

## BRITISH PAEDIATRIC SURVEILLANCE UNIT

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### CHILDHOOD IDIOPATHIC INTRACRANIAL HYPERTENSION

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#### Abstract

Idiopathic intracranial hypertension (IIH), previously known as pseudotumor cerebri or benign intracranial hypertension, is a rare condition of increased intracranial pressure without any identifiable pathology. The clinical definition and associations of this unique condition have evolved with time and the advances in neuroimaging. Despite intervention, the clinical course of IIH is often prolonged and recurring with potential complications of distressing headache and blindness.<sup>1-3</sup> The overall (children and adult) annual incidence of IIH is estimated to be 1-3 per 100,000 population,<sup>4-5</sup> however the epidemiological data on childhood IIH to date are sparse and limited to hospital based retrospective case series.<sup>6,7</sup> The principle objective of this BPSU study is to determine a contemporary national annual incidence of IIH in children. Furthermore, up to date clinical information will be collected to devise current best practice to guide clinicians in future management of paediatric IIH cases.

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#### Background

Idiopathic intracranial hypertension (IIH), first described in 1897, is a rare condition of increased intracranial pressure (ICP) without any identifiable pathology. In adulthood, IIH is most common in obese young women, however in childhood both genders are equally affected. Although numerous risk factors for IIH have been described, the exact pathophysiology of IIH remains unclear. Moreover, the characteristics of childhood IIH have changed over recent decades, otitis media<sup>8</sup> is no longer the main association and obesity is increasingly being recognised as a possible risk factor in recent paediatric literature.<sup>9</sup> This raises the concern that the recent rise in the prevalence of childhood obesity will be associated with an increased incidence of IIH. The overall annual incidence of IIH is estimated to be 1-3 per 100,000 population,<sup>4-5</sup> however the epidemiological data on childhood IIH to date are sparse and limited to hospital based retrospective case series.<sup>6-7</sup>

The key features of IIH are headache and papilloedema, but the spectrum of clinical presentation varies with age. The diagnosis of IIH is currently based on the clinical presentation, neurological, ophthalmological, neuroimaging and cerebrospinal fluid (CSF) findings of raised ICP and the exclusion of other causes of generalised intracranial hypertension. The level for raised ICP in diagnosing children with IIH is often considered as above 20cm CSF, based on the lower range of the upper normal limit in normal adult data. The normal ranges of ICP for children in fact vary with age. It is well recognised that movement, posture and emotions can affect CSF pressure during lumbar puncture (LP) and ICP can also vary considerably with time. The momentary manometric assessment of the CSF column by LP in children is merely a crude recording of ICP.<sup>10</sup> Hence, the opening lumbar CSF pressure is not invariably raised in some cases of childhood IIH.<sup>2,11</sup>

Treatment may include rectifying underlying predisposing condition or withdrawal of precipitating factor, medication (analgesia, diuretics or steroids), therapeutic lumbar puncture(s) or neurosurgery to divert the drainage of CSF. However there, is yet no consensus on how best to treat IIH in children due to a lack of evidence in therapeutic effectiveness. Despite intervention, the clinical course of IIH is often prolonged and recurring with potential complications of distressing headache and blindness.<sup>1-3</sup> Prompt diagnosis with close monitoring of vision are of paramount importance in managing IIH cases. Through this BPSU surveillance we will gain a contemporary national incidence figures of IIH in children to fill the current epidemiological gap of this rare and potentially serious condition. It will also provide up to date clinical information to devise current best practice and to guide clinicians in the future management of paediatric IIH cases.

**Coverage** United Kingdom and Republic of Ireland

**Duration** July 2007 – July 2008 (13 months)

**Objectives** To determine the

1. annual incidence of IIH in children aged 1 to 16 years in the UK and Ireland
2. spectrum of clinical presentation of IIH children across various age groups
3. national incidence of various established associations of IIH in children in particularly with obesity at presentation
4. frequency and spectrum of visual disturbances in children presenting with IIH
5. current clinical management of children with IIH
6. the outcome of headache and spectrum of the visual disturbances in this national cohort one- year post diagnosis following various treatment modalities.

**Case definition** Any child aged 1 to 16 years (not including 17<sup>th</sup> birthday) who fulfils at least two of the key features and all of the three essential criteria.

At least TWO Key Features:

1. Symptoms of raised intracranial pressure (such as headache, nausea, vomiting or irritability) **and/or** visual symptoms diplopia, blurring vision or transient visual loss
2. Papilloedema, unilateral or bilateral
3. Raised opening cerebrospinal fluid pressure above 20 cm by lumbar puncture

AND all THREE Essential Criteria:

1. Normal level of consciousness
2. Cranial imaging (including CT or MRI and MR or CT venography) does not reveal a structural cause such as ventricular dilatation, cerebral mass, vascular lesion or sinus venous thrombosis\*, to explain the presenting symptoms or signs of raised intracranial pressure
3. Normal cerebrospinal fluid contents (for atraumatic tap: white cell count < 6 x 10<sup>6</sup> /L, protein < 0.4 g/L and ratio of cerebrospinal fluid glucose to plasma glucose > 0.5)

**Excluding** \*sinus venous thrombosis whose neuroimaging appearances can be difficult to distinguish from venous obstruction related to raised intracranial pressure. **Please report if in doubt or if case was excluded due to sinus venous thrombosis.**

**Caution** Optic nerve head Drusen (a degenerative condition consists of hyaline deposits within the optic nerve head which results in an apparent elevation or swelling of the optic disc) can mimic papilloedema. However papilloedema and optic nerve head Drusen can occur concurrently, their differentiation can be made by optic ultrasound and/or orbital CT scan. **If in doubt, please report suspected cases.**

**Reporting instructions** Any child aged 1-16 years (not including 17<sup>th</sup> birthday) seen in the past month who fulfils at least two of the key features and all of the three essential criteria

**Methods** Paediatricians who have notified a case will be asked to complete (1) a short case notification questionnaire to obtain demographic, clinical and initial management details, and (2) a brief follow-up questionnaire which will be sent a year after initial diagnosis to seek data on subsequent management and the clinical outcome for each notified case.

**Ethics Approval** This study has been approved by the MREC (East London and the City Research Ethics Committee, Ref:07/Q0603/47), PIAG/BPSU 2-05(FT3)/2007 and North East Wales NHS R&D

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**Patient Support Group** Association for Spinal Bifida and Hydrocephalus (<http://www.asbah.org>)

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