Evaluation and management of adult idiopathic intracranial hypertension

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ABSTRACT
This paper summarises the first consensus guidelines for idiopathic intracranial hypertension as an infographic. Following a systematic literature review, a multidisciplinary specialist interest group met and established questions relating to population, interventions, controls and outcomes (PICO). A survey was sent to doctors who manage idiopathic intracranial hypertension (IIH) regularly. Statements were reviewed by national professional bodies, specifically the Association of British Neurologists, British Association for the Study of Headache, the Society of British Neurological Surgeons and the Royal College of Ophthalmologists. This is the first consensus guidance for optimal management of IIH.

Identification of papilloedema can be challenging, and clinicians should be aware of the differential diagnosis of pseudopapilloedema (figure 1). Once papilloedema is confirmed, it requires urgent investigations, including lumbar puncture, where the patient experience could be greatly improved. Symptoms of IIH are not pathognomonic, and hence it is essential to apply the diagnostic criteria, including excluding secondary causes, for a definite diagnosis. The lumbar puncture opening pressure was one key area of debate. Within the wider Delphi group, it was clear that there is a ‘grey zone’ of lumbar puncture opening pressures between 25 cm cerebrospinal fluid (cmCSF) and 30 cmCSF, as to what each expert considered to be pathological, and this is reflected within the infographic thermometer for lumbar puncture opening pressure (figure 1).

Principles of management need to address both the rapidity of the disease that may lead to visual loss in some and require surgical intervention and the morbidity of the headache that can develop in the majority, which substantially affects the quality of life. Weight loss is currently the only established disease-modifying therapy and is notoriously difficult to achieve and maintain.

Evaluation of the headache phenotype is essential to target treatment and to help identify medication-overuse headache. Where there are features of migraine, topiramate may be the first line in treatment.
Consensus Guideline in Adult Idiopathic Intracranial Hypertension: an infographic summary

Investigation of Papilloedema

Papilloedema identified
- Assess vision
- Record visual acuity
- Pupil examination
- Formal visual fields
- Dilated fundoscopy

Other causes of bilateral disc swelling
- Infection
- Inflammatory
- Hypertension crisis
- Trauma
- Metabolic
- Hereditary
- Ocular

Pseudopapilloedema
- Congenital anomalies
- Optic nerve head drusen
- Hypertensive disc
- Meyerian disc
- Myelinated nerve fibres

Imaging features
- Increased intracranial pressure
- Enlarged optic nerve sheath
- Flattened posterior globe/retina
- Intracranial proptosis of optic nerve
- Abrupt attenuation of ophthalmic veins
- Increased likelihood of being pathologic

Urgent Brain imaging within 24 hours (CT/MR)
+ Venography essential

Consensus Guideline in Adult Idiopathic Intracranial Hypertension

Diagnosis

Symptoms
- Headache
- Visual obscurations
- Pulsatile tinnitus
- Back pain
- Dizziness

Diagnostic criteria
- A. Papilloedema
- B. Normal neurological examination (except sixth nerve palsy)
- C. Neuroimaging: normal brain parenchyma. Venous thrombosis excluded
- D. Normal CSF constituents
- E. Elevated LP pressure >25cmCSF

Principles of management

Management strategies

Protect vision
- Atypical ICH
  - Patients who are not female, or not of childbearing age or who have a BMI below 30kg/m²
- Typical ICH
  - Patients who are female, of childbearing age and who have BMI >30kg/m²

Manage underlying disease
- Weight management for all
- Consider medical therapy with acetazolamide

Reduce headache morbidity
- Fulminant ICH
  - Patients with declining visual function within 4 weeks of diagnosis of ICH
  - Temporising lumbar drain if surgery planned >24 hours
  - CSF diversion
  - Optic nerve sheath fenestration

Weight management
- Headache assessment
  - Ongoing visual assessment
  - Significant deterioration of visual function

Acute Exacerbation of Headache in ICH

- Papilloedema
- Assess vision
- If vision threatened

- Exclude secondary causes and red flags
- No papilloedema

- Emergency room attendance due to headache
- Mandatory assessment of papilloedema

- No papilloedema
- No history required
- Papilloedema flare-up

- No papilloedema

Figure 1  Consensus Guideline in Adult Idiopathic Intracranial Hypertension: an infographic summary.
and recent evidence indicates that it has a significant intracranial pressure-lowering effect in rodents. Acute exacerbation of headache often leads to reinvestigation with lumbar puncture, and the collective expert opinion reflected that lumbar puncture provides only temporary relief, can lead in some to longer term complications and exacerbation of headache. In those with acute exacerbation of headache, optic nerve examination is essential, and in those found not to have papilloedema, investigation with lumbar puncture and brain imaging is not required, so long as no other secondary causes of headache are suspected. The infographic illustrates the management of acute exacerbation of headache in IIH (figure 1).

