Patient

View this article online at: patient.info/brain-nerves/idiopathic-intracranial-hypertension-leaflet

Idiopathic intracranial hypertension

In idiopathic intracranial hypertension 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.

What is idiopathic intracranial hypertension?

Intracranial hypertension means raised pressure inside the skull and around the brain. Idiopathic intracranial hypertension (IIH) is a condition where raised pressure inside the skull occurs without an identifiable cause. Idiopathic means that the cause of this raised pressure is unknown.

There are many other causes of raised intracranial pressure. When there is a clear cause, such as a brain tumour, it's sometimes called secondary intracranial hypertension. However, in IIH, there is no clear cause.

The brain is surrounded by a fluid called cerebrospinal fluid (CSF). It's thought that, in IIH, there is a problem where too much CSF is produced, not enough CSF is re-absorbed, or both. We don't know exactly why this happens in IIH, although there are several theories.

Having too much CSF around the brain causes high pressure inside the skull. This raised pressure leads to the symptoms of idiopathic intracranial hypertension. 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 Swelling of the optic nerve is called papilloedema. If papilloedema is not recognised and treated, it can cause damage to the optic nerve. This can lead to vision problems, including blindness.

Idiopathic intracranial hypertension used to be called benign intracranial hypertension. However, this name is not being used as much now. This is because IIH 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.

How common is idiopathic intracranial hypertension?

Idiopathic intracranial hypertension 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.

Idiopathic intracranial hypertension occurs more frequently in some groups of people. Possible risk factors include:

- Gender. Amongst adults, more than 9 out of 10 people with idiopathic intracranial hypertension are women. It's thought that female hormones may play a role in the condition.
- Overweight and obesity. This is very common amongst people with idiopathic intracranial hypertension. Hormonal changes from obesity might be linked to the condition.
- Rarely, certain medications: stopping steroids can trigger idiopathic intracranial hypertension, and other medications such as tetracycline antibiotics, lithium, and vitamin A have all be linked with it.
- Obstructive sleep apnoea.

Idiopathic intracranial hypertension symptoms

There may be many different symptoms associated with idiopathic intracranial hypertension (see Further Reading and References, below). However, the typical symptoms include:

- Headache.
- Tinnitus.
- Changes in vision.

The most prominent symptom of idiopathic intracranial hypertension is headache. This can be severe and is a long-term (chronic) headache. It can vary in its location and may come and go. Often - but not always - the headache is worse in the morning, worse on lying flat, and worse when coughing, sneezing, or straining (all of these things can increase the pressure inside the skull slightly).

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 long does idiopathic intracranial hypertension last?

This differs from person to person, but idiopathic intracranial hypertension often takes several months to improve with treatment. In some people, it goes away completely after months or years. In other people, it may go away but return again in future. For some people, it is a lifelong condition.

How is idiopathic intracranial hypertension diagnosed?

If you visit your doctor, complaining of headaches and/or sight (visual) symptoms, your doctor will usually discuss your symptoms with you, and examine the nerves in your arms, legs, and face.

Ophthalmoscopy

They may examine the back of the eyes with a hand-held instrument for looking in the back of the eye (an ophthalmoscope). This can also be done at an optician, who will often take photos of the back of the eye. This may show swelling at the back of the eye (papilloedema). However, not everyone with idiopathic intracranial hypertension has papilloedema.

Papilloedema is a sign of raised pressure within the skull (raised intracranial pressure). Therefore, the main thing when diagnosing idiopathic intracranial hypertension 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.

Eye examination

You 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 idiopathic intracranial hypertension.

Brain scans

A brain scan is likely to be performed to rule out other causes of raised intracranial pressure, such as a brain tumour. In idiopathic intracranial hypertension, brain scans are usually normal, although sometimes there are subtle changes of high pressure that can be seen.

This is likely to be an MRI scan, but sometimes a CT scan is used instead.

Lumbar puncture

This is one of the main tests to confirm high pressure inside the brain. CSF is found around the brain, but also around the spinal cord. A lumbar puncture (sometimes called a spinal tap) involves putting a needle into the lower part of the spine to sample some of the fluid around the spine. In idiopathic intracranial hypertension, measurements will show that the CSF is under raised pressure. A sample of CSF can also be taken and analysed to look for any other causes of raised pressure.

See the separate leaflet called Lumbar Puncture (Spinal Tap) for more details.

