# Pediatric Idiopathic Intracranial Hypertension: Clinical and Demographic Features

**Sindičić Dessardo, Nada; Dessardo, Sandro; Sasso, Antun; Vukelić Šarunić, Alenka; Šubat Dežulović, Mirna**

*Source / Izvornik:* **Collegium antropologicum, 2010, 34 supplement 2, 217 - 221**

**Journal article, Published version**
**Rad u časopisu, Objavljena verzija rada (izdavačev PDF)**

*Permanent link / Trajna poveznica:* https://urn.nsk.hr/urn:nbn:hr:184:919791

Coll. Antropol. **34** (2010) Suppl. 2: 217–221
Professional paper

# Pediatric Idiopathic Intracranial Hypertension: Clinical and Demographic Features

**Nada Sindičić Dessardo, Sandro Dessardo, Antun Sasso, Alenka Vukelić Šarunić and Mirna Šubat Dežulović**

Pediatric Department, University Hospital Rijeka, Croatia

**A B S T R A C T**

*Idiopathic intracranial hypertension (IIH) is a clinical condition characterized by elevated intracranial pressure and absence of clinical, laboratory or radiographic evidence of central nervous system infection, vascular malformation, intracranial space occupying lesion or hydrocephalus. In the last years the raising understanding of pediatric IIH, especially concerning its demographics and epidemiology, has brought up to a redefinition of diagnostic criteria and reevaluation of pathogenesis and treatment. The authors reviewed the records of nineteen consecutive children with newly diagnosed IIH in order to compare demographic characteristics, clinical pictures and ophthalmologic aspects as optic disc evaluation and visual field evaluation, as well as treatment modalities and follow up. Beside obesity and female gender, potential alternative risk factors remains to be investigated, which need a good collaboration between neuro-ophthalmologists and pediatric neurologists.*

***Key words:*** *idiopathic intracranial hypertension, children, obesity, epidemiology*

## Introduction

Idiopathic intracranial hypertension (IIH) is a clinical condition characterized by elevated intracranial pressure and absence of clinical, laboratory or radiographic evidence of central nervous system infection, vascular malformation, intracranial space occupying lesion or hydrocephalus[1]. The incidence of the syndrome is about 1:100,000, with higher prevalence rates, reflecting the chronic nature of the condition. The pathogenesis of idiopathic intracranial hypertension is yet to be clarified but most theories focuses on the compromised venous blood flow within the intracranial compartment. In the last years the raising understanding of pediatric IIH, especially concerning its demographics and epidemiology, has brought up to a redefinition of diagnostic criteria and re-evaluation of treatment modalities. With advances in neuroimaging and better recognition of etiopathogenetic mechanisms the diagnostic criteria have been updated: 1) signs and symptoms of raised ICP or papilledema, 2) CSF opening pressure greater than 250 mm $H_2O$ (with normal composition), 3) absence of hydrocephalus, mass lesion and structural or vascular lesion on MRI or contrast enhanced CT and 4) no other identifiable cause of intracranial hypertension[2]. Pediatric IIH is further divided into prepubertal and pubertal forms, based on development of secondary sexual characteristics or age. According to the most of published papers there is a net distinction between the groups in demographic and clinical characteristics. Pediatric IIH patients may present with systemic complaints such as headache, lethargy, poor appetite and somnolence while vision complaints may be overlooked, especially in younger children. Until recently, VI cranial nerve palsy was the only accepted neurological abnormality permitted in diagnosing IIH. Papilledema has generally been considered as a hallmark physical finding of idiopathic intracranial hypertension and its severity is positively correlated with the risk of visual loss. The incidence and type of visual problems in children are similar to the ones observed in adult patients. Vision loss in children with IIH is usually mild and reversible although occasionally it could be devastating and permanent[3]. In general, children with idiopathic intracranial hypertension have a good prognosis and complete resolution of symptoms, if diagnosed promptly and treated adequately.

