DB

- 6 year old boy with severe headache
- BHX- 6lb 9 oz
  - Maternal hypertension, stress, and four seizures
- Normal Development
- Lives with Grandmother
• Surgery – T&A 2010

• ROS- Rickets, allergies, eczema, asthma

• Meds- albuteral, Singulair, Flovent, Nasonex, Ventolin, Vit D, Calcium, K-Phos-Naproxen, lortab
• FHX- Headaches – several family members, Vit D resistant rickets and HBP (maternal), SSD (paternal)
“My head won't stop hurting”

- **Headaches**
  - Began age 4 ½ but Worse since 2010
- **Location?**
  - All over and not able to describe
- **Severity?**
  - Severe – cries and leaves school
- **Frequency?**
- **5-6X/week and Vomits 1-2X/week**
DB

- **Time?**
  - Primarily around noon
- **Better?**
  - Dark room, lying down +/- lortab
- **Precipitants?**
  - Unclear
- **Tried multiple medications**
  - Exact ones unclear
Examination

• Lying in a dark room
• Flat affect, tired appearing wanting to be in his Grandmother’s arms

• Gen PE normal except bowing of legs and in-toeing
• CN normal- Fundi benign and disc sharp
• Motor, sensory, coordination examination normal
Next Step?

• Medications?
  – Begun on Periactin
    • Some minor help
    • Too sleepy
  – Added Topamax
  – Added Zofran
Next Step?

- Contact Endocrine
- Ophthalmology
  - Dr Sessums
  - Normal optic nerves- “No eye etiology of headaches”-Plan “Back to you”
- Imaging?
  - MRI/MRA/MRV normal
Next Step?

- April 2013-Spinal tap
  - OP=37-38 cm water
  - CP= 20.5 cm water
  - 16 cc removed

- Begun on Diamox 250 bid

- DRAMATIC IMPROVEMENT
Pediatric Pseudotumor Cerebri
From Diagnosis to Treatment
Historical Timeline

- First described by Quincke in 1897
  - Called “meningits serosa”
  - Otitic hydrocephalus
- Nonne coined pseudotumor cerebri in 1904
- Benign intracranial hypertension in 1955
- Idiopathic Intracranial Hypertension (IIH)
What are the Criteria?

- Increased intracranial pressure (>25 cm of water)
- Nonfocal neurologic exam (except CN)
- Unremarkable CSF analysis
- No identifiable cause
What Causes it?

• Consensus = result of impaired CSF absorption

Two schools of thought:
1. Reduced CSF absorption at arachnoid villi
2. Increased CSF flow resistance from raised venous pressure from outflow obstruction
Presentation

- Daily pulsatile frontal headache
- Worse lying down and or in am
- Worse with cough
- Tinnitus (whooshing sound)
- Increased HA with eye movements
- Blurred vision or transient obscurations
- Neck stiffness, vertigo, nausea & vomiting
Demographics

- Ask a medical student:
  - Obese
  - Middle age
  - Woman
  - Symptoms of headache and blurred vision
Demographics

- Actual statistics vary: annual incidence in US 0.9 per 100,000
  - More common in females (3.5/100,000)
  - In obese females (12.19/100,000)
- Female : Male 4:1
- Now seen more in younger patients

Obesity or recognition??
Signs and Symptoms

• Presentation:
First Task: rule out other lesions

Requires exclusion of tumor, hydrocephalus, SAH, etc
Secondary IIH: Dural Sinus Thrombosis

- Symptoms due to ICP
- Transverse sinus is most common
- Risk factors: mastoiditis, coagulopathy, post-partum
Risk Factors: Medications

- Growth hormone
- Corticosteroids
- Minocycline
- Retinoic acid
- Vitamin A
- Vitamin D
- Oral contraceptives
- Nalidixic acid
- Cyclosporin
- Cytarabine
- Lithium
- Tetracycline
Risk Factors: Systemic Issues

- Obesity
- Post-malnutrition
- Sleep apnea
- Down syndrome
- Adrenal insufficiency
- Thyrotoxicosis
- CHF
- Anemia
- Lead
Signs and Symptoms: When to suspect it

