Understanding the Complex Pathophysiology of Idiopathic Intracranial Hypertension: A Comprehensive Review of the Literature and Proposed Management Algorithms

Nisha Giridharan; Smruti K. Patel MD; Amanda Ojugbeli BS; Aria Nouri MD MSc; Peyman Shirani MD; Aaron W Grossman M.D. PhD.; Joseph S. Cheng MD MS; Mario Zuccarello MD; Charles J. Prestigiacomo MD FACS

Departments of Neurosurgery and Neurology, University of Cincinnati College of Medicine, Cincinnati, Ohio

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Introduction

Idiopathic intracranial hypertension (IIH), formerly known as pseudotumor cerebri or benign intracranial hypertension, is a disease defined by elevated intracranial pressure without established etiology. The most common clinical symptoms of IIH include headache and visual complaints. Many current theories regarding the etiology of IIH focus on increased secretion or decreased absorption of CSF and on cerebral venous outflow obstruction due to venous sinus stenosis. Whereas there is now consensus on the definition of the disorder, its complex pathophysiology remains elusive. Several treatments including optic nerve sheath fenestration, CSF diversion with ventriculoperitoneal or lumboperitoneal shunts, and more recently venous sinus stenting have been described for medically refractory IIH. Despite the availability of these treatments, no guidelines or standard management algorithms exist for the treatment of this disorder.

Methods

In this paper, the authors provide a review of the literature of IIH, its clinical presentation, pathophysiology and evidence supporting treatment strategies, with a specific focus on the role of venous sinus stenting. Based this review, the authors provide proposed workup and management algorithms for patients with IIH.

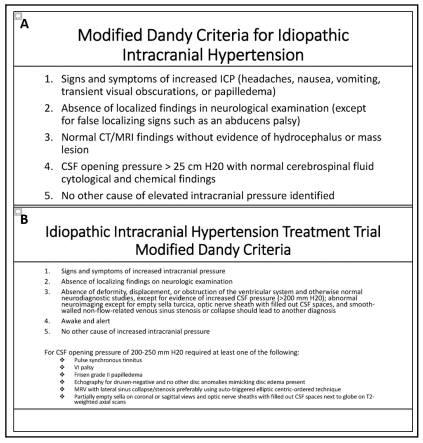

Figure 1: A: Modified Dandy Criteria initially proposed in 1985. Adapted from Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology 2002(59): 492-1495. B: Idiopathic Intracranial Hypertension Treatment Trial updated version of the Modified Dandy criteria with further revisions allowing for the "probable" diagnosis of IIH syndrome in certain circumstances. Adapted from Wall M, Corbett JJ. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology 2014;83(2): 198-200.

Results

For patients with IIH who do not tolerate or experience persistence of symptoms despite maximal medical management, several interventions exist. Based on an extensive literature review, the authors provide a proposed initial workup for idiopathic intracranial hypertension (Figures 1 and 2) and treatment algorithm (Figure 3) for patients with medically refractory IIH.

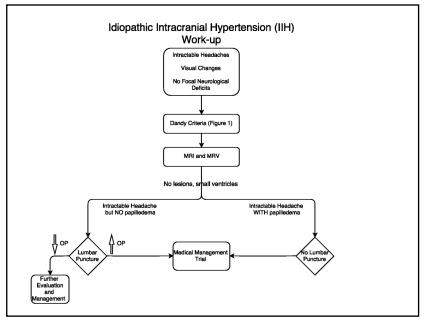

Figure 2: Proposed flow-chart algorithm for initial diagnostic work-up of IIH based on the tenets of the Modified Dandy criteria. We suggest that for patients who meet clinical criteria, imaging workup with MRI and MRV is completed; if negative, then further workup with lumbar puncture is based on the presence or absence of papilledema. All patients with who are found to meet criteria with elevated ICP should undergo a medical management trial. For those patients with clinical symptoms suggestive of IIH without an elevated opening pressure, further evaluation and management is warranted.

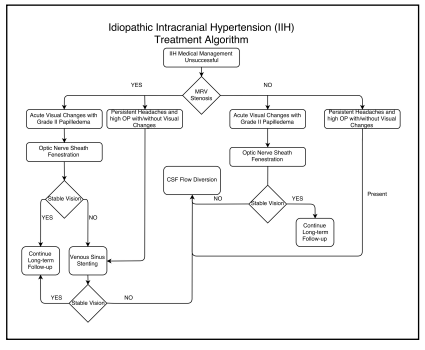

Figure 3: Proposed flow-chart algorithm for treatment and management of medically refractory IIH. For patients with IIH who do not tolerate or experience persistence of symptoms despite maximal medical management, several interventions exist.

Figure 3 (cont'd): As part of the proposed initial workup for IIH (Fig. 2), patients should have an MRV to assess for venous sinus stenosis. For those patients who have evidence of venous sinus stenosis on MRV, persistent headaches, and elevated opening pressures objectively measured by lumbar puncture, with or without visual changes, the provider should consider consulting an endovascular expert to discuss the option of venous sinus stenting (VSS) as a treatment.

If after VSS, the patient experiences resolution of headache and stable vision, then long-term follow-up can be continued. If the patient's vision continues to deteriorate, then CSF diversion should be considered as the next therapeutic option. For those patients who have no evidence of venous sinus stenosis on MRV and persistent headaches and high opening pressure, with or without vision changes, we recommend CSF flow diversion as the most appropriate surgical option.

In all patients with acute visual changes and grade II papilledema, we recommend proceeding with optic nerve sheath fenestration (ONSF). If the patient's visual complaints remain stable, continued long-term follow-up is recommended.

If visual complaints persist after ONSF for patients with evidence of venous sinus stenosis on MRV, practitioners should consider a trial of VSS. If VSS is selected as the therapeutic option and fails to control visual changes, CSF flow diversion can alternatively be offered to the patient for symptomatic relief. For those patients with acute visual changes whose symptoms are not relieved by ONSF and who have no evidence of venous sinus stenosis on MRV, we recommend CSF flow diversion.

Conclusions

While the current evidence appears promising regarding venous sinus stenting in a subset of patients with idiopathic intracranial hypertension, future studies are required with a focus on investigating the long-term outcomes for venous sinus stenting, comparing its efficacy to cerebrospinal fluid diversion and optic nerve sheath fenestration, as well as establishing the best candidates for stent placement in order to create specific best-practice guidelines for the treatment of medically refractory idiopathic intracranial hypertension.

Learning Objectives

1) To understand the current proposed theories regarding the pathophysiology of IIH

2) To understand the current literature and resultant outcomes in IIH patients treated with various surgical strategies

3) To provide proposed workup and treatment algorithms for management of patients with IIH in a more standardized fashion