**Pediatric Idiopathic Intracranial Hypertension: A major Review**

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**Introduction and Goals**

Idiopathic Intracranial hypertension (IIH) is a syndrome of elevated intracranial pressure (ICP) without clinical, laboratory and radiographic evidence of intracranial etiology¹. Medical therapy is initiated with a carbonic anhydrase inhibitor such as acetazolamide; diuretics such as furosemide or headache prophylactic agents such as topiramate may be added as well². Neurosurgical interventions may be necessary if patients are refractory to medical management. Prior studies have reported visual acuity loss in 6-20% and visual field loss in 90% of pediatric cases at presentation³⁴. Despite treatment permanent loss of visual acuity occurred in 0-10%² and visual field loss persisted in 17% cases⁵. Even though it is recognized among pediatric population the clinical characteristics in this population are not well described. We performed a multi-team approach to better understand the demographics, presenting features and treatment outcomes of pediatric patients diagnosed with Idiopathic Intracranial hypertension.

**Methods**

We performed a retrospective chart review of pediatric patients diagnosed with IIH between January 1, 2007 and July 31, 2014. Inclusion criteria were as follows: 1) patients age 1 day to 18 years at the time of diagnosis, 2) patients with a primary diagnosis of IIH, and 3) patients with a recorded lumbar puncture (LP) opening pressure. Patients 19 year and older and those without a documented LP opening pressure were excluded from this study. Main outcome variables included demographic characteristics, body mass index (BMI), comorbid diagnosis, presenting symptoms, medical/ surgical treatment, long-term visual outcomes and recurrences.

**Results**

Of the 78 patients, 70.5% were females with a strong female preponderance in adolescents (92.5%) but no significant gender predilection in patients younger than 12 years (Figure 2). The mean age of onset was 11.92 ± 4.09. Ninety-percent of adolescent patients were overweight, moderately or severely obese. A significant association of obesity was found both in elementary (p=0.007) and adolescent age group (p<0.0001) (Figure 3) but there was no significant correlation in pre-K aged patients. Presenting symptoms included headaches (83.3%), blurry vision (48.7%), nausea (41.02%), and photophobia (38.46%) (Figure 4). On physical exam papilledema was noted on presentation in 91% patients (Figure 5). Of the 30 patients with reliable visual fields at presentation 27 patients (90%) had significant visual field changes. Nine percent had cranial nerve VI palsy. Eleven patients (14.1%) had incidental findings of papilledema on routine eye exam that led to their diagnoses of IIH. The average cerebrospinal fluid opening pressure was 304 mmHg. All patients attempted medical management and 14 patients (18.0%) subsequently underwent surgical interventions (Figure 6). Seven (10.9%) of 64 patients who did not undergo surgical intervention experienced recurrence. Permanent visual-field changes at the time of last follow up were seen in 25% patients that had reliable visual field exam. Seven (10.9%) of 64 patients who did not undergo surgical intervention experienced recurrence.

**Conclusions**

Idiopathic Intracranial Hypertension is a condition of preventable irreversible blindness secondary to increased intracranial pressure. In our study the pre-k children with IIH had lower incidence of obesity with no gender predilection. Obesity was found to be of statistical significance in the elementary and adolescent age groups, while the female gender was only found to be significant in the adolescent group. Children with IIH usually complain of headaches and blurred vision and are at risk for permanent visual loss. Majority of children had papilledema and whenever reliable, significant visual field changes at presentation. Despite aggressive management significant visual field changes persisted in some patients. Early diagnosis and prompt management is essential in preventing permanent vision changes in patients with pediatric IIH. Patients should be followed up carefully as recurrence of the disease can be seen despite resolution of signs and symptoms.

**References**