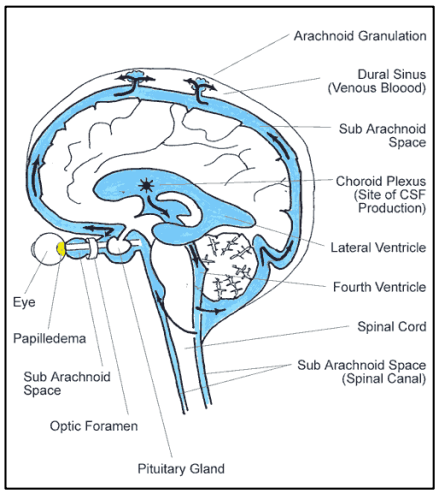
Pseudotumor Cerebri

Anesthetic Pearls: Anesthetic Implications and Management of Pseudotumor Cerebri



AKA: Idiopathic Intracranial Hypertension & Benign Intracranial Hypertension

Incidence:

A. 3.5 / 100,000 women 15-44 years.
B. 13 / 100,000 women 20-44 years and 10% above ideal body weight.
C. **19 / 100,000 women 20-44 years & > 20% above ideal body weight.**
D. Rare in patients > 45 years old, men, and slim adults.

Pathophysiology: Early theories believed that cerebral edema caused the increased ICP. However, there is no altered level of alertness, cognitive impairment or focal neurological findings (it may have been that the cerebral edema was secondary to tissue fixation rather than in vivo edema). Current theories include increased resistance to CSF outflow at the arachnoid granulations or that occult cerebral venous outflow abnormalities can cause it.

Mortality / Morbidity: There is no known specific mortality risk separate from the increased mortality of morbid obesity. The only permanent morbidity is vision loss from papilledema with progressive optic nerve atrophy. The frequency and degree to which visual loss occurs is difficult to establish (with current literature disagreeing at 22%-96% incidence).

Signs / Symptoms: primary presentation from elevated ICP and papilledema.

1. **Headache** – non-specific and varies in type, location, and frequency.
2. **Pulsatile tinnitus** – a rhythmic sound, heard in one or both ears synchronous with the pulse that may be exacerbated by the supine or bending position
3. **Horizontal diplopia** – a symptom of a false-localizing 6th cranial nerve palsy.
4. **Radicular pain** – a rare symptom that occurs in the arms (usually).
5. **Transient visual obscurations** – dimming or blackout of vision in one or both eyes lasting for a few seconds (may be predominately or uniformly orthostatic).
6. **Metamorphopsia** – blurring / distortion of central vision from macular wrinkling.
7. **Peripheral vision loss** – slowly progressive to total vision loss
8. **Total visual loss** (acute) from intraocular hemorrhage secondary to peripapillary subretinal neovascularization related to chronic papilledema.

Diagnosis: CBC, ESR, FE / TIBC, ELISA, ANA profile, Anticardiolipin antibodies / lupus anticoagulant, a full procoagulant profile (protein S, protein C, homocysteine levels, AT-3 levels). CSF studies (opening pressure, WBC count, RBC count, protein, AFB, RPR, Glucose level, tumor markers, cryptococcal antigen, and protein electrophoresis). MRI with Gadolinium for enhancement of hydrocephalus, tumors, meningeal infiltrative and / or inflammatory processes, and dural venous sinus thrombosis. MRV or CT scans can be used but not as sensitive.

Medical Treatment: Patients without visual loss are usually treated with a carbonic anhydrase inhibitor (Acetazolamide) to lower the ICP. Some authors also use Digoxin for the same effect (and it may have fewer side effects). Patients with severe symptoms (early visual field loss or failed standard medical therapy) with get a short course of high dose corticosteroids. Patients also are usually placed on weight-loss programs. There is some evidence that weight loss is associated with improvement of the papilledema.

Surgical Options: 1) CSF shunting procedure; or 2) optic nerve sheath fenestration. CSF is drained via lumbo-peritoneal (LP), ventriculo-peritoneal (VP), or ventriculo-atrial (VA) shunts. Optic nerve sheath fenestration involves cutting slits or rectangular patches in the dura surrounding the optic nerve immediately posterior to the globe. This allows CSF to drain into the orbital fat and then be recirculated. Note: Headache is not reliably relieved nor is the ICP lowered, but there may be regression of the papilledema in both eyes following fenestration of one optic nerve.

Drug interactions:

1. **Acetazolamide** – Reduces CSF production by ~50%, which leads to a reduction in the ICP (may not be sustained effect). Many patients develop side effects severe enough to hinder compliance (perioral / digital parasthesias, anorexia, N/V, and metallic taste). The drug is contraindicated in patients with severe renal disease, hepatic disease (may become comatose), adrenocortical insufficiency, or pulmonary obstruction. Therapeutic levels of Lithium may be decreased and can alter excretion of other drugs (Amphetamines, Phenobarbital, Salicylates) by alkalinizing urine. Rarely, patients may develop aplastic anemia.
2. **Digoxin** – May reduce CSF production up to 80% by inhibiting the Na^+-K^+ ATPase pump. The drug is contraindicated in patients with hypertrophic obstructive cardiomyopathy (HOCM / IHSS), dysrhythmias caused via accessory nerve tracts, and ventricular fibrillation. Digitalis toxicity may occur due to interactions with potassium-depleting diuretics or corticosteroids.
3. **Prednisone** – Mechanism of action to lower ICP is unknown. The drug is contraindicated in patients with PUD, hepatic dysfunction, and infections of connective tissue or fungal / tubercular skin infections. The hepatic metabolism is enhanced with Phenytoin, Phenobarbital, Ephedrine, and Rifampicin (drugs that augment the cytochrome P-450 system).

Other Complications:

A. **Post-Dural Puncture Headache** – may develop intractable low-tension headaches following lumbar puncture for diagnosis and may require a blood patch for alleviation.
B. **Hypotension** may lead to poor optic nerve perfusion if the ICP is elevated (keep in mind while Propofol is being rapidly pushed).