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Clinical Observations

Use of Ocular Coherence Tomography in Children With Idiopathic Intracranial Hypertension—A Single-Center Experience

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ABSTRACT

BACKGROUND: Idiopathic intracranial hypertension is a disorder characterized by elevated intracranial pressure without an identifiable etiology. Detection of papilledema may be challenging and leads to diagnostic uncertainty in evaluating a child for possible idiopathic intracranial hypertension. Ocular coherence tomography has the potential to add accuracy to the diagnosis of idiopathic intracranial hypertension. The goal of the present study was to determine if there was a relationship between severity of papilledema (as determined by direct ophthalmoscopy and confirmed by fundus photography) and measures of ocular coherence tomography in a pediatric population with idiopathic intracranial hypertension. **METHODS:** Thirteen pediatric patients were recruited prospectively after diagnosis of either definitive idiopathic intracranial hypertension (with papilledema) or possible idiopathic intracranial hypertension (without papilledema) at Children's Hospital of Michigan over a period of one year. Clinical data and results of initial ocular coherence tomography and visual field testing were collected and statistically analyzed. **RESULTS:** The Frisén scale of papilledema significantly correlated with average retinal nerve fiber layer thickness of each eye ($r = 0.633$, $P = 0.02$ in right eye and $r = 0.868$, $P = 0.001$ in left eye). The retinal nerve fiber layer thickness (mean ± SD) was significantly higher in the definitive group than in the possible group (189 ± 65 μm vs 104 ± 10 μm in right eye, 165 ± 42 μm vs 106 ± 9 μm in left eye, $P < 0.01$ in both eyes). **CONCLUSIONS:** Ocular coherence tomography may be used as a supplementary method to aid in the reliable detection of papilledema in evaluating a child for idiopathic intracranial hypertension.

Keywords: idiopathic intracranial hypertension, ocular coherence tomography, fundus, papilledema

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Idiopathic intracranial hypertension (IIH) or primary intracranial pressure is characterized by elevated intracranial pressure in the setting of normal cerebrospinal fluid (CSF) composition, without ventriculomegaly or evidence of a mass lesion on imaging.[1-4] Application of diagnostic principles, such as the modified Dandy criteria and most recently the criteria proposed by Friedman et al., assist in making the diagnosis with unambiguity in most cases.[2,3] The publication of a study, albeit in the form of a letter, outlining normal values for opening pressures following lumbar punctures in children sought to shed further light on this matter.[4] However, an aspect of IIH that defies objectivity is reliable detection of papilledema, which constitutes a crucial consideration in establishing the diagnosis.

Examination of the ocular fundus is one facet of the neurological examination that many emergency department physicians, pediatricians, and residents in training are

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unfamiliar with and is consequently ignored or poorly performed.[5] Furthermore, there are limitations to the direct fundus examination, especially when performed in nonmydriatic pupils,[6] leading to diagnostic uncertainty in many clinical scenarios, and is probably best exemplified in the setting of evaluating a child for possible IIH. The introduction of ocular coherence tomography (OCT) has the potential to add accuracy and reduce interobserver variability to this step of the diagnostic algorithm.

The goal of the present study was to determine if there was a relationship between papilledema (as determined by direct ophthalmoscopy and confirmed by fundus photography) and measures of OCT in a pediatric population with IIH, which for reasons of clarity will be referred to as primary intracranial pressure in this article.

Materials and Methods

Subjects

Children who had never been diagnosed with primary intracranial pressure were recruited prospectively over a period between March 2014 and February 2015 from the clinics and inpatient units at Children's Hospital of Michigan. Subjects who fulfilled the criteria set forth by Friedman et al.[2] for IIH were classified as definitive cases, i.e., group 1 (all children had definitive papilledema per the examining neurologist). Presence of papilledema, with or without abducens nerve palsy, constitutes an essential feature of primary intracranial pressure per the guidelines of this group. Those who had questionable or no papilledema and no abducens nerve palsy but had elevated opening pressure following a lumbar puncture and had imaging evidence suggestive of IIH were labeled as possible cases, i.e., group 2. None of the subjects in group 2 were found to have papilledema per subsequent examination by a neuro-ophthalmologist. There were no children in our cohort who lacked papilledema but had abducens nerve palsy and fulfilled other diagnostic criteria for IIH. Potential causes of intracranial hypertension ("secondary" intracranial pressure) such as use of implicating medications, endocrine disorders, hypercoagulable states, anemia, mastoid infections, and previous disease states that might lead to scarring of arachnoid villi were ruled out by appropriate investigations and by obtaining relevant historical data.

