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Incidence of Papilledema and Obesity in Children Diagnosed With Idiopathic "Benign" Intracranial Hypertension: Case Series and Review

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Abstract

Idiopathic intracranial hypertension is an important cause of headaches in the pediatric population and can lead to permanent blindness if not diagnosed in a timely manner. The aim of this study was to characterize the incidence of papilledema and obesity in children with idiopathic intracranial hypertension. We retrospectively analyzed 27 patients followed at The University of Texas Houston Pediatric Neurology Clinic. Papilledema was absent in 13 (48%) patients. The majority of our patients were nonobese (70%). Our results are contrary to the current medical practice of associating papilledema and obesity with idiopathic intracranial hypertension in childhood and highlight the importance of revised diagnostic criteria in this population needed to detect and manage this condition.

Keywords

idiopathic intracranial hypertension; pseudotumor cerebri; papilledema; obesity

Idiopathic intracranial hypertension, more commonly known as pseudotumor cerebri, is a well-known condition of unknown etiology. Idiopathic intracranial hypertension is defined by increased intracranial pressure in the absence of intracranial pathology with normal cerebrospinal fluid and no identifiable underlying systemic cause. Idiopathic intracranial hypertension is most commonly found in obese women of child-bearing age who present with chronic headache, papilledema, and visual impairment.[1,2] Current diagnosis of idiopathic intracranial hypertension is based on the hallmark physical finding of papilledema on ophthalmological examination with confirmation by means of elevated cerebrospinal fluid opening pressure on lumbar puncture. Persistently raised intracranial pressure can result in diplopia, visual field defects, and, if not treated, irreversible blindness.

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Recent publications suggest that pediatric patients with idiopathic intracranial hypertension have a clinical presentation that differs from adults.[3–6] Younger children are less likely to be obese[3,6–9] and a gender difference is not always identified. Descriptions of ophthalmological findings in children with idiopathic intracranial hypertension are lacking. The goal of this retrospective study was to determine the incidence of papilledema and obesity before the diagnosis of idiopathic intracranial hypertension in children.

Methods

This study is a retrospective review of 27 pediatric patients (ages 4–18 years) diagnosed with idiopathic intracranial hypertension at The University of Texas Houston Medical School and Children's Memorial Hermann Hospital in Houston, Texas, during the period of January 2007 through June 2009. Before chart review, idiopathic intracranial hypertension was defined as an opening cerebrospinal fluid pressure of ≥200 mm H_2O in nonobese and ≥250 mm H_2O in obese patients.[10,11] Subjects were excluded from the study if any known cause for elevated cerebrospinal fluid pressure was identified.

Height, weight, and presence of papilledema were recorded at, or just before, diagnosis of idiopathic intracranial hypertension. Body mass index for age and sex was defined by the Centers for Disease Control, and patients with a body mass index > 95% percent were categorized as obese. All others were classified as nonobese (Table 1). Visual fields were assessed by perimetry.

Subjects were selected for lumbar puncture if they had refractory headaches, as evidenced by failure of at least 2 trials of standard migraine prophylactic therapy, or had signs and symptoms suggesting increased intracranial pressure. All patients were analyzed by means of magnetic resonance brain imaging (MRI) or magnetic resonance brain imaging with venography (MRV) before undergoing lumbar puncture. As is standard at our institution, lumbar punctures were performed by board certified interventional neuro-radiologists. Patients were sedated using a long- or short-acting benzodiazepine or chloral hydrate and placed in a prone position to measure opening cerebrospinal fluid pressure.

Results

As shown in Table 1, 27 patients met the diagnostic criteria for idiopathic intracranial hypertension with female predominance (24/27). Age at presentation ranged from 4 to 18 years, with a mean age of 13 years; 20 females were over 10 years of age. Duration of symptoms before presentation ranged from 4 weeks to 5 years, with the majority of patients being diagnosed less than 1 year from symptom onset. Headaches were present in all patients. Additional symptoms included nausea, vomiting, blurred vision, diplopia, limb paresthesias, back or shoulder pain, fatigue, vertigo, syncope, or photophobia (Table 2).

A total of 70% of the patients were nonobese (body mass index < 95%) and 30% were obese (body mass index > 95%). Patients were further classified as being prepubescent or pubescent based on age (prepubescent: girls < 10 years, boys < 12 years; pubescent: girls > 10 years, boys > 12 years). Eighty-six percent of the prepubescent and 65% of the pubescent patients were nonobese.

MRI of the brain was performed in all 27 patients and venography in 8. Normal intracranial venous flow was found in all. The majority of patients (16/27) had normal brain imaging. One patient had an arachnoid cyst, 1 patient had a pineal cyst, and 9 patients presented with a history of Chiari 1 malformation who were later diagnosed with idiopathic intracranial hypertension.

Fundoscopy revealed papilledema in 13 patients bilaterally and unilaterally in 1; papilledema was absent in 13 patients. Optic disc abnormalities varied from blurring of the disc margins to gross papilledema with hemorrhages. One patient presented with complete visual loss.

