Idiopathic Intracranial Hypertension: a review of diagnosis and management

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Abbreviations
IIH – Idiopathic Intracranial Hypertension
CSF – Cerebrospinal fluid
OCT – Optical Coherence Tomography
IIHWOP – Idiopathic Intracranial Hypertension Without Papilloedema
ICP – Intracranial Pressure
BMI – Body Mass Index
LPS – Lumboperitoneal shunt
VPS – Ventriculoperitoneal shunt
ONFS – Optic Nerve Sheath Fenestration
VSS – Venous Sinus Stenting
VF – Visual Fields
VA – Visual Acuity
HA – Headache

Abstract
With the increasing prevalence of obesity, the incidence of idiopathic intracranial hypertension (IIH) is rising. Headache and threat to vision are the predominant features and the principal cause of morbidity and reduced quality of life. Identification of papilloedema must prompt urgent investigation to exclude any underlying cause and management should be multi-disciplinary, focusing on protecting vision and reducing headache burden. Weight loss is the most effective and only disease modifying treatment for IIH but surgical interventions may need to be considered in some patients. Whilst optic nerve sheath fenestration and CSF diversion have established roles in protecting vision, there is increasing interest in venous sinus stenting and bariatric surgery as additional interventions that may have efficacy in the treatment of this condition.

Idiopathic Intracranial Hypertension (IIH) is a disorder of uncertain cause, characterised by raised intracranial pressure in the absence of hydrocephalus or a mass lesion. It typically affects obese women of childbearing age and although a self-limiting condition in the majority of patients, it can be the cause of significant psychological morbidity. Previously known as Benign Intracranial Hypertension, this misnomer failed to recognise the small proportion of patients who develop irreversible, life-changing visual loss, the prevention of which should be the primary concern of those involved in the diagnosis and treatment of the condition. The management of IIH requires a multidisciplinary approach with specialist input from neurology, ophthalmology, dietetics, radiology and neurosurgery. Patients with IIH will present via primary care and acute hospital services, and therefore a broad understanding of the condition is essential for both General Practitioners and Emergency Physicians. The purpose of this review is to summarise the diagnostic and management challenges and to highlight best practice guidelines as well as current trends and controversies.

Epidemiology

The incidence of IIH varies around the world and is related to the prevalence of obesity in the general population. The estimated global incidence is 0.14 per 100,000 per year in populations with an obesity prevalence of <10% but rises to 1.48 per 100,000 per year in populations where the prevalence of obesity is >20% [1]. Recent large studies from the UK further highlight the relationship of both obesity and deprivation with IIH. In Wales, where obesity prevalence has increased from 29% in 2003 to 40% in 2017, the prevalence of IIH has increased six-fold to 76 per 100,000 and the incidence three-fold to 7.8 per 100,000 per year. This peaks at 180 per 100,000 and 23.5 per 100,000 per year in obese women [2]. A similar trend is found in England [3,4]. More than 80% of patients are female and 85% are under 45 years old at diagnosis (mean 28-30 years) [1-3]. In men, the mean age at diagnosis is slightly higher (32-33 years) but the proportion of cases diagnosed in childhood is also higher (approximately 30% of male cases, 10% of female cases). Social deprivation, even after adjusting for obesity, has an association with IIH, with fewer cases identified in the least deprived geographical areas. Interestingly, whilst the association with obesity is seen across both female and male patients, the relationship with deprivation is only a feature of female cases [2,3]. Based on current trends, the cost of treating IIH in English hospitals is projected to be $462million by 2030, nearly a 50-fold increase since 2002 [3].

Diagnostic criteria

Both the modified Dandy [5] and Friedman criteria [6] are widely used in the diagnosis of IIH. A modified version of the Friedman criteria, adopted by a consensus of UK experts [7] offers a pragmatic approach for clinicians (Table 1).

Clinical features and investigations

Headache is the predominant symptom and the major cause of morbidity and reduced quality of life [5,7-9]. The typical raised intracranial pressure headache (positional, early morning, exacerbated by Valsalva manoeuvre) is not universal and studies indicate that migraine (chronic or episodic) is the most common phenotype [8]. Headache of IIH may coexist with chronic daily headache, medication over-use headache and migraine and is frequently associated with other symptoms including photophobia, phonophobia, visual obscurations and diplopia (with or without abducens palsy). Pulsatile tinnitus (thought to be caused by turbulent flow in the intracranial venous system) is also commonly reported, as is neck, back and radicular pain (Table 2) [9].

