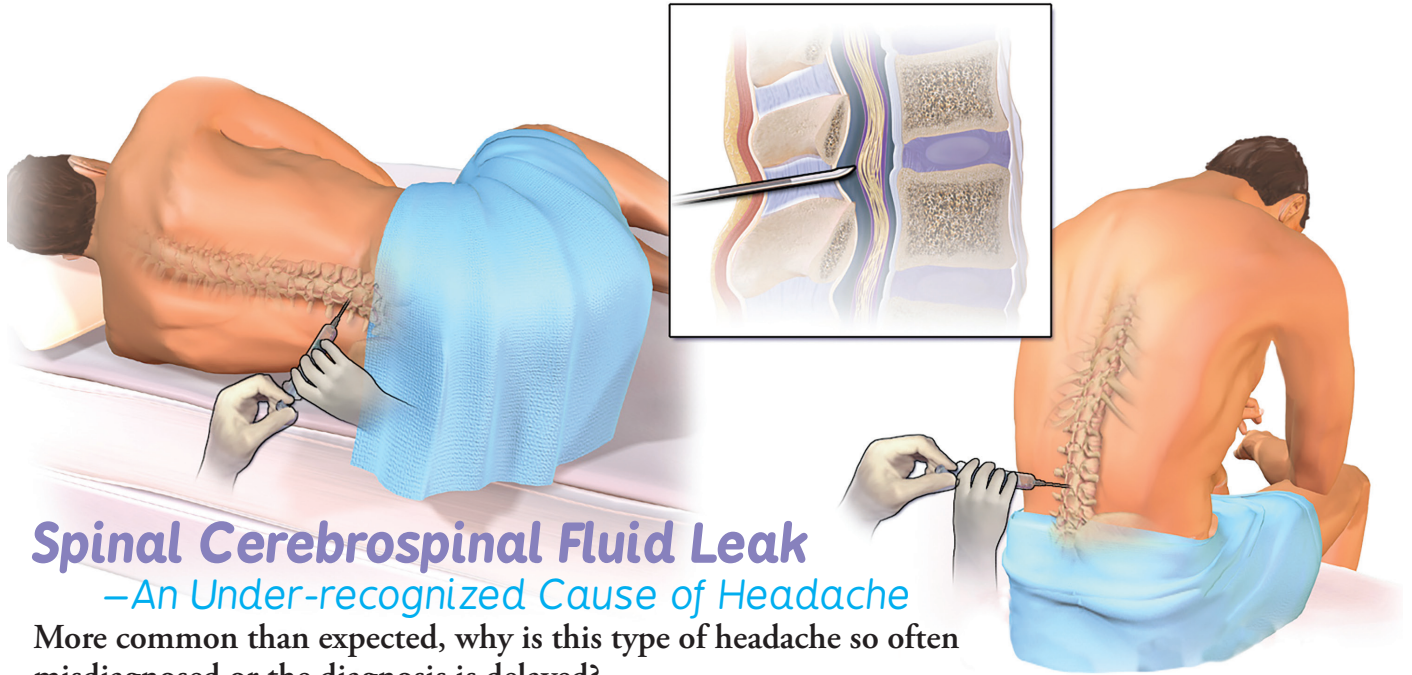


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Spinal Cerebrospinal Fluid Leak

—An Under-recognized Cause of Headache

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Spinal Cerebrospinal Fluid Leak

– An Under-recognized Cause of Headache

Connie Deline, MD,
Spinal CSF Leak Foundation

Spontaneous intracranial hypotension, or low cerebrospinal fluid (CSF) pressure inside the head, is an under-recognized cause of headache that is treatable and in many cases, curable. Although misdiagnosis and delayed diagnosis remain common, increasing awareness of this condition is improving the situation for those afflicted.

