= Idiopathic intracranial hypertension =

Idiopathic intracranial hypertension (IIH) , sometimes called by the older names benign intracranial hypertension (BIH) or pseudotumor cerebri (PTC) , is a neurological disorder that is characterized by increased intracranial pressure (pressure around the brain) in the absence of a tumor or other diseases . The main symptoms are stroke @-@ like headache , nausea , and vomiting , as well as pulsatile tinnitus (sounds perceived in the ears , with the sound occurring in the same rhythm as the pulse) , double vision and other visual symptoms . If untreated , it may lead to swelling of the optic disc in the eye , which can progress to vision loss .

IIH is diagnosed with a brain scan (to rule out other causes) and a lumbar puncture ; lumbar puncture may also provide temporary and sometimes permanent relief from the symptoms . Some respond to medication (with the drug acetazolamide , Topiramate , Furosemide , others or a combination) , but others require surgery to relieve the pressure . The condition may occur in all age groups , but is most common in women aged 20 ? 40 .

= = Signs and symptoms = =

The most common symptom of IIH is headache , which occurs in almost all (92 ? 94 %) cases . It is characteristically worse in the morning , generalized in character and throbbing in nature . It may be associated with nausea and vomiting . The headache can be made worse by any activity that further increases the intracranial pressure , such as coughing and sneezing . The pain may also be experienced in the neck and shoulders . Many have pulsatile tinnitus , a whooshing sensation in one or both ears (64 ? 87 %) ; this sound is synchronous with the pulse . Various other symptoms , such as numbness of the extremities , generalized weakness , loss of smell , and loss of coordination , are reported more rarely ; none are specific for IIH . In children , numerous nonspecific signs and symptoms may be present .

The increased pressure leads to compression and traction of the cranial nerves , a group of nerves that arise from the brain stem and supply the face and neck . Most commonly , the abducens nerve (sixth nerve) is involved . This nerve supplies the muscle that pulls the eye outward . Those with sixth nerve palsy therefore experience horizontal double vision which is worse when looking towards the affected side . More rarely , the oculomotor nerve and trochlear nerve (third and fourth nerve palsy , respectively) are affected ; both play a role in eye movements . The facial nerve (seventh cranial nerve) is affected occasionally ? - the result is total or partial weakness of the muscles of facial expression on one or both sides of the face .

The increased pressure leads to papilledema , which is swelling of the optic disc , the spot where the optic nerve enters the eyeball . This occurs in practically all cases of IIH , but not everyone experiences symptoms from this . Those who do experience symptoms typically report " transient visual obscurations " , episodes of difficulty seeing that occur in both eyes but not necessarily at the same time . Long @-@ term untreated papilledema leads to visual loss , initially in the periphery but progressively towards the center of vision .

Physical examination of the nervous system is typically normal apart from the presence of papilledema , which is seen on examination of the eye with a small device called an ophthalmoscope or in more detail with a fundus camera . If there are cranial nerve abnormalities , these may be noticed on eye examination in the form of a squint (third , fourth , or sixth nerve palsy) or as facial nerve palsy . If the papilledema has been longstanding , visual fields may be constricted and visual acuity may be decreased . Visual field testing by automated (Humphrey) perimetry is recommended as other methods of testing may be less accurate . Longstanding papilledema leads to optic atrophy , in which the disc looks pale and visual loss tends to be advanced .

= = Causes = =

" Idiopathic " means " of unknown etiology " . Therefore , IIH can only be diagnosed if there is no

alternative explanation for the symptoms . Intracranial pressure may be increased due to medications such as high @-@ dose vitamin A derivatives (e.g. isotretinoin for acne) , long @-@ term tetracycline antibiotics (for a variety of skin conditions) and hormonal contraceptives . There are numerous other diseases , mostly rare conditions , that may lead to intracranial hypertension . If there is an underlying cause , the condition is termed " secondary intracranial hypertension " . Common causes of secondary intracranial hypertension include obstructive sleep apnea (a sleep @-@ related breathing disorder) , systemic lupus erythematosus (SLE) , chronic kidney disease , and Behçet 's disease .

