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Original Article

Incidence and Demographics of Pediatric Intracranial Hypertension

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ABSTRACT

OBJECTIVE: We analyzed the incidence and demographic features of pediatric intracranial hypertension. **METHODS:** Inpatient and outpatient encounters of children aged 18 years or younger who were diagnosed with primary (idiopathic) or secondary intracranial hypertension between January 2010 and December 2013 were identified. Data were collected from a subspecialty clinic devoted to intracranial hypertension and the sole children's hospital in a large Midwestern city of the United States. Estimated incidence rates were calculated based on the number of newly diagnosed patients in our hospital's primary service area, which includes seven central Ohio counties. Sex, race, body mass index, socioeconomic status, and geographic distribution were also noted. **RESULTS:** A total of 74 pediatric patients were diagnosed with intracranial hypertension (49 primary/idiopathic and 25 secondary) between January 2010 and December 2013. Using census data, we determined the pediatric population in our service area during the four-year period. The Ohio Hospital Association's database indicated that 92.3% of patients aged 0 to 17 years residing in the region sought care at our institution. By combining these data, we calculated an annual incidence of primary and secondary intracranial hypertension of 0.63 and 0.32 per 100,000 children, respectively. **CONCLUSIONS:** The estimated annual incidence of pediatric primary intracranial hypertension in our seven county service area in central Ohio is similar to previous pediatric reports from other countries and is 67% of that reported in the US adult population.

Keywords: pseudotumor cerebri, intracranial hypertension, pediatric, headache, incidence

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Introduction

Primary/idiopathic intracranial hypertension (PIH) is traditionally viewed as a disorder affecting obese women of childbearing age.[1] However, PIH also occurs in the very young, the very thin, and in the absence of headache or papilledema.[2-6] The reported annual incidence in adults in US population-based studies has varied from 0.9 per 100,000 to 1.07 per 100,000 in Iowa and Louisiana, respectively.[7] Internationally, annual incidence rates have been reported as 2.2 per 100,000 persons in Libya and, more recently, 2.36 per 100,000 persons in Northern Ireland.[8,9] In contrast, there is little information about the incidence of intracranial hypertension in children in the United States. Similarly, there is limited information about nonweight-based demographic features of pediatric intracranial hypertension such as socioeconomic status (parental education level, household income, and degree of poverty) and geographic location.

Nationwide Children's Hospital is a tertiary care facility in central Ohio, and its physicians provide most of the pediatric neurological and ophthalmologic care in the region. In 2010, we established the first pediatric multidisciplinary

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intracranial hypertension clinic in the country to manage children with PIH or secondary intracranial hypertension (SIH). The clinic was developed to facilitate joint management of patients with this condition. The raised awareness of intracranial hypertension among providers across all disciplines in our area has facilitated timely referral of children with suspected intracranial hypertension. The team consists of a dedicated nurse, two neurologists, a headache specialist, and four ophthalmologists; nutrition, psychology, and neurosurgery services are available when needed.

The aims of this study were to estimate the incidence of pediatric intracranial hypertension in our Midwestern area and to expand our understanding of the demographic characteristics of pediatric intracranial hypertension in central Ohio.

Methods

We identified patients aged 18 years or younger who were diagnosed with intracranial hypertension at Nationwide Children's Hospital from January 2010 through December 2013. The study was approved by Nationwide Children's Hospital Institutional Review board.

Both inpatient and outpatient encounters were searched for visit diagnosis of intracranial hypertension using ICD-9 codes of 348.2 (idiopathic intracranial hypertension), 377 (papilledema), and 781.99 (other symptoms involving nervous and musculoskeletal systems/intracranial hypertension). All patients included were newly diagnosed, followed in our multidisciplinary intracranial hypertension clinic, and resided in the hospital's primary service area (encompasses 40 different postal zip codes in seven counties). Each chart was reviewed to confirm that the patient met the age-based criteria derived from the modified Dandy criteria by one of our primary investigators (N.G. or S.C.A.) (Table 1).[5,10,11] All subjects were examined by an ophthalmologist at diagnosis and followed during treatment with formal examination including visual field testing in children who could participate.

