Spontaneous intracranial hypotension – a spinal disease

Spontane intrakranielle Hypotension – eine spinale Erkrankung

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ABSTRACT

Background Spontaneous intracranial hypotension (SIH) remains an underdiagnosed condition despite increasing awareness due to recent scientific advances. Diagnosis can be delayed by the broad clinical presentation and imaging pitfalls. This results in a high degree of physical impairment for patients, including social and psychological sequelae as well as long-term damage in the case of delayed diagnosis and treatment.

Method The study is based on a selective literature search on PubMed including articles from 1990 to 2023 and the authors' clinical experience from working in a CSF center.

Results and Conclusion SIH mostly affects middle-aged women, with the primary symptom being position-dependent orthostatic headache. In addition, there is a broad spectrum of possible symptoms that can overlap with other clinical conditions and therefore complicate the diagnosis. The causative spinal CSF loss can be divided into three main types: ventral (type 1) or lateral (type 2) dural leak and CSF-venous fistula (type 3). The diagnosis can be made using a two-stage workup. As a first step, noninvasive MRI of the head and spine provides indicators of the presence of SIH. The second step using focused myelography can identify the exact location of the cerebrospinal fluid leak and enable targeted therapy (surgical or interventional). Intrathecal pressure measurement or intrathecal injection of gadolinium is no longer necessary for primary diagnosis. Serious complications in the course of the disease can include space-occupying subdural hematomas, superficial siderosis, and symptoms of brain sagging, which can lead to misinterpretations. Treatment consists of closing the dural leak or the cerebrospinal fluid fistula. Despite successful treatment, a relapse can occur, which highlights the importance of follow-up MRI examinations and emphasizes the chronic nature of the disease. This paper provides an overview of the diagnostic workup of patients with suspected SIH and new developments in imaging and therapy.

Key Points

- SIH is an underdiagnosed condition with a wide range of possible symptoms.
- The first diagnostic step using MRI provides indications of the presence of SIH.
- The second diagnostic step using (dynamic) myelography can identify the CSF leak.
- Collaboration with a CSF center is advisable for further diagnosis and treatment.
- Prompt detection and treatment of SIH improves the outcome.

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ZUSAMMENFASSUNG

Hintergrund Die spontane intrakranielle Hypotension (SIH) ist ein unterdiagnostiziertes Krankheitsbild, das jedoch aufgrund neuer wissenschaftlicher Erkenntnisse zunehmend Aufmerksamkeit erfährt. Die Diagnosestellung kann durch ein breites klinisches Erscheinungsbild und Fallstricke in der Bildgebung verzögert werden. Daraus resultiert eine hohe körperliche Beeinträchtigung für die Patienten, einschließlich sozialer und psychischer Folgen sowie Langzeitschäden bei prolongierter Diagnostik und Therapie.

Methode Basierend auf einer selektiven Literaturrecherche auf PubMed mit Einschluss aller Arbeiten zwischen 1990 und 2023 sowie der klinischen Erfahrung aus der Arbeit der Autoren in einem CSF-Zentrum.

Ergebnisse und Schlussfolgerungen SIH betrifft meist Frauen im mittleren Alter, Leitsymptom ist dabei der lageabhängige orthostatische Kopfschmerz. Daneben gibt es ein breites Spektrum weiterer möglicher Symptome, die sich mit anderen Krankheitsbildern überlappen können und dementsprechend die Diagnosestellung erschweren. Der ursächliche spinale Liquorverlust lässt sich in drei Haupttypen unterteilen: das ventrale (Typ 1) und laterale (Typ 2) Duraleck sowie die Liquor-Vene-Fistel (Typ 3). Die Diagnostik kann über eine zweistufige Aufarbeitung erfolgen. Im ersten Schritt liefert die nicht invasive MRT von Kopf und Wirbelsäule Hinweise auf das Vorliegen einer SIH. Der zweite Schritt mittels gezielter Myelografie kann den genauen Ort des Liquoraustritts identifizieren und eine gezielte Behandlung ermöglichen (operativ oder interventionell). Eine intrathekale Druckmessung oder die intrathekale Gabe von Gadolinium ist dabei zur Primärdiagnostik nicht mehr notwendig. Ernstzunehmende Komplikationen im Verlauf der Krankheit bestehen z. B. in raumfordernden Subduralhämatomen, der superfiziellen Siderose und Symptomen im Rahmen eines „brain sagging“, was zu Fehlinterpretationen führen kann. Die Therapie besteht im Verschluss des Duralecks bzw. der Liquorfistel. Auch nach erfolgreicher Therapie können Rezidive auftreten, was die Wichtigkeit einer klinischen Nachsorgeuntersuchung einschließlich Follow-Up-MRT betont und den chronischen Charakter der Krankheit unterstreicht. Diese Arbeit liefert eine Übersicht über den diagnostischen Work-Up von Patienten mit Verdacht auf SIH und neue Entwicklungen in Bildgebung und Therapie.

Kernaussagen

- Die SIH ist ein unterdiagnostiziertes Krankheitsbild mit einer großen Variationsbreite möglicher Symptome.
- Der erste Diagnostikschritt mittels MRT liefert Hinweise auf das Vorliegen einer SIH.
- Der zweite Diagnostikschritt mittels (dynamischer) Myelographie kann das Liquorleck identifizieren.
- Eine Zusammenarbeit mit einem CSF-Zentrum ist bei weiterführender Diagnostik und Therapie sinnvoll.
- Eine zeitnahe Erkennung und Behandlung der SIH verbessert das Outcome.

