

Idiopathic Intracranial Hypertension (Pseudotumor Cerebri)

WHAT IS IDIOPATHIC INTRACRANIAL HYPERTENSION?

Idiopathic intracranial hypertension (IIH) is a disorder that results from an increase in the pressure of the Cerebro-Spinal Fluid (CSF) that cushions and protects the brain and spinal cord. The CSF is constantly produced in the brain and reabsorbed back into the bloodstream at a fairly constant rate. This allows the fluid pressure around the brain to remain constant. An older name for IIH that is still used is Pseudotumor Cerebri.

WHAT ARE THE SYMPTOMS OF IDIOPATHIC INTRACRANIAL HYPERTENSION?

- Headaches that are generally nonspecific in location, type and frequency and can be associated with nausea and vomiting.
- Pulsatile tinnitus is a rhythmic or pulsating ringing heard in one or both ears.
- Horizontal double vision can be a sign of pressure on the 6th cranial nerve(s).
- Nonspecific radiating pain in the arms or legs (radicular pain).
- Transient obscurations of vision (TOV), which are temporary dimming or complete blacking out of vision.
- Visual field defects. These defects can occur in the central as well as the peripheral vision.
- Loss of color vision.

WHAT CAUSES IDIOPATHIC INTRACRANIAL HYPERTENSION?

The cause is usually not known. A common explanation for increased pressure is a problem with the reabsorption of this fluid back into the body, which causes the pressure to increase. Sometimes the cause is found and then it is called secondary intracranial hypertension.

WHAT ARE THE RISK FACTORS FOR IDIOPATHIC INTRACRANIAL HYPERTENSION?

The most common risks in adults are being obese and female. In children, the causes for IIH are still being researched. Causes of secondary intracranial hypertension include certain medications (oral contraceptives, steroids, vitamin A, Isotretinoin, lithium, growth hormone, nitrofurantoin, phenytoin, sulfa drugs, minocycline, Tamoxifen, naladixic acid, thyroid replacement, tetracycline, and some chemotherapeutic drugs) and medical conditions (dural venous sinus thrombosis, kidney disease, head injuries, Lyme disease, lupus, acute sinusitis or mastoiditis, measles, blood clotting disorders, anemia, leukemia, periodic fever and meningitis).

CAN IDIOPATHIC INTRACRANIAL HYPERTENSION AFFECT CHILDREN?

Yes, and cases are divided into two groups. The first group includes prepubescent children and tends to be secondary and affects males and females equally. Teenagers make up the second group and is more typical of the adult form and is usually associated with weight gain and obesity.

HOW IS PEDIATRIC IDIOPATHIC INTRACRANIAL HYPERTENSION DIAGNOSED?

If idiopathic intracranial hypertension is suspected, an ophthalmologist usually examines the optic nerve for swelling (papilledema). Testing by the ophthalmologist may include visual fields and special photography including optical coherence tomography (OCT).

If symptoms of headaches or optic nerve swelling is found, an MRI of the brain and MRV of the venous system will be ordered. Then, if the MRI does not show structural changes, a lumbar puncture (spinal tap) is performed to determine the pressure as well as content of the CSF. High opening pressure on the lumbar puncture will diagnose IIH.

HOW IS IDIOPATHIC INTRACRANIAL HYPERTENSION TREATED?

After the MRI and the lumbar puncture, usually a diuretic medication called Diamox is used to decrease the pressure. Lasix and Topamax are sometimes used as well. In very severe cases of papilledema, sometimes steroids are used. Occasionally, in very severe cases, surgery to shunt the CSF fluid or surgery to protect the optic nerve (optic nerve sheath fenestration) are performed. Any secondary causes are also treated.

The ophthalmologist monitors vision and resolution of papilledema to determine if treatment is working. Neurology and sometimes neurosurgery teams will also be involved in managing the IIH.

ARE THERE TREATMENTS BESIDES MEDICINE AND SURGERY THAT CAN HELP?

The most important thing anyone can do to improve IIH is to lose weight if they are obese. Ask your doctor about local resources and clinics designed to assist you in your weight loss.

WHAT IS THE PROGNOSIS OF IDIOPATHIC INTRACRANIAL HYPERTENSION?

As many as 10 percent of the people with pseudotumor cerebri experience progressively worsening vision and may eventually become blind. Even if symptoms have resolved, a recurrence can occur months or even years later. It is important to have regular follow-up eye examinations.



Fig. 1: Normal optic nerve.

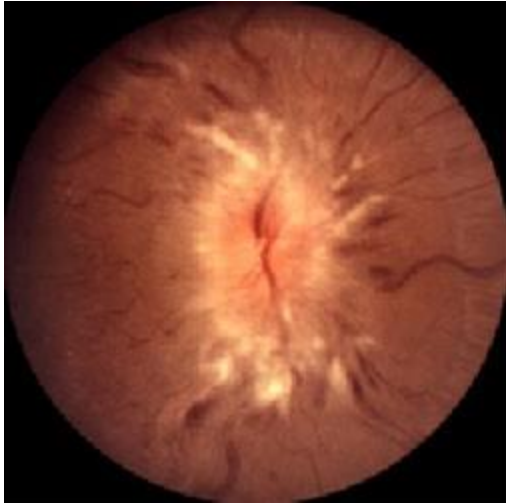


Fig. 2: Optic nerve with papilledema

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