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Idiopathic intracranial hypertension in pediatric and adolescent patients

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Abstract

Objectives: Pediatric idiopathic intracranial hypertension is a rare condition, but inappropriate diagnosis and management may lead to devastating outcome with loss of vision and lifelong handicap. Dandy criteria are used for diagnosis of idiopathic intracranial hypertension (IIH) in adult, but these criteria cannot be applied in all pediatric patients.
The aim of this study is to evaluate the diagnostic criteria and outcome of management of IIH in pediatric patients.

Methods: Nineteen patients with IIH and age less than 16 years old have been evaluated for the diagnostic criteria and outcome of management. The patients were classified according to the secondary sexual criteria into adolescent IIH and pediatric IIH. Full neurological and ophthalmological evaluation was completed in all patients. The patients were managed and followed up for a mean period of 12 ± 8.6 months at Zagazig University hospitals or Kingdom Hospital in the time period from 2009 to 2018.

Results: Nineteen patients (16 females and 3 males) had been diagnosed and treated with idiopathic intracranial hypertension, their age is between 4 and 15 years, and the patients have been divided into two groups. Group I (pediatric IIH) involved 9 patients (6 females and 3 males), and their age is ranging between 4 and 9 years with mean age 5.56 ± 1.9 years while group II (adolescent IIH) involved 10 patients, all of them are females and their age ranges between 12 and 15 years with mean age 13.5 ± 1.3. Diagnostic criteria of the patients are papilledema, symptoms and signs of intracranial hypertension, and elevated CSF opening pressure with normal MRI. Headache, delayed school performance, and sixth nerve palsy are the most common clinical finding in the patients of group I while headache is the most common presenting symptom in group II. Anemia and otitis media are the most common associated risk factors in group I while obesity and female gender were the most common associated risk factors in group II. The minimum value for opening pressure in group I is 180 mmH_2O while the minimum value in group II is 250 mm. 16 patients improved after the first lumbar puncture and drainage of CSF; two patients improved after repeated lumbar puncture while lumbo-peritoneal shunt was inserted for two patients, both of them were in group II. Recurrence was diagnosed in four patients (21%), one of them (11.1%) was in group I while three of them (30%) were in group II.

Conclusion: Pediatric idiopathic intracranial hypertension can be classified into two subtypes: pediatric type and adolescent type according to the secondary sexual criteria.
Modified Dandy criteria can be applied for the diagnosis of adolescent type, but pediatric type is not associated with obesity, has no female predominance, and usually responds to the initial lumbar puncture with a low rate of recurrence.

Keywords: Idiopathic intracranial hypertension, Papilledema, Lumbo-peritoneal shunt

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Introduction

Idiopathic intracranial hypertension (IIH) is a vision-threatening disorder [1, 2] that can be presented at any age but usually affects obese women during the child-bearing period [3–5]. Pediatric IIH has been reported long time ago [6–9] but it was considered a relatively rare condition [10] until 1992 when Lessell [1] refined the disorder in pediatric patients and later on an epidemiological study showed that the annual incidence of symptomatic IIH in pediatric age from 3 to 15 years is 0.9 per 100,000 [11].

The clinical presentation of IIH is variable and the diagnostic criteria have changed over time, but currently, Dandy criteria and its modifications which involve symptoms and signs of increased intracranial pressure with elevated CSF pressure without laboratory or radiological evidence of intracranial lesion, infection, or hydrocephalus are used for diagnosis of IIH in adult patients [2, 4, 5, 12]; however, there is no agreement for specific diagnostic criteria in pediatric IIH due to controversies about the diagnostic value of CSF opening pressure [13–16], in addition to the difference in the presentation and clinical finding between children and adolescent patients [2, 16].

Lumbar puncture and CSF drainage, carbonic anhydrase inhibitors, corticosteroid, and topiramate are usually used for treatment of IIH while CSF diversion procedures and optic nerve sheath fenestrations are used after failure of medical treatment or in fulminant cases with significant optic neuropathy at presentation or in cases with progressive visual deterioration during the follow-up [4, 5]; recently, venous sinus stenting is used for cases with transverse sinus stenosis, but the role of neurovascular stenting is not yet established [5, 17].

In this study, we are looking forward to evaluate the diagnostic criteria and outcome of management of IIH in pediatrics and adolescent patients.

Materials and methods

All cases with IIH who were diagnosed and treated at the Department of Neurosurgery, Zagazig University or Kingdom Hospital between 2009 and 2018 with age less than 16 years old were included.

