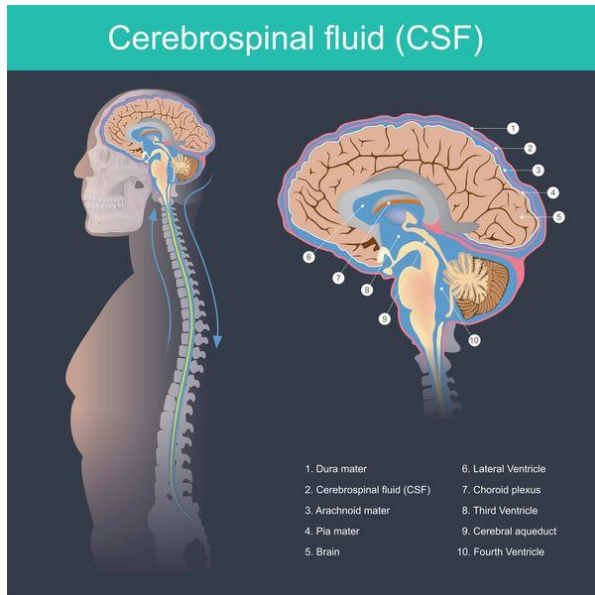


# About Spinal CSF Leaks



## A Primer

Cerebrospinal fluid (CSF) surrounds the brain and spinal cord, providing protection, nourishment, and waste removal. It continually circulates in pulsing waves, transferring nutrients, conducting signals, removing toxins, providing immunity, and keeping the brain buoyant. **CSF acts as a shock absorber**, reducing the effective weight of the brain and providing cushion to protect against gravity and to protect brain tissue, cerebral vessels, and spinal cord against

sudden movement.

The brain and spinal cord make up the central nervous system. The **CNS controls everything from thought, movement, and emotion to breathing, heart rate, body temperature, and hormone regulation.**

*CSF is vital to normal function.*

The skull contains a closed system of brain, blood, and cerebrospinal fluid. The skull is bone and therefore cannot expand, so to maintain intracranial pressure (ICP) and protect CNS function, a leak may open to act as a relief valve when pressure becomes too high.

**When something goes wrong (for instance, illness, epidural, or injury) the dura may become unable to seal naturally.** A CSF leak lowers the level of fluid in the skull, particularly when a person is upright (imagine a partially filled water bottle upright versus on its side). When the cause of the leak is not clear, it is referred to as a spontaneous leak or spontaneous intracranial hypotension (SIH). Iatrogenic spinal CSF leaks are caused by a medical procedure and traumatic spinal CSF leaks are caused by injury.

***Spinal CSF leaks are not rare but remain estimated to be misdiagnosed at a rate of more than 90%. Misinformation, misdiagnosis, and delayed diagnosis are common.***

To make matters worse, the diagnostics themselves may be flawed. One study found 94% of patients would have been misdiagnosed if using opening pressure alone as leak indicator. And the absence of MRI findings does not rule out a CSF leak. Some patients may even cycle between high and low pressure.

Intracranial hypotension (low intracranial pressure and leaks) and intracranial hypertension (high intracranial pressure) each have a variety of possible symptoms, many of which overlap. What's more, connective tissue may become damaged, creating its own complications including blood flow changes in the brain. **Postural orthostatic tachycardia syndrome (POTS) can present like intracranial hypotension** or patients may have both. Hypermobility type Ehlers-Danlos Syndrome (hEDS) patients have a higher prevalence of both POTS and CSF leaks, but patients with connective tissue disorders are only a portion of those affected by leaks.

*CSF leaks can happen to anyone.*

**Studies have found high rates of intracranial pressure issues in many illnesses including chronic fatigue syndrome (ME/CFS), fibromyalgia (FM), and Long Covid.** It may be the cause of an illness or contribute to the symptoms, and proper management may improve a patient's quality of life.

Loss of CSF fluid can cause the brain to sag, compromising the brain stem, affecting nerves, and complicating many bodily functions, and high intracranial pressure can cause issues of its own. Regardless of an initial onset that is post-illness or mechanical onset, pressure issues can create a variety of symptoms and complicate diagnosis.

