



What is IIH?

Idiopathic Intracranial Hypertension (IIH)

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Idiopathic intracranial hypertension (IIH), also known as benign intracranial hypertension or pseudotumour cerebri, is a condition with an unknown cause or causes. The condition is associated with raised fluid pressure around the brain. The fluid that cushions the brain is called cerebrospinal fluid (CSF).

It can cause disabling daily headaches and visual loss, which can be permanent. The raised brain pressure can press the nerves supplying the eye (also known as papilloedema) and this can affect vision.

This leaflet will help you to understand what IIH is.

Who gets IIH?

IIH can happen to anyone. It is a condition found more commonly in women (90%), but some men are also affected by it (10%). It is common in teenagers and young women, but can also affect children and adults of any age.

How common is IIH?

IIH is considered a rare disease. Recent medical reports show that IIH is happening more often. Somewhere between 1-3 people in every 100,000 get

this condition in the normal population, but it becomes more common in those with obesity with rates reported up to 20 per 100,000. Overall, it is thought that IIH is becoming more common.

What causes IIH?

We do not know what the actual cause or causes of IIH are. There is a striking association to being overweight. This is a sensitive issue. Medical studies have shown that recent weight gain can cause IIH, and that weight loss can put IIH into remission. There is ongoing research into the cause/causes of IIH.

Is IIH genetic?

The cause or causes of IIH are not known. Although genes play an important role in lots of conditions, it is not yet known whether they play a large role in IIH.

How is IIH diagnosed?

Doctors need to talk to you about what you have been experiencing and perform a physical examination. It is important that other conditions are ruled out before diagnosing IIH.

It is essential that other conditions such as venous sinus thrombosis (blood clot in brain), anaemia (lack of red blood cells) and certain drugs such as some antibiotics or vitamin A containing drugs are ruled out, as they require different treatment.

To be diagnosed with IIH you will need brain scans and a lumbar puncture (LP), sometimes called a spinal tap. It is vital that lumbar puncture reading is performed with you relaxed and lying on your side for the reading to be accurate. For more information, see the IIHUK Information on Lumbar Puncture Booklet.

For doctors to be able to diagnose IIH all the following 5 things need to be present:

1. Papilloedema (swelling of the eye nerves).
2. Normal neurological examination (sixth nerve palsy causing double vision is allowed).
3. Normal brain imaging. This is usually with computerized tomography (CT) or magnetic resonance imaging (MRI) scans. They should also include a scan of the veins of the brain to exclude venous sinus thrombosis.
4. Normal brain fluid (CSF) analysis.
5. Elevated lumbar puncture opening pressure above 25cm (for some people a pressure above 25cm may be normal for them)

(For the criteria for IIHWOP, see the IIHWOP leaflet)

What do people with IIH experience?

IIH affects people differently. Not everyone has all the symptoms that people report when they have IIH. The symptoms of IIH can have a significant impact on the quality of your life.

The commonest symptoms of IIH include:

- Headaches
- Visual obscurations
- Pulsatile tinnitus
- Back pain
- Dizziness
- Neck pain

Less common symptoms that are sometimes reported include:

- Blurred vision
- Memory problems
- Nerve pain
- Double vision

Headaches in IIH

Headache is the most common symptom in patients with Idiopathic Intracranial Hypertension (IIH). Although not everybody with IIH gets headache.

The headache may happen every day or less often. Some IIH headaches improve after lumbar puncture (but migraine headaches can also improve after lumbar puncture). It may be worse in the morning, on bending and on coughing (but other headaches can have these too). The exact feeling of these headaches is not well described, and vary considerably between people.

If you have IIH you can get a number of different types of headaches (for example migraine or medication overuse). In the context of IIH your health care professional will consider with you what types of headaches you have and how best to treatment them. For further information on headaches see the IIHUK Headache in IIH leaflet.

Vision problems in IIH

If your vision is getting worse, the doctors may recommend you need emergency treatment to save your vision. This is usually a surgery to release the brain pressure.

There are two main types of surgery:

1. Shunt surgery – one end of a flexible tube is put into the fluid filled space in your brain (ventricle) or your spine (lumbar) and the other end in another part of your body, such as your abdominal cavity (peritoneum).
2. Optic nerve sheath fenestration – a small window is created in the layer (sheath) around your eye nerve to allow the fluid to drain away.

