Review

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Diagnosis and treatment of idiopathic intracranial hypertension

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Naz Raoof[1,2] and Jan Hoffmann[3,4]

Abstract

Objective: To review and discuss the clinical presentation and treatment of idiopathic intracranial hypertension.

Discussion: Visual alterations and headache are the two main symptoms of idiopathic intracranial hypertension, although additional features including cranial nerve palsies, cognitive deficits, olfactory deficits and tinnitus are not uncommon. The headache associated with idiopathic intracranial hypertension frequently has a migrainous phenotype. The underlying cause of the disorder has not yet been elucidated. Several hypotheses have been postulated but none of them can explain the full clinical picture. Therapeutic options remain limited, focusing mainly on reduction in body weight and the reduction of CSF production with carbonic anhydrase inhibitors.

Conclusion: The accurate diagnosis of idiopathic intracranial hypertension is essential as visual deterioration due to papilledema may be irreversible. Given its phenotypic similarity and frequent overlap with chronic migraine it is essential to consider idiopathic intracranial hypertension in the diagnostic workup of chronic headache; in particular, when considering its increasing prevalence. Understanding in detail the pathophysiological mechanisms behind the associated headache would also allow study of current and future therapeutic options in a structured way.

Keywords

Idiopathic intracranial hypertension (IIH), headache, pain, papilledema

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Idiopathic intracranial hypertension (IIH) is caused by an elevation of intracranial pressure (ICP). The condition mainly affects obese young women of childbearing age. Its prevalence ranges between 0.5 and 2 per 100,000 of the general population and is expected to increase further given the worldwide increase in obesity (1). The underlying cause of the disease, as well as the gender preference and its pathophysiological relation to obesity, remain largely unknown. Several mechanisms have been proposed as the underlying cause of IIH, such as an overproduction of cerebrospinal fluid (CSF), outflow obstruction, elevated pressure in the venous sinuses and more recently a dysfunction in the glymphatic pathway as well as hormonal alterations. Given that none of these theories can explain the entire clinical picture, therapeutic approaches rely mainly on weight reduction and the decrease of CSF production with carbonic anhydrase inhibitors (2). As increasing evidence suggests that a CSF overproduction is unlikely to be the driving factor behind IIH, treatment with acetazolamide is likely to target a consequence, rather than the cause of this disorder.

The two most prominent symptoms of IIH are progressive visual deterioration resulting from papilledema and chronic headache, although additional symptoms including cranial nerve palsies, cognitive deficits,

[1]Department of Paediatrics, Strabismus and Neuro-ophthalmology, Moorfields Eye Hospital, London, UK
[2]Department of Ophthalmology, Royal London Hospital, Barts Health NHS Trust, London, UK
[3]Wolfson Centre for Age-Related Diseases, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, UK
[4]NIHR-Wellcome Trust King's Clinical Research Facility/SLaM Biomedical Research Centre, King's College Hospital, London, UK

Corresponding author:
Jan Hoffmann, Wolfson Centre for Age-Related Diseases, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London SE5 9PJ, UK.
Email: jan.hoffmann@kcl.ac.uk

tinnitus and olfactory dysfunction are frequently also part of the clinical presentation. While the visual dysfunction is known to largely result from a pressure-induced papilledema, the origin of the IIH-related headache is less clear and therapeutic approaches are less investigated.

IIH-related headache is defined in the Headache Classification of the International Headache Society (ICHD-3) (3). While the ICHD has had an enormous impact on advancing the understanding and treatment options of many headache disorders, as it allowed the creation of homogenous patient groups resulting in improved diagnostic accuracy and more effective clinical trials, ICHD has had less of an impact in IIH-related headache. The reason is a rather unspecific definition of IIH-related headache resulting from its highly variable clinical presentation that may frequently mimic or overlap with primary headaches, in particular migraine (4). As a result, while the understanding of IIH is advancing, unfortunately this is not so much the case in IIH-related headache.

The aim of this review is to discuss the most prominent visual symptoms and IIH-associated headache as well as the potential therapeutic options that exist to treat these two features of IIH.

