Learning objectives

The purpose of this poster is:

- to briefly present SIH,

- to elucidate the role and modalities of spinal imaging in this disorder,

and discuss the treatment options based on imaging findings.
Background

Orthostatic headache, the hallmark presentation of SIH has been known for more than a century. However, better understanding of this enigmatic entity only came with the widespread use of modern neuroimaging in the last 2 decades. Since then, knowledge of this disorder has been amassing rapidly. Recognition of broader clinical spectrum and numerous imaging features of SIH enables us to identify a much larger number of patients than previously.

A distinction must be made between SIH and intracranial hypotension (IH) due to trauma or surgery. Despite a similar cause and the hallmark presentation of orthostatic headache, these two groups of patients can differ significantly in the natural history of disorder, prognosis and optimal treatment. This presentation only focuses on patients with SIH. A similar distinction holds true for CSF leaks; some are spontaneous while other are posttraumatic/surgical. In this presentation, the term CSF leak is used to denote a spontaneous leak, unless indicated otherwise.

It has been established that the main pathogenic factor in SIH is actually CSF hypovolemia. Because of this, alternate terms for SIH have been proposed such as spontaneous CSF leak or CSF volume depletion.

Diagnosis

The first diagnostic criteria for SIH were provided by the International Classification of Headache Disorders - second edition (ICHD - II). Since they proved to be too restrictive for the varied clinical and radiological presentation, new criteria for SIH were proposed by Schievink et al. in 2008: criterion A, the demonstration of extrathecal CSF on spinal imaging. If criterion A is not met, criterion B, which is cranial MR imaging findings of spontaneous intracranial hypotension, follows, with at least one of the following: 1) low opening pressure, 2) spinal meningeal diverticulum, or 3) improvement of symptoms after epidural blood patch. If criteria A and B are not met, there is criterion C, the presence of all of the following or at least 2 of the following if typical orthostatic headaches are present: 1) low opening pressure, 2) spinal meningeal diverticulum, and 3) improvement of symptoms after epidural blood patch. Based on these criteria and advances in understanding of SIH new criteria for headache due to SIH have been presented and will most likely be included in the new ICHD classification (ICHD - III beta).

Epidemiology
SIH is known to occur in all age groups, however it is most common in female patients aged from 35 to 42. Incidence has been estimated to be from 2-5/100 000 people per year.

Clinical presentation

Most common presentation of SIH is an orthostatic headache; this type of headache typically worsens in the upright position, while lying down relieves the pain. It can be mild or severely painful, usually it is bilateral and frontal, however it can present in many different ways. In time, the orthostatic nature of the headache usually becomes less pronounced; it can take much longer than just a few minutes for the change of stature to affect the pain and sometimes chronic daily headache evolves. Orthostatic headache is therefore in no way pathognomonic for SIH - not all orthostatic headaches are caused by SIH, some patients with SIH never develop it and some have a different type of headache. Other clinical manifestations such as nausea, neck stiffness, radicular and cranial nerve symptoms, tinnitus, dizziness, vertigo and other are thought to arise from diminished CSF volume causing sagging of the brain and traction and/or compression on the various intracranial structure such as cranial nerves, brain lobes and brainstem. Increased volume of intracranial and epidural venous structures and changes in perilymph pressure in the middle ear may also account for a part of the symptoms.

Treatment

Numerous treatment options are available to treat SIH. However, the efficiency of many has not been undoubtedly confirmed; in addition, no randomized trials to assess the different methods have been conducted yet.

Conservative measures are often all that is needed - many CSF leaks close on their own - and include bed rest and possibly hydration. Medical treatment includes caffeine and theophylline (both increase CSF production), which have been proven to have an effect, although it is often mild and transient. Corticosteroids have been used, but are not routinely advised, since they only provide partial benefit to a subgroup of patients and have a large potential for side effects. Among the unproven methods are compressive abdominal binders, such as corsets.

In cases where conservative measures fail to induce a response in one to two weeks, more invasive methods come in consideration.

