

24.10.5 Idiopathic intracranial hypertension 6054

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section 24 Neurological disorders 6054 Chemotherapy of recurrent malignant glioma is associated with poor response rates, so new agents are constantly being investigated, including dendritic cell vaccines, immune checkpoint inhibitors, and antiangiogenesis agents. To date, no new treatment has been shown to be more effective than nitrosurea-based chemotherapy. Prognosis Overall survival from brain tumours has increased over the last 40 years but is still poor, around 40% at 1 year and 18% at 5 years. The four most important favourable prognostic factors for patients with gliomas are young age (less than 40 years), good performance status, low-grade oligodendroglial histology and molecular subtype (IDH1 mutation and, 1p/19q codeletion). The survival advantage for different treatments is modest in comparison. Any trial claiming a significant survival advantage for a new treatment therefore needs to show that this effect is independent of other prognostic factors. The median survival for patients with malignant gliomas varies from 6 months to 5 years, dependent on the aforementioned conditions. Generally, patients with glioblastoma survive for 1–2 years, whereas patients with anaplastic gliomas survive for 2 to 5 years, the exception being anaplastic oligodendrogliomas where survival can extend up to 10–20 years. The outlook for patients with low-grade gliomas is considerably better, with a median survival of 5–15 years depending on age, pre-operative performance status, histology, and tumour growth rate. Oligodendrogliomas have a more indolent course and are more chemosensitive than astrocytomas, so their prognosis is correspondingly better, with patients surviving 15–20 years after diagnosis, even with anaplastic histology. A recent genome wide analysis of almost 300 adult lower-grade gliomas correlating molecular data with clinical outcomes has shown that these tumours can be categorized into three molecular classes—those with IDH mutations and either 1p19q codeletions

(most favourable outcome) or TP53 mutations and those with IDH wild-type tumours, which behaved clinically more like glioblastoma. At least 40% of primary intracranial tumours are extra-axial (not arising from within the brain substance itself) and are thus readily treatable, if not curable. Some tumours, such as meningiomas and pituitary adenomas, are associated with 10-year survival rate of over 90% if diagnosed before irreversible neurological damage has occurred.

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24.10.5 Idiopathic intracranial hypertension

Alexandra Sinclair **ESSENTIALS** Idiopathic intracranial hypertension is a condition of raised intracranial pressure of unknown cause predominantly affecting obese women of childbearing age. Impaired absorption or increased production of cerebrospinal fluid, or raised venous pressure, may be contributory. Secondary causes include cerebral venous thrombosis, anaemia, endocrinopathies, and drugs (particularly tetracycline and vitamin A derivatives or supplements). Clinical features Characteristic presentation is with headache, which may be typical of raised intracranial pressure but is frequently nonspecific. Papilloedema is present, visual field defects are common, and (rarely) there may be sixth nerve palsy. Diagnosis, treatment, and prognosis Brain imaging, including venography, should exclude other causes of raised intracranial pressure. Lumbar puncture reveals pressure greater than 250 mm cerebrospinal fluid with normal constituents. Treatments aim to prevent permanent visual loss and manage

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6055 headaches. Therapy includes weight loss and acetazolamide, and other diuretics are sometimes used (without evidence of efficacy). For those with rapid visual decline, urgent surgical intervention (ventriculoperitoneal/lumboperitoneal shunt with a valve or optic nerve decompression) is essential. A temporizing lumbar drain should be considered if surgery is delayed. For most patients this is a chronic condition characterized by significantly disabling headaches and relapses, typically precipitated by weight gain. Introduction

Idiopathic intracranial hypertension (IIH) (also called pseudotumour cerebri and, previously, benign intracranial hypertension) is a syndrome of raised intracranial pressure in the absence of an intracranial mass lesion, enlargement of the cerebral ventricles, or venous sinus thrombosis. IIH affects predominantly obese women of childbearing age (>90%). The condition has considerable morbidity from permanent visual loss (up to 25% of cases) and chronic disabling headaches, which result in poor quality of life. Patients presenting acutely with papilloedema must be evaluated urgently for secondary causes of raised intracranial pressure (e.g. space occupying lesion and venous thrombosis). After this, the priority is to assess accurately the threat to vision. In most patients, the condition becomes chronic and the disease burden is mostly from chronic headaches, which need active management, alongside visual monitoring. This chapter does not cover paediatric IIH. The IIH management guidelines reflect the consensus from the Association of British Neurologists, The Royal College of Ophthalmologists, The Society for British Neurological Surgeons, key international opinion leaders and patients group (open access reference below) and are a key pragmatic resource for this condition.