= = Mechanism = =

The cause of IIH is not known . The Monro @-@ Kellie rule states that the intracranial pressure is determined by the amount of brain tissue , cerebrospinal fluid (CSF) and blood inside the bony cranial vault . Three theories therefore exist as to why the pressure might be raised in IIH : an excess of CSF production , increased volume of blood or brain tissue , or obstruction of the veins that drain blood from the brain .

The first theory , that of increased production of cerebrospinal fluid , was proposed in early descriptions of the disease . However , there is no experimental data that supports a role for this process in IIH .

The second theory posits that either increased blood flow to the brain or increase in the brain tissue itself may result in the raised pressure . Little evidence has accumulated to support the suggestion that increased blood flow plays a role , but recently Bateman et al. in phase contrast MRA studies have quantified cerebral blood flow (CBF) in vivo and suggests that CBF is abnormally elevated in many patients with IIH . Both biopsy samples and various types of brain scans have shown an increased water content of the brain tissue . It remains unclear why this might be the case .

The third theory suggests that restricted venous drainage from the brain may be impaired resulting in congestion . Many patients with IIH have narrowing of the transverse sinuses . It is not clear whether this narrowing is the pathogenesis of the disease or a secondary phenomenon . It has been proposed that a positive biofeedback loop may exist , where raised ICP (intracranial pressure) causes venous narrowing in the transverse sinuses , resulting in venous hypertension (raised venous pressure) , decreased CSF resorption via arachnoid granulation and further rise in ICP .

= = Diagnosis = =

The diagnosis may be suspected on the basis of the history and examination . To confirm the diagnosis , as well as excluding alternative causes , several investigations are required ; more investigations may be performed if the history is not typical or the patient is more likely to have an alternative problem : children , men , the elderly , or women who are not overweight .

= = = Investigations = = =

Neuroimaging , usually with computed tomography (CT / CAT) or magnetic resonance imaging (MRI) , is used to exclude any mass lesions . In IIH these scans typically appear to be normal , although small or slit @-@ like ventricles , dilatation and buckling of the optic nerve sheaths and " empty sella sign " (flattening of the pituitary gland due to increased pressure) and enlargement of Meckel 's caves may be seen .

An MR venogram is also performed in most cases to exclude the possibility of venous sinus stenosis / obstruction or cerebral venous sinus thrombosis . A contrast @-@ enhanced MRV (ATECO) scan has a high detection rate for abnormal transverse sinus stenoses . These stenoses can be more adequately identified and assessed with catheter cerebral venography and manometry . Buckling of the bilateral optic nerves with increased perineural fluid is also often noted on MRI imaging .

Lumbar puncture is performed to measure the opening pressure , as well as to obtain cerebrospinal

fluid (CSF) to exclude alternative diagnoses . If the opening pressure is increased , CSF may be removed for transient relief (see below) . The CSF is examined for abnormal cells , infections , antibody levels , the glucose level , and protein levels . In IIH , by definition all of these are within their normal limits . Occasionally , the CSF pressure measurement may be normal despite very suggestive symptoms . This may be attributable to the fact that CSF pressure may fluctuate over the course of the normal day . If the suspicion of problems remains high , it may be necessary to perform more long @-@ term monitoring of the ICP by a pressure catheter .

= = = Classification = = =

The original criteria for IIH were described by Dandy in 1937 .
They were modified by Smith in 1985 to become the " modified Dandy criteria " . Smith included the use of more advanced imaging : Dandy had required ventriculography , but Smith replaced this with computed tomography . In a 2001 paper , Digre and Corbett amended Dandy 's criteria further . They added the requirement that the patient is awake and alert , as coma precludes adequate neurological assessment , and require exclusion of venous sinus thrombosis as an underlying cause . Furthermore , they added the requirement that no other cause for the raised ICP is found .
In a 2002 review , Friedman and Jacobson propose an alternative set of criteria , derived from Smith 's . These require the absence of symptoms that could not be explained by a diagnosis of IIH , but do not require the actual presence of any symptoms (such as headache) attributable to IIH . These criteria also require that the lumbar puncture is performed with patient lying sideways , as a lumbar puncture performed in the upright sitting position can lead to artificially high pressure measurements . Friedman and Jacobson also do not insist on MR venography for every patient ; rather , this is only required in atypical cases (see " diagnosis " above) .