Abbreviations

CISS	constructive interference in steady state
cMRI	cranial magnetic resonance imaging
CSF	cerebrospinal fluid
CT	computed tomography
CTM	CT myelography
DSM	digital subtraction myelography
EBP	epidural blood patch
FLAIR	fluid attenuated inversion recovery
fs	fat saturation
HASTE	half acquisition single-shot turbo spin echo
MPRage	magnetization prepared rapid gradient echo
MRI	magnetic resonance imaging
POTS	postural orthostatic tachycardia syndrome
SIH	spontaneous intracranial hypotension
SLEC	spinal longitudinal extradural CSF collection
SPACE	sampling perfection with application optimized contrast using different flip angle evolution
SWI	susceptibility weighted imaging
T1w	T1-weighted
T2w	T2-weighted
TSE	turbo spin echo
2 D	two-dimensional
3 D	three-dimensional

Introduction

The diagnosis and treatment of spontaneous intracranial hypotension (SIH), which was first described in 1938 by a German neurologist, have undergone rapid development in recent decades [1]. However, in spite of increasing awareness of the disease, it is often initially misdiagnosed [2, 3]. Studies show that SIH is more common than initially assumed but there is minimal data regarding the incidence [4]. Rates of 5/100,000 patients per year have been described, which almost corresponds to the global incidence of aneurysmal subarachnoid hemorrhage [5, 6, 7].

The disease is caused by a spinal CSF leak resulting from a dural tear or a direct connection of the CSF space into a paravertebral vein (CSF-venous fistula) [8]. In contrast, a CSF leak at the base of the skull with CSF rhinorrhea or otorrhea usually does not cause any SIH symptoms [9]. Predominantly affected are women (f:m approx. 3:2) in middle age [6]. The resulting symptoms vary greatly and, in addition to orthostatic headache as the main symptom, can cause a broad spectrum of symptoms, with a comatose state being the most severe presentation [10]. Studies also show that the symptoms of low CSF pressure can have a significant effect on quality of life. Depression, anxiety, and suicidal thoughts are common in this patient population [11, 12]. An incorrect diagnosis can result in an incorrect therapeutic workup and even unnecessary surgical interventions [3].

In recent years, the diagnostic and therapeutic options have developed rapidly: even micro-pathologies can be visualized with high-resolution imaging and can be treated with various interventional therapies. As a result of the further development of the

▶ **Table 1** Modified diagnostic criteria of spontaneous intracranial hypotension (SIH) based on the International Classification of Headache Disorders 3rd edition [10, 13]. These criteria cannot be applied for patients in whom a lumbar puncture or previous trauma has led to the development of CSF loss.

Modified diagnostic criteria of spontaneous intracranial hypotension	
A.	Any headache fulfilling criterion C
B	CSF opening pressure < 6 cm H_2O and/or evidence of CSF leakage on imaging
C	Headache that has developed in temporal relation to low CSF pressure or CSF leakage or has led to its discovery
D	Not better accounted for by any other ICHD-3 diagnosis

diagnostic workup and treatment, the disease can be detected and properly treated increasingly earlier, thus avoiding serious complications. As a result of the growing awareness, increased detection of the disease and thus a greater incidence can be expected. This review provides guidelines for the care of affected patients.

Definition and pathophysiology

In the current third edition of The International Classification of Headache Disorders, the headache that occurs in association with SIH is described as an orthostatic headache usually accompanied by neck stiffness and hearing disturbances [13]. The diagnostic criteria for low-pressure headaches are summarized in ▶ **Table 1**. Due to the different origins and prognoses, it is necessary to differentiate between spontaneous hypotension and other types of low-pressure headache. For example, there are postdural puncture headaches (PDPH) as well as headaches caused by a CSF fistula following surgery or trauma [13].

Even if SIH was defined as a reduced cerebrospinal fluid opening pressure (< 6 cm H_2O) intially, recent studies have shown that the majority of patients have normal intrathecal pressure [14]. The term "spontaneous intracranial hypotension" is actually misleading since it is a condition of hypovolemia rather than true hypotension [15, 16]. Due to this new information and the high sensitivity of MRI in the primary diagnostic workup of SIH (see below), intrathecal pressure measurement is no longer necessary as a diagnostic intervention [13].

According to the Monro-Kellie doctrine, a loss of volume in a compartment of a closed system results in an increase in volume in another compartment. In the case of CSF loss in SIH, the venous sinuses become dilated with characteristic morphological changes on imaging [10, 17]. It is assumed that brain sagging and the resulting traction on the meninges cause the usually occipital headache [3].

Modified according to Schievink et al. and Farb et al., the following three main types of CSF leak have become established in clinical practice [8, 18]:

▶ **Table 2** Frequency and variance of associated symptoms of SIH. Table adapted to a meta-analysis by D'Antona et al. [6].

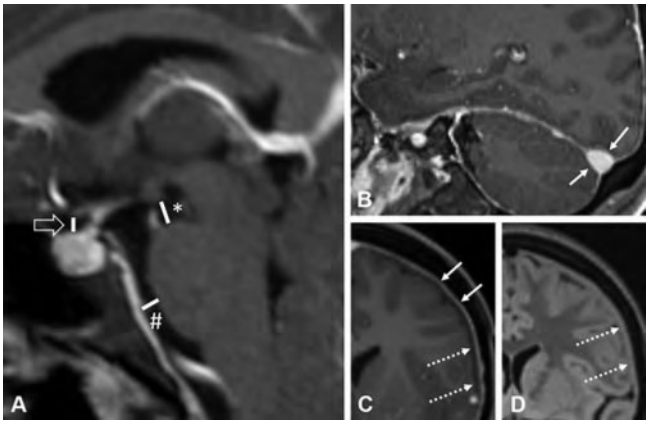
Symptoms of SIH	**Frequency**
Headache ▪ orthostatic headache ▪ nonorthostatic headache	94–99 % [6] 87–96 % [6] 4–13 % [6]
Nausea, vomiting	46–62 % [6]
Neck stiffness	32–53 % [6]
Dizziness	13–42 % [6]
Hearing disturbances	18–38 % [6]
Tinnitus	14–26 % [6]
Photophobia	5–16 % [6]
Diplopia	3–10 % [6]
Other visual symptoms	7–21 % [6]
Changes in consciousness	8–22 % [6]
Cognitive impairments	2–11 % [6]

