023;65(02):233–243
- 30 Schievink WI, Maya MM, Jean-Pierre S, Moser FG, Nuño M, Pressman BD. Rebound high-pressure headache after treatment of spontaneous intracranial hypotension: MRV study. *Neurol Clin Pract* 2019;9(02):93–100