Ashford and St. Peter's Hospitals
NHS Foundation Trust

Idiopathic Intracranial Hypertension In Paediatrics

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Guideline History		
Date	**Comments**	**Approved By**
15/02/2021		Paediatric Guidelines Group

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1.

Idiopathic Intracranial Hypertension In Paediatrics

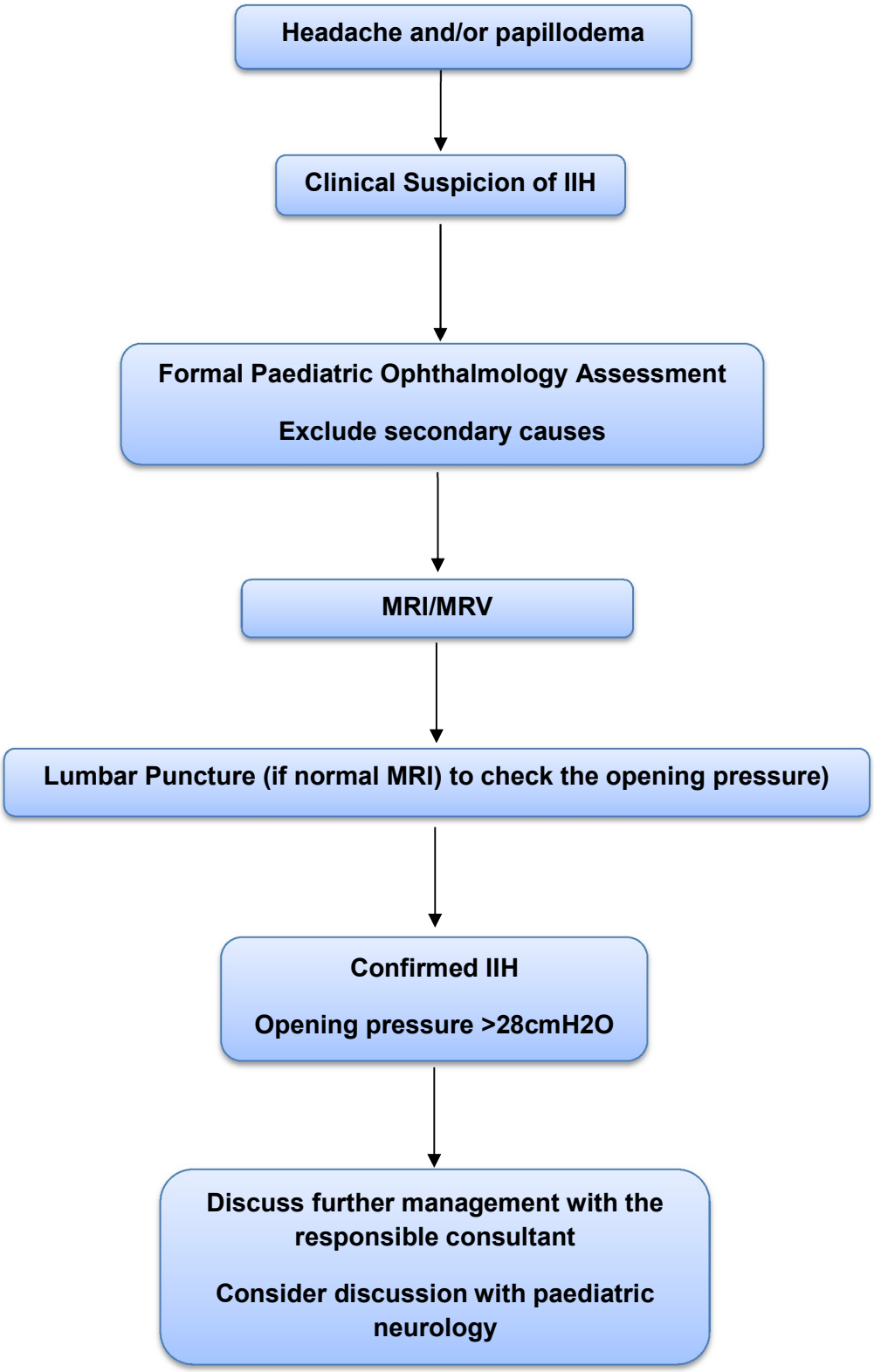
Introduction

This guideline has been created in response to several cases seen in our department over the last year. Children can present in many ways (not overweight, asymptomatic , only headache, others with cranial nerve palsies) thus this guideline aims to achieve an optimal strategy on the management of IIH.

