East Kent Hospitals University NHS Foundation Trust

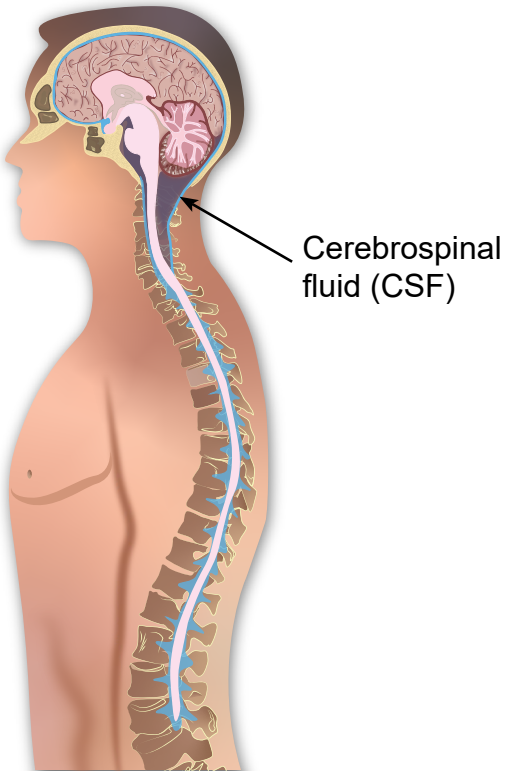
Idiopathic intracranial hypertension (IIH)

Information for patients

This leaflet is for patients already diagnosed with and also those being investigated for idiopathic intracranial hypertension (IIH). The leaflet will explain what IIH is, how it is diagnosed, and the best ways of managing the condition.

What is idiopathic intracranial hypertension (IIH), and how common is it?

- IIH is a neurological condition caused by an increase in pressure in the fluid that surrounds the brain. This fluid is called cerebrospinal fluid (CSF) and bathes the brain, some of your nerves, and your spinal cord.
- When the pressure gets too high it can affect the nerves which supply your eyes and can cause headaches.
- IIH affects approximately two to three people in every 100,000.
- The number of cases diagnosed each year is increasing.



We care

What causes IIH?

The medical term 'idiopathic' means that the exact cause is not clear. However, certain things are known to increase your risk of developing the condition, including the following.

- **Weight gain** is often the trigger in susceptible people, especially those who are already overweight or obese.
- **Obesity (where your BMI is greater than 30)** has been shown to be the biggest cause of IIH. Approximately nine in every 10 patients diagnosed with IIH are obese.
- IIH most commonly affects **women in their 20s and 30s**.
- Taking **certain medications** can increase your chance of developing IIH. These include the combined oral contraceptive pill, tetracycline antibiotic, an overdose or more than your recommended dosage of vitamin A, and anabolic steroids.
- Patients who have been diagnosed with **hormonal problems** such as Cushing's syndrome and hypothyroidism, are more likely to develop IIH.
- A blood clot in the veins draining CSF from the brain (venous sinus thrombosis) can mimic IIH. We will rule out this condition by arranging an MRI scan before your diagnosis is confirmed.

What are the symptoms of IIH?

Common symptoms include the following.

- Headaches are the most common symptom.
- Double vision (also known as diplopia).
- Temporary loss of vision in one or both eyes, especially when carrying out activities that increase CSF pressure such as bending over, straining, and coughing.
- Blurred vision.
- Whooshing noise in your ears, in time with your heartbeat (pulsatile tinnitus).

Rarer symptoms can include the following.

- Aversion (dislike) to light (photophobia).
- Loss of peripheral vision (you cannot see things you are not looking directly at).
- Decreased depth perception (you lose some ability to judge the distance between objects).
- Nausea (feeling sick) and vomiting.
- Fatigue (extreme tiredness).
- Back pain.

Sometimes there are no symptoms, but papilloedema (swelling of the optic disc) is picked up at a routine optician's check-up.

How is IIH diagnosed?

If we think that you have IIH we will usually carry out the following tests to help make a diagnosis and rule out other conditions that have similar symptoms.

- A physical examination. This will involve using an ophthalmoscope to look in the back of your eyes to check for swelling around the optic nerves (known as papilloedema).
- A general examination of your body to look for other causes of high pressure. We will record your height and weight, so we can calculate your body mass index (BMI), which will be checked at your follow-up appointments.
- Visual testing will involve testing how clear your vision is.
- CT and MRI brain scans will help rule out other causes of your symptoms and help confirm your diagnosis.
- A lumbar puncture is the only test which can physically confirm whether the CSF is under high pressure. For more information, ask a member of staff for a copy of the Trust's **Lumbar puncture** leaflet, or download a copy from the Trust web site www.ekhuft.nhs.uk/neurology/

Using an ophthalmoscope

What is the recommended treatment?

Your doctor will discuss each of the following options with you before you start any treatment. If you have any questions about any of the following, please speak to your doctor. Weight loss along with one medication is what we usually recommend.

Weight loss

The most important and successful treatment for IIH is weight loss. Research has shown that weight loss in IIH leads to a reduction in the pressure and reduces symptoms such as headaches and visual changes. The amount of weight you need to lose to stop symptoms is not yet known, but research suggests that a target of 15% weight loss can help to resolve papilloedema linked with IIH and preserve eyesight. Your doctor will discuss your target weight loss with you in clinic.

