What to do with the fainting or dizzy child?

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Learning Objectives

- define syncope, Postural Orthostatic Tachycardia Syndrome (POTS) and their causes
- Treatment options: what can be done in primary care to evaluate, diagnose, and treat
- When to refer these patients to specialists and which specialists to refer them to

The Definition of 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

POTS Defined

- Postural Orthostatic Tachycardia Syndrome- heart rate elevation of 40 points with change in position (lying to sitting or lying to standing) or heart rate above 120 bpm within 10 minutes of standing, with no orthostatic hypotension.
- Signs/Symptoms (to name a few):
  - tachycardia /palpitations with orthostatic change
  - light headed
  - headache
  - nausea
  - syncope
  - concentration issues (brain fog)
  - fatigue, exercise intolerance
  - purplish coloring in legs with standing

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, etc
- Common event in our patients' lives- neurally mediated syncope 35% by age 18 and 50% by age 21

Disclosures

- None

www.dysautonomiainternational.org

Paris, et. al JAH A. 2015. 1-13

Etiology of Syncope

Psych 3%
Idiop 18%
Hyp 5%
Card 59%
Neur 9%
HypoaDr 9%
NMS 39%
Why do we care about POTS?

- Estimated to impact 1,000,000 -3,000,000 Americans and many more worldwide

- Quality of Life: some mild decreases in their QOL others much more debilitated: some healthcare providers feel impairment of QOL of POTS patients comparable to those with chronic obstructive pulmonary disease or CHF, 25% are disabled/unable to work

Grubb, Circulation. 2008; 117: 2814‐2817

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


Another Way to Look At Syncope etiology:

- 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

- Vasovagal, vasodepressor, neurocardiogenic syncope= NM Syncope

NM Syncope - Continued

- Most frequent of all syncope
- Types of NMS: central (emotional reaction), Postural (most common), situational (hair brushing, defecation, postprandial, micturition),
- Symptoms:
  - Lightheadedness
  - Palpitations
  - Weakness
  - Dim/blurred vision
  - Nausea
  - Diaphoresis
  - Dilated pupils
  - Pale

Subsets of NM Syncope

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

POTS - chronic symptoms of orthostatic intolerance (>6 months), >40 pt HR increase with change in position within 10" standing, without orthostatic hypotension
- lying, sitting, standing BP/HR and standing at 5" and 10" BP/HR test =evaluates pt for POTS

POTS Defined...

- Primary- no other identifiable disorders cause: "idiopathic"
- Secondary- another medical condition that can contribute to or cause POTS

4 Types of POTS

- 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 OI 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 OI, GI dysfx, bladder retention and anhidrosis (Rare in Kids)
  - Causes: autoimmunity, diabetes

Many can have mixture of types.

What Causes POTS- the $1,000,000 or >?

- There are a number of factors and disorders that are associated and can cause POTS.
- POTS can be described as "primary"- no other identifiable medical condition, or "secondary": presence of another medical condition known to contribute towards POTS: i.e. Ehlers Danlos Syndrome- hypermobility type, Infections (Mono, Strep, EBV), Deconditioning, to name a few.

The Extensive List I have found:

- Autoimmune Disease
- Autoimmune Autonomic Ganglionopathy, Sjögren’s Syndrome, Lupus, Sarcoidosis, Antiphospholipid Syndrome
- Chronic Mediation
- Deconditioning
- Ehlers Danlos Syndrome
- Genetic Disorders/Mutations
- Infections such as Mononucleosis, Epstein Barr Virus, Lyme Disease, extra pulmonary Mycoplasma pneumonia and Hepatitis C
- Multiple Sarcoidos
- Myocardial Disorders
- Neutrophilic Enteritis
- Neuroendocrine Thyroid Disease
- Neuroendocrine Pancreatic Disease
- Paroxysms
- Paroxysmal SVT
- Pulmonary Hypertension
- Sarcoidosis
- Scleroderma
- Sturge-Weber-Sphenoid Syndrome
- Unique Syndromes: Neuronal Deficiency

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
- 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
  - Hypertrophic Cardiomyopathy
  - Delta Wave-WPW (SVT)
  - Causas EKG patterns for Brugada: 1. Covid 51, 2. add block, 3. combo of both
What can be done in primary care?

Take the History:
1. ask the lifestyle ’s: diet, hydration, sleep, headaches, school/home life, activities involved in, menstrual hx
2. Family: SCD, heart dysrhythmias, seizure, stroke, syncope, connective tissue
3. Symptoms with syncope: dizziness, vision changes, palpitations, chest pain, shortness of breath, loss of bowel/bladder, jerking motions pre syncope
4. History of dislocations or hypermobility, poor wound healing

Physical Exam in Primary Care

- Vital signs: do orthostatic HR/BP (lying, sitting, standing, Standing at 5 and 10")
- Neurologic: pupil reactivity, extra ocular motility, finger to nose, carotid bruits
- Cardiac: murmurs, heart rate, chest pain- reproducible, pulses
- Beighton score:

<table>
<thead>
<tr>
<th>Beighton Score</th>
<th>Normal (0-2)</th>
<th>Abnormal (3-6)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperextensible elbows</td>
<td>0-2</td>
<td>3-6</td>
</tr>
<tr>
<td>Hyperextensible knees</td>
<td>0-2</td>
<td>3-6</td>
</tr>
<tr>
<td>Hyperextensible fingers</td>
<td>0-2</td>
<td>3-6</td>
</tr>
<tr>
<td>Hyperextensible spine</td>
<td>0-2</td>
<td>3-6</td>
</tr>
</tbody>
</table>