The diagnostic criteria are symptoms and signs of increase intracranial pressure (ICP), normal brain magnetic resonance image (MRI) without a space-occupying lesion or hydrocephalus, and normal CSF analysis in addition to papilledema with CSF opening pressure above 180 mmH_2O, while in cases without confirmed papilledema, IIH was considered only if the CSF opening pressure was above 250 mmH_2O.

The patients were divided according to the presence of secondary sexual criteria into two groups.

Group I (pediatric type IIH) which involved 9 patients without secondary sexual criteria and group II (adolescent type IIH) which involved 10 patients with one or more secondary sexual criteria.

All patients were subjected to standardized management protocol including full comprehensive history taking through analysis of the symptoms and general medical, neurological, and ophthalmological examination. Radiological assessment included magnetic resonance imaging (MRI) and magnetic resonance venography (MRV).

Lumbar puncture was carried out for all patients in the lateral decubitus position under complete aseptic condition. The opening pressure was measured, and CSF sample was taken for chemical analysis (protein level, glucose level, and cell count).

Laboratory investigations involved CSF analysis, complete blood picture, liver function test, kidney function test, serum electrolyte, and thyroid-stimulating hormone (TSH) to exclude secondary causes of IIH as CSF infection, adrenocortical dysfunction, or hypothyroidism, while for other investigations, complete blood picture was used to diagnose risk factors as systemic infection or anemia.

Initial lumbar puncture and CSF drainage, carbonic anhydrase inhibitors, and corticosteroid were used in all patients for 2 weeks, and lumbar puncture have been repeated 3 days and 1 week for those patients without improvement of the symptoms while CSF diversion was considered for patients with resistant symptoms after three lumbar punctures or for those patients who developed visual deterioration.

Recurrence and follow-up

Weekly strict ophthalmological follow-up was mandatory for the initial 1 month until complete recovery of the optic disc from papilledema then every one month for 3 months then every 3 months in addition to family education and instructions for medical advice if there were any relapsing symptoms. Recurrence was considered when the symptoms and signs relapsed after complete recovery of papilledema and disappearance of the symptoms while early relapse of symptoms after the initial lumbar puncture without complete recovery was considered a resistant case needed repeated lumbar puncture or even CSF diversion procedures. The recurrence was diagnosed after MRI excluded any new intracranial findings while papilledema was confirmed by fundus photography, and elevated CSF opening pressure was confirmed by lumbar puncture.

Statistical analysis

The statistical analysis was performed using Stata/IC 15.1 software. The data were described using means, standard deviation, and proportions while a chi-square

test and two-sample *t*-test were used to compare the risk factors and clinical findings between groups.

Results

Nineteen patients (16 females and 3 males) with age ranging between 4 and 15 years were evaluated. The demographic data and clinical findings in both groups are listed in Table 1.

The most common symptom is headache, but the sixth nerve palsy is common in the patients of group I. Other manifestations of increased ICP in pediatrics as deterioration of the school performance, irritability, and irregular sleep were detected in most of the patients which may be due to both headache and visual abnormalities.

The mean opening pressure is 291 ± 79.5 mmH_2O, and the minimum value for opening pressure in group I is 180 mmH_2O while the minimum value of opening pressure in group II is 250 mmH_2O (Table 1).

Anemia and history of otitis media with or without upper respiratory tract infection are the most common associated disorders in group I while obesity and female predominance are common associated risk factors in group II (Table 2).

16 patients improved after the first lumbar puncture and drainage of CSF. All patients in group I improved after the first lumbar puncture while seven patients out of the ten patients improved after the first lumbar puncture, two of them improved after the second lumbar puncture, and one patient did not improve even after the third lumbar puncture and CSF diversion was created. Lumbo-peritoneal shunt was inserted for two patients in group II, one of them due to prolonged symptom and failure of repeated lumbar puncture and the second one was inserted due to recurrence with visual field affection.

Recurrence was confirmed in 4 patients (21%), one of them (11.1%) in group I while the other three cases (30%) in group II. The mean time for recurrence is 11.25 ± 3.78 months. A recurrent case in group I improved after lumbar puncture; in group II, two patients improved after lumbar puncture and medical treatment while the third one was treated by lumbo-peritoneal shunt due to deterioration of the visual functions (Table 1). During the follow-up, all recurrent cases were improved after the initial lumbar puncture and medical treatment and there was no recorded recurrence after lumbo-peritoneal shunt.

