What to do with the fainting or dizzy child?

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Conflict of Interest

• I have no conflicts of interest.

What I want to get through today…

• Define syncope and its causes.
• Define POTS and its causes.
• Treatment options
• Discuss what can be done in primary care to evaluate, diagnose and treat.
• When to refer and what I do.
Syncope

- Syncope comes from Greek term “to cut short” or “interrupt”
- All of below have to be fulfilled:
  - Loss of Consciousness
  - Loss voluntary muscle tone
  - Relatively rapid onset (may have 10-20 sec premonitory symptoms or none at all)
  - Recovery spontaneous and usually prompt
  - Underlying cause: transient global cerebral hypoperfusion

Why do we care about syncope?

- Presence and severity of structural heart disease is mortality predictor
- Bad things can happen when people pass out: Concussions, cardiac dysrhythmias, seizures, SCD, etc
- Common event in our patients’ lives - neurally mediated syncope 35% by age 18 and 50% by age 21

(Paris, et. al JAHA. 2015. 1-13)

Etiology breakdown of syncope

Pathophysiology: What happens when we stand...

- Change in posture: 500-800ml blood trapped in veins below heart, plasma out to interstitial fluid and venous return and CO and BP
- Any given moment 5% body’s blood in capillaries, 8% in heart, 12% pulmonary vasculature, 15% arterial system and 60% venous (Problem with any of these can cause orthostatic challenges - systemic hypotension, cerebral hypoperfusion - syncope)
- HR 10-15 bpm, little change in systolic BP
- Continued standing activates neurohormonal changes (depends on volume state)
- Volume depleted activates RAAS

NM Syncope

- Systemic arterial pressure and cerebral perfusion pressure drops if either CO or PVR drops: syncope

- Excess vasodilation main cause of syncope

- Inability to increase vascular resistance during standing principle cause of syncope in orthostatic hypotension

- Abnormality in peripheral veins (increase venous pooling)

- Skeletal muscle tone

- Situational etiology: combo of preload, vagal activity

The Big Picture

Nervous System- Dysautonomia can affect...
Neurally Mediated Syncope

Neurally mediated (AKA- neurocardiogenic, situational, reflex syncope, simple faint or vasovagal faint)

• Most frequent of all syncope
• Types of NMS: central (emotional reaction), Postural (most common), situational (hair brushing, defecation, postprandial, micturition)

Subset of NMS

Orthostatic Hypotension: inability to maintain arterial BP when standing up or impaired capacity of sympathetic nerves to increase vascular resistance

• decline in BP of at least 20mmHg systolic or 10mmHg diastolic in three minutes of standing
• Medication cause of:
  Vasodilators and diuretics

Orthostatic Hypotension:

• decline in BP of at least 20mmHg systolic or 10mmHg diastolic in three minutes of standing
• Medication cause of: Vasodilators and diuretics

Postural Orthostatic Tachycardia Syndrome (POTS)

- chronic symptoms of orthostatic intolerance (>6 months), >40 pt HR increase with change in position within 10" standing, no or minimal BP change in children (>30 pt HR increase in adults)
- lying, sitting, standing BP/HR and standing at 5" and 10" BP/HR test evaluates pt for POTS

4 Types of POTS:

"Most pediatric patients with POTS have a syndrome rather than a disease, in which no etiology is found in spite of extensive diagnostic testing" – Imad Jarjour

- Hypovolemic and deconditioned: studies have found POTS pts to have hypovolemia common (30%), postural swelling/edema common, chronic fatigue and fibromyalgia type symptoms with decreased exercise common
  - Causes: preceding viral illness, trauma or surgery reducing activity and DI symptoms with marked somatic hypervigilance
- Hyper adrenergic: sympathetic activation symptoms, >10 mmHg systolic BP within 10" standing, HR often >120 bpm
  - Causes: NE transporter def, pheochromocytoma, mast cell activation disorders, and baroreflex failure
- POTS with JHS: use Beighton score, check family history (EDS pts also can have just general dysautonomia- not all have POTS)
- Neuropathic: partial distal autonomic neuropathy (especially distal legs), symptoms involve DI, GI dyssynergia, bladder retention and anhidrosis (Rare in Kids)
  - Causes: autoimmunity

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Symptoms of Orthostatic Intolerance/POTS:

**The Numbers**
- Female: Male 4:1
- Ratio with POTS
- Age 12-40
- Diagnosis often delayed by 2 years

**Orthostatic Symptoms**
- Dizziness and lightheadedness
- Near faint
- Blurred vision
- “Whirlpool” of vision
- Weakness in legs
- Poor concentration
- Headache
- Nausea

**Sympathetic Overactivation**
- Palpitations
- Chest pain
- Migraine
- Tremulousness
- Anxiety
- Pallor
- Exercise intolerance

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What can be done in primary care...

