



Idiopathic Intracranial Hypertension (IIH)

What is Idiopathic Intracranial Hypertension?

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 increased 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 booklet will help you 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. 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 ongoing research into this. A sensitive issue is the marked association to being overweight. Medical studies have shown that recent weight gain can cause IIH and that weight loss can put IIH into remission. Regaining weight also causes the disease to relapse.

Is IIH genetic?

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 (the 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

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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 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. Brain imaging that shows no other cause. 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)

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:

1. Headaches
2. Visual obscurations (greying and blacking out of the vision in either eye that lasts seconds)
3. Pulsatile tinnitus (heart beat sound in one or both ears)
4. Back pain
5. Dizziness
6. Neck pain

Less common symptoms that are sometimes reported include:

1. Blurred vision
2. Memory problems
3. Nerve pain
4. 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 (although 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 features 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 treat them.

Vision problems in IIH

If your vision is getting worse, the doctors may recommend an 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)

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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. This is only done in few hospitals in the United Kingdom

How is IIH treated?

Management is a sensitive issue as 9 out of 10 people with IIH are overweight and weight loss is the most effective treatment. The majority of people will have weight loss advice and medical (drugs) treatment for IIH and headaches. Less than 1 out of 10 people with IIH will have surgical treatment.

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 and identify the best drug and surgical treatments for IIH. Acetazolamide (DiamoxTM) is often prescribed for IIH.

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. Your health care professional may use medications to help with the headaches.

Are there any warning signs of IIH getting worse?

Increase in the frequency and severity of headaches, and 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 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 (IIHWOP) is a rare condition where high pressure can trigger headaches but in this condition at diagnosis, there is no papilloedema. These people never develop papilloedema.

Where can I get more information?

www.IIH.org.uk

<http://jnnp.bmj.com>

Write notes or questions for your appointment here:

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For full details see the information leaflet that comes with the medicine.
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