

Spontaneous intracranial hypotension pathophysiology

The underlying cause of [spontaneous intracranial hypotension](#) is usually a spontaneous [cerebrospinal fluid fistula](#), however, some cases may be due to low [cerebrospinal fluid volume](#). Evidence supports an underlying weakness of the meninges as a contributing factor; for instance, connective tissue disorders like [Marfan syndrome](#), and [Ehlers-Danlos syndrome](#).

Spinal diverticula, at the cervicothoracic junction or thoracic spine (thoracic being more common), and excluding lumbosacral perineural cysts, are thought to be the source of CSF leak in most patients. No relationship has been found between cranial leaks and SIH.

Other causes of dural injury are degenerative disc disease, osteophytes, and bony spurs.

The orthostatic headache is believed to be caused by the descent of the brain, causing strain on intracranial structures sensitive to pain.

Considering that the skull is a rigid noncollapsible container, loss of CSF volume is typically compensated by subdural fluid collections and by increase in intracranial venous blood which, in turn, causes pachymeningeal thickening, enlarged pituitary, and engorgement of cerebral venous sinuses on magnetic resonance imaging (MRI). Another consequence of CSF hypovolemia is sinking of the brain, with descent of the cerebellar tonsils and brainstem as well as crowding of the posterior fossa noted on head MRI. The clinical consequences of these changes include headaches that are often but not always orthostatic, nausea, occasional emesis, neck and interscapular pain, cochleovestibular manifestations, cranial nerve palsies, and several other manifestations attributed to pressure upon or stretching of the cranial nerves or brain or brainstem structures. CSF lymphocytic pleocytosis or increase in CSF protein concentration is not uncommon. CSF opening pressure is often low but can be within normal limits. Stigmata of disorders of connective tissue matrix are seen in some of the patients ¹⁾.

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Mokri B. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. Headache. 2013 Jul-Aug;53(7):1034-53. doi: 10.1111/head.12149. Epub 2013 Jun 28. Review. PubMed PMID: 23808630.

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