

# Postural Headache? It's Not a Tumor!

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## ANNALS CASE

A 42 year old woman presents with 2 days of positional, gradual-onset, diffuse headache associated with vomiting. Her headache improves with lying down and worsens with sitting up. Her vital signs, physical examination, and head computed tomography (CT) results are unremarkable. The diagnosis is intracranial hypotension from a cerebrospinal fluid (CSF) leak through a traumatic dural tear.<sup>1</sup>

I know what you are thinking. We all remember from our medical school days and residency that a postural headache equals a brain tumor. In tumors, though, *postural* means the headache worsens on lying flat and improves on sitting up. Other headache features concerning for brain tumor include gradual onset, progression over time, and worsening in the morning.<sup>2</sup>

For our case, the postural symptoms are the opposite: the headache worsens with sitting up and improves with lying flat! Instead of elevated intracranial pressure from a mass, spontaneous idiopathic intracranial hypotension (IIH) results in low intracranial pressure or volume. The classic IIH case starts with minor trauma leading to a dural tear. CSF leakage from the tear then results in low volume or low pressure within the thecal sac. The headache presumably occurs because of traction on central nervous system structures from a less buoyant or downwardly displaced brain. Another theory involves pain from engorged sensing intracranial veins.<sup>3,4</sup>

## WHEN TO THINK ABOUT IIH

IIH is rare and there are limited data estimating incidence, which is likely part of the reason this diagnosis often is missed. In one case series, 94% of patients received an incorrect diagnosis on their initial presentation.<sup>5</sup> In one patient it took physicians 13 years to figure out the diagnosis!<sup>5</sup> That's a long time to have a headache. Rarely, IIH may result in more severe symptomatology, such as cerebral vein thrombosis, spontaneous subdural hemorrhage, and even coma.<sup>6</sup>

The key feature on history is the positional nature of the headache. Making this diagnosis more difficult, the positional component may become inconsistent over time. Asking about any positional exacerbation when the headaches first started may be the only clue on history for IIH. The headache descriptions can vary significantly. In fact, some patients present with a typical "thunderclap" (sudden and severe at onset) and others with an exertional headache.<sup>3,7</sup> In more severe cases, the intracranial hypotension may result in cranial nerve palsies (most commonly unilateral or bilateral horizontal diplopia from abducens nerve palsy), hypoacusis, tinnitus, or vertigo.<sup>3,8,9</sup> IIH also causes intracranial vascular congestion, which, when present in the pituitary gland, can lead to hyperprolactinemia and galactorrhea. Other reported clinical manifestations include quadriparesis, parkinsonism, and personality changes.<sup>9</sup> Lastly, risk factors are important and may help guide the clinician to the diagnosis. These include patients with Marfan's and Ehler-Danlos's syndromes, presumably because these conditions cause the dura to be more flexible, as well as autosomal-dominant polycystic kidney disease.<sup>9</sup>

In 35% of patients, minor trauma can be identified but it can be exceedingly minor, such as a positional change, reaching for a toy on the ground, chiropractic neck manipulation, sneezing, ground-level falls, roller coaster rides, or sexual activity.<sup>9,10</sup> Admittedly, some of those seem more "minor" than others.

## GETTING THE DIAGNOSIS

In most of the published literature, magnetic resonance imaging (MRI) of the brain with and without gadolinium is the primary diagnostic test. MRI of the spine without

contrast may also be useful to identify the site of a dural tear.<sup>7,11</sup> CT can help diagnose SIIH but has variable sensitivity.<sup>4</sup>

MRI results concerning for SIIH can be remembered with the mnemonic SEEPS (Figure 1).<sup>1</sup> As you may have guessed, “sagging of brain” is not normal. Although the sample size was small, a 2016 article in the *Journal of Emergency Medicine* found CT results indicative of intracranial hypotension in 5 of 7 patients who received a diagnosis of SIIH and 11 of 11 women with intracranial hypotension from an epidural-related CSF leak.<sup>12</sup> Like CT, MRI without gadolinium can diagnose SIIH but may still miss key findings.<sup>13,14</sup> In one case series of SIIH, 93 patients received MRI with gadolinium and 6 received MRI without gadolinium.<sup>13</sup> Of those 99 patients, 93% had at least 1 of 3 SIIH MRI findings, 33% had 2 of 3, and 43% had all 3. Of the 6 patients who received MRI without gadolinium, 5 showed at least 1 SIIH MRI finding. In this study, 47% of the 98 patients who had CT myelography demonstrated a high-flow leak and 8% had a low-flow leak. No leak was identified in the remaining patients (45%).<sup>13</sup>

Lumbar puncture with removal of CSF sounds as though it would worsen the intracranial hypotension and perhaps even cause another dural tear. Fortunately, lumbar puncture seems to only worsen symptoms in 5% of patients.<sup>4</sup> An opening pressure less than 6 cm H<sub>2</sub>O is consistent with the diagnosis of SIIH, but the opening pressure can also be normal.<sup>4</sup> A prolonged recumbent position can result in normal opening pressures, even in cases of MRI-positive SIIH.<sup>9</sup> Lumbar puncture can result in a “dry tap” if the pressure is quite low (I can hear future interns using this as an excuse already).<sup>9</sup>

