Idiopathic intracranial hypertension

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Idiopathic intracranial hypertension is common in obese women and can lead to significant visual impairment. First described more than 100 years ago, the cause of the disorder remains unknown. Despite a multitude of proposed links, the aetiology has never been established. Impairment of cerebrospinal-fluid reabsorption is the most likely underlying pathophysiological cause of the raised pressure, but this notion has yet to be proven. Cerebral venous sinus abnormalities associated with the disorder need further exploration. Although the major symptoms of headache and visual disturbance are well documented, most data for disease outcome have been from small retrospective case series. No randomised controlled trials of treatment have been done and the management is controversial. The importance of weight loss needs clarification, the role of diuretics is uncertain, and which surgical intervention is the most effective and safe is unknown. Prospective trials to examine these issues are urgently needed.

Introduction

Idiopathic intracranial hypertension occurs most commonly in overweight women and is encountered by most neurology and ophthalmology departments on a regular basis. Far from being benign, the condition causes blindness in 10% of cases; indeed, most affected patients show a degree of visual loss. The association with obesity suggests that the incidence will increase as the population in the developed world gains weight. Despite this theory, very little is known about the disorder. The cause is undetermined and treatments lack any convincing evidence base. This review will summarise current knowledge of the cause, differential diagnosis, and pathophysiology of idiopathic intracranial hypertension, drawing attention to dilemmas in treatment and the need for well-designed prospective trials.

Definitions

Idiopathic intracranial hypertension is the clinical syndrome of raised intracranial pressure, in the absence of space-occupying lesions or vascular lesions, without enlargement of the cerebral ventricles, for which no causative factor can be identified. Serous meningitis and pseudotumour cerebri were the terms previously used for idiopathic intracranial hypertension. In 1937, diagnostic criteria for intracranial pressure without brain tumour were first published by Dandy, and in 1955 Foley coined the term benign intracranial hypertension. More recently, the term benign has been dropped after reports of severe visual loss.

With the advent of sophisticated neuroimaging techniques, intracranial lesions and vascular pathologies have been identified in patients who might previously have been labelled as having idiopathic intracranial hypertension. Diagnostic criteria have undergone several modifications as a result. A strict definition now exists to ensure that idiopathic intracranial hypertension is only used when other causes of intracranial hypertension have been excluded (panel 1). Further confusion arises through the alleged association with various medical conditions and treatments, most of which have only been described in case reports or small series. When intracranial hypertension can be assumed to be due to an underlying disease or a historically associated drug, it is incorrect to apply the term idiopathic. Where there is convincing evidence that the drug or disease is causally or temporally related, the term secondary intracranial hypertension is more appropriate.

Epidemiology

In the USA, Durcan reported the annual incidence of idiopathic intracranial hypertension in the general population of Iowa to be 0·9 per 100 000 and in Louisiana to be 1·07 per 100 000. A prospective longitudinal study in Benghazi, Libya, reported an annual incidence of 2·2 per 100 000. In Hokkaido, Japan, only two cases were identified from the study in 1993, of a population of around 5·8 million, giving an incidence for that year of 0·03 per 100 000. The incidence of idiopathic intracranial hypertension is similar to that of Guillain Barré syndrome, pituitary tumours, and cluster headache. Confining studies to women aged 20–44 years, who are 20% or more above their ideal bodyweight, yields an incidence of 15–19 cases per 100 000 in the USA, approaching that of more common diseases such as motor neuron disease and multiple sclerosis.

Idiopathic intracranial hypertension is most common in women and obese individuals. Reported female to male ratios range from 4:1 to 15:1. Published frequencies of obesity in patients with the disorder are 71%, 88%, 91%, and 94%. In a North American prospective study of 50 patients by Wall and George, 47 were obese and there was an average weight gain of 7·7 kg in the 12 months preceding the onset of symptoms. The mean age at onset of the disorder has been reported

Panel 1: Diagnostic criteria for idiopathic intracranial hypertension

If symptoms or signs are present, they may only indicate those of generalised intracranial hypertension or papilloedema

Intracranial pressure, as measured in the lateral decubitus position, is raised

The composition of the cerebrospinal fluid is normal

There is no evidence of hydrocephalus, mass, structural, or vascular lesion

No other cause of intracranial hypertension has been identified
Idiopathic intracranial hypertension does occur in childhood, although no large epidemiological studies in this age-group have been undertaken. The disorder is rare in prepubertal children and has different characteristics to the adult form, including no apparent predilection for obese girls. In older teenage children, however, the rates of obesity seem to mirror those of the adult population with the disorder. Ethnic background has not been shown to affect the incidence, although few studies have addressed this question or involved sufficient numbers of patients. Familial cases are encountered, but there is little evidence to lend support to a genetic predisposition.