Table 1: Diagnostic criteria for IIH

Modified Dandy Criteria [5]

1. Signs and symptoms of increased intracranial pressure
2. Absence of localising findings on neurologic examination
3. Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neurodiagnostic studies, except for evidence of increased cerebrospinal fluid pressure (>200 mm water). Abnormal neuroimaging except for empty sella turcica, optic nerve sheath with filled out CSF spaces, and smooth-walled non flow-related venous sinus stenosis or collapse should lead to another diagnosis
4. Awake and alert
5. No other cause of increased intracranial pressure present

Modified Friedman Criteria [6]

1. Required for diagnosis of pseudotumor cerebri syndrome*
   - A. Papilloedema
   - B. Normal neurologic examination except for cranial nerve abnormalities
   - C. Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used
   - D. Normal CSF composition
   - E. Elevated lumbar puncture opening pressure (≥250mmCSF in adults and ≥280mmCSF in children [250mmCSF if the child is not sedated and not obese]) in a properly performed lumbar puncture

A diagnosis of pseudotumor cerebri syndrome is definite if the patient fulfills criteria A–E. The diagnosis is considered probable if criteria A–D are met but the measured CSF pressure is lower than specified for a definite diagnosis.

2. Diagnosis of pseudotumor cerebri syndrome without papilloedema
   In the absence of papilloedema, a diagnosis of pseudotumor cerebri syndrome can be made if B–E from above are satisfied, and in addition the patient has a unilateral or bilateral abducens nerve palsy
   In the absence of papilloedema or sixth nerve palsy, a diagnosis of pseudotumor cerebri syndrome can be suggested but not made if B–E from above are satisfied, and in addition at least 3 of the following neuroimaging criteria are satisfied:
   - i. Empty sella
   - ii. Flattening of the posterior aspect of the globe
   - iii. Distension of the perioptic subarachnoid space with or without a tortuous optic nerve
   - iv. Transverse venous sinus stenosis

**Pseudotumor cerebri syndrome: synonym for IIH used predominantly in the USA*

UK Consensus Criteria adapted from Friedman Criteria [7]

1. IIH Diagnostic criteria
   - A. Papilloedema
   - B. Normal neurological examination (except sixth cranial nerve palsy)
   - C. Neuroimaging: normal brain parenchyma (no hydrocephalus, mass, structural lesion or meningeal enhancement). Venous sinus thrombosis excluded in all.
   - D. Normal CSF constituents
   - E. Elevated lumbar puncture pressure ≥25cmCSF
2. IIH without Papilloedema (IIHWOP) Diagnostic criteria
   Presence of criteria B-E for IIH plus:
   Unilateral or bilateral sixth cranial nerve palsy
3. Suggestion of possible IIH without Papilloedema (IIHWOP) Diagnostic criteria
   Presence of criteria B-E for IIH plus:
   Three neuroimaging findings suggestive of raised intracranial pressure
   - Empty sella
   - Flattening of posterior aspect of the globe
   - Distension of the peri-optic subarachnoid space ± a tortuous optic nerve
   - Transverse venous sinus stenosis

Table 2: Symptom frequency in IIH adapted from Markey et al [9]

Symptom	Frequency (% of patients)
Headache	75–94%
Nausea with or without vomiting	42–73%
Photophobia, phonophobia, or both	72–75%
Transient visual obscurations	68–72%
Pulsatile tinnitus	52–60%
Back pain	53%
Dizziness	52%
Neck pain	42%
Visual loss or blurring	32%
Cognitive disturbances	20%
Radicular pain	19%
Horizontal diplopia	18%

By definition, papilloedema (optic disc swelling due to raised intracranial pressure) is present and its identification must prompt urgent action to find any treatable cause and protect vision. Initial assessment should include visual acuity, formal visual fields and dilated fundoscopy; early ophthalmology assessment including retinal photography and optical coherence tomography (OCT) is also recommended [7]. OCT provides quantitative, objective measurements of papilloedema which complements clinical assessment particularly as visual fields can be unreliable and non-organic visual loss can coexist [10,11]. It is important not to be falsely reassured by normal visual acuity which can be maintained despite significant loss of the peripheral visual field. Blindness from IIH is rare (0.42-2% [2,3,10] but milder visual impairment is more common. Visual loss may occur at any time, be insidious and asymptomatic so careful monitoring for detection of subclinical visual field loss is essential. The reported incidence of any reduction in visual acuity or field is wide-ranging [9] and the most common field abnormalities are blind spot enlargement, constriction, nasal loss and arcuate defects [12]. A subset of patients who meet all the other criteria for IIH but do not have papilloedema (IIHWOP) is described and these patients do not appear to have a risk to vision [6,7] but should be monitored for the development of papilloedema.