- Headache (am)
- Pain in neck and back
- Tinnitus ("Whooshing")
- Cranial neuropathy
- Visual symptoms
- Focal sign suggest alternate diagnosis
- Optic Disc edema
Mimickers of Papilledema

- No disk edema or intracranial ICP
  - Anomalous nerves
  - Drusen bodies
- Disk edema without ICP
  - Optic neuritis
  - Anterior ischemic optic neuropathy
  - Malignant cell infiltrate
Mimickers of Papilledema
Ophthalmologic Signs and Symptoms

- Blurred vision
- Diplopia, not always due to VI palsy
- Transient visual obscurations
- Peripheral field loss is often asymmetric
- Complete blindness
Visual Field Loss

- Peripheral visual field constriction due to increased ICP
- Macular vision is spared initially
Evaluation and Treatment

• Laboratory
  – CBC, CMP
  – Thyroid panel

• IF clinically supported:
  – Thrombotic profiles, ESR, ACE, Vitamin A and D levels, and ANA
Imaging

- All patients require imaging before LP

- HCT and/or MRI
  - MRI focus on orbits, MRA/MRV
  - Usually Normal—other findings can include:
    - Lateral venous stenosis (due to ICP), empty sella, slit ventricles, flattening of post globe, optic nerve edema
Lumbar Puncture

- Document OP and CP and volume removed
  - 10-15 cc
- Evaluate the CSF
- Culture
  - Viral cultures- arbo, EBV, CMV, HSV, lyme PCR if warranted
IC Pressure Norms

- **Adult <20 cm water**
  - For diagnosis > 25cm of water
- **Pediatric patients**
  - > 8 years use adult guidelines
  - < 8yr < 18 cm water
Treatment
Medical Management

• Weight loss
  - Studies have shown that except for discontinuing medications, weight loss is the MOST important
Acetazolamide

- Acts via carbonic anhydrase inhibitor
  - Lowers sodium transport across the choroid epithelium
- Children starting dose 10-25 mg/kg/day divided BID
- Adolescents 0.5-1 g/day divided BID
- Target 1-2g divided BID
- Caution if Sulfa allergy
Acetazolamide

• Side effects
  – Metallic taste
  – Transient anorexia
  – Paresthesias
  – Kidney stones
  – Metabolic acidosis
    • Check in 4-6 weeks
  – Aplastic anemia reported
Furosemide

• Works by increasing diuresis and reducing sodium transport
• 1-2 mg/k/day BID or TID
• Adolescent and adults 40-120 mg BID or TID
• Watch electrolytes and K supplement

• May be synergistic with acetazolamide
Topiramate

- Weak carbonic anhydrase inhibitor
- Same side effects as acetazolamide
- Also possible
  - Anhydrosis
  - Mild word finding problems
  - Appetite suppressant
Corticosteroids

- Not routinely used
- Can be used to STABILIZE rapid vision loss
- IV methylprednisolone at 15 – 20 mg/kg/d
  - Then PO prednisone 2 mg/k/d for 2 weeks and then taper over 2 weeks
Surgical Interventions

• Optic nerve fenestration or CSF diversion
• Considered if acute or progress vision loss
• Concern:
  – Optic nerve head is at a watershed so increased risk of ischemia with ONF
  – Use steroids and diversion
Optic nerve sheath fenestration

- Decreases pressure on nerve
- Series of slices along the sheath
- How it works?
  - Thought to be related to scarring of arachnoid
  - Can work if done unilaterally
- 4 weeks to resolve papilledema
Optic nerve sheath fenestration

- Via lateral orbitotomy approach
- Window of dura and arachnoid made
- Arachnoid excised
- CSF allowed to drain
CSF Diversion

- Lumbar and ventricular peritoneal shunts
- Primarily addresses the pain
- Resolves papilledema in 3 months
- Headache resolves sooner
Outcomes

- Up to 20% will already have some vision loss at diagnosis
- Permanent in only 10%
- Resolution of papilledema in 3-6mo
- Visual outcome better if pre pubertal

- Recurrence in 10-20%
  - Most common if no weight loss
- Common to have migraine
Take Home Points

- Obesity and gender are less important in children
- R/O Venous thrombosis
- ICP can be increased without papilledema
- ICP can be increased without symptoms
- Transient IIH is common in children
- Peripheral visual loss occurs first