Lumbar punctures were performed with a 20-gauge needle, in the right lateral decubitus position, with both legs extended and under general anesthesia using sevoflurane. The opening CSF pressure was measured in all patients using a column manometer, and fluid was sent for microbiological, biochemical, and cellular assays. Opening pressures equal to or greater than 28 cm H_2O were considered abnormal. Magnetic resonance imaging and magnetic resonance venography studies of the brain were obtained in all subjects, and interpretation was performed by a pediatric neuroradiologist who was blinded to the classification into the two groups.

All subjects were examined by neurologists (YAL, LS) and subsequently referred to the Kresge Eye Institute for specialized ophthalmologic examination after undergoing diagnostic lumbar punctures. Ophthalmologic examination included visual acuity (using Snellen visual acuity chart), color vision testing (Ishihara color plates), automated or manual perimetry, fundus photography, and peripapillary mydriatic OCT. The grading of papilledema was performed per the Frisén scale (Table 1) utilizing fundus photography.[7]

Visual field testing

Standard automated perimetry was performed using Humphery 750i visual field analyzer (Carl Zeiss Meditec, Inc, 24-2 SITA fast, Germany) or Goldman perimeter using the Octopus 900 (Haag Streit Diagnostics, Switzerland). Subjects performed a 1-minute trial before the test was initiated, to acclimatize them to the procedure. Criteria for reliable results were established as <33% fixation losses, <33% false negatives, and <33% false positives. Visual field index, mean deviation, and pattern standard deviation were recorded by trained personnel.

TABLE 1.
Frisén Classification of Papilledema

Frisén Grade	Description
0	Normal optic disc
1	Blurring of the nasal border, subtle grey halo with temporal gap
2	Circumferential halo without major vessel obscuration
3	Increased diameter of optic nerve head, obscuring one or more major blood vessels at disc margin
4	Elevation of whole optic nerve head including the cup, obscuration of major vessels on the disc
5	Protrusion of optic nerve head as a dome

Ocular coherence tomography

Two-dimensional (2D) spectral domain OCT was performed with the Cirrus HD-OCT (Model 4000, Software ver. 4.0, Carl Zeiss Meditec, Inc) on each eye of participants. Spectral domain OCT is a form of noninvasive, low-coherence spectrometer that produces high-resolution cross-sectional images of the retina without touching the eye. OCT is similar to ultrasonography but uses reflection of light waves that are directed at the retina to create a pattern thereafter based on the optical interface.

The protocol used for retinal nerve fiber layer (RNFL) thickness assessment was the optic disc cube protocol in which a 3.46-mm circular scan was placed around the optic disc to obtain peripapillary RNFL thickness data. Parameters automatically calculated by the Cirrus software and evaluated in this study included average and/or full circle thickness (360° measure), temporal quadrant thickness, superior quadrant thickness, nasal quadrant thickness, inferior quadrant thickness, disc area, rim area, and cup volume of each eye. The macular cube protocol was used to assess the thickness of the ganglion cell and/or internal plexiform layer of each eye. To ensure reliability each eye was measured twice consecutively. The best peripapillary scan was individually selected for each patient by the neuro-ophthalmologist.

Fundus photography

Optic disc photographs centered on the optic disc and focused on the retinal plane of highest disc elevation were obtained.

Approval from Wayne State University School of Medicine Institutional Review Board was obtained before start of the study.

Statistical analyses

All statistical techniques were analyzed and summarized using SPSS Version 22.0. Continuous variables were reported as means and S.D., whereas categorical variables were reported as counts and percentages. In skewed samples, the median and the range were employed. Student *t* test was used for comparison of data with equal variance, and Mann-Whitney *U* test was used for data with unequal variance. Paired-samples *t* test was used for comparison of means between two groups on the same continuous, dependent variable. χ^2 Test was used to examine differences with categorical variables. Correlations between continuous variables were evaluated using a nonparametric Spearman rank correlation coefficient. Differences were considered statistically significant when probability values were 0.05 or less.