

## Pseudotumor Cerebri or Idiopathic Increased Intracranial Pressure (IICP)

Pseudotumor cerebri (also called idiopathic increased intracranial pressure, IICP) is a disorder in which a person's pressure inside his/her brain is increased above normal. The cause is not known or "idiopathic." It is called "pseudo-tumor" because the pressure is elevated as may be seen in people with brain tumors but there is no brain tumor present. Pseudotumor cerebri is a diagnosis of exclusion, meaning tests are needed to excluding known causes of increased brain pressure (brain tumor, hydrocephalus known as "water on the brain" or other causes). Typically, people with pseudotumor cerebri have headaches and swollen optic nerves (eye nerves) called papilledema. The most serious issue with pseudotumor is potential injury of the optic nerves which could cause irreversible visual impairment. The diagnosis is made by a medical history, examination, an ophthalmologic evaluation (eye exam by a pediatric ophthalmologist), brain MRI/MRA/MRV and a lumbar puncture ("spinal tap").

If imaging is normal, then a lumbar puncture is required to measure the brain pressure. If the pressure is elevated then fluid (Cerebral Spinal Fluid or "CSF") is removed to lower pressure. Sometimes just one lumbar puncture resolves the disorder, but sometimes repeated lumbar punctures are needed. Rarely some patients need a neurosurgical procedure to permanently reduce pressure (shunt). Sometimes a medication, Acetazolamide or "Diamox" is tried to lower pressure in addition to the lumbar punctures. Diamox does have some annoying side effects in some people (nausea, headaches, numbness).

Usually headaches resolve after the lumbar puncture. Sometimes return of headaches (days or weeks or even months later) requires a repeat lumbar puncture to see if the pressure is elevated or not. The most important treatment for pseudotumor is repeated ophthalmologic (eye) examinations to assess for resolving or persistent papilledema and visual function or impairment.

If the LP and lowering of ICP results in resolution or marked improvement of headaches, with a history of normal imaging, then the diagnosis is likely IICP. Then, the treatment is monitoring optic nerves for decreasing papilledema and serial LPs to check on pressure and to lower pressure if headaches recur and/or papilledema persists. Acetazolamide is considered useful for lowering ICP but my experience is too many GI side effects. If papilledema persists or vision impairment persists despite repeated lumbar punctures, then a more permanent treatment should be considered such as:

- VP shunt placement to be discussed with a neurosurgeon.
- Nerve sheath fenestration to be discussed with a neuro-ophthalmologist (consider Dean McGee Eye Institute in Oklahoma City)

Pseudotumor is associated with some medicines (minicycline, excessive vitamin A) and obesity. Therefore, if the patient is on a medicine at time of diagnosis of pseudotumor, it is often discontinued if is safe to do so and the patient is strongly encouraged to begin an exercise and weight loss program.

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