Visual features of IIH

Transient visual obscurations (TVOs) refer to loss or clouding of the vision in one or both eyes, lasting for a minute or less. The cause has been suggested to be raised ICP at the optic nerve causing transient ischemia of the optic nerve head (5). While not specific for raised ICP, a questionnaire-based study of 50 patients with IIH and 100 controls reported that while this symptom was reported by both patients and controls (68% of IIH patients versus 21% of controls, odds ratio 8 (95% CI 3.7–17.1)), daily occurrence of these symptoms was much more common in IIH patients with an odds ratio of 66.7 (CI 60–550) (6). Postural changes were described as a provoking factor in 50% of cases. Similarly, 68% of 165 patients enrolled in the IIH Treatment Trial (IIHTT) reported TVOs, with a median of one episode per day (7). These studies suggest daily TVOs are useful clinical indicators of raised intracranial pressure and therefore also IIH.

Diplopia is another symptom encountered in patients with IIH. The IIHTT reports that 18% of their cohort experienced diplopia, but the nature of the diplopia is not further characterised in this study (7). Guiseffi et al. report a higher incidence of diplopia in patients with IIH at 38%, but again the nature of this is not stated. Sixth nerve palsy is thought to be the most common cause of diplopia in patients with IIH, with one study reporting 14 cases in a cohort of 101 patients (8). These patients describe horizontal diplopia that is binocular in nature (closure of either eye eliminates the diplopia). The long course of the sixth nerve, including the relationship to the apex of the petrous temporal bone as well as its ascending course between the pons and clivus to enter Dorello's canal, have been given as reasons for the vulnerability of the sixth nerve to being both compressed and stretched as a consequence of raised intracranial pressure. Third and fourth cranial nerve palsies have been described in patients with IIH, but are rarer. Patients can describe a non-specific visual blurring as "double vision", but a report of binocular diplopia should alert a clinician to the possibility of a disturbance in ocular motility associated with raised ICP.

Visual loss in IIH can manifest as loss in visual field or loss of visual acuity. An enlarged blind spot is well recognised as a common early visual field defect in raised ICP. In a cohort of 82 patients, where the majority had Frisen grade 2 papilledema, 80% of visual fields showed an enlarged blind spot while 72% had a nasal defect (9). The IIHTT describes similar co-existent visual fields defects in their baseline population, with an enlarged blind spot and inferior partial arcuate defects as the commonest changes in this cohort with mild disease (10). Loss of visual acuity is generally accepted to be a feature of advanced disease, although fulminant IIH (severe visual loss within 4 weeks from symptoms onset) is a well-recognised, albeit rare, form of the condition that clinicians must be alert to as it requires prompt surgical intervention to prevent marked permanent visual deficits (11). However, there is evidence from two large studies that visual acuity can be affected even in relatively mild disease. Pollack et al. report that 17% of eyes had a visual acuity of 0.2 logMAR or worse, while in the IIHTT 29% of study eyes had a visual acuity worse than 20/20 (equivalent to 0.0 logMAR) despite accounting for the patients' refractive error. Taken together, these findings broadly suggest that visual field assessment may be more sensitive than visual acuity assessment in detecting visual deficits in IIH. This is supported by Rowe et al. in a study of 35 patients with IIH, who documented an abnormal visual field assessment in 103 of 110 tests (94%) but abnormal visual acuity in 36 tests (33%), giving a value of $f = 0.0047$ (F-test) (12). It should be stated, however, that this was a smaller study based on 35 patients, a significant proportion of whom showed more severe disease (approximately 25% of patients showed moderate visual field loss on Goldmann visual field testing).

Papilledema is encountered in almost all patients with IIH and is therefore a key feature in assessing patients suspected of having this condition. A large retrospective study of 353 patients with IIH identified

only 20 patients who presented without papilledema (5.7%) (13). It is important for clinicians to be aware that while mostly bilateral, papilledema may be asymmetrical. Indeed, the IIHTT found that 7% of patients had a difference of 2 Frisen grades or more in the papilledema grading between the two eyes (7). When assessing an optic nerve for the likelihood of papilledema, the presence of spontaneous venous pulsations has traditionally been considered to exclude the possibility of raised ICP. However, a study by Wong et al. reports that SVP were visible in 12 of 20 IIH patients with mild papilledema, five of whom had an opening pressure on lumbar puncture of >30 cmH_2O (14). Although this represents a subgroup analysis and is limited by the small sample size, this study serves as a reminder that the presence of SVP does not invariably rule out raised ICP. If untreated, papilledema can result in optic atrophy in patients with IIH, although the associated visual deficits mean the diagnosis is often easy to make at this late stage.

