

Review Article

Spontaneous Intracranial Hypotension: 10 Myths and Misperceptions

Peter G. Kranz, MD ; Linda Gray, MD; Timothy J. Amrhein, MD

Objective.—To discuss common myths and misperceptions about spontaneous intracranial hypotension (SIH), focusing on common issues related to diagnosis and treatment, and to review the evidence that contradicts and clarifies these myths.

Background.—Recognition of SIH has increased in recent years. With increasing recognition, however, has come an increased demand for management by neurologists and headache specialists, some of whom have little prior experience with the condition. This dearth of practical experience, and lack of awareness of recent investigations into SIH, produces heterogeneity in diagnostic and treatment pathways, driven in part by outdated, confusing, or unsubstantiated conceptions of the condition. We sought to address this heterogeneity by identifying 10 myths and misperceptions that we frequently encounter when receiving referrals for suspected or confirmed SIH, and to review the literature addressing these topics.

Methods.—Ten topics relevant to diagnosis and treatment SIH were generated by the authors. A search for studies addressing SIH was conducted using PubMed and EMBASE, limited to English language only, peer reviewed publications from inception to 2018. Individual case reports were excluded. The resulting studies were reviewed for relevance to the topics in question.

Results.—The search generated 557 studies addressing SIH; 75 case reports were excluded. Fifty-four studies were considered to be of high relevance to the topics addressed, and were included in the data synthesis. The topics are presented in the form of a narrative review.

Conclusions.—The understanding of SIH has evolved over the recent decades, leading to improvements in knowledge about the pathophysiology of the condition, diagnostic strategies, and expanded treatments. Awareness of these changes, and dispelling outdated misconceptions about SIH, is critical to providing appropriate care for patients and guiding future investigations going forward.

Key words: spontaneous intracranial hypotension, cerebrospinal fluid leak, cerebrospinal fluid pressure, CSF hypovolemia, orthostatic headache

Abbreviations: CSF cerebrospinal fluid, MRI magnetic resonance imaging, NDPH new daily persistent headache, POTS postural orthostatic tachycardia syndrome, RIH rebound intracranial hypertension, SIH spontaneous intracranial hypotension

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INTRODUCTION

Spontaneous intracranial hypotension (SIH) is an important cause of secondary headaches resulting

from spinal cerebrospinal fluid (CSF) leaks that has received increasing attention over the past few decades. Although historically perceived as rare, SIH is now being recognized more commonly; the incidence of SIH has been estimated at 5 per 100,000. For comparison, aneurysmal subarachnoid hemorrhage, which is not generally perceived as a rare medical condition, occurs at a rate of 10 per 100,000.¹ The

From the Division of Neuroradiology, Department of Radiology, Duke University Medical Center, Durham, NC.

Address all correspondence to Peter G. Kranz, DUMC Box 3080, Department of Radiology, Durham, NC 27519, USA.

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true incidence of SIH is expected to be higher, as it is frequently misdiagnosed initially.²

Although awareness of this condition is increasing, exposure of providers to SIH remains infrequent in most practices, meaning that levels of practical experience in diagnosis and management are generally low. At the same time, a more complex understanding of the underlying pathophysiology of the condition has emerged in recent years, fed by numerous avenues of accelerating scientific investigation.³ As a consequence, conceptions about appropriate diagnosis and treatment of SIH are sometimes based on outdated or insufficient information.

The purpose of this narrative review is to present common myths and misperceptions about SIH that we encounter frequently in our practice at a large referral center. The review will also discuss the evidence that contradicts these misperceptions, and will provide an up-to-date summary of current knowledge about this condition.

METHODS

A list of 10 topics considered most relevant to diagnosis and treatment SIH was generated by the authors, reflecting common misconceptions that we frequently encounter in our practice. A search for studies addressing SIH was conducted using PubMed and EMBASE. The search was limited to English language only, peer reviewed publications from inception to 2018. Individual case reports were excluded. The resulting studies were reviewed for relevance to the topics in question.

RESULTS

The search generated 557 studies addressing SIH. Of these, 75 were excluded for being case reports, leaving 482 studies for review. The majority of included studies (284, 59%) were published in the last 10 years. Fifty-four studies were considered to be of high relevance to the topics addressed, and were included in the data synthesis.

DISCUSSION

A discussion of the literature most relevant to the 10 most common myths we encounter in dealing with SIH is presented in the narrative review

that follows. The most important points are summarized in the Box.

Myth 1: SIH Is Defined by Low CSF Pressure.—The original conception of SIH was as a condition caused by low CSF pressure. Early reports of the condition considered low pressure to be the defining pathophysiological disturbance, the *sine qua non*, responsible for SIH.^{4,5} Subsequent work, however, has clearly shown this conception to be incomplete.

