



How to Care for POTS at School

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Pediatric Cardiology

UAB
MEDICINE.

DYSAUTONOMIA INTERNATIONAL



AWARENESS



ADVOCACY



ADVANCEMENT

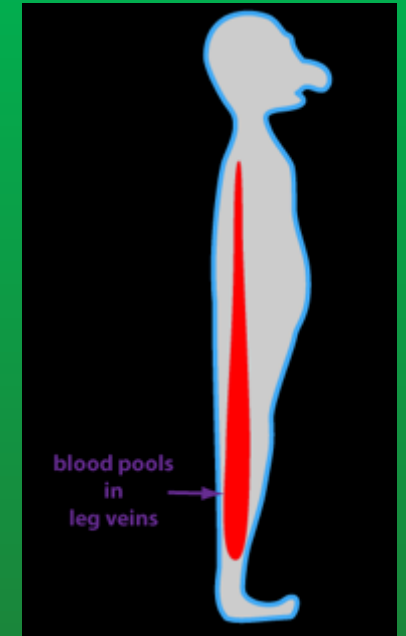
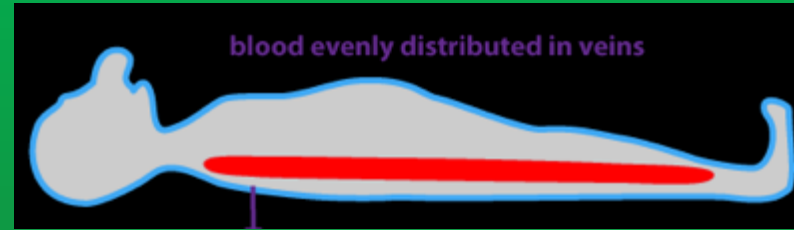
Today's Agenda - POTS

- Definitions and Pathophysiology
- Clinical Pearls
- Treatment Options
- Outcomes



Definitions

- **Orthostasis**
 - assuming the upright position
- **Orthostatic tachycardia**
 - sustained \uparrow HR ($> 30-40$ bpm) within 3 min of standing
- **Orthostatic hypotension**
 - sustained \downarrow SBP > 20 mmHg or diastolic BP > 10 mmHg within 3 minutes of standing



Definitions (2)

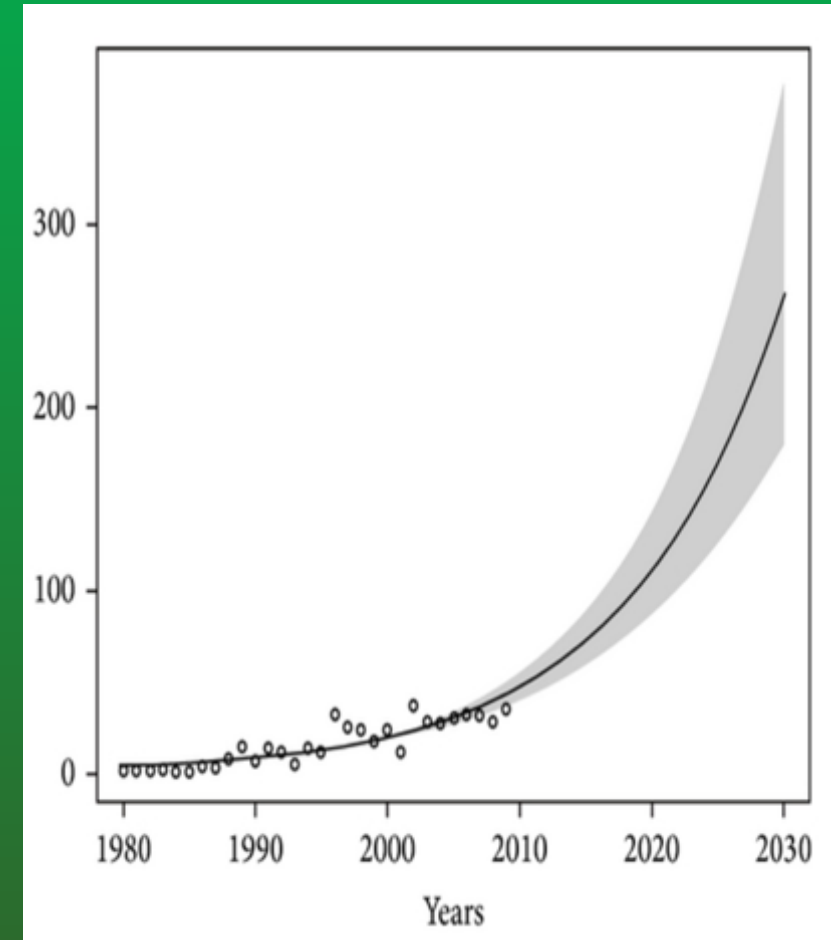
- **Orthostatic intolerance**
 - **Symptoms** of lightheadedness, dizziness, blurry vision, etc that occur with being upright (sitting/standing)
 - Symptoms resolve with lying down
 - Symptoms are principally due to initial decrease in cerebral perfusion and the circulatory response (\uparrow HR, \downarrow BP)
- **Vasovagal syncope**
 - Loss of consciousness and postural tone
 - Due to reflex decrease in HR and/or BP
 - Stimuli: dehydration, standing, pain, emotional startle

Postural Orthostatic Tachycardia Syndrome

- POTS is a clinical syndrome of orthostatic intolerance
- **Definition**
 1. **Daily / significant symptoms** of orthostatic intolerance
 2. Sustained increase in HR (**> 30-40 bpm**) within 10 minutes of upright posture / tilt table test
 3. Symptoms are **chronic** (> 6 months) and relieved by lying down
 4. **Diagnosis of exclusion**
- **Practical Additions**
 - Requiring treatment with medications
 - Symptoms are severe enough that the patient is missing school or work

Pertinent POTS Points

- Increasing incidence?
 - Individual institutions – 4x increased rate of diagnosis starting ~ 2012*
- **5:1 female-to-male ratio****
- Cause?
 - Many report onset of symptoms after a surgery, concussion, or illness (influenza, mononucleosis)
 - Hypermobility and EDS
- **Diagnosis of Exclusion**
 - Medications
 - Thyroid disease, anemia
 - Primary psychiatric
 - ***Screen for eating disorders!***



* Brinth L. Auto Neurosc 2018

** Sheldon. Heart Rhythm 2015

Dysautonomia and POTS

- “Sister” diagnoses

- **Dysautonomia**

- Significant orthostatic intolerance
- Prominent autonomic symptoms
 - Abnormal sweating
 - Heat/cold intolerance
 - Sleep disturbance
 - Gastroparesis
 - Urinary retention





Orthostatic Intolerance

POTS

Dysautonomia

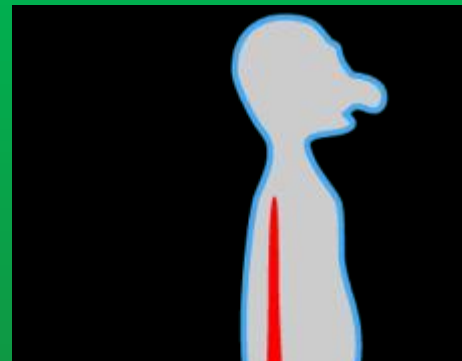
Vasovagal
Syncope

Orthostatic
Hypotension

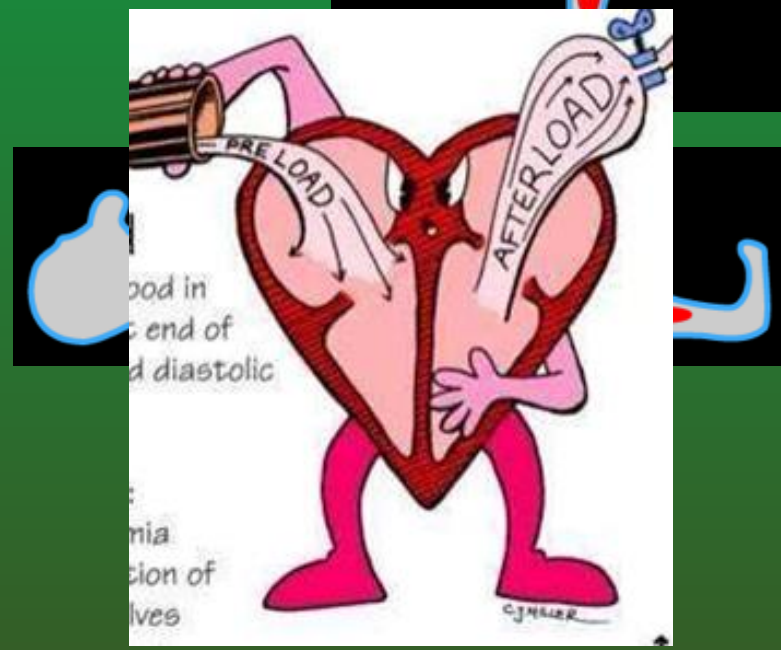
The Challenge of Standing

- Initial physiology of orthostasis
 - In adults, > 500 mL of blood is transferred caudally with standing
 - Decreased venous return to the heart → decreased stroke volume, CO, and ultimately BP
 - Symptoms (LH, nausea, vision change) coincide due to transient decreased cerebral perfusion