Headache in IIH

As described above, chronic headache is another key symptom of IIH. Headache represents the most common reason for IIH patients to seek medical advice as it has a profound impact on the patient's quality of life (15,16). While the definition of IIH-related headache of the International Headache Society is relatively unspecific (3), it frequently has a migrainous phenotype (7,17–20). Therefore, the differentiation between chronic migraine and IIH-related headache may be challenging (4,21). This is complicated even further due to the fact that in many IIH-patients, headache persists in the long-term despite a normalisation of CSF pressure (22,23). The IIHTT and other studies revealed that there is no correlation between CSF pressure and headache presence or intensity (7,17,24). The transient improvement after lumbar puncture is not exclusive to IIH-related headache, as it is also observed in other chronic headaches (24). When IIH-related headaches have a migrainous phenotype, the accompanying symptoms such as nausea, photophobia and phonophobia also tend to improve after lumbar puncture (25). For these reasons, a lumbar puncture-induced transient improvement of headache and accompanying symptoms does not allow the pathophysiological origin of IIH-related headache to be distinguished. It is clear that attributing it simply to the direct effect of elevated CSF pressure falls short as a plausible explanation, at least in the majority of IIH patients. Understanding the pathomechanism of IIH-related headache, including the involved neuronal networks and molecular mediators, would allow us to understand and reliably distinguish whether, in an individual patient, IIH-related headache is its own entity or if it is the result of a CSF pressure-induced aggravation of a pre-existing migraine, or if it may even unmask a migraine that had been previously been clinically inapparent. This differentiation may also allow understanding of why headache persists in some patients after normalisation of CSF pressure while it does not in others. It may be speculated that the subgroup of patients in which headache persists despite normalisation of CSF pressure may in fact have an additional migrainous biology, which may be the driving force behind headache chronification and maintenance of post-IIH headache. Unfortunately, to date there is no structured clinical study that answers this question in detail although this knowledge would be essential for the therapeutic management of IIH-related headache. When assessing the efficacy of a specific medication for the treatment of IIH, in contrast to the improvement in papilledema and visual function, the effect on IIH-related headache is much more difficult to assess given the complexity around the origin of IIH-related headache and its frequent overlap with chronic migraine.

Treatment of IIH

As stated in the IIH Guidelines (26,27), the main principles of management can be summarised as i) treatment of the underlying disease, ii) protection of vision, and iii) minimisation of the headache morbidity.

While the cornerstone of treating the underlying disease is weight loss, acetazolamide is typically used to lower ICP in most cases of IIH, relieving pressure on the optic nerves and allowing time for weight loss to occur. Acetazolamide inhibits carbonic anhydrase, thereby reducing production of CSF by the choroid plexus. The IIHTT was a randomised, multicentre, double-blinded trial designed to investigate the efficacy of acetazolamide (up to the dose of 4 g) versus placebo in IIH patients with mild visual loss (2). To date, it remains the only randomised-controlled trial for the pharmacological treatment of IIH. All patients were also on a low-sodium weight reduction diet. Perimetric mean deviation (PMD), a measure of global field loss, was used as the primary outcome variable in this study. This is the mean deviation from age-corrected normal values, with larger negative values indicating greater visual loss. Individuals receiving acetazolamide had a greater improvement in mean PMD compared with those receiving placebo at 6 months, with a statistically significant difference of 0.71 dB (95% CI, 0 to 1.43 dB; $p=0.050$). There was also a statistically significant improvement in mean papilledema grade (acetazolamide: -1.31, from 2.76 to 1.45; placebo: -0.61, from 2.76 to 2.15; treatment

effect, −0.70; 95% CI, −0.99 to −0.41; $p < 0.001$). Of note, both groups also lost weight, although the group receiving acetazolamide lost more weight than those receiving placebo (−7.50 kg vs. −3.45 kg; $p < 0.001$). Headache severity was assessed using the six-item Headache Impact Test (HIT-6), but no significant treatment effects were noted in this study. At 6 months, headaches were reported by 69% of the acetazolamide group versus 68% of participants in the placebo group ($p = 0.80$). One other randomised open-label study of acetazolamide versus no acetazolamide has been conducted but had only 25 patients in each arm and is therefore not sufficiently powered (28).