Received for publication January 10, 2010

## Patients and Methods

Nineteen consecutive patients age ≤16 years were evaluated for a new diagnosis of idiopathic intracranial hypertension at the Pediatric department of the University Hospital in Rijeka, Croatia, during a 12 years period. Their medical records were reviewed, and age at diagnosis, sex, height and weight, as well as signs and symptoms at time of initial examination were recorded. All of them met the modified Dandy criteria for the diagnosis of IIH:

1. cranial CT and/or MRI were performed in all cases in order to confirm the absence of a mass lesion or hydrocephalus

2. all of them had an elevated CSF opening pressure with normal fluid content

3. all patients had a normal result of neurologic examination with the exception of papilledema, visual disturbances or VI nerve palsy

The patients were defined as »pubertal« or »prepubertal« according to the presence of secondary sexual characteristics, and the term prepubertal was reserved for patients who had yet to develop any secondary sexual characteristics.

A standard Body Mass Index was used to determine obesity[4]. Patients were categorized as overweight/obese if their BMI at the time of diagnosis of IIH was over the 85 or 95 percentile value respectively.

All patients underwent complete neuro-ophthalmic examination including visual acuity and visual field testing. The visual filed was evaluated using the KOWA automatic visual field plotter. The optic nerve assessment and grading of papilledema were performed using direct opthalmposcopy and optic disc edema graded according to the Frisen scale[5].

## Results

### *Demographics*

During a twelve years period nineteen children under 16 years of age were admitted to our University Hospital fulfilling the diagnostic criteria for idiopathic intracranial hypertension. The mean age was 10.7+/–4.07, and the male-to-female ratio was 6:13. In the prepubertal group (n=7) the male-to-female ratio was 4:3, whereas in the pubertal group it was 2:10. We haven't found any remark in the perinatal and family history of our patients.

Clinical features

The presenting symptoms of our study population are shown in Tables 1 and 2. There were no asymptomatic patients. Papilledema was found in eighteen patients. Neurologic evaluation was normal in fifteen children while patients had VI cranial nerve paresis. In our study population we haven't found any other neurologic abnormality.

**TABLE 1**
CLINICAL SYMPTOMS

| SYMPTOMS | NUMBER | % |
|---|---|---|
| Headaches | 18 | 94 |
| Nausea/vomiting | 10 | 52 |
| Diplopia | 3 | 16 |
| Visual obscuration | 9 | 47 |
| Somnolence | 2 | 10 |
| Tinnitus | 2 | 10 |
| Neck/shoulder pain | 1 | 5 |

**TABLE 2**
WEIGHT DIFFERENCES BETWEEN GROUPS

| WEIGHT | TOTAL (19) | PREPUBERTAL (7) | PUBERTAL (12) |
|---|---|---|---|
| Normal Weight | 5 (26 %) | 2 (28 %) | 3 (25 %) |
| Overweight | 5 (26 %) | 2 (28 %) | 3 (25 %) |
| Obesity | 9 (48 %) | 3 (44 %) | 6 (50 %) |

### *Associated conditions*

All patients underwent a thorough diagnostic work-up in order to exclude any associated medical condition that could result in a diagnosis of secondary intracranial hypertension. By that means we have excluded in all of them any systemic diseases, endocrinology problems, hematologic conditions as well as electrolyte and/or fluid dysbalance and renal failure.

### *Diagnostic procedures*

Lumbar puncture was done on all patients. Opening pressure ranged from 220 to 410 mm Hg with a mean of 295+/–37 mm $H_2O$, with normal cerebrospinal fluid content in all patients. Four patients had an opening CSF pressure less than 250 mm $H_2O$. Visual field analysis were performed in 19 patients, the results are shown in Table 3. Computed tomography (CT) was done in 5 patients, magnetic resonance imaging (MRI) was done in all 19 patients. Visual evoked potentials (VEP) were analyzed in 14 patients, one patient had a bilateral prolonged latency, the others were normal. The EEG finding was normal in all patients.