Opening cerebrospinal fluid pressure on lumbar puncture ranged from 210 mm H_2O to 650 mm H_2O with a mean of 319 mm H_2O; cerebrospinal fluid content was normal in all patients. No correlations were identified between severity of symptoms, fundoscopic findings, opening pressure, and clinical course.

Twenty-five patients received pharmacological treatment. Acetazolamide was the first-line pharmacological agent followed rarely by ethacrynic acid. Lumboperitoneal shunts were placed in 2 patients due to refractory symptoms, and a single patient had a lumboperitoneal shunt placed at diagnosis. Serial therapeutic lumbar punctures were necessary in 3 patients.

Discussion

This retrospective study investigated 27 children (ages 4−18 years) diagnosed with idiopathic intracranial hypertension at our institution over a period of 2.5 years. The objective was to investigate the incidence of papilledema and/or obesity with childhood idiopathic intracranial hypertension.

Papilledema is the hallmark finding associated with idiopathic intracranial hypertension and is often considered a prerequisite for diagnosis. In our study, the incidence of papilledema was investigated in children diagnosed with idiopathic intracranial hypertension with an elevated opening cerebrospinal fluid pressure measured under controlled conditions. Although there have been case descriptions where papilledema is absent in idiopathic intracranial hypertension,[12–15] this study confirms that almost half of pediatric patients (13/27) do not have papilledema at presentation. The implications of this finding suggest that revised clinical criteria are necessary for diagnosis of idiopathic intracranial hypertension in a pediatric population with reduced emphasis on papilledema. Clinicians should not be reluctant to perform a lumbar puncture for assessment of opening pressure if clinical suspicion of idiopathic intracranial hypertension is present, even in the absence of papilledema.

In adults, obesity is closely related to idiopathic intracranial hypertension, but this correlation does not necessarily apply to children.[7–9] It has been well documented in the literature that children with idiopathic intracranial hypertension are not typically obese and our findings confirm this. Of the patients in our study, 70% were nonobese and 30% were obese. Based on physical maturity, 86% of the prepubescent and 65% of pubescent children were nonobese.

Conclusions

Increased intracranial hypertension has long been associated with obesity and papilledema. Our results indicate that neither papilledema nor obesity is as common in children as adults. We showed that half of our idiopathic intracranial hypertension pediatric patients did not have clinical signs of papilledema at the time of diagnosis. The absence of papilledema should not exclude the consideration of idiopathic intracranial hypertension. Our findings raise concern that many cases of idiopathic intracranial hypertension can be overlooked in childhood because of the association described between the presence of papilledema and idiopathic intracranial hypertension in the adult population.

Previous reports have shown that obesity is not as common in children as it is in adults diagnosed with idiopathic intracranial hypertension; our study confirms this finding, showing that most pediatric idiopathic intracranial hypertension patients were nonobese (70%) at the time of diagnosis. Further studies incorporating larger patient populations are required to delineate the mechanisms of idiopathic intracranial hypertension in the pediatric population, including the interesting association between Chiari 1 malformations and idiopathic intracranial hypertension.

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Table 1

Patient Characteristics

Age (years)	Gender	Papilledema	HA Onset (years)	Opening Pressure (mm H_2O)	Body Mass Index	Weight Category[a]
4	F	+	3	210	16	Healthy
6	F	+	6	260	17	Healthy
7	F	+	6	320	17	Healthy
8	F	+	8	320	23	Obese
8	F	+	7	240	18	Healthy
10	F	+	9	240	17	Healthy
12	F	+	11	310	23	Overweight
12	F	−	8	460	31	Obese
12	F	+	11	270	18	Healthy
13	F	+	12	380	28	Obese
13	F	−	12	300	17	Healthy
13	F	−	10	240	28	Obese
14	F	−	12	330	18	Healthy
14	M	−	12	270	27	Obese
15	M	−	12	245	19	Healthy
15	F	−	14	340	41	Obese
16	M	+	15	280	22	Healthy
16	F	−	5	400	15	Underweight
16	F	−	10	280	17	Healthy
16	F	−	14	230	25	Overweight
16	F	+	16	650	35	Obese
16	F	−	16	470	20	Healthy
16	F	−	15	280	27	Overweight
17	F	+	16	490	24	Healthy
17	F	+	12	250	27	Overweight
18	F	−	16	270	26	Overweight
18	F	+	14	280	31	Obese

[a] Age-adjusted weight category based on data obtained from the Centers for Disease Control (obese body mass index > 95%; overweight body mass index 85%–95%; healthy body mass index 5%–85%; underweight body mass index < 5%). HA, headache.

Table 2

Symptoms at Time of Presentation

Symptom	Number of Patients (N = 27)	Percentage of Patients
Headache	27	100
Headache only	5	19
Nausea and/or vomiting	5	19
Blurred vision	3	11
Diplopia	3	11
Limb paresthesias	3	11
Back pain	2	7
Fatigue	2	7
Shoulder pain	2	7
Vertigo	2	7
Blackout	1	4
Photophobia	1	4