= = Treatment = =

The primary goal in treatment of IIH is the prevention of visual loss and blindness , as well as symptom control . IIH is treated mainly through the reduction of CSF pressure and , where applicable , weight loss . IIH may resolve after initial treatment , may go into spontaneous remission (although it can still relapse at a later stage) , or may continue chronically .

= = = Lumbar puncture = = =

The first step in symptom control is drainage of cerebrospinal fluid by lumbar puncture . If necessary , this may be performed at the same time as a diagnostic LP (such as done in search of a CSF infection) . In some cases , this is sufficient to control the symptoms , and no further treatment is needed .
The procedure can be repeated if necessary , but this is generally taken as a clue that additional treatments may be required to control the symptoms and preserve vision . Repeated lumbar punctures are regarded as unpleasant by patients , and they present a danger of introducing spinal infections if done too often . Repeated lumbar punctures are sometimes needed to control the ICP urgently if the patient 's vision deteriorates rapidly .

= = = Medication = = =

The best @-@ studied medical treatment for intracranial hypertension is acetazolamide (Diamox) , which acts by inhibiting the enzyme carbonic anhydrase , and it reduces CSF production by six to 57 percent . It can cause the symptoms of hypokalemia (low blood potassium levels) , which include muscle weakness and tingling in the fingers . Acetazolamide cannot be used in pregnancy , since it has been shown to cause embryonic abnormalities in animal studies . Also , in human beings it has been shown to cause metabolic acidosis as well as disruptions in the blood electrolyte levels of newborn babies . The diuretic furosemide is sometimes used for a treatment if acetazolamide is not

tolerated , but this drug sometimes has little effect on the ICP .

Various analgesics (painkillers) may be used in controlling the headaches of intracranial hypertension . In addition to conventional agents such as paracetamol , a low dose of the antidepressant amitriptyline or the anticonvulsant topiramate have shown some additional benefit for pain relief .

The use of steroids in the attempt to reduce the ICP is controversial . These may be used in severe papilledema , but otherwise their use is discouraged .

= = = Venous sinus stenting = = =

Venous sinus stenoses leading to venous hypertension appear to play a significant part in relation to raised ICP , and stenting of a transverse sinus may resolve venous hypertension , leading to improved CSF resorption , decreased ICP , cure of papilloedema and other symptoms of IIH .

A self @-@ expanding metal stent is permanently deployed within the dominant transverse sinus across the stenosis under general anaesthesia . In general patients are discharged the next day . Patients require double antiplatelet therapy for a period of up to 3 months after the procedure and aspirin therapy for up to 1 year .

In a systematic analysis of 19 studies with 207 cases , there was an 87 % improvement in overall symptom rate and 90 % cure rate for treatment of papilloedema . Major complications only occurred in 3 / 207 patients (1 @.@ 4 %) . In the largest single series of transverse sinus stenting there was an 11 % rate of recurrence after one stent , requiring further stenting .

Due to the permanence of the stent and small but definite risk of complications , most experts will recommend that patients with IIH must have papilloedema and have failed medical therapy or are intolerant to medication before stenting is undertaken .

Consultation with a neurologist , neurophthalmologist and / or ophthalmologist in combination with a neurointerventionalist who performs the procedure is generally recommended .

= = = Surgery = = =

Two main surgical procedures exist in the treatment of IIH : optic nerve sheath decompression and fenestration and shunting . Surgery would normally only be offered if medical therapy is either unsuccessful or not tolerated . The choice between these two procedures depends on the predominant problem in IIH . Neither procedure is perfect : both may cause significant complications , and both may eventually fail in controlling the symptoms . There are no randomized controlled trials to guide the decision as to which procedure is best .

Optic nerve sheath fenestration is an ophthalmological operation that involves the making of an incision in the connective tissue lining of the optic nerve in its portion behind the eye . It is not entirely clear how it protects the eye from the raised pressure , but it may be the result of either diversion of the CSF into the orbit or the creation of an area of scar tissue that lowers the pressure . The effects on the intracranial pressure itself are more modest . Moreover , the procedure may lead to significant complications , including blindness in 1 ? 2 % . The procedure is therefore recommended mainly in those who have limited headache symptoms but significant papilledema or threatened vision , or in those who have undergone unsuccessful treatment with a shunt or have a contraindication for shunt surgery .

