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Overview of CSF Leaks and Intracranial Hypotension

A cerebrospinal fluid (CSF) leak is an underdiagnosed, yet often hugely debilitating, medical condition comprising a small tear or hole in the outer membrane that contains the fluid surrounding the spinal cord and brain (known as the [dura mater](#)).

When the fluid leaks out of the dura, particularly at a point along the spine, the overall pressure of fluid within the skull drops (known as [intracranial hypotension](#)), its cushioning effect is reduced and the brain is displaced downwards. This slumping effect, also called 'brain sag', results in severe pain, pressure, blurred/double vision, eye pain, neck pain, hearing impairment, tinnitus, spasm, seizures, cognitive impairment and a wide range of other symptoms.

A key characteristic of a CSF leak is that the vast majority of symptoms, if not all, worsen markedly upon sitting or standing, but lessen substantially, and in some cases disappear completely, when lying down. The longer a leak goes untreated, the less prominent this orthostatic characteristic of symptoms may becomeⁱ.

Many people develop a leak as a result of a lumbar puncture or an epidural injection or anaesthesia (iatrogenic), while others develop them following trauma - ranging from major trauma and whiplash, to minor trauma which may otherwise have gone unnoticed.

Sometimes a CSF leak can be an unintentional consequence of an over-draining shunt, placed for the treatment of [hydrocephalus](#) or [intracranial hypertension](#) (high pressure within the skull), or as a result of underlying intracranial hypertension that has gone undiagnosed and/or untreated (where the high pressure may stress the dura and cause a hole to form).

Cranial leaks may occur in the nose following surgery ([rhinorrhea](#)), from the external auditory canal ([otorrhea](#)) or from an operative defect in the skull. They can also be a common complication following severe head injury, where CSF may be observed draining from the nose, ear or discharging to the back of the throat (a [Beta-2 transferrin](#) test can be used to test for CSF).

Spinal CSF leaks, particularly those resulting from trauma or occurring spontaneously, are commonly found at the [thoracic spine](#) and [cervicothoracic junction](#)ⁱⁱ. Spinal leaks have also been reported due a variety of conditions affecting the vertebrae, and increasingly are seen in association with [osteophytes](#) (small spurs of bone) and calcified discs.

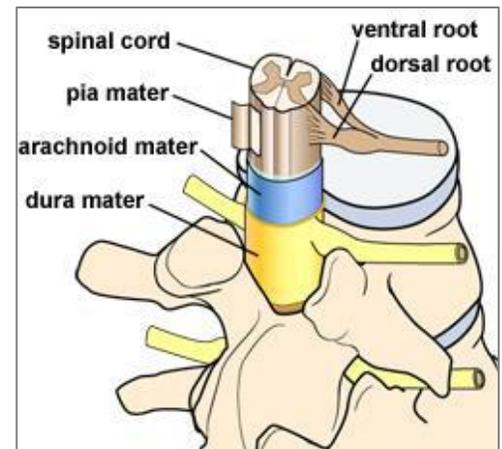


Figure 1 – Anatomy of a Spinal Cord

Questions? Need more information?
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[Spontaneous CSF leaks](#), where the onset of symptoms comes out of the blue and no obvious trigger is noted, were once considered rare, but are now known to be far more prevalent than first thought. Spontaneous CSF leaks are increasingly attributed to an underlying congenital disorder, such as [Ehlers-Danlos Syndrome](#) and other [connective tissue diseases](#)ⁱⁱⁱ.