Associated conditions
Apart from female sex and obesity there are no proven associations in idiopathic intracranial hypertension. Whether the raised intracranial pressure is a result of, or occurs by chance in association with, the factor in question is important to establish.

Vitamin A deficiency
The fat-soluble vitamin retinol (vitamin A) affects the structure of the arachnoid villi. Studies of retinol-deficient animals have shown increased cerebrospinal fluid (CSF) pressure, although the exact relation between the structural changes and the mechanism of raised pressure is uncertain. Cases have been reported of intracranial hypertension occurring in infants deficient in vitamin A, which resolved with replacement therapy. Excessive dietary intake of vitamin A has also been associated with the disorder. Raised serum retinol concentrations have been reported in some human cases of idiopathic intracranial hypertension, although not to the levels seen in true hypervitaminosis A. This finding has caused speculation that a non-specific alteration in retinol metabolism contributes to idiopathic intracranial hypertension in predisposed individuals. More recently, significantly heightened concentrations of retinol have been reported in the CSF but not serum of patients with idiopathic intracranial hypertension compared with controls.

Medication
Case reports have implicated several drugs in intracranial hypertension (panel 2). There have been reports of idiopathic intracranial hypertension in a few patients taking tetracycline antibiotics, particularly minocycline and doxycycline, but prospective case-controlled studies have not firmly established a causative relation. Similarly, oral retinoids used in dermatological conditions have earned a reputation for causing pseudotumour cerebri despite a lack of convincing evidence. Abrupt withdrawal of corticosteroids has been reported to cause a syndrome indistinguishable from idiopathic intracranial hypertension, with clinical improvement after reintroduction of the drug. However, idiopathic intracranial hypertension, defined according to strict diagnostic criteria, has not been conclusively associated with corticosteroids in large controlled studies. The oral contraceptive pill has been historically connected with the disorder, but more recent research has shown no significant increase in the proportion of women taking the pill in those with idiopathic intracranial hypertension compared with the general population. Early studies may have included undiagnosed cases of cerebral venous sinus thrombosis among women taking hormonal medication; the high prevalence of oral contraceptive users in idiopathic intracranial hypertension cohorts in the past could reflect the higher risk of cerebral venous sinus thrombosis in women taking oral contraceptives.

Diseases
Systemic arterial hypertension has been reported as occurring in 14–32% of patients, and in one study blood pressure was significantly higher in people with idiopathic intracranial hypertension than in controls. Whether this finding is a true disease association or simply reflects the high incidence of raised blood pressure in a population with increased bodyweight is unclear. Polycystic ovary syndrome seems to occur with increased frequency in idiopathic intracranial hypertension. Other comorbid conditions reported include diabetes mellitus, thyroid disease, hypoparathyroidism, stroke, chronic migraine, iron deficiency anaemia, ulcerative colitis, systemic lupus erythematosus, sickle-cell disease, cystinosis, and renal
transplant, although undiagnosed cerebral venous sinus thrombosis or simply chance co-occurrence cannot be excluded in most cases.

**Pregnancy**

Pregnancy has been traditionally regarded as carrying an increased risk of idiopathic intracranial hypertension. In recent years, this association has been subjected to more rigorous investigation and has not been shown to be significant. As with oral contraceptive use, a high number of pregnancies are to be expected in a condition that favours women of reproductive age.

**Menstrual dysfunction**

A history of menstrual irregularities is more common in idiopathic intracranial hypertension than in unaffected women. In one questionnaire study of 40 patients with idiopathic intracranial hypertension, a change in menstrual pattern was more frequently reported just before diagnosis than in the reference period in 39 controls. Onset of menses at or before age 13 years was also significantly more likely in women with the disorder.

Menarche, oligomenorrhea, or amenorrhea have preceded the onset of idiopathic intracranial hypertension in case reports, and larger studies have listed menstrual dysfunction among reported symptoms. However, there has been no published evidence of specific hormone dysfunction to explain this finding, and obesity itself is known to be associated with menstrual irregularities.