**Take the History:**
1. Ask the lifestyle ?’s: diet, hydration, sleep, headaches, school: grades, attendance, home life: drama, friends, activities involved in, menstrual hx.
2. Family: SCD, ❤ rhythms, seizure, stroke, syncope, connective tissue
3. Symptoms with syncope: dizziness, vision changes, palpitations, chest pain, shortness of breath, headaches, concussions

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**Physical Exam in Primary Care**

- Vital signs
- Cardiac: murmurs, heart rate, chest pain reproducible, pulses
- Beighton scale:
- Neuro eval

Testing in Primary Care

- Cardiac: EKG, Holter Monitor, or Event Monitor, Echocardiogram
- Neurally Mediated: Orthostatic BP/HR (lying, sitting, standing, standing 5 and 10")
- Labs: CBC, BMP, Fe, Fe Sat, Ferritin, Transferrin, Soluble Transferrin, Transferrin receptor, TSH, Urine (24 hour urine <100 common POTS)
- EEG?/MRI?
- CT- Concussion concern

Treatment in Primary Care

Neurally Mediated
Lifestyle recommendations:
1. Hydration: 64 ounces of water or low calorie electrolyte solution (G2 or Powerade Zero)
2. Salt: increase salt (2,000mg or greater)- caveat can’t have hypertension
3. Activity: aerobic (jogging, swimming, rowing machine), and anaerobic (planks, squats, sit ups)
4. Sleep: sleep hygiene talk, melatonin
5. Iron: multi vit or increase diet, possible Iron Supplement
6. Compression Stockings: 30 mmHg

Syncope Mimics

- Epilepsy- EEG, Brain CT or MRI
- Subclavian Steal Syndrome- syncope like attacks provoked by physical exercise of the arms (use US to look for)
- Strokes and TIA- doesn’t look like syncope, carotid TIA- neuro deficit hemiparesis or aphasia
- Hyperventilation Syndrome- attacks of anxiety with somatic complaints (SOB, tight chest, tingling fingers)
- Syncope of unknown origin- be wary of psych disorders/sexual abuse, conversion disorder
When To refer on...

- Cardiac: hx of exertional syncope, fam hx for SCD, EKG abnl, Echo abnl, Murmur
- Neurology: headache component of syncope, migraines, ? Seizures, concussions, strokes
- Genetics: EDS concern
- Neuropsych: post-concussive syndrome, ADHD/ADD, school issues
- Psychiatry/Psychology

Cardiac Syncope

Cardiac: Associated with heart block, cardiac arrhythmias, or structural heart disease
- Sudden onset and absence of premonitory warning symptoms (sweating and palpitations may occur)
- May happen in erect or prone position

Cardiac Syncope and kids

- Things to think of:
  - Long QT syndrome
  - Hypertrophic cardiomyopathy
  - Right ventricular dysplasia
  - Brugada Syndrome
  - Polymorphic VT with short or normal QT
  - Congenital aortic stenosis
  - Paroxysmal SVT

[Diagram of Classic ECG patterns for Brugada 1-2-3: Coved ST, saddleback, combo of both]

[Diagram of Hypertrophic Cardiomyopathy]
Treatment options beyond PCP

**MIDODRINE**
- **Background:** Midodrine Hydrochloride (Orvaten, ProAmatine) is used to treat orthostatic hypotension. It is an alpha-1 agonist. This means it causes constriction of blood vessels in arteries and veins and increases blood pressure and increases vascular tone. This helps decrease pooling of blood in lower extremities.
- **Forms:** 2.5 mg, 5 mg, and 10 mg tab
- **Dosing:** 5 mg three times daily starting dose (no more frequent than every 3 hours, and do not give after 6pm), onset of action 30 minutes–1 hour, lasts 3–4 hours, max dose 20 mg three times daily.
- **Side Effects:** Supine hypertension, flushing of face, confusion, dry mouth, anxiety, headaches, rash, and piloerection (goose bumps).
- **Prior to Starting Medication:** Ensure no supine hypertension and assess for renal function.

