



NANOS

Patient Brochure

Idiopathic Intracranial Hypertension

(Pseudotumor Cerebri)

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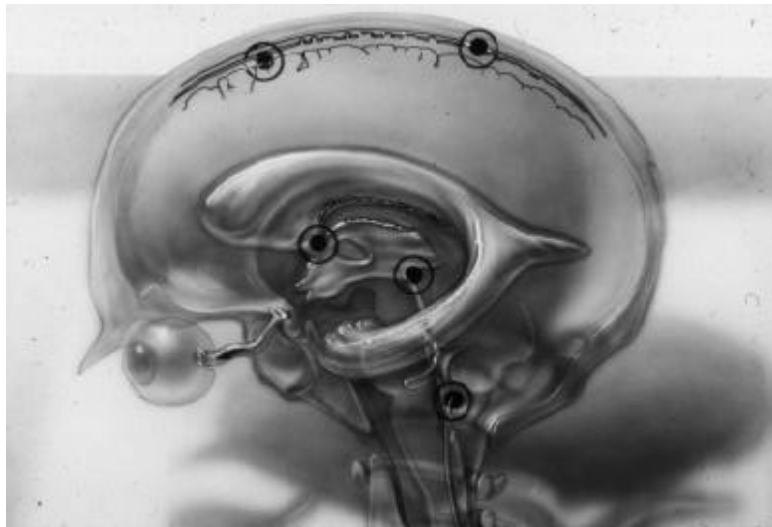


IIH Idiopathic Intracranial Hypertension (IIH)

Idiopathic intracranial hypertension (IIH), also called pseudotumor cerebri, is a condition in which high pressure inside your head can cause headache and problems with vision. While the cause of the condition is not known, we know much about the condition. IIH is predominantly a disease of young women in the childbearing years. Less commonly, IIH may occur in children, men, and patients who are not overweight. It is characterized by elevated pressure in the fluid filled spaces surrounding the brain.

Anatomy:

The brain and spinal cord are bathed in a clear fluid called cerebrospinal fluid (CSF).



This supplies oxygen and nutrients to portions of the brain that do not have their own blood supply. CSF also cushions the brain against traumatic injury. Cerebrospinal fluid is produced from blood flowing through the choroid plexus within the ventricles (open cavities within the brain substance). The fluid is eventually absorbed through the arachnoid granulations mostly at the superior sagittal sinus that collects venous blood at the very top of the cranial cavity returning it to the heart.

Physiology:

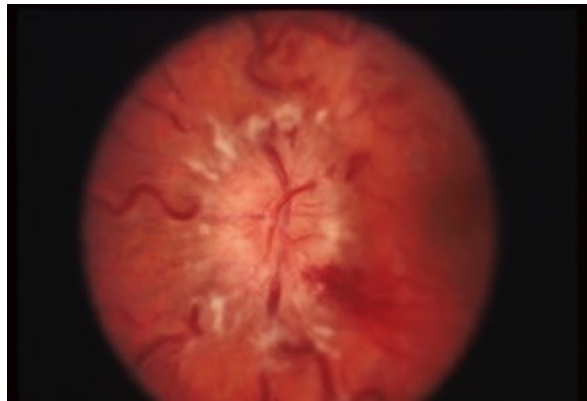
In IIH CSF outflow is thought to be partially blocked. This leads to high pressure inside your head. The pressure is transmitted to the optic nerve head at the back of the eye through the optic nerve sheath (surrounding each of the optic nerves) producing the swelling seen at the optic disc (papilledema). The reason for decreased CSF outflow is not clear. The elevated pressure inside the head may lead to headache. Swelling of the optic disc (optic nerve head) can damage the optic nerve fibers producing decreased vision.

