

Skull Base CSF Leaks: Potential Underlying Pathophysiology and Evaluation of Brain MR Imaging Findings Associated with Spontaneous Intracranial Hypotension

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ABSTRACT

BACKGROUND AND PURPOSE: Cerebrospinal fluid (CSF) leaks of the skull base and spine share a common process of CSF volume loss, and yet only the latter has been associated with spontaneous intracranial hypotension (SIH). Despite published claims that only spinal leaks cause SIH, no prior studies have evaluated brain MR imaging in patients with skull base leaks for findings associated with SIH such as dural enhancement. The purpose of our study was to use a validated brain MR imaging scoring system to evaluate skull base CSF leak patients for findings associated with SIH.

MATERIALS AND METHODS: We included patients with confirmed skull base CSF leaks and contrast enhanced pre-operative brain MRI. The pre-operative MR images were reviewed for findings associated with SIH using the Bern score. Patient age, presenting symptoms and their duration, and leak site were also recorded.

RESULTS: 31 patients with skull base CSF leaks were included. Mean Bern score was 0.9 (range 0-4, SD 1.1), and only 1 patient (3%) had dural enhancement. Mean age was 53 years (range 18-76). Mean symptom duration was 1.3 years, with 22 patients presenting within one year of symptom onset. 23 patients (74.2%) had intraoperative confirmation of leak from the middle cranial fossa, involving the temporal bone, while 7 (22.6%) had leaks from the anterior skull base. One patient, who had dural enhancement, had an infratentorial CSF leak along the petrous segment of the internal carotid artery.

CONCLUSIONS: Our study provides further evidence that skull base and spinal CSF leaks represent distinct pathophysiologies and present with different brain MRI findings.

ABBREVIATIONS: SIH = Spontaneous Intracranial Hypotension; IIH = Idiopathic Intracranial Hypertension

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SUMMARY SECTION

PREVIOUS LITERATURE: Prior studies have described that spinal CSF leaks cause spontaneous intracranial hypotension (SIH), but that skull base CSF leaks do not. This is based on the absence of skull base CSF leaks in a cohort of patients with SIH, but without direct study of skull base CSF leaks.

KEY FINDINGS: We specifically studied patients with skull base CSF leaks and found that they do not have brain MR imaging findings that are associated with SIH.

KNOWLEDGE ADVANCEMENT: Our study adds to the body of literature advocating that SIH is caused by spinal CSF leaks but not skull base CSF leaks.

INTRODUCTION

Spontaneous intracranial hypotension (SIH), caused by a CSF leak, can present with debilitating symptoms including orthostatic headache, dizziness, and even dementia-range neuropsychiatric changes or coma.¹ As the name implies, SIH was initially thought to be due to low intracranial CSF pressure.² Since that time, however, other publications have shown that the majority of SIH patients have normal CSF pressure, and thus the underlying etiology has been attributed to low CSF volume.³ Per this mechanism, any site of CSF loss with subsequent low CSF volume might be expected to lead to the same disorder. Abnormal brain MR imaging findings can be explained on

the basis of the Monro-Kellie doctrine⁴, in that the loss of CSF is replaced by venous blood with resultant dural enhancement, venous engorgement, subdural fluid collections, and venous engorgement of the pituitary. Spontaneous skull base CSF leaks, frequently associated with intracranial hypertension and often present with CSF leakage through the nose or ear, should therefore have overlapping sequelae as spinal CSF leaks. The contrary has been described, predominantly based on one study by Schievink et al. that reviewed a collection of SIH patients, none of whom were found to have a skull base CSF leak.⁵ The dissociation of skull base leaks from the clinical symptoms and brain MRI findings of SIH has been propagated in the literature despite the absence of prior studies specifically evaluating skull base CSF leaks.^{6,7} The purpose of the current study was to evaluate skull base CSF leak patients for SIH clinical symptoms and brain MRI findings using a validated scoring system.

MATERIALS AND METHODS

We reviewed a historic database of patients with skull base CSF leaks seen at our referral center from 2009-2017. Individual patients were reviewed and included if they had a confirmed skull base CSF leak and pre-operative contrast enhanced brain MRI. Each brain MRI was reviewed for cerebellar tonsil descent below foramen magnum and Bern score components⁸ (Table 1) which include dural enhancement, small suprasellar cistern (≤ 4 mm), and venous engorgement (2 points each) as well as for a small prepontine distance (≤ 5 mm), small mamillopontine distance (≤ 6.5 mm), and subdural fluid collections (1 point each). Electronic medical records were reviewed for age, symptoms and their duration, location of CSF leak, history associated with CSF leak, and lumbar puncture with opening pressure. Descriptive statistical analysis including mean, standard deviation, and range were reported.

Table 1: Bern Score criteria and scoring system.

Brain MRI Findings	Points
Diffuse dural enhancement/thickening	2
Suprasellar cistern ≤ 4 mm	2
Engorged venous sinus	2
Subdural fluid collection	1
Prepontine distance ≤ 5 mm	1
Mamillopontine distance ≤ 6.5 mm	1

Probability of spinal CSF leak	Total Points
Low	≤ 2
Intermediate	3-4
High	≥ 5

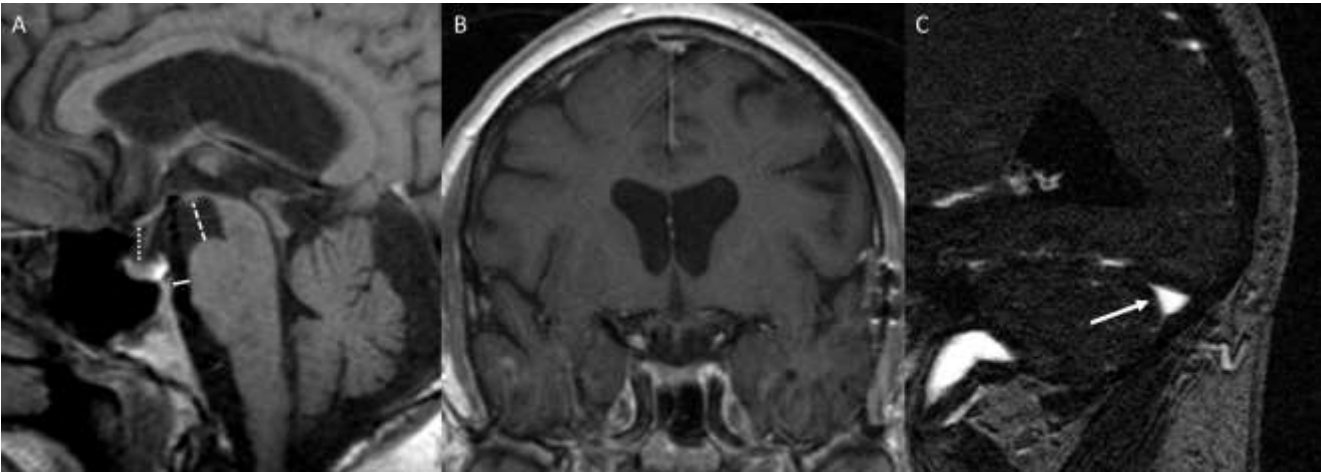


FIG 1. 57-year-old male with a surgically proven right tegmen tympani CSF leak without brain MRI findings associated with SIH (Bern score = 0). (A) Sagittal T1-weighted image shows normal suprasellar (dotted line), mamillopontine (broken line), and prepontine distances (solid line). (B) Contrast enhanced coronal T1-weighted image shows normal dura without thickening or enhancement. (C) Sagittal image from an MRV shows a non-distended dominant transverse sinus (arrow).

RESULTS

31 patients (mean age 53 years, range 18-76) with skull base CSF leaks and pre-operative contrast enhanced brain MRI were identified and included in this study (Fig 1). All cases had intraoperative confirmation of a skull base CSF leak. Eight patients (25.8%) presented with headaches: 1 was constantly orthostatic, 3 were intermittently orthostatic, 1 overlapped with diagnosed infectious meningitis, 1 was worse when supine, and 2 were not further characterized. Excluding the patient with meningitis, no patients reported photophobia, phonophobia, or cognitive issues. 18 patients presented with otorrhea, 8 with rhinorrhea, and 5 had neither otorrhea nor rhinorrhea but had brain MRI findings of a meningoencephalocele and an intraoperative dural defect. The mean symptom duration was 1.3 years, with 22

patients presenting within one year of symptom onset.