Patients with an identifiable cause of intracranial hypertension were classified as SIH. Those who lacked an identifiable cause were classified as PIH. Exclusion criteria were (1) incomplete records (lack of testing or followed by other physicians); (2) absence of complete ophthalmologic examination; (3) diagnosis before 2010; (4) diagnosis at an outside institution, (5) age greater than 18 years at the time of diagnosis; and (6) no signs or symptoms consistent with intracranial hypertension.

Lumbar puncture procedures were performed according to accepted convention and most were performed under sedation. All patients were in the lateral decubitus position with head and legs extended during pressure measurement. Sedated outpatient lumbar punctures used nitrous oxide and propofol for induction and sevoflurane for maintenance. Emergency department procedures used ketamine.

TABLE 1.
Criteria for Primary Intracranial Hypertension

1. Signs and symptoms of raised intracranial pressure (headache, nausea, vomiting, transient visual obscurations, pulsatile tinnitus, or papilledema)
2. No focal neurological signs (with the exception of cranial nerve IV or VI palsy)
3. Normal cerebrospinal fluid composition
4. Cerebrospinal fluid opening pressure of >18 cm H_2O in children <8 years or >25 cm H_2O in children >8 or <8 years without optic disc edema
5. Normal to small ventricles as demonstrated by neuroimaging

The 2000 Centers for Disease Control and Prevention growth charts for the United States were used to calculate the age-based body mass index (BMI) percentile at the time of diagnosis for each patient.[12] Pediatric population estimates for each county from 2010 to 2013 were obtained from the Ohio Hospital Association's database.[13] The frequency of intracranial hypertension was calculated as the number of patients with intracranial hypertension diagnosed each year divided by the reported pediatric population in each respective county for that year. The frequencies of PIH and SIH were calculated in a similar manner for all counties combined.[14] The annual frequency was calculated from the average over the four year period. The frequency of children with PIH who presented with papilledema was calculated in a similar fashion.

For each county, information regarding income, poverty percentage, highest level of parental education, race, and sex is based upon 2010 US population census bureau statistics (Table 2). Demographic data gathered for each patient included age at the time of diagnosis, height, weight, gender, and race, which are readily available in our electronic system. Age was used to determine age-based cutoff for the opening pressure (OP). Gender was examined as there have been differences in female to male ratios depending on pubertal status.[6,15] Race was recorded because obesity rates are lower in African American adult patients.[16]

Results

We identified 235 unique encounters for children matching the ICD-9 codes who resided within the institution's primary service area. Seventeen subjects with the ICD-9 code 781.99 were excluded because of diagnoses other than intracranial hypertension. Another 144 subjects were excluded because of diagnosis before 2010, incomplete records, no signs or symptoms of intracranial hypertension, or diagnosis at another institution. Two patients were excluded for age greater than 18 years. The remaining 74 patients were used in our analysis. The indications for initiating diagnostic evaluation were at the discretion of the provider and consultant managing the patient (Table 3).

Forty-nine subjects were classified as PIH, and the remaining 25 were characterized as having SIH. Of the individuals with SIH, six had cerebral sinus venous thrombosis, five had cancer (three with acute lymphoid leukemia, one with non-Hodgkin's lymphoma, and one with B cell lymphoma), two were on tetracycline antibiotics, four had a central nervous system viral infection, four were undergoing growth hormone replacement therapy, two had Muckle-Wells syndrome, one had an intracranial hemorrhage, and one subject had two systemic inflammatory conditions (membranoproliferative glomerulonephritis and Crohn disease). Patients with Muckle-Wells syndrome and systemic inflammatory conditions were treated as secondary because of literature reports of the same or similar conditions resulting in intracranial hypertension.[17,18] We also anecdotally noted that, after stabilization of their intracranial hypertension, some patients experienced exacerbation of symptoms and sometimes of their papilledema during flares of their underlying condition and corresponding improvement with adjustments to the therapeutic regimen for their underlying condition.

The frequency of pediatric PIH was 2.42 per 100,000 children across the four year period, the aggregated county-specific incidences are displayed in Table 2. We found the annual frequency of PIH to be 0.63 per 100,000 children. There were 1.90 per 100,000 PIH children (37 of 49) who presented with papilledema over the four year period, equating to an annual frequency of 0.48 per 100,000