*(Dive further into cerebrospinal fluid mechanics here or check out the research page for more.)*

## Symptoms

Leak symptoms may vary in number and severity. It's important to note that **not every patient has a headache**, not every patient experiences the positional aspect (especially those who have been leaking long-term), and normal opening pressure or MRI does not rule out a leak. Also take into account that other diagnoses may be secondary to a leak (some patients present with stroke, dementia, POTS, or movement disorders).

**In general, CSF leak symptoms worsen when sitting or standing and improve or resolve when lying down.** Having a variety of symptoms may also be significant. When the cause is a known injury (traumatic) or medical procedure (iatrogenic) it is not referred to as spontaneous intracranial hypertension and should be addressed at the time of injury.

**A recent diagnostic guideline** notes spontaneous intracranial hypotension should be considered in any patient presenting with **orthostatic headache**; 'end of the day' or '**second half of the day**' headache with improvement of the headache on lying flat; **thunderclap headache** which is followed by orthostatic headache; and **new daily persistent headache** with an initial orthostatic quality. The presence of associated symptoms below should increase the suspicion of SIH.

**The guideline lists common associated symptoms as:**

- Dizziness or vertigo (50.5%)
- Nausea and vomiting (49.0%)
- Disequilibrium (42.6%) *unsteadiness, imbalance, or loss of equilibrium often accompanied by spatial disorientation*
- Muffled hearing or aural fullness (37.1%)
- Posterior neck pain (34.2%)
- Cognitive impairment† (31.7%) *most commonly non-specific problems with concentration and word finding*
- Tinnitus (27.7%) *ringing ears*
- Hypoacusis (26.2%) *hearing acuity issues*
- Fatigue (24.3%)
- Photophobia or phonophobia (20.3%) *sensitivity to light or sound*
- Visual blurring (17.8%)
- Facial numbness, paraesthesia (*tingling or pricking*) or pressure (15.8%)

## The guideline lists less associated symptoms as:

- Interscapular pain (10.9%) *pain between the shoulder blades*
- Dysgeusia (7.4%) *taste disorder*
- Hyperacusis (5.9%) *sound intolerance*
- Behavioral variant frontotemporal dementia syndrome (2.5%)
- Reverse orthostatic headache (2%) *headache improves in upright position*
- Bibrachial amyotrophy (1.5%) *upper limb motor disorder*
- Superficial siderosis (1.5%) *hearing loss, movement abnormalities (ataxia), and motor difficulties*
- Cerebral venous thrombosis (1%) *venous sinus blood clot*
- Abducens nerve palsy (1%) *ocular motor paralysis*
- Spinal cord herniation (1%)
- Coma (0.5%)
- Syringomyelia (0.5%) *fluid-filled cyst within the spinal cord*
- Hemifacial spasm (0.5%) *spasms of the muscles on one side of the face*

Headache may present as pressure or nonstandard head or neck pain. **Some leakers have also reported POTS-like symptoms** of heat intolerance, tachycardia, and shortness of breath, as well as exercise intolerance, post-exertional malaise, weakness or heaviness in limbs, nerve pain in jaw, neck, or back, disturbed or unrefreshing sleep, watery/runny nose, irritated throat, scent sensitivity, gut issues, low appetite, or food sensitivities that may be related to leaking. Some severe leakers have also reported experiencing a low tolerance for movement such as car rides, muscle weakness, inability to walk or grip, stinging pinprick pain, strange electrical jolts, handwriting/fine motor control issues, skin changes, issues communicating or slurred speech, numbness or tingling in face, hands, legs, or feet, muscle spasms, dry cough, gag reflex or swallowing issues, burning pain at top of head; tender, bruised-feeling scalp, temperature issues such as chills or low fever, mood changes including anxiety, depression, anger, impending doom, and more, or gait issues, and seizure.

Many of these symptoms can be misleading when searching for a diagnosis or dismissed due to outdated information regarding the diagnosis of and symptoms related to intracranial pressure.

Individual symptoms can vary and may change over the course of one's illness.

***It is important to note the orthostatic component may become less apparent over time. Consider whether position had any effect at illness onset.***

## Things to Consider When Seeking Diagnosis

CSF pressure issues can be aggravated by flow problems such as stenosis, bone spurs, cysts, or many things mechanical, and made worse by connective tissue disorders (like hEDS) or medications (like antibiotics or steroids). While **the most common cause of spinal CSF leaks are medical procedures such as lumbar puncture, epidural, surgery, or chiropractic manipulation** (and may be made severe when not treated in a timely manner), **leaks can have many causes.**

Some causes may include:

- Epidural (often during childbirth)
- Lumbar puncture or spinal tap
- Accident or injury involving the head or spine
- Surgery
- Bone spur
- Venous fistula
- Spinal manipulation (such as chiropractor)
- Weakened dura due to illness or CTD

It should be noted that **even minor events** such as sneezing or coughing, straining with bowel movements or by heavy lifting, jumps or falls, stretching/yoga, or similar activities can lead to leaking.

