NANOS Patient Brochure

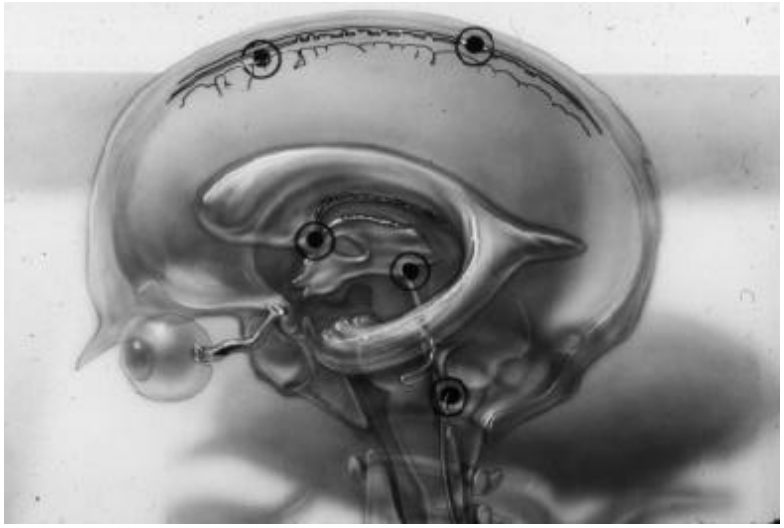
Pseudotumor Cerebri

Pseudotumor Cerebri

Your doctor thinks you may have pseudotumor cerebri. This is a condition in which high pressure inside your head can cause problems with vision and headache. In the days before CT and MRI scans, doctors who noted swelling of the optic disc (the beginning of the optic nerve in the back of the eye) were concerned about the possibility of a tumor within the head. Patients with optic disc swelling but no evidence of a tumor were said to have "Pseudotumor."

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 made from blood flowing through the choroid plexus within the ventricles (open cavities within the brain substance). The fluid is eventually absorbed through the superior sagittal sinus that collects venous blood at the very top of the cranial cavity returning it to the heart.

Physiology:

In pseudotumor CSF outflow is blocked. This leads to high pressure inside your head. The pressure is transmitted to the back of the eye via the optic nerve sheath (surrounding each of the optic nerves) producing the swelling seen at the disc (papilledema). The reason for decreased outflow is not clear. As it does seem to occur more often in young women who are overweight there is the possibility of a hormonal influence. In some cases antibiotic or steroid use may be associated with pseudotumor. High doses of vitamin A may also lead to increase in intracranial pressure. Pseudotumor may occur in children, men, and patients who are not overweight. The elevated pressure in side the head may lead to headache. Swelling of the optic disc can damage (possibly permanently) the optic nerve producing decreased vision.

Symptoms:

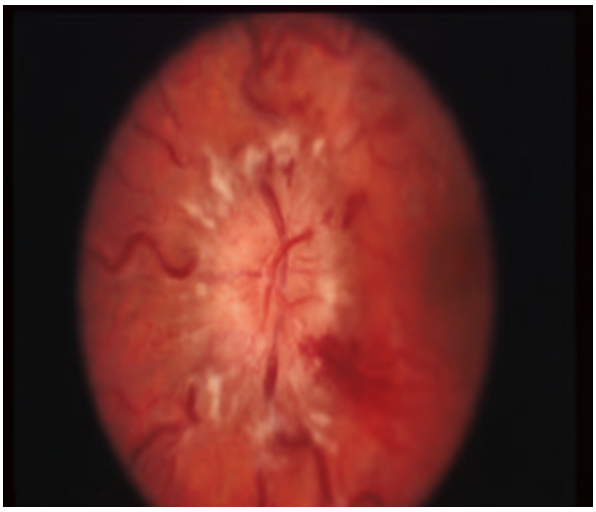
The most common symptoms of high intracranial pressure are headache and visual loss. The headache may be located anywhere; frequently in the back of the neck. It is usually steady but may be pounding. It may be very severe, and unlike migraine, it may awaken the patient in the middle of the night. It also may worsen with bending or stooping. The optic nerve swelling may eventually lead to loss of

vision seen as dimming, blurring or graying of vision. Patients may be aware of difficulty seeing to the side. Frequently patients notice visual disturbance lasting for a few seconds (often associated with bending or stooping). These visual "obscurations" may be very disturbing but do not increase the risk of visual loss.

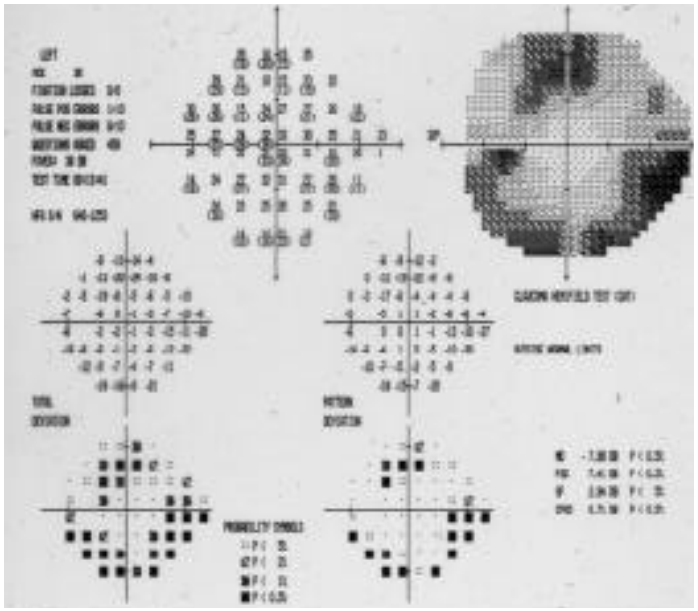
High pressure may cause damage to the nerves that move the eyes resulting in double vision. Patients may also be aware of a rushing noise in their ears. Nausea and vomiting may occur if the pressure is high and especially with a severe headache.