All patients with papilloedema should undergo cranial imaging (MRI with venography or CT with venography) within 24 hours to rule out a mass lesion, hydrocephalus or venous sinus thrombosis before proceeding to lumbar puncture [7]. Lumbar CSF pressure correlates well with intracranial pressure [13] but should be performed in the lateral decubitus position and allowed to settle with the legs extended prior to manometry to avoid an artificially raised pressure. An opening pressure of greater than 25cmH_20 is required to meet the diagnostic criteria although, given diurnal variations in ICP, a single reading should be interpreted with caution when it does not fit the clinical picture [7]. Where doubt exists or lumbar puncture contraindicated (e.g. due to coexisting Chiari malformation), intracranial pressure monitoring can be considered.

Other secondary and potentially reversible causes of raised ICP, including endocrine dysfunction, iron deficiency anaemia and medications (Table 3) should be ruled out, particularly when the patient is not the typical phenotype (non-Caucasian, male or BMI<30Kg/m^2). Associated conditions including sleep apnoea, polycystic ovary syndrome and risk of cardiovascular disease should also be considered [4-7].

Imaging findings

Radiological stigmata are summarised in Table 4 and have high specificity but relatively low sensitivity for IIH, other than transverse sinus stenosis which is both sensitive and specific. However, imaging

Table 3: Associations and secondary causes of raised ICP, adapted from Markey et al. [9] and Mollan et al. [7]	
Secondary causes of raised intracranial pressure	
Iatrogenic	Antibiotics Tetracycline, minocycline, doxycycline, nitrofurantoin, sulphonamides, and nalidixic acid Hormones Levonorgesterel implant, thyroxine, growth hormone, corticosteroids and tamoxifen Vitamin A (including isotretinoin) Other drugs Lithium, ciclosporin, indomethacin, cimetidine
Haematological	Anaemia Polycythaemia vera
Respiratory	Obstructive sleep apnoea Hypercapnia, Chronic Obstructive Pulmonary Disease
Endocrine	Addison's disease Cushing's syndrome Adrenal insufficiency Thyroid & parathyroid dysfunction
Renal	Chronic kidney disease/renal failure
Autoimmune	Systemic Lupus Erythmatosus Sjögren's syndrome
Syndromic	Down Syndrome Craniosynostosis Turner Syndrome
Venous outflow obstruction	Central venous sinus thrombosis Jugular vein thrombosis Superior Vena Cava syndrome

Table 4: MRI signs diagnostic of IIH, adapted from Kwee and Kwee [14]		
MRI Sign	Sensitivity	Specificity
Empty Sella	62.2	90.7
Posterior displacement of pituitary stalk	41.2	84.0
Meningocoeles	9.3	99.2
Posterior globe flattening	56.3	95.3
Optic nerve head protrusion	29.1	97.0
Optic nerve enhancement	13.2	95.9
Optic nerve sheath distension	68.6	86.1
Optic nerve tortuosity	36.9	88.4
Transverse sinus stenosis	84.4	94.9
Slit-like ventricles	14.5	89.9
Tight subarachnoid spaces	6.1	97.1
Inferior position of cerebellar tonsils	19.2	92.8

findings should be correlated with clinical features of IIH as these signs may be present in the absence of raised intracranial pressure. For example, pituitary height and globe configuration may persist after normalisation of ICP and resolution of papilloedema [14] and primary empty sella has been reported in 8-35% of the general population [15].

Medical and non-surgical management

The only disease modifying treatment in typical IIH is weight loss. Up to 15% weight loss may be required to achieve remission so patients should receive support from dietetics and weight management programmes [7]. There is increasing evidence for the role of bariatric surgery, which is discussed below.

Medical therapies are used to protect vision and reduce headache burden. Acetazolamide, a carbonic anhydrase inhibitor that reduces CSF production, is frequently used in an attempt to protect vision but side effects including paraesthesia, fatigue, diarrhoea, alterations in taste (dysgeusia), nausea and vomiting can result in the drug being poorly tolerated. In the IIH Treatment Trial, a double-blind randomised placebo-controlled study, acetazolamide in combination with a low-sodium weight-reduction diet, had a modest effect on visual field function and improved quality of life measures [5] however the evidence to either recommend or reject its use remains insufficient [16]. Other diuretics including furosemide are sometimes used although efficacy remains uncertain [7]. Topiramate, which has some inhibitory effect on carbonic anhydrase, is a mild appetite suppressant and is efficacious in episodic migraine, has a role in some patients [8].

