A Review of Pediatric Idiopathic Intracranial Hypertension

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KEYWORDS

• Pediatric idiopathic intracranial hypertension • Pseudotumor cerebri • Papilledema

KEY POINTS

- Many children who suffer with headaches first present to their pediatrician for evaluation. Intracranial hypertension must be included in the differential diagnosis. Missing the diagnosis could result in permanent and severe visual loss.
- Idiopathic intracranial hypertension occurs in both children and adults.
- Making the diagnosis of idiopathic intracranial hypertension in a child can now be done with more confidence because of newly outlined diagnostic criteria.
- A significant percentage of pediatric patients with intracranial hypertension will have an identifiable secondary cause.
- Most children diagnosed with idiopathic intracranial hypertension will respond well to medical management alone.
- Surgical intervention is reserved for those who fail to respond medically.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is defined as elevated intracranial pressure (ICP) without clinical, radiologic, or laboratory evidence of a secondary cause. The most frequently cited incidence data for IIH in the general population of the United States are from a study by Durcan and colleagues,[1] who reported the incidence to be 1 in 100,000 individuals. When restricting the inclusion criteria to women aged 20 to 44 years, who are 20% or more above their ideal body weight, the annual incidence increases to 15 to 19 cases per 100,000 in the United States.[1] Although it has historically been described as a condition affecting obese women of childbearing age, it can occur in all age groups and genders in both obese and nonobese individuals and is becoming more recognized in the pediatric population.

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NOMENCLATURE

There has been much discussion and debate about the appropriate nomenclature to describe this condition. Historically, the condition has been referred to by terms such as meningitis serosa,[2] otitic hydrocephalus,[3] hypertensive meningeal hydrops,[4] pseudotumor cerebri,[5] benign intracranial hypertension,[6] idiopathic intracranial hypertension,[7] and, most recently, pseudotumor cerebri syndrome.[8] With so many names, confusion and misunderstanding can occur. The more descriptive term "intracranial hypertension," which is then further classified as either "idiopathic" or "secondary," is used in this discussion. The term "idiopathic" is reserved for those cases in which known secondary causes have been excluded. The term "secondary" is used for those cases in which an underlying cause is identified. These terms are simple, descriptive, and understood by all clinicians. It is important to recognize that little is known about the pathophysiology of IIH. It is quite possible that what is referred to today as "idiopathic" may likely become "secondary" as more is learned about the disease process.

DEMOGRAPHICS

More information is becoming available regarding the demographics of pediatric IIH. Current data suggest that prepubertal children affected with intracranial hypertension are more likely to be secondary rather than idiopathic in nature. In addition, the association with obesity and female gender does not hold true in this population.[9,10] Studies also suggest that IIH is infrequent in children less than 10 years of age[9] and extremely rare in infants less than 3 years old.

ASSOCIATED CONDITIONS

There are a myriad of secondary causes of intracranial hypertension and knowledge of these associated conditions will aid the clinician during the history and examination of the patient and can assist in guiding the subsequent workup. It is worth mentioning again that pediatric intracranial hypertension is more likely to be secondary in nature than idiopathic. Systemic conditions, cerebral venous abnormalities, drugs, endocrine abnormalities, and infectious causes have been identified as secondary causes of intracranial hypertension in the literature (**Box 1**). The exact mechanism by which these conditions result in secondary intracranial hypertension is not entirely understood in all cases.

CLINICAL PRESENTATION

The clinical presentation of pediatric IIH includes many of the same symptoms and objective findings as the adult patient. Headache is by far the most common symptom of IIH and occurs in more than 90% of cases.[11] There is no specific headache pattern that is pathognomonic of IIH; therefore, a thorough history and physical examination are needed to try and determine the need for further evaluation. Other symptoms include neck, shoulder, or arm pain, nausea, vomiting, pulsatile tinnitus, diplopia, blurred vision, and transient obscurations of vision.[12,13] Diplopia can be secondary to unilateral or bilateral sixth nerve palsies. The sixth cranial nerve is susceptible to damage from high ICP alone. Infrequently, patients may present with no suggestive symptoms at all and are only diagnosed when papilledema identified on routine eye examination prompts a further workup. For example, a thin 14-year-old girl presented for her regular eye examination and was found to have papilledema. She had been started on clindamycin for acne 8 weeks previously. Only retrospectively when prodded did she complain of new headaches. Within 2 weeks of stopping the clindamycin,