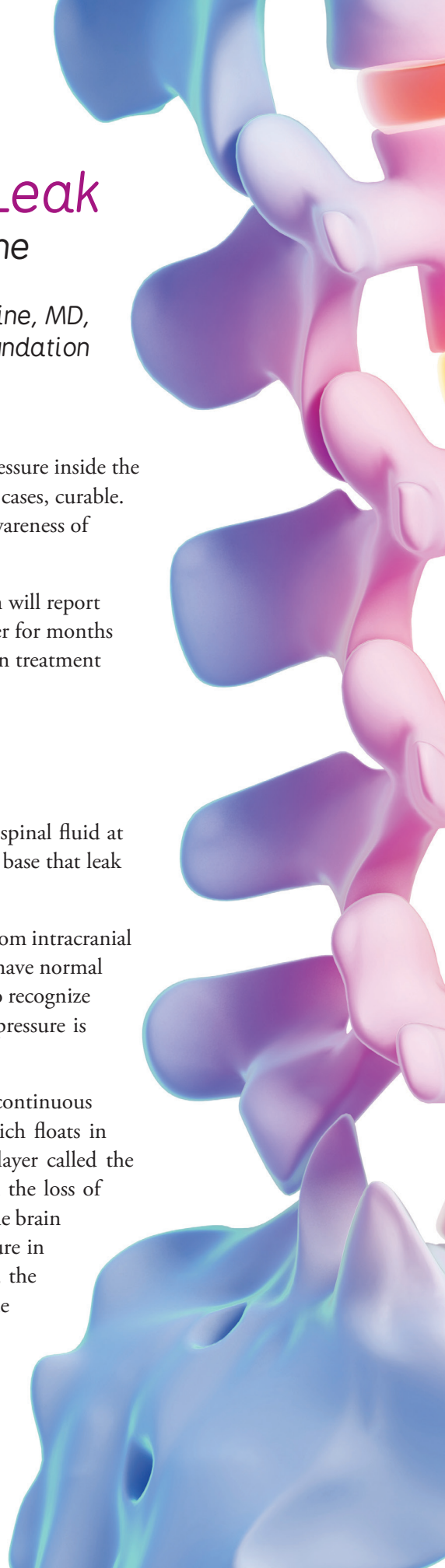
Frequently, patients with a confirmed diagnosis of intracranial hypotension will report that they have been treated for chronic migraine or another headache disorder for months or years. This type of headache rarely responds to medications; however, when treatment is directed at the appropriate underlying cause, most patients respond well.


Anatomy of Intracranial Hypotension

In almost all cases, intracranial hypotension results from a leak of cerebrospinal fluid at the level of the spine. Spontaneous CSF leaks located at the level of the skull base that leak from nose or ear are not causally associated with intracranial hypotension.

Low pressure headache is a term commonly used for the headache resulting from intracranial hypotension. It is important to note that a significant percentage of patients have normal cerebrospinal fluid pressure measurements. Our understanding has evolved to recognize that a low volume of the cerebrospinal fluid is the core problem although pressure is related to volume.

The cerebrospinal fluid surrounds the brain and spinal cord in one continuous compartment. One of its many functions is to cushion the fatty brain which floats in this watery fluid. The fluid is held in place by a tough connective tissue layer called the dura mater, or simply the dura. A hole or tear of the spinal dura results in the loss of cerebrospinal fluid volume around the brain and spinal cord, and sinking of the brain inside the head. When the patient is lying flat, the cerebrospinal fluid pressure in the head and the spine are equal. However when the individual is upright, the cerebrospinal fluid pressure in the head is much lower than the pressure in the lower spine, as with any column of fluid. When a loss of cerebrospinal fluid volume occurs, this results in a larger than normal drop in pressure in the head when the patient goes from lying flat to being upright. This pressure change during positioning explains why symptoms are usually worse when the patient is upright.





It is very common for patients with intracranial hypotension due to a spinal CSF leak to be misdiagnosed with migraine headache, cervicogenic headache, occipital neuralgia, or other headache disorders.

The Headache

The most common symptom associated with intracranial hypotension is an “orthostatic” or positional headache. When the patient rises from lying flat, the pain often worsens within seconds to hours and improves with lying flat again. When severe, the patient may be unable to raise their head even one inch off the bed or may need to position their head lower than their spine for relief. This positional aspect tends to be more abrupt soon after the onset of the leak, and may become less evident over time. The headache may evolve into a “second-half-of-the-day headache” or a chronic daily headache without an obvious postural component. It is very common for patients with intracranial hypotension due to a spinal CSF leak to be misdiagnosed with migraine headache, cervicogenic headache, occipital neuralgia, or another headache disorder.

Usually, the headache is daily but in some cases, may be intermittent. The pain is located most often at the back of the head, but may be at both temples, across the front of the head, or all over the head. Severity can range from mild to excruciating. The headache quality may be variably described as pulling, pressure, or throbbing.

Some patients remain quite functional despite their symptoms while others have very limited ability to be functional when sitting or standing upright. Patients may be largely bedridden. The degree of disability is often under-appreciated.