- Type 1 (approx. 50 % of cases) [18]: ventral dural tear usually caused by a bone spur or rarely by a soft disc herniation [19]. ⅔ of type 1 leaks are located in the upper thoracic region [20].
- Type 2 (approx. 20 % of cases) [18]: lateral dural tear in the region of the nerve root sheath, with the exact cause of the dural tear being unclear to date. Dural weakness and micro-traumas are being discussed [21]. Type 2 leaks are usually located between the mid-thoracic spine and upper lumbar spine.
- Type 3 (approx. 25 % of cases) [18]: CSF-venous fistulas, i. e., a direct connection from the CSF space to the internal or external spinal venous plexus. Tears of spinal arachnoid granulations may be involved in the pathogenesis [22]. Type 3 leaks have a right-sided predilection and are primarily located in the mid and lower thoracic spine [23].

In addition to these three types, there are indications of a fourth group in the sacral region (approx. 6 % of cases), with this being almost exclusively seen in women [24]. Further information about sacral leaks and etiology is not yet available.

Clinical manifestation

The main symptom of SIH is position-dependent, orthostatic headache with improvement of symptoms in a lying position [3]. Accordingly, patients often report an increase in headache primarily in the second half of the day (so-called "second half of the day headache" [25]), while the symptoms are only mild or absent upon waking up in the morning [25, 26, 27]. It is not uncommon for patients to be able to name the exact onset of the symptoms [3]. Typical symptoms are mental impairment ("brain fog") as well as a feeling of water in the ears (aural fullness) [10, 26, 27, 28]. Moreover, there is significant variability in potential associated symptoms (▶ **Table 2**).



▶ **Fig. 1** SIH score assessing the probability of the presence of SIH. Sagittal, fat-saturated T1w sequence after contrast shows the measurement of the suprasellar distance (open white arrow in **A**; ≤ 4 mm gives 2 points), mamillopontine distance (asterisk in **A**; ≤ 6.5 mm gives 1 point), and prepontine distance (diamond in **A**; ≤ 5 mm gives 1 point). Same sequence in **B** shows a dilated, convexly configured transverse sinus (solid white arrows in **B**; yields 2 points); the normal, non-dilated sagittal sinus is triangularly concave in configuration. The coronal fat-saturated T1w sequence after contrast shows pachymeningeal enhancement (solid white arrows in **C**; yields 2 points). Below this, a hypointense rim in the sense of a hygroma (dashed white arrows in **C**; results in 1 point) can be seen, which appears as a narrow, hyperintense band in the coronal FLAIR sequence (white dashed arrows in **D**). 0–2 indicates low, 3–4 intermediate, and ≥ 5 high probability for the presence of SIH.

Important differential diagnoses of SIH include postural orthostatic tachycardia syndrome (POTS), migraine, and cervicogenic headache [29, 30]. However, differentiation from other diseases like Chiari malformation or frontotemporal dementia can also be difficult (see below).

As a rule, the symptoms of SIH can change over the course of the disease, with the headache or the postural dependence of the headache becoming less prominent and other symptoms becoming more significant [3, 31].

Step-by-step diagnosis

Indications of SIH (MRI and SIH score)

The first step in the diagnostic workup is to evaluate the probability of a loss of CSF. Noninvasive MRI examination with suitable sequences is sufficient for this purpose [32].

MRI of the head

A standard protocol should include at least one sagittal T1w sequence after contrast administration, ideally a 3 D sequence (e. g. MPRage, "magnetization prepared rapid acquisition with gradient echoes"), and a non-contrast FLAIR sequence ("fluid attenuated inversion recovery") of the head [30].

The "Bern SIH score", which has become established in the literature for evaluating the presence of a CSF leak (▶ **Fig. 1**), is calculated based on these sequences [17, 33]. Different SIH signs are weighted differently in this score. Thus, pachymeningeal enhancement, dilation of the venous sinus, and a reduced suprasellar distance are very sensitive signs of SIH and are therefore each given 2 points [17]. Knowledge of the ubiquitous pachymeningeal contrast enhancement in SIH is decisive for the differentiation from inflammatory or neoplastic conditions (usually leptomeningeal enhancement) [29]. Minor criteria include a reduced prepontine and mamillopontine distance and the presence of hygromas or subdural hematomas (each given 1 point) [17]. Adding the points results in a point scale between 0 and 9. A score of ≤ 2 has a low probability of a CSF leak, a score of 3–4 a moderate probability, and a score of ≥ 5 a high probability. The scores help with decisions about the further approach [17].