Aetiology Elevated intracranial pressure (ICP) is caused by alterations in the volume of either cerebral blood, cerebrospinal fluid (CSF), or brain tissue. CSF volume is tightly regulated and is dependent upon the balance between CSF secretion and drainage. The mechanisms involved in regulation of CSF dynamics are poorly understood.

Epidemiology Idiopathic intracranial hypertension is comparatively rare in the general population, with an annual incidence of approximately 1 in 100 000, but this figure rises to 19 in 100 000 in obese women of childbearing age. Although more than 90% of patients are obese women, IIH can also occur in childhood and is rarely observed in men.

Pathogenesis The underlying pathogenesis is not fully understood, but is driven by disordered CSF dynamics. This may be through either excessive CSF production at the choroid plexus, reduced CSF drainage (predominantly by the arachnoid granulations) or elevated venous sinus pressure, or a combination of more than one of these factors. The mechanisms underlying the elevated intracranial pressure are not fully understood. As typical patients are obese and female, a pathogenic role for sex hormones and adipokines has been speculated.

Clinical features Characteristic presentation is with headache (94%) and papilloedema (although rarely patients can be diagnosed with IIH without papilloedema (IIHWOP)). Other symptoms include transient visual obscurations, pulsatile tinnitus, visual disturbance, double vision and, in some, nonspecific back pain, neck pain, and dizziness.

Headache This is the most common symptom and is present to some degree in almost every case. In those with significantly raised intracranial pressure (typically at presentation) the headache phenotype typically reflects that of raised intracranial pressure (worse in the mornings, on lying down, on bending down, and with Valsalva manoeuvres). The International Headache Society criteria for the diagnosis of headache associated with IIH (criteria 7.1.1) lists a headache with daily occurrence, which is diffuse and/or constant (typically non-pulsating) and aggravated by coughing and bending. These features are not exclusive to IIH (exacerbation of headache with coughing occurs in 70% of IIH patients and 35% of migraineurs while bending exacerbates 50% of IIH headaches and 44% of migraineurs). IIH headaches can resemble migraine and additionally may coexist with migraine. IIH headaches improve after lumbar puncture and CSF drainage in 72% (but improvement is also documented in 25% of migraineurs).

Papilloedema This is a virtually universal finding, but IIH without papilloedema (IIHWOP) is sometimes observed. Papilloedema results from swelling of the intraocular (prelaminar) portion of the optic nerve head. Severity of papilloedema can be classified using Frisen Grading (Fig. 24.10.5.1). Choroidal-retinal folds may be noted in IIH. Although typically identification of papilloedema is not challenging (particularly when there is moderate to severe swelling), distinguishing between mild papilloedema and pseudopapilloedema (e.g. due to

anomalous discs or optic nerve head drusen) can be a challenge. There is a risk that once a patient is labelled with papilloedema the diagnosis is then not questioned, which can lead to inappropriate investigations and treatment. An accurate assessment of the optic disc is, therefore, essential and if there is any doubt as to whether there is true papilloedema of the optic disc, an opinion by a senior ophthalmologist or neuro-ophthalmologist should be sought. Investigations that might be helpful include optical coherence tomography to quantify elevation of the retinal nerve fibre and identify drusen; orbital ultrasound B-scan can identify drusen and measure fluid in the optic nerve sheath and fluorescein angiography to look for early leakage from the blood vessels in papilloedema. Papilloedema and drusen may coexist in a minority of patients. Loss of spontaneous venous pulsations, particularly in a patient where they were previously noted, is an indicator of raised ICP. However, spontaneous venous pulsations cannot be identified in a large portion