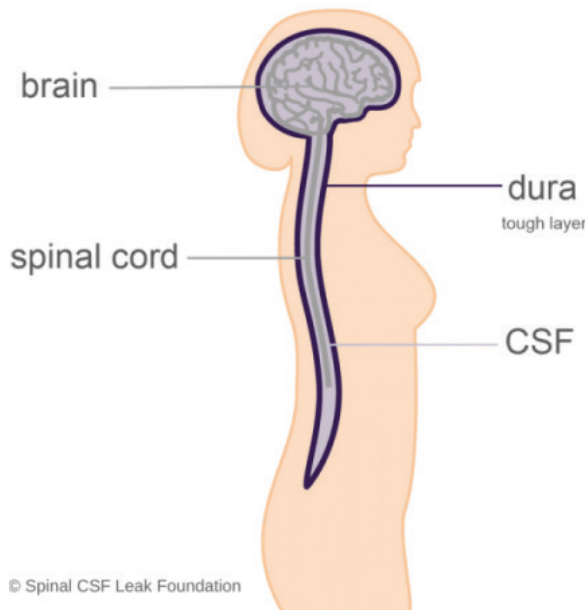
Headache is not universally prominent or even present. A range of other symptoms may be more troublesome than head pain for a subset of patients.

Other Symptoms

Patients with intracranial hypotension often report neck pain or stiffness, nausea with or without vomiting, pain or tightness between the shoulder blades, sense of being off balance, changes in hearing, dizziness or vertigo, sensitivity to light or sounds, cognitive difficulties, as well as arm pain or numbness. Less commonly reported symptoms include visual changes, facial pain or numbness, fatigue, changes in taste, pain at nerve root levels below the shoulders, or fluid discharge from the nipples. More rare presentations or complications include unsteady gait, tremor, dementia, quadriplegia, stroke, stupor/coma, and very rarely, death.

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Underlying Causes

Spontaneous

The cases that occur with no apparent precipitating event or a relatively minor physical factor, such as lifting or bending, are the type that most often remain unrecognized for months or years. There are two known associations with spontaneous spinal CSF leaks. Patients may have an underlying weakness of the spinal dura from an inherited disorder of connective tissue, including but not limited to Ehlers-Danlos syndromes and Marfan syndrome. The second known association is underlying bone spurs arising from calcified intervertebral (between the vertebrae) discs. These bone spurs can puncture the spinal dura.

Spontaneous leaks may occur at any age and in both genders but are diagnosed more often in women around age 40. An estimated incidence of 5 in 100,000 per year is based on a single study of patients presenting to an emergency department and is probably an underestimate since most patients are now diagnosed on an outpatient basis.

Medical Procedures

The most common cause of a spinal CSF leak is a lumbar puncture (spinal tap), in which the spinal dura is intentionally punctured for diagnostic or therapeutic reasons. A positional headache that develops after the procedure is usually recognized and treated promptly.

Another well-known cause of a spinal CSF leak is an inadvertent puncture of the spinal dura during an epidural anesthesia or an epidural steroid injection. These injections normally go into the space inside the spinal canal but outside the dura.

Spinal surgery can be complicated by nicks or tears of the spinal dura. These leaks may or may not be recognized promptly.

Over-draining CSF shunts are also known to cause intracranial hypotension.

Trauma

Cerebrospinal fluid leaks have been reported in association with injuries sustained in falls, motor vehicle accidents, or sports injuries.

Spontaneous intracranial hypotension, or low cerebrospinal fluid (CSF) pressure inside the head, is an under-recognized cause of headache that is both treatable and in many cases, curable.

Most patients who receive treatment for intracranial hypotension do well, with improved quality of life or complete cure.



Diagnosis

The diagnosis of intracranial hypotension relies primarily on the symptoms and history of the patient. The positional aspect of the headache is a key feature in most, but not all, cases. A patient may report a recent history of a lumbar puncture, epidural anesthesia or injection(s), surgery, or trauma. A previous diagnosis of postural orthostatic tachycardia syndrome (POTS) is not uncommon. POTS refers to a condition in which moving from a lying to standing position causes an abnormal increase in heart rate and a range of other signs and symptoms. A patient's positional symptoms may be due to POTS or due to a spinal CSF leak or both. Underlying inherited disorders of connective tissue (Marfan syndrome,

Ehlers-Danlos syndromes, others) may or may not be recognized prior to the onset of symptoms related to a spinal CSF leak. A few patients may have received a diagnosis of Chiari I due to the brain imaging finding of low-lying cerebellar tonsils (part of brain at back of head). Chiari I is a congenital condition in which the back part of the skull is abnormally small or misshapen, and part of the brain, the lower part of the cerebellum, extends into the spinal canal. In intracranial hypotension, the finding of low lying cerebellar tonsils and brain sag is due to the loss of CSF volume and is reversible with treatment of the leak, so it is often called pseudo-Chiari. It can be challenging for clinicians to sort out if this finding on brain imaging is from congenital Chiari or from low CSF volume of intracranial hypotension or from a combination of both. Minimal response to medications used for migraines can be an additional clue to the diagnosis.