The majority of patients (23/31, 74.2%) had intraoperative CSF leaks found in the middle cranial fossa involving the temporal bone. Seven (22.6%) had an intraoperative CSF leak found in the anterior skull base. One patient (3.2%) had infratentorial CSF leaking along the petrous portion of the internal carotid artery and had orthostatic headache. For medical history relevant to the skull base CSF leaks, 3 patients (9.7%) had the diagnosis of idiopathic intracranial hypertension (IIH), 6 (19.4%) had a history of trauma, 9 (29.0%) had prior temporal bone surgery, and 13 (41.9%) were spontaneous.

The mean Bern score was 0.9 (range 0-4, std dev 1.1). Lower Bern scores are associated with lower likelihood of finding a spine CSF leak or CSF-venous fistula on subsequent myelography (score 0-2 low and 3-4 intermediate likelihood).^{8,9} Brain MRI findings are summarized in Table 2. Only two patients had a Bern score higher than 2, one patient with headache and an infratentorial leak (Bern 4, dural enhancement and venous distension), and one patient whose brain MRI findings (Bern 4, small suprasellar, prepontine, and mamillopontine intervals) without headache were unchanged one year after CSF leak repair.

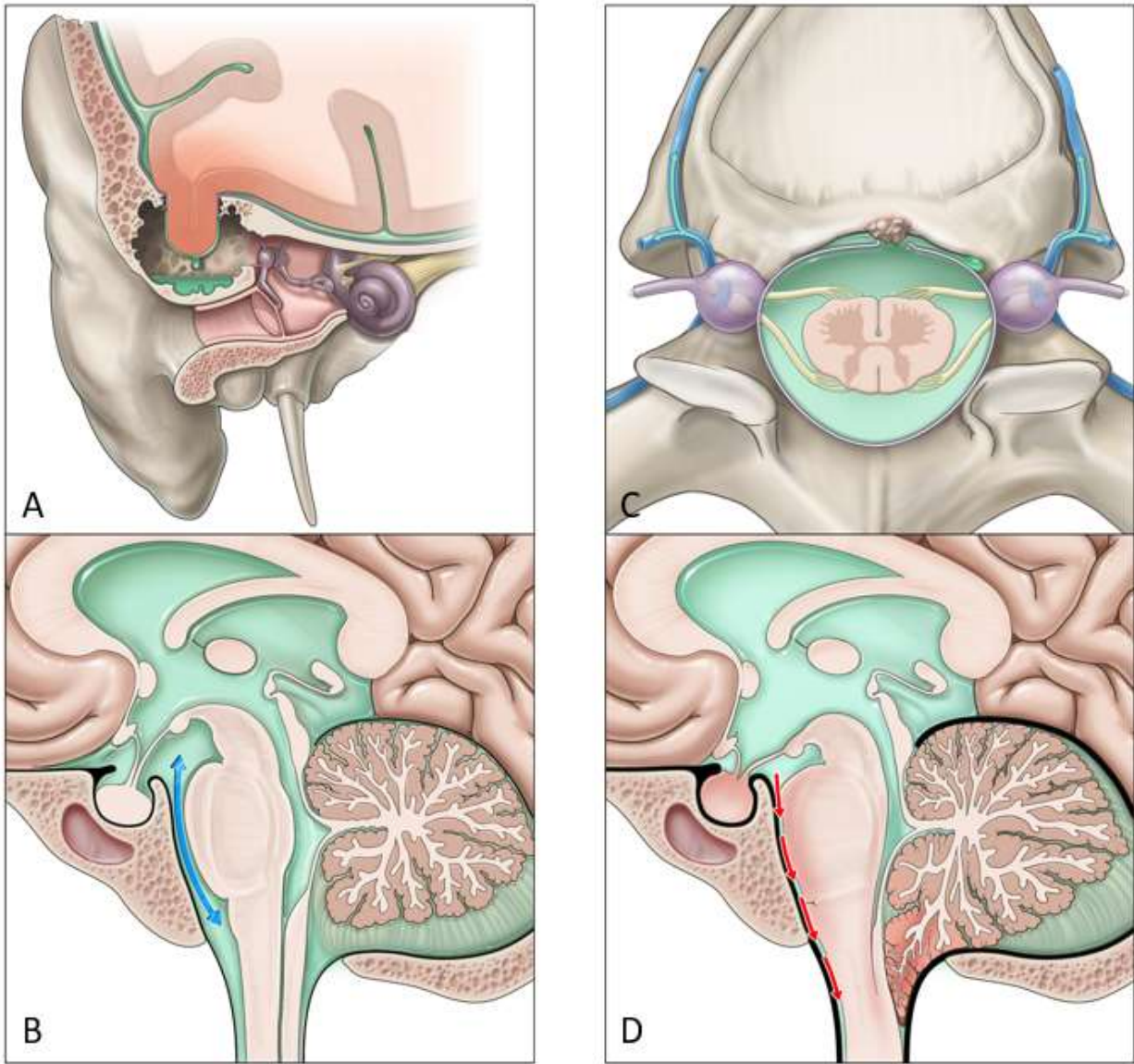


FIG 2. Illustration of a patient with a spontaneous supratentorial skull base CSF leak in the middle cranial fossa (A) with preserved CSF flow at the foramen magnum (B). This is in contrast to a patient with a spinal CSF leak (C) that could include a ventral dural tear or CSF-venous fistula, and potential sump effect with craniocaudal CSF flow at the foramen magnum (D) that contributes to findings associated with SIH including brain sag and pituitary engorgement shown here.

DISCUSSION

Our study used a validated brain MRI scoring system to assess for SIH in patients with skull base CSF leaks. Previously, skull base CSF leaks have been seen as distinct from SIH based on a single study that evaluated SIH patients and did not find any with skull base CSF leaks. Patients with skull base CSF leaks have not previously been studied for brain MRI findings of SIH. Of the 31 patients included in our study, the mean Bern score was less than 1, and only 1 patient had dural enhancement. Only 4 had headaches with orthostatic features. Our findings provide further evidence of the distinct underlying pathophysiology of skull base and spinal CSF leaks.