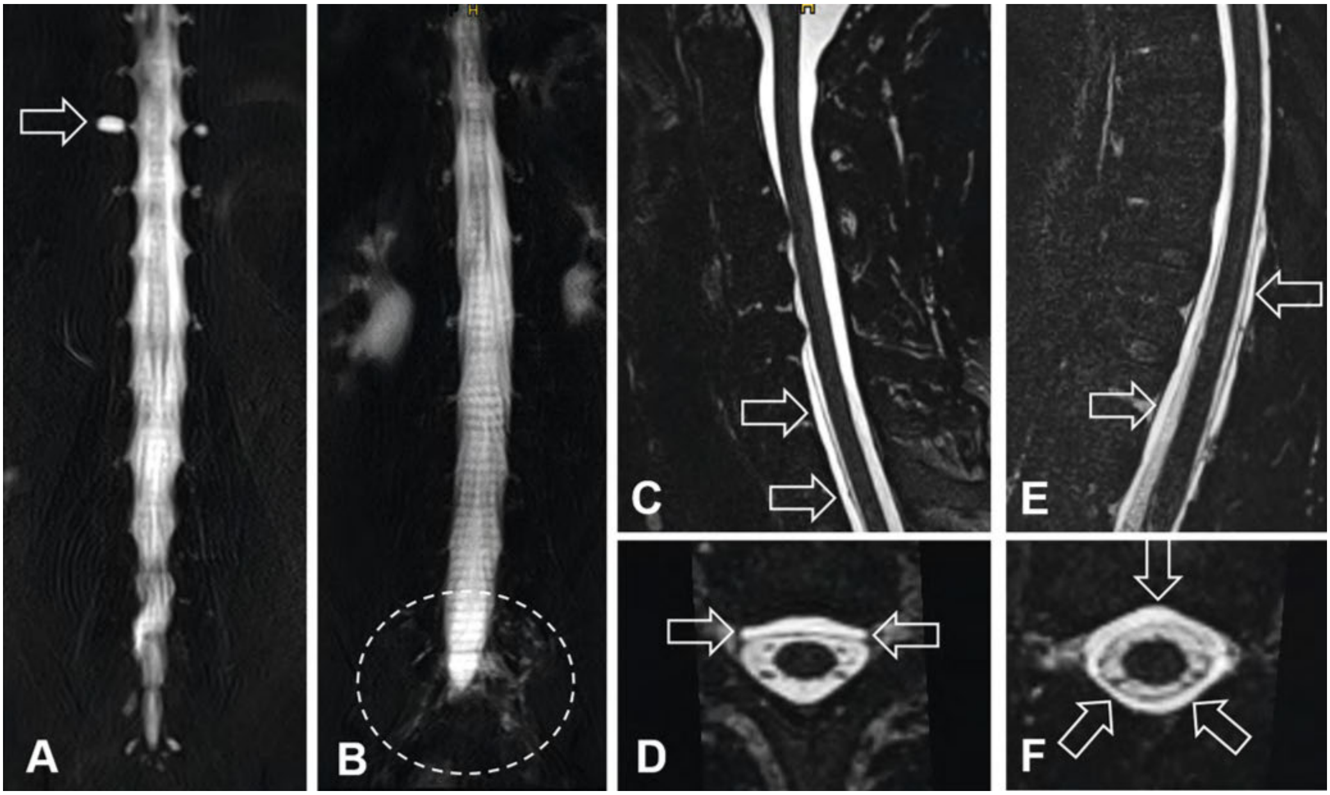
MRI of the spine

The importance of an MRI examination of not only the head but also the spine must be emphasized. As the disease progresses, the signs in the head and thus the SIH score can decrease and epidural fluid collections along the spine can be the only sign of SIH on MRI [18, 34, 35].

Heavily T2w 3 D sequences of the spine, e. g., T2 SPACE ("sampling perfection with application optimized contrast using different flip angle evolution") with fat saturation (▶ **Fig. 2C–F**) are suitable to detect or rule out this epidural fluid (as a result of a dural tear) with high sensitivity [30, 36]. Complete visualization of the sacrum should be ensured [30]. Fat saturation on T2w sequences is essential for differentiating between epidural fat and epidural fluid. Other T2w 3 D sequences like fat-saturated T2 TSE ("turbo spin echo") or T2 CISS ("constructive interference in steady state") are also possible but can sometimes have disturbing flow artifacts [30, 37]. At least isotropic layers should be acquired (slice thickness of ≤ 1.0 mm; duration approx. 5 minutes per examination block) to allow axis-corrected, three-dimensional visualization of the spinal canal [30, 36]. Coronal 2 D T2w HASTE ("half acquisition single-shot turbo spin echo") myelograms are acquired quickly (approx. 20 s per examination block) and are helpful for visualizing epidural fluid or prominent meningeal diverticula (▶ **Fig. 2A, B**) [24].

The significance of meningeal diverticula is a frequent point of discussion, especially because they are regularly found in healthy subjects. In addition, the available data regarding their significance in the case of low CSF pressure is minimal. At present, it can only be stated that CSF-venous fistulas originate from nerve root diverticulum in approximately 80 % of cases [38, 39] (which are often prominent in our experience; see ▶ **Fig. 2A**). Type 2 leaks seem to be more frequently associated with broad-based cysts adjacent to the dura (as a result of an arachnoid herniation through the lateral dural tear) [40]. However, the presence of meningeal diverticula alone does not indicate SIH [41].

Sequences of the spine after intravenous contrast administration are not needed and do not provide any additional information [30, 36]. The intrathecal administration of gadolinium is also not superior to non-contrast, heavily T2-weighted MRI of the spine with respect to the detection of epidural fluid. The same applies to the conventional CT myelography with intrathecal iodine-containing contrast agent administered with the patient in a supine position [32, 36, 42].



▶ **Fig. 2** Spinal MRI sequences in the SIH workup. Coronal, 2 D T2w HASTE myelogram of the middle thoracic spine down to and including the sacrum shows a conspicuous, prominent meningeal diverticulum at the level of thoracic vertebra (Th) 10/11 on the right (which in this case was the origin of a CSF-venous fistula, see also ▶ **Fig. 5C, D**) and example of a streaky sacral fluid collection (dashed oval white ring in **B**), which was later confirmed as a sacral dural leak. 0.74 mm 3 D isotropic, fat-saturated T2 SPACE sequence of the cervical/upper thoracic spine shows a "spinal longitudinal extradural fluid collection" (SLEC) ventrally in the sagittal slice (open white arrows in **C**) and in the axial reconstruction (open white arrows in **D**), which often indicates a ventral CSF leak. The same sequence in another case in the region of the middle and lower thoracic spine, shows a fluid collection with ventral, but also lateral and dorsal components (open white arrows in **E** and **F**), which often indicates a lateral cerebrospinal fluid leak. In this case, the dura can be clearly distinguished from the epidural fluid collection as a hypointense line (**D** and **F**).

Identification of the leak

When targeted treatment is intended, the second step in the diagnostic workup is the exact identification and localization of the CSF leak.

