Idiopathic Intracranial Hypertension (IIH)/Pseudotumor Cerebri

What is idiopathic intracranial hypertension/pseudotumor cerebri?

Idiopathic intracranial hypertension (IIH), also called **pseudotumor cerebri**, is a condition in which there is **high pressure in the fluid surrounding your brain, spinal cord, and optic nerves**. This can cause headaches and problems with vision. Although the cause(s) of the condition is not fully understood (the definition of “idiopathic”), we know much about the condition itself.

Do I have a tumor?

Patients with pseudotumor cerebri do *not* have a tumor. A brain tumor may also cause increased intracranial pressure, which is why this condition is called “pseudotumor” it can cause symptoms like a real tumor, but the tests do not show any tumors (pseudo- means “false,” so pseudotumor means “false tumor”).

Who can get IIH?

IIH can affect anyone, but is more common in women (90%) than men. It is more common in teenagers and young women but can affect people of any age. It is much more common in those who are overweight, obese or who have had a recent substantial weight gain.

What causes IIH?

We do not know what causes IIH. However, there is a clear association to being overweight or obese. Not all overweight or obese people develop this condition. This likely means that there are unique features that predispose some people to develop IIH that we have yet to discover. Medical studies have shown that recent weight gain can cause IIH, and that weight loss alone can achieve remission in most cases. There is ongoing research into the cause/causes of IIH. Certain medications increase a person’s chance of having IIH. These mostly include tetracycline antibiotics (e.g., minocycline, doxycycline), high doses of vitamin A and retinoids so

these should be discontinued and avoided in the future by people with the diagnosis. Hormonal contraceptives (oral and implants) have not been convincingly shown to cause IIH and should not necessarily be discontinued in IIH patients.

How does IIH affect me?

Not everyone will develop all the symptoms of IIH. The most common symptoms of IIH include:

- **Headaches**
    - **Headache is the most common symptom**, although not everybody with IIH has headaches. These headaches are often worse upon awakening or lying down
- **Brief visual changes** such as dimming, blurring or graying of vision
    - These changes only last for a few seconds (often with bending or stooping then rising).
    - Some patients notice blind spots to the side of their vision in each eye.
- Sounds of a whooshing heartbeat-like noise in the ears (**pulsatile or pulse-synchronous tinnitus**)
- **Neck pain**

Less common symptoms include:

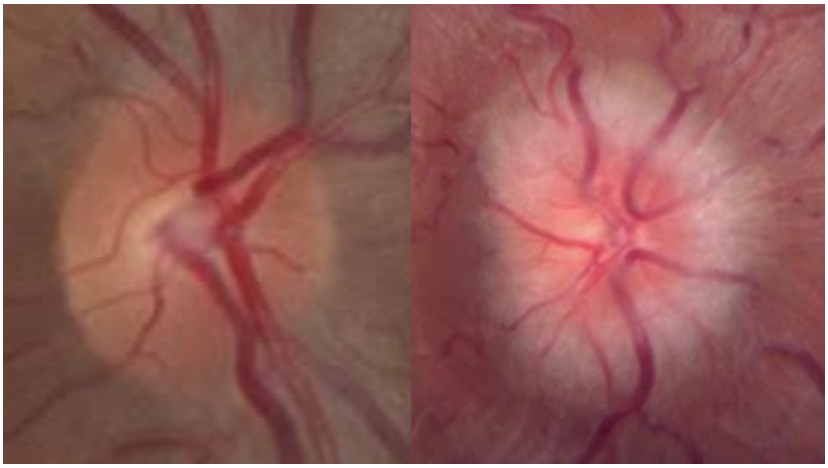
- Double vision
- Nerve or back pain
- Dizziness
- Nausea

In severe cases, IIH can cause severe peripheral vision loss and blurred central vision. If left untreated, there is a high risk of permanent disabling loss of vision.

How is IIH diagnosed?

There are three important steps that must be completed in order to diagnose IIH:

- A **complete eye exam** must be performed, including **special tests on your visual field** and **dilation of the pupils** to look for swelling of the optic nerves caused by high pressure in the brain (**papilledema**).



Courtesy of Dr. Valerie Biousse, M.D.

Normal optic nerve (left) and papilledema (right). The normal optic nerve looks like a flat orange-yellow disc. In papilledema, the edges of the disc become blurred and fuzzy, and the entire nerve eventually swells.

- **Brain scans**, in particular magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) must show that there is no tumor, blood clot, or other cause for the suspected high pressure. Occasionally if an MRI is not readily available, a CT (computed tomography) scan will be done initially. Sometimes the brain scan report will say that there are changes to the pituitary gland, fluid around the optic nerves or narrowing of the brain veins that suggest IIH. **These can be useful clues for your doctor to diagnose IIH but cannot replace a lumbar puncture (spinal tap) to check the pressure of the spinal fluid.**
- A **lumbar puncture** must be performed to confirm a high pressure and a normal spinal fluid (CSF). **The lumbar puncture must be performed with you relaxed and lying on your side with your legs straight, or in the prone position (on your belly) when done under radiographic guidance (fluoroscopy) for the reading to be accurate.**

Why do I need to see a neuro-ophthalmologist?

