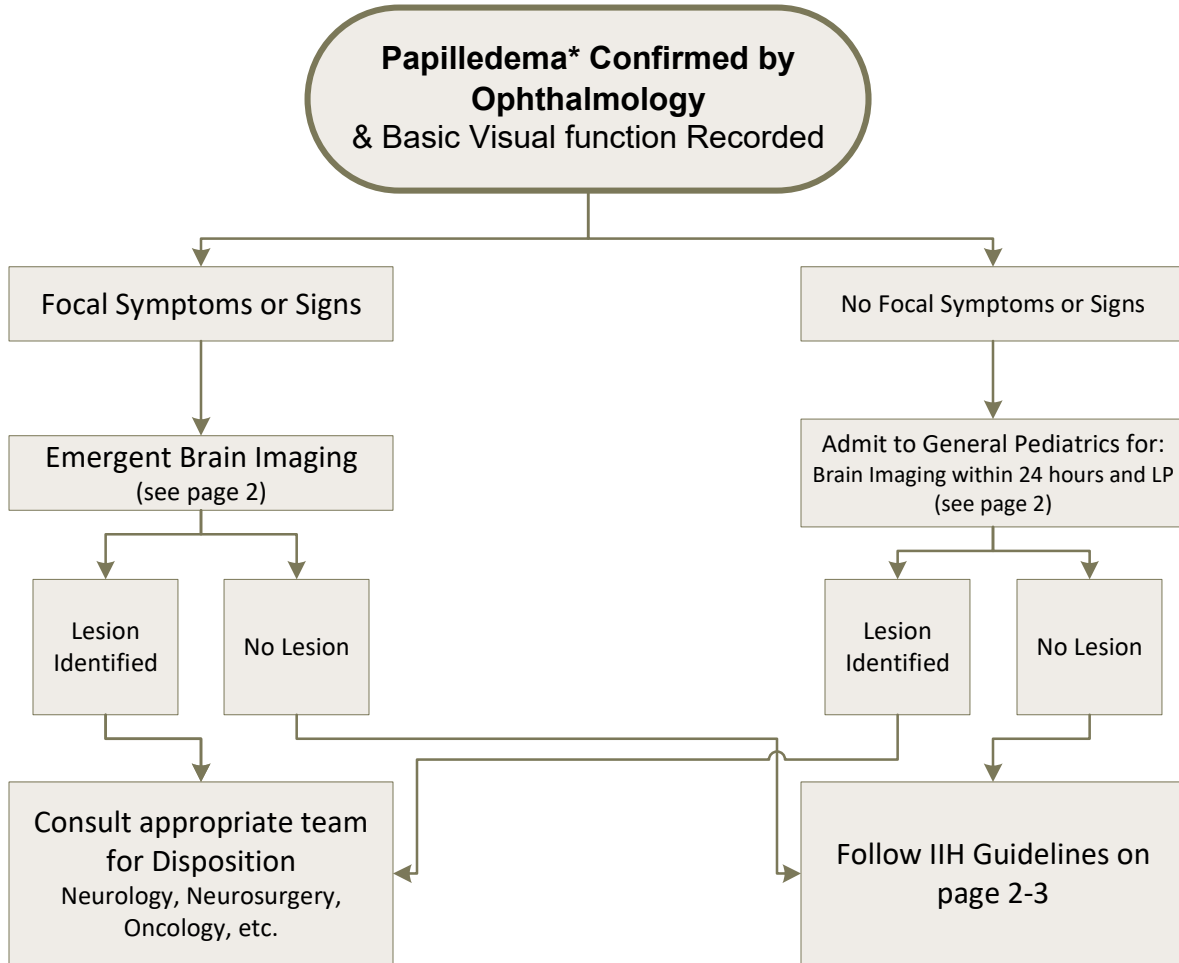


# IIH/Papilledema Guidelines for Pediatric Patients

Clinical Practice Guideline



## **\*\*Focal neurological symptoms and signs**

- Altered mental status
- Unilateral ocular findings
- Focal weakness or numbness
- Altered tone
- Gait abnormality
- Cranial nerve deficit, in addition to optic disc edema
- Abnormal vital signs
- New onset headache, worsening headache, or other headache red flags
- Vomiting not attributed to other causes

## Guidelines for the Management of Papilledema, including Idiopathic Intracranial Hypertension

Revised 12-14-21

### Guidelines:

1. If papilledema suspected and **there are focal neurological signs\*\*** → emergent imaging and Ophthalmology consult to confirm and assess for vision loss.

