

What is IIH?

Idiopathic Intracranial Hypertension (IIH) is a neurological condition defined by increase intracranial pressure around the brain without the presence of tumour or disease. IIH is also known by its earlier name: Benign Intracranial Hypertension (BIH) but this term is little used nowadays because the condition can cause visual loss and therefore is not harmless or benign. You may also occasionally see it referred to as Pseudotumor Cerebri (PTC) because some sufferers present with signs and symptoms of a brain tumour despite no tumour being present.

The space around the brain is filled with CSF, if due to a variety of factors the pressure around the brain rises then the space containing the fluid cannot expand. It is this excessively high pressure that produces the symptoms of IIH.

Common symptoms of IIH

Severe headaches, papilloedema (but not in all cases), transient visual obscurations, diplopia, pulsatile tinnitus and pain behind the eye and with eye movement.

Other symptoms reported by sufferers include: nausea, vomiting, fatigue, photophobia, problems with balance and spatial awareness, aphasia, memory loss, confusion, decreased depth perception and peripheral vision.

Some children are often too young to report their symptoms adequately and can present with many nonspecific symptoms such as mood swings and more. Although many sufferers have symptoms in common, each sufferer is an individual and should be treated accordingly.

Medical Conditions and Drugs with Increased Susceptibility to Developing IIH

Acute Sinusitis, Addison's Disease, Adrenal Insufficiency, Anaemia, Behcet's Disease, Blood Clotting Disorders, Chronic Ear Diseases, Chronic Kidney Disease, Gluten Intolerance, Guillain Barre Syndrome, Head Trauma, Hyperadrenalism, Hyperaldosteronism, Hypercoagulable States, Infections, Kidney Failure, Lactose Intolerance, Leukaemia, Lyme Disease, Malnutrition, Menopause, Obstructive Sleep Apnoea, Overactive & Underactive Thyroid/Hypothyroidism & Hyperthyroidism, Polycystic Ovary Syndrome, Pregnancy, Protein Malnutrition, Sarcoidosis, SLE, Vitamin D Deficiency.

All trans retinoic acid (used in the treatment of Promyelocytic Leukaemia), Amiodarone, Contraceptives, Corticosteroid treatment (including withdrawal), Cyclosporine, Excessive ingestions of Vitamin A (Hypervitaminosis A), Growth Hormone, Indomethacin, Isotretinoin (Accutane) for acne, Lithium, Minocycline, Nalidixic Acid, Nitrofurantoin, Phenytoin, Steroid withdrawal, Tetracycline, Thyroid Replacement, Vasopressin. (this is not an exhaustive list).

Causes

Narrowing of the Dural Venous Sinus is now known to cause Intracranial Hypertension, it is important that this has been ruled out in all patients thought to have IIH.

Helping your patient cope with symptoms

IIH is an invisible condition and it is hard to understand what the sufferer feels or is experiencing. For the patient with IIH the symptoms can be extremely debilitating and disabling. That said, IIH affects everyone differently, some sufferers may have more severe symptoms than others. Sufferers may no longer be able to enjoy social activities and may be limited in the type of activities they can do. They may feel unable to stay out for long periods of time or have to cancel or change plans at the last minute if they feel unwell. Many IIH patients report feeling fatigued, pain itself can be very tiring and draining. The headaches associated with IIH can be extremely painful and often do not respond to analgesics. People with IIH often suffer from pulsatile tinnitus which can be a symptom that occurs most of the time. Silence is the worst thing for tinnitus because the tinnitus simply sounds louder. Tinnitus can be extremely distressing especially if it interferes with sleep, sufferers may benefit from listening to music or audio books to aid sleep.

Patients with IIH describe the nausea and dizziness they feel as though the room is spinning or rocking, like feeling sea-sick on a boat and their balance problems as though they are 'drunk', or being pushed by an unseen force. The visual problems associated with IIH can be extremely disabling.

Working to relieve the pressure!

Many patients with IIH can experience a decrease in visual acuity. IIH sufferers may experience blurred and double vision, lack of depth perception and may find walking around without bumping into things or crossing roads difficult. They may also find reading difficult. Those who suffer from photophobia may find household lights too bright and may want to draw curtains and close blinds, they may benefit from having dimmer switches fitted to lights around the house. Problems with memory or confusion and disorientation can make it difficult for some sufferers with IIH to go outside alone. They may need help with many everyday tasks which most people take for granted. As well as the physical symptoms, sufferers also have to cope with the emotional side of the condition, worries about the future and their symptoms.