**TABLE 3**
VISUAL FIELD DESCRIPTION

| VISUAL FIELD | NUMBER | % |
|---|---|---|
| Normal | 5 | 26 |
| Nasal field deficit | 3 | 16 |
| Blind spot enlargement | 8 | 42 |
| Concentric constriction | 3 | 16 |

*Treatment modalities*

All nineteen patients were treated with acetazolamide. Additional twelve were treated with topiramat, chronologically the last 8 patients were treated with both acetazolamide and topiramat as initial treatment approach. Corticosteroids were adjunct in 4 patients. No child underwent any aggressive procedure as serial spinal taping, ventriculoperitoneal shunts or optic nerve sheath fenestreation.

*Clinical course and outcome*

Complete resolution of swollen disc findings was achieved in 12 patients following median treatment duraion of 5 months (4–6 months). Seven patients had peristent optic disc abnormalities. One child had a symptomatic recurrence of IIH resolving after an additional course of acetazolamide and topiramat. A six months ophthalmologic follow up has shown a complete regression of fundus abnormalities in ten patients, while in the other nine persisted just minor changes of the optic disk(six with indinstinct disk margins and 3 with pale optic disk).

## Discussion

The incidence of pediatric IIH resulting from our study is 1.2/100 000 which is consistent with the previously published epidemiologic data. The most common complaints were headache (18/19), visual disturbances (12/19) and nausea and vomiting (10/19). In general, headache is the most common symptom, present virtually in every patient with IIH. The generalized headache with morning worsening, typical for IIH, is often accompanied by retro-orbital, neck, shoulder and arm pain[6]. Headaches in idiopathic intracranial hypertension could resemble migrainous headaches, although in IIH headaches tend to be continuous and less severe than in migraine. It is still unknown why a small number of patients have an absence of headache, despite an increase in cerebrospinal pressure.

Three patients presented with sixth cranial nerve palsy at the time of hospital admission. The reported incidence of this neurological abnormality among children with IIH varies from 9% to 48%, while palsies of the 3rd, 4th, 7th,9th and 12th cranial nerves, which could be found in pediatric IIH patients are far less common[3,7]. It is still unclear why ocular nerves palsy as well as other ocular motility disorder like ophtalmoplegia or nystagmus are quite frequent among IIH patients since a raised intracranial pressure could not completely explain the mechanism of these abnormalities.

Papilledema, as a criterion for the diagnosis, is observed in nearly all patients and is the most important sign, although the absence of it is not an exclusionary criterion. Clinical examination and subjective grading of optic disc changes is still the most common approach for evaluation of optic nerve swelling in patients with idiopathic intracranial hypertension. New and emerging techniques as Optical Coherence Tomography (OCT) or Laser Scanning Tomography (LST) seems to be very effective in optic nerve evaluation[8,9]. Additional studies are needed in order to establish the role of these new diagnostic tools in the diagnosis and follow-up of papilledema in children with IIH.

Fifthteen of nineteen patients had some pattern of visual field defect. One of them had atypical scotoma whether the others presented with more common visual field defects as blind spot enlargement, global constriction and nasal field defects. Visual field testing has been shown to be more sensitive than visual acuity or contrast sensitivity in detecting visual deficits in adult IIH patients[10]. The automated perimetry makes the visual field assessment more standardized and accurate. Although most authors agree than the majority of children 4 years and older are able to perform reliable visual field testing some authors remain skeptical regarding the age in which children can cooperate and will not use this method in children below 8 years of age[11].

