Idiopathic intracran

Figure 1 Papilloedema from patient with idiopathic intracranial hypertension.

INTRODUCTION

We have recently completed a review for the Cochrane Library on interventions for idiopathic intracranial hypertension (IIH) but could not find any relevant randomised controlled trials (Lueck & McIlwaine 2002). There is therefore inadequate evidence on which to base the management of this condition, and yet patients still need to be treated. Many different treatments have been suggested over the years, and various clinicians have their own preferred method of management. What we are presenting here is really 'how we do it', with the reasons why we do what we do (where we can provide them!).

BACKGROUND

Idiopathic intracranial hypertension has had a number of different names in the century since it was first described by Quincke under the name of 'meningitis serosa' (Quincke 1897). The most commonly used synonyms are 'pseudotumour cerebri' (Nonne 1904) and 'benign intracranial hypertension' (Foley 1955). However, there are problems with both these names because the condition is not always benign (some patients go blind), and 'pseudotumor' is an archaic word dating from pre-imaging days and which is potentially frightening to patients. The label 'idiopathic' was therefore suggested in 1969 (Bucheit *et al.* 1969). Having said that, there is still considerable confusion in the literature because the clinical picture of 'IIH' has in some cases been associated with a number of potential underlying factors (e.g. sagittal sinus thrombosis and vitamin A toxicity), in which case the term 'idiopathic' obviously becomes inappropriate. However, IIH seems to be the generally preferred name at the moment (Miller 1998).

IIH is defined as raised intracranial pressure without ventricular enlargement or intracranial mass on imaging, with normal cerebrospinal fluid constituents (Miller 1998). It often presents with headaches, and is usually associated with bilateral papilloedema (Fig. 1).

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The overall incidence is 1–3/100 000 population/year (Durcan *et al.* 1988; Radhakrishnan *et al.* 1993a,b; Radhakrishnan *et al.* 1994). It is somewhere between four and 10 times as common in women as in men, particularly women between the ages of 15 and 44 when it is strongly associated with obesity (the incidence rises to 21/100 000 population/year in this group). Having said that, it is well described in children, where the gender difference is much less obvious and obesity is a considerably less-strongly-associated factor (see Digre & Corbett 2001).

In spite of the fact that several different symptoms and signs can be associated with IIH (Table 1), the only major complication is visual loss. Early series reported that some 10% of affected eyes could end up blind (Corbett *et al.* 1982). More recent studies have shown some degree of visual loss in up to 85% of cases if carefully sought (Rowe & Sarkies 1998). Therefore, on the grounds that intervention *should* prevent blindness, IIH is an important condition to diagnose and treat. Many different treatments have been suggested over the years (Table 2), partly because so many different types of specialist can be involved depending on which symptoms and signs are dominant in

the early stages. Each of these various specialists has their preferred method of treatment, for example a lumboperitoneal shunt is generally more expedient for neurosurgeons, while optic nerve sheath fenestration is more convenient for ophthalmologists. The problem is that many of the treatments have potential complications, many are expensive, and none of the available treatments has been subjected to a randomised trial (Lueck & McIlwaine 2002). This is, as they say, an 'evidence free zone'.

The cause of IIH remains unknown. The most popular theory is that there is some form of obstruction to cerebrospinal fluid (CSF) outflow, either at the level of the arachnoid villi (Pacchionian granulations), or downstream in the dural venous sinuses or draining veins. Venous sinus thrombosis is well-described, and the clinical picture is often very similar to that of IIH. However, by definition 'IIH' implies that there is no obvious sinus thrombosis present. Alternative theories include an excess of CSF production, cerebral oedema, and increased cerebral blood volume. We do not propose to go into causation in any further detail here, other than to point out that IIH may well prove to be the final result of more than one mechanism (Digre & Corbett 2001).