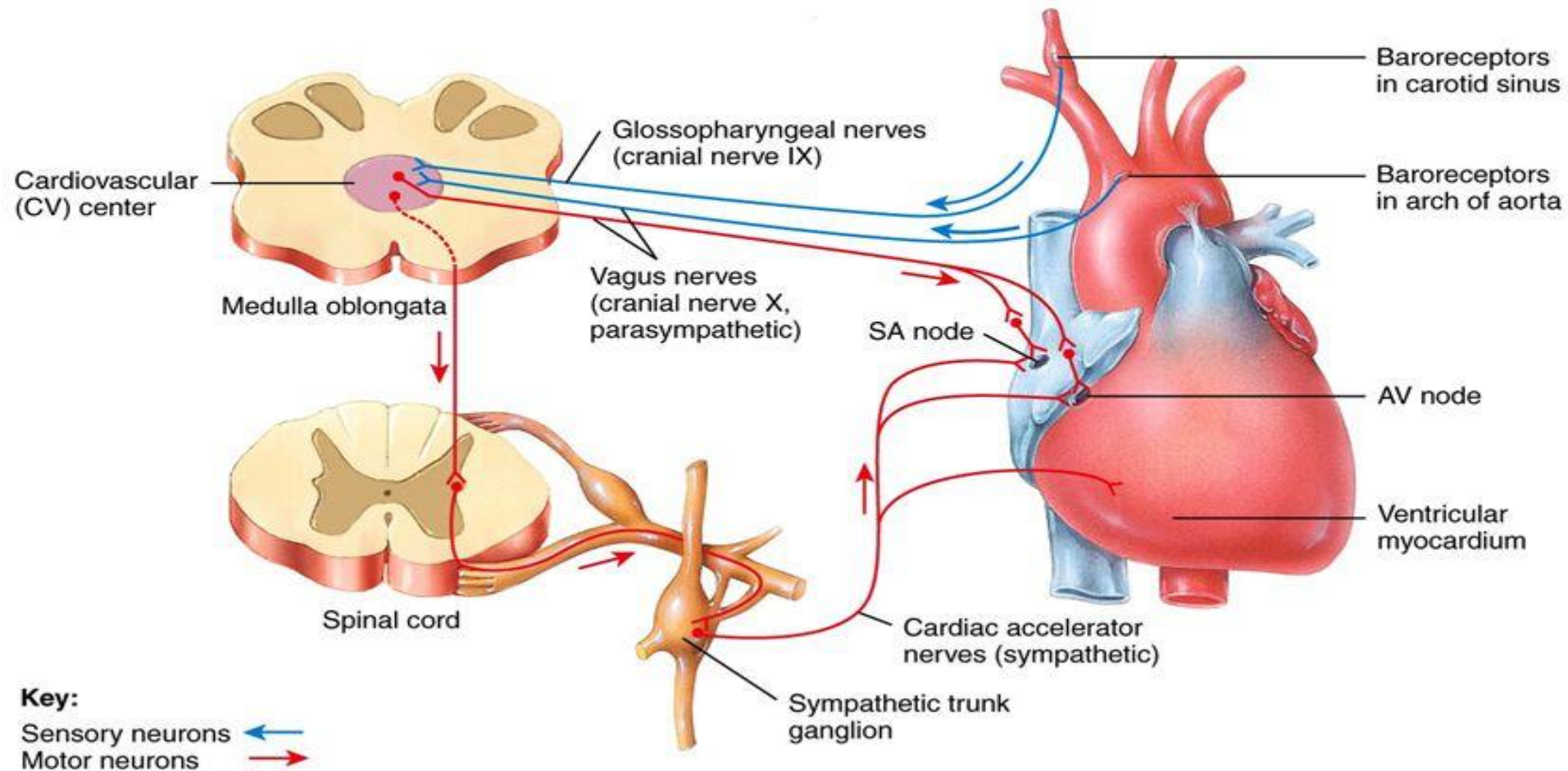
$$BP = CO * SVR$$

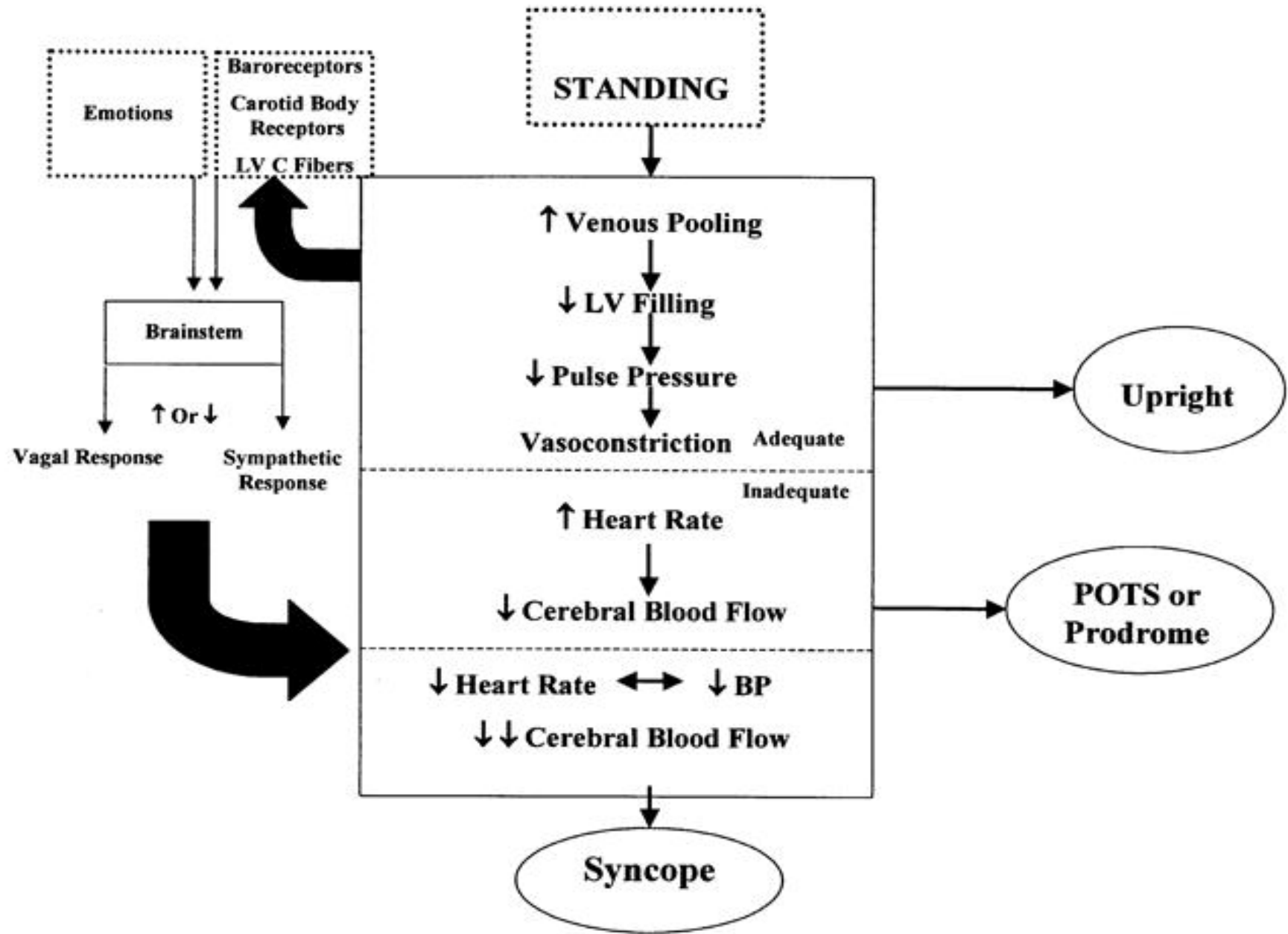


blood pools in leg veins

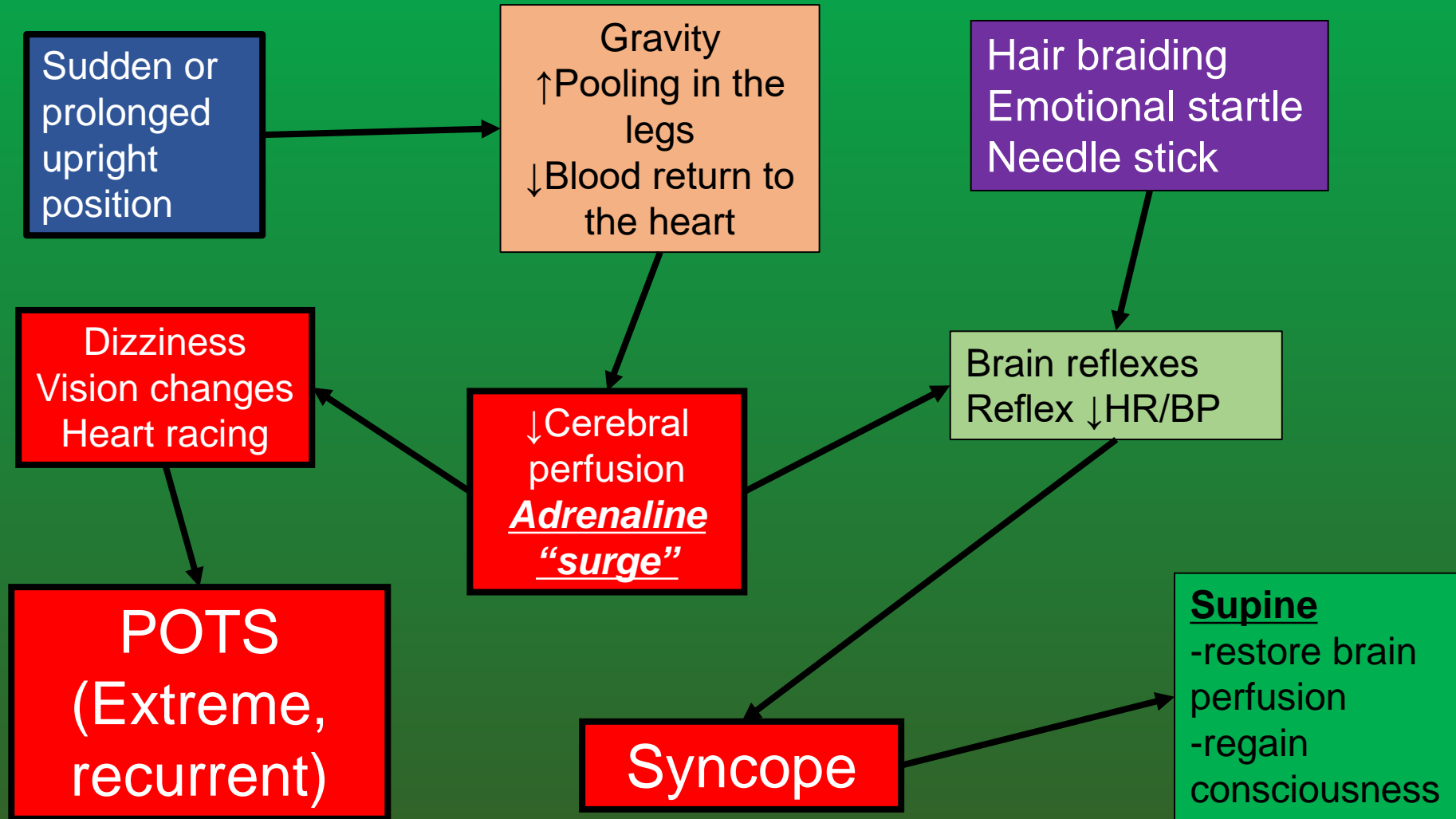


Autonomic Nervous System Regulation of Heart Rate





How I Explain It To Patients



Susceptible individual

- Unknown genetic predisposition
- Highly active person



Inciting event:

- Infection
- Surgery
- Concussion
- Pain/headache
- Enforced bedrest



DEVELOPS SYMPTOMS:

- Orthostatic intolerance (occurs early even after brief bedrest)
- Exercise intolerance
- Dizziness



DIMINISHED ACTIVITY +/- FURTHER BEDREST



CARDIOVASCULAR DECONDITIONING:

- Decreased blood volume
- Decreased stroke volume
- Cardiac atrophy



SYMPATHETIC ACTIVATION (in upright position):

- Increased heart rate
- Decreased GI motility
- Headaches



The “downward spiral” of POTS

-Several factors culminate (? susceptible individual, inciting event, etc.), ultimately leading to **diminished activity, bedrest, deconditioning, and potentiation of symptoms**