Studies of patients with SIH conducted in the 1990s found that a low opening pressure (<6 cm H₂O) was present in the majority of cases—more than 80% in 2 series.^{6,7} Since that time, several larger studies have found that the prevalence of low CSF pressure in SIH is not nearly as high as was originally reported. In a series of 106 patients, Kranz et al found only 34% of patients with confirmed SIH had an opening pressure <6 cm H₂O; most patients had pressures of 6-20 cm H₂O, and a few patients with active CSF leaks were found to have pressures >20 cm H₂O.⁸ Yao et al found that 45% of patients had opening pressure of >6 cm H₂O in a series of 206 patients,⁹ and Luetmer et al found that only 21% of patients had a pressure of ≤5 cm H₂O in a series of 76 patients, whereas 43% had pressures between 10 and 18 cm H₂O.¹⁰ These data indicate that while opening pressure is often low in SIH patients, normal pressure is also frequently found, and a normal pressure should not be used to exclude the diagnosis.

At least part of this discrepancy between earlier and more contemporary descriptions of SIH is probably attributable to spectrum bias. Initial reports of SIH likely selected for the most severe and most acute patients with the most stereotypical symptoms. Over the recent decades, it has been recognized that SIH does not always present with such stereotypical symptoms and imaging findings,¹¹ and that symptom duration may affect presentation and diagnostic testing. For example, it has been shown that CSF pressure increases with time even if an active CSF leak is still present.^{8,12} More recent investigations into CSF pressures in this condition are thus likely more representative of the true spectrum of disease, and hence demonstrate broader phenotypes. Regardless of the explanation, the empiric evidence now unequivocally

Text Box**SUMMARY OF IMPORTANT CONSIDERATIONS IN THE APPROACH TO SIH**

- Currently, there is no single diagnostic test that excludes SIH with a high level of sensitivity. When SIH is suspected, a combination of brain imaging, spine imaging, and CSF pressure measurement may be necessary for a complete evaluation.
- Orthostatic headache is the most common, but not the only, clinical presentation of SIH. The lack of orthostatic headache should not exclude the diagnosis when there is objective imaging evidence of SIH.
- Orthostatic headache can be caused by other conditions such as POTS, cervicogenic headache, and craniocervical instability. Some cases of chronic daily headache with orthostatic features may defy easy classification.
- Diffuse, smooth dural enhancement on brain MRI is a highly specific finding for SIH.
- Cerebellar tonsillar ectopia can be seen with both SIH and Chiari I malformation. Other imaging signs of brain sagging can help differentiate these entities.
- Spinal imaging is often useful in detecting CSF leaks, which can help to confirm the diagnosis of SIH and direct treatment efforts.
- Causes of CSF leaks include meningeal diverticula, ventral dural tears caused by disk protrusions or osteophytes, and CSF-venous fistulas.
- SIH is not typically caused by skull base CSF leaks.
- Recovery from spontaneous spinal CSF leaks differs from lumbar puncture related headaches, typically taking longer.
- Recovery from SIH can be complicated by RIH. Awareness of this entity is important for appropriate management after treatment for a spinal CSF leak.

indicates that SIH is not excluded by a normal CSF opening pressure.

If low CSF pressure is not the common underlying pathophysiology of SIH, then what is? The evidence suggests that low CSF volume, not pressure, is the principal problem.¹¹ The relationship between pressure and volume is governed by the physiologic concept of compliance. Compliance, however, is not a fixed attribute with regards to the CSF compartment. It changes between upright and recumbent posture,¹³ changes with removal of CSF,¹⁴ and is altered by body habitus.¹⁵ Compliance may be influenced by the spinal epidural venous plexus, which surrounds the thecal sac and is found to be dilated in some, but not all, patients with SIH.¹⁶ The rate of CSF leakage, and by extension, the degree of CSF volume depletion, is also different between SIH patients.¹⁷ This individual variation may explain the

fact that not all patients with SIH will have low CSF opening pressure, despite a common unifying problem of low CSF volume.

Myth 2: SIH Is Always Characterized by Orthostatic Headache/Orthostatic Headache Is Always SIH.—The classic symptom of SIH is headache that worsens with upright posture (termed orthostatic or positional headache). It is often described as being relieved quickly upon lying down and aggravated within minutes of standing up. Headaches are commonly occipital in location and abrupt in onset. Tinnitus is a common complaint, as are neck pain and interscapular pain. Valsalva maneuvers often elicit a sudden worsening of headache.

