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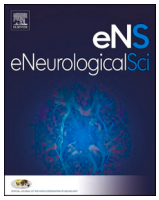

IIH, SIH and headache: Diagnosis and treatment update

Shuu-Jiun Wang [a,b,c,*,1]

[a] Brain Research Center, National Yang Ming Chiao Tung University, Taipei, Taiwan
[b] College of Medicine, National Yang Ming Chiao Tung University, Taipei, Taiwan
[c] Department of Neurology, Neurological Institute, Taipei Veterans General Hospital, Taipei, Taiwan

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ABSTRACT

Idiopathic intracranial hypertension (IIH) and spontaneous intracranial hypotension (SIH) are two major secondary headache disorders resulting from abnormal intracranial pressure (ICP). This short communication outlines the pathophysiology, diagnostic criteria, and management strategies for IIH and SIH. IIH predominantly affects obese women of childbearing age and presents with daily headaches, visual disturbances, and papilledema. It is characterized by elevated cerebrospinal fluid (CSF) pressure, with diagnosis supported by imaging and lumbar puncture. Treatment includes weight reduction, medications, and surgical interventions in refractory cases. SIH, conversely, is caused by spontaneous spinal CSF leaks and presents with acute orthostatic headaches. Diagnosis is supported by neuroimaging and lumbar puncture, revealing low CSF pressure. Treatment includes supported care, (targeted) epidural blood patch, with surgical repair considered in refractory cases. Advances in imaging and treatment have significantly improved outcomes for both conditions.

1. Introduction

Idiopathic intracranial hypertension (IIH) (pseudotumor cerebri) and spontaneous intracranial hypotension (SIH) are two major secondary headache disorders attributed to change of the intracranial pressure (ICP). This article provides a concise overview of the pathophysiology, diagnostic criteria, and management approaches for both IIH and SIH.

1.1. Idiopathic intracranial hypertension (IIH)

IIH predominantly affects obese women of childbearing age [1] with the annual incidence of 1.8 per 100,000 individuals but varies significantly across different populations [2]. The risk factors for IIH include obesity, female sex, and certain endocrine disorders [3]. IIH commonly presents with severe and daily headache. Other common symptoms include transient visual obscurations and pulsatile tinnitus. Papilledema, a hallmark of IIH, may lead to visual disturbances or even permanent visual loss.1 Notably, a subgroup of IIH patients may present without papilledema, making the diagnosis even more challenging.

The pathophysiology of IIH remains not fully understood; however, the dysfunctioned veno-dural junction has been suggested recently, which reduces cerebrospinal fluid (CSF) absorption and, in turn, increases venous pressure and then raises ICP [4].

The diagnosis of IIH is primarily clinical-based and is supported by imaging and lumbar puncture findings. The International Classification of Headache Disorders (ICHD-3) defines IIH as the presence of elevated CSF pressure (>250 mm CSF in adults or > 280 mm CSF in children) with normal CSF composition and a new or worsening headache [5]. Neurological examinations are usually normal except for abducens nerve palsy. Neuroimaging findings such as empty sella, distention of the perioptic subarachnoid space, flattening of the posterior sclera, protrusion of the optic nerve disc, and transverse sinus stenosis can support the diagnosis, especially in the absence of papilledema.

IIH is a significant cause of morbidity due to its impact on quality of life and the risk of permanent visual loss. Early diagnosis and appropriate management, including lifestyle modifications and medical or surgical treatments, are essential to prevent long-term sequelae. Weight loss is the cornerstone of treatment because even modest weight reduction can significantly lower ICP. Medications like acetazolamide and topiramate are commonly prescribed to reduce CSF production. Surgical interventions such as CSF shunting, optic nerve sheath fenestration, or venous sinus stenting are suggested in refractory cases or when vision is threatened [1]. Some new treatments are emerging. Weight watch is important to prevent the relapse of IIH.

* Corresponding author at: Brain Research Center, National Yang Ming Chiao Tung University, Taipei, Taiwan.
E-mail address: sjwang@vghtpe.gov.tw.
[1] Address: No. 201, Sec. 2, Shi-Pai Rd., Beitou Dist., Taipei 11217, Taiwan.

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1.2. Spontaneous intracranial hypotension (SIH)

SIH is a condition characterized by spinal CSF leaks without a history of trauma or medical intervention [5]. The annual incidence is 3.7 per 100,000 individuals, but it is believed to be more common than previously thought because some patients can solely present with atypical symptoms [5]. It affects both sexes with female predominance (2:1) [5]. The hallmark of SIH is acute severe orthostatic headache, which worsens upon upright position and improves with recumbency [6]. Subdural hematoma is the most common (20 %–25 %) and life-threatening complication of SIH [7], followed by rare occurrences of venous sinus thrombosis and superficial siderosis [7].

The primary cause of SIH is a spinal CSF leak, often at the level of the cervical or thoracic spinal dura, leading to decreased CSF volume and pressure [8]. CSF-venous fistula is an uncommon but increasingly recognized cause of SIH, especially in patients where spinal CSF leaks cannot be identified [8,9].

The diagnosis of SIH requires demonstrating typical cerebral or spinal neuroimaging findings or reduced CSF pressure (<60 mm CSF) through a lumbar puncture, along with clinical symptoms [5]. However, a low CSF pressure is not always present or necessary for diagnosis [8]. Brain MRI findings of SIH include diffuse pachymeningeal enhancement, venous distension sign, brain sagging, and subdural fluid collection [7]. Compared to conventional spinal MRI, heavily T2-weighted magnetic resonance myelography (MRM) is a noninvasive technique that does not require intravenous or intrathecal contrast and offers better visualization of spinal CSF leaks [7]. Typical findings of MRM include periradicular leaks, epidural CSF collections, and high-cervical retrospinal CSF collections [7]. Periradicular leaks can be used as a guide for targeted epidural blood patching (EBP) [8].

Early recognition and treatment are crucial to prevent complications such as subdural hematomas or persistent neurological deficits. Conservative treatments for SIH include bed rest, hydration, and caffeine, but have a low success rate of 28 % [6]. Epidural blood patching (EBP) is the treatment of choice [9]. Targeted EBPs with larger injected blood volume ($\geq$ 22.5 mL) are generally more effective [7,10]. In refractory patients who fail at least three or four EBPs, surgical repair of the dural leaks may be considered [9]. For patients with subdural hematoma, burr hole evacuation immediately after EBP is recommended when the hematoma is $\geq$10 mm in thickness or if there is cognitive impairment [7]. The advances in imaging and treatment modalities have improved outcomes for patients with SIH.

2. Conclusion

Proper clinical evaluations, utilizing neuroimaging as well as lumbar puncture, is critical in diagnosing IIH and SIH. Effective management strategies, such as weight loss and pharmacotherapy for IIH, and targeted EBP for SIH, are crucial for improving patient outcomes and preventing complications. Early recognition and targeted treatments remain key to mitigating the morbidities associated with these conditions.

CRediT authorship contribution statement

Shuu-Jiun Wang: Conceptualization, Writing – original draft, Writing – review & editing.

Declaration of competing interest

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