Symptoms:

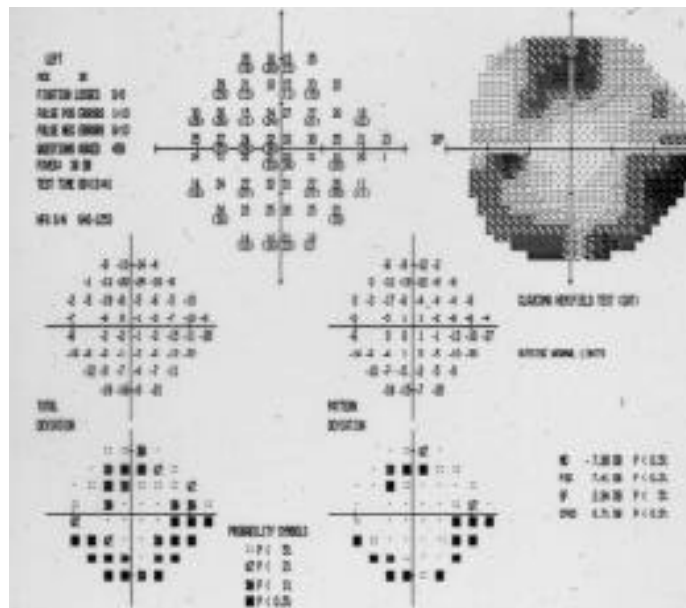
The most common symptoms of raised intracranial pressure are headache and visual loss. Other common symptoms are neck pain, back pain, pulsatile tinnitus and double vision. The optic nerve swelling may eventually lead to loss of vision seen as dimming, blurring or graying of vision. Frequently patients notice visual disturbance lasting for a few seconds (often associated with bending or stooping). High CSF pressure may cause damage to the nerves that move the eyes side to side resulting in double vision. Patients may also be aware of a rushing noise in their ears (pulsatile or pulse synchronous tinnitus). Nausea and vomiting may occur if the pressure is high and especially with a severe headache.

Signs:

The most important clue to the presence of IIH is the finding of optic disc swelling seen when looking in the back of the eye.



This is done after the pupil has been dilated. The optic disc swelling is usually present in both eyes and is usually associated with retained central vision. Peripheral vision (detected on visual field testing) is usually abnormal.



and is one of the most important means of judging both the necessity for and effectiveness of treatment.

Diagnosis:

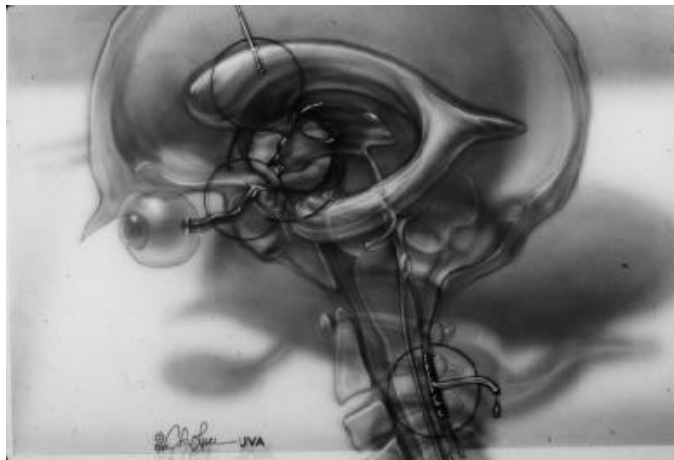
Because brain tumors, and a blood clot in the veins draining blood from the head, some medications and excessive intake of vitamin A and other conditions cause the signs and symptoms of raised ICP, the diagnosis of IIH requires a brain MRI scan. The diagnosis also requires a spinal tap, both to document that the CSF pressure is high and to examine the CSF for evidence of other diseases causing increased intracranial pressure. The finding of abnormal CSF cells, inflammatory cells, or elevated protein may indicate a previous infectious, inflammatory, or tumor related cause of elevated intracranial pressure.

Treatment:

Weight loss: Weight reduction programs that include lifestyle modification and a diet low in sodium may be effective in treating IIH. The goal is 5-10% weight loss and improvements may take months to begin. Extremely overweight patients (BMI greater than 40) may consider bariatric surgical procedures as a treatment.