All patients had a CSF opening pressure of 200 mm $H_2O$ or more, while fifthteen of nineteen had the CSF pressure over the discriminating value of 250 mm $H_2O$, according to the modified Dandy criteria. Nevertheless we are considering all of them over the upper limit for normal opening pressures, since the majority of published data concerning children values, as cited by Fishman[12] consider 180 mm $H_2O$ the upper limit of normal opening pressure for children less than 8 years of age. Nevertheless, we haven't observed in any of them a dramatic clinical picture of the so called »fulminant IIH« which usually could be seen in about 2–3% of patients with IIH and is characterized by extremely severe visual loss and need some kind of surgical procedure (optic nerve sheath fenestration, lumboperitoneal or ventriculoperitoneal shunting) to obtain a satisfactory clinical improvement[13]. Although no controlled trial has evaluated surgical treatments in IIH there is a general agreement that surgery is indicated in cases of progressive visual loss[14].

In our patients we have found a marked predominance of obesity and overweight, even in the prepubertal group. According to previous publications it seems that pubertal and prepubertal IIH patients differs in demographic and clinical characteristics, especially concerning the impact of overweight which is so accentuated in the pubertal patients, similarly to the adults[15,16]. Unlike the results of Balcer et al. this investigation, although the number of patients is restricted, does not provide strong evidence that in the pediatric age group, younger patients with IIH are less likely to be obese than are older children or adults.

Many case reports and case series describe various associations between intracranial hypertension and certain medical conditions and medications. The most reported association of intracranial hypertension is with systemic diseases as lupus erythematosus[17], thyroid gland dysfunction and other endocrinology problems[18,19], hematologic conditions[20] as well as chronic renal failure[21].

These associations should be described as a secondary pseudotumor cerebri[22] and the term IIH should be reserved for the purely idiopathic condition. In our case series we haven't found any associated medical condition or pharmacologic treatment so we could say that all of our patients had a »pure« idiopathic intracranial hypertension. Anyhow, much more information has yet to be learned regarding the risk factor profile for idiopathic intracranial hypertension in children. Larger multicentric trials will be needed to examine the potential roles of metabolic, hormonal or any other physiologic factor in determining the relation between age and overweight in pediatric IIH, as well as to search for potential alternative mechanisms for the development of IIH in nonobese children.

The patients were treated with acetazolamid and topiramat while five patients needed steroid therapy for severe visual disturbances. Traditionally, oral acetazolamide, a carbonic anhydrase inhibitor, has been the first-line medication for inhibiting CSF production. Oral topiramat is a useful adjunct which was found to be equally efficacious in improving vision, visual fields and optic disc edema, with a benefit of being a promoter of substantial weight loss. In our institution, according to a greater amount of published experiences as well as on personal experience, we place our IIH patients on both acetazolamide and topiramate together[15]. Parenteral corticosteroid treatment is reserved for patients with severe visual problems since they can show some temporary benefit by minimizing further nerve damage in situations of severe papilledema.

Complete resolution of papilledema was achieved in a majority of our patients within six months. Nevertheless, we should take account of seven of our patients with persistent optic disc abnormalities and of one child with symptomatic recurrence of IIH. According to previously published paper the optimal duration of follow up is not yet established. The« Iowa experience« with patients followed over ten years shows that IIH can be either a subacute and remitting disorder or take a more chronic course which justifiy a long-term follow-up care in this group of patients[23].

## Conclusions

Among children IIH is a rare condition with a reported incidence of 1/100 000, and a slighter biger prevalence indicating the somewhat chronic caracter of the condition. Potential alternative risk factors remains to be investigated, which need a good collaboration between neuroophthalmologists and pediatric neurologists.

