

Symptomatic vs. Asymptomatic Idiopathic Intracranial Hypertension in Children

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Introduction

- Idiopathic Intracranial Hypertension (IIH) is a disease of elevated intracranial pressure (ICP) with normal neuroimaging and cerebrospinal fluid (CSF) analysis
- Certain pediatric IIH patients are incidentally found to have papilledema and elevated ICP without symptoms of IIH (e.g. headache, vision blurring, tinnitus)
- This study aims to characterize the clinical, ocular, and neuroradiological features of asymptomatic IIH in children

Methodology

- Retrospective, case-control study review at one academic institution from 2005-2016
- Charts reviewed to find patients with ICD-9 or ICD-10 diagnosis of IIH in the study period

Inclusion criteria

1. Age 0-18 at time of diagnosis
2. Meets the Modified Dandy Criteria for IIH diagnosis (**Table 1**).
3. No cause of elevated ICP discovered in study period

Subjects were classified as the following:

- **Symptomatic:** ≥1 symptom of IIH at initial presentation (headache, visual blurring, diplopia, tinnitus, nausea/vomiting)
- **Asymptomatic:** zero symptoms of IIH at initial presentation

Bivariate analysis performed. Significance assigned if $p < 0.05$.

Table 1. Modified Dandy Criteria for IIH Diagnosis in Adults and Children
I. If symptoms or signs are present, they only reflect increased intracranial pressure
II. No localizing signs with exception of Abducens (sixth) nerve palsy
III. Patient is awake and alert
IV. Normal CT/MRI findings with no evidence of thrombosis, hydrocephalus, mass, structural, or vascular lesion
V. Documented elevated intracranial pressure with opening pressure of >25cm H2O
VI. Normal CSF composition
VII. No other explanation for raised ICP

Results

53 pediatric subjects diagnosed with IIH → 12 (22.6%) were asymptomatic

Compared to symptomatic IIH, asymptomatic IIH had:

- Younger age of onset (10.00 vs 12.54, $p < 0.05$) (**Fig 1**)
- Lower opening pressure on lumbar puncture at diagnosis (34.25 vs. 46.24, $p < 0.05$) (**Fig 2**)
- Lower optic nerve edema grades bilaterally ($p < 0.05$) (**Fig 3**)
- Lower frequency of globe flattening on brain MRI (6 vs. 17 subjects, $p < 0.05$) (**Fig 4**)
- Lower required dosage of Acetazolamide for optic nerve edema resolution ($p < 0.05$)

