

Idiopathic Intracranial Hypertension *without* Papilloedema (IIHWOP)

What is Idiopathic Intracranial Hypertension *without* Papilloedema?

Idiopathic intracranial hypertension (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 Idiopathic Intracranial Hypertension *without* Papilloedema and Idiopathic Intracranial Hypertension?

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 Idiopathic Intracranial Hypertension (IIH) less is known about it, and there are no large studies to help guide treatment specifically for IIHWOP. This leaflet will help you to understand IIHWOP.

Who gets IIHWOP?

IIHWOP 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 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. This 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 (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 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. For more information, see the IIHUK Information on Lumbar Puncture Leaflet.

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. 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.
3. Normal brain fluid (CSF) analysis.
4. Elevated lumbar puncture opening pressure above 25cm (note for some people a pressure above 25cm may be normal for them)
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 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, termed 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
- Visual obscurations
- Blurred vision
- Photopsias
- Double vision

Headaches in IIHWOP

Headache is the most common symptom in patients with Idiopathic Intracranial Hypertension without papilloedema.

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 (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 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 leaflet.

Pulsatile tinnitus in IIHWOP

Just under a half 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 termed 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 commonest issues with the vision in IIHWOP are photopsias, blurred vision, and double vision.

In IIHWOP people report blurred vision and when tested in the eye clinic often the field of vision test may not be normal. This is not thought to be a permanent problem.

What are photopsias?

This is an experience of flashes of light, sometimes coloured or white, in the vision. They are very short lived or brief. They do not cause any permanent problem with the vision. Another word to describe this is called phosphenes.

Photopsias can occur in many other condition as well as IIHWOP. There is no treatment for photopsias. Often they settle when the IIHWOP settles into remission.

What is double vision?

Double vision (termed diplopia) is often hard for people to describe. It is seeing two images of one image (scene or object) overlapped. This can happen some or all the time.

Sixth nerve palsy typically happens in 1 in 4 people with IIHWOP and is the cause of the double vision. Double vision settles when the IIHWOP settles into remission.

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. For further information on weight loss see the IIHUK Weight and IIH leaflet.

Acetazolamide (Diamox™) tablets may be prescribed for IIHWOP, for further information on acetazolamide see the IIHUK acetazolamide drug information leaflet.

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. For further information see the IIHUK shunt surgery leaflet.

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 (Newborg 1974; Koppersmith 1998; Johnson 1998; Sinclair 2010) 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. For further information on headaches see the IIHUK Headache in IIH leaflet.

Are there any warning signs of IHWOP getting worse?

Increase in the frequency and severity of headaches, and increase in pulsatile tinnitus, may be signs that IHWOP 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 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 IHWOP?

- If you are considering becoming pregnant, tell your doctor as some of the medicines used in IHWOP 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 IHWOP. 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



I want
to know
more

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 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 (Reviewed August 2021) review by July 2023.



info@iih.org.uk

Registered Charity Number – England
& Wales 1143522, Scotland SC043294