section 24 Neurological disorders 6056 of normal individuals, making absence of this sign an unreliable indicator of elevated intracranial pressure. Significant compression of the optic nerve can result in permanent loss of retinal nerve fibres leading to optic atrophy. An atrophic optic nerve head cannot subsequently swell if there is disease recurrence. Papilloedema is typically bilateral but can be more severe in one eye or very rarely unilateral (unilateral papilloedema requires a more extensive imaging to exclude a lesion compressing the optic nerve). Visual symptoms In patients with papilloedema, transient visual obscurations (blackening or greying out of the vision, usually in both eyes for a few seconds, particularly on Valsalva or bending) likely result from intermittent ischaemia of the optic nerve. These episodes do not correlate with visual loss. Diplopia is most frequently horizontal due to a sixth cranial nerve palsy (false localizing sign resulting from elevated ICP). Monocular diplopia is extremely rare and can occur due to macula oedema (early) or epiretinal membrane (late) in the setting of significant papilloedema. Symptoms of visual loss are common and variable (e.g. dark area (scotoma), tunnel visual from peripheral constriction).

Pulsatile tinnitus Occurring in 60% of patients with active disease, this is usually bilateral but can be unilateral. It may be described as whooshing or akin to a heartbeat. Jugular venous compression can temporarily eliminate the sound. Diagnosis Terminology The term pseudotumour cerebri denotes elevated intracranial pressure in the absence of space occupying lesion, which may be due to several causes. If no underlying cause is identified, then the term IIH is appropriate. If an underlying cause is identified, the term secondary pseudotumour cerebri may be used with a description of the underlying cause (e.g. pseudotumour cerebri secondary to anaemia). The term fulminant IIH is used to describe patients with rapidly deteriorating vision of less than four weeks' duration. The term benign intracranial hypertension is no longer used. Diagnostic criteria See Table 24.10.5.1. Differential diagnosis IIH is a diagnosis of exclusion. Once papilloedema has been confirmed and space occupying lesion, hydrocephalus, and venous thrombosis excluded on imaging, a full history, drug history, and system enquiry are vital to elicit any treatable or underlying causes (Table 24.10.5.2). Investigations Once papilloedema has been confirmed, urgent imaging must be conducted to exclude hydrocephalus, space occupying lesions, and venous sinus thrombosis. MRI or CT with MR venography or CT venography is recommended. Imaging features which can be associated with IIH include: small ventricles, empty pituitary sella, optic nerve sheath fluid, and tortuosity and flattening of the posterior aspect of the optic globe. Cerebral venous sinus narrowing is often VFI: 98% MD: 1.98 DB P<10% PSD: 1.84 DB P<10% (g) (e) VFI: 100% MD: +0.09 DB PSD: 1.54 DB (h) (f) VFI: 96% MD: -3.75 DB P<1% PSD: 2.72 DB P<2% 30 30 (c) (a) (d) VFI: 85% MD: -3.51 DB P<2% PSD: 2.95 DB P<2% (b) LEFT RIGHT BASELINE 12MONTHS Fig. 24.10.5.1 A 32-year-old lady presented to her local optician with a three-month

history of headaches. She was diagnosed with papilloedema and sent to the local casualty department. On further questioning she admitted to pulsatile tinnitus and occasional bilateral blacking-out of her vision for a few seconds, particularly on bending down. Secondary causes of raised intracranial pressure were excluded (including normal MRI and MR venography). Examination revealed bilateral papilloedema (a, b), visual acuity was maintained at 6/4 bilaterally, Humphrey visual fields showed an enlarged blind spot particularly on the right (c, d), lumbar puncture pressure 41 cmCSF, and BMI 41. She was treated with a weight loss programme and at 12 months her BMI was 34, lumbar puncture pressure 28 cmCSF, and the papilloedema and perimetry had significantly improved (e, f, g, h).

