Spinal cerebrospinal fluid (CSF) leak is an important and under-diagnosed cause of new 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 defect, hole or tear, the result is a loss of CSF volume, known as intracranial hypotension.

Causes of spinal CSF leaks:

Iatrogenic – caused by a medical procedure intentionally or inadvertently.
(a) Post lumbar puncture CSF leak (often known as Post Dural Puncture Headache = PDPH) is the most common cause of a spinal CSF leak. The lumbar dura is intentionally punctured for various diagnostic and therapeutic reasons. Most often these holes heal over quickly, but in some cases, they do not. It is not yet common practice to use a less traumatic type of lumbar puncture (LP) needle (pencil-tip vs sharp), even though these reduce the risk of post-LP headache.
(b) Dural tears may occur inadvertently at the time of epidural injections (epidural space is in spinal canal outside of dura and spinal cord).
(c) They may occur at the time of surgery.
(d) CSF shunt over-drainage may cause intracranial hypotension.

Traumatic – caused as a result of an injury.
Traumatic leaks have been reported in association with brachial plexus injuries, spinal injuries, sports injuries and chiropractic neck manipulation.

Spontaneous – often occurring with minimal or no clear precipitant. Common reported events or mechanical factors associated with the onset of symptoms include lifting small or large items, straining, stretching, positional changes, sporting activities, roller coaster rides and falls. Some of these cases might be categorized as traumatic.
(a) Spontaneous spinal CSF leaks may be associated with spinal pathology such as calcified disc material or bone spurs. These leaks are usually ventral or anterior to the spinal cord.
(b) There is a growing evidence base suggesting that a significant proportion of spontaneous spinal CSF leaks occur as a result of pre-existing dural weakness. A range of dural defects have been reported at surgery. Electron microscopy of dura has revealed abnormalities in a substantial proportion of cases. Heritable Disorders of Connective Tissue occur at a higher frequency in affected individuals; intracranial hypotension may be the first noted manifestation. Marfan syndrome, Ehlers Danlos syndrome (both classic and hypermobility type) and unspecified heritable disorders of connective tissue have been reported. Patients with spontaneous spinal CSF leaks have been shown to have higher risk of intracranial aneurysms, bicuspid aortic valve and thoracic aortic aneurysms and should be evaluated for Heritable Disorders of Connective Tissue.

Spontaneous intracranial hypotension secondary to spinal CSF leak is uncommon, but not rare. One estimate of annual incidence is 5 in 100,000, however this has not been well-studied. Unfortunately, misdiagnoses and delayed diagnoses are common in this subset.

Key Points
- Spontaneous spinal CSF leaks are recognized less readily than iatrogenic spinal CSF leaks
- Spontaneous spinal CSF leaks are often related to an underlying Heritable Disorder of Connective Tissue; intracranial hypotension may be the first noted manifestation