**FLUDROCORTISONE ACETATE** (Florinef)
- **Background:** Fludrocortisone Acetate (Florinef) is a mineralocorticoid (hormone involved in retention of sodium). It helps kidneys retain sodium. It is used to treat fainting due to low blood pressure, because it promotes fluid retention through kidney retention and fluid absorption.
- **Dosing:** 0.1-0.2mg daily (Do not suddenly discontinue medication- may become ill.)
- **Side Effects:** Change in appetite, upset stomach, headache, trouble sleeping, mood swings, growth suppression, inhibition of the body’s natural steroid production.
- **Avoid:** Vaccinations during therapy unless approved by provider. Avoid exposure to chickenpox or measles. Florinef contains lactose so if you are intolerant to some sugars you might not tolerate.

**PROPANOLOL** (Inderal)
- **Background:** Propranolol is a nonselective beta-blocker (it blocks the action of epinephrine and norepinephrine at specific protein receptors in the nervous system). This medication helps regulate your heart rate, and can be used to treat high blood pressure, elevated heart rate or migraines.
- **Formulations:**
  - 10mg, 20mg, 40mg, 60mg and 80 mg tablets
  - Extended Release has not been found to be helpful for POTS- tachycardia complaints
- **Dosage:**
  - Start most of my patients at 20 mg three times daily
- **Side effects:** The most common side effect is tiredness (fatigue, lack of energy)- for many this will stop in 2 weeks of starting Propranolol. It may help to take the medicine at bedtime. It fatigues/lethargy is a problem. Other common side effects: nausea, depression, abdominal pain, constipation and worsen asthma.
- **Contraindications:** Propranolol should not be used for patients with severe asthma, or those with 2nd or 3rd degree heart block. Alternatives should be considered in patients with diabetes, bradycardia (slow heart rate), kidney or liver disease, and depression.
- **Precautions:** Avoid abrupt withdrawal of medication- needs to be weaned off. Propranolol should be held during periods of dehydration (diaphoresis, vomiting, and inability to take oral intake). Propranolol can be retained at normal dose when return to hydrated state.
Exercise Prescription

They will meet with Ashley Kimball my exercise physiologist for specific exercise prescription.

- **TILT STANDING TRAINING**
  Standing training should be done daily. This allows the body to better adapt to the upright position and gravity. Start at 10 minutes 3 times a day. You can stand at a kitchen counter for balance or using a wall.

- **Aerobic Exercise Guidelines**
  **Frequency:** 4-5 days per week  
  **Intensity:** 75-80% Max Heart Rate, RPE 13-15  
  **Time:** 30 minutes  
  **Type:** Seated / Supine Modalities, upright activities as tolerated  
  **Progression:** 10% increase in intensity in a 2 week period does goal duration of 30 minutes is achieved.

- **Resistance Training Prescription**
  **Frequency:** 2-3x/week  OR 4-5x/week**  
  **Intensity:** RPE of 15  
  **Time:** ~30 minutes  
  **Type:** All large muscles groups, squats, lunges, push-ups, bridges, planks, hoop curls  
  **Progression:** 10% increase in weight as RPE decreases to 12 or as suggested by exercise professional.

**Resistance training can be done as a whole body workout sessions 2-3 days per week or can be broken down into muscle group days.**

Other Testing:

- **Tilt Table Testing:**
  - Positive-end-point: syncope or reproducibility of symptoms associated with near syncope
  - Patient has to fast for 2 hours prior to test
  - Good for diagnostic purposes, but not recommended for treatment efficacy

- **Electrophysiology Study:**
  - Useful when looking at atrial automaticity (HR, sinus node recovery and intrinsic HR)
  - Pacing atria at higher rate to stress adequacy of AV conduction system—highly predictive of impending AV block
  - Examine for Paroxysmal SVT or SVT (AVNRT, WPW, AVRT, atrial flutter) and ablate

- **Implantable Loop Recorder:**
  - Used to identify heart rhythm during Syncope/Presyncope if Holter or Event monitor have been able to document rhythm, placed left of sternum, small subcutaneous pocket, battery life 18-24 months, diagnostic yield 25-40%

References: