Intracranial hypotension secondary to spinal CSF leak: diagnosis

Spinal cerebrospinal fluid (CSF) leak is an important and underdiagnosed cause of new onset headache that is treatable. Cerebrospinal fluid (CSF) bathes and supports the brain and spinal cord. When the connective tissue known as dura that holds CSF in around the spinal cord has a hole or tear, the result is a loss of CSF volume, known as **intracranial hypotension**.

Patient history

The most important clues to the diagnosis of intracranial hypotension lie in the patient history.

Symptoms – The hallmark symptom is a headache that is worse upright and improved with horizontal positioning. It is important to note that not all headaches related to spinal CSF leaks are positional and not every patient with a positional headache has a spinal CSF leak. Common and uncommon associated symptoms are listed on our **Symptoms** page.

History of medical procedures or trauma – Lumbar punctures, epidural injections and spinal surgery occurring prior to onset of symptoms may be responsible for a spinal CSF leak. Significant trauma may also result in a spinal CSF leak. See our page on **Causes**.

History of Heritable Disorders of Connective Tissue (HDCT) – Spinal CSF leaks that develop spontaneously or with minimal precipitant may be related to an underlying weakness of the spinal dura. Marfan Syndrome, Ehlers-Danlos Syndrome, other HDCTs, as well as Polycystic Kidney Disease have been associated with spontaneous leaks. In many cases, the symptoms related to intracranial hypotension are the first recognized evidence of a HDCT.

Known spinal problems – Spontaneous leaks are also associated with calcified discs or bone spurs that may or may not be known prior to the onset of symptoms.

Previous diagnosis of POTS (Postural Orthostatic Tachycardia Syndrome) – The intolerance of upright posture associated with POTS can mimic intracranial hypotension, but the two may also coexist. Patients with the hypermobile type of Ehlers-Danlos Syndrome are known to have a higher prevalence of POTS as well as a greater risk of spontaneous intracranial hypotension.

Previous diagnosis of Chiari - Low-lying cerebellar tonsils on cranial imaging can be the result of intracranial hypotension (and reversible) and may not be recognized as such. To complicate matters, congenital Chiari is also more common in patients with some types of HDCT.

Physical signs

Physical findings may be entirely absent but physicians may note that their patient prefers to be lying flat. Additional findings and presentations may include:

Evidence of HDCT – There are a number of physical signs of HDCT that may be noted, such as joint hypermobility, high arched palate, dental crowding, tall stature, scoliosis, easy bruising, atrophic scars and a number of others.

Neurologic Findings – There are many findings including involvement of cranial nerves, cognitive changes, gait disturbance, sensory and motor changes, but the exam may be entirely normal.

Clinical mimics – There are a number of disorders that intracranial hypotension may mimic due to the wide range of neurologic presentations. Patients may present with dementia, ataxic gait, Parkinsonism and other movement disorders.

Serious neurologic presentations – Emergency Medicine physicians may see patients presenting clinically with quadriplegia, stupor, coma or other serious neurologic presentations. Death is a reported complication, albeit rare.

Diagnostic testing includes lumbar puncture (not required), cranial (brain) imaging, spinal imaging and occasionally a nuclear medicine study called a radioisotope cisternogram.

Lumbar Puncture (LP) may be done to measure the opening pressure and to collect CSF for analysis but this is not required to make the diagnosis. An opening pressure is often measured at the time of LP for myelography. Normal opening pressure has a reference range of 65 to 195 mm of water. The opening pressure is often <60, and can be unmeasurable or subatmospheric. Normal pressure is not uncommon, however, and does not rule out CSF leaking. High pressures also occur.

Cranial imaging

Cranial MRI should be done in ALL cases where a spinal CSF leak is suspected. This should be done without and with gadolinium contrast enhancement. There are 5 findings, remembered by the mnemonic **SEEPS**, however the absence of these findings does not rule out CSF leaking. About 20% of patients have a normal cranial MRI. The lower prevalence of dural enhancement associated with longer duration of symptoms can contribute to underdiagnosis.

Subdural fluid collections
Enhancement of pachymeninges
Engorgement of venous structures
Pituitary hyperemia
Sagging of the brain

Cranial CT is not infrequently ordered when a patient presents to the emergency department with acute onset of severe headache. Subdural fluid collections or subdural hematomas may be seen. MRI is, however, the preferred imaging of the brain for diagnosis.

Spinal imaging

A variety of spinal imaging techniques are used to localize leaks. It should be emphasized that spinal imaging may not be necessary since many patients respond well to lumbar epidural blood patching. MRI without intrathecal contrast and CT myelography are used most commonly. Digital subtraction myelography is used increasingly. It should be noted that extradural CSF at C1-2 and the cervicothoracic junction are common false localizing signs. Spinal imaging continues to be refined but remains insufficiently sensitive to image about half of suspected spinal CSF leaks. More than one type of spinal imaging and/or repeat imaging is often necessary.

Spinal MRI is MR imaging of the spine without the use of intrathecal contrast, is non-invasive and is often the first type of spinal imaging used.

MR myelogram usually refers to MR imaging of spine with the use of specific sequencing/weighting but without intrathecal contrast.

Intrathecal gadolinium enhanced spinal MRI is a spinal MRI that is done following injection of gadolinium into the intrathecal space via lumbar puncture.