24.10.5 Idiopathic intracranial hypertension 6057 noted, typically in the transverse sinus; these usually reverse with CSF drainage after lumbar puncture or shunting. Blood pressure measurement to exclude malignant hypertension and blood tests (blood count, ESR, CRP, and renal function) are recommended. Cerebrospinal fluid pressure Lumbar puncture (LP) should be performed in the lateral de- cubitus position. More than one manometer may be needed to accurately measure the pressure if it is above 40 cmCSF. Once the LP needle is in position, the legs should be straightened slightly to prevent the legs applying pressure to the abdomen which can falsely elevate pressure. Sufficient time must be allowed to let the pressure settle before a reading is taken (the pressure should fluctuate with respiration and can sometimes take a few minutes to settle). Draining the CSF to a reading of 17 cmCSF is undertaken in some centres but is not evidence based, nor is the importance of recording a closing pressure. Certainly, in those with significantly elevated pressure, draining CSF may have a temporary therapeutic effect. In some patients, LP is challenging, and image guidance may be needed. CSF should be analysed for microscopy, protein, and glucose (with paired serum glucose) to facilitate identification of secondary causes. A definitive cut-off for elevated LP pressure is debated. Those with pressure greater than 25 cmCSF should be evaluated for IIH. Only a weak, nonsignificant relationship exists between LP pressure and obesity. In normal individuals, pressures up to 28 cmCSF have been recorded. It should be remembered that LP pressure is a snapshot recording. LP pressure varies over 24 hours and the LP technique can impact on the reading; multiple LP attempts which puncture the dura may falsely lower the pressure while abdominal compression from the legs or Valsalva (breath-holding or crying) can falsely elevate the pressure. If the LP reading is out of keeping with the clinical picture it should be questioned and, in some cases, repeated. Atypical patients In patients with an atypical phenotype (men, children, and non-obese women) more extensive investigations are recommended to look for a secondary cause. MRI imaging with intravenous contrast (to exclude meningeal infiltration) and potentially MRI imaging of the proximal jugular veins (to identify proximal stenosis) may be indicated. A more extensive blood workup (vitamin A, endocrine testing) and sleep apnoea monitoring may identify alternative pathologies. Management Patients given a diagnosis of idiopathic intracranial hypertension are usually bewildered and frightened. It is important to provide a simple explanation of the nature of the condition and the rationale for treatment. Most patients are obese and weight loss is disease modifying; however, discussions regarding the importance of weight loss need to be sensitively managed. Table 24.10.5.1 Diagnostic criteria for idiopathic intracranial hypertension (IIH) Diagnostic criteria for adult IIH Diagnostic criteria for adult IIH without papilloedema (IIHWOP) A. Papilloedema • B-E satisfied with additional sixth nerve palsy B. Normal neurological examination except for cranial nerve abnormalities C. Neuroimaging: normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement or venous sinus thrombosis

on MRI and magnetic resonance venography; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used • In the absence of sixth nerve palsy, IIHWOP can be suggested if B–E are satisfied and at least 3 of: • Empty sella • Flattening of the posterior aspect of the optic globe • Optic nerve sheath distention +/- tortuous optic nerve • Transverse sinus stenosis D. Normal cerebrospinal fluid (CSF) composition E. Elevated lumbar puncture opening pressure (≥ 25 cmCSF) in a properly performed lumbar puncture A diagnosis of IIH is definite if A–E are fulfilled. The diagnosis is probable if A–D are met, but the CSF pressure is lower than specified. Table 24.10.5.2 Causes of raised intracranial pressure Secondary causes of raised intracranial pressure Drug-related causes of raised intracranial pressure a Venous sinus thrombosis (rarely internal jugular vein thrombosis) Fluoroquinolones and Tetracycline class antibiotics (this will save room over listing them) Anaemia Sulphonamides (e.g. trimethoprim/nitrofurantoin) Obstructive sleep apnoea Lithium CSF Hyperproteinaemia/Hypercellularity (e.g. spinal cord tumour/meningitis/Guillain Barré syndrome /subarachnoid haemorrhage) Depo provera and combined oral contraceptive pill Renal failure Vitamin A excess and retinoids Endocrine diseases (e.g. Addison's/Cushing's /hypothyroidism) Corticosteroids (and withdrawal)/Beclomethasone Ciclosporin a The most common drug-related causes are listed but others have been documented in case reports.