Table 1 Symptoms and signs of idiopathic intracranial hypertension

	COMMON (> 10% CASES)	UNCOMMON/OCCASIONALLY REPORTED
Symptoms	headache (typically daily, retrobulbar, worsened with eye movement; may be throbbing with nausea, vomiting and photophobia) visual obscurations (few seconds, partial/complete, no correlation with extent of papilloedema) visual blurring visual loss double vision	tinnitus, hearing loss, vertigo photopsia, retrobulbar pain on eye movement gaze-evoked blindness paraesthesiae, spinal & radicular pain facial pain, facial numbness nausea & vomiting hemifacial spasm, torticollis altered taste & smell CSF rhinorrhoea cognitive disturbance, disturbance of consciousness, depression irritability/drowsiness, intermittent ataxia (in children)
Signs	papilloedema (may be unilateral/absent) visual field defect, particularly enlarged blind spot reduced visual acuity reduced colour vision afferent pupillary defect 6th nerve palsy obesity	3rd nerve palsy 4th nerve palsy skew deviation/strabismus (children)/nystagmus facial nerve palsy (unilateral >> bilateral) hearing loss hemiparesis optic atrophy subretinal haemorrhage choroidal neovascular membrane

Table 2 Treatments proposed for idiopathic intracranial hypertension**Weight loss**

- low calorie diet
- surgery for obesity

Diuretic therapy

- acetazolamide
- thiazide diuretics
- loop diuretics

Other pharmacological therapy

- corticosteroids
- glycerol
- analgesics

Repeated lumbar puncture**Surgical intervention**

- optic nerve sheath fenestration:
 - medial
 - lateral
- lumboperitoneal shunting
- other neurosurgical procedures:
 - subtemporal decompression
 - ventriculo-peritoneal shunt ($\pm$ stereotaxic placement)
 - cisterno-peritoneal shunt

Other suggested treatments

- low salt diet
- low tyramine diet
- hyperbaric oxygen
- vasopressin
- D1 agonists
- external negative abdominal pressure devices

In this article, we propose to discuss two main areas in detail. First, making the diagnosis of IIH and the difficulties associated with doing so, and, second, managing the patient once diagnosed.

THE DIAGNOSIS OF IDIOPATHIC INTRACRANIAL HYPERTENSION

Most readers will be familiar with the overweight young woman who presents with a few weeks' history of headaches and who has gross bilateral papilloedema. Brain imaging shows no significant abnormality, and a lumbar puncture (often performed with some difficulty as the patient is obese) reveals a grossly elevated CSF pressure but is normal in all other respects. On the whole, there is little doubt about the diagnosis in this situation, but a number of points need to be considered, particularly if the case is not so straightforward.

Brain imaging

Is it really normal? It is very easy to miss sinus

thrombosis on CT or MR scan unless formal venography is performed. Nowadays our neuroradiologists are generally happy to exclude significant sinus thrombosis on CT or MR angiography, so we now rarely proceed to formal catheter angiography in this condition. Nevertheless, *all* patients presenting with apparent IIH need some form of angiography to exclude an underlying sinus thrombosis.

CSF pressure

Measurement

As mentioned above, it is often difficult to obtain CSF in obese individuals, and this frequently results in agitation on the part of the patient. Likewise, the headaches and general misery of the condition add to the overall stress, which in turn has the effect of increasing CSF pressure. It is important that the CSF pressure is measured with the patient horizontal (yes, we have seen pressures recorded with the patient sitting up!), legs extended (the patient is asked to straighten their legs after the needle has been passed into

the CSF space), and with the patient as relaxed as possible. In some cases it is necessary to perform the lumbar puncture under X-ray screening to make it easier on the patient.

The normal range?

What is the normal range of CSF pressure, anyway? For the purposes of diagnosing IIH, most clinicians rely on the study by Corbett & Mehta (1983) who found that CSF pressure in normal patients was less than 200 mmH₂O, but if they were obese the normal pressure could be as high as 250 mmH₂O. Anything higher is therefore regarded as abnormal. Although the CSF pressure does vary with weight, the degree of correlation is not clear and so the upper limit of normal may not be precise. This needs to be borne in mind when making a diagnosis of IIH.