**Pathogenesis**

**Mechanisms of raised intracranial pressure**

The pathophysiology underlying the raised intracranial pressure is unclear. Hypotheses have developed around the main determinants of CSF pressure.

**Excess CSF production**

CSF is produced in human beings at a rate of about 500 mL in 24 h, the whole CSF volume being renewed every 6–8 h. The early suggestion that idiopathic intracranial hypertension resulted from CSF hypersecretion has been refuted by studies showing no significant differences in the production rate between patients and controls or rates even lower than previously quoted normal values. The only condition in which excessive secretion of CSF is known to occur (choroid plexus papilloma of childhood) does not give rise to a similar clinical picture to idiopathic intracranial hypertension.

**Brain water content**

Evidence of diffuse brain oedema in idiopathic intracranial hypertension is slight. Early histological studies suggestive of intracerebral and interstitial oedema have been re-examined and repeated without confirmation of the initial claims. Some MRI studies, which used high-field strength and diffusion-weighted imaging, have suggested increased brain water in idiopathic intracranial hypertension, whereas other investigators have found no differences between patients and controls. Patient numbers in these studies have been too small for robust conclusions to be drawn.

**Reduced CSF absorption**

A more popular hypothesis is that idiopathic intracranial hypertension is a syndrome of reduced CSF absorption. An early study used the intrathecal saline infusion test to show resistance to drainage of CSF in four of five patients with pseudotumour cerebri. Johnston and Paterson published similar work using isotope cisternography showing a substantial delay in CSF circulation, with hold up of technetium isotope in the subarachnoid space. Altered patterns of CSF clearance have been reported with this and other techniques, such that resistance to CSF outflow seems to be implicated in the pathogenesis. However, little is known about the exact site or mechanism, such as how CSF egress is regulated at the level of the arachnoid villi.

**Increased cerebral venous pressure**

Whether the raised CSF pressure is the primary problem or the consequence of high pressure in the cerebral venous sinuses, due either to anatomical narrowing or generalised venous hypertension, is widely debated. In patients with proven venous sinus thrombosis, the high intracranial pressure is relieved by drainage or diversion of CSF, although it is uncertain how this happens. Venography shows narrowing of the cerebral venous sinuses leading to functional outflow obstruction in many cases of idiopathic intracranial hypertension in the absence of occlusion by thrombus. In one prospective blinded study that used magnetic resonance venography, substantial bilateral sinovenous stenoses were seen in 27 of 29 cases leading the authors to conclude that dural venous sinuses are anatomically different in idiopathic intracranial hypertension. Raised pressure in the absence of thrombosis within superior sagittal and transverse sinuses has also been shown. Whether such findings result from or cause the high CSF pressure is not known, but there is evidence to lend support to both explanations.

The notion of apparent venous stenosis as a secondary effect requires that the vessel walls are externally compressed or the lumen is partly obstructed by enlarged arachnoid granulations. Both types of filling defect have been reported in idiopathic intracranial hypertension. There have been reports of stenotic lesions in the lateral sinuses resolving after CSF diversion procedures, and of transverse sinus pressures dropping when CSF is directly removed by cervical puncture. However, a more recent study has shown persistence of transverse sinus stenoses in idiopathic intracranial hypertension after normalisation of CSF pressure. The findings of other studies lend support to the opposite claim that venous stenoses are the primary lesion in idiopathic intracranial hypertension. Stenoses are the primary lesion in idiopathic intracranial hypertension.
Review

hypertension. Dilatation and stenting of the lateral venous sinuses has produced clinical improvement in some patients with idiopathic intracranial hypertension who have high venous sinus pressures at manometry.57 Karahalios and colleagues58 reported high pressure with normal anatomy in the venous sinuses of five patients with idiopathic intracranial hypertension, and showed that the right atrial pressures were increased, leading them to suggest a universal mechanism of raised venous pressure. Others have focused attention on a venous hypertension model in which raised intracranial pressure is a direct result of intra-abdominal pressure, via elevation of the diaphragm, raised pleural pressure, impeded cerebral venous return, and consequently sustained elevation of the venous pressure.59 This theory requires the presence of central obesity to increase the intra-abdominal pressure and does not account for the preponderance of women with the disorder, since women are less likely than men to have a central androgenic distribution of body fat. Idiopathic intracranial hypertension would also be expected to occur more commonly in pregnancy, which it does not. Furthermore, clinical evidence of right-heart failure is usually lacking in patients with the disorder. Nevertheless, the possibility of some shared pathophysiological mechanism in idiopathic intracranial hypertension and venous sinus disease remains enticing. Unrecognised non-occlusive thrombus lining dural vessels has been suggested as a possible mechanism for impairment of CSF absorption,60 which could fundamentally change the current definition of idiopathic intracranial hypertension. Thrombophilic haematological abnormalities have been reported in the disorder,56,58–62 and more recently in consecutive case studies of 65 female63 and ten male patients.64 However, a prospective study did not find epidemiological evidence to link idiopathic intracranial hypertension with a prothrombotic state,65 and there is little evidence of a specific abnormality.

