Diagnosis and management of benign intracranial hypertension

D Soler, T Cox, P Bullock, D M Calver, R O Robinson

Benign intracranial hypertension (BIH) is a headache syndrome characterised by (1) raised cerebrospinal fluid (CSF) pressure in the absence of an intracranial mass lesion or ventricular dilatation; (2) normal spinal fluid composition; (3) usually normal findings on neurological examination except for papilloedema and an occasional VI nerve palsy; and (4) normal level of consciousness. The appellation “benign” means not fatal. The syndrome can, however, disrupt normal life and cause significant visual failure. It is an uncommon condition in childhood presenting about once or twice a year in a large referral hospital. Early recognition is important as timely intervention may preserve vision and enables the doctor to start the appropriate treatment to control headaches. Children as young as 4 months can be affected; sex distribution is equal.1 2

Which intracranial compartment is primarily responsible for raising CSF pressure in the absence of ventricular dilatation is still unclear. Theories of BIH pathophysiology have been based on neuroradiological studies on patients with BIH (computed tomograms, magnetic resonance imaging (MRI), magnetic resonance diffusion scans, and radionuclide cisternography) and CSF hydrodynamic studies. These include increased venous sinus pressure, decreased spinal fluid absorption, increased spinal fluid secretion, increased blood volume, and brain oedema.3 4

Since the first large report on childhood BIH in 1967, reports subsequently show a changing clinical picture over time in terms of possible aetiology and clinical presentation.5 6 Diagnosis is not always simply achieved. BIH can occur in the absence of papilloedema; a “normal resting” CSF pressure does not exclude the diagnosis in the presence of suggestive symptoms and signs.7 8 Review of our cases over the past 10 years confirms the wide clinical spectrum of this condition. Of the 22 cases seen, 15 presented with the classical picture of headaches, papilloedema, and a raised CSF pressure of more than 20 cm CSF; four patients showed an increased CSF pressure in the absence of papilloedema, and three patients showed fundoscopic evidence of papilloedema with “normal” to “borderline” CSF pressures of 7–13 cm CSF.

On the basis of our experience we have developed a standard and logical approach to diagnosis and treatment of children with BIH.

Diagnosis
Diagnostic process is one of exclusion based on clinical symptoms, neurological, ophthalmic, radiological, and CSF findings.

SYMPTOMS
Symptoms in BIH are non-specific and are those of increased intracranial pressure. Headaches, nausea/vomiting, and visual disturbances are the most common presenting symptoms.9 Headaches are predominantly frontal in location, become worse on lying down, and may wake the child at night. Increased intracranial pressure can exacerbate migraine. Some with a “mixed headache syndrome” are able to differentiate between the continuous daily headache of BIH which is worse on awaking from associated more severe, but intermittent, migrainous headaches.

Children describe a variety of visual disturbances—diplopia, transient visual loss/blurring of vision, photophobia, and “shimmering lights with coloured centres”. Other symptoms include lethargy and tiredness, dizziness, mood change, and intracranial buzzing sounds. Sleep and behaviour disturbances are often reported by parents in the young preverbal child. In contrast with patients with an intracranial mass lesion, the level of consciousness and intellectual functioning remains normal in BIH.

NEUROLOGICAL EXAMINATION
By definition, the neurological examination is normal apart from papilloedema or a sixth nerve palsy. Sixth nerve palsy is the most common neurological abnormality reported in 9–48% of children with BIH.10 Like others, we have seen an occasional III or IV nerve paresis. Other neurological abnormalities reported have included facial paresis, neck pain, seizures, hyperreflexia, bruit, hypoglossal nerve palsy, nystagmus, and choreiform movements.10 11 but these features are sufficiently rare that diagnosis of BIH should seriously be considered only after exclusion of an underlying intracranial mass lesion, an infectious or inflammatory process.

Although there are no case-control studies of aetiology in paediatric benign intracranial hypertension, various case studies have reported a number of associated conditions. Drug related cases and several endocrine abnormalities in children are among the most...
common reported associations. Treatment with tetracycline and isotretinoin for acne, nitrofurantoin prophylaxis for urinary infection, oral contraceptives (which are now given at younger and younger ages), and corticosteroid withdrawal including use for eczema have all been implicated. Hypothyroidism, hyperthyroidism, thyroid replacement, and chronic hypocalcaemia secondary to vitamin D deficiency or hypoparathyroidism need to be considered in selected cases.