Most patients with IIH have CSF pressures well in excess of 300 mmH₂O, in which case there is little room for doubt. However, it is not unknown for obese women with migraine and optic nerve head drusen (see below) to have an opening pressure of, for example, 260 mmH₂O and be inappropriately labelled by the unwary as

having IIH. Headaches and obesity are common conditions and therefore it would not be unusual for them to be associated with optic nerve head drusen, or an anomalous optic disc.

Papilloedema

This is potentially one of the most difficult areas. If the disc looks like that in Fig. 1, there is seldom any diagnostic doubt. However, life isn't always that easy, and many patients have 'borderline' papilloedema, i.e. their discs look 'a bit swollen'. The differential diagnosis of swollen discs is listed in Table 3 (see also Fig. 2). Therefore, in conjunction with a borderline CSF pressure, there is considerable scope for diagnostic error.

How to sort this out? The first move is to get an opinion from an ophthalmologist or neuro-ophthalmologist. Four useful clues that an apparently swollen disc may not actually represent straightforward papilloedema are:

- the blind spot is not enlarged (on formal perimetry);
- the presence of spontaneous pulsation in the retinal veins, or venous pulsation on minimal digital pressure applied to the eye;

Table 3 Differential diagnosis of papilloedema

DIAGNOSIS	COMMENTS
Myelinated nerve fibres	not usually too difficult to distinguish from papilloedema
'Crowded' optic disc 'bunching up' of vessels in a small disc gives the appearance of swelling absence of optic cup	seen in hypermetropia
Tilted optic disc	can be associated with visual field loss
Optic nerve head drusen (Fig. 2) often associated with anomalous vasculature optic cup often absent drusen may be visible or not ('buried drusen') surface drusen 'autofluoresce' on fluorescein angiography can sometimes be seen as calcification on CT scan or signal void on MRI ophthalmic ultrasound often helpful for diagnosis	can be associated with visual field loss and occasionally haemorrhage at the optic disc
Other causes of optic disc swelling history unlikely to be confused with IIH pattern of visual loss usually different from that of papilloedema other investigations likely to be abnormal therefore unlikely to be a problem in the differential diagnosis of IIH	ischaemia, granulomata, Leber's, neoplastic infiltrations, etc.

Figure 2 Optic nerve head drusen.

- the absence of an optic cup in a mild-to-moderately 'swollen' disc;
- the presence of anomalous vasculature at the disc, e.g. trifurcations.

If there is still diagnostic doubt, the next move is to obtain a fluorescein angiogram of the discs (usually with help from an ophthalmologist). It is important to also ask for 'autofluorescence', as optic nerve head drusen often fluoresce before dye is injected. Another useful trick is to ask the medical photographer to photograph the discs while the patient is having their angiogram: this can prove invaluable in future management for the purposes of comparison.

Some patients with IIH have unilateral papilloedema (Sher *et al.* 1983; Wall & White 1998), and a small number have no papilloedema at all (Lipton & Michelson 1972; Mathew *et al.* 1996). Why this should be is not clear, but it may reflect anatomical variations at the ocular end of the optic nerve sheath: in particular there may be more effective lymphatic drainage channels in some nerves than others (Killer *et al.* 1999),

thereby protecting those eyes from the development of papilloedema in the presence of high CSF pressure. In patients with unilateral papilloedema, visual failure only occurs in those eyes that have papilloedema (Maxner *et al.* 1987) implying that it is the presence of the papilloedema rather than the raised CSF pressure *per se* that is responsible for the visual loss. If this is the case, then IIH without papilloedema becomes a completely different management issue: it is simply a matter of controlling the headache symptomatically as there is no threat to sight, and this is probably better done without recourse to invasive procedures for fear of potential complications (see below).

Risk factors

A huge number of potential risk factors has been reported, many of which are based simply on case reports of conditions probably associated by chance. A few case-control studies have tried to make sense of this but the studies are rather small (Ireland *et al.* 1990; Giuseffi *et al.* 1991).

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To date, the only factors found to be significant are female sex, reproductive age, menstrual irregularity, obesity, recent weight gain and hypertension. Curiously, there does not seem to be a significant effect of the oral contraceptive pill. Given that the pill has potentially prothrombotic effects, this seems intrinsically unlikely, and we are still recommending that patients avoid all hormonal forms of contraception. Likewise, tetracyclines, vitamin A analogues and uraemia are almost certainly associated considering the number of cases of IIH with these factors in the literature. Presumably much larger studies are needed to detect real and possibly causal associations.

