Idiopathic Intracranial Hypertension

In idiopathic intracranial hypertension (IIH) there is raised pressure within the skull (raised intracranial pressure), which puts pressure on the brain. Idiopathic means that the cause of this raised pressure is unknown. The main symptoms are headache and loss of sight (visual loss). It mostly affects women of childbearing age who are overweight or obese. Treatment is aimed at preventing permanent visual loss and includes treatment with medicines. Brain surgery (neurosurgery) may also be used. For many people, a combination of medical and surgical treatment can help to control their symptoms well.

Some anatomy around the brain

The meninges form the protective lining that surrounds the brain within the skull, and the spinal cord within the backbone (vertebral column). There are three layers of meninges:

- The outermost layer that lies next to the skull or the vertebral column is called the dura mater.
- The middle layer is called the arachnoid mater.
- The inner layer that is closest to the brain or the spinal cord is called the pia mater.

There are also three spaces between the layers of meninges:

- The epidural space - the space between the vertebral column and the dura mater. (This is only a potential space in the head between the skull and the dura mater.)
- The subdural space - the space between the dura mater and the arachnoid mater.
- The subarachnoid space - the space between the arachnoid mater and pia mater.
Understanding cerebrospinal fluid

The brain and the spinal cord are bathed in a clear, watery fluid called cerebrospinal fluid (CSF). This fluid helps to cushion the brain from contact with the skull when the head is moved vigorously. CSF is made by a network of blood vessels inside the ventricles of the brain. The ventricles are essentially four cavities within the brain. The CSF circulates through the brain ventricles into the subarachnoid space. Eventually, CSF is absorbed into the bloodstream through some one-way valves called the arachnoid villi.

What is idiopathic intracranial hypertension (IIH) and what causes it?

In IIH there is raised pressure within the skull (raised intracranial pressure). Idiopathic means that the cause of this raised pressure is unknown. Various theories exist as to what may be the cause. Essentially, for some reason, there is too much CSF. The subarachnoid space that contains the CSF cannot expand and, because of this, the pressure around the brain rises.

This raised pressure leads to the symptoms of IIH. It can cause headaches and also swelling of the first part of the optic nerve - the optic disc - at the back of the eye (this is known as papilloedema). If papilloedema is not recognised and treated, it can lead to a condition called optic atrophy where there is deterioration and loss of function (degeneration) of the optic nerve. Severe sight impairment (blindness) can occur as a result of this.

IIH is also known as benign intracranial hypertension. However, this name is not being used as much now. This is because the condition isn't harmless (benign). It can cause some quite disabling symptoms and can lead to loss of vision if it is not treated. Another old name is 'pseudotumour cerebri', as it can lead to some signs and symptoms of a brain tumour, without a brain tumour actually being present.

Who gets idiopathic intracranial hypertension (IIH)?

IIH is rare. It affects 1 or 2 people in every 100,000. It mostly affects women of childbearing age who are overweight or obese. However, men and children can sometimes be affected as well as people who are not overweight.

More than 9 out of 10 people with IIH are obese women in the reproductive age range. However, in someone who is not overweight, there are some 'risk factors' that are thought to be associated with the development of IIH. There are many such rare 'associations' but some of these include:

- Taking (or after stopping) certain medicines such as steroids, some antibiotics and oral contraceptive pills.
- Other diseases such as systemic lupus erythematosus, sarcoidosis and kidney disease.
- Pregnancy.

What are the symptoms of idiopathic intracranial hypertension (IIH)?

There may be many different symptoms associated with this condition (see Further help & information and references, below). However, the following three are typical: headache, tinnitus and changes in vision. The most prominent symptom is headache. This can be severe and is a long-term (chronic) headache. It can vary in its location and may come and go. Some people can feel sick or be sick (vomit) with the headache. You may also notice tinnitus in one or both of your ears. This is typically a pulsating, rhythmic sound that you can hear in your ear.

You may also notice some temporary sight (visual) disturbance or temporary loss of vision. For example, you may have dimming or loss of your vision in one or both of your eyes, lasting for a few seconds. This can sometimes come on after bending over. You may have some double vision when looking from side to side, or pain behind the eyes on eye movement. You may also notice a progressive permanent loss of vision in one or both of your eyes.
How is idiopathic intracranial hypertension (IIH) diagnosed?

