



Idiopathic Intracranial Hypertension without Papilloedema (IIHWOP)

What is Idiopathic Intracranial Hypertension without Papilloedema (IIHWOP)?

IIHWOP is a very rare type of Idiopathic Intracranial Hypertension (IIH).

IIH has been known by other names such as Benign Intracranial Hypertension or pseudotumour cerebri. It 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).

When the brain pressure is high, the majority of people will have eye (optic) nerve swelling called papilloedema, but in IIHWOP there is no papilloedema.

What is difference between IIH and IIHWOP?

People with IIH have swelling of their eye (optic) nerves at diagnosis, called papilloedema. Those with IIHWOP never have eye (optic) nerve swelling.

As IIHWOP is much rarer than IIH, less is known about it and there are no large studies to help guide treatment specifically for IIHWOP.

This booklet will help you understand IIHWOP.

Who gets IIHWOP?

IIHWOP can happen to anyone. It is a condition found more commonly in women (90%) than in men (10%). It is common in young women but can also affect children and adults generally below the age of 50 years.

How common is IIHWOP?

It is not known how common IIHWOP is. In large hospitals that look after IIH, IIHWOP occurs in less than 6% of all their IIH patients. It is considered very rare.

What causes IIHWOP?

We do not know what the actual cause or causes of IIHWOP are. There is a striking association with being overweight which is a sensitive issue.

Is IIHWOP genetic?

The cause or causes of IIHWOP 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 IIHWOP.

How is IIHWOP 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 IIHWOP. It is essential that other conditions such as venous sinus thrombosis (the blood clot in brain),

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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 IIHWOP, 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.

It is important that the doctors look carefully for small signs of papilloedema (eye nerve swelling) to ensure they do not miss IIH.

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

1. Normal neurological examination (sixth nerve palsy causing double vision is allowed)
2. Brain imaging showing no other cause of raised brain pressure. 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
3. Normal brain fluid (CSF) analysis
4. Elevated lumbar puncture opening pressure above 25cm (note that for some people, a pressure above 25cm may be normal)
5. Sixth nerve palsy on one or both sides

Possible IIHWOP can be made if 1–4 above are present, without a sixth nerve palsy as long as there are 3 features of raised brain pressure seen on brain scanning such as an empty sella; flattening of the eye globe; widening of the space around the eye nerve and/or transverse venous sinus.

What is the difference between IIHWOP and IIH in ocular remission?

When the raised intracranial pressure settles in IIH, the papilloedema settles. This is called IIH in ocular remission. Although they are in remission from their IIH, often those with IIH will have ongoing headaches.

This should not be confused with IIHWOP, where in IIHWOP they will never have had papilloedema.

What do people with IIHWOP experience?

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

The commonest symptoms of IIHWOP include:

- Headaches
- Pulsatile tinnitus (heart beat sound in one or both ears)
- Visual obscurations (greying and blacking out of the vision in either eye that lasts seconds)
- Blurred vision
- Photopsias (sparkles or lights in vision)
- Double vision

Headaches in IIHWOP

Headache is the most common symptom in patients with IIHWOP.

The headache may happen every day or less often. They can happen on one side of the head or both sides. Some IIHWOP 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 too). The exact feeling of these headaches is not well described and vary considerably between people.

If you have IIHWOP, you can get a number of different types of headaches (such as raised pressure headache, migraine and medication overuse headache). In the context of IIHWOP your health care professional will consider with you what types of headaches you have and how best to treat them.

For further information on headaches see the IIHUK Headache in IIH booklet.

Pulsatile tinnitus in IIHWOP

Less than 50% of patients with IIHWOP get 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 IIHWOP settles, the pulsatile tinnitus disappears.

Vision problems in IIHWOP

In IIHWOP there is no swelling of the eye (optic) nerves (no papilloedema). Therefore, doctors are not worried that you will have or will develop a permanent problem with your vision. Also, no emergency treatment would be required to protect the vision in IIHWOP.

The most common issues with the vision in IIHWOP are blurred vision, photopsias, and double vision.

How is IIHWOP treated?

The majority of people will have medical (drugs) treatment for IIHWOP and headaches. There are no large studies to help guide treatment in IIHWOP.

9 out of 10 people with IIH are overweight and weight loss may be the most effective treatment. Acetazolamide (DiamoxTM) tablets may be prescribed for IIHWOP.

Surgical treatment, such as shunt surgery, has been used to treat raised brain pressure in some cases, but there is little evidence in this area and long-term headache outcomes for shunting is not good.

How do you treat headaches caused by IIHWOP?

There are no drugs specially designed for IIHWOP headaches. There is evidence that weight loss improves headaches in IIH and this has been reported in IIHWOP cases too.

It is essential to withdraw medications that may contribute to medication overuse headache. Your health care professional may use medications to help with the headaches. These can include drugs that are mainly used for migraine.

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Are there any warning signs of IIHWOP getting worse?

Increase in the frequency and severity of headaches and increase in pulsatile tinnitus may be signs that IIHWOP is getting worse. This can be frightening. If this is happening it is important to inform your doctor.

What is the long-term outlook?

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 IIHWOP?

If you are considering becoming pregnant, tell your doctor as some of the medicines used in IIHWOP 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 IIHWOP. If you are worried, speak to your doctor.

Some drugs used to control headaches in IIHWOP 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 IIHWOP. If you are worried, speak to your doctor.

Where can I get more information?

IIH UK website

www.IIH.org.uk

Write notes or questions for your appointment here:

A team of people contributed to this booklet. It was written by S. Mollan. 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 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.

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

Version 1.0 (1st June 2021) Review by June 2023.

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