OPHTHALMIC FINDINGS

The hallmark of BIH is papilloedema which may be bilateral, asymmetrical, or even unilateral. One of the main difficulties in diagnosis is differentiation of papilloedema from pseudopapilloedema. Optic nerve drusen, or an anomalously raised disc, in the presence of headaches can mimic papilloedema leading to a mistaken diagnosis of BIH. To add to the confusion, transient visual loss, haemorrhages on the disc, and visual field defects can be seen with drusen. We have seen drusen associated with papilloedema. Fluorescein angiography can help clarify the diagnosis, as in papilloedema the disc leaks diffusely, but with drusen there may be spots of autofluorescence before fluorescein is injected and no diffuse leakage is seen. These finer ocular subtleties clearly require the expertise of an ophthalmologist who must be involved early as an integral member of the team.

IMAGING

Normal imaging is a prerequisite for the correct diagnosis of BIH. Computed tomography and MRI confirm one of the pathognomonic features of BIH: undilated ventricles in the presence of intracranial hypertension. A computed tomogram and MRI imaging can supply important and predictive information about the state of the optic nerves in BIH. Thin section computed tomogram sections of the orbits may show hydrops of the optic nerve sheath and reversal of the optic nerve head. Severe visual loss in BIH patients is correlated with more frequent and more severe reversal of the optic nerve head. Because of the risk of radiation damage to the lens, however, high resolution images of the optic nerves are no longer used as widely as they once were. Hydrops of the optic nerve is also visible on MRI (fig 1). Orbital ultrasound is said to be another useful investigation in assessing the diameter of the optic nerve in relation to the CSF pressure. Magnetic resonance venography (MRV) is the procedure of choice for diagnosis of dural venous sinus thrombosis in BIH. Limited intracranial thrombosis, typically of the transverse sinus can present with BIH without localising neurological signs. It is important to establish the presence or otherwise of clot in the venous sinuses as steroid treatment in this situation may exacerbate the condition. Venous sinus thrombosis may be the presenting feature of a hypercoagulable state or may be caused by adjacent infection which may require treatment in its own right.

Figure 1 Fast spin echo T2 weighted axial MRI of the optic nerves in a patient with BIH.

CSF FINDINGS IN BIH—WHAT IS NORMAL CSF PRESSURE IN CHILDREN?

Increased intracranial pressure with normal CSF chemical and cellular analysis confirms the diagnosis of BIH. Obtaining reliable CSF pressure readings in children requires skill and often sedation. CSF pressure measurement via the lumbar route is always done after imaging has excluded a mass lesion. As there may be a wide diurnal fluctuation in CSF pressure, establishing an increased pressure is not always straightforward. For this reason, “normal” levels can be recorded in patients with elevated optic discs. In this situation, we advocate repeating the lumbar pressure measurement. When clinical suspicion is sufficiently strong, prolonged pressure monitoring may be indicated. The optimum technique for this is arguable. While the Camino catheter in the subarachnoid space is invasive, a catheter in the lumbar subarachnoid space connected to a pressure transducer may be less reliable. The upper limit of what may be regarded as a normal CSF pressure in children is not well defined. Data on normal values of CSF pressure in children are sparse and little is known of the characteristics of an intracranial pressure recording in healthy people. Most reviews on BIH in children consider 20 cm CSF as the upper limit of normal. Studies on intracranial pressure in infants, however, report that the upper limit of normal intracranial pressure is 7.5 cm CSF below the age of 2 years and 13.5 below the age of 5 years. The only controlled study on intracranial pressure found the upper limit of normal CSF pressure ranged between 20–25 cm CSF in normal non-obese and obese adults, whereas the majority of patients with acute BIH showed concentrations above this range. The age at which transition occurs to the pressure appropriate to that of adults is unknown.

