

LRI Children's Hospital

Diagnosis and Management of Idiopathic Intracranial Hypertension.

Staff relevant to:	Medical & Nursing staff caring for Children & Young people with Idiopathic Intracranial Hypertension.
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1. Introduction and Who Guideline applies to

Idiopathic intracranial hypertension (IIH) is a rare condition of increased **intracranial** pressure (ICP) without any identifiable pathology. Despite intervention, the clinical course of IIH is often prolonged and recurring with potentially serious complications of distressing headache and blindness (1). The overall (including children and adult) annual incidence of IIH is estimated at 1–3 per 100000 population. Obesity has become increasingly dominant in recent paediatric reports, affecting 43–91% of teenage IIH cases.

This guideline applies to Medical & Nursing staff caring for Children & Young people with Idiopathic Intracranial Hypertension.

Related Documents:

This guideline needs to be used in conjunction with relevant infection control and consent policies to ensure the child receives safe care and children and families are able to understand the reasons for care to facilitate co-operation.

1. [Lumbar Puncture UHL Childrens Hospital Guideline UHL C82/2007](#)
2. [Swollen Optic Discs pathway Children's UHL Childrens Hospital Guideline UHL D3/2022](#)

2. Definition

Headache syndrome characterized by:

- Raised CSF pressure in the absence of ventricular dilatation or structural cause such as intracranial mass, vascular lesion or cerebral sinus venous thrombosis on MRI or CT cranial imaging and MR or CT venography—that is, no identifiable cause of intracranial hypertension or obstruction of CSF drainage i.e. NORMAL brain imaging.
- Normal spinal fluid composition (normal CSF biochemistry and cell counts on microscopy for atraumatic tap: white cell count $<6 \times 10^6/l$, protein <0.4 g/l and glucose CSF/plasma ratio >0.5).
- Usually normal neurological examination except for papilloedema with or without VIth nerve palsy
- Normal level of consciousness.
- IIH can occur in the absence of papilloedema.

2.1 Aetiology

Drugs	Endocrine disorders
Tetracycline	Hyperthyroidism, Hypothyroidism
Corticosteroid withdrawal	Vitamin D Deficiency
Nalidixic acid, Nitrofurantoin	Hypoparathyroidism
Oral Contraceptives	Adrenal Insufficiency
Isotretinoin	Head Trauma
Thyroid replacement	Infections like ear and sinus disease
Growth Hormone replacement	Venous sinus thrombosis
Vasopressin, Phenytoin	Systemic disorders like SLE, Leukemia,
Indomethacin, Cyclosporin	GBS, Iron-deficiency, hypercalcemia.

2.2 Clinical presentation

Symptoms of intracranial hypertension. (General)	Vision related symptoms and signs
Headache, often pulsatile, may wake from sleep, worse on lying down and with coughing, bending over or physical activity (a common and chief complaint in older children)	Papilloedema (unilateral or bilateral if present)
Nausea, vomiting	Transient visual loss
Lethargy, mood change	Blurring of vision
Irritability, apathy, somnolence, dizziness, ataxia, sleep and behaviour disturbance (all are common in young children)	Diplopia

Facial paresis, back and neck pain (common in pre-adolescents)	Squint
Neck stiffness	Sixth cranial nerve palsy
Retroocular pain worse with eye movement	Visual acuity loss
Tinnitus	Visual field defect
Asymptomatic except from papilloedema	

2.3 Investigations

Neuroimaging

To exclude a mass lesion/hydrocephalus, MRV may be warranted to exclude an occult venous sinus thrombosis.

Blood investigations: FBC, Vitamin D, TSH, Bone profile and others if necessary as mentioned in the aetiology table.

CSF pressure measurement

Best done in a co-operative non-sedated child but if not possible, consider sedation. It must be noted that sedating agents can cause falsely high CSF pressure measurements.

Current diagnostic criteria for IIH in children uses a lumbar puncture opening pressure more than 25 cm H₂O.

Cerebrospinal fluid composition (protein, cells, glucose) is normal in patients with IIH. If it is not normal, consider alternative diagnosis to IIH.