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Assessment of the Patient with Orthostatic Intolerance

The best "test" is a detailed medical history

- Symptoms
 - Dizziness
 - Palpitations
 - Headaches
 - Fatigue
 - "Brain fog"
 - Syncope – preceded by dizziness?
 - Anxiety
 - Joint pain, GI intolerance
- Frequency, duration, and **severity** of symptoms – meeting criteria for **POTS**?
- Body position at time of symptoms? Relieved by recumbence? Timing (morning, before lunch)?
- Stressor at onset of symptoms (viral illness, concussion, surgery, etc.)
- Fluid and salt intake? Breakfast? Exercise? Missing school?

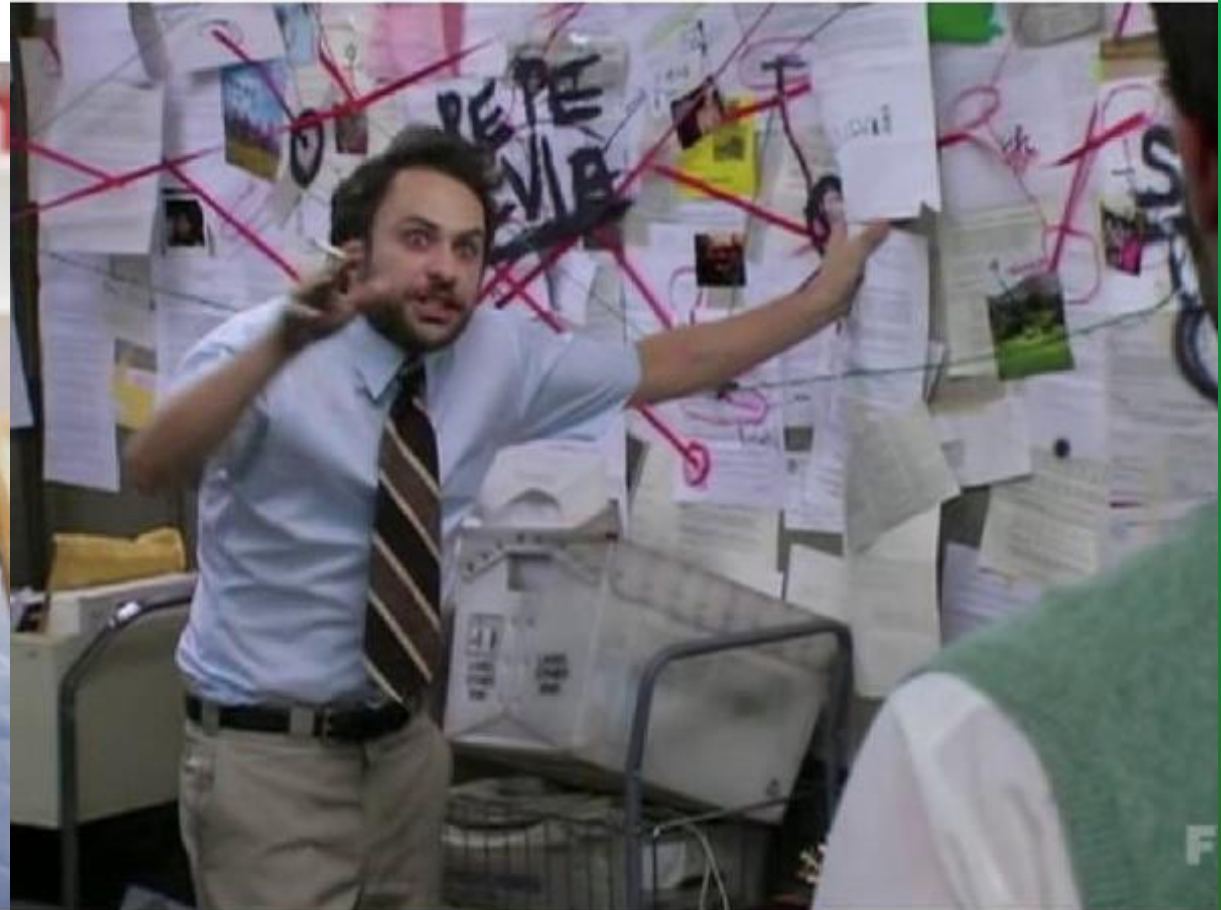
Abdominal pain
Raynaud
Pseudoseizures
Joint pain
Missed school
Anxiety
Hopelessness
Iron deficiency
Palpitations
Syncope
Vitamin D deficiency
Can't concentrate
Insomnia
Dizziness
Fatigue
Leg discoloration
Positive orthostatic vital signs
Nausea
Can't exercise
Tachycardia
Chest pain
Shortness of breath
Constipation/Diarrhea
Depression
"Somethings wrong"
Fear of Missing Out (aka FOMO)



