You Spin Me Right Round, Baby

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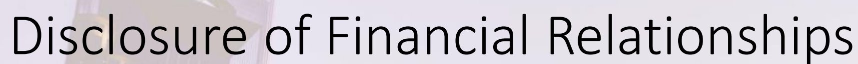
Disclosure of Financial Relationships



- No financial relationships to disclose relating to this presentation

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History of Present Illness

- 16 year old male with APECED (autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy)
- Presents with intermittent episodes of vertigo, dizziness, headaches, blurry vision, that have increased in frequency over the last 5 months
- Episodes range from 1 day to 2 weeks and are self-resolving
- Mom has to help him ambulate during episodes due to unsteady gait
- Have presented symptoms to many specialists over the last few months with no answers

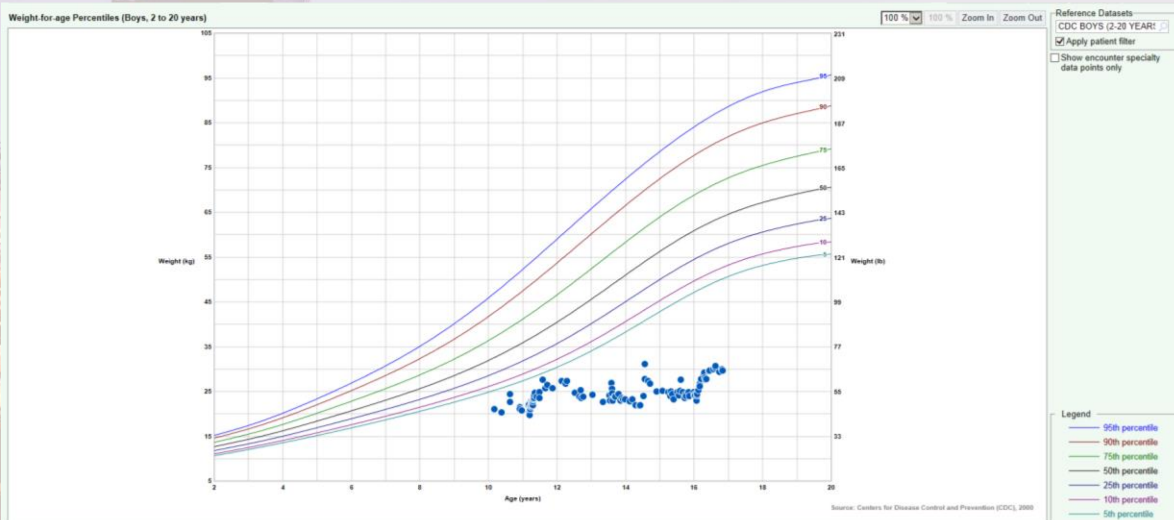
Past Medical History

- As a result of his APECED, he has:
  - Chronic autoimmune hepatitis
  - Chronic pancreatitis
  - Enteritis: has a g-tube to maximize his nutrition
  - Failure to thrive
  - Chronic kidney disease - stage II
  - Nephrolithiasis
  - Cytopenias of unknown etiology
  - Prolonged QTc
  - JRA
- On multiple medications including tacrolimus, ursodiol, and prednisone
- Previously on growth hormone

Physical Exam

- T: 98.1, BP 110/80, HR 71, RR 16, O2 97%, Wt 22.5kg
- No orthostatic hypotension
- General: small for age, chronically malnourished, otherwise comfortable
- Neuro:
    - Mentation intact
    - Dizziness reported with standing
    - Unsteady, shuffling, wide based gait
    - Cranial nerves were intact
    - No nystagmus or papilledema
    - Normal strength and reflexes in extremities

Growth Chart



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Initial Differential Diagnosis?