## REFERENCES

1. FRIEDMAN DI, JACOBSON DM, Neurology, 59 (2002) 1492. — 2. RANGWALA LM, LIU GT, Surwey of Ophthalmology, 52 (2007) 597. — 3. BABIKIAN P, CORBETT J, BELL W, J Child Neurol, 9 (1994) 144. — 4. KOLCIĆ I, BILOGLAV Z, ZGAGA L, JOVIĆ AV, CURIĆ I, CURIĆ S, SUSAC J, VELAGIĆ V, MATEC L, ZOBIĆ I, ZEDELJ J, STRNAD M, Coll Antropol, 33 (2009) 135. — 5. FRISEN L, J Neurol Neurosurg Psychiatry, 45 (1982) 13. — 6. GIUSEFFI V, WALL M, SIEGEL PZ, ROJAS PB, Neurology 41 (1991) 239. — 7. YOUROUKOS S, PSYCHOU F, FRYSSIRAS S, J Child Neurol 15 (2000) 453. — 8. SALCHOW DJ, HUTCHESON KA, J Pediatr Ophthalmol Strabismus, 44 (2007) 335. — 9. HECKMANN JG, FASCHINGBAUER F, LANG C, J Neurosurg, 107 (2007) 543. — 10. ROWE F, SARKIES NJ, Eye, 12 (1998) 111. — 11. STIEBEL-KALISH H, LUSKY M, YASSUR, Ophthalmology, 111 (2004) 1673. — 12. FISHMAN RA, Cerebrospinal fluid in diseases of the nervous system. In: SAUNDERS PA (Ed) (Philadelphia, 1992). — 13. THAMBISETTY M, LAVIN PJ, NEWMAN NJ, BIOUSS V, Neurology, 68 (2007) 229. — 14. FRIEDMAN DI, JACOBSON DM, J Neuro-ophthalmology, 24 (2004) 138. — 15. WOLF A, HUTCHESON KA, Current Opinion Ophtalm, 19 (2008) 391. — 16. BALCER LJ, LIU GT, FORMAN S, VOLPE NJ, GALETTA SL, MAGUIRE MG, Neurology, 52 (1999) 870. — 17. PADEH S, PASSWELL JH, Journal of Rheumatology, 23 (1996) 1266. — 18. HUSEMAN CA, TORKELSON RD, American Journal Disease in Childhood, 138 (1984) 927. — 19. CONDULIS N, GERMAIN G, CHAREST N, LEVY S, CARPENTER TO, Clin Pediatr, 36 (1997) 711. — 20. CAPRILES LF, Arch Neurol, 9 (1963) 147. — 21. ČORAK M, BAĆANI B, CVITANOVIĆ-SOJAT L, NOVAK-LAUS K, ZRINSĆAK O, MANDIĆ Z, Coll Antropol, 29 (2005) 133. — 22. CHANG D, NAGAMOTO G, SMITH WE, Cleve Clin J Med, 59 (1992) 419. — 23. SHAH VA, KARDO RH, LEE AG, CORBETT JJ, WALL M, Neurology, 70 (2008) 634.

*N. Sindičić Dessardo*

*Pediatric Department, University Hospital Rijeka, Istarska 43, 51000 Rijeka, Croatia*
*e-mail: sindin2@medri.hr*

## PEDIJATRIJSKA IDIOPATSKA INTRAKRANIJSKA HIPERTENZIJA: KLINIČKA I DEMOGRAFSKA OBILJEŽJA


### S A Ž E T A K

Idiopatska intrakranijska hipertenzija (IIH) kliničko je stanje karakterizirano uvećanim intrakranijskim pritiskom uz odsustvo kliničkih, laboratorijskih ili radioloških elemenata u prilog infekcije središnjeg živčanog sustava, krvožilne malformacije, intrakranijske ekspanzivne tvorbe ili hidrocefalusa. Posljednjih godina je sve veće razumijevanje pedijatrijske IIH, poglavito na polju epidemiologije i demografskih obilježja, dovelo do redefiniranja dijagnostičkih kriterija kao i reevaluacije patogeneze i liječenja. Autori analiziraju demografske značajke te kliničke karakteristike kao i oftalmološke aspekte grupe od devetnaestoro djece s idiopatskom intrakranijskom hipertenzijom, uz analizu terapijskih obilježja. Uz prekomjernu tjelesnu težinu te ženski spol kao poznate čimbenike rizika, potrebna su daljnja istraživanja mogućih rizičnih faktora za IIH, što zahtjeva dobru suradnju neurooftalmologa i neuropedijatra.