Much of the difficulty getting treatment is due to the medical community being informed by outdated material and many assume (incorrectly) that an immediate headache with epidural is the chief or only cause and diagnosable symptom, and that it will always heal on its own with time. (Of note: the Spinal CSF Leak Foundation remarks *the use of pencil-point LP needles reduces the risk of post-dural puncture headache.*)

A few of the more common misdiagnoses are headache disorders or migraine,

sinus or allergy issues, occipital neuralgia or trigeminal neuralgia, post-traumatic headache (post-concussion or whiplash), POTS, depression, and anxiety.

Within the CSF leak patient groups, cases may range from highly functioning to severe. Physicians unfamiliar with the true scope of leak symptoms may be at a loss, risking unnecessary or harmful medications and surgeries. Brain sag has been mistaken for Chiari in some cases, many MRIs return normal, opening pressures on lumbar punctures may appear normal, and even neurologists and neurosurgeons may not take the suggestion of a leak seriously because they're considered rare (they are not—only underreported). ***It is essential to find a specialist experienced in leaks and armed with the most current information.***

Read the [Multidisciplinary consensus guideline for the diagnosis and management of spontaneous intracranial hypotension](#) for more information and check the [Spinal CSF Leak Foundation Physician Directory](#) for doctors using the best practices for leak diagnosis and repair.

## Important Clues

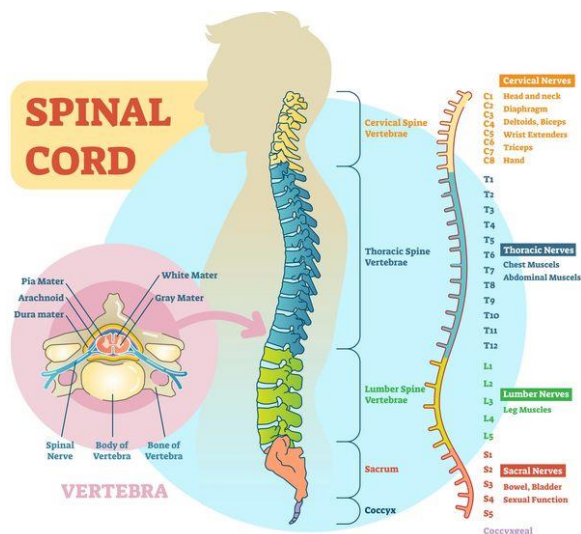
**Pressure may be the cause of your illness, not just a symptom.** It may be difficult to discern whether your symptoms are positional or pressure related. Below are a few questions to help pinpoint when and where symptoms occur.

- *Do you feel worse—either immediately or within days –after strenuous activity such as lifting, running, high-stress events, sneezing/coughing, strenuous bowel movements, coitus, or bending or twisting? A worsening of symptoms after straining may be an indicator of a leak or high pressure.*
- *Have you had an unusual (good or bad) reaction to doxycycline, prednisone, or any medicine that affects cranial pressure? Does caffeine, salt/processed foods, Vitamin A, or B1 affect the severity of your symptoms? (Google the medicine or food with "cranial pressure" to find connections.)*
- *Do you feel worse in the evenings or have vision or head or neck pain changes as the day progresses? Do you feel worse at different elevations or with barometric pressure swings?*
- *Do you find your mood shifts with your symptoms after these activities or events? Are your cognitive or neurological symptoms brought on by bending, lifting, twisting, jumping, straining, or Valsalva maneuver? Is some part of your pain positional?*

- Do you have scoliosis, bruise easily, have unusual scarring, stretchy skin, or have joint hypermobility in your family? These may be signs of a connective tissue disorder, making leaks and pressure issues more likely.
- Do you ever feel anxious or restless when lying down? This may be high pressure urging you to get upright to ease pressure in the skull. Do you ever find relief by lying flat? This may be low pressure returning needed fluid to your brain.