Currently, the most commonly employed invasive method of SIH treatment is the epidural blood patch (EBP). In this procedure autologous blood (10 - 80 ml) is injected in the epidural space which results in early effect due to compression of dural sac - apparent increase in CSF volume and late effect of sealing the site of the CSF leak.
EBP can be performed as targeted in the area of the diagnosed leak or blindly at the lumbar level, on a single or on multiple levels. After the procedure, patients should rest in horizontal or Trendelenburg position for up to 24 hours. It has been shown that maintaining Trendelenburg position after the procedure facilitates the spread of epidural blood in cranial direction. Therefore even in the cases of CSF leaks in the thoracic and cervical spine, EBP may be done in the lumbar region, where the procedure is easier and associated with less risk.

The success rate of single EBP has been estimated between 30 and 70%. In case of inefficiency the procedure can be repeated up to a total of three times. When three consecutive EBPs have failed to provide an improvement, additional imaging is warranted to diagnose the site of the leak (if not already known).

If the leak is discovered, targeted EBP is the next treatment option. When EBP prove ineffective and the location of the CSF leak is known, CT guided application of fibrin glue (sometimes combined with blood) can be considered.

In a select group of patients with identified leak site and severe symptoms refractory to other measures surgical procedures can be performed.
Findings and procedure details

Imaging plays two roles in patients with SIH; first, in diagnosing the condition by identifying typical imaging abnormalities and second, in finding the exact CSF leak site. Brain MRI is the standard investigation in diagnosing SIH. Depicted abnormalities are the result of CSF hypervolemia, which results in sinking of the brain and, according to Monro-Kellie doctrine, results in compensatory dilatation of venous spaces. Most commonly described intracranial MRI signs of SIH are diffuse pachy-meningeal enhancement, subdural fluid collections (hygromas and hematomas), engorgement of venous structures (dural sinuses - venous distension sign), enlarged pituitary gland, sinking of the brain which results in descent of cerebellar tonsils, reduced size of cisterns and flattening of optic chiasm.

The role of spinal MRI in patients with SIH is not yet clearly established so that it is not yet routinely recommended in diagnosing SIH. However, it has the potential to both detect typical abnormalities needed for diagnosis and sometimes even diagnose the CSF leak site - the cause of SIH. This has been indicated by Watanabe et al. in a small study which demonstrated higher sensitivity of spinal MRI than brain MRI for SIH specific imaging abnormalities. The results of this study stressed the usefulness of spinal MRI in diagnosis of SIH in early stage in patients without brain MRI findings. Spinal MRI finding in SIH patients are:

1. Distended spinal epidural veins (75-100% sensitivity); like most of the spinal manifestations, the distension of epidural veins (as well as sometimes intradural spinal veins) is a consequence of a collapsed intraspinal dural sac (due to reduced CSF volume), which is not firmly adherent to the overlying bone. A distended anterior epidural venous plexus should always be considered abnormal in the high cervical spine, where it is normally not seen.

2. Dural collapse is seen as a hexagonal shape of dural sack in axial plane, commonly described as “festooned” appearance. The specific shape is the result of reduced CSF pressure within the sack and attachments of it to the surrounding ligamentous structures.

3. Epidural fluid collections are the result of CSF leakage. Fluid collection mostly span more than one segment; in the cervical spine most are located anteriorly, in the thoracic region, posterior are somewhat more common, while both are common in the lumbar area - locations correlate to the amount of epidural space at different levels.

4. Abnormalities in paraspinal soft tissues (along the nerve sleeves) seen as fluid collections, are an uncommon finding and may point to approximate location of CSF leak.

5. Spinal dural contrast enhacement.

6. Meningeal diverticula, which may be the site of leakage.
7. Site of CSF leakage and dural tears, a rare finding on plain spinal MRI.

The main pathological factor same to all SIH patients is, as already mentioned, CSF hypovolemia. In most cases this is attributable to leakage of CSF through a dural defect on the level of the spine. Since CSF leak is not always demonstrable, other possible causes of CSF hypovolemia have been proposed - by pooling of CSF in dilated dural sack or loss of CSF due to diffusely increased permeability of meninges. Determination of exact site of CSF leak is difficult and often requires specialized, invasive imaging procedures. However localizing the leak is not always necessary. Since most patients experience symptom relief after conservative treatment or non-targeted EBP, current management guidelines only recommend localizing the leak site in patients who do not respond to three non-targeted EBP.

