Just diagnosed - What is IIH?
An Introduction to Idiopathic Intracranial Hypertension
Introduction

If you have been given this booklet you will have just been diagnosed with Idiopathic Intracranial Hypertension (IIH). Or you could be the partner, relative or friend of someone who has just received this diagnosis.

As someone who has just been diagnosed, you may be experiencing relief from finally getting a diagnosis, especially if it has taken quite some time to get to this point. It is quite natural if you feel overwhelmed at this point and have lots of questions that you thought you should have asked at your appointment.

If you are a partner, relative or friend this will help you to understand what IIH is and the effects it has on someone who has it.

This booklet will answer some, if not all, of the questions you have about what IIH is and what it means now that you have it.

What is Idiopathic Intracranial Hypertension?

Your brain is protected by fluid called cerebrospinal fluid (CSF), and it is produced in the Choroid Plexus, which is found in the ventricles of the brain. CSF is also located in the subarachnoid space around the brain and spinal cord.

The Choroid Plexus has several important roles, one of them being to generate around one pint of CSF every day. CSF washes over the brain and spinal cord until it is finally absorbed into what is known as the venous blood system. There is a continuing cycle of CSF production, and when this functions normally, the volume of the CSF and pressure of the fluid stays at a consistent level. For adults this pressure is usually below 200 mm H₂O.

In the case of IIH, the body is unable to competently absorb or drain the CSF. When this takes place, the intracranial pressure increases, which then causes the debilitating, painful headaches and other symptoms. The average adult has replenished CSF 3-4 times a day. When this is either interrupted or too much is produced, the painful headaches and symptoms are apparent. Pressure higher than 250 mm H₂O or above is a marker for IIH.
If you look at how the brain is contained within the skull, there is very little space between the brain and the skull, and this means CSF, which is under high pressure, presses on significant brain structures. This then restricts the flow of blood into the brain. There is no room for the additional CSF to go and so it compresses the brain.

Chronic daily headache becomes a dominant factor caused by raised intracranial pressure. The optic nerves become swollen. This causes visual obscurations such as blurring, double vision, blind spots and the visual fields to decrease. Your visual fields are what enable you to see above, below, left and right, while looking in a forward direction.
Are Pseudotumour Cerebri and Benign Intracranial Hypertension the same as IIH?

Pseudotumour Cerebri (PTC) and Benign Intracranial Hypertension (BIH) are the names that IIH were previously known as, but are still used by some doctors. It is now called Idiopathic Intracranial Hypertension. Idiopathic means no known cause.

Who gets IIH?

Anybody can get IIH, it doesn’t matter what gender, age, body type, race and ethnic background. You can develop it at any time in your life, and you won’t know you have it until you are diagnosed after having several tests, the main one being a Lumbar Puncture. Unlike some conditions you can’t be screened for IIH, so until you start exhibiting its symptoms you wouldn’t know you have it. It affects 1-2 people in every 100,000, which makes it a rare disease.

At this point no one knows why an individual has IIH, although it is known that genes do factor in a lot of conditions and diseases, it has neither been ruled in nor out, as a factor in IIH. One of the characteristics better known about this condition is that it affects overweight women of childbearing age. Although rare, men can also have this condition.

There is evidence to show that weight loss can be beneficial to some patients with this condition who are overweight. This can reduce the optic nerve swelling, intracranial pressure and headaches, which can lead to remission. For some people, a loss of about 6% of their total body weight can make a difference and improvement in symptoms. Should weight fluctuate or increase again then there is a risk of remission ending, and recurrence of the symptoms of IIH.

Is there a cure?

There is no cure for IIH but there are treatments that can help send the condition into remission. Remission can be permanent or temporary depending on the individual and their medical history. As with any medical condition research is ongoing.
The Symptoms
Headache is the main common symptom of IIH and can be mistaken for chronic headache and migraine. The characteristics of an IIH headache are the unbearable pain which isn’t relieved by lying down and is worse in the morning. However, some patients have no headache and still have other symptoms indicative of IIH.

Different to a chronic headache or a migraine, an IIH headache is accompanied by a noise often referred to as a “whooshing” sound, and the reason it happens, is because high intracranial pressure causes the compression of blood vessels. It can accompany the headaches, or just present on its own and is a form of tinnitus, called pulsatile tinnitus.