Topiramate is gaining increasing popularity as a therapeutic option for IIH, in light of its actions as an anti-migraine and appetite suppression agent, as well as its ability to inhibit carbonic anhydrase (1). To date, there are no controlled studies into the role of topiramate, although one open-label study comparing topiramate and acetazolamide has been conducted (29), in which 41 patients with IIH were alternately assigned to receiving either acetazolamide (1000–1500 mg per day) or topiramate (100–150 mg per day). One patient had rapidly worsening vision and dropped out of the topiramate group after 1 week to undergo CSF diversion surgery. Visual field grading was used to assess patients 3, 6 and 12 months after treatment. In both groups, there was a statistically significant difference in visual field grading at 12 months when compared with the initial visual field. There was no statistically significant difference between the two arms at 12 months, however.

For the management of IIH-related headache it has to be considered that a migrainous phenotype is the most common presentation of IIH-related headache. From a headache perspective, these patients may be treated with a migraine preventive (26,27). In general, when selecting an appropriate migraine preventive for these patients, the use of medications that may induce weight gain should be only be used with caution. In the context of IIH, beyond its efficacy as a migraine preventive, topiramate inhibits carbonic anhydrase thereby reducing CSF pressure, as outlined above, and frequently causes weight loss. Therefore, topiramate would be particularly attractive as it would target IIH-related headache while at the same time reducing CSF pressure. However, it has to be taken into consideration that topiramate may induce cognitive side effects and depression, two common comorbidities in IIH. In these patients, its use should be considered with caution.

When managing headache in IIH, the overuse of acute medication (pain killers) should be assessed as this is a common observation in IIH patients (17,23). While there is no clinical study proving that medication overuse aggravates IIH-related headache, given the current mechanistic understanding of medication overuse in the context of migraine, and the fact that IIH-related headache commonly presents with a migrainous phenotype or comorbid migraine, it is likely that medication overuse may play a role in the chronification of IIH-related headache.

Surgical treatments

Surgical management of IIH is required when there is a rapid or progressive decline in visual function (26,27). Surgical treatments include CSF shunting (including ventriculoperitoneal (VP), lumboperitoneal (LP), ventriculojugular and vetriculoatrial shunts), venous sinus stenting and optic nerve sheath fenestration (ONSF). The treatment offered is often dependent on the local expertise available. These should occur in conjunction with weight loss.

CSF diversion surgery

The scientific literature around CSF diversion neurosurgery remains limited by the fact that it is mainly observational and often based on case series. A randomised-controlled trial comparing medical management alone to medical management plus either CSF diversion surgery or ONSF was abandoned due to low enrolment, with only 16 of a planned 180 patients recruited (30). One large meta-analysis looking at surgical management of refractory IIH included 435 patients who had undergone CSF shunting (31). This report did not consider the type of shunt that was placed but reported that papilledema improved in 70% of patients (107/153), with an improvement in visual acuity in 54% of patients post-shunt (104/193). The paucity of visual data for over half the cases included in this meta-analysis is surprising, given the primary aim of this intervention should be to protect vision. Similarly, there was improvement in headache in 80% of patients, although there was only information on headache available for 287 of the total 435 cases. However, 43% of patients required more than one surgical procedure, with 428 additional surgical procedures performed in 154 of the total 435 patients. The commonest reason for a repeat procedure was shunt obstruction (41%). They report that the rate of major complications (shunt infection, tonsillar herniation, subdural hematoma, and CSF fistula) was 7.6% (33/435), while minor complications (including abdominal pain, shunt disconnection, valve dysfunction and low-pressure headache) occurred in 32.9% (143/435).