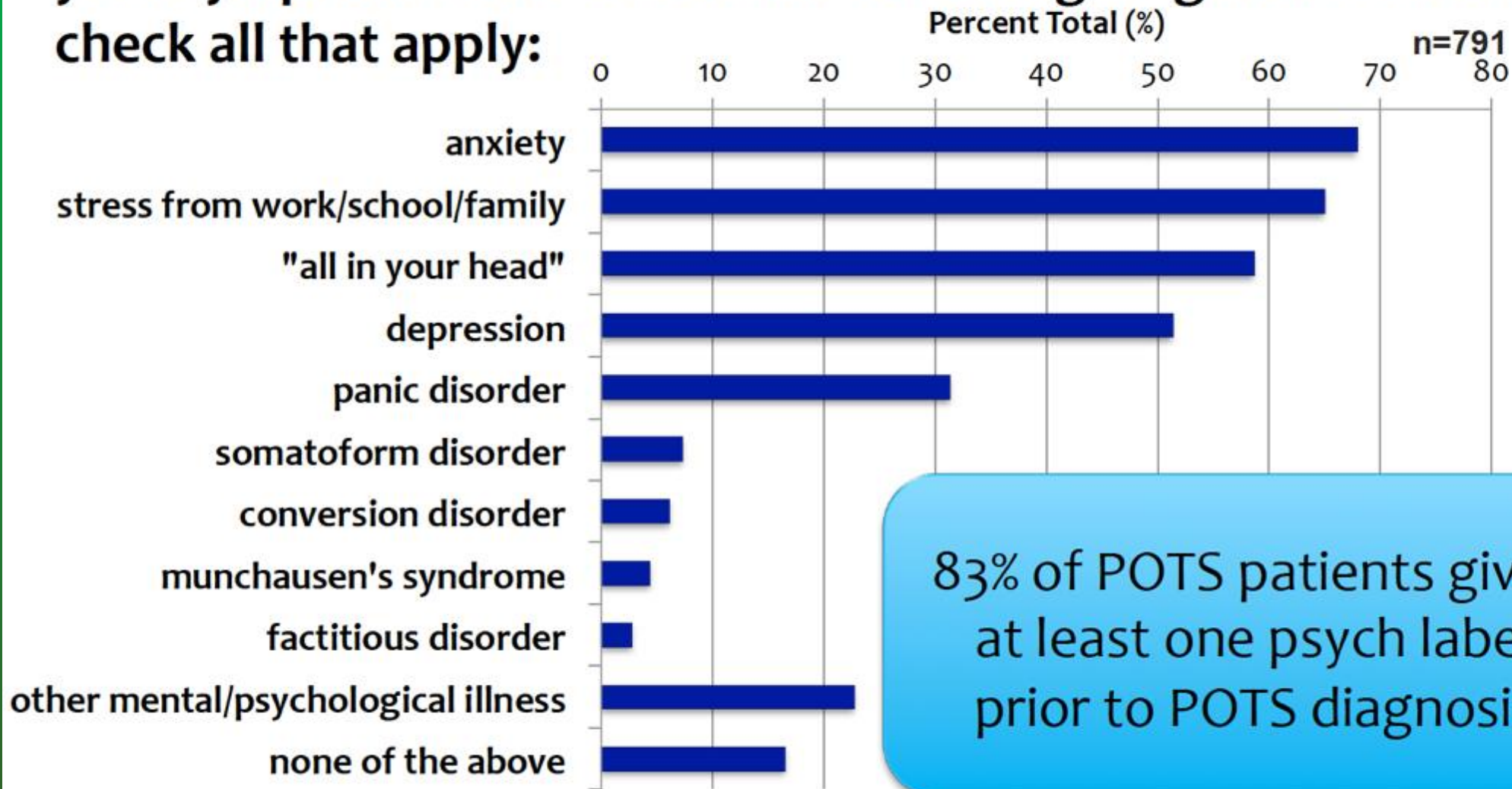
What I think I sound like when I'm talking to a new doctor about my chronic illnesses:



What the doctor hears:



BEFORE your POTS diagnosis, were you ever told by a doctor that your symptoms were due to the following diagnoses? Please check all that apply:



Vasovagal syncope

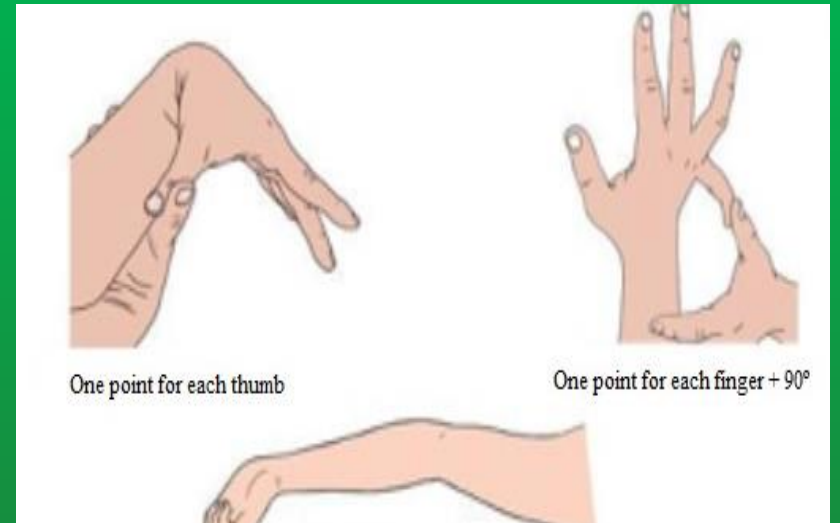
- NOT so prominent a symptom in POTS patients
- Only described in $\sim 30\%$ of patients with chronic symptoms related to POTS*

-Lots of syncope?

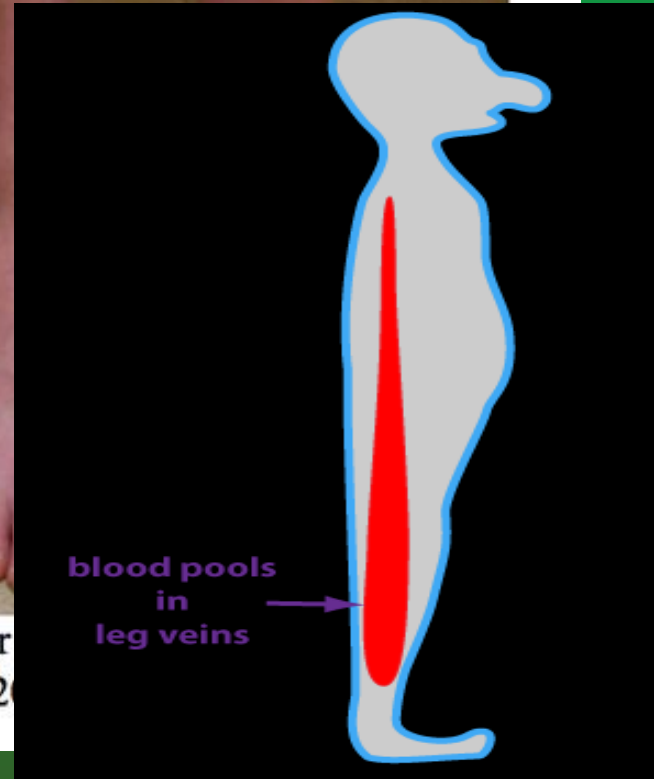


*Shen. JACC 2017

Physical Exam – Orthostatic Intolerance



Before and After Standing

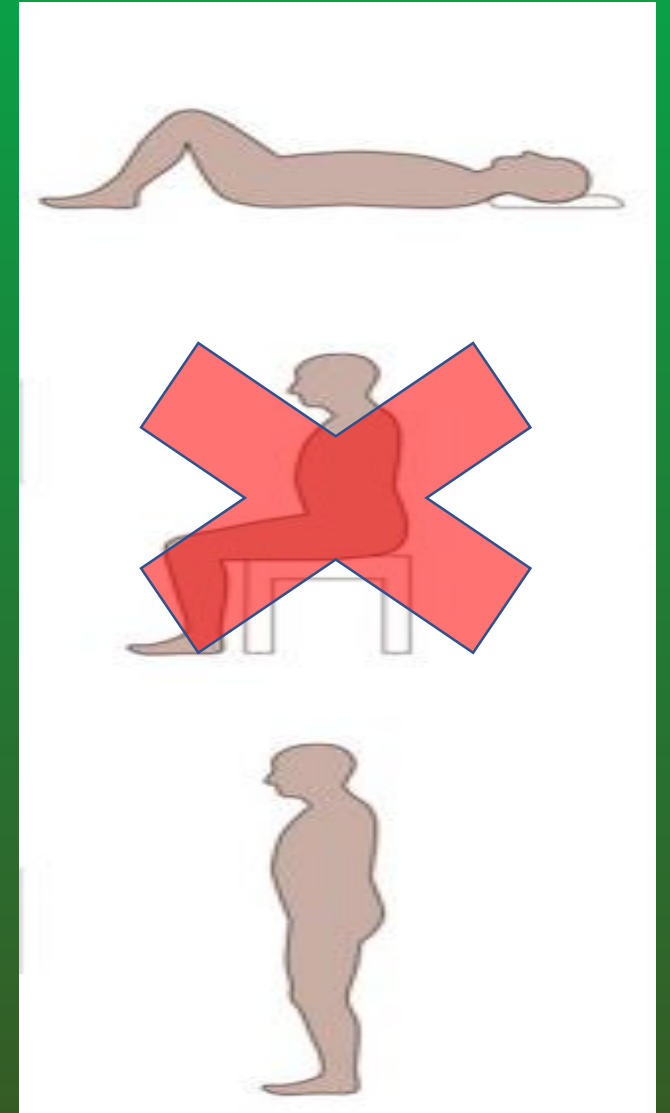


Sitting	After
Pulse: 80	Pulse: 120
Blood Pressure: 100/72	Blood Pressure: 100/72

Orthostatic Vitals – 3 Minute Standing Test

• Recommendations

1. Baseline HR, BP – lying down
2. Then stand, no need for sitting vitals
3. While standing, wait and take HR and BP at:
 - 1 min, 2 min, 3 min
 - Patient should stand as still as possible