For further information on surgery in IIH see the IIHUK shunt surgery in IIH and optic nerve sheath fenestration leaflets.

What are visual obscurations?

This is a short lived loss of vision, that returned to normal within seconds. The vision may “grey-out” or become dark. It can happen in one or both eyes. It can be triggered by bending down.

What is pulsatile tinnitus?

Tinnitus is a term for hearing sounds that come from inside your body, rather than from outside. Pulsatile tinnitus is the noise of your heart beat or pulse in one or both of your ears. For some they only hear this when things are quiet (for example in bed at night). For others it can be a disabling distracting noise they hear all the time which can affect their concentration, and can cause difficulty sleeping.

There is no proven treatment for pulsatile tinnitus. For the majority as their IIH settles the pulsatile tinnitus disappears.

How is IIH treated?

The majority of people will have medical (drugs) treatment for IIH and headaches. Less than 1 out of 10 people with IIH will have surgical treatment.

9 out of 10 people with IIH are overweight and weight loss is the most effective treatment. For further information on weight loss see the IIHUK Weight and IIH leaflet.

Medical treatments for IIH have been assessed by a medical review body (the Cochrane review) and they have suggested that more trials are required to understand what are the best drug and surgical treatments for IIH. Acetazolamide (DiamoxTM) is often prescribed for IIH, for further information on acetazolamide see the IIHUK acetazolamide drug information leaflet.

How do you treat headaches caused by IIH?

There are no drugs specially designed for IIH headaches. There is evidence that weight loss improves headaches in IIH (Newborg 1974; Koppersmith 1998; Johnson 1998; Sinclair 2010).

Your health care professional may use medications to help with the headaches. For further information on headaches see the IIHUK Headache in IIH leaflet.

Are there any warning signs of IIH getting worse?

Increase in the frequency and severity of headaches, increase in visual obscurations and pulsatile tinnitus can all be signs that IIH is getting worse. This can be frightening. If this is happening it is important to see your doctor.

What is the long-term outlook?

In some people, after diagnosis, IIH can settle itself. For the majority weight loss and combined with medical treatment, will control the symptoms well. However, some people may continue to have disabling symptoms despite treatment.

Can I get pregnant if I have IIH?

- If you are considering becoming pregnant, tell your doctor as some of the medicines used in IIH and headache can potentially harm the unborn baby.

I take the pill (oral contraceptive), what do I need to know?

- There are many different types of pills for contraception. They have different amounts of hormones in them. More often they have no relationship to the development of IIH. If you are worried, speak to your doctor.
- Some drugs used to control headaches in IIH can affect how well the oral contraceptive pill works, so you could be at risk of becoming pregnant. Tell your doctor if you take oral contraceptives.

I have a contraceptive injection or the contraceptive implant, what do I need to know?

- These types of contraceptives use the hormone progestogen. There is no clear relationship between using these types of contraceptives to the development of IIH. If you are worried, speak to your doctor.

What is IIH without papilloedema?

IIH without papilloedema, is a rare condition where high pressure can trigger headaches but in this condition at diagnosis there is no papilloedema and these people never develop papilloedema. For further information see the IIHUK IIHWOP leaflet.


Where can I get more information?

IIH UK website

www.iih.org.uk

IIH Consensus Guidelines

<http://jnnp.bmj.com>



I want to
know more
about IIH....

Write notes or questions for your appointment here:

A team of people contributed to this booklet. It was written by S. Mollan. Critically reviewed by S. Mollan and A. Sinclair. It was assessed in the draft stage by the ophthalmology nursing team at University Hospitals Birmingham (UHB). It was reviewed by a group of patients who have IIH, and also assessed by friends and family that attended the Joint Idiopathic Intracranial Hypertension clinic at UHB. It was critically reviewed by the IIHUK trustees. S Mollan is responsible for the final version. The views expressed in this booklet are of the authors and not their employers or other organisations.

Please note we have made every effort to ensure the content of this is correct at time of publication, but remember that information about the condition and drugs may change. This information booklet is for general education only.

Version 4.0 (13 June 2018). Review by June 2020.



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Registered Charity Number - England &
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