CT myelogram involves the injection of radiographic contrast into the intrathecal space via lumbar puncture, followed by a CT of the spine. Several variables can be manipulated, depending on the circumstances, such as timing of the imaging after the injection of contrast (**dynamic**, early, delayed), the intrathecal injection of normal saline to raise the intrathecal pressure, or the use of **digital subtraction imaging**.

Dynamic CT myelogram involves the performance of the CT as the contrast is being injected. This is most commonly used to localize rapid or high-flow spinal CSF leaks.

Digital subtraction myelogram (DSM) is a myelogram (injection of intrathecal contrast + imaging) done under fluoroscopy with the additional ability to digitally subtract a pre-contrast image to enhance the visualization of the contrast. This is a dynamic form of imaging. This can be done under general anesthesia to stop breathing for a short time, thereby preventing movement artifact on the imaging. A CT can be done soon after or with a delay. This is used to image rapid leaks, ventral (front of spinal cord) leaks or leaks not associated with an obvious extra-thecal CSF collection, such as a CSF-venous fistula.

Radioisotope cisternogram is a nuclear medicine study that is occasionally used. This can help to confirm the presence of CSF leaking, but is insensitive in localizing leaks.

Most radiology departments can perform these procedures, however, in cases where a leak cannot be localized, neuroradiology departments that see larger volumes of spinal CSF leak patients may offer the additional expertise with interpretation as well as with optimization of imaging sensitivity. The understanding of anatomic types of leaks has resulted in refined imaging strategies to characterize a leak in a specific patient to guide treatment. See the **Classification of Spontaneous Spinal CSF Leaks.**

Diagnostic criteria for spontaneous intracranial hypotension

Proposed diagnostic criteria for spontaneous spinal CSF leaks
(Headache 2011;51:14421444)

A. orthostatic headache;
B. the presence of at least one of the following:
   - low opening pressure (≤60 mm H2O),
   - sustained improvement of symptoms after epidural blood patching,
   - demonstration of an active spinal cerebrospinal fluid leak,
   - cranial magnetic resonance imaging changes of intracranial hypotension (e.g., brain sagging or pachymeningeal enhancement);

C. no recent history of dural puncture; and
D. not attributable to another disorder.

ICHD-3-beta, the International Criteria of Headache Disorders, 3rd edition, beta version, 2013

7.2.3 Headache attributed to spontaneous intracranial hypotension

Description:
Orthostatic headache caused by low cerebrospinal fluid (CSF) pressure of spontaneous origin. It is usually accompanied by neck stiffness and subjective hearing symptoms. It remits after normalization of CSF pressure.

Diagnostic criteria:
A. any headache fulfilling criterion C
B. low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
C. headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
D. not better accounted for by another ICHD-3 diagnosis.
(full comments available online at ichd3.org)

Diagnostic difficulties

Misdiagnosis and delayed diagnosis of spontaneous spinal CSF leak remain common, largely due to low familiarity by health care professionals, as we see with all uncommonly recognized disorders. There is considerable variability in clinical presentations and many diagnostic challenges. Severity of symptoms and degree of disability are often underappreciated. Not surprisingly, many patients suffer for months or years before a correct diagnosis is made.

Diagnoses often considered

- primary headache disorders such as migraine headache, tension headache
- sinus headache
- cervicogenic headache (arising from neck problems)
- meningitis
- occipital neuralgia, trigeminal neuralgia
- psychogenic disorder (arising from psychological issues), depressive disorder, anxiety disorder

Differential or concurrent diagnoses

- POTS (postural orthostatic tachycardia syndrome) - may be an alternate or concurrent diagnosis
- Chiari malformation (congenital, with abnormal posterior fossa) - low-lying cerebellar tonsils in intracranial hypotension (reversible with treatment) may be mistaken as congenital Chiari. Intracranial hypotension may also worsen cerebellar tonsillar descent in patients with congenital Chiari
- patients with Ehlers-Danlos Syndrome often have multiple comorbidities
- in some cases, intracranial hypotension may lead to a diagnosis of a Heritable Disorder of Connective Tissue not previously recognized; screening for vascular complications such as thoracic aortic aneurysm or intracranial aneurysm should be considered on a case by cases basis

Clinical presentations that may be unrecognized as possibly secondary to intracranial hypotension

- subdural hematoma
- dementia
- Parkinsonism or other movement disorders
- stupor or coma
- stroke
- reversible cerebral vasoconstriction
- posterior reversible encephalopathy syndrome
- spinal manifestations (radiculopathy, myelopathy, syringomyelia, quadriplegia, bibrachial amyotrophy) – large extradural fluid collections usually evident

Additional diagnostic challenges

- not every patient with intracranial hypotension has headache; there are many clinical presentations
- not every headache related to a spinal CSF leak is positional
- not every patient with a positional headache has a spinal CSF leak
- lumbar puncture is not required to make the diagnosis
- normal or high CSF pressures do occur and do not rule out the diagnosis
- interpretation of imaging requires experience / training
- normal cranial MRI does not rule out the diagnosis
- extradural CSF at C1-2 and the cervicothoracic junction are common false localizing signs
- sensitivity of spinal imaging is inadequate to localize a significant percentage of leaks, even with the most refined techniques, which limits treatment options
- more than one type of spinal imaging and/or repeat imaging is often necessary

Improving the diagnostic process

Education tailored to both health care professionals and a general audience is helping to shorten diagnostic delays and access to appropriate diagnostic testing along with accurate interpretation, which impacts treatment and outcomes.

Research funding to accelerate the refinement of diagnostic imaging is expected to improve sensitivity in locating elusive leaks for targeted treatment.