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

- Assess vision
  - Record visual acuity
  - Pupil examination
  - Formal visual fields
  - Dilute fundoscopy
- Check BP
  - Exclude malignant hypertension
  - Venography essential
- URGENT
  - Brain imaging within 24 hours (CT/MRI)
  - + Venography essential
- Lumbar puncture
  - No lesion identified

Other causes of bilateral disc swelling
- Infective
- Inflammatory
- Hypertensive crises
- Torsion
- Neurofibromatosis
- Chordoid meningiomas

Exclude secondary causes
- Anemia
- Venous thrombosis
- Drugs
- Endocrine disease syndromes
- Drugs such as fluoroquinolones, tetracyclines, lamotrigine, and who with withdrawal of long term glucocorticoids

Idiopathic Intracranial Hypertension

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

Symptoms

- Frequency of symptoms reported
  - >65%
  - 40-65%
  - <30%

Diagnostic criteria

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

Principles of management

- Protect vision
- Manage underlying disease
- Reduce headache morbidity
- Full re-evaluation

Weight management

- Temporising lumbar drain
- If surgery planned ≥4 hours

Optic nerve sheath fenestration

Acute Exacerbation of Headache in IIH

- Emergency room attendance due to headache
- Mandate assessment of papilloedema
- No papilloedema
- Assess vision
- Exclude secondary cause and red flags a meningioma

If vision threatened

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.8 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.10 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.11 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.

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References