Discussion

Though IIH in children have been reported long time ago [1, 2, 9, 18], there is no specific diagnostic criteria and the currently used modified Dandy criteria [12] for diagnosis of adult cases cannot be applied for most of the pediatric patients as children are not able to articulate their symptoms effectively in addition to the controversies about the diagnostic value of CSF opening pressure and in addition to the presence of various conventions applied in subdividing the pediatric population by age.

In this study, we reviewed 19 patients with IIH and their age is less than 16 years old at the time of diagnosis; typical patients with IIH in adults are female obese patients during the child-bearing period [3–5, 17] while in pediatric IIH obesity and female predominance are not a common finding [6–8], but in this study, there is an overall female predominance (84.2%) with more significant female predominance in group II (Table 1) and this female predominance in pediatric IIH was also observed in a study of Mosquera Gorostidi et al. [15] and in the study of Jirásková and Rozsíval [9]. Multiple potential risks for the development of IIH in adult as female sex and obesity were reported [4, 15, 19], while in pediatric patients, infection, malnutrition, and pharmacological causes may be significant risk factors [2, 15], and in this study, anemia and infection which involved pharyngitis, otitis media, and

Table 1 Epidemiological and clinical findings

	Total	Group I	Group II	*P* value
Age (in years)	9.7 ± 4.4 (4–15)	5.56 ± 1.9 (4–9)	13.5 ± 1.3 (12–15)	< 0.001
Gender (F/M)	16/19 (84.2%)	6 /9 (66.7%)	10/10 (100%)	0.04
Follow-up (in months)	12 ± 8.6 (4–36)	12.1 ± 10.6(6–36)	11.8 ± 6.7(4–24)	0.9
Recurrence	4/19 (21%)	1/9 (11.1%)	3/10 (30%)	0.3
Clinical findings				
Sixth nerve palsy	4/19 (21%)	4/9 (44.4%)	0/10 (0%)	0.018
Headache	12/19 (63.2%)	4/9 (44.4%)	8/10 (80%)	0.03
Blurring of vision	8/19 (42.1%)	3/9 (33.3%)	5/10 (50%)	0.4
Visual obscurations	5/19 (26.3%)	1/9 (11.1%)	4/10 (40%)	0.16
Deterioration of the school performance	12/19 (63.16%)	4/9 (44.4%)	8/10 (80%)	0.1
Irritability and irregular sleep	8/19 (42.1%)	5/9 (55.5%)	3/10 (30%)	0.27
ICP (mmH_2O)	291 ± 79.5 (180–460)	258 ± 69 (180–350)	320 ± 80 (250–460)	0.09

Table 2 Associated risk factor

Risk factors	Total	Group I	Group II	*P* value
Female predominance	16/19 (84.2%)	6/9 (66.7%)	10/10 (100%)	0.049
Obesity	3/19 (15.8%)	1/9 (11.1%)	2/10 (20%)	0.05
Anemia	5/19 (26.3%)	4/9 (44.4%)	1/10(10%)	0.04
Infection	3/19 (15.8%)	3/9 (33.3%)	0/10 (0%)	0.05

bronchitis were the potential risk factors in group I (Table 2) while obesity was found in only one patient in group I and 2 patients in group II; also, Mosquera Gorostidi and his colleagues [15] did not report obesity as associated risk factor in patient less than 15 years old.

The clinical findings of IIH are variable [4, 15], but headache and papilledema are the main clinical findings in adult with or without additional symptoms and signs of increased ICP [4, 9, 14, 19, 20]; however, a typical IIH was diagnosed without headache [12] or papilledema [21]. In this study, the patients of group II have quite similar clinical findings and diagnostic criteria to adult IIH as headache is the most common symptom while non-specific symptoms of increased ICP as deterioration of the school performance, irregular sleeping and irritability, and loss of appetite were significant in group I (Table 1). In pediatric IIH, strabismus, due to the sixth nerve palsy, is the most common presenting symptom in the prepubertal patients [7], and in our study, it was reported in 4 patients (44.4%) in group I but not reported in group II; also, the sixth nerve palsy was reported in one case out of the 12 pediatric patients in the series of Mosquera Gorostidi and his colleagues [15]. Neuroimaging is usually normal in cases with IIH [2], but venous sinus stenosis, small ventricles, empty sella, and optic nerve tortuosity were reported in patients with IIH [15, 17, 22–26]. In our series, transverse sinus stenosis was found in one case in group I and one case in group II while optic nerve tortuosity was found in only one case in group I, but the radiological findings in this series underestimated the findings in IIH cases because our main target of radiological work-up was exclusion of hydrocephalus and mass lesion; specific radiological details need high-resolution MR imaging with special MRI protocol with thin slice which improves the visualization and findings in the pituitary gland and optic nerves [26] while magnetic resonance venography in pediatrics IIH may be questionable, as partial occlusions may be unnoticed [15].

