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 Royal College of Ophthalmologists and by international experts. Key areas are represented based on the guideline, namely: (1) investigation of papilloedema and diagnosis of IIH; (2) management strategies; and (3) investigation and management of acute exacerbation of headache in established IIH. We present an infographic as an aide-mémoire of the first consensus guidelines for IIH.

IIH is commonly associated with obesity, younger age and females. 1, 2 Patients present acutely to many different specialties and often have multiple acute visits through the course of their disease. The investigation and management of IIH is complex involving many specialities. 3 This infographic summarises three key pathways based on the recommendations of a multidisciplinary, patient-involving and multiprofessional specialist interest group on the investigation and management of IIH. 4

The basis of the specialist interest group included representation from neurology, neurosurgery, neuroradiology, ophthalmology, nursing, primary care doctors and patient representatives. Questions on PICO were defined and through a large Delphi group exercise; expertise was captured from a wide-reaching group of clinicians, thus reflecting practice from across the UK and internationally. The statements were then critically reviewed by key opinion leaders and by 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. 4

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 therapy 7 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
Direct fundoscopy

Other causes of bilateral disc swelling
Infectious
Inflammatory
Hypertensive crises
Toxic
Metabolic
Hereditary
Drugs
Obstruction (corpus callosum)

Check BP
Exclude malignant hypertension
>180/120mmHg

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

No lesion identified

Exclude secondary causes

Anaemia
Venous thrombosis
Drugs
Enterine disease/syndromes
Drugs e.g. fluoroquinolones, tetracyclines, intent A analogues, withdrawal of long term glucocorticoids

Lumbar puncture

CSF opening pressure of >25cmCSF normal constituents

Idiopathic Intracranial Hypertension

Symptoms

Headache
Visual obscurations
Pulsatile tinnitus
Back pain
Dizziness

Diagnostic criteria

A. Papilloedema
B. Normal neurological examination (except sixth nerve palsies)
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

Weight management

Atypical IH
Patients who are not female, or not of child bearing age or who have a BMI below 30kg/m²

Typical IH
 Patients who are female, of child bearing age and who have BMI >30kg/m²

Evaluate headache phenotype

Migraine
Medication overuse

Address medication overuse

NSAID/Paracetamol
Antidepressants

Migraine prevention

Options
Topiramate
Carbonate
Beta blockers
\% per nects

Treat migraine headache

Acute migraine attack

Triptan
NSAID/Paracetamol
Antidepressants

Acute Exacerbation of Headache in IH

No papilloedema
No red flags or other secondary causes
No imaging required
No LP required

Emergency Room Attendance due to headache

Mandatory assessment of papilloedema

Papilloedema

Assess vision

If vision threatened

Patient involvement

No papilloedema
Exclude secondary causes and red flags e.g. meningitis

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 intracranial pressure-lowering effect in rodents. Acute and recent evidence indicates that it has a significant exacerbation of headache in IIH (figure 1). 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 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), Sateesh Ramalingam (University Hospitals Birmingham NHS Foundation Trust), Julie Edwards (University Hospitals Birmingham NHS Foundation Trust and Sandwell and West Birmingham NHS Trust), Krystal Hemnings (IIH UK Charity), Shelly Williamson (IIH UK Charity), Mike Burdon (University Hospitals Birmingham NHS Foundation Trust), Ghaniah Hassan-Smith G(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 Hoffman (University Medical Centre Hamburg).

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**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.

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**Competing interests**

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**Patient consent**

Not required.

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REFERENCES