If papilledema suspected and **there are no focal neurologic signs\*\*** → Ophthalmology consult to confirm papilledema, and to assess visual function/vision loss.

2. Ophthalmology to assess visual function (best corrected, or pinhole, visual acuity, Amsler grid exam, confrontation visual fields), and grade papilledema using the modified Frisen scale 0-5. If the technology is available, obtain fundus photos and/or OCT.

3. Imaging study:

In children: Avoid CT head/CTV head, except in extenuating circumstances: Instead obtain MRI brain + orbits w/wo + MRV w/wo (includes 3D T1 post contrast sequences). 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.

4. Lumbar puncture:

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, erroneously, the CSF opening pressure may be normal 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 protractedly therapeutic; if multiple LPs are required consider a temporizing lumbar drain.*

5. Check BP to evaluate for accelerated hypertension; also evaluate for other causes of elevated intracranial pressure, e.g., anemia, BMI, cerebral/jugular vein thrombosis, OSA, offending meds, etc.

6. Indications for Admission:

#### A. For Aggressive Medical Therapy:

If ophthalmology determines moderate visual loss in either eye: e.g., 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 mean deficit (MD) of -7 to -15 dB.

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

Indications for surgery at the discretion of the physicians/services involved:

If the presentation is fulminant (rapid onset with progression in the last 3-4 weeks), or there is significant visual loss in either eye (in the presence of Frisen scale 3-5 papilledema) such as:

- i. Constricted confrontation visual field < 30° from fixation (using a small target such as a Q-tip) or, if available, reliable Humphrey visual fields with a mean deficit of >-15 dB.
- ii. Encroachment on Amsler grid (central 20°).
- iii. Significant central visual loss in the absence of an ocular cause such as globe flattening, macular edema, or macular exudates.

C. Admit children to a Pediatric inpatient team.

Medical Therapy:

Postpubescent: IV acetazolamide 500 -1000 mg 6 hourly

Steroids, if used at all, should be short-term only if surgery imminent.

Prepubescent: 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 temporizing lumbar drain (while continuing medical therapy).

7. If there is progressive visual loss despite maximally tolerated medical therapy surgical intervention:  
If visual loss is the major issue optic nerve sheath fenestration or shunt, at the discretion of the physicians/services involved.  
If catastrophic visual loss, consider both fenestration and shunt.
8. If vision is not compromised significantly (MD <-7), start oral acetazolamide (Diamox) provided there is no serious contraindication. Alternatives: topiramate, other diuretics (e.g., methazolamide, HCTZ, chlorthalidone, spironolactone, etc.).  
*Note: The effect of a single LP is transient and not protractedly therapeutic.*  
**Minimal starting dose of oral acetazolamide\*\*\*** (For adults, use 250 mg tablets):  
Adults: 500 mg bid (average maintenance dose 1-2 Gm/d, maximum 4 Gm/day\*\*\*\*).  
Prepubescent: 25 mg/kg/day, max 100 mg/kg/ day (or max 2 Gm/day in 3-4 divided doses)
9. Follow-up in pediatric ophthalmology clinic) within 1-4 weeks depending on severity. At discharge the ophthalmology resident involved will send a message to the VEI access Center. The patient will need visual acuity, color vision, Humphrey visual fields 24–2, fundus photographs, BMI, and OCT at that visit.
10. Follow-up in the ophthalmology is imperative despite cause, to monitor vision and optic disc changes. The interval of follow-up visits will depend on the severity of the illness and progress.
11. Neurology follow-up will depend on the cause, and the need for management of headaches.

\* **Concerns:** Papilledema (bilateral optic disc edema due to elevated intracranial pressure) can lead to permanent blindness. Note: Visual acuity of 20/200, or visual field less than 20° despite preserved central vision = legal blindness.

\*\* **Focal neurological symptoms and signs:**

Altered mental status  
Unilateral ocular findings  
Focal weakness or numbness  
Altered tone  
Gait abnormality  
Cranial nerve deficit in addition to optic disc edema  
Abnormal vital signs  
New onset headache, worsening headache or other red flags for headache  
Vomiting not attributed to other causes

\*\*\* Increase acetazolamide as the situation dictates. For mild visual field loss, the I<sup>2</sup>HT<sup>2</sup> initiated 500 mg bid and increase 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.