Although these symptoms are the most stereotypical manifestations of spinal CSF leak, the actual range of headache phenotypes are quite broad. The degree of relief provided by recumbent position

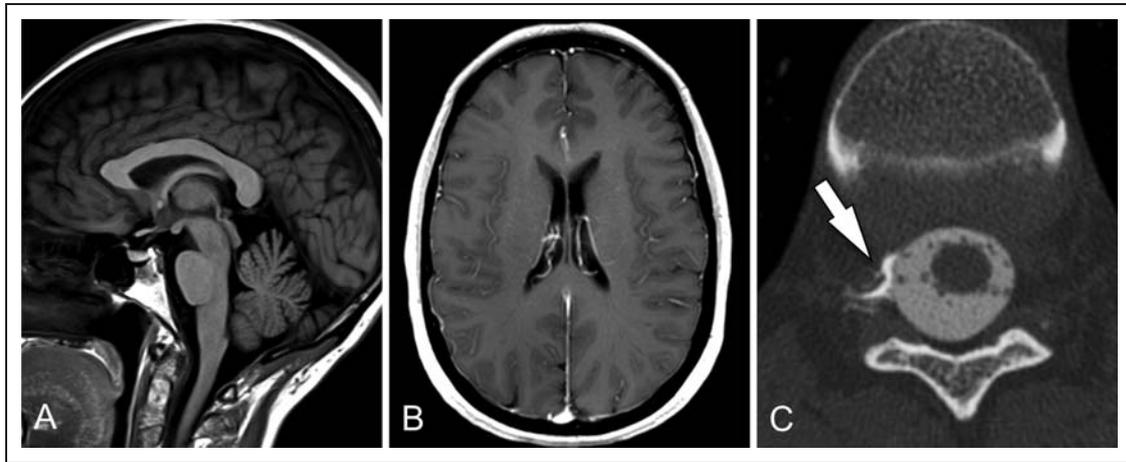


Fig. 1.—Fifty-five-year-old woman with abrupt-onset orthostatic headache. (A) Sagittal T1-weighted brain MR image shows no brain sagging. (B) Axial T1-postcontrast brain MR image shows no dural enhancement. (C) Axial post-myelogram CT image of the thoracic spine shows a focal CSF leak, confirming the diagnosis of SIH, despite a normal brain MRI.

varies from complete relief when lying down to only mild decrease in headache. Headaches that gradually worsen over the course of the day resulting in increasing pain in the afternoon and evening hours, so-called “second half of the day” headaches, are quite common.¹⁸ A minority of patients may have “wake up” headaches that are present even before getting out of bed in the morning. Other patients may have orthostatic features initially that evolve into non-positional headache, and headache might disappear entirely despite the persistence of objective evidence of CSF leakage and other related symptoms, such as tinnitus or vertigo.¹⁹ Uncommonly, headache might be non-orthostatic or absent entirely.¹¹ In a study of 70 patients with SIH, Mea et al found that 23% of patients with SIH lacked typical orthostatic headache.²⁰ When headaches are absent (termed the “acephalgic” form), auditory symptoms, such as muffled hearing or tinnitus, and ear pain or pressure are common complaints.¹¹

Headaches that worsen when upright should prompt suspicion for SIH, but SIH is not the only type of headache that exhibits an orthostatic component.³ Postural orthostatic tachycardia syndrome (POTS) has been well documented to cause orthostatic headache in some patients.²¹ POTS should be suspected particularly when headache is accompanied by other symptoms of orthostatic intolerance including dizziness, syncope or pre-syncope, or

palpitations. Cervicogenic headache can also cause headache when upright due to axial loading of the spine,²² and is often occipital in location. It may be aggravated by mechanical stimuli such as head turning.²³ Craniocervical instability can cause headache, and is more common in patients with connective tissue disorders such as Ehlers-Danlos.²⁴ Finally, some patients may have orthostatic headaches with no demonstrable signs of intracranial hypotension or spinal CSF leak, and no response to epidural blood patching. Such patients with chronic daily orthostatic headache may defy easy diagnostic categorization, and might be classified as having new daily persistent headache (NDPH).

Myth 3: A Negative Brain MRI Excludes SIH.—Brain imaging findings of SIH have been extensively described in the literature.²⁵ Perhaps the most characteristic is the presence of diffuse, smooth enhancement of the dura.^{26,27} Other signs include sagging of the midbrain, dilation of the dural venous sinuses, and hyperemia of the pituitary gland.²⁵ Subdural effusions may occasionally be present bilaterally.²⁶

These signs reflect the physiology of low CSF volume, and yet paradoxically, they are not always present even when an active spinal CSF leak is proven or CSF pressure is low (Fig. 1). In one study of confirmed cases of SIH, dural enhancement, brain sagging, and venous distention were present in 83, 61, and 75% of patients, respectively.¹²

Furthermore, the presence of the imaging signs correlated poorly with the presence of a low CSF pressure. Together, these findings suggest that brain imaging, while highly specific, is only moderately sensitive for the detection of SIH.

To be clear, brain MRI plays a very important role in the diagnosis of SIH, and should be the first test performed whenever this condition is suspected. Performing the study with IV administration of a gadolinium-based contrast agent is necessary, since dural enhancement is the most sensitive of the brain imaging signs. However, one must be cautious not to over-estimate the sensitivity of brain imaging; the diagnosis of SIH should not be excluded based on a negative brain MRI. In instances where brain imaging is negative, further testing with spinal imaging and CSF pressure measurement should be pursued if SIH is suspected.

Myth 4: Patients With Dural Enhancement Should Be Worked Up for Meningitis.—When present, dural enhancement is a highly specific indicator of SIH. To those unfamiliar with the imaging of SIH, however, the presence of dural enhancement may prompt an evaluation for other infectious, granulomatous, or neoplastic disorders. These investigations may delay treatment of SIH, and may result in invasive diagnostic procedures, such as dural biopsy. In our experience, the most common error of this type is attribution of dural enhancement to infection, resulting in extensive testing of the CSF for pathogens.

Such investigations are usually unnecessary, however, because the pattern of dural enhancement caused by low CSF volume is quite different from abnormalities of the dura caused by other etiologies. Specifically, dural enhancement in SIH has 2 cardinal features: it is smooth, and it is diffuse.²⁶

Other disease processes may involve the dura, but they produce a pattern on imaging that is either not smooth (ie, nodular or plaque-like) or not diffuse (ie, localized or unilateral; Fig. 2). For example, granulomatous diseases such as sarcoidosis or granulomatosis with polyangiitis (formerly known as Wegener granulomatosis) usually show nodular, plaque-like, or mass-like dural thickening.²⁸ Metastatic disease similarly shows multiple dural masses,

not smooth dural thickening. Dural enhancement due to a previous subdural hematoma or craniotomy may be smooth, but are localized to the affected side of the skull. Idiopathic hypertrophic pachymeningitis, a rare cause of dural enhancement, is typically irregular and does not involve all the dura uniformly.²⁹

Importantly, infectious meningitis does not typically cause dural enhancement at all because it involves the leptomeninges rather than pachymeninges (dura). The dura may enhance in some cases of infection where the infectious source is external to the dura and invades inward, such as in complicated sinusitis, mastoiditis, or infection of a craniotomy site, but in these cases the dural enhancement is localized at the site of infection, and is not diffuse.

In summary, the presence of smooth, diffuse dural enhancement is essentially pathognomonic for decreased intracranial CSF volume, especially in the context of orthostatic headache. Pursuing extensive diagnostic testing for infectious, granulomatous, or neoplastic disorders in this context is unlikely to reveal alternate diagnoses and may delay care.

Myth 5: Chiari I Is a Feature of SIH.—Brain sagging is a common imaging finding in SIH,¹² and may also be present in iatrogenic or post-traumatic causes of CSF leak. Descent of the brain results in several identifiable anatomic changes: effacement of the suprasellar cistern, downsloping of the third ventricular floor resulting in descent of the mammillary bodies, narrowing of the prepontine cistern, narrowing of the vertical distance between the mammillary bodies and the pons (the so-called mammillo-pontine distance), and descent of the cerebellar tonsils toward the foramen magnum (Fig. 3).^{27,30} This latter imaging finding may cause confusion because cerebellar tonsillar ectopia is also a feature of Chiari I malformation, and both SIH and Chiari I may cause headache.

Chiari I malformations are congenital malformations that are thought to be the result of an abnormally small posterior fossa.³¹ As a result, they would not be expected to develop later in adulthood. The presence of new tonsillar ectopia that

