

Information for Patients

Idiopathic Intracranial Hypertension (IIH)

What is IIH?

Idiopathic Intracranial hypertension (IIH) is a rare condition affecting about one or two in every 100,000 people, most of them women, but men and children can also be affected.

The space around the brain is filled with water like fluid known as cerebrospinal fluid (CSF). IIH is a neurological condition in which there is too much CSF present, which causes pressure around the brain. This causes headaches, swelling of the optic nerves (papilloedema) and can result in loss of vision or blindness.

IIH was formerly known as Benign Intracranial Hypertension (BIH) although this name is not used very often now as the condition can cause permanent visual loss and therefore is not harmless or benign. It is also sometimes known as Pseudo Tumour Cerebri (PTC), as there are some of the signs and symptoms of a brain tumour without a tumour being present (*pseudo* meaning false).

What are the symptoms of IIH?

The most common symptoms of IIH are:

- Severe headache.
- Loss of field of vision and rarely of central vision.
- Transient blurred vision.
- Double vision.
- Photophobia (light sensitivity).
- Pulsatile tinnitus ('whooshing noise' in the ears in time with the pulse).

How is it diagnosed?

Diagnosis of IIH is made by identifying the typical symptoms of the disease and ruling out other conditions that can cause similar signs and symptoms. Neurological examination is often normal apart from the presence of papilloedema in some patients but not all. A CT scan (computerised tomography) or MRI scan (magnetic resonance imaging) of the brain is usually performed and the results are often normal. A CT scan is a special kind of X-ray machine but allows much more detail to be seen. A MRI scan uses magnetic and radio waves and is able to take pictures from many angles. If a scan is required, your doctor will decide which one is best.

Definitive diagnosis of IIH is made by performing a procedure called a lumbar puncture, which shows the pressure of cerebrospinal fluid (CSF) to be higher than normal.

Visual field testing may show an enlarged blind spot and peripheral field constriction. Repeated visual field testing is carried out to monitor the disease.

What causes IIH?

The causes of IIH are unknown. However, withdrawal of steroids, taking large doses of vitamin A or intake of foods containing large amounts of vitamin A, the use of body building-type steroids and possibly hormonal changes, obesity and taking certain drugs can cause raised intracranial pressure. Blood clots in the veins draining the brain can also cause increased intracranial pressure.

What is the treatment of IIH?

The aim of treatment is to relieve the symptoms of raised intracranial pressure and prevent the progression of optic nerve damage. If a patient is overweight, most doctors recommend losing weight. In addition, other treatments, either medical or surgical, may be necessary depending on the severity of the condition.

- **Medical treatment**

The most commonly prescribed medication is Acetazolamide (Diamox) which acts by reducing the production of CSF. Whilst this is generally a safe treatment, most patients will experience tingling of the fingers and toes as a side effect of the medication and it has been known to cause birth defects. It is therefore important to inform your doctor if you are pregnant or planning a pregnancy.

- **Surgical treatment**

If visual problems persist in spite of treatment, surgical treatments may be considered. These procedures aim to relieve pressure on the optic nerve and drain excess CSF in order to reduce the intracranial pressure.

The most common procedure is the surgical insertion of a shunt. Surgical intervention is usually only used as a last resort to protect vision. This is due to the potential complications associated with surgery and anaesthesia, including infection, blockage and over-drainage of the shunt.

While for some patients their symptoms may disappear as quickly as they came on, for others, a combination of medical and/or surgical treatments are required to control their condition. In some cases both medical and surgical treatments can be limited in their effectiveness, and symptoms may remain. For these patients, other medications may be required.

Because each patient is different, the information contained in this leaflet is a general guide only. Should you require any further information please contact your GP.