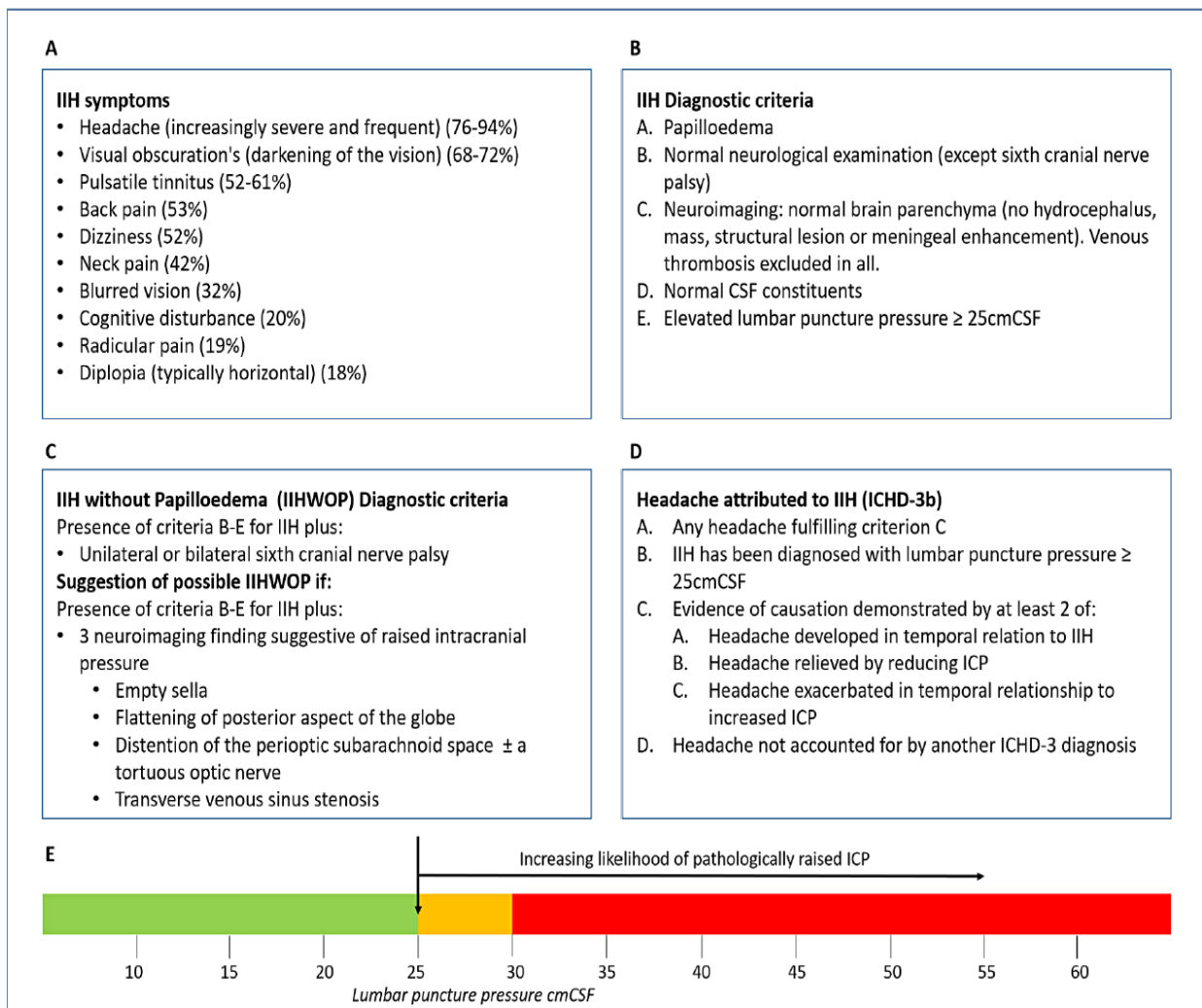


Figure 1 Consensus in diagnosing IIH. (A) Frequency of IIH symptoms reported, adapted from Markey *et al.*³ (B) IIH diagnostic criteria, adapted from Friedman *et al.*⁴ (C) IIHWOP diagnostic criteria, adapted from Friedman *et al.*⁴ (D) Headache attributed to IIH, as described by the International Classification of Headache Disorders, 3rd edition (beta version) (ICHD-3 beta).⁶ (E) Line figure detailing the consensus of the interpretation of LP opening pressure. Uncertainty: it needs to be recognised that this is a single LP OP measurement; and after raised ICP what is then a normal ICP for this population on repeat LP readings is unknown. CSF, cerebrospinal fluid; IIH, idiopathic intracranial hypertension; LP, lumboperitoneal.