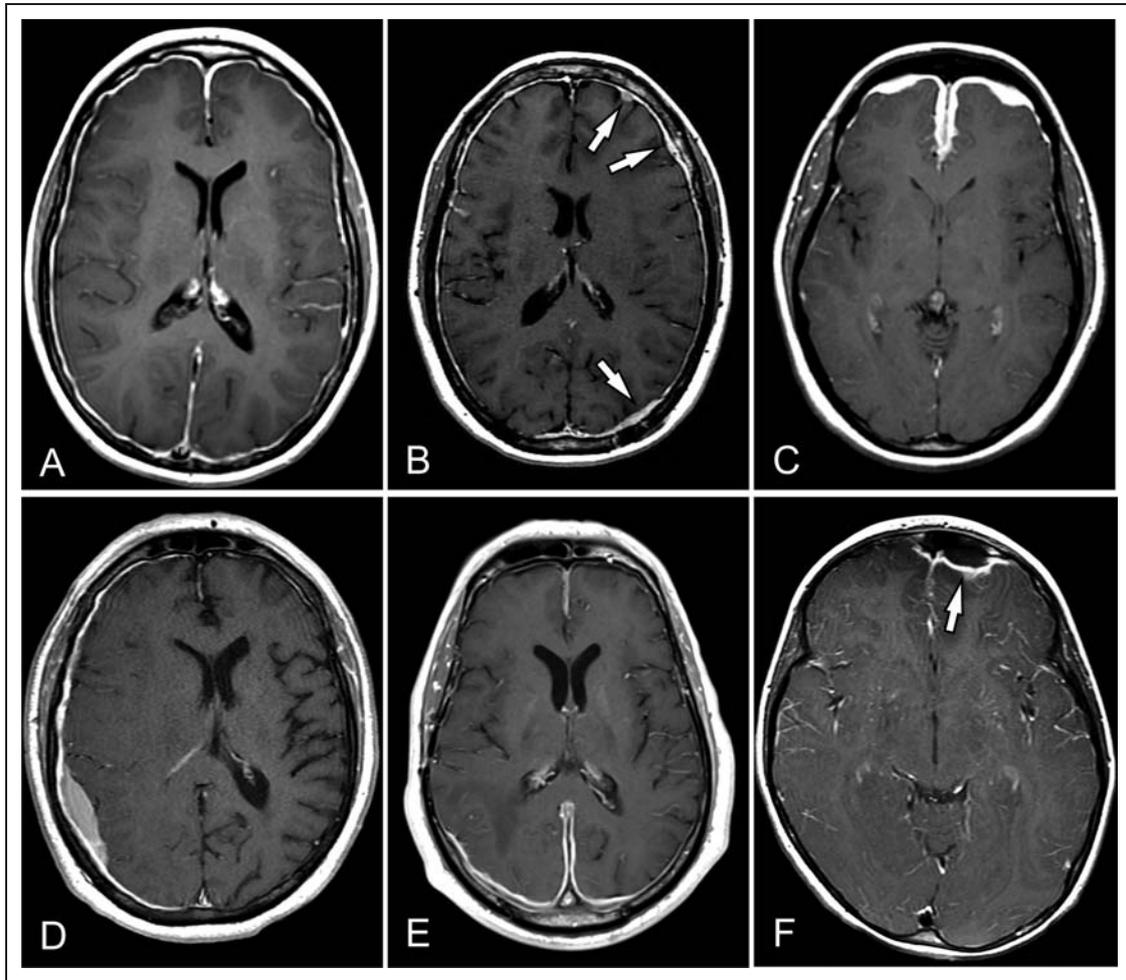


Fig. 2.—Axial T1-postcontrast images from several patients with abnormal dural enhancement. Only the patient with SIH (A) shows dural enhancement that is both diffuse and smooth. Patients with other causes of dural enhancement including dural metastatic disease (arrows) (B), granulomatosis with polyangiitis (C), subdural hematoma (D), idiopathic hypertrophic pachymeningitis (E), and empyema (arrow) due to acute frontal sinusitis (F), all show patterns of dural enhancement that are either nodular, plaque-like, or nondiffuse.

was not present previously likely indicates SIH. The practice of describing the tonsillar descent associated with SIH as “acquired Chiari” should be avoided, since it is potentially misleading. Chiari I malformations are treated differently than SIH; symptomatic Chiari I malformations are treated with suboccipital craniectomy to decompress the posterior fossa. Patients with brain sagging due to SIH should not undergo suboccipital craniectomy, as this will not stop spinal CSF leaks, which are the underlying cause of their headaches.

When cerebellar tonsillar ectopia is encountered on imaging, it is important to examine the third ventricular floor and mammillo-pontine

distance to assess for brain sagging. If these features are present, it indicates a spinal CSF leak. Chiari I malformation, however, is a congenital malformation that is not associated with the other signs of brain sagging. Recognition of this distinction is critical to correct diagnosis and appropriate therapy.

Myth 6: All Spinal CSF Leaks Are Caused by Tarlov Cysts/Spinal Diverticula.—One of the earliest recognized causes of spontaneous CSF leaks was spontaneous dural defects, often associated with fragile meningeal diverticula.^{25,32} These diverticula represent herniation of the leptomeningeal layer through dural tears or areas of dural