Consider how these and other positional factors affect your symptoms. Don't let the incorrect notion that leaks are rare dissuade you from finding treatment--if you fit the profile and your physician dismisses the possibility, consider a second opinion from a doctor experienced in spinal CSF leaks and pressure.

**If you believe you have an orthostatic component to your illness, if you have any relief staying still and flat (or find you are unable to stay still and flat), look into CSF pressure issues.**



## Treatment

CSF is held in by layers of connective tissue. The outer layer is called the dura mater (from the Latin for tough mother). A hole in the dura can allow cerebrospinal fluid to escape (called a spinal CSF leak) and the low CSF level causes the brain to be without its protective fluid (called intracranial hypotension).

To determine if a spinal CSF leak is present, a specialist (often an interventional radiologist) will look at a patient's symptoms and medical history, and if a leak is suspected, may order tests such as a head MRI with and without contrast. Note that the **initial testing may appear normal**. An experienced leak specialist will know when to proceed with further treatment or testing.

Spinal MRI, CT myelogram, or digital subtraction myelogram may be used to locate spinal leaks, tears, or other issues. Equipment may vary depending on the medical facility, and there are not currently a great number of leak specialists available

worldwide. (See the physician's directory link at the bottom of this page for a list of doctors using best practices and with knowledge of CSF leaks and [learn more about the imaging types here.](#))

If imaging fails to identify the precise location of the leak, in some cases non-targeted "blind" treatments may be used. Remember that every case is different, and while many leakers only require a quick blood patch to be repaired, some may have a more complicated condition due to bone spurs, cysts, stenosis, or other issues that need resolved for a patch to do its work.

**Spinal CSF leaks are repaired by epidural blood patch** and can vary in post-procedure care. In cases where an epidural anesthesia injection has gone wrong and the attending physician is aware that a puncture was created (and, key here, it's a small and fresh puncture), conservative treatment may be an option, probably consisting of bed rest, water, and caffeine. During childbirth an anesthesiologist may unintentionally puncture the dura, and if the patient has an upright headache, they may be given a quick blood patch with little to no restrictions or follow up.

But in cases where the puncture was not recognized, the leak symptoms can continue or worsen over time. Leaks can have many causes in addition to epidural, spinal tap, or surgery, and in the case of long-term or severe leakers, the dura has not healed on its own and may need additional support.

Epidural blood patches use the patient's own blood to "seal" the puncture. Instead of going through the dura, blood will be injected into the epidural space just outside/over the dura to form a patch, often by an interventional radiologist using fluoroscopic guidance. Some patients may feel immediate improvement due to the blood preventing further CSF escape (in turn raising pressure). It is thought that the additional pressure of the blood can ease symptoms temporarily, then the blood encourages wound healing at the leak site. Over time, the blood that makes up the patch will dissolve and symptoms may fluctuate, but if successful, the puncture will scab over and seal, and be on its way to fully healed scar tissue.

If imaging was able to find the precise location of a leak, blood will be injected in that spot using image guidance (called a targeted blood patch). If not, a blind blood patch may be performed (injecting blood into the suspected area with the hope that the leak is covered). When the blood is injected, the patient may feel pressure or other sensations. The patient may be kept awake via conscious sedation so that



they are able to confer with their doctor during the procedure and report any unusual sensations or changes in pain.

*To learn more about venous fistulas, dural tears, bone spurs, or other more complicated repairs, visit <https://spinalcsfleak.org/dr-schievink-explains-dura-mater/>*

It has been estimated that over 80% of patients respond favorably to a blood patch, though some patients may require more than one procedure to fully seal.

Fibrin glue (a pooled blood product) may be used instead of or in addition to the patient's own blood, as the glue creates a longer lasting seal. More complicated issues may require surgical repair using stitches or clips, and if bone spurs caused the puncture, then those can be removed prior to repair. The volume or amount of blood injected can range from 10 to 100 mL.

While many anesthesiologists may be able to perform blood patch procedures, it's important to find a physician experienced in long-term leaks or more severe cases. Bear in mind that many leaks begin when an anesthesiologist accidentally punctures the dura, and the follow up care for long-term leakers is much different than the standard epidural puncture recommendation of "take it easy for a day or so."

## Post Patch

Each specialist may have their own post-patch care guidelines, and recommendations may vary dependent on the case. Most recommend no bending, lifting, twisting (called BLT among leakers), or straining/Valsalva for 6-8 weeks following the procedure.