Another common symptom experienced is visual obscurations, which consist of blurring, grey spots sometimes described as cobwebs or wisps, and blobs in one or both eyes. There can also be episodes of dimming vision, photophobia (light sensitivity) and double vision. These visual changes are caused by swollen Optic nerves, also called Papilloedema. This can be what is termed “bilateral”, meaning both optic nerves, and “unilateral”, meaning one optic nerve. There is evidence to show that Papilledema doesn’t have to be present and so IIH shouldn’t be ruled out when other symptoms are present.

One of the essential reasons for monitoring and treating IIH is to prevent deterioration in vision. A very small minority of patients (1-2%) go on to lose their sight, so following your prescribed treatment is essential, to ensure you are doing all you can to preserve your sight, or to resolve any early temporary vision loss you have already experienced.

Other symptoms that can also be experienced are:

- Nausea & vomiting
- Neck stiffness
- Dizziness
- Depression
- Lethargy
- Memory issues
- Exercise intolerance
- Heat intolerance
- Pain in arms, legs & back
- Tingling in hands & feet
- Spinal fluid leaks from nose (Cerebral Fluid Rhinorrhea - this is rare)
As with any illness it can affect people differently, some people find that it disappears spontaneously or once they have had a lumbar puncture, but others can have relapses and continue with this condition into the chronic stage. There are several medications that successfully treat this condition by controlling cerebral spinal fluid levels, and they can lead to a successful remission.

Only a small number of people with IIH will need a surgical intervention, and this is only done as a last resort, and not before all other treatments are explored. Your doctor will go through all the best treatment options suitable and discuss them with you.

Self Awareness
The longer you have IIH the more aware of your symptoms and triggers you’ll become. Keeping a symptom log will help you to recognise the symptoms and when they appear. Being informed can help you to manage your IIH and its effects and cope better with this condition.

Your GP
Your GP may not have heard of or treated IIH before, and because of this may not be able to understand your symptoms. You might not see the same GP everytime you have an appointment. Always be clear and precise when giving them information about your IIH if you are seeing them because of headache, how it is treated and by whom.

Keeping a record can be very helpful in assisting your doctor to have a clear picture of your symptoms. This may include information on:

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<thead>
<tr>
<th>Headache - when did it start?</th>
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<tr>
<td>What kind of pain you are experiencing?</td>
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<td>Where is the pain?</td>
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<tr>
<td>Vision - what changes have you noticed?</td>
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<tr>
<td>Hearing – what problems are you having?</td>
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<td>Are there any other symptoms - nausea/tiredness?</td>
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Types of Intracranial Hypertension

There are different types of Intracranial Hypertension.
  - Chronic IIH
  - Severe IIH
  - Secondary Intracranial Hypertension (SIH)

Chronic IIH
Most patients who have Chronic IIH have their symptoms managed by medication to control cerebral spinal fluid (CSF) levels, and also have occasional lumbar punctures if necessary. Losing weight can help lead into remission, if weight is a factor. There is no particular known cause.

Severe IIH
When IIH becomes severe, the symptoms have become harder to manage by medication and lumbar puncture. Vision will be decreasing rapidly and a surgical intervention will be necessary to control the CSF levels, reduce optic swelling and prevent visual loss.

Secondary IH
This form of intracranial hypertension is usually caused by a known cause. Some medications are known to cause SIH such as a group of antibiotics called Tetracyclines, the Oral contraceptive pill, Vitamin A and Progesterone. There are also other diseases known as Systemic diseases, which are diseases that affect a number of organs, tissues or the whole body.
Coping with your diagnosis

There is no rule to how you cope with your diagnosis. Everybody reacts differently and you may feel quite shocked, scared and numb which is a completely natural reaction. You may not be able to take in what your doctor is telling you, and wonder how you are going to tell your friends and family. It is important to remember that IIH is not a life threatening disease only a life changing one. Take time to digest what you have been told and to accept your diagnosis. Some people are in denial when first told they have Idiopathic Intracranial Hypertension.

The I Have IIH Foundation has a website where you can connect to other people who have this condition, as well as providing information, support and practical help. Some people who have IIH, describe it as “losing the person they were” but have found that by being able to connect to others with this condition, that it has helped them to cope a lot better and helped them to feel less isolated.

Telling people you have IIH

Once you have emotionally and mentally dealt with your diagnosis, you may want to inform the people closest to you. The best way to do this is to make sure you have all the information about IIH, and are prepared for any questions they may ask you. It is possible that they may still not understand your condition and provide the support and understanding that you expected. This is usually because they haven’t heard of IIH before, and as it is an invisible condition, may not have realised you are ill. People with IIH are often told they “don’t look sick”.

