**Definition**

Spontaneous intracranial hypotension is defined as a marked reduction of intracranial cerebrospinal fluid (CSF) volume from a dura tear without any ascertained cause such as trauma or lumbar puncture.1,2

**Epidemiology**

Incidence of spontaneous intracranial hypotension (SIH) is approximate 5 per 100,000 of the population.1 Women are affected more frequent than men in a 2:1 ratio. Incidence of SIH is higher in the fourth and fifth decade of life.5

**Pathophysiology**

The etiology of SIH is not clear. It is believed SIH is resulted from a leaking of CSF from a tear in the dura mater.1 Two major contributing factors to CSF leak are weakness of the dural sac and mild trauma.3,4 The most common causes of weakness of the dura are connective tissue disorder, meningeal diverticula, and a weakened thecal sac. Schievink & Louy5 identified three precipitating factors for CSF leakage: 1) rapid positional changes such as sudden arising into the upright position from a bending position. This sudden position change shift intracranial CSF down to the spinal canal rapidly, which increases the spinal CSF pressure and leads to a CSF leak; 2) Valsalva maneuvers such as coughing, lifting, labor and delivery. Valsalva maneuvers increase intracranial pressure and subsequently increase spinal CSF pressure that result in CSF leak; 3) a trivial nonpenetrating spinal injury such as cervical manipulation. Cervical manipulation may cause a tear in the fragile meningeal diverticula or attenuated dura and result in CSF leakage.1,4,6,7,8

Most CSF leaks are typically found at either the cervico-thoracic junction or the upper thoracic region.1,7,9 It is because the meningeal sheaths in these areas are thinner than in other regions.10 The less common sites are lumbar and sacral regions. In most cases, CSF leakage sites are unidentifiable.8,11,12

Complications of SIH include cortical vein thrombosis, subarachnoid hemorrhage, subdural hematoma, brainstem compression, and cerebellar tonsillar herniation.1

According to Kellie-Monroe doctrine, decreases in volume of one intracranial component will result in increases in volume of other component. In SIH, decreases in CSF volume will increase venous blood volume especially the bridging veins. Due to the downward displacement of brain when patient is in the upright position, it increases stretch to the engorged bridging veins and increases risk of bridging vein rupture and subdural hematoma.13 Radiographic results may also find venous sinus engorgement, pachymeningeal enhancement, subdural effusion, and enlargement of the pituitary gland.14

**Manifestations**

Manifestations of SIH are related to the intracranial hypovolume which leads to the brain shifting towards the foramen magnum.1,12

Orthostatic headache is common and occurs in 76% of patients with SIH. The majority of these patients developed headache within 15 minutes after a change from supine to a standing position. This is relieved when patient return to a recumbent position. Other presentations include nausea, vomiting, neck stiffness, hypacusia (hearing impairment), tinnitus, and photophobia.1,2,6,7,15

Orthostatic headache is related to intracranial hypovolemia. When a patient is in the upright position, the brain descents to the bottom of the cranial vault and creates traction of the pain fibers located on the cerebral blood vessels and meninges resulting in headache.7 Dilation of the cerebral venous blood vessels in SIH is another cause of headache.4 Fifty percent of patients present with subdural fluid collection.1,15

Descent of the brain may lead to distortion and compression of the brain stem and some of the cranial nerves and result in various cranial nerve palsies. Ophthalmoplegia may occur as a result of cranial nerve palsy. Approximate 30-35% of intracranial hypotension patients presented with ophthalmoplegia. Eighty percent of these patients had abducens nerve paresis. Abducens nerve is more vulnerable due to its long intracranial course. Occular manifestations include decreased visual acuity, visual field deficits, nystagmus, and photophobia.14

**Diagnostic Tests**

The criteria established by the International Classification of Headache Disorders (ICHD) for diagnosis of SIH include: A) orthostatic headache that is associated with neck stiffness, tinnitus, hypacusia, photophobia, or nausea; B) Evidence of low CSF pressure or leak by conventional myelography, CT myelography or cisternography, or lumbar puncture CSF opening pressure <60 mmH2O in sitting position; C) No history of dural puncture or other cause of CSF fistula.1,16

Both magnetic resonance imaging (MRI) and CT scan have been used to diagnose SIH. Researches have shown CT scan is not effective in diagnosis for SIH but may be helpful to rule out other causes of headache.5,17

Magnetic resonance imaging is the preferred test for SIH.10 Brain MRIs are able to reveal any anomalies such as subdural hematomas or hygromas, brain herniation, descent of cerebella tonsils, and diverticula. The compensatory venous vasodilation in SIH causes greater concentration of gadolinium in the dural vasculature and interstitial fluid.5,18
MRI myelogram shown gadolinium leaking from the C1-2 level

Treatment Options

Depending on the severity of symptoms and whether the CSF leak site can be identified, treatments for SIH include strict bed rest, epidural blood patch (EBP), and surgery.11

Conservative treatment includes bed rest, hydration of patient, caffeine intake, abdominal binder, analgesics, theophylline, and steroid therapy.7 If conservative treatments are ineffective, other treatment options such as EBP, injection of fibrin sealant, epidural saline infusion, or surgery may be required.7,15 Currently, EBP is the most commonly used treatment modality when conservative management has failed.12

Epidural blood patch is to inject 10 to 20mL of autologous blood into the spinal epidural space.7 The injected blood increases the intraspinal pressure and creates a tamponade effect that “plugs” the dural hole and prevent CSF leak. After EBP, a series of healing processes occur, they are fibroblastic remodelling, collagen deposition, and scar formation. The plugged dura hole will be sealed in approximate 3 months.13 More than one EBP may be required to stop the CSF leak.6,19

Intrathecal infusion of saline is usually through a lumbar catheter. Saline infusion rate is between 10mL/h and 30mL/h. Mechanism of epidural saline injection is similar to EBP. However, increases in spinal pressure after infusion is transient, and adiadditional intervention may be required.13

If both conservative and EBP failed, patients are disabled by their symptoms, and the location of CSF leak can be identified, surgical treatment is suggested. Surgical interventions include partial or bilateral laminectomy and/or facetectomy, ligation of the leaking meningeal diverticulum, epidural packing with blood soaked Gelfoam, muscle pledges, or fibrin glue.20

Nursing Implications

When patients are on conservative treatment, provide different types of fluid choices to encourage oral fluid intake. Administer caffeine and analgesia to alleviate headache and discomfort.

Patient may be discharged on the day of EBP procedure. Patient should be advised to report back if fever, back or radicular pain, or any abnormal symptoms occurs.21

Reference


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