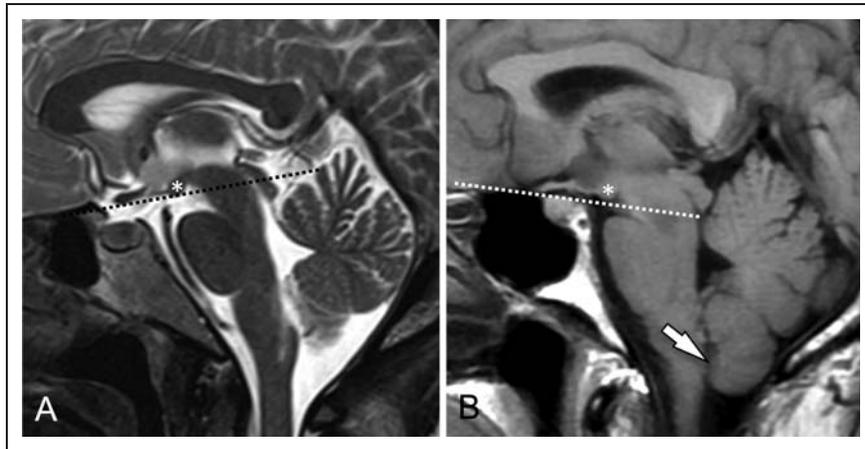


Fig. 3.—Imaging indicators of brain sagging in SIH. (A) Sagittal T2-weighted image in a normal patient shows a expected upsloping of the third ventricular floor (dashed black line) when a line is drawn between the optic chiasm and the mammillary bodies (*). (B) In a patient with SIH and brain sagging, there is a downward slope of the third ventricular floor (dashed white line), and narrowing of the distance between the mammillary bodies (*) and the pons. Note that the cerebellar tonsils (arrow) are low-lying in the patient with SIH; the presence of other signs of brain sagging distinguishes SIH from Chiari I malformation.

dehiscence, often along the lateral aspect of the thecal sac in close proximity to a spinal nerve root sleeve, producing an outpouching that is prone to rupture.³² However, there are also normal perispinal cystic structures that may mimic the appearance of these fragile diverticula on imaging. Perineural cysts, which occur along spinal nerve roots, are normal structures found most commonly along the lower cervical, and thoracic nerve roots.³³ Tarlov cysts, a histopathologically distinct entity, are common structures found incidentally in the lumbosacral spine and may also have a similar imaging

appearance.³⁴ As a result, there is often confusion about the diagnostic significance of perispinal cystic structures in patients with suspected or known SIH.

When actively leaking meningeal diverticula are identified, they are most commonly encountered in the thoracic spine or upper lumbar spine.^{32,33} Lower lumbar and sacral spontaneous leaks are distinctly uncommon, and sacral Tarlov cysts rarely, if ever, are the source of spontaneous CSF leakage. Cervical and thoracic perineurial cysts are common incidental findings in normal patients, and, in isolation, the presence of such cysts does

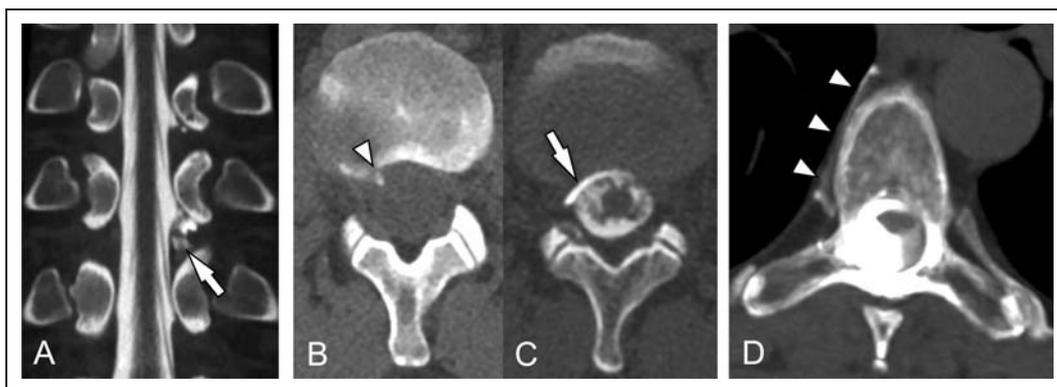


Fig. 4.—Various causes of spinal CSF leak in SIH illustrated with CTM. (A) Coronal CTM image shows contrast leakage (arrow) inferior to an irregular nerve root diverticulum. Axial CTM image from before (B) and after (C) intrathecal contrast injection shows a sharp osteophyte (arrowhead) causing a CSF leak (arrow). (D) Axial CTM image shows myelographic contrast filling a paraspinous vein (arrowheads), indicating the presence of a CSF-venous fistula.

not necessarily indicate or increase the likelihood of a diagnosis of SIH if no active leakage is identified on imaging.³³

It should be emphasized, however, that meningeal diverticula are only one cause of spontaneous CSF leak (Fig. 4). Ventral dural tears are a major source of spontaneous CSF leaks, and are often associated with calcified thoracic disk herniations or osteophytes.^{17,35} One series of over 500 patients identified ventral dural tears as the cause of SIH in approximately one-quarter of patients.³⁶

Another recently identified cause of SIH is the CSF-venous fistula.^{37,38} In this entity, a direct connection between the CSF and paraspinal veins allows for unregulated loss of spinal fluid into the circulatory system, resulting in CSF hypovolemia. Because the leak is not into the epidural space, they may be occult on spinal imaging, although specialized type of myelography may show the fistula.^{39,40} This leak type has been increasingly identified over the past few years.

Although more work is needed to investigate what anatomic characteristics differentiate leaking meningeal diverticula from normal perispinal cystic structures, one should avoid the temptation to assume that all such cysts are pathologic, especially if they are not found to be actively leaking on imaging. Furthermore, one should be aware of other causes of CSF leaks, such as calcified osteophytes and CSF-venous fistulas, since the identification of the source of CSF leak can help target treatment.