A special myelography or angiography suite (ideally with a tilting table) is needed for digital subtraction myelography (DSM). Any conventional CT unit can generally be used for CT myelography (CTM), with certain equipment being helpful for good patient positioning. A customized tiltable table placed on the CT patient table is used for this purpose in our hospital in order to achieve a head-down position (which can also be achieved by placing pillows under the stomach or pelvis). By acquiring multiple consecutive scans in the suspicious section of the spine, a dynamic examination can be achieved (necessary in the case of type 1 or type 2).

Examination in prone position

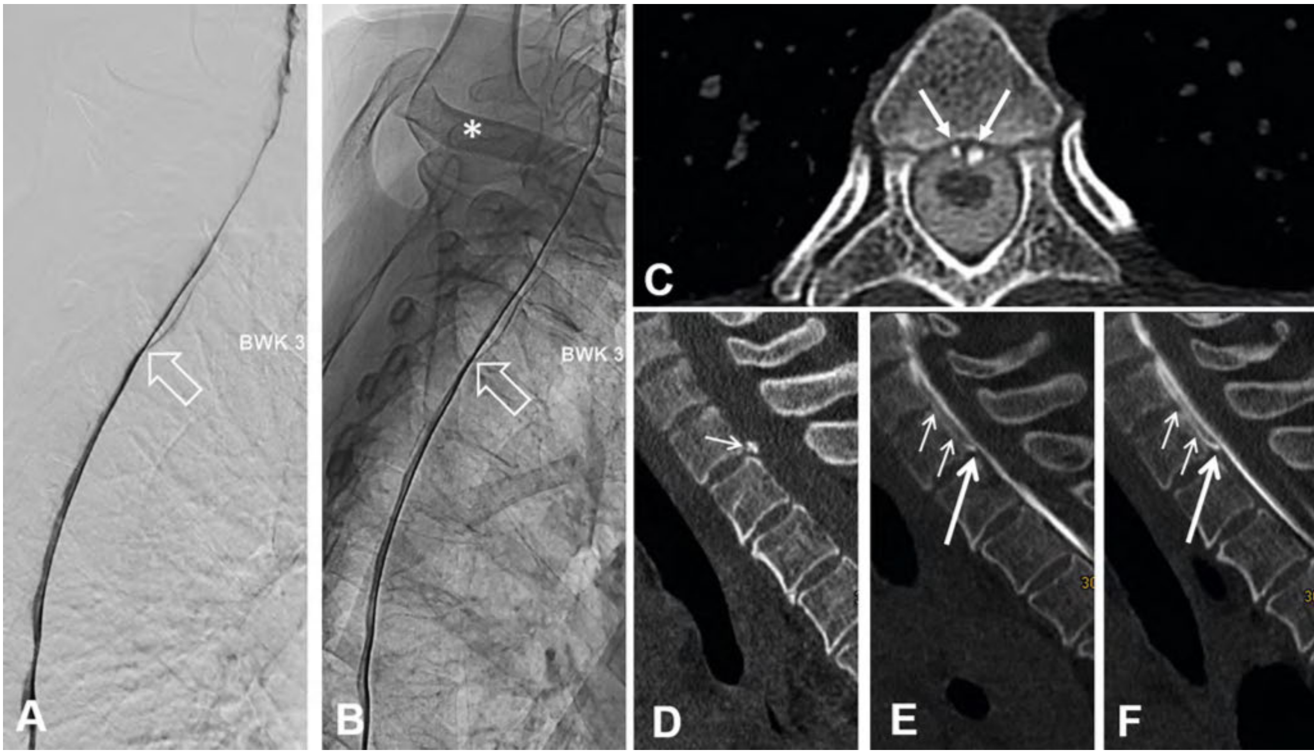
Correct positioning is decisive for sufficient diagnostic workup: If there is suspicion of a ventral leak (type 1) based on the MRI images, patients should be positioned in a prone position with the head sufficiently low (approx. 10–20°).

This significant tilting in the prone position is necessary for the contrast agent to be able to overcome the natural kyphosis of the spine and distribute uniformly to the base of the skull [20]. In our experience, ventral leaks usually have a high flow rate so that a dynamic DSM with very high temporal resolution is essential. Since the contrast agent leakage can often be seen within a couple of seconds, the site of the CSF leak can be easily missed in the case of later visualization. Alternatively, dynamic CT myelography can be performed.

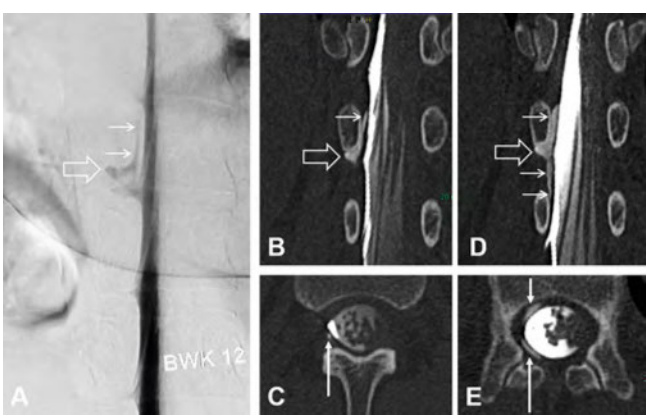
Examination in lateral decubitus position

In the case of suspicion of a lateral leak (type 2) or a CSF-venous fistula (type 3), examination in a lateral decubitus position (DSM or CTM) is necessary. A slight head-down position (approx. 6–7°) is usually sufficient to distribute the contrast agent along the spine and particularly along the diverticula. Analogous to ventral leaks, a dynamic examination is needed (DSM or CTM) for lateral leaks.

Visualization of CSF-venous fistulas (type 3) is often challenging. In general, both modalities are suitable. However, CTM was shown to be more sensitive than DSM for this purpose in a current study [43]. A dynamic examination is not necessary. The scan should be performed immediately after intrathecal contrast administration.