Signs:

The most important clue to the presence of pseudotumor is the finding of disc swelling upon looking in the back of the eye.



This is done after the pupil has been dilated. The disc swelling should be 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. The doctor will also want to check for asymmetric optic nerve involvement by looking at the swinging flashlight test. Eye movement problems may occur and be noted by the patient as double or blurred vision.

Diagnosis:

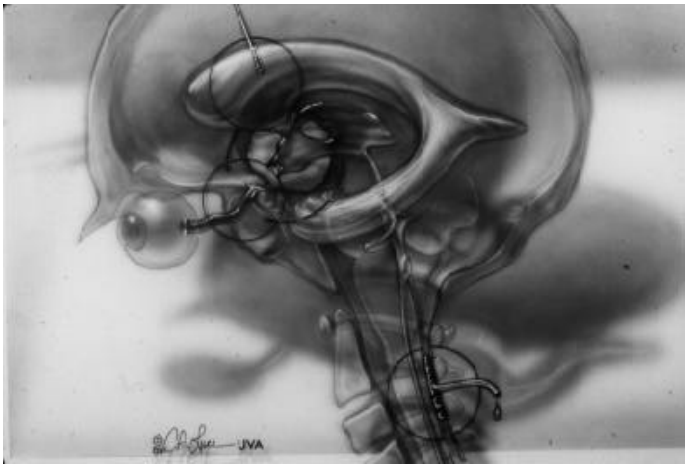
Because tumors, abnormal connections between arteries and veins, and a clot in the veins of the head may produce similar signs and symptoms, the diagnosis of pseudotumor requires a normal MRI scan. The diagnosis also requires a spinal tap. This will document elevated pressure inside your head and make sure there are no other abnormalities in the CSF. The finding of abnormal cells, inflammatory cells, or elevated protein may indicate a previous infectious, inflammatory, or tumor related cause of elevated intracranial pressure. In rare cases, an angiogram, where a catheter is placed in the arteries and veins going to the head, may be necessary to exclude an abnormality of the blood vessels. Headache may persist in spite of treatment. Since headaches may be do to other causes it may be necessary to recheck intracranial pressure. A repeat spinal tap fail may indicate persistent pressure elevation. It is possible that pressure is only elevated transiently. In unusual circumstances a small pressure sensor may be inserted into the skull (requiring hospitalization) providing a continuous pressure read out over 1-2 days.

Treatment:

Reduction in CSF production or increase in its outflow may reduce intracranial pressure. Weight reduction programs (in overweight patients) may be effective. If vitamin A is elevated its intake should be limited.

Diamox (acetazolamide), a pill used for treating glaucoma, can lower pressure by reducing CSF production. It can cause side effects, including tingling of fingers and toes, loss of appetite, and intolerance of carbonated beverages. It may alter taste and causes frequent urination and fatigue. Much more rarely, it may predispose the patient to kidney stones or even cause bone marrow blood problems. Other agents similar to Diamox, such as Neptazane (methazolamide), may produce fewer side effects but may not be as effective. Diuretics, such as Lasix, may also be prescribed. Steroids (prednisone or dexamethasone) have been used to protect the optic nerve but have limited long term use and may produce significant side effects.

Pressure may also be lowered by draining off 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. Continuous drainage may be surgically accomplished by placing a catheter between the spinal canal and the abdomen (lumbo-peritoneal shunt). Potential problems include local back pain and future obstruction of the shunt leading to the need for further intervention.



In patients with worsening visual fields or decrease in central acuity, who do not have severe headaches, an optic nerve sheath fenestration 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 should 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. This procedure may not be successful in all cases and if the patient has persistent or recurrent vision problems, re-operation may be indicated.

Over the counter pain medications may be partially effective in relieving headache but should not be over used as rebound worsening may occur. Medications used to treat migraine may also be effective.

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

Frequently Asked Questions

Do I have a tumor?

While the most commonly used term "pseudotumor," has that word in it, by definition patients with pseudotumor cerebri specifically do not have a tumor. A tumor may cause increased intracranial pressure and therefore be mistaken for pseudotumor but this should be seen on an MRI scan.

When will this go away?

It was thought in the past that pseudotumor was a self-limited disease that resolved over 1-2 years. While it is possible for pressures to vary over time, prolonged problems with CSF outflow may result in long-term increased pressure.

Do I need to be treated?

If you have no significant headaches or evidence of vision loss (including visual fields) no treatment may be necessary (weight reduction is always a good idea). The decision to start treatment or to alter treatment from dietary to medical to surgical intervention depends on the function of the optic nerve and the status of headaches. Headaches that do not respond to over-the-counter medications and, even more importantly, evidence of damage (particularly progressive damage) to the optic nerve function are major indicators that treatment is necessary.

I hate those visual fields. Can't you just look at the back of the eye?

Unfortunately the appearance of the optic nerve (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 need another spinal tap?

In the past we treated pseudotumor with repetitive spinal taps. This is not effective. While we would like to know the intracranial pressure, re-measuring becomes important only when there is evidence of further damage to the optic nerve (worsening visual field or central vision) or worsening headaches. It is then important to distinguish between inadequately treated intracranial pressure and some possible additional cause of worsening symptoms. As mentioned, if the pressure on the repeat spinal tap is low there still may be a need for further monitoring. Fortunately this form of problem producing worsening of symptoms is rare.