DIAGNOSTIC CHALLENGES

- low awareness contributes to delayed diagnosis and misdiagnosis
- not every headache due to intracranial hypotension is positional
- not every positional headache is due to intracranial hypotension
- not every patient with intracranial hypotension has a headache
- common and uncommon presentations and findings may not be recognized as secondary to intracranial hypotension (subdural hematomas, tremor, unsteady gait, dementia, low lying cerebellar tonsils, coma, stroke, spinal cord manifestations)
- normal CSF pressures are not uncommon
- imaging interpretation requires experience and training
- brain MRI is normal in ~ 20 %
- spinal imaging is negative in ~ 50%
- more than one type of spinal imaging is often needed

A diagnostic lumbar puncture to determine pressure measurement or CSF analysis is not usually performed unless another disorder such as meningitis is being ruled out. Minor CSF abnormalities may be noted and cerebrospinal fluid pressure may be low, normal, or even high.

An MRI of the brain with contrast should be done in all suspected cases to determine several classic findings although imaging findings are absent in about 20% of patients. Spinal imaging is used to locate leaks or other abnormalities for targeted treatment – but may be negative in up to one-half of suspected cases due to the limits of sensitivity. A full spine MRI without contrast is often the initial choice because the testing is non-invasive. Myelography, which involves lumbar puncture for the injection of contrast, uses CT, MR, or digital subtraction techniques. More than one type of spinal imaging is often needed.

Treatments

Spinal CSF leaks associated with medical procedures are amenable to targeted treatment approaches.

It is suspected that a substantial percentage of spontaneous cases resolve within days to weeks of onset without any intervention. A brief course of a conservative approach is often recommended if symptom severity and complications do not preclude it. Consuming extra fluids and generous amounts of caffeine, as well as bedrest may reduce symptom severity. Medications often used for migraine headaches are largely ineffective for the head pain. Treatment directed to the underlying cause of spinal CSF leak is needed when conservative measures fail.

Urgent treatment will be needed in cases with serious complications, such as coma or a large subdural hematoma (blood clot pressing on brain inside the skull).

Epidural blood patch (EBP) is a procedure used routinely for post-dural puncture headache and is also the mainstay of treatment for spontaneous spinal fluid leaks. This may be performed after brain MRI with contrast but with or without imaging of the spine. This imaging-guided procedure involves the injection of the patient's blood into the epidural space (space just outside the dura) in the lumbar and/or thoracolumbar region of the spine. It may be repeated several times.

If the response to one or more epidural blood patches is partial or if the symptoms relapse, spinal imaging is performed to try to locate the leak for targeted treatment. If evident on imaging, the leak location and characteristics will dictate the best approach, whether that be epidural patching targeted to the level of the leak or open surgical repair. Targeted epidural patching is done with blood, fibrin sealant (type of glue made from blood products), or both. When spinal imaging is negative, non-targeted epidural blood patches may be repeated or epidural patching may be targeted at suspected leak locations.

Surgical repairs are necessary for some patients depending on factors such as the leak type, leak location, or for patients in whom other measures have failed

Prognosis

Following successful treatment, some patients develop rebound intracranial hypertension or elevated intracranial

pressure. This scenario is usually self-limited but may last for weeks or months, rarely for years. Treatment with medications to lower intracranial pressure is occasionally necessary.

While study of long-term outcomes remains limited, most patients who receive treatment for intracranial hypotension do well, with improved quality of life or a complete cure. Negative spinal imaging, however, limits treatment options for those with partial or temporary response to epidural patching procedures. There are patients, however, that endure persistent symptoms despite multiple procedures.

Summary

A significant percentage of individuals with chronic daily headache may actually be suffering from intracranial hypotension secondary to a spinal CSF leak. These individuals tend to have minimal response to treatments normally used for primary headache disorders. Treatment directed at the underlying cause can lead to improved quality of life or a complete cure. As awareness of this disorder rises, diagnostic delays are becoming shorter. Earlier diagnosis and treatment are critical to reducing the burden of suffering. **HW**



Recommended Reading

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