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. A British Paediatric Surveillance Unit (BPSU)-supported study has estimated the UK annual incidence of IIH to be 0.8 (1.0 for girls, 0.5 for boys) per 100 000 child population, with a median age of presentation was 12 years, and obese girls are at most risk. Obesity is a risk factor for IIH in adults and older children. In one case series, 91% of IIH patients aged 15–17 years were obese, although the association was much weaker in prepubertal children. **[1]**

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

Summary – An algorithmic approach for management of IIH



1. Diagnostic Criteria: [2]

1- IIH with papilloedema (Definite Diagnosis)
A- Papilloedema B- Normal Neurologic Examination. C- Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and MR venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used. D- Normal cerebrospinal fluid (CSF) composition. E- Elevated lumbar puncture opening pressure (≥25 cm of H2O, or >28 cm of H2O if obese or sedated) in a properly performed lumbar puncture.
2- IIH without Papilloedema (Probable Diagnosis)
A- If B – E from above are met and satisfied, in addition, the patient has a unilateral or bilateral 6th cranial nerve palsy. B- If B–E from above are met and satisfied, and additionally, at least 3 of the following neuroimaging criteria are satisfied: • Empty Sella. • Flattening of the posterior aspect of the globe. • Distention of the peri-optic subarachnoid space with or without a tortuous optic nerve. • Transverse Venous Sinus Stenosis.

A diagnosis of IIH is definite if the patient fulfils criteria A–E.

The diagnosis is considered probable if criteria A–D are met but the measured CSF pressure is lower than specified for a definite diagnosis.

3. History/Signs and Symptoms:

a) Headache:
• Exacerbation with Valsalva maneuver, lying flat or bending down are useful clues.
• Pulsatile tinnitus that is described as a 'whooshing sound' in the ears is commonly reported.

b) Vision:
• Asymptomatic visual field impairment is usually an early sign.
• The blind spot is commonly affected first followed by the peripheral fields.
• Visual disturbance is an alarming symptom and warrants immediate treatment (Malignant IIH).
• Blurred Vision, Diplopia, squint (features of 6th cranial nerve palsy).

c) History suggestive of analgesics/drugs overuse headache should be sought.

d) Sleep difficulties or snoring or features of nocturnal hypoventilation.

e) Irritability, somnolence, dizziness, ataxia, sleep and behavior disturbance (all are common in young children).

f) Back and neck pain.

4. Examiation:

By definition, the neurological examination should be normal apart from the following exceptions:

- papilloedema
- reduced visual acuity
- visual fields defects
- unilateral or bilateral sixth nerve palsy

General examination should include:

- Blood pressure.
- Weight/height measurements with calculation of the Body Mass Index (BMI) (kg/m2), and BMI centile or BMI standard deviation score (SDS).
- Ear, nose and throat examination for signs of otitis media, mastoiditis, or sinusitis.
- A search for clinical signs of medical conditions causing secondary intracranial hypertension is recommended.

5.Investigations

1- Formal Paediatric ophthalmological assessment is essential, to ensure that true papilledema is diagnosed and differentiated from optic disc crowding and optic nerve head drusen. Examination and investigations would include:

- Visual fields and visual acuity
- Blind Spot
- Fundal photo
- B scan, Optical coherence tomography

2- Neuroimaging, MRI brain to exclude a mass lesion/hydrocephalus, MR venography (MRV) should also be done (discuss with the radiologist), to identify cerebral venous system abnormalities

3- LP must only be performed after satisfactory neuroimaging (Opening and closing pressure, microscopy, culture, virology, glucose). Patients and parents should be warned about post procedure low-pressure headache which can last for a few days, and advised to take extra fluids.