▶ **Fig. 3** Ventral leak (type 1), visualized with DSM and CTM. Digital subtraction myelography (DSM; **A**) in prone position and unsubtracted images of the same series (**B**) showing a ventral CSF leak at the level of Th 3/4 (white open arrow in **A** and **B**), with the contrast agent spreading cranially in the epidural space. This was caused by two paramedian bone spurs (solid white arrows in **C**), which can be seen in the subsequent CT myelography (CTM) in the supine position. Dynamic CTM in the prone position (several CT scans): In this case, the first native CT scan shows a bone spur Th 1/2 (solid white arrow in **D**). In the second CT scan, a faint contrast egress can be seen leaking into the ventral epidural space at the level of Th 1/2, which becomes clearer on the third CT scan (large solid white arrow in **E** and **F**). Cranial distribution of the contrast in the epidural space (small solid white arrows in **E, F**).



▶ **Fig. 4** Lateral leak (type 2), visualized with DSM and CTM. Digital subtraction myelography (DSM) in the right decubitus position showing a contrast egress at the level of Th 10/11 on the right (open white arrow in **A**) and linear spread of the contrast agent in the epidural space cranially (small solid white arrows in **A**). Dynamic CT myelography (CTM) in the right decubitus position to visualize a lateral CSF leak in another example: In the first CT scan at the level of Th 12/L1 on the right, an epidural contrast egress is seen coronal (open white arrow in **B**), which spreads cranially in the epidural space (solid white arrow in **B** and in the axial slice in **C**). On the immediately following second CT scan, the epidural contrast agent leakage increases (solid white arrows in **D** and **E**).

Examples of the visualization of a type 1, 2, and 3 CSF leak by DSM, CTM, and cone beam CT are shown in ▶ **Fig. 3**, ▶ **Fig. 4**, ▶ **Fig. 5**.

DSM and CTM have different advantages and disadvantages (▶ **Table 3**) that should be adjusted to the different types of leaks depending on the suspicion on the basis of the MRI finding and depending on the experience and options on-site (available examination equipment). While high doses can occur in some cases particularly during dynamic CT (multiple consecutive scans) [20], DSM has a general advantage due to the relatively lower radiation exposure [44]. Recent developments at the fluoroscopy suite include conebeam CT (rotation of one tube) to check questionable findings or to acquire ultra-high-resolution 3 D images (▶ **Fig. 5C, D**) [21, 45, 46].

The flowchart in ▶ **Fig. 6** shows the diagnostic workflow. A certain term that effectively describes the workflow has become established in scientific and clinical discourse. Patients with epidural fluid on MRI are characterized as SLEC + (spinal longitudinal extradural CSF collection), with this fluid indicating a dural tear (type 1 and 2 or sacral), while a high Bern score (Head +) with SLEC neg. spine indicates a type 3 CSF-venous fistula [18, 26].

As a result of pitfalls regarding correct positioning and the timing of contrast administration, both DSM and CTM are technically challenging and require a specific routine to achieve a high-quality result. Otherwise, there is a risk of subjecting patients to repeated examinations and unnecessary radiation exposure.

Future possibilities

Technical advances like the development of photon counting CT and the use of hybrid systems (angiography suite + CT) will potentially be important in the future: The high spatial resolution of photon counting CT with a relatively low radiation dose and spectral analysis can help to improve diagnosis and to reduce radiation exposure [47, 48]. Hybrid devices have the advantage of the ability to directly couple the two modalities DSM and CTM on-site [49].

Therapeutic approach

Conservative measures and blood patch

In the early phase of SIH, conservative treatment methods like bed rest, hydration, and caffeine (even without preceding localization of the leak) can generally achieve a temporary reduction of symptoms [4, 35]. However, there is only minimal evidence for the use of these measures for lasting treatment. After two weeks without significant improvement at the latest, a non-targeted epidural blood patch (EBP) should be considered [30]. The patch works by increasing epidural resistance and thus providing temporary compensation of the loss of CSF (immediate effect). Potential closure of the defect as a result of granulation processes is being discussed [50, 51]. It is unclear if the EBP has a lasting effect due to the heterogeneous nature of the available data [52]. Smaller studies were able to show a closure rate of approx. ⅓ of patients [53, 54]. A volume of approx. 20 ml seems to have a good effect [55].

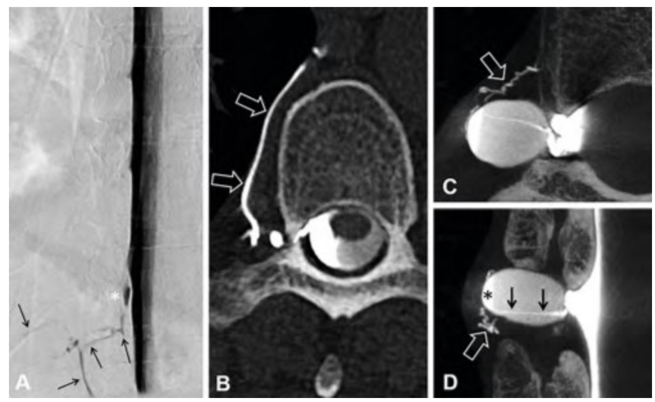
10–14 days after EBP, follow-up should be performed to evaluate the subjective clinical status [30]. If symptoms persist, a second EBP can be offered, or further diagnostic workup (step 2, see above) can be initiated in order to provide definitive treatment [30]. This approach is important particularly with respect to avoiding chronic manifestations and long-term damage [31, 56].

Even in the case of severe symptoms and complicated courses (e. g., evidence of subdural hematoma on imaging), prompt introduction of step two for leak identification is recommended [30]. In this case, early referral to a CSF center can be helpful [30].