A) Vestibular
B) Cardiac arrhythmia
C) Medication side effect
D) Infection: ADEM, encephalitis
E) Intracranial process

Initial Differential Diagnosis

- Vestibular etiology
- Vasovagal
- Arrhythmia
- Medication side effects
- Intracranial mass
- Sinus venous thrombosis
- Encephalitis
- Acute disseminated encephalomyelitis

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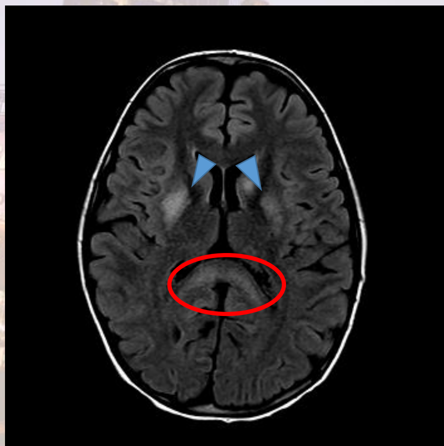
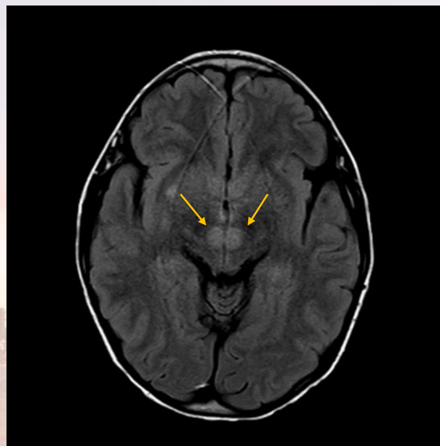
Hospital Course

- Admitted to the acute care floor
- Remained stable, at his baseline
- Symptoms improved by morning, but parental concerns persisted due to chronic, intermittent nature of symptoms
- Consult – Ophthalmology, Neurology, ENT
- MRI brain

New information/Clinical Course





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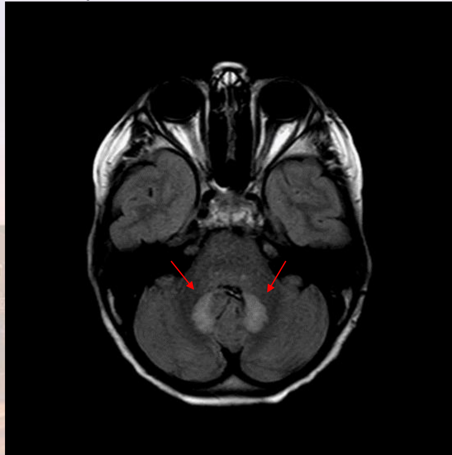
New information/Clinical Course



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Diagnostic Pause

- Differential based on imaging alone:
  - Vascular etiology (vasculitis, PRES)
  - Infection (viral > bacterial)
  - Inflammatory (ADEM)
  - Drugs/toxins (lead, cyanide, carbon monoxide)
  - Non-inherited metabolic (Wernicke's encephalopathy, hepatic or renal failure)
  - Mitochondrial disorders

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Final diagnosis?

A) Vascular etiology (vasculitis, PRES)

B) Infection (viral > bacterial)

C) Inflammatory (ADEM)

D) Drugs/toxins (lead, cyanide, carbon monoxide)

E) Non-inherited metabolic (Wernicke's encephalopathy, hepatic or renal failure)

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Final Diagnosis – Wernicke's Encephalopathy

- Rare disorder in the pediatric population
- Classic triad – confusion, ophthalmoplegia, ataxia
- Majority of WE cases in pediatrics is in oncology patients, followed by those who receive prolonged parenteral nutrition, bariatric surgery, or those with eating disorders

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Hospital Course

- Thiamine level was drawn (level was 60 nmol/L – normal is 70-180nmol/L)
- Patient was empirically started on IV thiamine immediately
- Symptoms were likely due to his long standing history of autoimmune enteritis and malabsorption
- Drastic improvement in symptoms

Wrap Up – Pathophysiology

- Thiamine is absorbed in the duodenum and proximal jejunum
- Majority of patients do not present with class triad
    - Ataxia or mental status changes
    - Ocular findings
- Important to check for other vitamin level deficiencies
- Glucose metabolism relies on thiamine

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Wrap Up – Final Thoughts

- Challenges in diagnosis
  - Complex medical history
  - Non-specific complaints
  - Intermittent, self-resolving nature of symptoms
- Nutrition status is often overshadowed by complex medical conditions
  - Integral to clinical status
  - Lack of exposure to nutritional deficiencies results in failed recognition

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