Vitamin D deficiency

- Associated with **orthostatic hypotension** in adults^{1,2}
 - Association in children has not been well-defined
- Also associated with **depression and anxiety**³⁻¹⁰ → frequent cohabitants!
- Goal: 25-Vitamin D > 40 ng/mL
- Dose: Vitamin D3 4000 IU daily in adults, 1000 IU per 25 lbs in kids



1. Annweiler C. J Intern Med 2014
2. McCarroll K. Age Ageing 2012
3. Parker G. J Affect Dis 2017
4. Anglin R. J Psych 2013
5. Balion C. Neurology 2012
6. Wu C. Medicine 2016
7. Huang J. J Women Health 2014
8. Armstrong D. Clin Rheum 2007
9. Kelley L. J Dev Orig Health 2016
10. Bicikova M. Phys Res 2015



Iron Deficiency

- Increased incidence in adolescents with POTS¹
- Correcting anemia can improve orthostatic tolerance²
- **Labs:** cbc with diff, ferritin
- Can contribute to symptoms EVEN IF the patient is not anemic
- **Normal ferritin: > 30 ng/mL**
- Iron and GI side effects
- Address underlying cause
 - **Menorrhagia**
 - Diet

1. Jarjour IT. Clin Auton Res, 2013

2. Low PA. Curr Opin Neurol, 1994





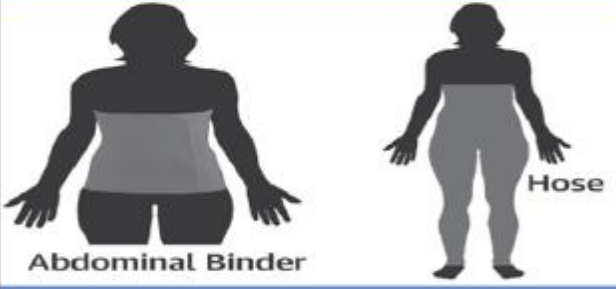
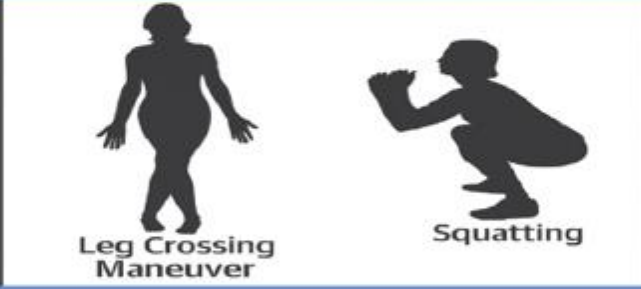




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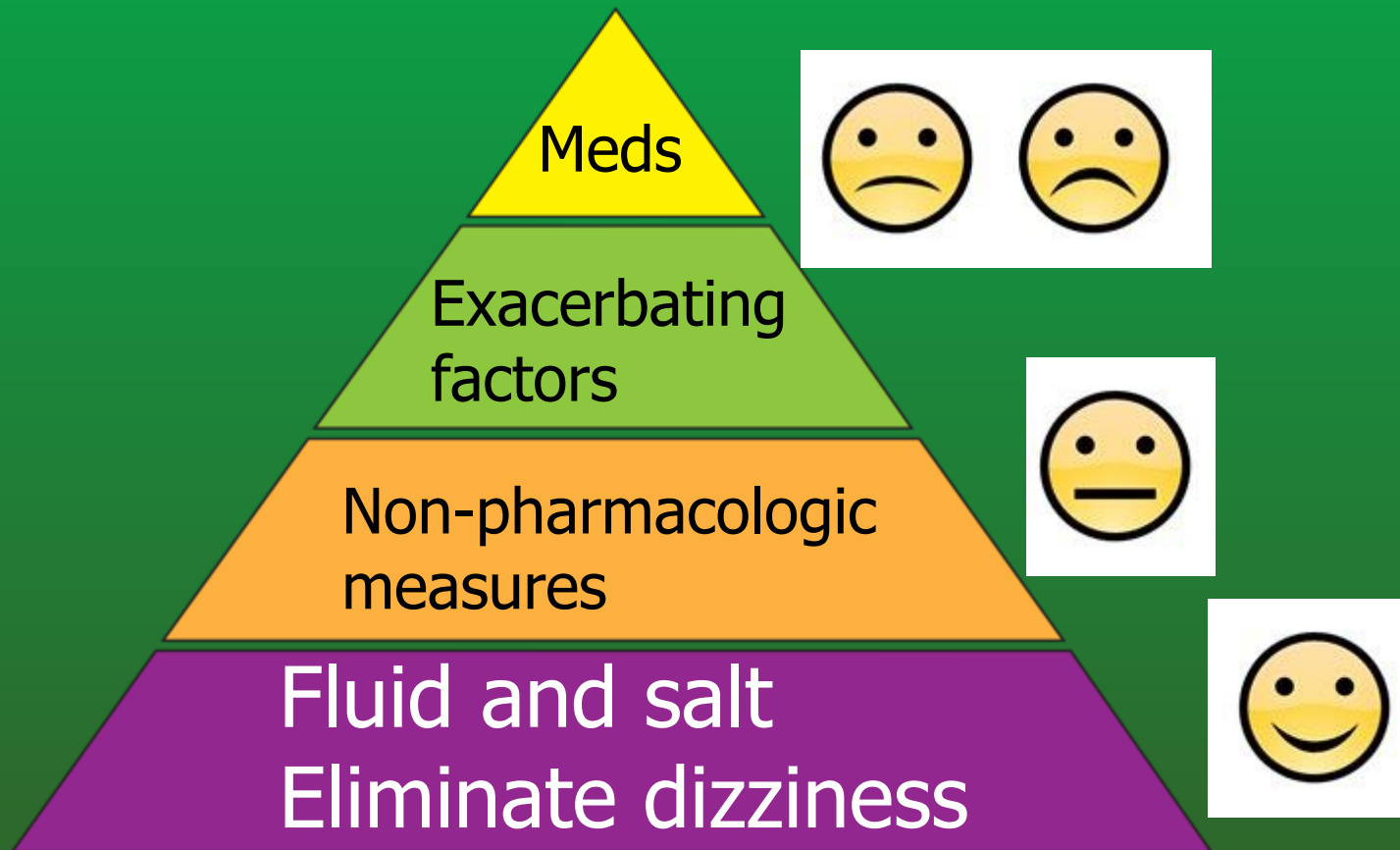


POTS often can be managed by nonpharmacological measures alone. These strategies can help increase blood volume and minimize orthostatic symptoms. In addition to boosting blood volume, an exercise regimen geared toward POTS can also increase stroke volume, increase left ventricular mass, and lead to longer lasting reduction in orthostatic symptoms. POTS = postural orthostatic tachycardia syndrome.

Bryarly M. J Am Coll Cardiol. 2019

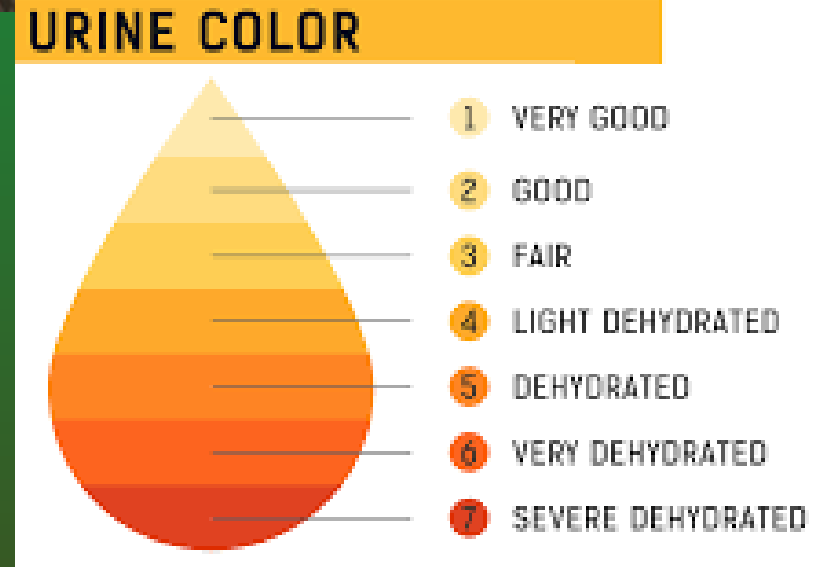
<p>Avoid Situations That Can Exacerbate Symptoms</p>	<p>Liberal Intake of Salt and Water</p>	<p>Sleep With Head of Bed Elevated</p>
 <p>Large/Heavy Meals</p> <p>Heat Exposure</p>  <p>Alcohol Intake</p>		 <p>Head posts should be elevated 4-6 inches</p>
<p>Use of Compression Garments</p>	<p>Physical Counter Maneuvers</p>	<p>Drinking Water Before Getting Up In The Morning</p>
 <p>Abdominal Binder</p> <p>Hose</p>	 <p>Leg Crossing Maneuver</p> <p>Squatting</p>	<p>Drinking a 16 oz glass of water quickly before getting out of bed in the morning or prolonged standing to minimize orthostatic symptoms</p> 
<p>Strategies to Avoid Upright Exercise</p>		
 <p>Seated Rower</p>	 <p>Swimming</p>	 <p>Recumbant Bicycle</p>

Treatment of Orthostatic Intolerance



You Must Stay Well Hydrated!