Management of headache can be challenging so involvement of a clinician with experience in headache management is important, particularly as multiple headache phenotypes, particularly migraines, may coexist [7]. Triptans, topiramate, non-steroidal anti-inflammatories, paracetamol, beta-blockers, tricyclic anti-depressants and sodium valproate are amongst the range of therapies used, however high-quality evidence to guide decision making in IIH is lacking [8]. Patient education about the use of analgesia to avoid medication over-use symptoms is essential and opioid medications should be avoided [7].

Whilst most patients will have relief of headache following therapeutic lumbar puncture, the effect is short lived as CSF is rapidly replenished. Serial lumbar puncture, which can lead to chronic back pain, is not recommended for headache although may be used as a temporising measure when there is concern about rapidly progressing visual loss [7].

Surgical management

Surgical interventions are typically reserved for patients with visual loss refractory to medical treatments and include CSF diversion with a ventriculoperitoneal (VPS) or lumboperitoneal shunt (LPS) or with optic nerve sheath fenestration (ONSF). There is increasing evidence regarding the efficacy of endovascular venous sinus stenting (VSS) [17-19] and bariatric surgery [20-22], but these are not currently routine practice for IIH in most centres in the UK. There are no controlled trials comparing any of these interventions in IIH and the reported outcome measures in the literature are very heterogenous, making comparisons problematic [23].

Patients should be referred for urgent CSF diversion or ONSF when vision is imminently threatened (Fulminant IIH) or when there is a significant deterioration in visual function despite optimal medical management. CSF diversion for headache alone is rarely recommended and should only be considered as part of a multidisciplinary approach following a period of intracranial pressure monitoring [7].

Optic nerve sheath fenestration

The exact mechanism by which ONSF relieves papilloedema is not fully understood. It is thought that decompression of the perioptic subarachnoid space allows CSF egress and reaccumulation is prevented through scarring [24]. ONSF is very successful at improving papilloedema (90.5%) and visual fields (65.2%) but has less effect on improving visual acuity (44.1%) and headaches (49.3%). It has a favour-

Table 5: Summary of outcomes of surgical procedures for IIH, adapted from Kalyvas et al [23]. Figures are percentage of patients for each outcome

Procedure (pooled n)	Improved VF %	Improved VA %	Improved HA %	Improved Papilloedema %	Failure Rate %	Severe complications %
ONSF (n=818)	65.2	44.1	49.3	90.5	9.4	2.2
CSF Diversion (n=609)	66.8	55	69.8	78.9	43.4	9.4
VSS (n=825)	72.7	64.6	72.1	87.1	11.3	2.3
Bariatric surgery (n=50)	83.3	100	99.3	100	10.8	29.4

VF Visual Fields, VA Visual Acuity, HA Headache

able safety profile with only 2.2% suffering a severe complication, however a significant proportion (16.9%) go on to have additional interventions such as CSF shunting due to inadequate symptom control [23]. It is only performed in a few centres in the UK but is more widespread elsewhere in the world.

CSF diversion

Shunting is the most frequently performed surgical intervention in IIH, with approximately 8% of patients having a shunt procedure [2,3]. A shunt is a tube that diverts CSF from the ventricle or lumbar subarachnoid space to another body space where it is absorbed into the bloodstream. The shunt contains a valve which controls the amount of fluid passing through the tubing. Most commonly, the distal tube is placed in the abdominal cavity, although the pleural space and right atrium of the heart can also be utilised.

CSF diversion is effective in improving papilloedema (78.9%) visual fields (66.8%) and visual acuity (55%) but shunt failure and complications are high [23]. A lumboperitoneal shunt may appear to be the less invasive option and it does avoid the obligatory temporary disqualification from driving, however due to a lower revision rate, VPS is usually recommended for visual deterioration in IIH [7]. Furthermore, LPS can cause an iatrogenic Chiari malformation. Complications related to both VPS and LPS include infection, shunt obstruction, catheter migration/malposition/disconnection, over-drainage, intracranial haemorrhage, slit ventricle syndrome and abdominal pain [23]. Obese patients are at significantly higher risk of distal catheter migration, where the abdominal catheter coils in the subcutaneous tissue and requires surgical reimplantation [25]. Nearly 10% of patients suffer a severe complication and more than 40% have failure of their shunt. Many patients require multiple shunt operations, with a mean of 2.6 revisions per patient [2,23].