section 24 Neurological disorders 6058 Monitoring Visual monitoring is essential in active disease (Table 24.10.5.3). Imaging the fundus (e.g. colour fundus photography) is useful for longitudinal assessment. Optical coherence tomography (OCT) is increasingly being utilized to objectively monitor changes in the retinal nerve fibre layer. OCT must be interpreted cautiously as a reduction in elevation of the retinal nerve fibre layer may indicate resolving papilloedema and/or loss of retinal nerve fibres leading to optic atrophy. Treatment Weight loss Weight loss is the only current disease modifying therapy for IIH; however, the amount of weight loss needed is not fully elucidated. In the prospective study by Sinclair et al., weight loss (15% body weight) significantly reduced intracranial pressure and induced disease remission. Even modest weight loss of 5–10% can improve disease. Weight loss strategies are, however, notoriously difficult to achieve and maintain, and options for long-term weight management in IIH (such as bariatric surgery) need further evaluation. Drug treatments Pharmacological strategies aim to reduce CSF secretion and consequently ICP. The most frequently used is acetazolamide, a carbonic anhydrase inhibitor. There is class 1 evidence of efficacy to use acetazolamide in the subgroup of IIH patient with mild visual loss (the dose of acetazolamide used was higher than what is used in most centres; 40% were on doses of 4 g). In these patients, a small improvement in the visual fields was noted, although there was no improvement in visual acuity or headache disability. Studies have also demonstrated that up to 48% of patients may discontinue acetazolamide due to side effects (paraesthesia, altered taste, and nausea), despite moderate doses (1.5 g daily). The 2015 Cochrane review concluded that there was insufficient evidence to recommend or reject the efficacy of acetazolamide for treating IIH. In reality, prescribing practices vary and many patients are managed without acetazolamide. Modified release preparations may reduce patient-reported side effects and doses of 1 g daily are often adequate. If acetazolamide is not tolerated, other diuretics may be prescribed although there is no evidence of their efficacy. Topiramate is being increasingly prescribed as it is a useful migraine prophylactic agent, but also has weak carbonic anhydrase inhibitor activity and is an appetite suppressant in about 10% of patients. However, caution is needed as there are no randomized controlled trials supporting topiramate therapy in IIH and, additionally, the side effects of cognitive impairment and depression limit its use. Venous stenoses Venous sinus stenoses (narrowing), predominantly in the transverse sinuses, are noted in

IIH and are thought to arise as a consequence of the raised ICP. CSF drainage (through LP) with reduction in ICP has been shown to eliminate the stenoses. Consequently, the stenoses are not thought to represent an underlying cause of IIH but may exacerbate the condition by further impeding CSF drainage. Some specialist centres documented improvement in case series of IIH patients using stenting; however, recurrent stenoses adjacent to the stent and serious complications have been highlighted (venous sinus perforation, stent migration, in-stent thrombosis, subdural haemorrhage). Further evaluation in large clinical trials is required to evaluate short- and long-term efficacy before this treatment can be used routinely. Surgery

Surgical options are typically reserved for patients with rapidly declining visual function (fulminant IIH). Surgical options include shunting procedures (most commonly lumboperitoneal or ventriculoperitoneal shunts) or optic nerve sheath fenestration. These approaches can rapidly reduce intracranial pressure to help preserve vision acutely. However, in the longer-term shunt dysfunction is common and more than 50% of shunts need replacing; mostly in the first year after insertion (30% of patients will require multiple revisions). Low-pressure headaches after shunt insertion are also common (28%). Patients with ventriculoperitoneal shunts, particularly those with adjustable valves, which enable the degree of CSF drainage to be titrated, may have a better outcome but trial evidence is lacking in this area. CSF shunting in those with raised ICP but with unaffected visual function is not recommended. Shunting, as a treatment for headache, is rare due to documented shunt complications and development of secondary headaches post shunt. This may be an option for some patients in the setting of a specialist headache service. Shunting should be considered a temporary measure to save vision, but while the shunt is functioning the underlying disease still needs to be addressed, usually through weight loss.

