

## Guidelines for the Management of Idiopathic Intracranial Hypertension (Pseudotumor Cerebri Syndrome) in the Emergency Department.

Revised 10-23-17

**Concern:** Papilledema can lead to permanent visual loss, starting with decreased peripheral vision, to the point of legal blindness, despite preserved central vision (visual acuity).

### Guidelines:

1. If papilledema is suspected, check visual acuity, confrontation fields, and consult Ophthalmology.
2. Ophthalmology to assess visual function (best corrected visual acuity or pinhole acuity, Amsler grid, confrontation visual fields), and confirm and grade papilledema using the Frisen scale 0-5 (obtain fundus photos if technology available).

### 3. Imaging study:

In children: Avoid head CT or CTA, except in extenuating circumstances: instead obtain brain MRI/MRV, with and without IV contrast, including stealth post contrast sequence. If sedation or anesthesia is required try to get LP while the patient is still sedated. Weekend and evening emergent MRIs requiring anesthesia likely will require an attending to attending discussion. The attending ordering the study must board the MRI with the OR.

In adults: Ideally, obtain brain MRI/MRV with and without IV contrast, include stealth post contrast sequence. Alternatively, head CT, or CTA. If head CT done already, no need to get MRI unless the patient is atypical. If anesthesia or sedation is required, try to get LP while the patient is still sedated.

### 4. Lumbar puncture:

ED attending arranges LP.

If the opening pressure\* is >250 mm (>280 mm in sedated prepubescent children), lower by no more than 50%; send CSF for cells, protein, glucose, and infectious or neoplastic workup where appropriate.

Occasionally the CSF opening pressure\* may be normal, erroneously, in the presence of unequivocal optic disc edema. That should not deter from proceeding to treat (other options for diagnosis, at the discretion of the individual physician, include repeating LP, or ICP monitoring).

*Note: The effect of a single LP is transient and not therapeutic; if multiple LPs are required consider a lumbar drain.*

### 5. Indications for Admission:

#### A. For Urgent Surgery (or lumbar drain to temporize):

If ophthalmology determines significant visual loss in either eye (in the presence of Frisen scale 3-5 papilledema), i.e.: confrontation visual field constriction of less than 30° from fixation (using a small target such as a Q-tip), any encroachment on Amsler grid (central 20°), visual acuity 20/100 or worse, (or if available, reliable Humphrey visual fields with a deficit of 15 dB or worse), an obvious relative afferent pupillary defect (or light-near dissociation).

#### B. For Aggressive Medical Therapy and Observation:

If ophthalmology determines the following deficits in either eye: confrontation visual field constriction of less than 45° from fixation (using a small target such as a Q-tip), visual acuity of 20/50 or worse, or if available, a reliable Humphrey visual field deficit of -10 to 15 dB.

Admit adults to the General Neurology Service (unless surgery imminent)

Admit children to a Pediatric inpatient team, or Pediatric Neurology (**Fenichel Service**):

Medical Therapy:

Adults: IV acetazolamide 500 -1000 mg 6 hourly  
Steroids, if used at all, should be short-term only (if surgery imminent).

Prepubescent children: IV acetazolamide 25 mg/kg/d in 3-4 divided doses  
Maximum 100 mg/kg/ day, or 2 grams/day

Other options include neurosurgery consult for lumbar drain (while continuing medical therapy).

6. If there is progressive visual loss despite aggressive medical therapy → surgical intervention:  
If visual loss is the major issue → optic nerve sheath fenestration or shunt, at the discretion of the MD  
If catastrophic visual loss, consider both fenestration and shunt.  
If headache is the major issue → VP shunt
7. If vision is not compromised significantly, start oral acetazolamide (Diamox) provided there is no serious contraindication. Alternatives: other diuretics (e.g. methazolamide, furosemide, HCTZ, chlorthalidone, spironolactone, etc.) or topiramate.  
*Note: The effect of a single LP is transient and not therapeutic.*  
**Minimal starting dose of oral acetazolamide** (For adults, use 250 mg tablets):  
Adults: 500 mg bid, maximum 4 Gm/day<sup>1</sup>.  
Prepubescent children: 25 mg/kg/day, maximum 100 mg/kg/ day (or max 2 Gm/day)
8. Follow-up in neuro-ophthalmology clinic (children → pediatric ophthalmology clinic) within a week. The ophthalmology resident involved at discharge will send a message to the VEI access Center. Patient will need visual acuity, color vision, Humphrey visual fields 24–2, fundus photographs, BMI, and OCT at that visit.
9. Treat cause, e.g. cerebral/jugular venous sinus thrombosis, remove offending meds, BMI, OSA, etc.
10. Ophthalmology follow-up is imperative despite cause, and should include visual acuity, Humphrey visual fields 24–2, a dilated exam, fundus photographs/OCT, and further follow-up. The interval of visits will depend on the severity of the illness.
11. Neurology follow-up will depend on the cause, and the need for management of headaches.

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1. Increase acetazolamide as the situation dictates. For mild visual field loss the I2HT2 initiated 500 mg bid and increased it by 250 mg/d every week up to benefit or 4 g a day (the reference below can be used if the pharmacy balks at high doses).

Effect of Acetazolamide on Visual Function in Patients With Idiopathic Intracranial Hypertension and Mild Visual Loss. The Idiopathic Intracranial Hypertension Treatment Trial. JAMA 2014;311 (16): 1641-1651.