# Pseudotumor Cerebri (Idiopathic Intracranial Hypertension)

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## A Patient’s Guide

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What are the symptoms of Pseudotumor Cerebri?

Patients with Pseudotumor Cerebri typically experience headaches and episodes of blurred vision. It is common to have brief “grey-outs” of vision when standing up. Headaches may be accompanied by a “whooshing” sound or ringing in the ears. Pseudotumor can cause permanent visual loss. Typically, patients first develop loss of their peripheral vision. If Pseudotumor Cerebri is not treated, it can cause gradually progressing visual loss that affects central (straight-ahead) vision.

What causes Pseudotumor Cerebri?

The spaces within and around the brain are filled with cerebrospinal fluid that constantly circulates. This fluid needs to be produced and removed at the same rate. In patients with Pseudotumor Cerebri, this balance is disrupted, and they develop elevated pressure within the skull.

Increased pressure within the skull causes headaches and swelling of the optic nerves (which are the cables that connect the eyes to the brain). When the optic nerves are swollen, it is common to have brief episodes of visual blurring or “grey-outs.” Over time this also causes progressive loss of vision.

A number of risk factors are associated with developing Pseudotumor Cerebri. Recent weight gain is strongly linked with this condition. It occurs more commonly in women than in men.

There are several medications associated with the development of Pseudotumor Cerebri. These include certain antibiotics (in the tetracycline family) and medicines derived from vitamin A (including treatments for acne).

Other medications may also be associated with Pseudotumor Cerebri, but are not as well established as definite risk factors.

Symptoms

The symptoms of Pseudotumor Cerebri include:

- Headaches
- Ringing in the ears
- “Grey-outs” of vision
- Reduced peripheral vision

Pseudotumor Cerebri is associated with:

- Weight gain
- Pregnancy
- Medications
  - Tetracycline antibiotics
  - Vitamin A drugs (i.e., to treat
The diagnosis of Pseudotumor Cerebri usually depends on the following features:

- Symptoms
- Abnormalities on examination (including swelling of the optic nerve)
- Diagnostic tests (including MRI and lumbar puncture)

How is Pseudotumor Cerebri diagnosed?

The doctor will diagnose Pseudotumor Cerebri based on symptoms and the results of the examination and diagnostic tests.

Patients with Pseudotumor Cerebri have swelling of the optic nerve that is visible to the physician during examination with the ophthalmoscope. Sometimes areas of poor circulation or bleeding in the retina are also visible.

Examination of the visual fields (often using a computerized test) may demonstrate enlargement of the normal “blind-spot” in each eye. There may also be loss of peripheral vision, depending on the severity of the condition.

Unless severe visual loss has already occurred, the ability to read the eye chart and color vision are typically normal.

The optic nerves, seen in the back of the eye, are swollen and there are small areas of bleeding.
Diagnosis

What tests are required to confirm a diagnosis of Pseudotumor Cerebri?

In order to diagnose a patient with Pseudotumor Cerebri, the doctor will have to order additional tests. The first test is an MRI scan of the brain. An MRI uses a large magnet to take a detailed picture of the brain (it does not use X-ray radiation). The MRI will check if there is an abnormal growth in the brain or its coverings that is the reason for elevated pressure in the skull. It will also check for blood clots in the major veins in the brain.

The second test that is required is a lumbar puncture (also known as a “spinal tap.”) This test is performed by inserting a small needle into the back to measure the pressure of the cerebrospinal fluid. A small sample of the fluid is collected to check if there is any infection or inflammation that has caused elevated pressure. In patients with Pseudotumor Cerebri, the pressure is elevated but the cerebrospinal fluid itself is normal.
What are the treatments for Pseudotumor Cerebri?

The goal in treating Pseudotumor Cerebri is to use appropriate measures to reduce pressure within the skull and prevent further visual loss. Often this requires a combination of treatments.

If a patient with Pseudotumor Cerebri has had recent weight gain, one of the most important ways to treat this condition is weight loss. Losing approximately 10 lbs can have a significant impact, reducing the need for other medications or surgery. When necessary, a weight loss program can be developed under the guidance of a nutritionist or personal trainer.

Often the doctor will prescribe medications that help manage Pseudotumor Cerebri. They help to lower the pressure inside the skull, which can improve headaches and help prevent visual loss. A medicine that is used very frequently is Acetazolamide (Diamox®). Other potentially helpful medicines are topiramate (Topamax®), zonisamide (Zonegran®), and furosemide (Lasix®). The doctor will address drug allergies, drug interactions, and possible side effects when considering these medications.

In some cases of Pseudotumor Cerebri, there can be rapid, severe visual loss that requires more aggressive treatments. Patients may be admitted to the hospital to undergo a surgical procedure that aims to quickly reduce the elevated pressure inside the skull. A shunt or drain can be placed within the skull or spine to improve the flow of cerebrospinal fluid and reduce pressure. In some cases, a procedure called Optic Nerve Sheath Fenestration may be performed to help prevent visual loss by relieving pressure around the optic nerves.

Weight loss is one of the most important treatments of Pseudotumor Cerebri.

Medicines can help reduce elevated pressure within the skull. These include acetazolamide (Diamox®), topiramate (Topamax®), zonisamide (Zonegran®), and furosemide (Lasix®).

In severe cases, surgical procedures can improve the flow of cerebrospinal fluid and help prevent rapid visual loss.
What is the prognosis for patients with Pseudotumor Cerebri?

Many patients with Pseudotumor Cerebri have improvement in their symptoms after approximately several months to a year. Under the guidance of their doctor, they can usually stop taking medications at this time. In patients who are overweight, it is critical to achieve and maintain weight loss in order to improve the chance of getting better.

Some patients with Pseudotumor Cerebri have a complete recovery without any visual changes. Other patients, however, can have permanent visual loss, which can range from mild to severe.

Currently, there are no treatments to reverse the permanent injury to the optic nerves caused by Pseudotumor Cerebri. Treatment of this disease is focused on halting visual loss that has already occurred.

Where can I get more information on Pseudotumor Cerebri?

For more information on Pseudotumor Cerebri, consider the following sources:

- Intracranial Hypertension Research Foundation
  www.IHRFoundation.org

- North American Neuro-Ophthalmology Society
  www.nanosweb.org/i4a/pages/index.cfm?pageID=3283