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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 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 specialities 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 therapy⁷ and is notoriously difficult to achieve and maintain.

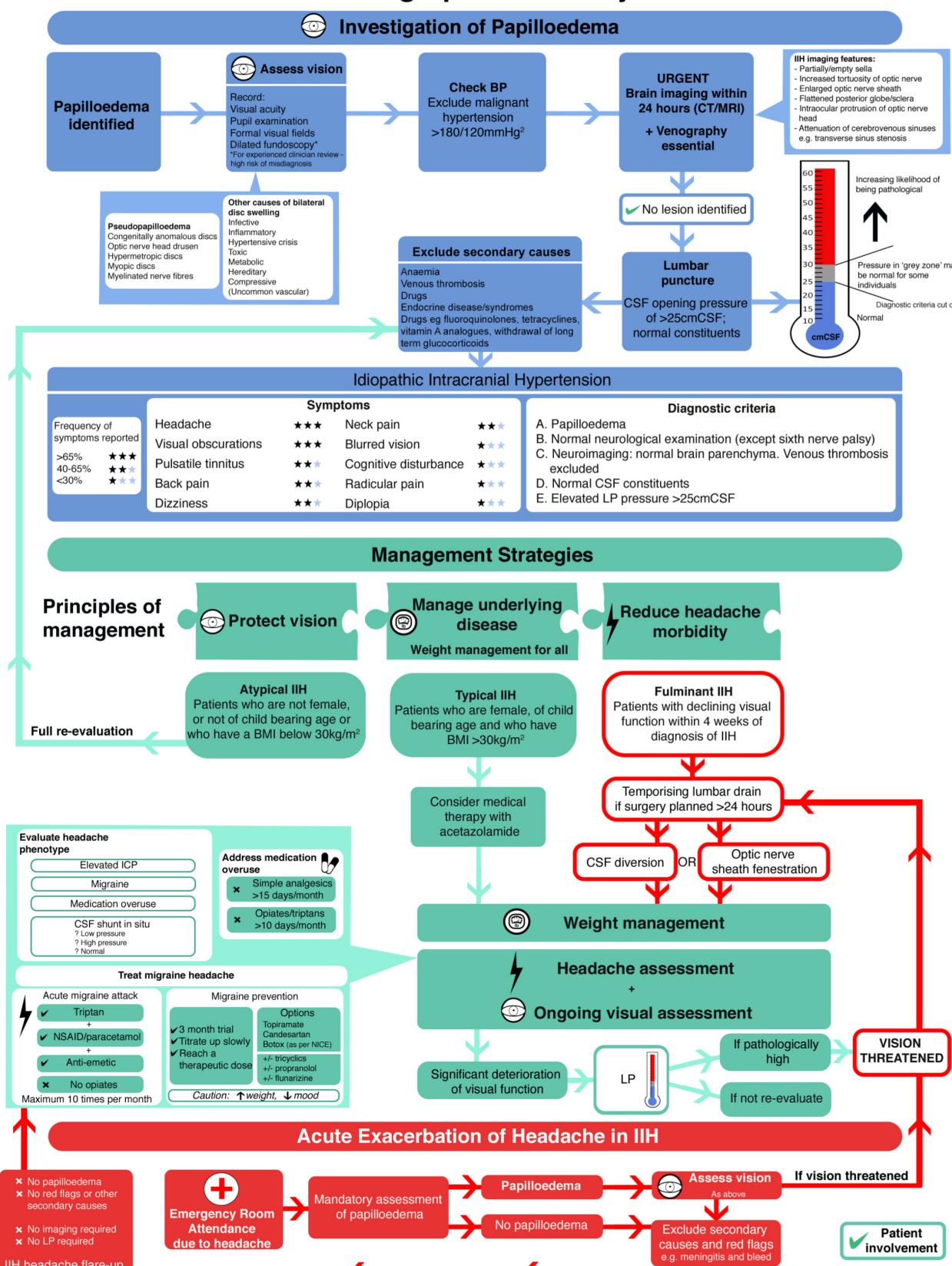
Evaluation of the headache phenotype is essential to target treatment and to help identify medication-overuse headache. Where there are features of migraine, topiramate may be the first line in treatment,



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Consensus Guideline in Adult Idiopathic Intracranial Hypertension: an infographic summary¹



1. Mullan SP, Davis B, Silver NC, Shaw S, Malucci C, Walkerley BR, Krishnan A, Chavda S, Remington S, Edwards J, Hemmings K, Williamson M, Burdon MA, Hassan-Smith G, Digre K, Liu GT, Jensen RH, Sinclair AJ. Idiopathic intracranial hypertension: International consensus guidelines on management. *J Neurol Neurosurg Psychiatry* 2018 (accepted 7/2/18). Endorsed by the Association of British Neurologists, Royal College of Ophthalmologists and Society of British Neurological Surgeons

2. Whelton PK, Carey RM, Aronow WS, et al. 2017 ACC/AHA/ABC/ACPM/AGS/APHA/ASH/ASPC/NMA/PCNA guideline for the prevention, detection, evaluation, and management of high blood pressure in adults: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Hypertension*

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 complications⁹ 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 recruitment¹²; a phase 3 randomised control trial investigating the best method for weight loss is underway¹³; 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 IIH⁴ (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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