

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**.

Symptoms

The most common symptom is a positional headache that is worse when upright and improved when lying down, but there are a number of other signs and symptoms. It is important to note that headache is not universally present nor universally positional.

Additional symptoms and signs vary considerably. Common symptoms include nausea, vomiting, neck pain or stiffness, imbalance, pain between shoulder blades, arm pain, sensitivity to sound and/or light, hearing changes, dizziness and impaired thinking. Serious complications such as dementia, Parkinsonism with tremor and unsteady gait, coma, stroke, quadriplegia and death are rare but do occur.

Many patients are quite disabled by their inability to be functional while upright. Suffering for months or years before diagnosis is common.

Causes

Spontaneous – occurring with minimal or no clear precipitant. This is the type that usually goes unrecognized as a spinal CSF leak. These spontaneous leaks are often associated with:

- a. abnormal dura as is seen with a number of heritable disorders of connective tissue, such as Marfan syndrome or Ehlers-Danlos syndrome
- b. spinal pathology, such as calcified intervertebral discs or bone spurs

Iatrogenic – caused by a medical procedure such as:

- a. lumbar puncture to sample and test CSF or to inject contrast or medications
- b. epidural injections or anesthesia
- c. surgery
- d. chiropractic manipulation

Traumatic – caused as a result of an injury, such as a fall or motor vehicle accident.

How common?

We don't know. There has been limited research to date.

Study of the incidence of **traumatic** cases and **iatrogenic** cases has been very limited. Because **spontaneous** cases often go unrecognized, the incidence and prevalence remain challenging to measure. A study based on an urban emergency department estimated annual incidence for spontaneous cases at 5 per 100,000 per year, but this is thought to be an under-estimate.

Diagnosis

Symptoms – The diagnosis is suspected largely on the basis of symptoms.

Physical Findings – These may be largely absent, or there may be neurologic findings and/or signs of heritable disorders of connective tissue.

Brain imaging – MRI of the head without and with contrast should be performed with few exceptions. There are several typical findings seen in about 80% of cases although normal imaging does not rule out the diagnosis.

One of the findings, cerebellar tonsillar descent, is often mistaken as congenital Chiari, occasionally leading to inappropriate and unsuccessful posterior fossa decompression.

Spinal imaging – Since some patients will respond well with one or more EBPs, spinal imaging may not be required in all cases. A range of myelographic imaging (MRI, CT, digital subtraction) is used to locate a CSF leak. Unfortunately, spinal imaging lacks the sensitivity to visualize a leak in a large percentage of cases. This can impact treatment options and outcomes negatively.

Treatment

Conservative – Bedrest, fluids and caffeine are used if the symptoms are not severe.

Epidural blood patch (EBP) – This mainstay of treatment may be performed without spinal imaging in some cases. This need not be directed at a known or suspected level of the leak and can be repeated a number of times. Many cases respond well and are cured with one or more EBPs.

Epidural patch with fibrin glue +/- blood – Treatment is usually directed at known or suspected leak locations.

Surgery – Some patients will require one or more neurosurgical procedures.

Prognosis

Most patients do well, but some patients continue to suffer with residual symptoms of variable severity for years or decades.

The Challenges

Often very disabling – Associated disability may be modest or profound, which is troubling given that treatment options exist and many patients can be cured.

Low awareness - Misdiagnosis and delayed diagnosis of spontaneous spinal CSF leak is common and is largely related to a lack of familiarity among physicians.

Few centers with expertise – This limits access to timely and appropriate care.

Insensitive spinal imaging – This adversely impacts treatment options and outcomes.

More research needed – A greater understanding of all aspects of the disorder is needed. Refinement of diagnostic imaging and treatment approaches should contribute to improved short and long-term outcomes for patients.