More recent advances in shunt technology may contribute to lower shunt failure and complication rates in the future. However, it is important to stress that CSF diversion surgery is not generally a recommended treatment for headaches in the absence of visual involvement (26,27). A relatively large single centre retrospective study of 50 patients undergoing CSF diversion surgery for IIH describes that headache remained a problem for 56% of these patients, with 42% requiring medication for headache management post-shunt (32).

Optic nerve sheath fenestration (ONSF)

As with CSF diversion surgery, the literature around ONSF in IIH is largely retrospective, with small sample sizes. Different mechanisms through ONSF exerts its action have been proposed, including dural fistula formation (33) or scarring of the subarachnoid space acting as a barrier to the transmission of intracranial pressure to the optic nerve (34). It is often considered in patients with asymmetrical papilledema and traditionally has been used in patients where headache is a less prominent feature. It is considered to be associated with fewer serious complications than CSF diversion surgery, particularly life-threatening problems. In their meta-analysis, Satti et al. included 1153 eyes of 712 patients with IIH who had undergone ONSF, with a mean follow-up of 21 months (compared with 41 months in the CSF diversion group) (31). They found that visual acuity improved in 59% (152/257 eyes), a similar figure to that reported for the CSF diversion group, with 95% of eyes either showing either an improvement or stable visual function post-ONSF. Papilledema improved in 80% (76/95 patients). While some improvement in headache can be seen in patients following ONSF, the proportion of patients who experience this is less than with CSF diversion surgery at 44%. The rate of major complications following ONSF was found to be 1.5% based on this group and included retinal artery occlusions, retrobulbar haemorrhage, traumatic optic neuropathy, and manifest strabismus. The rate of minor complications was 16.4%, and included transient double vision, anisocoria, conjunctival problems/cysts and optic nerve haemorrhages. A failure rate of 14.9% was found. It has been observed that unilateral ONSF can cause an improvement in the grade of papilledema seen in both eyes, although the mechanism for this is not certain (35). Patients with ONSF require ongoing monitoring as visual worsening is possible even after a period of visual stability, at a rate that could be as high as 45% at 3 years (27).

Venous sinus stenting

Many patients with IIH can be observed to have stenoses in the cerebral venous sinuses. While these may be constitutional, the possibility of raised intracranial pressure causing sinus compression remains and venous sinus stenting has been used in this scenario with some success. Satti et al. included 136 patients in their meta-analysis who had undergone venous sinus stenting for refractory IIH, with a mean follow up time of 22.9 months (31). This intervention appears to be very effective, with improvement in papilledema seen in 97% of patients (104/108 patients). Headache improvement was reported in 83% (101/121 patients). Complications were seen in 7.4% (10/136 cases), the most serious being related to vessel perforation and acute subdural haemorrhage, stent migration and thrombosis. It was found that 14/136 patients needed a repeat procedure (10.3%), with eight patients requiring a stent to be placed at or near the original treatment site for a restenosis. However, as Mollan et al. state in the IIH Consensus guidelines, long-term data concerning safety and efficacy is lacking (27). They also comment that there is a lack of data regarding the role of venous sinus stenting in patients with rapidly worsening vision. The advice is that this treatment should be reserved for carefully selected patients, with clear evidence of an elevated pressure gradient across a stenosis.

Clinical implications

- The main features of IIH are chronic headache and progressive visual disturbances, although other features including cognitive deficits, cranial nerve palsies, olfactory disturbances and tinnitus are not uncommon.
- Visual disturbances include visual loss, transient visual obscurations resulting from CSF pressure-induced papilledema and diplopia due to cranial nerve palsies.
- The pathophysiological basis has not been fully elucidated. Therefore, treatment strategies focus on the reduction of body weight and inhibition of CSF production with carbonic anhydrase inhibitors.
- IIH-related headache often has a migrainous phenotype and may persist even if CSF pressure has been normalised. In this context, acetazolamide is the only substance that has been investigated in a randomised-controlled trial. If headache has a migrainous phenotype, migraine preventives may be used.

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ORCID iD

Jan Hoffmann https://orcid.org/0000-0002-2103-9081

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