Endocrine dysfunction
Studies of endocrine function in idiopathic intracranial hypertension have involved only small numbers of patients. Conventional tests of pituitary function and peripheral target glands in one study of 15 patients were normal, apart from a slight reduction in growth hormone response to hypoglycaemia.66 Subnormal growth hormone response was also reported by Reid and colleagues67 in three of five patients with chronic idiopathic intracranial hypertension, but was also seen in one of the obese control patients, as can occur.68 Raised concentrations of vasopressin in CSF have been noted in patients with idiopathic intracranial hypertension compared with controls, despite similar plasma concentrations and osmolality;69,70 but the importance of this finding to the pathogenic processes in the disorder is not clear.

The predilection of the disorder for obese women of childbearing age suggests that sex hormones might be involved, but this notion is unproven. Adipose tissue has been identified as an actively secreting endocrine organ, and attention has been given to one of its products, leptin, as having a possible role in the disorder.69 The metabolism of sex steroids is affected by adipose tissue and the metabolic effects of obesity vary according to sex and menopausal status. More work is needed in this area.

Clinical features
Patients with idiopathic intracranial hypertension can be asymptomatic, but more commonly they complain of the symptoms listed in panel 3. Headache is consistently the most common symptom, occurring in 68–98% of patients3,8,13 and featuring as the presenting complaint in many.3 Headache seems to be less common in children than in adults with the disorder.20 Headache can fluctuate and be progressive or permanent. The pain may have similarities to migraine and tension-type headache71 or show features of raised intracranial pressure—ie, exacerbation by coughing, straining, or the Valsalva manoeuvre.1

Disturbance of vision is the second most prevalent symptom. Transient obscurations, variously described as shadows, dark patches, or black spots affecting one or both eyes and resolving after a few seconds or minutes, are reported in 57–72% of patients.1,3,13 Other less common visual abnormalities are diplopia and sparkles (photopsia) or the sensation of flashes of light. Even less frequently, patients present with visual loss and central vision is usually spared until late in the course of the illness.3

Intracranial noises are frequent, usually described as “whooshing” or “roaring” in the ear. The sounds are often pulsatile. In prospective analyses they affect as many as 60% of patients.1 Rare symptoms in adults include neck, back, shoulder, or radicular pain. Signs of meningeal irritation sometimes occur, including nausea, vomiting, and photophobia.9

Panel 3: Symptoms of idiopathic intracranial hypertension

| Headache |
| Visual disturbance |
| Visual loss and blurring of vision |
| Transient obscurations |
| Diplopia |
| Photopsia |
| Tinnitus |
| Pain |
| Neck |
| Shoulder |
| Radicular pain |
| Nausea with or without vomiting |
| Photophobia |
Papilloedema is almost universal in idiopathic intracranial hypertension and its absence should cause the diagnosis to be questioned. There have been a few reports of the disorder without papilloedema in patients with headache and increased CSF opening pressure.\(^7\)\(^-\)\(^7\)\(^4\)

The symptom may occasionally be unilateral, as in four of 57 patients in one series\(^1\) (figure 1). Slight oedema of the disc may be difficult to detect using the direct ophthalmoscope, so slit-lamp examination is needed. In equivocal cases, fluorescein angiography might be needed to show the leakage of dye from retinal vessels that is typical of papilloedema. Congenital anomalous variations in the architecture of the optic nerve head (eg, drusen) can mimic papilloedema.\(^4\)

Testing of visual fields by confrontation in patients with idiopathic intracranial hypertension elicits few

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**Figure 1:** Asymmetric papilloedema in a 42-year-old woman with headaches and tinnitus and transient visual obscurations on the left side only. CSF opening pressure was 30 cm water. A: Left fundus shows swelling of the optic disc. B: Right fundus shows no papilloedema.