BIH WITHOUT PAPILLOEDEMA

Various reports have confirmed that BIH can occur in the absence of papilloedema in adults and children. Recognition of this important headache syndrome has therapeutic implications in that these headaches respond to
Diagnosis and management of benign intracranial hypertension

pressure lowering treatment, including lumbo-peritoneal shunting. To date, there is no evidence that BIH without papilloedema is a threat to vision. Again, if clinical suspicion is sufficiently strong, repeat lumbar puncture is justified if the initial CSF pressure is normal.

Management
It is not possible to make evidence-based recommendations for the management of BIH because there are no randomised, controlled, double blind prospective studies of treatment, the natural history of the untreated condition is still unknown, and the underlying pathophysiology remains elusive. Although recovery is often gauged as resolved papilloedema, and is thought to be synonymous with the return of CSF pressure to normal, CSF pressure can be persistently increased for years after the initial episode of BIH which implies that BIH is a chronic condition.17 In addition, asymptomatic papilloedema with progressive visual loss has been reported months to years after the initial episode of increased intracranial pressure, thus emphasising the lack of a direct relation among papilloedema, symptoms of headache, visual disturbances, and increased CSF pressure. Therefore, at present, it is difficult to make rigid recommendations on how treatment is best assessed.

The various treatment modalities used in children have included corticosteroids, acetazolamide, frusemide, repeated lumbar punctures, and surgery. Most cases respond to non-surgical management. The goals of treatment are symptom relief and preservation of vision.

Acetazolamide, a carbonic anhydrase inhibitor, is perhaps the most commonly used drug of first choice. In adult patients, an oral dose of 1 g/day has been shown to resolve papilloedema and 4 g/day to decrease CSF pressure.15 Side effects are dose related, which may limit its use if high doses are required. These include gastrointestinal upset, perioral and digital tingling, loss of appetite, acidosis and electrolyte imbalance, and rarely nephrocalcinosis. Continuous medication may result in “low” pressure headaches, which are initiated or exacerbated by moving from the lying position to sitting or standing. In the absence of papilloedema, a trial of medication may help to clarify the situation.

STEROIDS
Evidence of the effectiveness of steroids in treating BIH relies on retrospective clinical analysis of patients with this condition. Clinical experience has shown that decrease of symptoms and resolution of papilloedema can be expected in the first two weeks of treatment. Our practice is to use steroids in those unresponsive or intolerant to acetazolamide treatment. Symptomatic relief occurred in three patients out of the eight treated with steroids.

REPEATED LUMBAR PUNCTURES
Although lumbar puncture can be used to lower CSF pressure, this has a short lived effect. CSF pressure can return to pretap concentrations within one to two hours. Spinal taps may be technically difficult and distressing to the child, especially if done repeatedly. This, together with the theoretical risk of developing intraspinal epidermoid tumours and the low back pain after the procedure, has discouraged us from using this option of treatment except as a temporary measure in a child with severe headaches.

SURGERY
Surgical management is indicated in those with deteriorating visual function and/or severe incapacitating headaches interfering with daily activities despite vigorous medical management. Currently, lumboperitoneal shunting (LPS) and optic nerve sheath fenestration (ONSF) are the two surgical procedures employed.

LUMBOPERITONEAL SHUNTING
LPS effectively lowers intraventricular pressure and relieves headaches and papilloedema. Unfortunately, it is fraught with problems. Shunt obstruction and low pressure headaches are the most common complications. Other complications include acquired cerebellar tonsillar herniation, syringomyelia, lumbar radiculopathy, and infection.25 The development of a lumboperitoneal catheter with a fixed resistance may prevent low pressure headaches or cerebellar tonsillar herniation. In addition, LPS has failed to halt progressive vision loss in documented cases.25 It may, however, be a treatment option in the patient whose documented increased intracranial pressure fails to respond to medical management.25 The long term outcome of visual function after LPS has not been reported. Our experience showed that headaches and visual function improved after LPS in all five of our patients who failed to respond to medical management.