And beware....

To make matters really difficult, there has been one case report of a patient whose clinical features were those of IIH and who responded to appropriate treatment, but whose CSF pressure was not elevated ('normal pressure pseudotumor') (Green *et al.* 1996). We have not come across this ourselves, but it is worth bearing in mind in case a similar patient appears. Likewise, it is worth remembering that a firm diagnosis of pseudopapilloedema does not necessarily preclude a diagnosis of IIH (Katz *et al.* 1988). However, in this situation one would expect abnormalities on fluorescein angiography (see above) so there should not be too much diagnostic confusion.

THE MANAGEMENT OF IDIOPATHIC INTRACRANIAL HYPERTENSION

Having made a diagnosis of IIH, there is then the major problem of what to do about it. As

mentioned above, there are numerous different forms of treatment that have been advocated (Table 2), and there are no randomised trials to help make sense of matters. We offer a few pointers in the hope that they might be of help.

Principles of follow-up

A small number of patients with IIH will experience sudden visual loss without any obvious warning. One assumes that this is less likely if the CSF pressure has been lowered by appropriate treatment. However, many of the non-invasive treatments (e.g. weight loss) require time to take effect, sometimes many months. If patients present with visual loss, invasive treatments need to be considered at the outset. However, if vision is normal, we believe that patients should be started on conservative treatment (e.g. diuretics and weight loss) and followed up. Follow up *must* include formal visual field assessment: rather like glaucoma, patients will probably be unaware of peripheral field loss, and clinical confrontation is simply inadequate to detect subtle field loss. Our practice is to perform full visual assessment, including visual fields, at diagnosis, again at 1 month and at 3 months, and then 3–6 monthly thereafter depending on what happens. Obviously the patient is seen more frequently if there are new symptoms or anything worrying develops on visual field testing.

Principles of treatment – general points

On the whole, treatments are either conservative (e.g. weight loss, drugs), or invasive (e.g. shunts, optic nerve sheath fenestration). We generally manage patients without visual loss conservatively. Patients who have visual loss at any point need consideration of more invasive therapy. The question of anticoagulation for venous sinus thrombosis is beyond the scope of this article.

Conservative treatment

Weight loss

Though there is no firm evidence, it is generally believed that weight loss (if it can be achieved) offers the potential for long-term cure in those patients who are overweight (Johnson *et al.* 1998). In our experience, this is extremely difficult to achieve, even though we strongly exhort all our patients to keep trying. The fact that patients are not particularly unwell means that it is often difficult to motivate them to suffer by

dieting. One suggestion is that patients are more likely to succeed if they are given a target weight to aim for rather than just being told to 'lose weight'. We have started doing this, but have yet to see any major impact.

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Diuretics

There are no comparative trials to suggest that any one diuretic is more effective than another. Traditionally, acetazolamide is the drug of first choice as it reduces the rate of CSF production by the choroid plexus (Rubin *et al.* 1966; McCarthy & Reed 1974). In the UK, we tend to start with 125 mg b.d., and increase the dose depending on clinical response. In our experience, patients tend to be easily troubled by acroparaesthesiae and altered taste and may not tolerate doses much above 250 mg t.d.s. This contrasts with the doses used in the USA where between 1 g and 4 g per day are recommended (Gucer & Viernstein 1978; Digre & Corbett 2001). We have also come across a number of patients who become very depressed on acetazolamide, an effect that is almost guaranteed to increase weight in those who started out obese, and we normally try to use a different diuretic (usually frusemide) in this situation.

Other diuretics that could be considered instead of acetazolamide (or in combination with it) include thiazide or loop diuretics. This is really a matter of trial and error. Like acetazolamide, frusemide has been shown to reduce the rate of CSF production (McCarthy & Reed 1974) so we tend to use it as our next choice.