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**Figure 2:** Visual field (Goldmann perimetry) from the right eye of a 54-year-old woman with idiopathic intracranial hypertension showing marked constriction. Snellen visual acuity was 6/9 and there was pallor of the optic disc at fundoscopy.
abnormalities. Formal perimetry is mandatory as it reveals visual-field defects and enlargement of the blind spot in nearly all patients. In Wall’s study, at the initial visit Goldmann perimetry was abnormal in 87% of eyes and Humphrey automated perimetry in 92%. Abnormalities of other visual function tests occurred far less frequently. Peripheral rim constriction most often occurs with nasal steps and arcuate inferior field loss (figures 2 and 3). The grade of visual field defect has not been shown to correlate with the severity of papilloedema.

Reductions in visual acuity, although less likely than field defects, can also occur. Contrast sensitivity and assessment of colour vision reveal more abnormalities than standard Snellen acuity. It is a requirement of the modern diagnostic criteria for idiopathic intracranial hypertension that neurological examination is normal except for the presence of signs of generalised intracranial hypertension and papilloedema. Unilateral or bilateral sixth cranial nerve palsies and afferent pupillary defects are sometimes seen.

Investigations

Idiopathic intracranial hypertension is a diagnosis of exclusion. Although there are no pathognomonic radiological signs in the disorder, empty sella turcica and variable diminution in ventricular size have been reported, although the latter is debated. CT became the investigation of choice until it was superseded by MRI. Posterior scleral flattening (relative to the normal curvature of the globe), prelaminar enhancement of optic nerves, vertical tortuosity of the orbital optic nerves, and distension of the perioptic subarachnoid space, as well as empty sella, have been shown, in a retrospective MRI series, to be sensitive and specific markers of raised intracranial pressure in patients with idiopathic intracranial hypertension.

To comply with recent diagnostic criteria, cerebral venous sinus thrombosis must be excluded. Thrombosis of the cerebral venous circulation can present with an identical clinical picture to idiopathic intracranial hypertension and may even fulfil the old modified Dandy criteria for idiopathic intracranial hypertension.

It is possible to overlook sinus thrombosis on CT or MRI unless formal study of the cerebral vasculature is undertaken by computed tomographic venography or magnetic resonance venography.

Examination of CSF pressure in patients with suspected idiopathic intracranial hypertension is mandatory and lumbar puncture is a safe procedure in the fully conscious patient with no focal neurological deficit and normal brain imaging, even in the presence of papilloedema. The diagnostic criteria require CSF constituents to be normal. Pressure is measured in the lateral decubitus position with a value of over 20 cm of water deemed abnormal. Historically, this threshold has been increased to 25 cm in obese patients, but the theory that obesity is a cause of mildly raised CSF pressure is now contested. An opening pressure of 25 cm has been shown to be abnormal irrespective of weight.

Continuous recording of CSF pressure shows that intracranial pressure changes over time and may occasionally drop to normal values in idiopathic intracranial hypertension. Overnight monitoring of patients shows that baseline pressure is usually increased and that plateau pressure (sustained above 50 mm Hg for 5–20 min) and B waves (rhythmic, 5–50 mm Hg, 0.5–2 cycles per min) are often present. Hence, in theory, a single lumbar puncture might lead to normal pressure recordings in idiopathic intracranial hypertension.
Management
There have been no sufficiently large studies of any treatment modality in idiopathic intracranial hypertension so the evidence base for management decisions is poor. A Cochrane systematic review in 2005 did not find any randomised controlled trials of treatments for the disorder.\textsuperscript{46} The aim of treatment is to halt or prevent visual loss. Current practice includes a combination of medical therapy, advice for weight reduction where appropriate, and surgical intervention for severe cases. Repeated lumbar punctures to directly reduce the intracranial pressure formed the mainstay of treatment for many years. In one study, removal of 15–25 mL of CSF reduced the pressure to below 10 cm of water, but it took an average of only 82 min for the pressure to return to predrainage levels.\textsuperscript{39} Although repeated lumbar puncture can result in holes in the dura, maintaining lowered pressure via small amounts of chronic CSF leakage, the risks of low-pressure headache, and the unpopularity of the procedure with patients led to regular lumbar punctures being phased out. However, the procedure could still have a role in the immediate treatment of acute visual loss in idiopathic intracranial hypertension while further treatment is organised, and also in the management of pregnant patients.\textsuperscript{79}