In this study, the mean opening pressure is 291 ± 79.5 cmH_2O and the minimum value for opening pressure in group I is 180 cmH_2O, while the minimum value in group II is 250 cm (Table 1); however, the diagnostic limit of the CSF opening pressure is considered a matter of debate in the diagnosis of pediatric IIH, but in the presence of typical IIH, symptoms and signs with papilledema low opening pressure could not exclude the diagnosis as the lumbar puncture may be done during the nadir of a pressure wave [12, 14]. Intracranial hypertension is considered in children with age less than 8 years if there is papilledema and cerebrospinal fluid opening pressure greater than 18 cmH_2O or no papilledema with opening pressure greater than 25 cmH_2O while in children above 8 years old the diagnosis is confirmed if the opening pressure is greater than 25 cmH_2O with papilledema [2]. In another retrospective observational study, the highest opening pressure in pediatric IIH was between 20 and 24 cmH_2O in 11% of the patients while it was between 25 and 39 cmH_2O in 48% and more than 40 cmH_2O in 42% of the patients [16]. In a newer study, opening pressure ≥ 28 cmH_2O is mandatory for diagnosis of intracranial hypertension for obese patients and 25 cmH_2O if the patient is not obese [13].

IIH in pediatric patients usually respond to pharmacological treatment with early clinical improvement after the initial lumbar puncture [2, 9, 10, 15, 16], and in our study, out of the 19 patients, 15 patients improved after the first lumbar puncture and drainage of CSF while repeated lumbar puncture was carried out for three patients with satisfactory symptom release in two of them while the third one needed lumbo-peritoneal shunt.

In this study, lumbo-peritoneal shunt was inserted for two patients in group II, one of them was due to prolonged symptom and failure of repeated lumbar puncture and the second one was inserted due to recurrence with visual field affection.

The treatment and outcome results in this this study are similar to the results of Mosquera Gorostidi and his colleagues [15] who recorded early clinical improvement in 66% of the pediatric IIH after the initial LP while in another study out of 9 pediatric patients good improvement after pharmacological treatment with one lumbar puncture in was seen in 5 patients and without LP in 3 patients, and only one patient needed optic nerve sheath fenestration [9].

The risk of recurrence of symptomatic IIH in adults is 40% [27] while the recurrence rate of symptomatic IIH in pediatrics is between 6 and 24% [2, 15, 28] and in this study the overall recurrence was confirmed in four patients (21%), one of them (11.1%) in group I while the other three cases (30%) in group II, which is quite similar to the risk of recurrence in adult IIH.

Conclusion

Pediatric idiopathic intracranial hypertension can be classified into two subtypes: pediatric type and adolescent type according to the secondary sexual criteria.

Modified Dandy criteria can be applied for the diagnosis of adolescent type, but pediatric type is not associated with obesity with less female predominance and usually responds to the initial lumbar puncture with a low rate of recurrence.

Abbreviations

CSF: Cerebrospinal fluid; ICP: Intracranial pressure; IIH: Idiopathic intracranial hypertension; MRI: Magnetic resonance image; MRV: Magnetic resonance venography; TSH: Thyroid-stimulating hormone

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Not applicable.

Authors' contributions

SA contributed to the idea of the research, collection of the data, collection of the references, writing of the article, and surgical intervention. AG contributed to the clinical part of the research and performed the ophthalmologic evaluation. MEM contributed to the clinical part of the research, collection of the data, and writing of the article. EAD contributed to the clinical part of the research, collection of the data, and surgical intervention. All authors read and approved the final manuscript.

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Availability of data and materials

Please contact author for data requests.

Ethics approval and consent to participate

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee (Zagazig Faculty of Medicine IRP committee and Kingdom Hospital ethical committee) and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Consent for publication

For this type of study, formal consent is not required.

Competing interests

The authors declare that they have no competing interests.

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