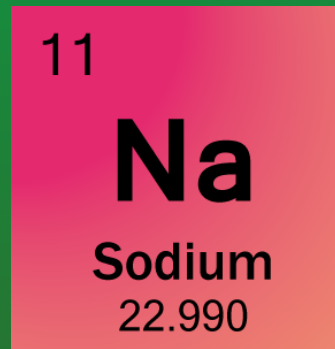
- Hypovolemia worsens symptoms
 - Decreased venous return to the heart
 - Decreased cerebral perfusion
- **2-3L or 64-100 fluid oz/day**
 - Tank up in the AM – NPO overnight!
 - Water > Sport drinks >>> Caffeinated drinks
- **How can you tell its enough for you?**
 - Clear / very light yellow urine color
 - Effect on dizziness with standing
- Literature support
 - Improves symptoms and hypotension¹⁻³



1. Shannon JR. Am J Med, 2002
2. Low, PA. Curr Opin Neuro, 1994
3. Jordan, J. Lancet, 1999

The One Time in Cardiology When Salt is a Good Thing!

- Increasing salt intake leads to increased fluid retention
- Focus on Sodium
- **3-5 g Na/day if severe symptoms**
- Will not cause weight gain
- Hypertension?
- Salt tablets
- Literature support – symptom improvement*



Nutrition Facts	
Serving Size 1 can (163 mL)	
Servings per Container 3.5	
Amount per serving	
Calories 30 Calories from Fat 0	
Total Fat	0g 0%
Saturated Fat	0g 0%
Cholesterol	0mg 0%
Sodium	520mg 22%
Total Carbohydrate	6g 2%
Dietary Fiber	1g 4%
Sugars	5g
Protein	1g

*Raj SR. Circulation, 2013

High Salt Diet

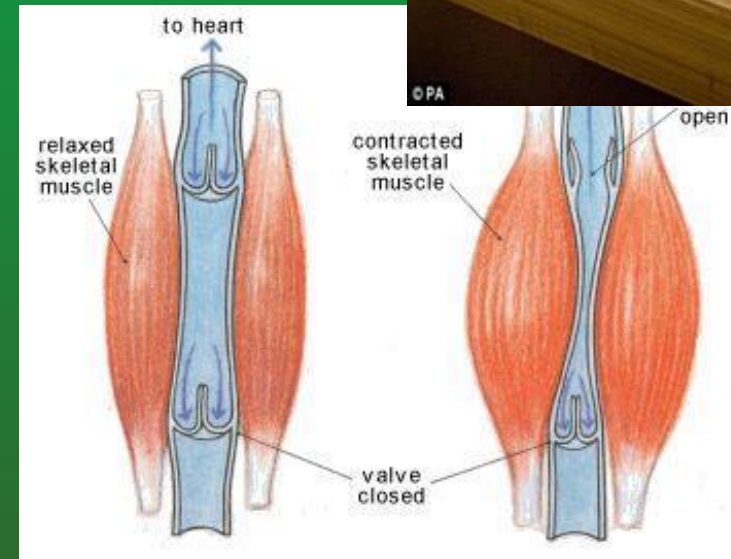
(3000-5000 mg Na/day)



Food Item	Mg of Sodium (Google)
Bacon (3 slices)	390
Table salt (1 tsp)	2300
Pretzel snack	1400
Salted nuts (1/2 cup)	420
Hamburger	690
Salt tablet	250 mg / tablet
Dill pickle	1430
Soy sauce (1 tbsp.)	870
Beef Jerky	420

Exercise Improves Orthostatic Tolerance

- Effects of prolonged bedrest¹
 - Decreased plasma volume
 - Muscle atrophy → **loss of skeletal muscle pump**
- Leg muscle training and improved venous compliance,² can expand blood volume 20-25%³
- **Trials** in teens with POTS have shown efficacy⁴
- PT sometimes to start
 - Modified Dallas Protocol
 - Exercise with weights while lying flat
- Ramp up over time
- Swimming, recumbent bike

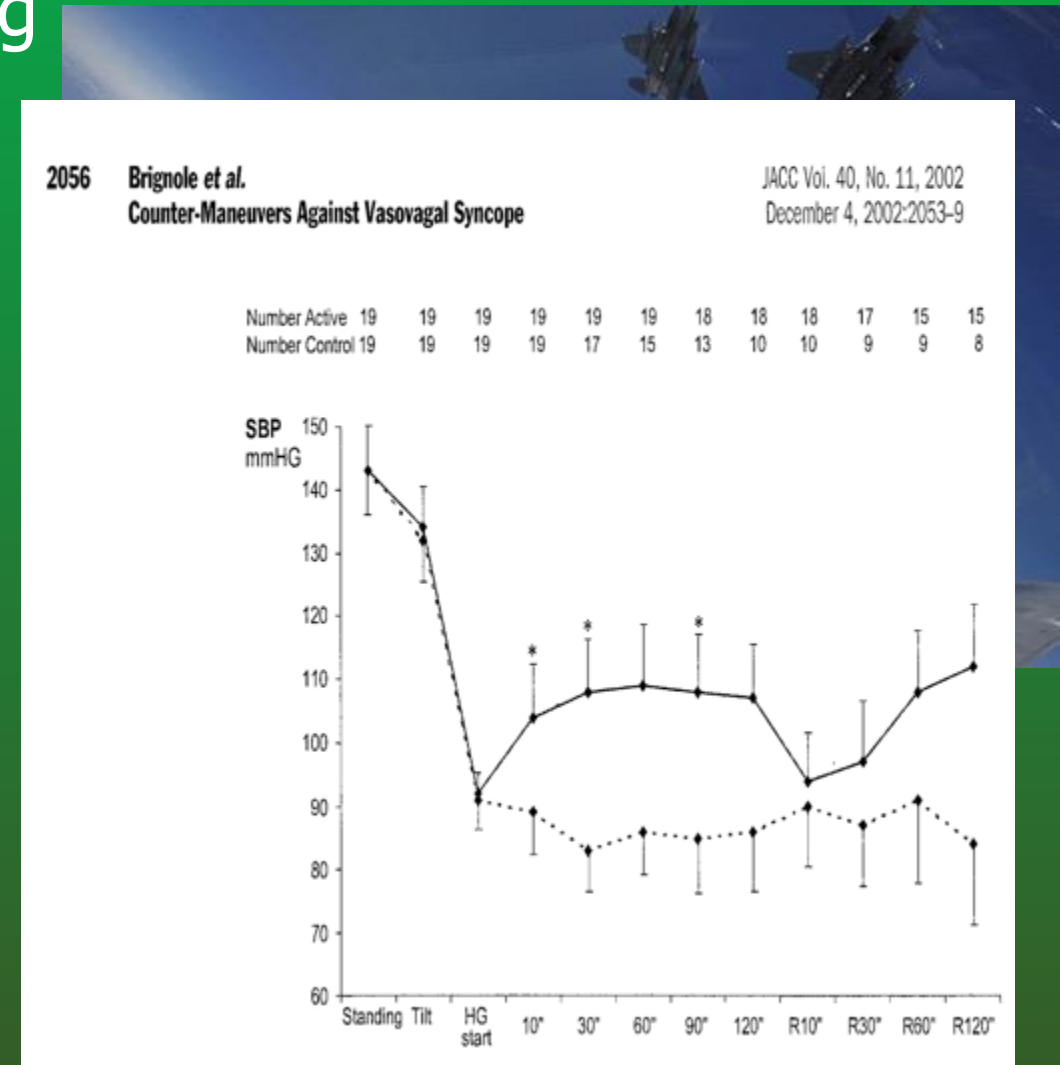


1. Greenleaf JE. Exerc Sport Sci Rev 1982
2. Hernandez JP. J Appl Physiol 2005
3. Convertino V. Am J Med Sci 2007
4. Bruce B. Clin Pediatr 2016

Counter pressure maneuvers

- Improve cerebral perfusion by improving venous return to the heart or **directly increasing BP**
 - Standing with legs crossed
 - Tightening pelvic muscles
 - Handgrip
 - Biceps curl
 - Avoiding standing passively
- **Literature support**
 - Tensing the leg muscles while standing → improved NIRS and MCA blood velocity by ultrasound*

*van Lieshout. Stroke, 2001



Compression socks

- Goal: decrease LE venous pooling
- Strength: 20-40 mmHg
- Take off during exercise and at night
- \$20-50.00 per
- At least knee high, closed toe
- brightlifedirect.com



Medications to Avoid*

- Vasodilators
 - ACEi, CCB
- Many medications used for “migraines” or chronic pain
- Diuretics
- Opiates
- Antiepileptic medications
- Decongestants
- Stimulants?