Definitive treatment methods

Definitive treatment (surgical or interventional) requires precise localization of the CSF leak on myelography.

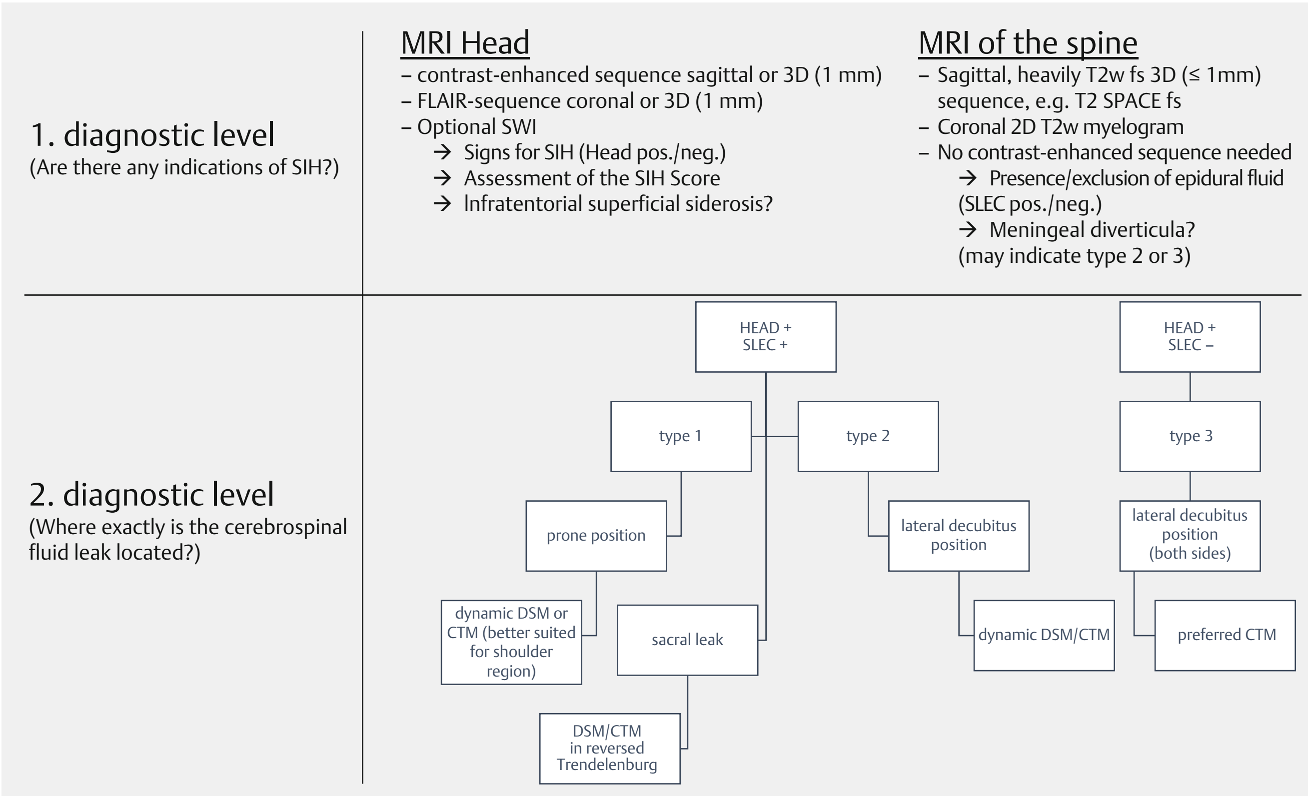
In the case of a targeted blood or fibrin patch, a puncture needle is placed as close to the exact location of the CSF leak as possible under CT guidance in order to inject blood or fibrin (approx. 2–4 ml) [57, 58, 59]. This method also has good results in patients who have not benefited from non-targeted EBP [60].



▶ **Fig. 5** CSF-venous fistula (type 3), visualized with DSM, CTM and ultra-high resolution cone-beam CT. Digital subtraction myelography (DSM) in the right decubitus position shows a small meningeal diverticulum (asterisk in **A**), from which paravertebral venous vessels contrast (solid black arrows in **A**) in the sense of a CSF-venous fistula Th 12/L 1 on the right. Another example of a CSF-venous fistula, shown in CT myelography (CTM), which was performed about 40 s after intrathecal injection of contrast agent in the right decubitus position (no dynamic CTM) and shows a contrast drain into the paravertebral vein Th 10/11 on the right (open white arrows in **B**). Ultra-high-resolution and highly zoomed image of another CSF-venous fistula Th 10/11 on the right, which originates from a prominent meningeal diverticulum (see also ▶ **Fig. 2A**), using cone-beam CT at a resolution of 0.14 mm (open white arrows in axial slice in **C** and in coronal slice in **D**). Within the meningeal diverticulum, a fine jet of contrast agent (black solid arrows in **D**) is visible between the nerve root sheath and the fundus of the cyst (black asterisk in **D**), which fills the cyst with contrast medium (not part of the actual CSF-venous fistula).

▶ **Table 3** Characteristics of the different investigation techniques in SIH workup.

Modality	Advantages	Disadvantages
DSM	▪ High temporal resolution (dynamic investigation) ▪ Especially suitable for high-flow leaks (type 1 and 2) ▪ Relatively lower radiation exposure than CTM ▪ Additional option to use cone-beam CT	▪ Limited area can be visualized (usually 49 cm detector field) ▪ Complex investigation ▪ Susceptible to artifacts (breathing, movement) ▪ Projection imaging (superposition phenomena) ▪ Small lesions can be missed ▪ Major learning curve for the examiner is to be expected
CTM	▪ Temporal resolution/dynamic investigation possible ▪ High local resolution due to 3 D imaging (thus also displaying small findings, e. g. in type 3 leaks) ▪ CT is widely used	▪ Relatively higher radiation exposure ▪ Lower temporal resolution than DSM ▪ Major learning curve for the examiner is to be expected

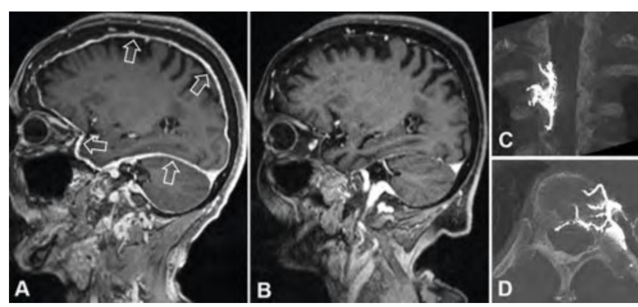


▸ **Fig. 6** Two-stage diagnostic concept in the SIH workup.