Methods used to diagnose leak sites are:

1. MR myelography (MRM) has been shown to be effective for detecting leaks, and is a non-invasive, does not require contrast injection and does not expose the patient to radiation, however has lower spatial resolution than CTM.

2. Radionuclide cisternography (RNC) with Indium-111 labeled DTPA injected intrathecally is sensitive in diagnosing indirect signs of SIH (early demonstration of tracer in urinary system, slow ascent along the neural axis and the absence of activity over the hemispheres at 24 hours) and can aid in detecting the leak. The weaknesses of this technique are low resolution, lack of correlation with morphology, possibility of iatrogenic leaks and radiation exposure. Sensitivity can be increased with the use of simultaneous CT/SPECT imaging.

3. CT myelography (CTM) with iodinated contrast agent administered intrathecally is considered by many as the study of choice for localization of CSF leaks. Beside the CSF leak discoverable anomalies include epidural and extraspinal fluid collections with possible contrast extension and meningeal diverticula (which may be the sites of the leak). Based on the rate of leakage, CSF leaks are divided in high- and low-flow leaks. Demonstrating each kind requires different imaging approaches. High flow leaks are best visualized using dynamic CTM, the aim of which is to shorten the interval between contrast administration and imaging by scanning during and immediately after contrast injection. Low-flow leaks are best shown by delayed imaging from three to 72 hours after contrast administration. Additional imaging strategies in low-flow leaks are positive pressure CTM or scanning after the patient was in upright stature or walking.

4. Digital subtraction myelography (DSM) has been successfully used for high-flow leaks, particularly in conjunction with other imaging modalities.

5. MR myelography (MRM). Gadolinium contrast agents have not yet been approved for intrathecal use, therefore MRM is an off label use of MR contrast. Despite this, several studies using diluted contrast agent demonstrated no notable side effects of this procedure. If such use of contrast proves safe, MRM will provide an alternative to CTM, with better contrast resolution and lack of ionizing radiation.
CSF leaks may be located anywhere in the spine; traditionally it has been assumed that the majority of leaks lie at the cervicothoracic junction or in thoracic spine. This view has been challenged by recent reports, which established myelographic extravasation of contrast at the level of C1-2 and at the cervicothoracic junction to be a common false localizing sign. Surgery and targeted percutaneous treatment at these levels are therefore not advised. Spontaneous CSF leaks located at the level of the skull do not induce SIH.

The exact cause of CSF leak usually remains unclear. Most papers agree that a preexisting dural weakness can spontaneously or after a minor trauma produce a CSF leak. Different dural abnormalities are more common in SIH patients than in general populations such as meningeal diverticula. Patients with disorders of connective tissue matrix are more likely to develop SIH due to meningeal diverticula or dural ectasia commonly present in this population. Spondylotic spurs and herniations of intervertebral disci have been recognized to cause ventral CSF spinal leaks.

Despite numerous reports and obviously wide clinico-radiological range of findings no serious attempt has been made to categorize or classify SIH patients until 2016, when Schievink et al. published the first categorization of SIH patients depending on the type of discovered CSF leak. Their classification recognizes three types of leaks and a fourth group for patients without demonstrable leak.

Type 1 CSF leaks (about a quarter of their cases) are leaks through a dural tear and are subdivided in 1a (ventral, vast majority) and 1b (dorsal, minority of cases) groups. In practically all cases extradural fluid collection was associated with these leaks.

Type 2 CSF leaks (less than half of cases) consist of meningeal diverticula; subgroups a and b denote the type of diverticula (a - simple single or multiple and b - complex diverticula). Only about fifth of these patients had identifiable extradural collection.

Type 3 CSF leaks (rare cases) represent a direct CSF venous fistula. Predictably no extradural collections were noted in this group.

Type 4 CSF leak (about a quarter of cases) is an indeterminate type of leak; about half of patients from this group had a spinal fluid collection.

Management of SIH in light of the new classification

How does a known location of CSF leak influence management?

Knowing the location of the CSF leak is vital if the first line treatment (EBPs) fails. Application of fibrin glue can only be performed as a targeted procedure and surgical procedures are more effective if the surgeon is able to directly visualize and repair the leak (which is often only possible on the base of radiological findings) instead of non-targeted procedures.
Known leak location also enables targeted EBPs if non targeted ones fail.

What are possible influences of CSF leaks classifications on SIH management?