If you lose enough weight your symptoms may improve and you will not need to take medication. However, if you put the weight on again IIH can return.

Even if your CSF pressure is normalised with weight loss or medication, you may still have headaches. Your neurologist will manage these headaches differently.

Medication

- **Acetazolamide**
  The most commonly used medication in IIH is acetazolamide (Diamox). This is a water tablet that works by reducing the amount of CSF you produce.

  Side effects include diarrhoea, changes to your taste, fatigue (extreme tiredness), nausea (feeling sick), paraesthesia (pin and needles), tinnitus (a ringing sound in your ears), vomiting, depression, and rarely kidney stones. Your dose will be increased slowly to reduce these side effects.

  Acetazolamide should be avoided in pregnancy, especially the first 12 weeks as it may be harmful to your baby. If you plan to become pregnant you should tell your neurologist.

- **Topiramate**
  Another less commonly used tablet is topiramate, which acts in a similar way to acetazolamide.

  The most common side effects are reduced appetite and low mood. You may also feel drowsy and have reduced concentration. Some people can have a stomach upset. Topiramate can reduce the effectiveness of hormonal contraceptives (including the pill).

  Topiramate should not be taken in pregnancy as there is an increased risk of major congenital malformations, such as cleft palate during the first 12 weeks of pregnancy. If you plan to become pregnant you will need to tell your neurologist.

Painkilling medications

Wherever possible avoid taking regular painkillers (such as paracetamol or ibuprofen), as these can cause another type of headache called medication overuse headache. If you do need to take these painkillers, try to only take them for 10 days or less each month. Avoid strong opioid medications such as tramadol and morphine.

Lumbar puncture

In the past, repeated lumbar punctures have been used therapeutically to try and control IIH. However, recent evidence no longer supports this. We now understand that the rate of production of CSF leads to the CSF pressure returning to its previous level within a few hours of having a lumbar puncture. The current recommendation is that lumbar puncture should only be routinely used to diagnose IIH.

In rare cases therapeutic lumbar puncture may be performed as a holding measure in those with serious visual loss who are awaiting surgery within the next few days.

Surgery
If you have severe IIH with visual problems and the above treatments have not worked, surgery may be considered to protect your vision. The most common procedures are those which divert (redirect) and drain the CSF, these are called 'shunts' and include the following.

- **Subcutaneous lumbar drain** is a temporary measure where a plastic tube is placed in your lower back using local anaesthetic (the area is numbed but you are awake for the procedure). This is sometimes used whilst planning a permanent shunt procedure.

- **Lumbo-peritoneal (LP) shunt** is a permanent plastic tube inserted into the spinal subarachnoid space at one end and into a cavity in the abdomen called the peritoneum at the other end.

- **Ventriculoperitoneal (VP) shunt** is a tube placed from the CSF containing chamber in the brain, called the lateral ventricle, down through the neck into the peritoneal cavity in the abdomen.

- **Optic nerve sheath fenestration** is an alternative procedure used to reduce the swelling of the optic nerves, whilst you lose weight. A small window is cut into the optic nerve sheath (a layer of fibrous tissue surrounding the optic nerve). This procedure carries the risk of blindness, so is only used in those who are severely affected by visual loss in IIH.

Although shunt surgery can provide successful long-term relief from IIH symptoms, all of the above procedures have risks. These include malfunction (the shunt does not work), infection, and kinking, blockages, and over-draining of the shunts. Shunt repair surgery is often needed.

What health professionals may be involved in my care?

- **Acute physicians** are the doctors working in the Emergency Department and Acute Medical Units. They will often be the first people you come in to contact with in the hospital.

- **Neurologists** are doctors who specialise in the care of the nervous system. They will be the specialist who takes ownership of your care once you have been seen in clinic.

- **Ophthalmologists** are doctors who specialise in the care of your eyes. Neurology will refer you to an ophthalmologist for regular visual field assessments to monitor for any impairment.

- **Dieticians** are health professionals who specialise in dietary advice and help with weight loss.

- **Neurosurgeons** are doctors who specialise in surgery of the nervous system.

What are the risks of IIH?
Visual loss is the most worrying risk. If left untreated IIH can lead to permanent loss of vision. If there is any concern that your vision has got worse, please contact your GP for urgent medical advice.

Does IIH get worse during pregnancy?

The effects of pregnancy on IIH vary from one individual to the next. Some women have had improvement in their IIH symptoms when they are pregnant and then a worsening of symptoms after childbirth.

Will I have regular follow-up appointments?

This will depend on how bad your condition is. Your doctor will discuss this with you at your clinic appointment.

What if I have any questions or concerns?

If you have any questions or concerns about your condition, please contact your relevant health professional.

Further information

- **Idiopathic Intracranial Hypertension UK**
  Web: iih.org.uk

This leaflet has been produced with and for patients

If you would like this information in **another language, audio, Braille, Easy Read, or large print** please ask a member of staff. You can ask someone to contact us on your behalf.

Any complaints, comments, concerns, or compliments please speak to your doctor or nurse, or contact the Patient Advice and Liaison Service (PALS) on 01227 78 31 45, or email ekh-tr.pals@nhs.net

Patients should not bring in large sums of money or valuables into hospital. Please note that East Kent Hospitals accepts no responsibility for the loss or damage to personal property, unless the property had been handed in to Trust staff for safe-keeping.

Further patient leaflets are available via the East Kent Hospitals web site www.ekhuft.nhs.uk/patientinformation

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