Medical management
The carbonic anhydrase inhibitor acetazolamide is the drug most commonly used to treat idiopathic intracranial hypertension. It reduces aqueous humour production in the eye and intraocular pressure and produces a weak diuresis when absorbed systemically. The drug may be associated with blood disorders, rashes, and electrolyte disturbances. Teratogenic effects have been reported in animals, but there are no adequate human studies of its use in pregnancy and a review did not find any reliable reports of adverse pregnancy outcomes in women treated with the drug.\textsuperscript{86} Patients taking acetazolamide commonly complain of nausea, fatigue, altered taste, and sensations of pins and needles.\textsuperscript{78} Other diuretics, such as furosemide, are sometimes used if acetazolamide is poorly tolerated.

Corticosteroids have been associated with improvement in the symptoms and signs of raised intracranial pressure, but only in small case reports.\textsuperscript{86–89} The study by Wall and George\textsuperscript{3} of 50 patients included nine treated with corticosteroids; five improved and four worsened at follow up, but numbers were too small to be conclusive. In another small prospective study, steroids had no effect in three patients who did not respond to a combination of acetazolamide and diuretics.\textsuperscript{89} Concerns over side-effects, especially weight gain, greatly limit their use. Additional medication may be needed to treat the headache in idiopathic intracranial hypertension, such as analgesics or even antimigraine drugs.

Surgical management
Some patients require surgical intervention to control symptoms or prevent visual deterioration. In the past, subtemporal decompression was undertaken, but this procedure has been replaced with less hazardous interventions. The two main operations in idiopathic intracranial hypertension are CSF diversion procedures and fenestration of the optic-nerve sheath. No prospective trial has compared the two and the choice is often based on the local availability of expertise. Surgical treatment needs to be considered from the outset for patients presenting with visual loss and at any time when there is deterioration in visual function.\textsuperscript{95} Some authors argue that the decision to operate rests entirely on the quantitative visual-field status.\textsuperscript{96} Surgery might also be appropriate where close monitoring and effective medical treatment of the patient is not feasible or if there is intractable headache.

CSF diversion procedures
The insertion of ventriculoperitoneal or lumboperitoneal shunts to treat idiopathic intracranial hypertension has become standard clinical practice. However, shunts have a significant failure rate requiring revisional surgery and a high frequency of complications (ie, low pressure headaches, infections, obstruction, general operative complications).\textsuperscript{97 Less commonly, subdural haemorrhage, radiculopathy, tonsillar herniation, and syringomyelia have been reported.\textsuperscript{98} Nevertheless, undertaken by an experienced neurosurgeon, such operations can be sight-saving and effective for all manifestations of idiopathic intracranial hypertension.\textsuperscript{99}

Optic-nerve-sheath fenestration
The optic nerve is decompressed by making a fenestration or slit in the dural sheath, which follows the nerve along its course from skull to orbit. The exact mechanism of action is uncertain and could be due to egress of CSF forming a chronic fistula or scarring of the meninges preventing transmission of the high CSF pressure to the optic-nerve head.\textsuperscript{100} For an experienced ophthalmic surgeon, it is a minor procedure and, unlike shunts, involves no foreign body.\textsuperscript{101} Complications are usually transient and benign, but severe ocular problems can occur and the procedure is not always successful.\textsuperscript{102} Visual function may deteriorate many years later and, even after initially successful operations, it can be necessary to proceed to CSF diversion to treat intractable headaches or halt progressive visual loss.\textsuperscript{102–104}

Weight reduction
It is widely believed that weight reduction improves the course of idiopathic intracranial hypertension in overweight individuals. Resolution of papilloedema alongside weight loss through dietary modification has been reported by several authors.\textsuperscript{90–92} Surgically induced weight loss has also led to improvements in idiopathic intracranial hypertension.\textsuperscript{93–95} Evidence is limited and further research is needed to quantify the optimum amount, timing, and methods of weight loss in management of idiopathic intracranial hypertension.
Monitoring

There is no consensus about how to monitor patients with idiopathic intracranial hypertension or which clinical assessments most reliably identify those most at risk of visual loss. Regular measurement of CSF opening pressure is rarely used because the procedure is unpleasant and CSF pressure can vary widely. Current practice should now include assessment of symptoms, examination of optic discs, measurement of visual acuity, and formal documentation of visual fields. Formal perimetry seems the most precise method of identifying visual loss and has been shown to exhibit statistically greater sensitivity by comparison with acuity and contrast sensitivity. False positives do occur and visual fields can show substantial variation, so it is essential to verify abnormalities by auditing technique and repeating tests.