What are the aims of idiopathic intracranial hypertension treatment?

There are two main aims of treatment for idiopathic intracranial hypertension:

- To improve, and hopefully cure, the headaches.
- To prevent visual loss.

If you are diagnosed with idiopathic intracranial hypertension, 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 may mean that your treatment needs to be adjusted.

Idiopathic intracranial hypertension treatment

Weight loss

For people with overweight or obesity, weight loss can be a very effective treatment for idiopathic intracranial hypertension. Research shows that losing 5-10% of body weight can significantly improve symptoms of IIH, and sometimes causes the condition to go into remission.

Medical treatment

The most commonly-used medication for idiopathic intracranial hypertension is acetazolamide. Acetazolamide reduces CSF production. Acetazolamide is a safe medication, but commonly causes tingling of the fingers and toes as a side-effect. It may also cause birth defects in pregnancy; speak to your doctor if you are considering or planning a pregnancy.

Other medicines that are used less commonly include topiramate and furosemide.

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 idiopathic intracranial hypertension. 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.

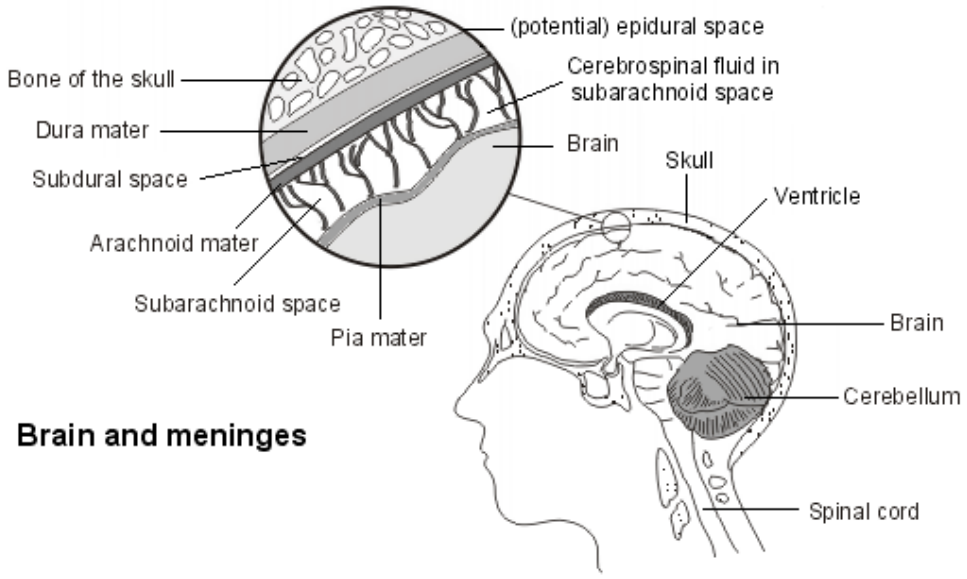
Some anatomy around the brain

The meninges form the protective lining that surrounds the brain and 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.



Brain and meninges

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 the outlook (prognosis)?

It is essential to detect idiopathic intracranial hypertension early and to start treatment early to prevent permanent loss of vision occurring.

In some people, idiopathic intracranial hypertension 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 reading

- Piper RJ, Kalyvas AV, Young AM, et al; Interventions for idiopathic intracranial hypertension. Cochrane Database Syst Rev. 2015 Aug 7;8:CD003434. doi: 10.1002/14651858.CD003434.pub3.
- Wall M; Update on Idiopathic Intracranial Hypertension. Neurol Clin. 2017 Feb;35(1):45-57. doi: 10.1016/j.ncl.2016.08.004.
- Mitchell JL, Mollan SP, Vijay V, et al; Novel advances in monitoring and therapeutic approaches in idiopathic intracranial hypertension. Curr Opin Neurol. 2019 Jun;32(3):422-431. doi: 10.1097/WCO.0000000000000690.
- Kalyvas A, Neromyliotis E, Koutsarnakis C, et al; A systematic review of surgical treatments of idiopathic intracranial hypertension (IIH). Neurosurg Rev. 2020 Apr 25. pii: 10.1007/s10143-020-01288-1. doi: 10.1007/s10143-020-01288-1.

Authored by:	Peer Reviewed by: Dr Colin Tidy, MRCGP	
Originally Published: 19/11/2023	Next review date: 19/09/2023	Document ID: doc_12396