The Swollen Optic Disc

As a manifestation of systemic disease in the pediatric patient

- I have no financial interests to disclose.

3 common causes of bilateral disc swelling in children
- Idiopathic Intracranial Hypertension
- Optic Neuritis
- Pseudopapilledema
- Other assorted....

Case #1
- 7 year old white female otherwise healthy presents to pediatrician with complaints of several week hx of double vision and headache
  - Afebrile, without other constitutional symptoms
  - No hx of trauma
  - ER doctor had seen abnormal optic discs with direct ophthalmoscopy - CT head negative

Exam
- Visual acuity: 20/20 OU
- Pupils – normal
- Motility – esotropia
- Anterior segment - normal

- CT head urgent – negative
- LP – opening pressure 36 cm H2O
  - protein, glucose, cell count - normal
Idiopathic Intracranial Hypertension

- Symptoms and signs if increased ICP without evidence of a mass or hydrocephalus
- Diagnosis of Primary IIH is established when the modified Dandy criteria are met.

Modified Dandy Criteria

- Signs and symptoms of ↑ICP
- Absence of localizing findings on neurologic exam
- Normal CT/MRI except for signs of ↑ICP
- Alert and oriented patient
- Normal CSF indices
- No other cause of increased ICP present

Symptoms

- Headache (50%)
- Lateral rectus paresis (esotropia)
- Transient visual obscurations on changing position (black outs, sparkles)
- Tinnitus/ear pain
- Once vision loss is recognized, it can progress quickly

Ophthalmic Signs

- Swollen optic disc(s)
- Loss of spontaneous venous pulsations at the disc
- Enlarged blind spot on visual field testing
- Accommodative spasm
- Loss of visual acuity and color vision

Tools to monitor visual function in children:

- Visual acuity (snellen, Allen figures, Teller acuity cards)
- Color vision plates
- Visual fields (automated, manual)
Secondary Pseudotumor Cerebri

- Neurologic disease (otic hydrocephalus, infectious/post infectious meningitis, Chiari malformation, subdural heme)
- Drug-related (steroid withdrawal, antibiotics, retinoids)
- Dural sinus stenosis/thrombosis
- Addison’s disease
- Sleep apnea
- post ECMO

Treatment (primary IIH)

- Medical - up to a year duration for visual loss pts
  - Topamax
  - Diamox
  - Steroids
- Surgical
  - Optic Nerve Sheath Fenestration
    - shunts

How do we follow disc swelling in IIH? Resolution or progression?

- Visual function is most important
  - Visual fields
- Appearance:
  - Fluorescein angiography
  - OCT

Case #2

- 10 yr old white male with 3 day hx of pain around the left eye when moving it and decreased vision OS. Found to have 20/100 vision left eye at pediatrician with optic disc “blurring” and hemorrhage by direct ophthalmoscopy
  - No recent illness
  - Negative past medical HX
Exam
- Vision: OD 20/30 OS 20/100
- Color vision: OD – normal OS – abnormal
- Slit lamp exam: normal OU

Optic Neuritis in Children
And... Neuro Retinitis

Optic Neuritis
- Inflammation of the optic nerve
  - Anterior (visible optic disc and 1 mm retrobulbar disc)
    - Optic disc is visibly swollen
  - Posterior (retrobulbar) – from 1 mm behind globe to optic chiasm
    - Optic disc appears normal acutely
    - Chiasmal neuritis – inflammation of optic chiasm seen on MRI

Optic Neuritis - Symptoms
- Progressive Loss of Vision over 2 days to 1 week
  - Central acuity
  - Color vision
  - Sound induced lights/colors (photisms)
- Pain around eye with eye movement
  - Annulus of Zinn
    - arrangement of EOM and optic nerve at orbital apex
Exam

• Decreased visual acuity
  ▫ Often as bad as “light perception” or NLP
• Decreased red/green color vision can be first
• Marcus Gunn pupil if unilateral or asymmetric
• Visual field
  ▫ Central scotoma/blind spot
• Optic disc: swollen or normal

Papilledema

• Use the term only if it is in reference to swollen discs caused by increased intracranial pressure
• May or may not be increased ICP, so call this “disc swelling”

Differences between Child and Adult optic neuritis

• Peds – 75% bilateral; Adults- 75% unilateral
• Peds – 75% swollen discs; Adults – 75% retrobulbar (normal disc appearance)
• Peds – mostly post infectious
  Adults – more common idiopathic or harbinger of multiple sclerosis

