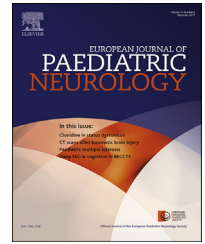




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Original article

Pediatric idiopathic intracranial hypertension — Is the fixed threshold value of elevated LP opening pressure set too high?

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ABSTRACT

Background: Idiopathic intracranial hypertension (IIH) in children is a rare condition of unknown etiology and various clinical presentations. The primary aim of this study was to evaluate if our pediatric IIH study group fulfilled the revised diagnostic criteria for IIH published in 2013, particularly with regard to clinical presentation and threshold value of an elevated lumbar puncture opening pressure. Additionally we investigated the potential utilization of MR-based and fundoscopic methods of estimating intracranial pressure for improved diagnosis.

Patients and methods: Clinical data were collected retrospectively from twelve pediatric patients diagnosed with IIH between 2008 and 2012 and revised diagnostic criteria were applied. Comparison with non-invasive methods for measuring intracranial pressure, MRI-based measurement (MR-ICP) and venous ophthalmodynamometry was performed.

Results: Only four of the twelve children (33%) fulfilled the revised diagnostic criteria for a definite diagnosis of IIH. Regarding noninvasive methods, MR-ICP ($n = 6$) showed a significantly higher mean of intracranial pressure compared to a healthy age- and sex-matched control group ($p = 0.0043$). Venous ophthalmodynamometry ($n = 4$) showed comparable results to invasive lumbar puncture.

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Conclusion: The revised diagnostic criteria for IIH may be too strict especially in children without papilledema. MR-ICP and venous ophthalmodynamometry are promising complementary procedures for monitoring disease progression and response to treatment.

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List of abbreviations

BMI	Body Mass index
CSF	cerebrospinal fluid
ICP	intracranial pressure
IIH	idiopathic intracranial hypertension
LP	lumbar puncture
MRI	magnetic resonance imaging
MR-ICP	MRI-based measurement of intracranial pressure
PTC	pseudotumor cerebri
PTCS	pseudotumor cerebri syndrome
VOP	venous outflow pressure
VP-shunt	ventriculo-peritoneal shunt

1. Background

Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri (PTC) is a rare condition in childhood with unknown etiology.^{1,2} Most PTC patients suffer from headache, papilledema, double vision, impairment of visual acuity, visual field defects and nausea. IIH in childhood may be less specific, especially in younger children: apathy and irritability may be the only presenting symptoms.³ Sometimes an incidental finding of papilledema in a routine ophthalmological examination leads to the diagnosis in a symptom-free child.

Pseudotumor cerebri may be primary (idiopathic intracranial hypertension – IIH) or secondary due to infections, metabolic diseases or medication.⁴ Diagnostic criteria for IIH include a normal neurological examination (except palsy of the sixth cranial nerve or visual field limitations), the presence of papilledema, an elevated lumbar CSF pressure but normal CSF composition, the absence of mass lesion, hydrocephalus and cerebral or sinus vein thrombosis on MRI and the absence of any underlying disease.⁵ Some MRI signs like empty sella or a distension of the perioptic subarachnoid space are suggestive of IIH.⁴

Avery et al. suggested a new threshold for an abnormally elevated LP opening pressure of 28 cmH₂O.⁶ This threshold is part of the recently published revised diagnostic criteria for the PTC syndrome in children (Table 1).⁴ These revised diagnostic criteria also include the diagnostic approach for IIH without papilledema.

Therapy of IIH is essentially important as damage of the optic nerve needs to be avoided. There are no pediatric treatment guidelines with a level of evidence I or II, so the

adult recommendations are usually adhered to. Treatment options include medications as first line treatment (especially acetazolamide), repeated lumbar punctures and, in case of severe papilledema and concern about an irreversible damage of the optic nerve, neurosurgical methods.^{1,3,5,7}

The primary aim of this study was to evaluate the applicability of the revised diagnostic criteria for IIH in pediatric patients diagnosed with IIH during 2008 and 2012 in a university children's hospital. Additionally, two noninvasive methods of measuring intracranial pressure, an MR-based method and venous ophthalmodynamometry, were used to obtain estimates of ICP in addition to the opening pressure in a subgroup of patients. The potential diagnostic efficacy of these methods was evaluated.

2. Methods

2.1. Study group

In this retrospective study design data was obtained for patients and control groups, patient records in the electronic file systems were searched for diagnosis of primary PTC (IIH). Fifteen pediatric patients were diagnosed with primary PTC (IIH) at the Hauner Children's Hospital of the Ludwig-Maximilian University of Munich between 2008 and 2012. Inclusion criteria were a diagnosis of IIH in the chart, absence of any other underlying conditions and an age below 18 years. Twelve subjects between 5 and 17 years of age (7 female) were included in the study cohort (see Table 2). The diagnosis of IIH in our patient cohort was based on clinical signs of elevated intracranial pressure (e.g. papilledema, visual symptoms, headache, dizziness, nausea), LP opening pressure >25 cmH₂O without cytological CSF abnormalities, absence of mass lesions, hydrocephalus or cerebral sinus vein thrombosis on MRI and no findings for a secondary cause of PTC. In two patients (see Table 2, No 4 and 11) LP opening pressure was <25 cmH₂O, but diagnosis of IIH based – beside normal MRI findings and no CSF abnormalities – on clinical signs of headache (no matching another headache type according to the international classification of headache disorder ICHD III-Beta) plus papilledema and clear improvement after draining cerebrospinal fluid. Three patients were not included due to a documented LP opening pressure <20 cmH₂O and a lack of papilledema in two of them, so diagnosis of IIH in these three patients was inconclusive (see Table 2).

2.2. Control group

A control group consisted of eight healthy volunteers, matched for age and sex, who underwent non-invasive MRI-

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