Since only a single study using (and establishing) the proposed CSF leak classification has been published, no definite recommendations for management can yet be made. The results of this study, however, show some interesting emerging patterns.

Percutaneous treatment seemed to be less effective in type 1 leaks; since most of these are ventral and often have associated calcifications these factors could be a part of the explanation.

Type 2 CSF leaks form a more complex group; in most of the patients with meningeal diverticula the exact leak site has not even been demonstrated; therefore the exact role of diverticula (as a direct leak site, sign of dural frailty or as by the process of CSF pooling) has not yet been established.

CSF venous fistulas (type 4 leaks) have been successfully treated with non-targeted (or targeted) EBPs; all of the cases were only successfully treated with targeted approaches - fibrin glue applications or (mostly) with surgery. This seems to be the rarest form of leak. However it has been the last type of leak discovered (2014) and not many cases have been reported yet.

In cases where exact leak locations have not been found with initial imaging, but the patients respond to non-targeted EBPs, more invasive and sensitive diagnostic imaging is usually not undertaken. Therefore leaks that could have been located and specified remain unknown. Better characterization of leaks in this patient group may reveal useful new information.
Fig. 1: Sagittal T1 weighted MR image of 31 year old male without history of trauma. Sagging of the brain with reduced pontomesencephalic angle is seen. The pituitary gland is marginally enlarged.

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**Fig. 2:** Coronal T2 weighted MR image of the same patient as in Figure 1. Bilateral large subdural hematomas (arrows).
**Fig. 3:** Sagittal T1 weighted brain MR image showing sagging of the brain with associated reduced pontomesencephalic angle, flattening of the pons (arrow), obliteration of basal cisterns, and descent of cerebellar tonsils (asterisk).
Fig. 4: Sagittal T1 weighted brain MR image of the same patient as in Figure 3; slightly more lateral view, showing mid-portion of dominant transverse epidural sinus with convex inferior margin - venous distension sign (arrow).
Fig. 5: Sagittal T1 weighted brain MR image of the same patient as in Figures 3 and 4 showing enlarged pituitary (arrow) and descent of cerebellar tonsils (open arrow).
**Fig. 6:** Axial (a) and sagittal (B) T2 weighted MR images of cervical spine show a large anterior epidural fluid collection and compression myelopathy at the site of the effusion.
Fig. 7: Sagittal T2 weighted MR image of thoracic spine of the same patient as in Figure 6 demonstrates large anterior epidural collections (asterisks). B: Sagittal T2 weighted MR image of thoracic spine in same patient slightly laterally shows ligamentous attachments (arrows) of anterior spinal dura (empty arrow) to vertebral column, which are responsible for the hexagonal festooned appearance of the dural sac in SIH (not shown).
**Fig. 8:** Sagittal T1 weighted image of thoracic spine (A) shows fluid collection (full arrows) in the posterior epidural fat and a possible site of leak (arrow) at the level of Th7. B, 3 months later, after treatment, absence of the fluid collection and leak is seen.
Fig. 9: Axial T1 weighted brain MR image after contrast injection shows diffuse pachymeningeal enhancement (arrows) and ventricular collapse (empty arrows) and thin subdural collections.

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Fig. 10: Sagittal T1 weighted MR images of cervical spine before (A) and after (B) contrast administration show distended anterior epidural venous plexus (arrow), as well as signs of brain sagging in the posterior fossa. C: The distended epidural veins (arrows) are appreciated as prominent flow voids in the anterior epidural space on the sagittal STIR weighted MR sequence (position slightly lateral to A and B.)
**Fig. 11:** Axial T1 weighted MR image of cervical spine on the level of C1/2 after intrathecal administration of gadolinium contrast agent showing extradural localization of contrast media posteriorly on the right (arrow) and retrospinally (open arrows); this could be interpreted as the CSF leak site. However, research has shown that extradural CSF collections at this level can be seen with confirmed leaks at lower cervical, cervicothoracic or even thoracic levels and no leakage at the C1/2 level. The pathophysiology of these "false localizing" collections is still subject to debate.

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Conclusion

Spinal imaging plays an important role in SIH patients. It enables localization and classification of CSF leaks. This allows a better understanding of different subgroups of SIH patients and more efficacious, targeted approach to treatment.
References