*Grubb & Karas. Pacing and Clin Electrophysiol, 1999

Fludrocortisone for the Prevention of Vasovagal Syncope

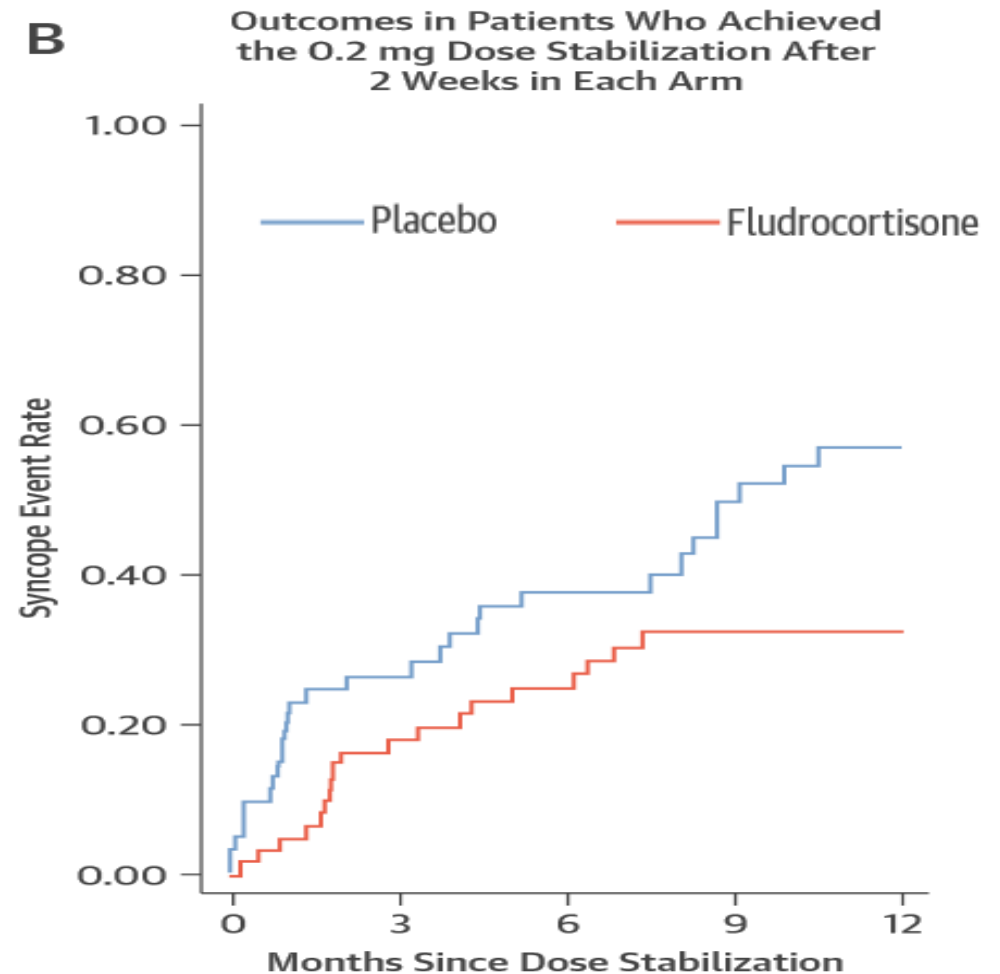


A Randomized, Placebo-Controlled Trial

J Am Coll Cardiol 2016

Robert Sheldon, MD, PhD,^a Satish R. Raj, MD, MScI,^b M. Sarah Rose, PhD,^a Carlos A. Morillo, MD,^c Andrew D. Krahn, MD,^d Eduardo Medina, MD,^e Mario Talajic, MD,^f Teresa Kus, MD, PhD,^g Colette M. Seifer, MD,^h Malgorzata Lelonek, MD, PhD,ⁱ Thomas Klingenhoben, MD,^j Ratika Parkash, MD,^k Debbie Ritchie, MN,^a

- N = 210
- Mean age = 30 years
- 1-year treatment period



$p = 0.019$

September 23, 2014; 83 (13) **ARTICLE**

Midodrine for orthostatic hypotension and recurrent reflex syncope A systematic review

Ariel Izcovich, Carlos González Malla, Matias Manzotti, Hugo Norberto Catalano and Gordon Guyatt

First published August 22, 2014, DOI: <https://doi.org/10.1212/WNL.0000000000000815>

Beta Blockers

- Can be ***great*** for POTS patients with lots of palpitations, sinus tachycardia, anxiety
- Careful!
 - Can make dizziness (thus everything) worse
 - Fatigue
- **Holter monitoring**
 - Average HR, HR range, upright HR
- **Propranolol**, atenolol, nadolol