If you visit your doctor, complaining of headaches and/or sight (visual) symptoms, your doctor will usually discuss your symptoms with you. They may examine your eyes with a hand-held instrument for looking in the back of the eye (an ophthalmoscope). This may show swelling at the back of the eye (papilloedema). However, not everyone with IIH has papilloedema.

Papilloedema is a sign of raised pressure within the skull (raised intracranial pressure). Therefore, the main thing when diagnosing IIH is to rule out other causes of raised pressure within the skull. These may include problems such as water on the brain (hydrocephalus) or a brain tumour. Your doctor will usually refer you to a specialist for investigations to rule out other causes. Investigations can include, for example, MRI or CT scanning of your brain.

Your may also have a more detailed eye examination. This will allow an eye specialist to examine the back of your eyes fully. You may have visual field testing to see if there are any signs of loss of vision in parts of one or both of your eyes. (Your visual field is the area in front of your eye in any part of which an object can be seen without moving your eye.) You may have your colour vision tested, as this can also be affected in IIH.

You may also have tests on your CSF by doing a lumbar puncture. This will show raised CSF pressure if you have IIH. A lumbar puncture - sometimes called a spinal tap - is a procedure where a sample of CSF is taken for testing. A needle is pushed through the skin and tissues between two vertebrae and into the subarachnoid space around the spinal cord which is filled with CSF. See separate leaflet called Lumbar Puncture for more details.

What are the aims of treatment?

If you are diagnosed with idiopathic intracranial hypertension (IIH), it is important that your vision be closely monitored to look for any changes or early signs of loss of vision. This can be done by regularly measuring your 'visual acuity' (the size of letters that can be read on a wall chart), along with checking your visual fields. Any signs of deterioration in your vision can mean that your treatment needs to be adjusted. Essentially, treatment for IIH aims to prevent any deterioration in vision. However, treatment also aims to reduce other symptoms such as headaches.

What treatments are available?

Medical treatment

If you do not have any loss of vision, the usual treatment for idiopathic intracranial hypertension (IIH) is with a group of medicines called carbonic anhydrase inhibitors. Commonly a medicine called acetazolamide is used. These medicines help to lower the pressure within the skull, probably by reducing the production of CSF. Furosemide is another medicine used. Various painkillers may also be used to help relieve headaches. A short course of steroid tablets is also sometimes used.

Treatment with medicines can work well for many people. However, if your symptoms do not improve with medical treatment or you have new loss of vision, surgery may be considered.

Surgical treatment

Surgery aims to reduce the pressure within the skull (intracranial pressure). There are two main procedures that are done. The first is to put in place a tube (called a shunt) to drain away the excess CSF. This is probably the most common surgical procedure that is used. The shunt is run from either of the following:

- The subarachnoid space in the lower part of the spine into the tummy (abdomen) - called a lumbar-peritoneal shunt.
- The ventricles in the brain into the abdomen (called a ventriculo-peritoneal shunt).

However, there can be problems with a shunt. It can become infected, it can drain away too much CSF, or sometimes it can become blocked. Therefore, someone who has had a shunt inserted needs regular check-ups to make sure that it is working normally.
The second type of surgical treatment is around the eye. A procedure called optic nerve sheath fenestration can be carried out. Small cuts are made in the protective sheath around the optic nerve. This allows CSF to escape and the pressure on the optic nerve is reduced. This procedure can be very good at helping sight (visual) symptoms associated with IIH. However, it may have little effect on other symptoms, including headache. This is because it tends to have little effect on reducing overall pressure within the skull.

Other treatment
Losing weight if you are overweight may help to improve symptoms in some people. For example, research suggests that a 6% decrease in body weight can help to resolve swelling at the back of the eye (papilloedema). However, many people find that weight loss does not help much.

What is the outlook (prognosis)?
It is essential to detect idiopathic intracranial hypertension (IIH) early and to start treatment early to prevent permanent loss of vision occurring.

In some people, IIH can get better by itself but recurrence (relapse) of symptoms is common. For many other people, a combination of medical and surgical treatment can help to control their symptoms well. However, some people can still have troublesome symptoms despite treatment.

Further help & information

**IIH (Idiopathic Intracranial Hypertension) UK**
Broadsands Drive, Gosport, Hants, PO12 2SD
Web: www.iih.org.uk

Further reading & references


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