Depression is quite common in people with chronic health conditions and should be treated accordingly. Sufferers who experience visual problems may be concerned about worsening vision or losing their vision, though only a small minority of sufferer will suffer substantial permanent visual loss. Sufferers of IIH may feel frustrated by the limitations of their symptoms. If they have difficulty getting around alone, they may feel isolated and distressed by their lack of independence or frustration and guilt that they have to rely on others to care for them.

As well as the physical and emotional effects of the symptoms of IIH, sufferers also have to cope with the treatments needed to control the condition. There is currently no cure for IIH and the success of treatment is variable. Medical treatment can have a variety of unwanted or severe side effects that come with the medications prescribed. Some doctors will only perform lumbar punctures for diagnostic and monitoring reasons, whereas some will perform them therapeutically to ease symptoms. Lumbar punctures can temporarily ease symptoms but can be uncomfortable and painful. If lumbar punctures are needed on a regular basis, they can cause long term back pain.

Drugs used to treat IIH

There is currently no 'cure' for idiopathic intracranial hypertension (IIH) but some medications can help to lower intracranial pressure and relieve some of the symptoms of IIH. There are two main types of medication that are prescribed - diuretics and analgesics. **Acetazolamide** is the most commonly prescribed diuretic. It is a carbonic anhydrase inhibitor used for glaucoma, some types of epilepsy and fluid retention. Acetazolamide can reduce CSF production in most patients and can be taken in a slow release form called Diamox SR if patients find they cannot tolerate the standard version. **Patients who are prescribed Acetazolamide must be monitored closely, have regular blood tests to check their electrolyte levels as Acetazolamide can deplete the body's stores of potassium.** If potassium levels become very low it may be necessary to prescribe potassium supplements. Advising your patients to eat potassium rich foods such as baked potatoes, tomato products, raisins, dried apricots and bananas can help increase potassium levels and lessen its side effects. Taking medication with food can help with some of the side effects experienced. **Topiramate** (Topamax®) is also used to treat IIH. Originally used as a weight loss medication Topiramate is used as an anti-seizure medication and can reduce the production of CSF. At this time there

have been no studies to show that it works any more efficiently than Acetazolamide. Again side effects are common and can be severe in some cases.

A side effect of all diuretics is the need to urinate more frequently, patients should be made aware that they should still drink plenty of fluids every day to prevent dehydration and to help the body flush away toxins and that drinking lots of fluids will help to lessen the side effects of diuretics.

Other diuretics that may be prescribed include **Furosemide**® (Frusol®, Lasix®, Duimide®, Frumil®) and **Bendrofluazide**. Sometimes more than one diuretic may be used in combination to achieve the desired effect and minimise side effects.

Sufferers of IIH will try anything to relieve themselves of the debilitating headache that many of them suffer from. It is not unheard of for some people with IIH to accidentally overdose on analgesics so it is important you are made aware of any over the counter analgesics they are already taking prior to prescribing a stronger analgesic. Stronger analgesics that have been prescribed to IIH sufferers include **Tramadol**, **Fentanyl** and **Morphine**. You will be aware of the risks of rebound headaches associated with some analgesics, therefore we suggest close monitoring of patients taking prescribed analgesics.

Working to relieve the pressure!

Other types of medication used to treat symptoms associated with IIH.

Antidepressants can also be used to treat IIH patients, drugs such as **Amitriptyline**, **Imipramine** (Tofranil), and **Diazepam** have all been used with varying success. Also drugs that are used to treat neuropathic pain such as **Gabapentin** (Neurontin®).

Sometimes a patient may complain of severe nausea, Anti-emetics prescribed for severe nausea associated with IIH are **Prochlorperazine** (Stemetil®, Buccastem®), **Domperidone** (Motilium®), and **Betahistine** (Serc®).

Surgical options for treating IIH

Shunts, both Ventricular peritoneal (VP) and Lumbo-peritoneal (LP). (If a patient has an LP shunt and comes to you complaining of low pressure headache symptoms please be mindful of the shunt overdraining. Overdraining LP shunts can cause a Chiari Malformation and an MRI of the head/neck should be requested for these patients. If a Chiari is found then it is usual for the LP shunt to be removed and a VP shunt to be inserted.)

Stenting of the Dural Venous Sinus

Optic Nerve Sheath Fenestration (rarely)

Subtemporal decompression

Surgical treatments for IIH

Lumbar puncture (LP)

Intracranial pressure (ICP) monitoring

IIH UK

IIH UK is a national charity working to support sufferers of IIH. We are currently forming a list of clinicians throughout the UK who have an interest in IIH should your patient need referral. In addition, we have a range of leaflets which have been medically verified on our website.

If you would like further information regarding IIH, please visit our website at www.iih.org.uk

Or email us at info@iih.org.uk