Evaluation and management of adult idiopathic intracranial hypertension

Susan P Mollan,1,2 Catherine Hornby,1,3 James Mitchell,1,3,4 Alexandra J Sinclair1,2,3,4

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.5 Symptoms of IIH are not pathognomonic, and hence it is essential to apply the diagnostic criteria, including excluding secondary causes, for a definite diagnosis.4 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.6 Weight loss is currently the only established disease-modifying therapy7 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**
  - Record visual acuity
  - Pupil examination
  - Formal visual fields
  - Diluted fundoscopy

- **Assess vision**
  - Optic nerve head (ONH)
  - Hypertensive discs
  - Narrow discs
  - Myelinated nerve fibers

- **Other causes of bilateral disc swelling**
  - Infection
  - Inflammatory
  - Hypertensive crises
  - Trauma
  - Hereditary
  - Toxic
  - Other causes

- **Exclude secondary causes**
  - Anemia
  - Venous thrombosis
  - Drugs
  - Endocrine disease/syndromes
  - Drugs eg fluoroquinolones, tetracyclines, estrogens A analogues, withdrawal of long term glucocorticoids

- **URGENT Brain imaging within 24 hours (CT/MRI)**
  + Venography essential

- **No lesion identified**

- **Exclude secondary causes**

- **Lumbar puncture**
  - CSF opening pressure of >25cmCSF
  - Normal controls

- **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

**Management Strategies**

- **Principles of management**
  - Full re-evaluation

- **Protect vision**
  - Address medication overuse
  - Use single agent
  - Dipyridamole
  - ≥10 days/month

- **Manage underlying disease**
  - Consider medical therapy with acetazolamide

- **Reduce headache morbidity**
  - Fulminant IH
  - Temporizing lumbar drain
  - CSF diversion OR Optic nerve sheath fenestration

**Acute Exacerbation of Headache in IHH**

- **Emergency Room Attendance due to headache**
  - Papilloedema
  - Assess vision

- **Mandatory assessment of papilloedema**
  - No papilloedema
  - No imaging required
  - No LP required

- **Headache flare-up**

**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.

Key opinion leaders: Brendan Davis (University Hospital North Midlands NHS Trust), Nick Silver (The Walton Centre NHS Foundation Trust), Simon Shaw S (University Hospital North Midlands NHS Trust), Conor Mallucci (The Walton Centre NHS Foundation Trust and Alder Hey Children’s NHS Foundation Trust), Ben Wakerley (Gloucestershire Hospitals NHS Foundation Trust), Anita Krishnan (The Walton Centre NHS Foundation Trust), Swarup 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 (Moran 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 Hoffmann (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.

Open access This is an open access article distributed in accordance with the Creative Commons Attribution 4.0 Unported (CC BY 4.0) license, which permits others to copy, redistribute, remix, transform and build upon this work for any

Correction 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

purpose, provided the original work is properly cited, a link to the licence is given, and indication of whether changes were made. See: https://creativecommons.org/licenses/by/4.0/.

REFERENCES