Do I need to tell anyone else?

You may need to tell your employer, if you have to have time off for appointments or a lumbar puncture. You may also need to tell them about any problems you are having especially, if you have visual problems, so that adjustments can be made for you. Having IIH doesn’t mean you have to give up your job. The I Have IIH Foundation website has advice about working with IIH and on what information you need to provide if necessary.

You may have to disclose your condition for life insurance purposes or to Mortgage companies for instance as an existing condition if they ask about any health conditions on their forms.
Living with IIH

It is understandable if you are feeling quite worried about finding out you have a condition that can affect your vision, requires daily medication and regular monitoring. Some people with IIH feel like their life is over, but it is possible to live and work with IIH. In most cases people with IIH lead a relatively normal life and continue as they did before, with the occasional flare ups of raised pressure. Again everyone is different, and their progress and recovery are different.

It is important that you still retain a sense of normality and social life as much as you can. Sometimes it may not be possible but it’s not impossible. It will be quite normal to worry about the effects IIH will have on your relationships, work life and home life. With any illness, adjustments have to be made in order to be able to cope with the condition. Life doesn’t stop with IIH.

Pregnancy

IIH can start anytime during pregnancy. If intracranial pressure is raised during pregnancy it can be treated. You may feel well through your pregnancy or you may have a flare up of raised intracranial pressure. As soon as you inform your doctor you are pregnant, they will be able to discuss with you how your IIH will be treated during your pregnancy should you experience raised pressure. As weight can increase risk of raised pressure you may be advised to watch your calorie intake while pregnant.

Can I fly with IIH?

There is no rule about not flying with IIH but some people may experience a high altitude headache. Make sure you take an ample supply of your prescribed medication with you in your bag, and a letter from your doctor to explain why you have the medication to avoid problems at customs for instance. It would be wise to inform your travel insurer too.
Can I drive?
There is no reason why you can’t drive unless you are having visual problems. If you are experiencing visual problems you will need to inform the DVLA. As with all medication you should be cautious when starting new medication or adjusting dosage, in case you are affected by drowsiness, in which case you should not drive.

What do I do if I have a high pressure headache?
If you experience a high pressure headache which feels worse in the morning and doesn’t ease with lying down, contact your neurologist via their secretary or the clinic you attend for your IIH appointments. You can ask your doctor about this when you see them in clinic.

Other headaches that can feel like a raised pressure headache
There are other headaches you may experience that can sometimes be confused with a raised pressure headache. By keeping a record of your symptoms you may be able to recognise other types and when they usually occur. This will help you determine whether or not you have a high pressure headache or if it is caused by something else.

Analgesic Rebound
When you take medication on a daily or near daily basis, you will find that analgesics can keep up the headache process. They may decrease the intensity of the pain for a few hours, but then, they appear to feed into the pain system in such a way that chronic headaches may result. Many times you could mistake this for a high pressure headache, when in fact it could be merely a rebound headache. Symptoms are a dull, tension-type headache or may be a more severe migraine-like headache and will continue unless you stop taking the analgesic, or it will become more severe and also include nausea and vomiting.
IIH medical specialists – the people you may see

**Neurologist** – A specialised doctor who diagnoses and treats patients with neurological disorders.

**Neuro-Ophthalmologist** – A specialised doctor in both neurology and ophthalmology, who specialise in eye conditions that start from the nervous system.

**Ophthalmologist** – A specialised doctor in the diagnosis and treatment of eye diseases.

**Radiology Staff** – These are the people who will perform and interpret your scans.

**Neurophysiologist** – A specialised doctor who tests electrical functions in the brain, spinal cord and nerves in limbs and muscles.
The I Have IIH Foundation

The I Have IIH Foundation is registered charity in England and Wales, no. 1155833.

We provide online support, information and practical help and advice. We support patients with IIH and their families, we also work with many other charitable organisations that can be found on our website, who provide valuable resources and information. We are also members of the RARE Foundation Alliance.

You can find us:
www.ihaveiih.com  I Have IIH Foundation  @IHaveIIH

Other booklets available in this series

Diagnostic Tests for IIH
Living with IIH – Carer’s Booklet
Medical Treatment and Monitoring in IIH
Surgical Treatments in Severe IIH

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