Horizon scanning for IIH shows that research is active and that metabolic concepts may potentially provide more understanding of the cause and provide evidence for innovative therapeutic opportunities. A phase 2 randomised control trial with the first novel drug treatment for IIH has finished recruitment; a phase 3 randomised control trial investigating the best method for weight loss is underway; other surgical trials are in planning.

This infographic highlights three areas that are covered by the consensus guideline for adult IIH, which are: (1) investigation of papilloedema and diagnosis of IIH; (2) management strategies; and (3) investigation and management of acute exacerbation of headache in established IIH (figure 1).

Key points

- Cerebral venography is an essential part of the work-up to exclude venous sinus thrombosis as a cause of papilloedema.
- Lumbar puncture opening pressure forms part of the diagnostic criteria; however, most clinicians feel there is a ‘grey zone’ between 25 cmCSF and 30 cmCSF, which may not be pathological.
- Those with fulminant or precipitous visual decline need urgent surgical treatment, preferably with a ventriculoperitoneal shunt.
- All patients diagnosed with idiopathic intracranial hypertension need sensitive and appropriate discussion regarding weight loss (the only disease-modifying treatment).
- Those with acute exacerbation of headache do not require further neuroimaging or repeat lumbar puncture, unless there are red flag symptoms/signs of infection, or papilloedema with precipitous visual decline.

Corruption notice This article has been corrected since it was published Online First. The provenance and peer review disclaimer incorrectly said the paper was not commissioned.

Acknowledgements We acknowledge all those who contributed to the surveys and critiqued the document in the wider Delphi group. The specialist interest group members and international key opinion leaders: Brendan Davis (University Hospital North Midlands NHS Trust), Nick Silver (The Walton Centre NHS Foundation Trust), Simon Shaw (University Hospital North Midlands NHS Trust), Conor Mallucci (The Walton Centre NHS Foundation Trust and Alder Hey Children’s NHS Foundation Trust), Ben Wakeley (Gloucestershire Hospitals NHS Foundation Trust), Anita Krishnan (The Walton Centre NHS Foundation Trust), Swapar Chavda (University Hospitals Birmingham NHS Foundation Trust), Sateesh Ramalingam (University Hospitals Birmingham NHS Foundation Trust), Julie Edwards (University Hospitals Birmingham NHS Foundation Trust and Sandwell and West Birmingham NHS Trust), Krystal Hemmings (IIH UK Charity), Shelly Williamson (IIH UK Charity), Mike Burdon (University Hospitals Birmingham NHS Foundation Trust), Ghaniah Hassan-Smith (University of Birmingham), Kathleen Digre (Morgan Eye Center, University of Utah) Grant Liu (Children’s Hospital of Philadelphia and Hospital of the University of Pennsylvania), Rigmor Jensen (Danish Headache Centre, Copenhagen).

Collaborators: Jane Anderson, (Cambridge University Hospitals NHS Foundation Trust), Peter Goadsby (King’s College Hospital), Tim Matthews (University Hospitals Birmingham NHS Foundation Trust) and Jan Hoffman (University Medical Centre Hamburg).

Contributors SPM and AJS drafted the infogram. CH executed the drawing of the infogram. SPM wrote the initial draft. JM and AJS critically reviewed the paper and infogram.

Funding AJS is funded by an NIHR Clinician Scientist Fellowship (NIHR-CS-011-028) and by the Medical Research Council, UK (MR/K015184/1).

Competing interests None declared.

Patient consent Not required.

Provenance and peer review Commissioned; externally peer reviewed by Joseph Anderson, Cardiff, UK and Mark Lawden, Leicester, UK. The guideline was also reviewed, critiqued and supported by the following professional bodies: The Association of British Neurologists, the Society of British Neurological Surgeons, the Royal College of Ophthalmologists and the British Association for the Study of Headache.

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