Medical treatment: Diamox (acetazolamide), a pill used for treating glaucoma, may lower pressure by reducing CSF production. It has been shown to significantly improve vision, optic nerve head swelling, quality of life and CSF pressure in a randomized placebo controlled clinical trial. It usually causes tingling of fingers and toes, loss of appetite, and a metallic taste to carbonated beverages. It may also cause fatigue. In 2-3% of cases, kidney stones occur. Diuretics, such as Lasix, may also be prescribed. Other related medications such as topiramate (Topamax) and methazolamide may also be used. Steroids (prednisone or dexamethasone) have been used to protect the optic nerve but have limited long term use and may produce significant side effects; they are also complicated by rebound increased intracranial pressure when they are tapered and discontinued. Therefore, they are seldom used.

Surgical treatment: CSF Pressure may also be lowered by draining off or diverting CSF. This may be accomplished with a spinal tap but continuing production will replace the lost volume within hours. If too much fluid is drained the patient may suffer a low pressure or post spinal tap headache. Stereotactic ventriculo-peritoneal shunts may be used to drain fluid from the space around the brain to the abdominal cavity.



In patients with worsening visual field examinations or decrease in visual acuity may have an optic nerve sheath fenestration (opening in the optic nerve covering) that may protect the optic nerve from further damage. A small hole or multiple slits are placed in the optic nerve sheath just behind the eye using an operating microscope. Patients may be able to return home the same day. Complications include eye redness and double vision (which usually goes away). In rare cases vision may get worse.

Over-the-counter pain medications such as Aleve (Naprosyn) may be effective in treating headache but should not be over used as rebound worsening may occur. Medications used to treat migraine may also be effective, especially low dose amitriptyline.

It is not rare for a migraine component to exist in a patient with IIH. Thus, correction of the increased CSF pressure may not relieve all headaches.

Frequently Asked Questions

When will IIH go away?

IIH may resolve over months or it may be a lifelong medical problem like arterial hypertension.

Do I need to be treated?

If you have no significant headaches or evidence of vision loss by visual field examinations, no treatment may be necessary (weight reduction is always a good idea to prevent the disease from worsening). The decision to start treatment or to alter treatment from dietary to medical to surgical intervention depends on the presence and progression of visual loss and the status of headaches. Headaches that do not respond to over-the-counter medications and, even more importantly, evidence of worsening visual loss are indicators that treatment is necessary.

I don't like those visual field examinations. Can't you just look at the back of the eye?

Unfortunately swelling of the optic nerve head (papilledema) does not tell us how well your optic nerve is working. To determine whether there is further damage to the optic nerve acuity and visual field testing is necessary.

Do I have a tumor?

While some call the disease "pseudotumor," by definition, patients with pseudotumor cerebri specifically do not have a tumor. A tumor may cause increased intracranial pressure and therefore be mistaken for IIH but the tumor should be readily seen on a brain MRI scan.

Do I need another spinal tap?

Generally not. We can get a better estimate of the elevation of CSF pressure by examining the optic disc for papilledema.

Will I go blind from IIH?

Unless there is marked visual field loss, blindness is preventable in most IIH patients. If medical treatment does not stop progressive visual loss, a surgical procedure is needed. Only a few percent of patients go blind with treatment.

Will my IIH recur?

Yes, IIH is a life-long disease like high blood pressure. When recurrence happens, it often is due to weight gain.

Is IIH hereditary?

There is a hereditary component to IIH. In the Idiopathic Intracranial Hypertension Treatment Trial 5% of participants had a family member with IIH.

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

While weight loss seems very difficult, a variety of minor lifestyle changes (like walking a half hour a day) and changing eating habits to eat healthily most of the time are often accomplished. Motivation for some patients comes from fear of going blind and with adequate weight loss (5-10% total body weight) medication doses can be lowered and sometimes discontinued.