Although it seems as though the CSF in SIIH should be normal, engorged veins can become “leaky,” leading to xanthochromia, elevated protein levels, or even WBC pleocytosis!<sup>4</sup> This may lead to confusion with spontaneous

SEEPS:
Subdural fluid collection and presence of extrathecal CSF
Enhancement of the pachymeninges
Engorgement of venous structures
Pituitary hyperemia
Sagging of brain or downward displacement of brain

**Figure 1.** MRI findings in SIIH.<sup>3</sup>

subarachnoid hemorrhage and even meningitis in some cases, and both diagnoses may need to be entertained until more definitive diagnostic data are available. The current diagnostic criteria for SIIH are summarized in Figure 2. The helpful acronym “positional LEAK” may help you remember that SIIH is positional plus one of the following: *l*ow opening pressure, *e*pidural blood patch helps, *a*ctive CSF leak, and (*k*) cranial MRI showing SIIH.<sup>11</sup>

## DISPOSITION AND TREATMENT

Although to our knowledge no randomized controlled trials exist to guide management, first-line therapy is typically conservative, with a combination of bed rest, antiemetics, fluid repletion, abdominal binders, and caffeine.<sup>7,9</sup> Yes, caffeine, as in “coffee to our headache patient stat” caffeine. One case report showed complete resolution of symptoms after 48 hours of “200 mL of tea every 2 hours.” The authors noted that because “tea is a...low cost drink...it seems that [it] is a promising method of treatment for SIIH. Despite all these advantages, however, drinking this amount of tea could be unpleasant for the patient.”<sup>14</sup> In the United States, a combination pill of acetaminophen, butalbital (a barbiturate), and caffeine can be used. As for dosage, one review recommended 200 to 300 mg of caffeine 2 to 3 times per day, or approximately 48 ounces of Starbucks’ featured dark roast daily.<sup>9,15</sup>

Another treatment option is an epidural blood patch. A catheter is placed in the epidural space, and 15 to 20 mL of the patient’s own blood is injected. Theoretically, an epidural blood patch helps by increasing pressure in the epidural space, interfering with CSF resorption, as well as fibrosis or scar formation over the dural tear.<sup>4</sup> Headaches typically resolve immediately with an epidural blood patch, although that timing does not fit with the presumed

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| <p>A) Orthostatic headache</p> <p>B) The presence of at least one of the following:</p> <ol style="list-style-type: none"> <li>1. Low opening pressure (60 mm H<sub>2</sub>O)</li> <li>2. Sustained improvement of symptoms after EBP</li> <li>3. Demonstration of an active spinal CSF leak</li> <li>4. Cranial MRI changes of intracranial hypotension (eg, brain sagging or pachymeningeal enhancement)</li> </ol> <p>C) No recent history of dural puncture</p> <p>D) Not attributable to another disorder</p> |
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**Figure 2.** Diagnostic criteria for headache caused by spontaneous intracranial hypotension.<sup>11</sup>

pathophysiology.<sup>4,6,16</sup> MRI performed after an epidural blood patch does show that the epidural clot extends primarily cephalad approximately 3 to 5 spinal segments.<sup>17</sup> If unsuccessful, the epidural blood patch can be repeated with larger volumes of 20 to 100 mL.<sup>4</sup> As for timing and location of an epidural blood patch, some authors recommend starting with an epidural blood patch in the lumbar region for all patients. If the lumbar EBP is unsuccessful, they perform MRI or CT myelogram to try to identify the leak to perform an epidural blood patch directed to that site.<sup>4,6</sup> Others perform MRI or CT myelogram first and start with a directed epidural blood patch.<sup>16</sup> In rare cases, open evaluation is necessary to surgically close the dural tear.<sup>4,7</sup> Other, more controversial options include steroids and theophylline.<sup>4</sup> Typically, cranial nerve deficits resolve with these therapies.<sup>8</sup>

If the patient's pain is tolerable and he or she can tolerate *per os*, discharge seems reasonable with close follow-up for definitive diagnostic testing and reassessment. If the headache is intractable, these patients may benefit from admission, inpatient MRI with contrast, and more aggressive therapies.

## BOTTOM LINE

Remember there are 2 types of positional headache. If it's worse with lying down and better with sitting up, think tumor or intracranial hypertension. If it's worse sitting up and better lying down, think intracranial hypotension! If you don't ask about positional exacerbations, you might miss these treatable, and potentially harmful, causes of headache. SIIH can be diagnosed by orthostatic headache plus either low opening CSF pressure, improvement after an epidural blood patch, active CSF leak, or cranial MRI demonstrating the findings described in [Figure 2](#). Treat with intravenous fluids, caffeine, bed rest, or an epidural blood patch.

## CASE CONCLUSION

In our case, MRI of the brain with gadolinium revealed 2 typical findings of SIIH: enhancement of the dura and prominence of the pituitary gland. Thoracic and lumbar MRI were also obtained, showing extensive extra-axial fluid collections. An epidural blood patch at the fourth to fifth lumbar interspace resulted in complete resolution of the patient's headache.<sup>1</sup>

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