Etiology

• Post infectious/infectious
  ▫ Rubeola, paramyxovirus, varicella zoster, pertussis, lyme, Epstein-Barr, bartonella hensalae, syphilis toxocara, toxoplasmosis, TB, Rickettsia, brucella, influenza
• Post vaccination
  ▫ Live or attenuated virus (polio, smallpox, rubella, influenza)
  ▫ Reported after Anthrax vaccine
• Post insect bite
  ▫ Bee sting
  ▫ Spider bite

• All post infectious/inflammatory causes are felt to represent a localized form of encephalitis
  (Hence,... more common bilaterally)
Optic Neuritis + other neurologic signs
- Acute Disseminated Encephalomyelitis
- Devic’s Disease (neuromyelitis optica)
- Multiple Sclerosis

Acute Disseminated Encephalomyelitis
- Uncommon inflammatory demyelinating disease of the CNS that usually follows a viral illness or vaccination by days to weeks
  - children 6-10
  - Acute onset of motor signs, seizure activity, and sometimes altered consciousness and ataxia
  - Gray and white matter lesions on MRI
- Can have concurrent optic neuritis

Devic’s Disease (NMO)
- Severe, usually bilateral optic neuritis
  - often permanent visual loss
- Progressive ascending sensorimotor myelitis affecting lower or upper and lower limbs
  - Transverse lesion of the cord (2 segments affected)
- CSF shows pleocytosis and increased total protein
- NMO antibody (serum and/or CSF) – antibody to aquaporin 4 water channels
- Tx: steroids initially → immune-modulating agents

MS and Pediatric Optic Neuritis = Controversy
- No large multicentered trial like ONTT (Optic Neuritis Treatment Trial)
  - Pediatric Optic Neuritis Treatment Trial pending
    - North American NeuroOphthalmology Society
    - A lot of pediatric treatment is extrapolated from ONTT
  - ONTT
    - Multicentered trial with 454 patients; 15 yr follow up
      - Placebo vs oral steroids vs IV steroids in patient with first case of optic neuritis

Things we think we know
- Kids more often have bilateral involvement with anterior optic disc swelling following antecedent viral illness
- Visual prognosis poorer than that of adults, but development of MS is less common
  - 17% vs 50% at 2 years
  - MRI lesions were predictive of development of MS in both children and adults
- Children who do present with unilateral involvement have a better visual prognosis, but develop MS at a greater rate than bilateral.

Course of Disease
- Initial vision loss often profound
- 33% recurrence rate in children
- Prognosis for visual recovery is good (>75%)
  - Improvement begins after 3rd week and lasts for up to 6 months
- Optic atrophy is sequelae.
Work up

- MRI (brain and orbits with GAD)
  - Look for optic nerve hyperintensity on Flair and T2 and enhancement
  - Look for brain lesions (plaques)

ONTT
- @ 15 years:
  - 25% of pts with NO MRI lesions developed MS
  - 72% of pts with 1 or more brain lesion developed MS

- Lumbar Puncture
  - opening pressure
  - Cell count, culture, glucose, protein, cytology, Oligoclonal bands, myelin basic protein

- CBC, serologies (see infectious cause list)
  - CBC usually normal in isolated optic neuritis

- Chest xray (sarcoidosis, TB)

Remember drugs:

Etanercept (Enbrel – TNF blocker)
Infliximab (Remicaid – TNF antibody)

Both have been known to cause optic neuritis.

(JIA)

Treatment

- No controlled studies; most recommendations are largely anecdotal

- ONTT:
  - Neither oral or IV steroids changed the visual outcome measured at 1 yr after onset
  - IV steroids gave a more rapid recovery of vision
  - Oral steroids alone had 2x the number of recurrent demyelinating events in the 2 yr following onset then placebo
  - IV steroids gave ½ the number of demyelinating events in 2 yr period as oral or placebo

Case # 3

- 12 yr old female referred from optometrist for “swollen optic discs” and visual field defect

  - Found on routine exam for glasses after reports of transient vision loss on hiking trip

  - Otherwise negative past medical hx

  - No trauma
Pseudopapilledema - Optic Disc Drusen

Optic Disc Drusen (German - ‘crystal-filled space in rock’)

- Yellow globular concretions situated deep in the optic disc
  - Take up calcium salts
  - Grow forward into the visible disc with time
  - “ball under the rug” elevation of disc
- Progressive process causing vision loss over time

Systemic Associations

- Retinitis pigmentosa
- Pseudoxanthoma elasticum
  - 30 times greater incidence of disc drusen
- Migraine headaches
- Growth Hormone deficiency
  - GH also causes true papilledema

Also Associated with Pseudopapilledema:

- Down Syndrome
  - Anomalous discs are common
  - IH is more common
  - Myelodysplastic disease can cause infiltration and thus swelling
    (Presents a diagnostic conundrum. Often undergo imaging and LP to confirm diagnosis.)

- Alagille Syndrome