The initial work from Dobrocky et al.⁸ described a probabilistic brain MRI scoring method (0-9, 9 with the highest probability) to measure the likelihood of a spinal CSF leak, frequently referred to as the Bern score, and reported an 83% rate of dural thickening/enhancement in SIH patients. Subsequent studies have described a similar rate of dural enhancement in SIH of 79%¹⁰ and 81%¹¹. Our study population largely did not demonstrate findings of SIH, with a mean Bern score of less than 1 and only 1 case (3%) with dural enhancement. Kranz et al. found that increasing symptom duration is associated with decreased rate of dural enhancement and that SIH patients without dural enhancement had a mean symptom duration of 45.3 weeks.¹² In our study, 22 patients had symptom duration of 1 year or less (1 had dural enhancement), including 10 with symptom duration of 6 months or less (none had dural enhancement).

The major prior study that separated skull base CSF leaks from spinal CSF leaks causing SIH was performed by Schievink et al., who reviewed 273 SIH patients from a single institution and did not find any cases with a skull base CSF leak.⁵ Schievink later reported a single case of SIH caused by a skull base leak from the posterior fossa.¹³ Tai et al. reported on the treatment and outcomes in patients with spontaneous versus nonspontaneous CSF leaks, and included 6 patients with spontaneous sphenoid sinus leaks.¹⁴ They stated that all 6 patients had MRI findings of intracranial hypotension but did not use a validated scoring system for MR imaging results, nor did they list what imaging findings were abnormal. Their methodology included focal dural enhancement as a sign of SIH, which in a different study has been described as an SIH mimicker¹⁵ and is likely distinct from the diffuse dural enhancement that is widely associated with SIH. Our study evaluated the MR imaging findings in skull base CSF leaks using a validated scoring system and did not find brain MR imaging results associated with SIH.