Testing In Primary Care

- Cardiac: EKG, Holter Monitor, or Event Monitor, Echocardiogram
- If Syncope sounds like either orthostatic hypotension or POTS: Orthostatic BP/HR (lying, sitting, standing, standing 5 and 10")
- Labs: CBC, BMP, Iron Panel and Ferritin, TSH, Vit D
- Syncope due to seizures- EEG
- CT for concussion evaluation

Treatment Recommendations- Primary Care

- Lifestyle:
  - increasing fluid intake (64 ounces or 2 liters daily)
  - increasing salt consumption (2,000 mg/day-9,000mg/day)
  - wearing compression stockings (30-40 mmHg)
  - healthy diet (no skipping meals, breakfast every morning)
  - aerobic activity daily: any activity that can increase vascular tone: walking, swimming, rowing machine, biking, jogging along with core strengthening (planks, squats, sit ups)
  - Counter maneuver pressure activities
  - Good sleep hygiene

Treatment in Primary Care- Medications

Iron- if mildly low multi vit or increase diet
- If really abnl: 4-6 mg/kg/day divided 2-3 times/day, start stool softener, if nl in 3 months than go to multi vit with Fe

Ferritin- if patient has sleep issues consistent with restless leg syndrome goal to get Ferritin to > than 50

Vitamins- Vit D: levels should be at least 20, if patient has complaints of chronic pain recommend 1,000-2,000 IU/day

Treatment in Primary Care- Medications

Midodrine Hydrochloride (Orvaten or ProAmatine)
- alpha-1 agonist: peripheral arterial/venous vasoconstrictor
- helps increase vascular tone in periphery (decreases venous blood pooling in LE), and increases blood pressure

Dose: start at 5 mg three times daily can go as high as 20 mg three times daily

Side Effects: pilorection, if take after 6pm can affect ability to fall asleep, skin rashes, paresthesias, supine hypertension
- It has a very short half- life (3-4 hrs) so have to take frequently for all day coverage
**Treatment in Primary Care - Medications**

**Fludrocortisone Acetate (Florinef) - Mineralocorticoid** (helps kidney retain sodium, and promotes fluid retention)

**Dose:** 0.1-0.2 mg daily (Max 0.2) - do not stop abruptly

**Side effects:** Change in appetite, upset stomach, headache, trouble sleeping, mood swings, growth suppression, inhibition of the body's natural steroid production. Use long term questionable.

**Avoid:** Vaccinations during therapy unless approved by provider, exposure to chickenpox or measles. Contains lactose so if you are intolerant to some sugars you might not tolerate.

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**Propranolol:** nonselective beta-blocker

This medication helps regulate heart rate, elevated heart rate or migraines.

**Dose:** 20 mg three times daily

**Side effects:** fatigue (for many this will stop in 2 weeks of starting), nausea, depression, abdominal pain, constipation and worsened asthma.

**Avoid:** Not be used for patients with severe asthma, or those with 2nd or 3rd degree heart block. Avoid abrupt withdrawal of medication - needs to be weaned off. Should be held during periods of dehydration (diarrhea, vomiting, and inability to take oral intake). Can be restarted at normal dose when return to hydrated state.

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**Treatment for POTS**

<table>
<thead>
<tr>
<th>Type of POTS</th>
<th>Volume Expansion</th>
<th>Physical Activity</th>
<th>Pharmacotherapy</th>
<th>Level E/M</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neuropathic</td>
<td>+ + + +</td>
<td>+ + +</td>
<td>Midodrine</td>
<td>I (likely)</td>
</tr>
<tr>
<td>Hyperadrenergic</td>
<td>+ + + +</td>
<td>+ + +</td>
<td>Volume Expansion, Propranolol</td>
<td>I (likely)</td>
</tr>
<tr>
<td>Deconditioned/Hypovolemic</td>
<td>+ + + + +</td>
<td>+ + + + +</td>
<td>Volume Expansion, Sustained exercise program</td>
<td>II (important)</td>
</tr>
</tbody>
</table>

*Levels: ++ (slightly), +++ (moderately), ++++ (very important)*

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**When to refer to Pediatric Specialty Service?**

- **Cardiology:** Syncope with exertion, complaints of palpitations or heart racing prior to syncope, abnl EKG, family history of SCD, heart dysrythmias, or sudden unexplained death
- **Neurology:** Syncope or dizziness with headaches/migraines, loss of bowel or bladder with syncope, jerking/rhythmic motions of body prior to syncope
- **Genetics:** Beighton Score of 5 or more, chronic joint pain, easily dislocates
- **GI:** n/v, constipation, diarrhea, reflux, appetite suppression
- **Neuropsych and/or Psychology:** post-concussive syndrome, ADHD/ADD, school issues, short term memory loss/brain fog

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**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

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**Resources for families**

- Dysautonomia Youth Network of America, INC: www.dynakids.org
- POTS - A World Tour - Dr. Satish Raj: https://vimeo.com/72346576
- Postural Tachycardia Syndrome, Blair P. Grubb, Circulation, 2008; 117: 2814-2817
References


• Moya, Angel et. al. (2009). Guidelines for the diagnosis and management of syncope. European Heart Journal. 30: 2631-2671