Myth 7: Spinal Imaging Rarely Reveals a Leak in SIH.—Spinal imaging is an important tool in evaluating suspected SIH. Demonstration of active CSF leakage both confirms the diagnosis of SIH and allows for directed treatment (targeted epidural patching or surgery), which is more effective than “blind” blood patching.^{41,42}

The rapidity of CSF leakage varies from patient to patient. Some patients may have “high-flow” leaks where CSF rapidly pours out of the thecal sac, while others may have slower “low-flow” leaks, and still others may have leaks that are not apparent on imaging.^{17,36} In one investigation of 99 subjects with confirmed SIH, a CT myelogram showed clear CSF leakage 55% of the time.¹² In another

series of 102 SIH subjects evaluated with spine MRI, a leak was detected in 26% of patients.⁴³ A large series of 568 patients who underwent imaging with either CT myelography (CTM) or spinal MRI reported finding a CSF leak in 51% of cases.³⁶ Although the precise yield of spinal imaging may vary by imaging modality, and depend on the population studied, it is clear that spinal imaging can contribute to the diagnosis of SIH in a substantial proportion of patients.

Despite this, there is a perception among some providers that spinal imaging has a very low yield in SIH. This incorrect perception may be due to several factors. First, some leak types are subtle, particularly low-flow leaks and CSF-venous fistulas, a newly recognized cause of SIH.^{37,38} Such leaks may not be apparent to those who are less familiar with the condition. Second, because not all orthostatic headache is due to SIH, some patients presumed to have SIH may be misdiagnosed, and will therefore not show evidence of spinal CSF leakage. Third, inadequate attention to imaging technique may decrease detection. Factors such as thin section imaging, proper patient positioning, and scanning immediately after intrathecal contrast injection have all been reported to promote leak identification on CTM.^{17,38,44,45} Finally, some leaks may be intermittent or may cease when the patient lies down to be scanned, and may be seen on one scan, but not another, despite similar scan technique.³⁸ Consequently, repeated spinal imaging may be useful in some cases when the leak is not initially visualized.

In summary, although the sensitivity of spinal imaging for detecting CSF leak in SIH patients is only moderate, a sizable proportion of patients will show evidence of a leak. Since leak detection has both diagnostic and therapeutic implications, spinal imaging should be performed in patients with suspected but unconfirmed SIH or SIH refractory to conservative therapy.

Myth 8: Skull Base CSF Leaks Cause Intracranial Hypotension.—Spontaneous CSF leaks occur in 2 major forms: spinal CSF leaks and skull base CSF leaks. Despite a superficial similarity in the underlying pathophysiology, these 2 types of CSF leaks typically manifest with separate clinical syndromes.

Skull base CSF leaks occur when there is dehiscence or fracture in the bone that separates an air-containing space of the skull from the intracranial compartment. This most commonly involves the cribriform plate in the anterior skull base, the roof of the middle ear or mastoid air cells (ie, tegmen tympani), or rarely, the otic capsule surrounding the inner ear (resulting in a perilymphatic fistula). CSF rhinorrhea or otorrhea is the result of these defects. Recurrent meningitis or headaches may be present, but headaches are not typically orthostatic. In a series of 273 patients with SIH, no causal association was found between skull base CSF leaks and intracranial hypotension.⁴⁶ Quite to the contrary, skull base CSF leaks have been found to be positively associated with idiopathic intracranial hypertension, not SIH.⁴⁷

The difference in symptoms between spinal and skull base CSF leaks can be explained by the normal physiology of CSF pressure in the standing position. When upright, gravity induces a gradient of CSF pressure that increases as one moves caudally down the spine. Under normal conditions, the pressure in the intracranial compartment is less than atmospheric pressure when upright, and the pressure in the lumbar spine is greater than atmospheric pressure.⁴⁸ The “zero-pressure point” is the spinal level where the CSF pressure transitions from negative to positive relative to the atmosphere, and is typically found in the upper cervical spine. Skull base defects occur above the level of the zero-pressure point, and thus do not leak CSF when the patient is upright. Rather, such leaks are typically aggravated by lying down or bending over with the head below the level of the waist.

Spinal CSF leaks occur below the zero-pressure point, are driven to leak most when the patient is upright, and thus cause orthostatic headache. Because spinal CSF pressure decreases in the recumbent position, lying down characteristically improves headache in spinal CSF leak. The brain imaging findings associated with intracranial hypotension are associated with spinal CSF leaks only.⁴⁶

As a result, fluid leaking from the nose should not be considered suspicious for SIH, and skull base imaging is not generally necessary in evaluation of patients with orthostatic headache.