4- Blood (FBC, U&E, LFT, Bone Profile, TFT, CRP, ESR, Vitamin D, PTH, and lactate and glucose paired with CSF, consider early morning cortisol and Lyme serology).

6. Risk factors (2nd causes of IIH)[3-4-5]

Drugs	Medical Conditions
Tetracycline	Hyperthyroidism, Hypothyroidism
Corticosteroid Therapy / Withdrawal	Vitamin D Deficiency
Nalidixic acid, Nitrofurantoin	Hypoparathyroidism
Oral Contraceptives	Adrenal Insufficiency / Cushing syndrome
Isotretinoin/ Vitamin A	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.
Amiodarone	Cystic Fibrosis
Lithium	Polycystic Ovary Syndrome
	Down syndrome / Turner syndrome

7. Management

1- Identification and correction of presumed/overt predisposing factors:

● Potential agents that might cause or worsen IIH (e.g; tetracycline derivatives, Isotretinoin) should be discontinued. Treatment may be needed for comorbid sleep apnoea and anaemia if present.

● Advice regarding weight reduction in obese patients should be offered as there is evidence of benefit in adults.

2- Medical:

- Main indicators of a successful treatment are improvement in headaches and resolution of papilloedema and visual symptoms and signs. The length of treatment varies and may last, in some cases, 9–14 months. Weaning should be considered as soon as improvement is evident.**[3-6]**

- **Before starting the medication discuss with the responsible consultant.**

Drug	Mechanism of action	Dose	Side Effects	Comments
Acetazolamide	* Carbonic anhydrase inhibitor. * May reduce CSF production by reducing sodium ion transport from the choroid plexus cells.	* Prepubertal children: start at 25 mg/kg/d in 2–3 divided doses and increase by 25 mg/kg/d to a maximum of 100 mg/kg/day. * Adolescents (>11 years) : start at 1 g/day in 2–3 divided doses and if needed increase by 250 mg/d till a maximum of 4 g/day.	* Metallic taste, tingling sensation in the mouth, Paraesthesia and nausea. * If baseline U&E + HCO3 normal, then repeating them routinely are not recommended, unless high dose (> 1gm /day) or symptomatic. * Hemorrhage, metabolic acidosis, nephrolithiasis.	Used as firstline treatment. Start NaHCO3 or Potassium citrate if if levels fall below 18 mmol/l.*

- Other medications which can be used are Topiramate, Furosemide and steroid.

3- Lumbar puncture (LP): [4]

A- At the initial diagnostic LP, CSF pressure is brought down to 15 to 20 cm of H_2O.

B- There is no evidence that serial lumbar punctures lead to long-term remission of IIH. LP decreases CSF pressure only temporarily. Unless there is a CSF leak, CSF reforms in six hours.

C- Serial LPs could be useful as a temporary measure in fulminant IIH threatening vision, as a temporary measure as prelude to surgical treatment.

4- Surgery:

Indications for surgery are:

A- Fulminant IIH presenting with acute visual deterioration or loss.

B- Failure of medical treatment which is reflected by recent deterioration or loss of visual acuity or fields.

C- Severe debilitating headaches not responding to medical treatment.

2. Supporting References

1- Babiker MOE, et al. *Arch Dis Child Educ Pract Ed* 2014;99:166–172.

2- Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology* 2013;81:1159–65.

3- Rangwala LM, Liu GT. Pediatric idiopathic intracranial hypertension. Surv Ophthalmol 2007;52:597–617.

4- Matthews YY. Drugs used in childhood idiopathic or benign intracranial hypertension. Arch Dis Child Educ Pract Ed 2008;93:19–25.

5- Esmaili N, Bradfield YS. Pseudotumor cerebri in children with down syndrome. Ophthalmology 2007;114:1773–8.

6- Kesler A, Hadayer A, Goldhammer Y, et al. Idiopathic intracranial hypertension: risk of recurrences. Neurology 2004;63:1737.

