

PSEUDOTUMOR CEREBRI

Pseudotumor cerebri is a poorly understood condition. The fluid that bathes and protects the brain and spinal cord (the *CSF* or *cerebrospinal fluid*) accumulates faster than it drains, causing the pressure of the CSF to rise. Normally, this pressure is less than 200 mm (or 20 cm) of water. In pseudotumor cerebri, it may rise into the 300's or even 500's. High CSF pressures often cause headaches, and in some individuals may cause visual disturbances or vision loss.

Pseudotumor cerebri is treated first with medications. Diamox and Lasix (diuretics) are commonly used. Some people also try steroids. If the medications do not help, or a patient is unable to take these medications, then surgery might be considered.

SURGERY

Reasons to consider surgery include:

- horrible headaches that can not be treated with medications
- vision loss
- threat of vision loss

Horrible headaches are best treated by a synthetic shunt (VP or LP shunts) implanted by a neurosurgeon. This is a tube that allows the CSF to drain into the belly. Vision loss may be treated by either a shunt or an optic nerve sheath fenestration (ONSF).

Optic Nerve Sheath Fenestration

The eyes are connected to the brain by nerves called optic nerves. The optic nerves are delicate structures that can be damaged by high CSF pressure. Optic nerve sheath fenestration (ONSF) is a microsurgical procedure developed to protect the optic nerves. Although quickly performed (about 1 hour), it requires considerable knowledge, skill, and experience.

The surgery is performed under general anesthesia through a skin incision less than an inch long. The incision is well hidden in the laugh lines at the corner of the eye. Through meticulous dissection, the optic nerve is identified, and a small window is cut in the covering over the nerve using microsurgical techniques. The CSF escapes through this window and is then absorbed in the fat that lies behind the eye. Some patients may be kept in the hospital overnight for observation, but many go home the same day.

With current techniques, it is extremely rare for the fenestration (window) to close. In most cases, only one nerve is operated on at a time. About 1/3 of patients require fenestration on only one side. This will become apparent within 6 weeks of the first surgery. Most patients regain vision with this surgery, some stabilize and lose no more; and only very rarely will vision loss progress after ONSF. As an added benefit, about 1/2 of patients stop having headaches. Following surgery, there will be some swelling for 1-3 weeks, depending upon each person's healing ability. Walking, reading, etc may be performed the day after surgery, but strenuous activity is discouraged for 7-10 days.

As with any surgery, there are risks. With ONSF, the risks are quite rare, but potentially serious. Infection (with spread to the brain), bleeding behind the eye, and partial or complete vision loss are all possible. Thankfully, we have never had any of these complications, but we always worry about them. Perhaps 1 in 5 patients may have a change in the shape or size of their pupil, a problem that is usually not a concern, but one that can be corrected with a special contact lens, if necessary. Some patients have disturbance of eye movement for a week or two, until the swelling is completely gone.

Given the fact that there are risks, when should ONSF be performed? Most physicians would agree that if there is clear evidence that vision is being lost, and a patient is taking as much medicine as they can, then an ONSF should be considered. Some physicians also argue that if there is no vision loss, but the CSF pressure is very high and the optic nerves (seen by someone looking into the eye) are swollen for many months despite medications, or there are hemorrhages (bleeding episodes) within the eye next to the nerve, then an ONSF should be considered before vision is lost. Thus, there is no simple answer as to when an ONSF should be performed, and in the end, the final decision must be made by each patient in an informed fashion.

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In your case, right now, our recommendation is:

- _____ Careful observation and medical management by your Ophthalmologist and other physicians.
- _____ Consideration of Optic Nerve Sheath Fenestration, as we have discussed during your visit.
- _____ Optic Nerve Sheath Fenestration
- _____ Neurosurgical shunting procedure