Spinal and skull base CSF leaks both result in CSF loss, but they are differentiated by the leak location. Why would the location of the CSF leak cause such different symptoms and brain imaging findings? It is likely that brain MRI findings in SIH are related to downward CSF shift in the cranial to caudal direction from a sump effect of the spinal CSF leak. Wolf et al.¹⁶ recently used phase contrast MRI in SIH patients (spinal leaks) to show that CSF flow was increased at C2-C3 compared to normal controls. They also found that SIH patients had larger craniocaudal spinal cord motion at C2-C3. Comparatively, in our study, skull base CSF leaks would theoretically have preserved physiologic CSF flow at the skull base, without resultant downward sump effect that would cause brain sag or venous engorgement (Fig 2).

Most interesting, the one skull base CSF leak patient in our cohort with dural enhancement had an infratentorial CSF leak. Prior to surgical repair of the skull base CSF leak, the patient underwent a conventional CT myelogram that did not show extra-dural fluid. Three years later, the patient was found to have a ventral spinal CSF leak from a calcified disc osteophyte complex. It is unknown whether the patient's symptoms were due to an occult spinal CSF leak, or if this occurred after the skull base leak repair. All other patients in this study without dural enhancement had supratentorial CSF leaks, suggesting that supra- versus infratentorial leak location could contribute to the brain MRI findings. One prior case report described a cranial CSF leak presenting with orthostatic headache and brain MRI findings that included diffuse dural enhancement, and this patient also had a posterior fossa CSF leak into the mastoid air cells.¹⁷ Bonomo et al.¹⁸ reported a single case of a small sphenoid-ethmoidal meningoencephalocele causing SIH symptoms and brain MRI findings, but no other studies have described similar findings. Therefore, the notion that SIH is caused by a spinal CSF leak might possibly be extended to leaks from the infratentorial skull base, however the current data is insufficient to fully support this claim.

Our study has limitations, starting with the retrospective nature and small sample size. Spontaneous skull base leaks are rare, but to our knowledge, we present the largest study focusing on brain MRI findings. Our theory, building on what Wolf et al.¹⁶ reported in SIH patients, requires testing. Further studies evaluating upper cervical CSF flow and cord motion in skull base CSF leaks could help to validate our hypothesis. Nonetheless, it has been suggested that skull base CSF leaks do not present with SIH, and our study largely supports this claim. One of the two patients with an intermediate Bern score of 4 in our study had brain sag and low-lying cerebellar tonsils that were unchanged 1 year after surgical repair of the CSF leak. The lack of improvement of the brain MRI findings, if the causative leak site was repaired, would be unexpected based on prior work by Dobrocky et al. studying post-operative brain MRI findings in patients with surgically repaired spinal CSF leaks¹⁹. It is possible that our patient's findings represented a Chiari 1 malformation rather than sequela of a CSF leak. Additionally, outside of the infratentorial CSF leak, none of the patients with headache in our study were evaluated for a spinal CSF leak, and therefore a spinal cause was not excluded.

Table 2: Brain MRI findings in patients with skull base CSF leaks.

MRI Findings	# patients (%) n=31
Dural enhancement	1 (3%)
Small suprasellar distance (≤ 4 mm)	2 (7%)
Engorged venous sinus	1 (3%)
Subdural fluid collection	0
Small prepontine distance (≤ 5 mm)	10 (32%)
Small mamillopontine distance (≤ 6.5 mm)	9 (29%)
Mean Bern score (SD)	0.9 (1.1)
Low lying cerebellar tonsils	2 (7%)

CONCLUSIONS

Prior studies have suggested that skull base CSF leaks, as opposed to spinal CSF leaks, do not present with SIH. This idea was largely based on observations of spinal CSF leaks only. We approached the question from the perspective of skull base CSF leaks and found further evidence for the same conclusion. Skull base leaks and spinal CSF leaks may indeed represent distinct disorders of CSF dynamics.

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