Myth 9: Epidural Blood Patch Immediately Cures SIH.—Epidural blood patching, adapted from the treatment of postdural puncture CSF leaks, is considered the first line treatment for SIH.^{3,25,45} While epidural patching usually causes rapid clinical improvement in postdural puncture headache, our experience has been that improvement in symptoms may take longer in cases of spontaneous CSF leak. While immediate improvement certainly does occur in some SIH patients after epidural patching, the absence of instantaneous postprocedural improvement should not prematurely be interpreted as a negative prognostic sign.

One factor that may contribute to this difference in treatment response is the time interval between onset of CSF leak and treatment. Postdural puncture headache is often diagnosed quickly because it occurs immediately following a lumbar puncture. In contrast, SIH is often misdiagnosed initially, and patients often are symptomatic for longer periods before being treated.² During this delay, there are certain physiologic changes such as dilation of intracranial and paraspinal veins, and possibly changes in CSF production rates, that occur in order to attempt to compensate for the chronic loss of CSF. These venous changes can be observed on imaging.⁴⁹ Such changes may not instantly reverse even after successful closure of a dural defect, potentially contributing to the lag in improvement after epidural patching.

We have found that it is often a source of anxiety for patients if they are told to expect immediate improvement, but do not feel it right away. Instead, we counsel patients to expect an improvement in symptoms over the first week after their epidural patch.

Myth 10: The Job Is Done After the Epidural Patch.—Data on treatment outcomes in SIH are very limited, with most investigation employing a retrospective study design and small numbers of patients.³ Some early reports emphasized a high rate of immediate response with low recurrence rates, leading to a perception that SIH is easily treated with a single blood patch, with little to no after care required.

Subsequent experience with larger patient samples have shown much more varied results. Reported

success rates for an initial epidural blood patch in SIH range from 30 to 70%.^{41,50,51} Although often cited as highly effective in treating SIH, conservative therapy consisting of bed rest and hydration in reality offers much more uncertain benefits, and comes at a high social and economic cost to patients. One study found that among patients with SIH treated with conservative therapy, two-thirds were still symptomatic at 6 months, and one-third remained symptomatic at 2 years.⁵² For patients with debilitating symptoms, such a course can hardly be considered benign.

In practice, repeated blood patching is often required to obtain remission of symptoms. Furthermore, some patients may relapse after initial improvement, necessitating repeated treatment. In some cases, surgical treatment may be needed to obtain a permanent cure, particularly in cases of high-flow CSF leaks due to calcified osteophytes or disk herniations, and CSF-venous fistulas.^{22,38}

A frequent but under-reported complication of CSF leak closure is rebound intracranial hypertension (RIH).⁵³ In this phenomenon, CSF pressure increases above normal levels after a leak is sealed, leading to a new type of headache than can range in intensity from mild-severe. The most consistent clinical symptom exhibited in SIH is headaches change from orthostatic to those that are worse when recumbent or nonpositional, which allows discrimination between recurrent SIH and RIH.⁵⁴ Often the location of the headache changes from occipital to retro- or peri-orbital, and nausea and blurry vision may be present.⁵⁴ Papilledema is often absent, even in cases where CSF pressure are markedly elevated. Symptoms may be very brief in duration, but can persist in some patients for weeks to months. This phenomenon can be seen with both epidural patching and surgical repair of CSF leaks. Treatment is with acetazolamide or topiramate; if severe, therapeutic lumbar puncture of CSF diversion may be needed.³ In our experience, some degree of RIH is present in over half of patients undergoing epidural patching, with approximately half of symptomatic patients requiring treatment for one week or longer.

The absence of high quality, prospective data on outcomes and complications in SIH highlight

the need for greater research in this area, to select the most appropriate therapy and better define prognosis. Nevertheless, it is clear that continuing care is often needed beyond the initial epidural patch.

CONCLUSION

SIH is a condition that is identified with increasing frequency, but also one in which outdated or incomplete understandings of the disease can hamper appropriate management. A more nuanced understanding of the role of CSF pressure and an expansion of the understanding of underlying causes of CSF leak in SIH has emerged in recent years. Initial simplistic views of the underlying pathophysiology and clinical presentation of SIH have been shown to be incomplete by this new research, yet these conceptions have stubbornly persisted. Awareness of such myths and misperceptions about SIH will help direct more appropriate diagnostic and therapeutic approaches to the condition in the future.

STATEMENT OF AUTHORSHIP

Category 1

(a) Conception and Design

Peter G. Kranz

(b) Acquisition of Data

Peter G. Kranz

(c) Analysis and Interpretation of Data

Peter G. Kranz, Timothy J. Amrhein, Linda Gray

Category 2

(a) Drafting the Manuscript

Peter G. Kranz

(b) Revising It for Intellectual Content

Peter G. Kranz, Timothy J. Amrhein, Linda Gray

Category 3

(a) Final Approval of the Completed Manuscript

Peter G. Kranz, Timothy J. Amrhein, Linda Gray

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