Ophthalmic assessment

Loss of visual function is the serious permanent complication of IIH. **Visual Field testing** remains the most sensitive indicator of incipient visual loss. Visual outcome apparently unrelated to duration of symptoms, degree of papilloedema, presence of visual obscurations or incidence of recurrent increased ICP.

2.4 Treatment options

The various treatment modalities used in children have included corticosteroids, acetazolamide, repeated lumbar punctures, and surgery. Most cases respond to non-surgical management. **The goals of treatment are symptom relief and preservation of vision.**

Medical Management.	Surgical interventions	Other options
<ul style="list-style-type: none"> • Lumbar puncture • Medications 	<ul style="list-style-type: none"> • Lumbo-peritoneal shunt • Optic sheath fenestration 	<ul style="list-style-type: none"> • Migraine treatment in mixed headaches

Lumbar puncture

Removal of sufficient fluid to lower CSF pressure of no more than 20 cm – it may be therapeutic as well as diagnostic.

Acetazolamide (1st line)

As per BNFC. Side effects: GI upset; perioral and digital tingling; loss of appetite; acidosis and electrolyte imbalance; nephrocalcinosis.

Topiramate: It is dosed at 1.5–3 mg/kg/day in 2 divided doses. Side effect: loss of appetite and on a long term can cause kidney stones.

CSF pressure monitoring

Consider for: Failure of medical treatment and assessment pre-consideration to shunting. Very young children with persistent symptoms and when visual field testing cannot be undertaken and evaluation of children with unremitting symptoms in the absence of papilloedema and who are unresponsive to medication.

Should be taken off medication for a trial period to exclude low pressure headache.

Patient information leaflet: <http://www.cafamily.org.uk/Direct/b14.html>

Identification and correction of presumed/overt predisposing factors

- Potential agents that might cause or worsen IIH (e.g., tetracycline derivatives) should be discontinued. Treatment may be needed for comorbid sleep apnoea and anaemia if present.
- Reducing weight loss in all obese patients with IIH: While the reported benefit for IIH is anecdotal; weight loss is associated with other health benefits.

3. Education and Training

None required

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Management of suspected and confirmed IIH	Audit	Consultant	Annually	Local clinical practice group

5. Supporting References

1. T Soler, T Cox, P Bullock. Diagnosis and management of benign intracranial hypertension. *Arch Dis Child* 1998; **78:89-94** doi:10.1136/adc.78.1.89.
2. Y Y Mathews, Drugs used in childhood idiopathic or benign intracranial hypertension. *Arch Dis Child Educ Pract Ed* 2008;**93:19-25** doi:10.1136/adc.2006.107326.
3. Wraige E, Chandler C and Pohl1 KRE (2002) Idiopathic intracranial hypertension: is papilloedema inevitable? *Archives of Disease in Childhood* 87: 223-224.
4. Mollan SP, et al. Idiopathic intracranial hypertension: consensus guidelines on management. *J Neurol Neurosurg Psychiatry* 2018;**89:1088–1100**. doi:10.1136/jnnp-2017-317440.

6. Key Words

CSF pressure measurement, **Idiopathic intracranial hypertension**

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs. As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

Contact and review details	
Guideline Lead (Name and Title) D Baskaran – Trust Grade in Paediatric Neurology N Hussain - Consultant in Paediatric Neurology	Executive Lead Chief Medical Officer
Details of Changes made during review: Added blood investigations required Amended - Diagnostic criteria for IIH in children uses a lumbar puncture opening pressure more than 25 cm H ₂ O Identifies Acetazolamide as (1 st line), added Topiramate, removed steroids and frusemide treatment options	