The effect has been shown in larger studies, particularly for type 1 and 3 [58, 59].

Alternatively, surgical closure is available as an option for all described leak types. A microsurgical approach, which has a high success rate and very low rate of complications (neurological complications < 2 %) if performed at a center with significant expertise, is suitable for this purpose [61]. The success rate after surgical leak closure is very high (over 90 %). However, patients with a CSF leak that has been present > 3 months can expect full recovery to take approx. 3–6 months in many cases [28]. Moreover, in the case of successful surgical closure, residual symptoms can persist in approx. one fourth of patients, which highlights the chronic nature of the disease [28].

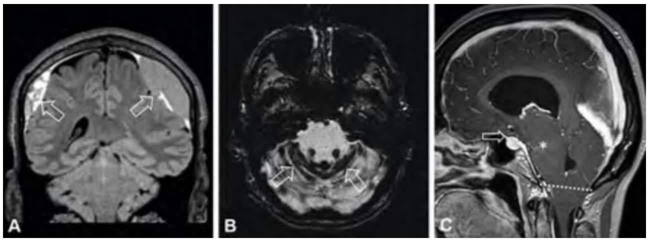
The endovascular option, which was first described in 2021, is available in addition to surgery for the treatment of CSF-venous fistulas. The fistula can be permanently closed by means of transvenous embolization of the draining, paraspinal vein using a liquid embolic agent (onyx) [62]. Given the high success rate, this method also provides a safe and minimally invasive alternative [63]. An example of MRI scans before and after transvenous embolization is shown in ▸ **Fig. 7**.



▸ **Fig. 7** Treatment response after CSF-venous fistula embolization. Preinterventional sagittal fat-saturated T1w sequence after contrast administration in an 88-year-old female patient with two synchronous CSF-venous fistulas shows pronounced pachymeningeal enhancement (open white arrows in **A**). After transvenous embolization of the CSF-venous fistulas at the level of Th 2/3 on the right and Th 10/11 on the left (coronal and axial hyperdense Onyx-cast in native cone-beam CT in **C** and **D**), the pachymeningeal enhancement is no longer detectable 4 months after treatment (**B**).

Long-term consequences and complications of diagnostic workup and symptoms

Complications are an important aspect of SIH that should not be underestimated (▸ **Fig. 8**) [10, 64]. Associated subdural hematomas (▸ **Fig. 8A**) occur relatively frequently. Particularly in younger patients with chronic subdural hematoma, CSF leaks seem to make up a relevant percentage of cases [64]. A cerebellar tonsillar herniation morphologically mimicking a Chiari malformation on imaging (▸ **Fig. 8B**) can result in the development of a syringomyelia [3, 65]. Sinus vein thrombosis or a clinical presentation similar to frontotemporal dementia (brain sagging dementia) is less commonly associated with SIH. In contrast to true frontotem-



▶ **Fig. 8** Complications of SIH. Coronal FLAIR sequence showing bilateral, space-occupying chronic subdural hematomas (open white arrows in **A**) in a 58-year-old patient with already impaired consciousness; the cause was a ventral CSF leak (type 1). Axial SWI sequence shows a streaky, infratentorial siderosis (open white arrows in **B**) in a 78-year-old patient with severe chronic progressive ataxia with tinnitus; the cause was a ventral CSF leak (type 1). Sagittal T1w sequence after contrast shows prominent "brain sagging" with no longer visible suprasellar (open white arrow in **C**) and mamillopontine distance (asterisk) as well as a tonsillar descent ("secondary Chiari", dashed white line). The cause of the 56-year-old female patient with symptoms of frontotemporal dementia, as well as gait disturbances, dysphagia, and dysarthria over many years, was a CSF-venous fistula (type 3).

poral dementia, the symptoms can completely regress after treatment provided if the leak was found [10, 66]. In the case of a persistent dural tear, irreversible long-term damage, like paresis and muscle atrophy in bibrachial amyotrophy or gait ataxia and hearing loss due to infratentorial superficial siderosis (▶ **Fig. 8C**), can occur [67, 68]. To visualize these hemosiderin deposits that presumably occur due to the chronic dural violation, the cMRI examination can be supplemented by a T2* or SWI (susceptibility weighted imaging) sequence [30, 67].

A known complication after definitive treatment of the leak is called rebound hypertension, which occurs in up to one fourth of patients [69]. This is associated with headache when in a lying position (and improvement when standing) and can usually be effectively treated with acetazolamide (lowers CSF production) [28, 70].

Conclusion

Spontaneous intracranial hypotension is a severe, underdiagnosed disease with a broad spectrum of symptoms that far exceed headaches. Knowledge of the quick (< 14 d) and step-by-step diagnostic workup and treatment is extremely important for improving long-term treatment success and avoiding long-term damage and complications for patients. Based on the complex and potentially radiation-intense diagnostic workup, collaboration with a center with appropriate expertise is helpful for ensuring targeted and successful treatment (surgical or interventional).

Conflict of Interest

H. Urbach received honoraria for lectures from Biogen, Eisai, Mbits, Lilly, Bayer, is supported by German Federal Ministry of Education and Research

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