Lumbar puncture

Repeated lumbar puncture (LP) is advocated by many clinicians. The rationale is that it can lower CSF pressure immediately and, supposedly, if performed often enough, turns the dura into a 'sieve', allowing CSF to escape continuously. In our experience, LP in obese patients is often difficult, and they do not like it, and

therefore we do not generally plan to do repeated LPs. In some patients, LP is symptomatically useful to relieve headache, but there are two problems with this. First, LP can undoubtedly have a placebo effect, and we have come across patients who have been treated this way for what eventually turned out not to be IIH at all (e.g. migraine with pseudopapilloedema in an obese individual). Second, some patients develop 'low pressure' headaches, which then generate considerable diagnostic confusion. In our opinion, the symptom of headache is best treated conservatively (diuretics, weight loss) and with additional symptomatic treatment (e.g. neck physiotherapy, amitriptyline and, if appropriate, antimigraine therapy) if at all possible. Having said this, lumbar puncture clearly has a place in the immediate management of acute visual failure while awaiting more definitive treatment. Similarly, it is useful in the management of IIH in pregnancy (see below).

IIH without papilloedema

As mentioned above, if patients are thought to suffer from this condition, there is evidence that their vision is not at risk (Maxner *et al.* 1987). Accordingly, we treat these patients conservatively with diuretics, analgesics and, if appropriate, neck physiotherapy and anti-migraine treatment. For the reasons given above, we would be wary of regular lumbar punctures, and we do not think that the risks of invasive procedures can be justified in this situation.

Invasive treatment

The two most common forms of invasive treatment are lumbo-peritoneal shunting and optic nerve sheath fenestration. A brief summary of the advantages and disadvantages of each is presented in Table 4.

Shunting is used by some clinicians to treat intractable headache in IIH, even in the absence of visual failure. We try to avoid this because the complication rate of shunts is not inconsiderable. Furthermore, the treatment effect can be difficult to interpret if headache is the only measure of efficacy. As mentioned above, we treat the symptom of headache conservatively. However, for the patient who develops visual failure, common sense would suggest that either form of treatment is potentially appropriate. There are no randomised trials of either intervention against conservative management, let alone a head-to-head comparison (Lueck &

Table 4 Optic nerve sheath fenestration vs. lumboperitoneal shunting

	ADVANTAGES	REPORTED COMPLICATIONS
Optic nerve sheath fenestration	relatively minor procedure no insertion of foreign body significant chance of improving headache	oculomotor disorders pupillary abnormalities infection orbital haemorrhage intraoperative glaucoma visual field defects/blindness death
Lumbo-peritoneal shunting	no direct involvement of visual pathways readily obtained in neuro-surgical departments	shunt obstruction low-pressure headache subdural haematoma catheter migration/dislocation radiculopathy/sciatica back pain/arachnoiditis shunt-related infection/meningitis abdominal pain/infection/haemorrhage tonsillar herniation syringomyelia death

McIlwaine 2002). Practically speaking therefore it is a matter of choosing a treatment based on the ease of obtaining it and an assessment of risk, i.e. is the procedure frequently performed by the relevant department in your hospital or not? We tend to opt for shunting because it is easier to organise and there is greater local experience of performing this procedure.

Other treatments

As listed in Table 3, a large number of other treatments have variously been advocated for IIH. Until they have been subjected to randomised trials, we do not see that they have a place in the routine management of IIH.

Pregnancy

Although IIH is not more common during pregnancy (Digre *et al.* 1984), many patients do develop it while pregnant. Management is made more difficult by the fact that acetazolamide, thiazides and loop diuretics are contraindicated in pregnancy. This is therefore a situation in which repeated lumbar punctures becomes a sensible first line management. But, as mentioned above, many patients find repeated lumbar punctures unpleasant, and we have had a number of patients who have preferred to undergo lumboperitoneal shunting.

SUMMARY

There is a desperate need for properly-designed clinical trials in IIH so that the various benefits and hazards of the different available treatments can be properly assessed. In the meantime, clinicians are left with patients to manage as best they can. We hope that some of our suggestions might prove helpful – they reflect what we do in the absence of good evidence.

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