- **Neuro-ophthalmologists are the experts with the most experience and knowledge about IIH.** Though many neurologists and ophthalmologists may also see and manage people with IIH, neuro-ophthalmologists receive specific training to help take care of all forms of IIH. Many specialists may be involved in your treatment:
- **Neuro-ophthalmologists** evaluate and guide the medical and/or surgical treatment.
- **Ophthalmologists** or neuro-ophthalmologists monitor your vision by performing **dilated eye exams**, **visual field testing**, **photographs and/or a special imaging test called optical coherence tomography (OCT).** Some ophthalmologists or neuro-ophthalmologists are specifically trained to perform **optic nerve surgery**, which may be one of several alternatives for management of severe cases of IIH.
- **Neurologists** may help coordinate the testing for diagnosing IIH and help treat headaches.
- **Neurosurgeons** may be involved if surgery (shunt) is necessary to control the high pressure.

How is IIH treated?

Most people with IIH are treated with medications and weight loss. Less than 1 out of 10 people (< 10%) with IIH will require surgical treatment.

Relatively few people become blind from IIH and most people recover if they are treated early. Surgery is needed if medical treatment and weight loss does not stop the visual loss. In very rare cases, blindness may occur even with correctly administered medications and surgery.

Main treatments of IIH

Do I need treatment?

If you do not have significant headaches or vision loss, no treatment may be necessary, although weight reduction is always a good idea to prevent the disease from worsening. The decision to treat or not treat is based on the **clarity of your vision (visual acuity)**, **peripheral vision (visual fields)**, **presence of papilledema**, and **whether or not you have headaches**.

Weight loss is hard to do. How much weight do I need to lose?

Weight loss can be a sensitive issue for both the doctor and the patient. Weight loss is difficult to achieve and maintaining a lower weight long-term is challenging. **Almost all people with IIH are overweight, and weight loss can be a very effective component of treatment.** Weight reduction programs that include lifestyle modification and a diet low in sodium have been shown to be effective in treating IIH. The goal is weight loss of 5-10% of your starting weight; for example, someone with a starting weight of 250 lbs (113 kg) would have a target weight loss of 12.5 lbs-25 lbs (5.6 kg-11.3 kg). In some cases, weight management (bariatric) surgery may be helpful.

What medications are used for treating IIH?

Diamox (acetazolamide) is the most common medication used for treating IIH. It is thought to lower brain pressure by reducing fluid (cerebrospinal fluid, or CSF) production. Research shows that Diamox significantly improves vision, papilledema, quality of life and CSF pressure.

Common side-effects of Diamox include:

- Tingling of fingers and toes
- Loss of appetite
- Metallic taste when drinking fizzy drinks (carbonated beverages)
- Tiredness (fatigue)
- Kidney stones may occur in rare cases (2-3%)

If you are not able to take acetazolamide, your doctor may prescribe another medication called Topamax (topiramate) which may help the headache and also assists with weight loss. Side effects of Topamax may include mild slowing of thought processing.

What medications help for headache in IIH?

There are no medications specifically designed for IIH-related headaches, although many neurologists specifically use topiramate because it may also lower brain pressure and may act as an appetite-suppressant. Over-the-counter analgesics such as non-steroidal anti-inflammatory drugs (NSAIDS) and/or migraine medications may be helpful if you have headaches even after the IIH treatment is successful in lowering the intracranial pressure and resolving the papilledema.

Using pain medications >2-3 days per week for more than a few months can make you dependent on the medications, so that when you don't take the pain medications you get a withdrawal headache (**medication-overuse headache**). This is a background daily headache that may be confused with high pressure and can make the severe headaches worse. Stopping these medications is helpful. Consulting with a neurologist may be useful if headaches are a persistent concern.

Surgical treatments

Surgery is recommended when vision is getting worse from high pressure despite aggressive treatment with medications. Because surgery has small but potentially dangerous risks to your life and/or vision, surgery for IIH is not recommended when your vision is good. The goal of surgery is to release pressure around the optic nerves. There are three main types:

1. **Shunt surgery:** a **neurosurgeon** places one end of a flexible tube into one of the normal fluid-filled spaces in your brain (ventricle) or into your spine (lumbar) and the other end into another part of your body, such as your abdomen (peritoneum).
2. **Venous stenting:** A **neurovascular surgeon** inserts a device that holds open a vein that drains blood from the brain. It is a newer technique for lowering brain pressure; hence, specialists are still considering where this approach fits into the management.

3. **Optic nerve sheath fenestration:** an **orbital or neuro-ophthalmic surgeon** creates a small window in the lining (sheath) around your optic nerve to allow the fluid to drain behind your eye.

When will the IIH go away? Can the IIH come back?

IIH may resolve over months to years or it may be a lifelong medical problem. IIH can return, and is often linked to regaining weight.

What Should I watch out for after being diagnosed with IIH?

You should let your neuro-ophthalmologist know about any changes in symptoms such as worsening headache. You should report changes in vision immediately since that could indicate worsening optic nerve swelling, which if not treated could lead to permanent loss of vision.

Will I need another spinal tap in the future?

Generally not, unless there is a new problem. IIH is rarely treated with multiple spinal taps.

Does IIH run in families?

IIH rarely runs in some families (approximately 5%).

Additional Reading/Resources

Other Websites

- **National Eye Institute**
    - https://www.nei.nih.gov/learn-about-eye-health/eye-conditions-and-diseases/idiopathic-intracranial-hypertension
- **American Academy of Ophthalmology**

    - https://www.aao.org/eye-health/diseases/what-is-idiopathic-intracranial-hypertension
- **Mayo Clinic Website**
    - https://www.mayoclinic.org/diseases-conditions/pseudotumor-cerebri/symptoms-causes/syc-20354031

Support Groups

- Links to two major support groups are listed below.
    - https://www.dailystrength.org/group/pseudotumor-cerebri
    - https://www.facebook.com/iihsupportgroup/community/

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