Fig 1. Comparison of Age of Onset

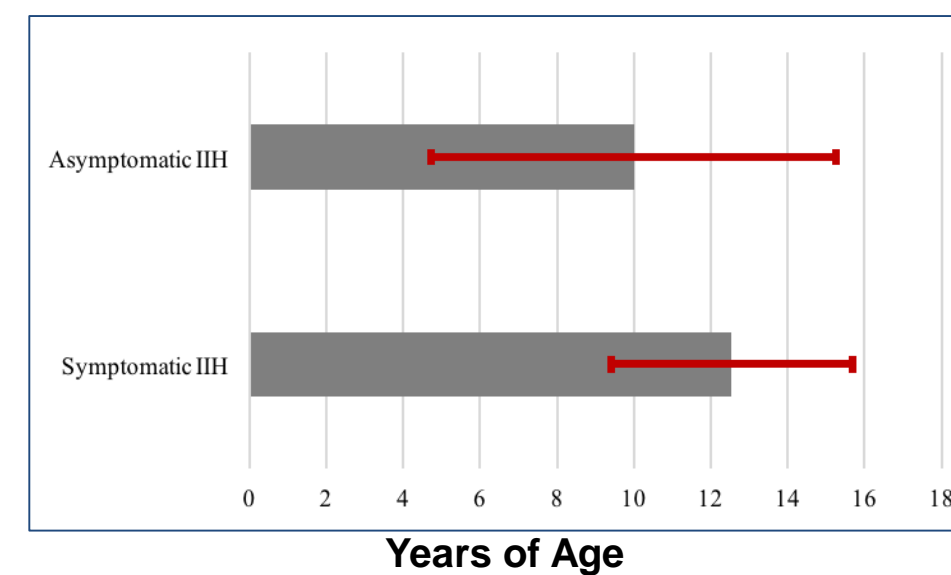


Fig 2. Comparison of Opening Pressure (cm H2O) at Initial Presentation

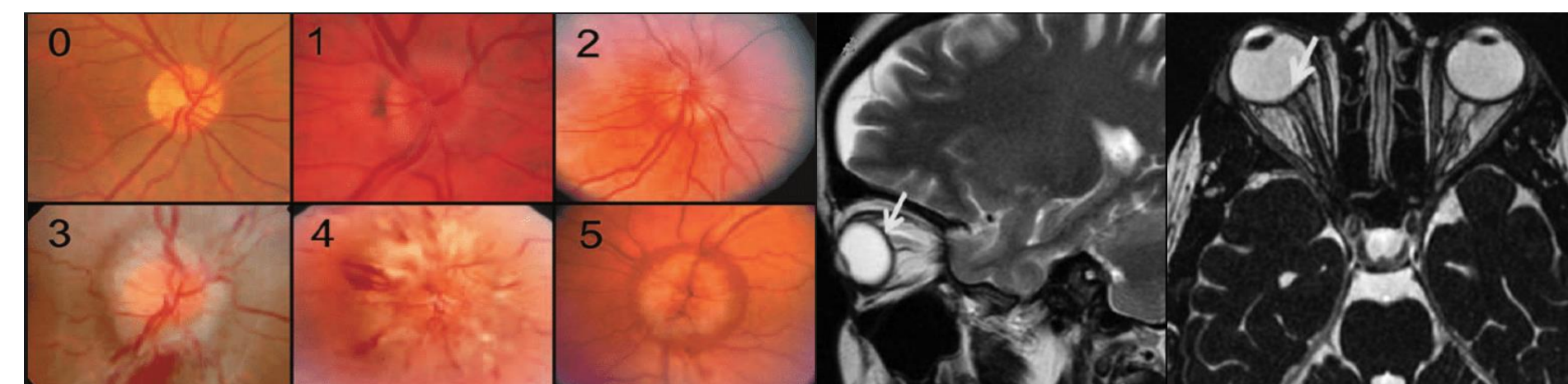
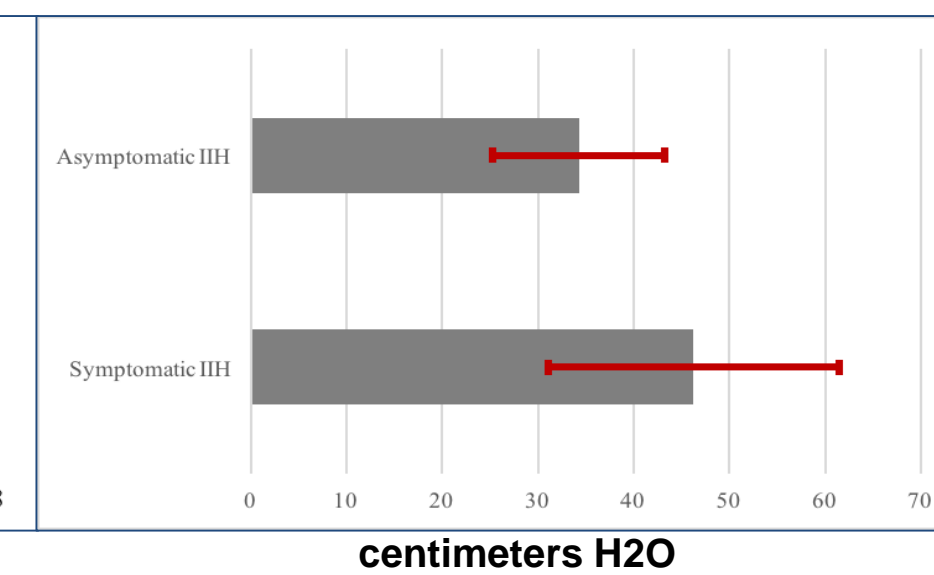


Fig 3. Frisen Optic Nerve Edema Grades

Optic nerve edema is indicative of elevated intracranial pressure. Fundus exam features include: venous engorgement, hemorrhages, blurring of optic margins

Fig 4. Orbital MRI Findings of Elevated ICP

Elevated intracranial pressure may cause compression on the orbital globe, resulting in the appearance of posterior flattening on orbital MRI

Conclusions

Asymptomatic IIH is common among pediatric patients with incidental papilledema, and may exist as:

1. A milder form of IIH that presents at a younger age, where ICP is not high enough to create symptoms or features of elevated ICP
2. A precursor, pre-symptomatic disease state that evolves into symptomatic IIH later in childhood

Possible explanations for asymptomatic disease exist:

- Nutritional or hormonal deficiencies/excess
- Anatomic variations (e.g. CSF leaks, number of arachnoid granulations)

Limitations:

1. Retrospective design with inconsistent follow-up intervals
2. Incidental diagnosis of asymptomatic disease
3. Small sample size due to rarity of IIH diagnosis

Future studies include:

1. Multi-center studies to increase sample size
2. Prospective designs to elucidate the natural disease course

Disclosure/References

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