Patients who have been leaking long-term or have connective tissue disorders may do well to extend those restrictions even further. The dura takes time to heal, with some estimates of it being up to a year before the scar tissue is fully healed.

Some helpful tips include: be prepared for a possible 24-48 hour completely flat recovery period before the looser restrictions begin, get a grabber tool to avoid bending over, ask for help with whatever tasks might need completed, plan to not lift children or dogs, and move whatever you might need to chest level before your patch procedure date.

*Some leaks may occur due to an existing high pressure issue. Refer to the intracranial hypertension page if you believe an existing high pressure condition needs addressed.*

Post-patch symptoms may vary greatly and it's common to be confused about what is happening as the body tries to resume normal. There seems to be no real data about what to expect, and because the interventional radiologists are focused primarily on the procedure while a good deal of the medical community seems at a loss regarding leak symptoms, the newly sealed may feel a bit adrift. Follow up care may be needed if there are any severe issues or signs of infection (for instance, temp over 101, neck stiffness, new cognitive issues, numbness or weakness in legs or feet, or bladder issues).

A few of the more common questions may be: *did it work?* and the answer is often that time will tell, because as noted above, the blood patch is only meant to encourage wound healing so until that period has passed a person may not know and should remain on full restrictions; *is this normal?* and the answer is often  $\neg(\text{ツ})\neg$  because no real data is available, but, also often, yes; and *why do I feel worse lying down now?* and the answer may be...

## **Rebound High Pressure**

Rebound intracranial hypertension (RIH) can occur after a blood patch. In long-term leakers, the body has been compensating for the loss of CSF fluid. When the leak is suddenly sealed, it may continue to produce at that volume for some time, creating excess pressure.

High intracranial pressure can also cause symptoms, including headache (though the presentation may differ from low pressure headache), cognitive issues, nerve pain, blurry or double vision, nausea, tinnitus, and more.

**Unlike low pressure/leak symptoms, high pressure symptoms may worsen when flat.** Many leak specialists may prescribe acetazolamide (Diamox), furosemide (Lasix), topiramate, or other medications until fluid production levels out.

It is ideal to discuss the possibility of rebound high pressure before your procedure to ensure, if it occurs, steps can be taken promptly.

As with leak symptoms, high pressure can feel confusing. Pressure can fluctuate and can be affected by food or activity, and the medicines prescribed for high pressure work by lowering pressure (so if a dose is too high, low pressure symptoms could potentially occur) or side effects may occur from the medicine. Patience is important, because keeping the pressure low may help prevent the reopening of (or additional) leaks.

### ***Tips to help manage rebound high pressure***

- Limit sodium, caffeine, and sugar.
- Sleep on an incline with a wedge pillow or raise the head of your bed.
- Get upright if possible (depending on your restrictions and physician's recommendation) and practice gentle walking through the house as often as is comfortable.
- Be careful of Vitamin A, temperature changes, and other personal triggers.

Or check the [intracranial hypertension](#) page and <https://csfleak.uk/resource/aftercare> for more.

*Please check the Spinal CSF Leak Foundation Physician Directory for doctors using best practices for leak diagnosis and repair at <https://spinalcsfleak.org/directory/>*

**Advances in spinal CSF leak testing and diagnosis are moving fast. It is critical that imaging is read by and patients are treated by an experienced leak specialist with a proven track record.**

## **Resources**

Spinal CSF Leak Foundation <https://spinalcsfleak.org/about-spinal-csf-leaks/treatment/>

CSF Leak Association <https://csfleak.uk/>

Directory of Imaging and CSF Leak Specialists—where to go for help  
<https://spinalcsfleak.org/directory/>

*Please note that spinal CSF leaks differ from cranial CSF leaks. Cedars Sinai lists cranial symptoms as drainage from the nose (rhinorrhea), salty or metallic taste in the*

*mouth, sense of drainage down back of throat, drainage from the ear (otorrhea), cutaneous sinus tract drainage (CSF leaks into the sinus tract, which then creates a pathway to drain through the skin), loss of sense of smell (anosmia), change in hearing or ringing in the ears, and less frequently, cognitive changes. Cranial and spinal leaks may occur at the same time. Cranial CSF leaks carry an increased risk of meningitis.*