Pediatric Idiopathic Intracranial Hypertension (IIH)

Pediatric IIH Definition and Causes

Proposed Criteria for Diagnosis (Prepubertal)

1. General Signs and symptoms of increased intracranial pressure or Optic disc edema
2. Elevated CSF pressure in lateral decubitus position
    1. 250 mm H2O in age Age 8 or above
    2. 250 mm H20 < Age 8 without disc edema
    3. 180 mm H2O < Age 8 with papilledema
    4. > 76 mm H2O in Neonates
3. Normal CSF composition except in neonates (up to 32 WBC/mm3 and protein up to 150 mg/dl)
4. no evidence of hydrocephalus, mass, structural or vascular lesion on MRI & MRV- narrowing of the transverse sinus is allowed
5. Reversible cranial nerve palsies

Associations

1. Growth hormone replacement (recombinant and synthetic)
2. Addison’s disease Wikipedia
3. Steroid withdrawal
4. Thyroxine replacement (dose change) or hypothyroid state
5. Levongestrel implant
6. Desmopressin nasal spray
7. Cytarabine
8. Retinoid acid
9. Acute Sinusitis
10. Varicella
11. Miller Fisher Syndrome (Guillan-Barré) Wikipedia

Clinical Guidelines

CHOP Clinical Pathway

Treatment

- Diamox (Acetazolamide) for Intracranial Hypertension

- Furosemide for Pediatric Patients

Andy Lee

- Starting dose: 25 mg/kg per day with a maximum dose of 100 mg/kg or 2 g per day.
- The sustained release formulation (Diamox sequels) may be better tolerated by patients who are intolerant of generic acetazolamide, but are substantially more expensive

Grant Liu

Acetazolamide and furosemide are the drugs most often used in the medical management of pediatric IIH. Acetazolamide, a carbonic anhydrase inhibitor, is thought to reduce the rate of cerebrospinal fluid production, and it is generally the first-line treatment choice in patients with IIH. In children, we usually use an oral dose of 15 mg/kg/day in two to three divided doses, until headache, disk swelling, and visual field abnormalities resolve-typically in 3 to 9 months. Many pharmacies can prepare a syrup formulation for younger children. Common doserelated side effects include GI upset; paresthesias involving the lips, fingers, and toes; anorexia; and electrolyte imbalance (metabolic acidosis). Kidney stones are rare, and aplastic anemia is exceedingly uncommon. We do not monitor electrolytes as children are usually asymptomatic from the acidosis. When the side effects become intolerable, the dose is lowered, or acetazolamide is replaced with furosemide 0.3-0.6 mg/kg per day. There are reports of combining acetazolamide with furosemide to produce additive results and reduce pressure more effectively than just acetazolamide alone. If acetazolamide or furosemide fail, topiramate (1.5-3.0 mg/kg per day in two divided doses, and no more than 200 mg/day), may be used, particularly when the child is obese. Topiramate is an anti epileptic medication with secondary carbonic anhydrase activity. The use of this medication in IIH is relatively new, and it is unclear whether it is superior to acetazolamide in reducing CSF pressure. However, topiramate has the added benefit of appetite suppression and weight loss in many patients, it is excellent for treatment of chronic daily headache, and it has been used safely for years in children with epilepsy. The dosage should be built up slowly over weeks (25 mg/week) to reduce the risk of cognitive side effects, which are more likely to occur with rapid dose escalations and at doses higher than 200 mg/day. Zonisamide, another drug with secondary carbonic anhydrase activity and appetite suppression, may be used in similar doses if the side effects of topiramate are not tolerated. In acute situations when visual loss is severe, the combination of oral acetazolamide and IV methylprednisolone 15 mg/kg can be used when surgery is not immediately available. The use of chronic steroids, however, should be avoided. - Surv Ophthalmol 52:597-617, 2007.

References

- Lubaina M. Rangwala, MD, and Grant T. Liu, MD. Pediatric Idiopathic Intracranial Hypertension. Survey of Ophthalmology 2007;55(6):597-617
- Peds In Review

- UpToDate

IIH, drugs