Table 24.10.5.3 Visual monitoring

Visual acuity Should not be used in isolation as it is not sensitive to the type of visual loss seen in IIH. Rapidly declining acuity occurs late in the fulminant disease course with severe optic nerve ischaemia and should be urgently evaluated. Colour vision Typically evaluated with Ishihara plates and indicates optic nerve dysfunction. Pupil examination A relative afferent pupillary defect may be seen if there is asymmetrical optic nerve dysfunction. Extraocular muscles A unilateral or bilateral sixth nerve palsy maybe noted. Visual field assessment Formal automated visual field test must be performed (e.g. Humphrey or Goldmann visual field). Typical defects include enlarged blind spot, inferior nasal loss, and generalized constriction. An unreliable field may reflect poor patient technique and may improve with familiarization or changing to an alternate test such as Goldmann. If the perimetry is not in keeping with the rest of the visual assessment it should be interpreted cautiously as medically unexplained visual loss can occur. Confrontational visual fields are not sensitive and only pick up gross defects. Dilated fundus examination To document optic nerve head and macula findings, and to exclude ocular causes of disc swelling. Ideally assessed using a slit lamp (stereoscopic view). Contrast sensitivity is also a useful marker of optic nerve function. Visual evoked potentials are not useful and loss only occurs very late as optic atrophy develops.

24.10.5 Idiopathic intracranial hypertension 6059 For patients with fulminant IIH, urgent shunting is sight saving. If there is any delay to surgery, admission for a lumbar drain to lower ICP can help to preserve vision until a definitive shunting procedure is performed. Headache management For many patients, headache is the most disabling aspect of the disease and may continue for many years. The headache phenotype is often highly variable and multiple headache types can coexist (migraine, tension type, and medication overuse). Patients with cerebrospinal fluid shunts may have low-pressure headaches (due to shunt overdrainage) and cough headaches can occur (due to cerebellar tonsillar descent post lumboperitoneal shunting). There is a lack of evidence to guide

treatment of this aspect of the condition. Identifying and treating the principal headache types is suggested. Medication overuse is common and needs to be considered and managed. Migraine prophylactic agents may benefit those with predominant migraine phenotype headache (but the drug side effects such as weight gain (pizotifen, β -blockers, tricyclic antidepressants), depression (β -blockers, topiramate, flunarazine), and cognitive slowing (topiramate) can be counterproductive). ICP monitoring can be diagnostically helpful. Special circumstances Pregnancy IIH may develop de novo in the setting of a pregnancy. More often a patient with existing IIH becomes pregnant. Acetazolamide has teratogenic effects in animal studies; effects in patients are not extensively studied but no adverse events were noted in a case series of 50 women using acetazolamide in the first trimester. A discussion of the potential risks and benefits of acetazolamide is thus essential prior to conception. In those with stable disease, omitting acetazolamide, particularly during the first trimester is a reasonable approach. Dietetic advice on the amount of weight gain to target during pregnancy is very helpful, as rapid excessive weight gain may precipitate a flare up of the IIH (5–9 kg weight gain during the pregnancy is suggested for those with a body mass index (BMI) ≥ 30 kg/m²). Normal vaginal delivery is suitable in the majority unless there is rapidly declining vision, in which case a caesarean section may be considered to avoid a prolonged second stage of labour. There is no contraindication to spinal or epidural anaesthesia in IIH. For those with rapidly declining visual function during the first trimester, serial LPs may help control the ICP and allow CSF shunting surgery to be delayed until the second trimester. Contraception IIH has been documented secondary to contraceptive use, particularly combined oral preparations. For most patients with established stable disease, starting contraceptives does not affect disease course. IIH without papilloedema (IIHWOP) IIH has been documented in the absence of papilloedema but is rare. In the absence of papilloedema other features suggestive of raised ICP should be sought (symptoms and imaging features). The diagnostic criteria are shown in Table 24.10.5.1. In those with borderline raised pressure, particularly in the absence of other features of raised ICP, the diagnosis of IIHWOP is much less likely. As these patients do not have and do not develop papilloedema, visual monitoring is not needed. Prognosis The course of disease in IIH is variable. In a minority, ICP will settle following a single diagnostic LP. Incipient visual loss needs to be aggressively managed with surgical CSF diversion. For the majority, IIH is a chronic condition with disabling headaches that need active management. Fluctuations in weight may be reflected in changes in ICP and visual function. Poor visual prognosis is associated with male patients, severe papilloedema, decreased visual acuity at presentation, and diagnostic delay. MRI features are not predictive of visual outcome (Fig. 24.10.5.1).

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