Shunt surgery , usually performed by neurosurgeons , involves the creation of a conduit by which CSF can be drained into another body cavity . The initial procedure is usually a lumboperitoneal (LP) shunt , which connects the subarachnoid space in the lumbar spine with the peritoneal cavity . Generally , a pressure valve is included in the circuit to avoid excessive drainage when the patient is erect . LP shunting provides long @-@ term relief in about half the cases ; others require revision of the shunt , often on more than one occasion ? usually due to shunt obstruction . If the lumboperitoneal shunt needs repeated revisions , a ventriculoatrial or ventriculoperitoneal shunt may be considered . These shunts are inserted in one of the lateral ventricles of the brain , usually by stereotactic surgery , and then connected either to the right atrium of the heart or the peritoneal

cavity , respectively . Given the reduced need for revisions in ventricular shunts , it is possible that this procedure will become the first @-@ line type of shunt treatment .
It has been shown that in obese people , bariatric surgery (and especially gastric bypass surgery) can lead to resolution of the condition in over 95 % .

= = Prognosis = =

It is not known what percentage of people with IIH will remit spontaneously , and what percentage will develop chronic disease .
IIH does not normally affect life expectancy . The major complications from IIH arise from untreated or treatment @-@ resistant papilledema . In various case series , the long @-@ term risk of ones vision being significantly affected by IIH is reported to lie anywhere between 10 and 25 % .

= = Epidemiology = =

On average , IIH occurs in about one per 100 @,@ 000 people , and can occur in children and adults . The median age at diagnosis is 30 . IIH occurs predominantly in women , especially in the ages 20 to 45 , who are four to eight times more likely than men to be affected . Overweight and obesity strongly predispose a person to IIH : women who are more than ten percent over their ideal body weight are thirteen times more likely to develop IIH , and this figure goes up to nineteen times in women who are more than twenty percent over their ideal body weight . In men this relationship also exists , but the increase is only five @-@ fold in those over 20 percent above their ideal body weight .
Despite several reports of IIH in families , there is no known genetic cause for IIH . People from all ethnicities may develop IIH . In children , there is no difference in incidence between males and females .
From national hospital admission databases it appears that the need for neurosurgical intervention for IIH has increased markedly over the period between 1988 and 2002 . This has been attributed at least in part to the rising prevalence of obesity , although some of this increase may be explained by the increased popularity of shunting over optic nerve sheath fenestration .

= = History = =

The first report of IIH was by the German physician Heinrich Quincke , who described it in 1893 under the name serous meningitis . The term " pseudotumor cerebri " was introduced in 1904 by his compatriot Max Nonne . Numerous other cases appeared in the literature subsequently ; in many cases , the raised intracranial pressure may actually have resulted from underlying conditions . For instance , the otitic hydrocephalus reported by London neurologist Sir Charles Symonds may have resulted from venous sinus thrombosis caused by middle ear infection . Diagnostic criteria for IIH were developed in 1937 by the Baltimore neurosurgeon Walter Dandy ; Dandy also introduced subtemporal decompressive surgery in the treatment of the condition .
The terms " benign " and " pseudotumor " derive from the fact that increased intracranial pressure may be associated with brain tumors . Those patients in whom no tumour was found were therefore diagnosed with " pseudotumor cerebri " (a disease mimicking a brain tumor) . The disease was renamed " benign intracranial hypertension " in 1955 to distinguish it from intracranial hypertension due to life @-@ threatening diseases (such as cancer) ; however , this was also felt to be misleading because any disease that can blind someone should not be thought of as benign , and the name was therefore revised in 1989 to " idiopathic (of no identifiable cause) intracranial hypertension " .
Shunt surgery was introduced in 1949 ; initially , ventriculoperitoneal shunts were used . In 1971 , good results were reported with lumboperitoneal shunting . Negative reports on shunting in the 1980s led to a brief period (1988 ? 1993) during which optic nerve fenestration (which had initially been described in an unrelated condition in 1871) was more popular . Since then , shunting is

recommended predominantly , with occasional exceptions .