There is little awareness of Spontaneous intracranial hypotension caused by a spontaneous spinal cerebrospinal fluid (CSF) leak among the public and within the medical community in Canada.

Raising awareness and educating the public and the medical community means more efficient diagnoses and better access to treatment for SIH patients.
It is now recognized that this condition is more common than once believed.

Spontaneous Intracranial Hypotension (SIH)

Spontaneous intracranial hypotension (SIH) is a debilitating and often invisible medical condition resulting from a spontaneous spinal cerebrospinal fluid (CSF) leak. CSF is a clear fluid that surrounds and protects the brain and the spinal cord, and cushions it from injury or damage. A CSF leak stems from a tear or abnormality in the dura, the fibrous tissue that covers CSF circulating around the brain and the spinal cord. A loss of CSF will usually cause the brain to sag in the skull, resulting in a severe headache that is typically positional. This means that the headache is much better lying down compared to being upright. A CSF leak can also lead to other possible neurological symptoms.

Currently, there is no medical diagnosis code for this disorder in Canada.

Symptoms

One of the most common symptoms of a spinal CSF leak is a positional/orthostatic headache. This means that the patient feels partial or complete relief of the headache when he or she lies down and the patient's headache worsens considerably when the patient is upright. Although classic symptoms of CSF leaks are severe headaches that improve when lying down, other signs and symptoms are: head pressure, a feeling of a grabbing sensation in the back of the head, extreme fatigue, nausea and vomiting; neck stiffness or pain, brain fogginess– feeling disconnected, photophobia, phonophobia etc.

Causes

A spinal CSF leak can be caused by a sudden traumatic event or can occur spontaneously. Some predisposing factors linked with a spinal CSF leak include: 1. Connective Tissue Disorders 2. Spine Disorders, 3. Medical Procedures (Iatrogenic), 4. Trauma, 5. Unknown.

Diagnosis

Under-diagnosis and/or misdiagnosis of a spinal CSF leak/ SIH is extremely common, resulting in little or no treatment for some patients suffering from it. Currently, there is not a single diagnostic test that can rule out SIH with a high degree of certainty; therefore, detailed assessment and investigation of symptoms by a specialist is crucial. Some patients with spinal CSF leaks may require a few scans to diagnose a leak, while others require several different types of tests or repeated testing over time to establish the diagnosis. Spinal CSF leaks can exist without any evidence of a leak on imaging.

A negative MRI of the brain or spine does not rule out a spinal CSF leak.

Symptoms are usually key in diagnosing a leak.

- About 15-20% of patients with spinal CSF leak(s) have normal MRI brain imaging.
- A Lumbar puncture is not a helpful diagnostic tool and is not required to make a diagnosis. A normal opening pressure does not exclude the diagnosis.
- Interpretation of imaging requires experience and training. There is a need for higher resolution imaging able to detect leaks that cannot be detected with traditional testing.

Treatments

When a minor spinal CSF leak occurs, sometimes no specific treatments are needed, and the leak will seal itself with bedrest, caffeine and fluid consumption. If conservative treatment fails, an epidural blood patch or epidural patching with fibrin glue are treatments options. Surgery may be considered when the site of the leak is identified, and epidural blood patches have failed.

Prognosis

Spinal CSF leaks are treatable. Once a leak has been successfully sealed, patients can be optimistic about the prognosis. Many times, successful outcome will be measured in months rather than days.

Rebound Intracranial Hypertension (RIH)

RIH is not often reported but it is a common complication of epidural blood patching. RIH is caused by a rebound increase in CSF pressure experienced by many patients after a CSF leak is successfully sealed. In other words, the pendulum may swing rapidly in the opposite direction leading to excess volume of CSF. Patients with increased head pain/pressure when they are flat following an epidural patch should seek medical attention. Relief from a high-pressure headache can often be achieved through the administration of certain medications designed to reduce CSF pressure.