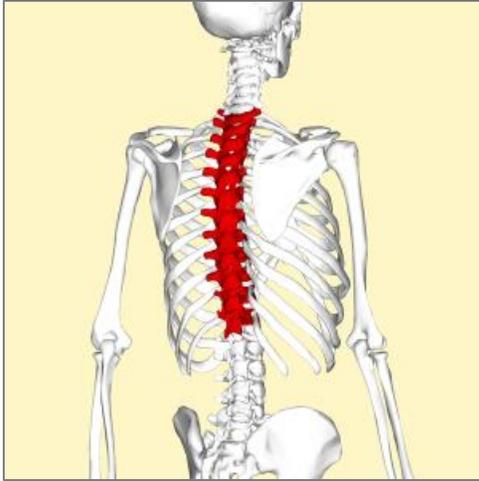


Figure 2 - The Thoracic Spine

Anatomography <http://lifesciencedb.jp/bp3d/?lng=en>

In many cases, a CSF leak will heal on its own accord following conservative treatment, including bed rest, increased fluid intake and caffeine. Some patients may require a clinical procedure known as an [epidural blood patch](#) in order to promote healing; low-volume if the leak location is known following [MRI](#), [CT Myelography](#) or [MR Myelography](#) imaging or high-volume if it remains undetected.

Where a blood patch is not successful, a surgical sealant known as [fibrin glue](#), or for more complex cases, neurosurgery, can be used to good effect if the site of the CSF leak is known. In a lesser, but certainly not insignificant, number of cases, cerebrospinal fluid may continue to leak despite repeated attempts at patching, fibrin glue application or neurosurgery.

Where a leak continues, either through unsuccessful treatment or delayed diagnosis, long-term disability is not uncommon due to severe chronic pain, visual disturbance, seizures and nerve damage. Accordingly, sufferers may be unable to work, undertake any form of physical exercise or carry out basic day-to-day functions that able-bodied people would take for granted.

While knowledge of CSF leaks may be limited within the general public and some sectors of the medical profession, such as general practice and emergency medicine, they are not uncommon and spontaneous leaks alone affect at least 5 in 100,000 people every year^{iv}. While the average age for a leak developing spontaneously is 42, and women are thought to be twice as likely as men to develop the condition, they can occur at any age through trauma.

CSF leaks were first documented in the 20th century by German neurologist Georg Schaltenbrand (1938) and later by American physician Henry Woltman of the Mayo Clinic (1950s). Despite the passing of time, CSF leaks are not fully understood, remain commonly misdiagnosed (particularly as migraine or sinusitis) and, as an area of clinical study, are significantly under-researched.

Other conditions which share some of the positional symptoms of a CSF leak are [orthostatic hypotension](#) (which can cause dizziness on standing), [positional vertigo](#) due to inner ear disturbances, and [orthostatic tremor](#). It has been suggested that orthostatic hypotension can sometimes develop secondary to a CSF leak; particularly where symptoms result in long periods of bed rest and physical deconditioning occurs.

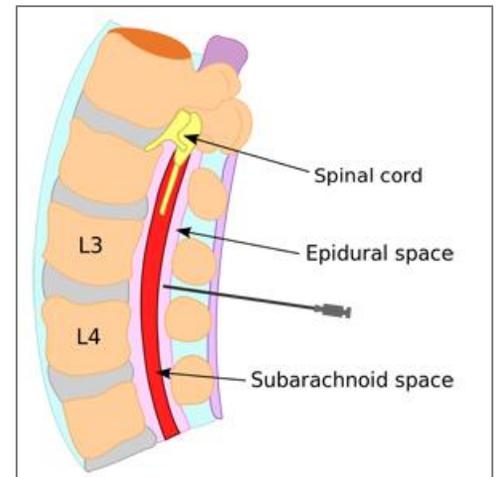


Figure 3 - Epidural Blood Patch

Further Information

For more information on CSF Leaks and Intracranial Hypotension, including causes, symptoms, diagnosis and treatment, or to download copies of our other factsheets, please visit our website: www.csfleak.info

ⁱ Schievink, W. 'Spontaneous Spinal Cerebrospinal Fluid Leaks and Intracranial Hypotension'. JAMA, May 17, 2006—Vol 295, No. 19: 2288

ⁱⁱ Schievink, W. 'Spontaneous Spinal Cerebrospinal Fluid Leaks and Intracranial Hypotension'. JAMA, May 17, 2006—Vol 295, No. 19: 2292

ⁱⁱⁱ Schievink W et al. Connective tissue disorders with spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. Neurosurgery. 2004;54:65-70

^{iv} Schievink, W. 'Spontaneous Spinal Cerebrospinal Fluid Leaks and Intracranial Hypotension'. JAMA, May 17, 2006—Vol 295, No. 19: 2286

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