OPTIC NERVE SHEATH FENESTRATION
ONSF is currently the favoured treatment for BIH in adults with deteriorating visual function despite medical management. The procedure successfully relieves papilloedema, rapidly reversing visual loss in most cases. The mechanism is not entirely clear, as pressure as measured by lumbar puncture in the immediate postoperative period is persistently increased. Despite this, two thirds of patients have improved headaches and few develop headaches requiring LPS after the procedure.23 24 The key to success with ONSF is early intervention and the appropriate expertise. Undoubtedly, better visual outcome is reported with ONSF after surgery for acute rather than chronic papilloedema. Thus, patients with BIH and vision threatening papilloedema should be offered ONSF without delay. Results are favourable in terms of visual outcome, there is an infrequent need for repeated surgery, and in expert hands the procedure is safe with few intraoperative or postoperative complications and no reported mortality. Eyes that have more than one ONSF, however, rarely stabilise or improve after surgery.25 Presently, there are no large reports
resolved after initial lumbar puncture

Continuing symptoms after lumbar puncture

Resolved on acetazolamide

Continuing symptoms on acetazolamide

Resolved after steroids

Continuing symptoms after steroids

Resolved after LPS

Continuing symptoms after LPS

Figure 2 Flowchart of results of treatment in our 22 patients with BIH.

Monitoring vision
Loss of visual function is the only serious permanent complication of BIH. Visual field loss or decreased visual acuity in children has been reported in 13–27%. This may be visible at presentation, progress during treatment, or be observed at transient visual loss in five out of the 22 children we followed up, and none had permanent visual impairment secondary to BIH.

Proposed management protocol
Having reviewed the various options, we offer our current schedule for management. While we would not claim that it is the best or even the most effective, it is at least consistent with what is known. We offer it as a base upon which others can improve. Figure 2 shows the result of treatment in our 22 patients.

(1) Investigations
Computed tomography/MRI are essential first investigations to exclude a mass lesion. MRV is done to exclude an occult venous sinus thrombosis if symptoms persist after initial lumbar puncture.

(2) Pressure measurement
After MRI/computed tomography, lumbar CSF pressure is measured carefully in the sedated child on spinal tap by manometry/pressure transducer. We recognise that volume loss to fill the manometer may lower the final pressure reading. Connecting the spinal needle to a pressure transducer is a counsel of perfection. If the pressure is increased, sufficient fluid is removed lower CSF pressure to 12–15 cm.
Diagnosis and management of benign intracranial hypertension

93

after the first presentation and unrecognised recurrence could result in irreversible visual loss.

(10) ANTIMIGRAINE TREATMENT
Persistent headaches with stable visual function may respond to antimigraine medication especially in those with a mixed headache syndrome.

(11) WEIGHT REDUCTION
Loss of weight has been shown to improve symptoms in adult patients.

(12) INDICATION FOR CSF—PRESSURE MONITORING
Preoperative evaluation of the very young child with persistent symptoms should be undertaken when visual fields cannot be tested.

Preoperative evaluation of children with unremitting symptoms in the absence of papilloedema who are unresponsive to medication should also be undertaken and they should be taken off the medication for a trial period. This process is essential in order to exclude low pressure headaches.

Conclusion
The correct diagnosis of BIH relies on the recognition of the typical symptoms, radiological exclusion of a mass lesion, and recognition of the possible diagnostic pitfalls. Visual impairment does occur in children and can occur at any stage. The incapacitating effect of headaches which interfere with the child’s daily activity cannot be ignored, however. Both factors have to be considered when deciding on the best treatment strategy. At the moment, it is difficult to make recommendations on how long to follow up children with BIH. This is because we do not understand the natural history of the condition and which factors predispose to a poor visual outcome. Meanwhile, all children regardless of age or ability to cooperate need careful neurological and ophthalmic follow up with the aim of preventing secondary optic atrophy. Future prospective studies on treatment will provide a scientific basis for a rational treatment plan for this condition.

Diagnosis and management of benign intracranial hypertension

D Soler, T Cox, P Bullock, D M Calver and R O Robinson

Arch Dis Child 1998 78: 89-94
doi: 10.1136/adc.78.1.89

Updated information and services can be found at:
http://adc.bmj.com/content/78/1/89

These include:

References
This article cites 28 articles, 5 of which you can access for free at:
http://adc.bmj.com/content/78/1/89#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections

- Headache (including migraine) (129)
- Pain (neurology) (598)
- Child health (3922)
- Hypertension (369)
- Eye Diseases (174)
- Clinical diagnostic tests (1133)
- Radiology (976)
- Radiology (diagnostics) (760)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/