2. Guideline Governance

a. Scope

This guideline is relevant to all staff caring for all children from 0-18 years old across the emergency department, inpatient ward and outpatient department.

b. Purpose

i. This guidelines aims to facilitate a common approach to the management of children. At times deviation from the guideline may be necessary, this should be documented and is the responsibility of the attending consultant.

ii. This guideline is subject to regular review to ensure ongoing evidence based practice.

c. Duties and Responsibilities

All healthcare professionals responsible for the care of all children 0-18years should be aware of practice according to this guideline.

d. Approval and Ratification

This guideline will be approved and ratified by the Paediatric Guidelines Group.

e. Dissemination and Implementation

i. This guideline will be uploaded to the trust intranet 'Paediatric Guidelines' page and thus available for common use.
ii. This guideline will be shared as part of ongoing education within the Paediatric Department for both medical and nursing staff.
iii. All members of staff are invited to attend and give comments on the guideline as part of the ratification process.

f. Review and Revision Arrangements

a. This policy will be reviewed on a 3 yearly basis by the appropriate persons.
b. If new information comes to light prior to the review date, an earlier review will be prompted.
c. Amendments to the document shall be clearly marked on the document control sheet and the updated version uploaded to the intranet. Minor amendments will be ratified through the Paediatric Guidelines Group. A minor amendment would consist of no major change in process, and includes but is not limited to, amendments to documents within the appendices.

g. Equality Impact Assessment

Background • Who was involved in the Equality Impact Assessment
Author and the supervising consultants.
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) • The data sources and any other information used • The consultation that was carried out (who, why and how?)
All groups of staff and patients were taken into consideration and there is no bias towards or against any particular group.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
There is no evidence of discrimination.
Conclusion • Provide a summary of the overall conclusions
There is no evidence of discrimination.
Recommendations • State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
This guideline is appropriate for use.

h. Document Checklist

To be completed (electronically) and attached to any document which guides practice when submitted to the appropriate committee for approval or ratification.

Title of the document: Idiopathic Intracranial Hypertension In Paediatrics

Policy (document) Author: Dr Clare Hill , Dr Mohamed Osman , Dr Alex Nowak

Executive Director: N/A

		Yes/No/ Unsure/NA	Comments
1.	**Title**		
	Is the title clear and unambiguous?	**yes**	
	Is it clear whether the document is a guideline, policy, protocol or standard?	**yes**	
2.	**Scope/Purpose**		
	Is the target population clear and unambiguous?	**yes**	
	Is the purpose of the document clear?	**yes**	
	Are the intended outcomes described?		
	Are the statements clear and unambiguous?	**yes**	
3.	**Development Process**		
	Is there evidence of engagement with stakeholders and users?	**No**	
	Who was engaged in a review of the document (list committees/ individuals)?	**yes**	**Paediatric Guidelines Group**
	Has the policy template been followed (i.e. is the format correct)?	**yes**	
4.	**Evidence Base**		
	Is the type of evidence to support the document identified explicitly?	**yes**	

		Yes/No/ Unsure/NA	Comments
	Are local/organisational supporting documents referenced?	**yes**	
5.	**Approval**		
	Does the document identify which committee/group will approve/ratify it?	**yes**	
	If appropriate, have the joint human resources/staff side committee (or equivalent) approved the document?	**NA**	
6.	**Dissemination and Implementation**		
	Is there an outline/plan to identify how this will be done?	**yes**	
	Does the plan include the necessary training/support to ensure compliance?	**yes**	
7.	**Process for Monitoring Compliance**		
	Are there measurable standards or KPIs to support monitoring compliance of the document?	**yes**	
8.	**Review Date**		
	Is the review date identified and is this acceptable?	**yes**	
9.	**Overall Responsibility for the Document**		
	Is it clear who will be responsible for coordinating the dissemination, implementation and review of the documentation?	**yes**	
10.	**Equality Impact Assessment (EIA)**		
	Has a suitable EIA been completed?	yes	

Committee Approval (Paediatric Guidelines Group)			
If the committee is happy to approve this document, please complete the section below, date it and return it to the Policy (document) Owner			
Name of Chair	**Dr Claire Mitchell**	**Date**	**26/03/2021**
Ratification by Management Executive (if appropriate)			
If the Management Executive is happy to ratify this document, please complete the date of ratification below and advise the Policy (document) Owner			
Date: n/a			