Outcome

For most patients, idiopathic intracranial hypertension lives up to its old name as a benign condition, without impairment of vision in the long term. However, a small but significant number of patients with idiopathic intracranial hypertension follow a more aggressive course. Blindness has been shown to occur in at least one eye in 8–10% of patients and some sustained loss of vision has been reported in almost half of patients in some studies (table). No consistent correlations have been found between visual outcome and age at diagnosis, sex, bodyweight, oral contraceptive use, steroid treatment, CSF opening pressure, duration or type of symptoms, and chronicity of papilloedema. A significant association was found between visual loss and the occurrence of systemic hypertension in one study.

Idiopathic intracranial hypertension can recur months to years after the original attack has resolved. No predictors of recurrence have been identified. Loss of sight might occur gradually, in a stepwise fashion, rapidly, or slowly over months or years. Sometimes visual loss is most severe at presentation. For many patients, symptoms are so minimal that the visual loss goes undetected until it is severe. This finding draws attention to the need for watchful monitoring of patients with idiopathic intracranial hypertension, many of whom may be asymptomatic until visual disaster occurs.

Future developments

Many questions remain unanswered about idiopathic intracranial hypertension. Its association with female sex and obesity is striking and its co-occurrence with symptoms of hormonal dysfunction intriguing. Although only a few overweight people develop idiopathic intracranial hypertension, the well-recognised increasing trend towards obesity in today’s society suggests that the incidence is likely to increase. Despite the large numbers of people affected, especially obese women of childbearing age, effective management lacks an evidence base. Further research should be directed towards improving our understanding of the disease process since the identification of underlying pathology could identify opportunities for therapeutic advances. There is an even more urgent need for well-designed randomised controlled trials in idiopathic intracranial hypertension to determine the most effective way both to monitor and to treat this sight-threatening condition.

Contributors

AKB and CEC co-wrote the review. AKB provided the figures.

Conflicts of interest

We have no conflicts of interest.

References


Search strategy and selection criteria

References for this review were identified by searches of MEDLINE and PubMed from 1955 to February 2006, with the terms “idiopathic intracranial hypertension”, “benign intracranial hypertension”, “pseudomotor cerebri”, and “otic hydrocephalus”. Additionally, we identified early references by hand searches of journals. Only papers published in English were reviewed. The final reference list was generated on the basis of originality and relevance to the topics covered in the review.

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<thead>
<tr>
<th>Study type</th>
<th>Number of patients</th>
<th>Number with visual loss</th>
<th>Comment</th>
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<tr>
<td>Boddie et al, 1974</td>
<td>Retrospective 34</td>
<td>4 (12%)</td>
<td>Three patients had visual loss at presentation</td>
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<tr>
<td>Corbett et al, 1982</td>
<td>Retrospective 57</td>
<td>14 (25%)</td>
<td>24 eyes in 14 patients were blind or profoundly visually impaired</td>
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<td>Orcutt et al, 1984</td>
<td>Retrospective 68</td>
<td>21 (31%)</td>
<td>Five eyes in four patients had severe visual loss</td>
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<td>Sorensen et al, 1988</td>
<td>Prospective 24</td>
<td>1 (4%)</td>
<td>Chronic papilloedema in 42%</td>
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<tr>
<td>Wall et al, 1999</td>
<td>Prospective 50</td>
<td>5 (10%)</td>
<td>Two patients were blind in both eyes, three were blind in one eye</td>
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<td>Radhakrishnan et al, 1993</td>
<td>Prospective 81</td>
<td>16 (20%)</td>
<td>Moderate to severe loss; mild loss in a further 23%</td>
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<td>Rowe et al, 1998</td>
<td>Prospective 35</td>
<td>6 (17%)</td>
<td>Three patients had visual loss and optic atrophy at presentation</td>
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Table: Published reports of visual loss as an outcome of idiopathic intracranial hypertension