Headaches frequently initially improve following CSF diversion although iatrogenic low-pressure headaches occur in 28% of patients [9]. Programmable valves can be adjusted using an external magnet allowing the opening pressure to be titrated to patient symptoms. The addition of an anti-gravity (which alters the opening pressure according to body position) or anti-siphon (which prevents excessive drainage during postural changes) device can further protect against over-drainage and low-pressure symptoms. A telemetric pressure sensor can be added proximal to the shunt valve to help guide valve programming and provides useful diagnostic data in the assessment of suspected shunt malfunction. Although meta-analysis indicates CSF diversion is effective for the treatment of headaches (Table 5) [23] this may not be sustained and patients can continue to have significant headache morbidity despite a functioning shunt and being in ocular remission. In one series 79% of shunted patients had persistent headaches at 2 years [26], with chronic daily headache, migraine, medication over-use, and low pressure symptoms contributory [7,8]. A third of patients have multiple hospital admissions in the year following diagnosis [3] and unscheduled hospital admissions are twice as high in those who have undergone CSF diversion compared to those who have not [2]. Severe headache is the reason for presentation in 79% [27] and unfamiliarity with the unique challenges of managing IIH can lead to unnecessary imaging and interventions. Current UK consensus guidelines advise against CT, shunt x-ray or lumbar puncture in shunted patients in the absence of papilloedema or suspicion of infection, with a recommendation to focus on evaluating the headache phenotype, eliminating medication over-use aspects and offering appropriate medical therapies [7].

Endovascular venous sinus stenting (VSS)

VSS has emerged as a possible treatment in medically refractory IIH based on the belief that venous outflow obstruction from transverse sinus stenoses is at least partly contributory to its aetiology. This has been contentious however as CSF diversion or removal (with reduction in ICP) has been demonstrated to reverse venous sinus stenosis indicating that stenosis is the result of raised ICP not the cause [9]. There may, in fact, be two distinct types of venous sinus stenoses: extrinsic compression (termed non-venogenic) as a consequence of raised ICP and less commonly, intrinsic focal venous stenosis (venogenic type) typically due to arachnoid granulation hypertrophy, fibrosis or deposition. As it is now widely accepted that venous hypertension and stenosis may play a role in the pathophysiology there is a compelling argument that when surgical intervention is considered necessary, VSS should be considered above ONSF or CSF diversion as it is the only intervention to act directly on venous sinus haemodynamics [28]. Careful patient selection is critical and requires venography, manometry and measurements of pressure gradients across the stenotic sinus to determine suitability for stent placement [18,23]. Outcomes from uncontrolled studies are very favourable in terms of visual function and headache improvements, particularly given the lower complication and failure rate compared to CSF diversion (Table 5) [17,18,23] but selection bias in these studies is likely. Whilst VSS is an important part of the armamentarium in some neurosurgical centres it is not universally available in the UK and high quality evidence is required to further support its use in the treatment of either visual deterioration or headache in IIH [7,28].

Bariatric surgery

There has been much interest in a possible role for bariatric surgery in IIH given that it has been demonstrated to be a successful, cost-effective and safe treatment for severe obesity when conservative strategies fail [21,22]. Meta-analyses [20,21,23] of small uncontrolled studies in IIH have reported significant improvements in BMI, CSF opening pressure, headache and papilloedema however complication rates are high and evidence quality inadequate to draw firm conclusions from. Most recently however, the IIH Weight Trial, a multi-centre parallel-group randomised clinical trial has reported that bariatric surgery is superior to community weight management intervention in lowering intracranial pressure. Importantly this effect, as well as significantly lower weight in the surgery group, is sustained at two years and is associated with improved quality of life measures. Whilst there was no significant difference between the two groups in terms of headache disability or visual function the additional weight loss achieved in the surgery group has a favourable impact on disease remission as well as a wide range of additional health benefits [22].

Conclusions

With the increasing prevalence of obesity, the incidence of IIH will continue to rise presenting both clinical and service provision challenges.

Clinicians should be mindful of the principles of protecting vision and reducing headache burden and awareness of current guidance will facilitate more consistent and evidence-based care. A multidisciplinary approach involving clinicians with experience in managing IIH is required for the diagnosis, surveillance and treatment of this condition and management strategies should focus on disease modification through weight loss. CSF diversion remains the principal surgical treatment to protect vision when non-surgical strategies fail but the long-lasting implications of this in terms of hospital admissions, complications and revision surgery should not be underestimated. There is however increasing evidence for the role of venous sinus stenting and bariatric surgery which may support a wider range of surgical interventions. High quality research is needed to better understand both medical and surgical treatment options with a view to improving visual outcomes, headache morbidity and quality of life.

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