# 두개내압저하의 진단과 치료



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# Diagnostic and therapeutic challenges in patients with intracranial hypotension

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Alterations of intracranial pressure may be present in various conditions. Spontaneous intracranial hypotension (SIH) is a rare disabling condition whose main clinical manifestation is orthostatic headache. Severe cases may cause serious complications including decreased level of consciousness and coma. The clinical symptoms and imaging findings of SIH are variable, and diagnosis and therapy are discussed controversially. In this review, we summarized diagnostic and therapeutic approaches associated with SIH.

Key Words: Intracranial hypotension, Headache

# Introduction

Spontaneous intracranial hypotension (SIH) is a syndrome in which a cerebrospinal fluid leak within the spinal axis. A postural or orthostatic headache is the most common of symptoms.<sup>1</sup> The estimated annual incidence is about 5 per 100,000. The SIH usually develop in the fourth to fifth decades of life with a peak incidence around age 40 and affects women more than men by a ratio of 1.5:1.<sup>2</sup> The SIH shows various clinical consequences and imaging findings. Therefore, the diagnosis and management of SIH can be challenging. The aim of this review is to summarize important aspect of SIH, both as far as diagnosis and management options.

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#### Clinical features

The most common manifestation of SIH is an orthostatic headache. It can occur within seconds to minutes of taking an upright position, but can be delayed by hours. The headache usually improves or resolves after lying down, typically within 30 minutes. The initial onset of headache in the majority of patients is gradual or subacute, and headache severity can vary from mild to severe.<sup>1-3</sup> The headache is usually diffuse, but may be localized to one region of the head or may be asymmetric.<sup>1,3,4</sup> Other symptoms of SIH include posterior neck pain or stiffness, and nausea and vomiting. These may be secondary to meningeal irritation. Some patients complain of changes in hearing, tinnitus, and a disturbed sense of balance, secondary to traction of cranial nerves. Rare symptoms include visual symptoms such as visual blurring, visual field defects. and diplopia. Distortion of the pituitary stalk may lead to hyperprolactinemia, galactorrhea and severe brain

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displacement may lead to diencephalic herniation with stupor or coma.<sup>1-3</sup>

Some patients may not have a postural headache and others have exertional headaches or headaches at the end of the day. Spontaneous intracranial hypotension can be an important cause of new daily persistent headache, and should be considered in the differential diagnosis of thunderclap headache.<sup>1-3, 5-7</sup>

# Diagnosis

For diagnosis, the history and physical exam remain important. The International Headache Society published diagnostic criteria (International Classification of Headache Disorders,  $3^{rd}$  edition, beta version), which was shown in Table 1.<sup>8</sup> Typically, CSF opening pressure is  $\langle 60 \text{ nm} \text{ in} \rangle$ SIH, but it can be immeasurable or even negative. It can also be normal in some patients. Majority patients performed magnetic resonance imaging (MRI) of the brain and radionuclide cisternography (RNC) and CT myelography (CTM) remain important tools. Although MRI is used to identify signs of reduced intracranial pressure, RNC and CTM are used to determine the exact site of the CSF leak.

Brain MRI in SIH may show subdural fluid collections, enhancement of the pachymenings, engorgement of venous structures, pituitary hyperemia, and sagging of the brain.<sup>1</sup> Most of the findings of SIH can be explained by compensatory changes related to loss of CSF volume. Subdural fluid collections are commonly observed and may represent hygromas, as compensatory fluid accumulation resulting from the spinal loss of CSF and consequential sagging of the brain, or hematomas due to rupture of bridging veins.<sup>9</sup> Diffuse supratentorial and infratentorial pachymeningeal enhancements result from the dilation of subdural blood vessels.<sup>10,11</sup> However, not all patients with SIH have brain MRI abnormalities. Spinal MRI can be used to locate extrathecal CSF collections but their use for leak localization is limited.

RNC can be helpful for detecting the leak and is a meth-

od to detect indirect signs of SIH. The most common findings on RNC are early accumulation of tracer in the kidneys and bladder, slow ascent of the radionuclide along the spinal axis, paucity of activity over the cerebral convexities at 24 hours, and abnormal root sleeve visualization.<sup>1,2</sup> The main limitation of RNC is the low spatial resolution and the exact site of the CSF leak remains obscure in around one third of the patients.<sup>12</sup> CTM far is the most accurate study for demonstrating the exact site and extent of the spinal CSF leakage.<sup>13</sup> In addition to its accuracy in revealing the site of the leak, it can show meningeal diverticula, dilated nerve root sleeves, extra-arachnoid fluid collections, and extra dural egress of contrast into the paraspinal tissues.<sup>14</sup>

#### Treatment

No major studies have been done on treatment outcomes for SIH. Because most cases of SIH resolve spontaneously, conservative treatment such as bed rest, hydration, caffeine intake, and abdominal binders are recommended for SIH.<sup>4</sup> Some patients may report benefit from theophylline and corticosteroids.<sup>14</sup> It is reasonable to try conservative treatment for one to two weeks before proceeding invasive treatment modalities.

The mainstay of treatment for SIH is the epidural blood patch (EBP). If the EBP is successful, relief of symptoms can be instantaneous or can take hours.<sup>1,2</sup> Initial EBPs are usually in the lumbar region. The mechanism by which the EBP works is not known. It likely forms a dural tamponade, thereby sealing the leak. It may also restrict CSF

**Table 1.** Diagnostic criteria of spontaneous intracranial hypotension

A. Anv	heac	lache	fulfilling	criterion	(
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- B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

 Table 2. Treatment of spontaneous intracranial hypotension

- 1. Conservative measures bed rest, coffee, hydration
- 2. Medications analgesics, caffeine, theophylline, corticosteroids
- 3. Abdominal binder
- Epidural injections homologous blood (epidural blood patch, "EBP"), fibrin glue
- . .

5. Surgical repair or the leak

flow and interfere with CSF absorption, and/or change dural resistance/stiffness.<sup>1-3,15</sup> Localized epidural blood patches may cause sciatica, and even bowel and bladder dysfunctions have been described.<sup>16</sup> Intracranial hypertension following EBP also has been described.<sup>17</sup> If the CSF leak can be located to a specific region of one or two (thoracic) segments or directed EBP is unsuccessful, a CT-guided injection of fibrin glue is an appropriate measure.18 Fibrin glue (fibrin sealant) mimics blood coagulation by forming a stable fibrin clot that can assist hemostasis and wound healing.<sup>18</sup>

Surgery should be considered as follows: (1) Symptoms are severe enough to warrant surgical intervention (2) Site(s) of leak have been identified (3) Symptoms have been refractory to other measures.<sup>19</sup> Surgical therapy consists of ligation of meningeal diverticulae by suture or aneurysm clips, of suturing dural tears or rents, or of packing the epidural space with gelfoam or fibrin glue.<sup>20</sup> Treatment of SIH was summarized in Table 2.

## Conclusions

Although recognition of SIH has increased in the past decade, SIH continues to be frequently misdiagnosed. To improve clinical symptoms of SIH, precise diagnosis and proper treatment are important.

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