Day vs. Night HR → “Orthostatic” Tachycardia

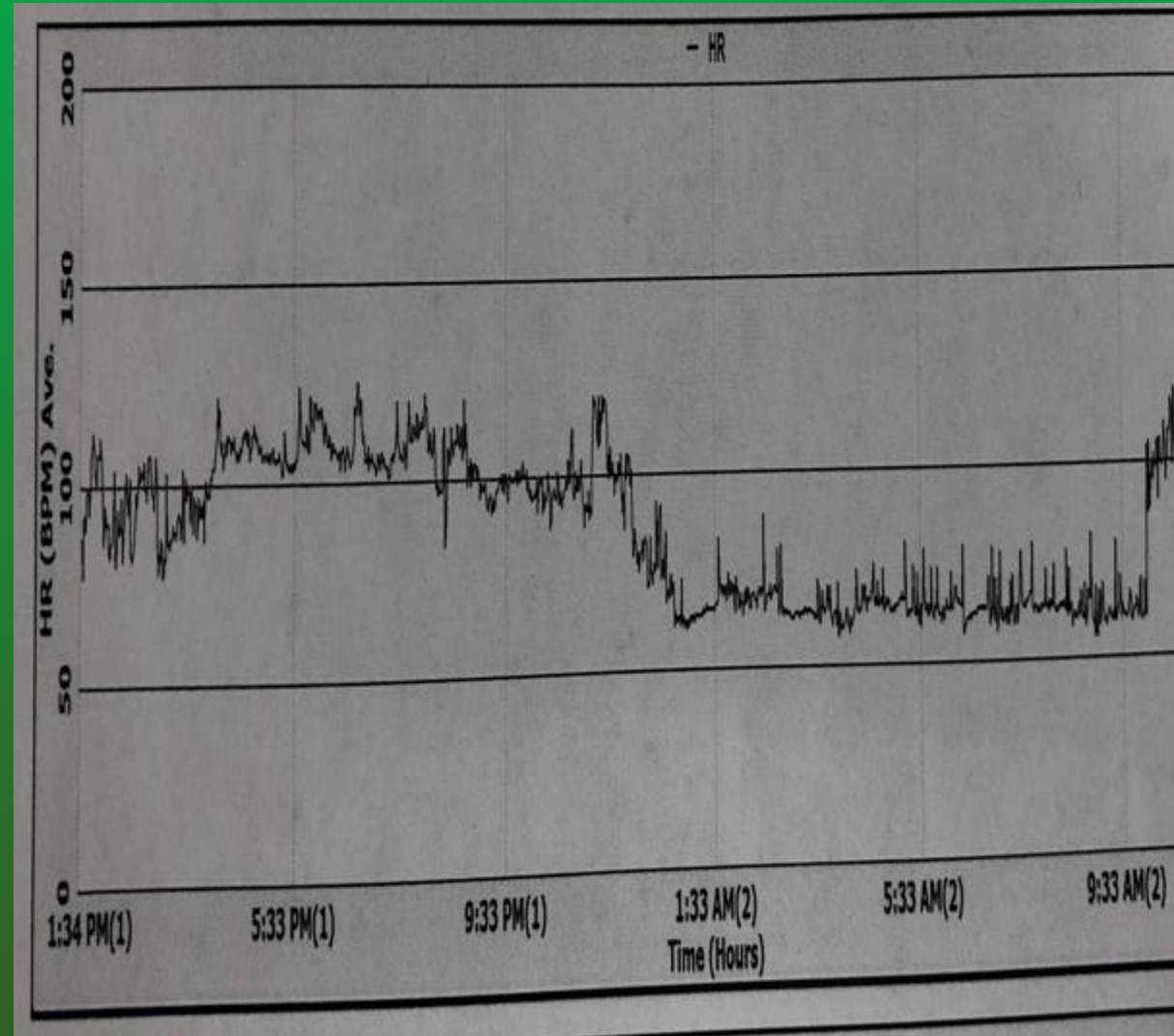


TABLE 2 Medications

Medication	Dose	Side Effects	Comments
Circulatory support			
Fludrocortisone	0.1–0.2 mg qAM	Peripheral edema, acne, headache, hypokalemia, hypomagnesemia	Monitor basic metabolic panel and magnesium at higher doses ^{90,136}
Midodrine	2.5–10 mg TID q4h	Tingling, goosebumps, headache, hypertension	Check supine BP 30–60 min after a dose ^{137–140}
Desmopressin	0.1–0.4 mg BID	Hyponatremia, headache ¹⁴¹	—
Octreotide	25–100 µg subcutaneously BID	Injection site discomfort, diarrhea, thyroid derangement	Decreased gastrointestinal transit time may be beneficial for some patients ^{138,142,143}
Erythropoietin	10 000–20 000 IU subcutaneously weekly	Hypertension, arthralgias	Ensure hematocrit <50%, ensure adequate iron intake ^{144,145}
Acute normal saline infusion	1–2 L intravenous every 5–7 d	Repeated phlebotomy can lead to scarring of veins	Intermittent rescue use may be beneficial in acute management ¹⁴⁶
Ivabradine	2.5–10 mg BID	Bradycardia without hypotension	Inhibits I _f sinoatrial node, FDA approved for adult CHF. Small trials showed benefit in POTS ^{147,148}
Autonomic modulation			
Metoprolol succinate	12.5–100 mg daily	Lightheadedness, decreased exercise tolerance, fatigue, worsening asthma, depression	Nighttime dosing may decrease lightheadedness ^{139,149}
Metoprolol tartrate	12.5–50 mg BID		
Atenolol	12.5–50 mg BID	Same as metoprolol succinate	—
Nebivolol	2.5–10 mg daily	Same as metoprolol succinate	Fewer overall side effects because of decreased blood–brain barrier penetration
Propranolol	—	Same as metoprolol succinate	—
Citalopram	10–40 mg daily	Nausea, headache, fatigue, increased appetite, suicidal ideation requiring early and frequent monitoring	Causes central sympathetic modulation, reduces abnormal autonomic response ¹⁵⁰
Escitalopram	5–20 mg daily	Same as citalopram	—
Sertraline	25–200 mg daily	Same as citalopram	—
Clonidine	0.1–0.3 mg transdermal every 7 d	Contact dermatitis with adhesive, fatigue, dry mouth, headache	Centrally acting α-agonist, may also be used for insomnia ^{151,152}
Pyridostigmine	30–120 mg BID to TID	Abdominal pain, muscle twitch, decreased intestinal transit time	May also be helpful for early satiety and constipation ^{153–155}

BID, twice daily; CHF, congestive heart failure; FDA, Food and Drug Administration; I_f, sinus node inward “funny” pacemaker channel; q4h, every 4 hours; qAM, every morning; TID, thrice daily; —, not applicable.

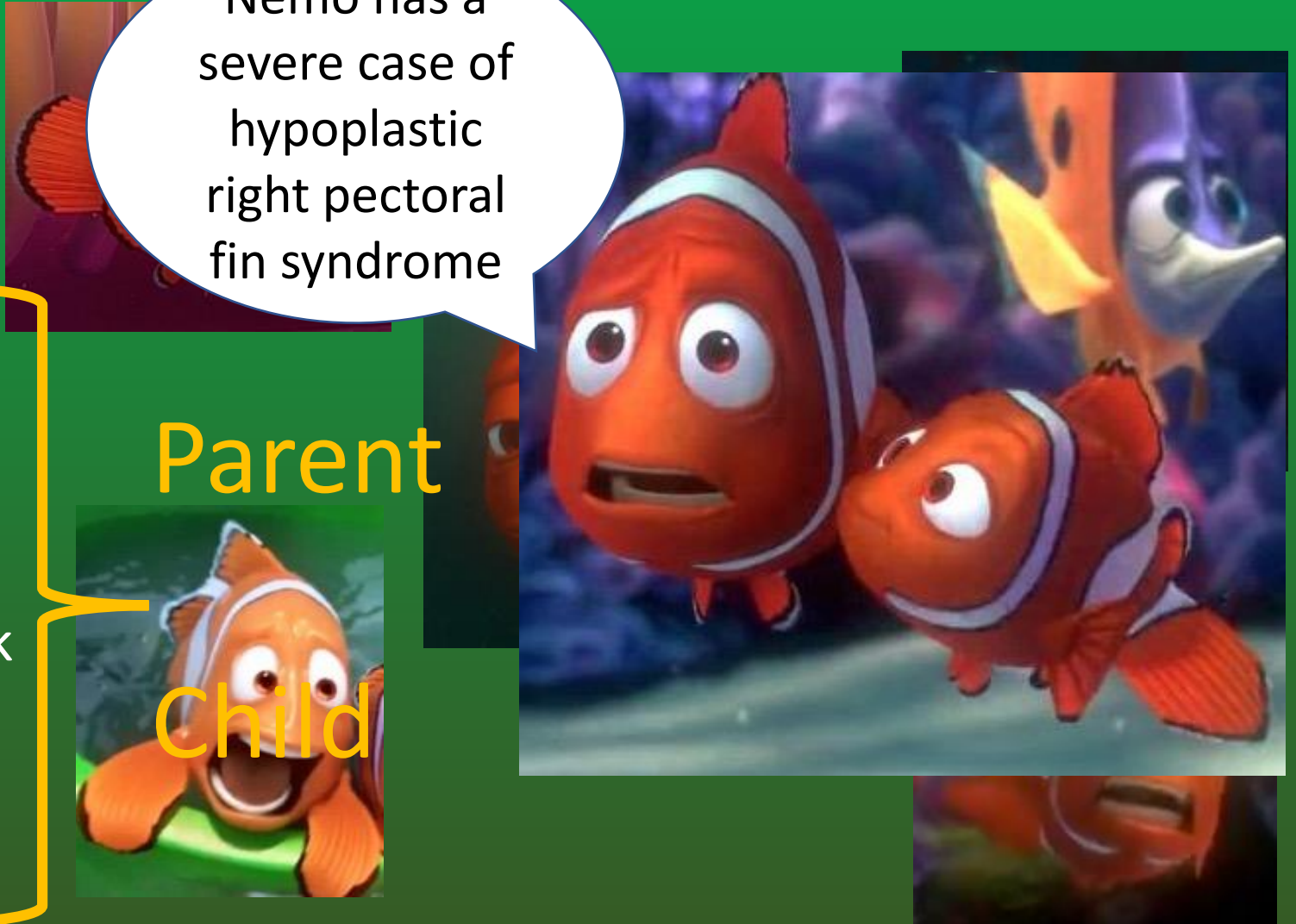
Why do some patients not get better (yet)?

- Super POTS?
- In many . . .
 - Symptom amplification
 - Catastrophizing
 - Somatic hypervigilance
 - Organizational skills (or lack thereof)
 - Depression

Nemo has a severe case of hypoplastic right pectoral fin syndrome

Parent

Child



Positivity, Prognosis, and Coping Skills

- Patients need to hear they will get better
- Literature discussion – Mayo, European studies, UAB experience
- Patients need to know that anxiety and poor coping make things **worse** in the meantime
- Working on this, just like the rest of the plan, is part of taking care of the complete person

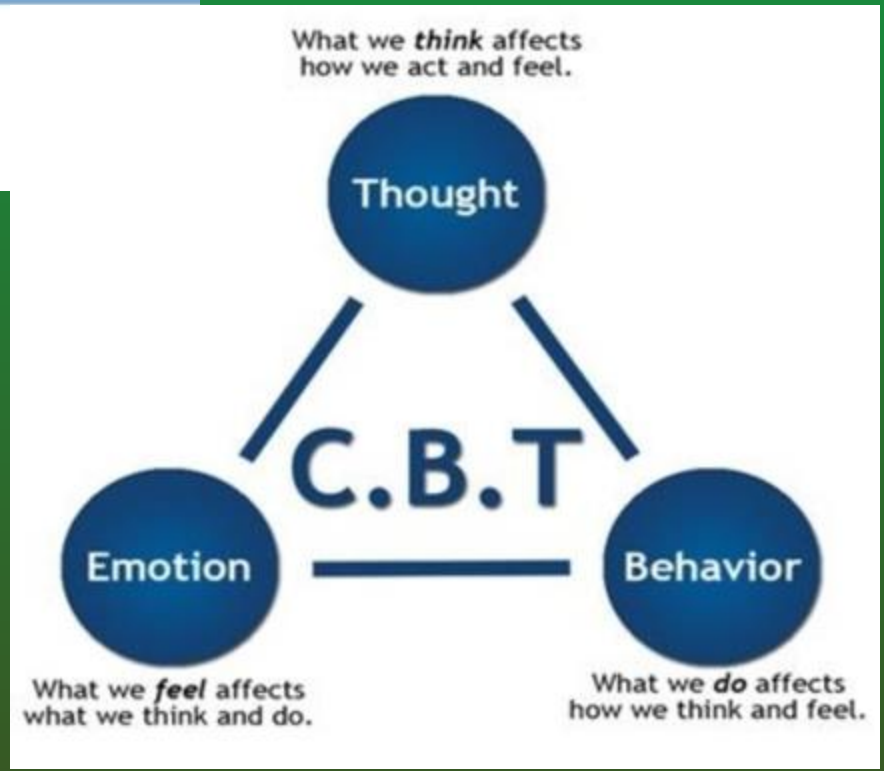




GP Guide
Journal Articles

Cognitive-Behavioural Therapy for Chronic Health Conditions

- Adjusting to the unpredictability of illness
- Improved coping with physiologic sensations
- Activity pacing
- Shaking off “illness identity”
- Managing associated anxiety and/or mood disorders



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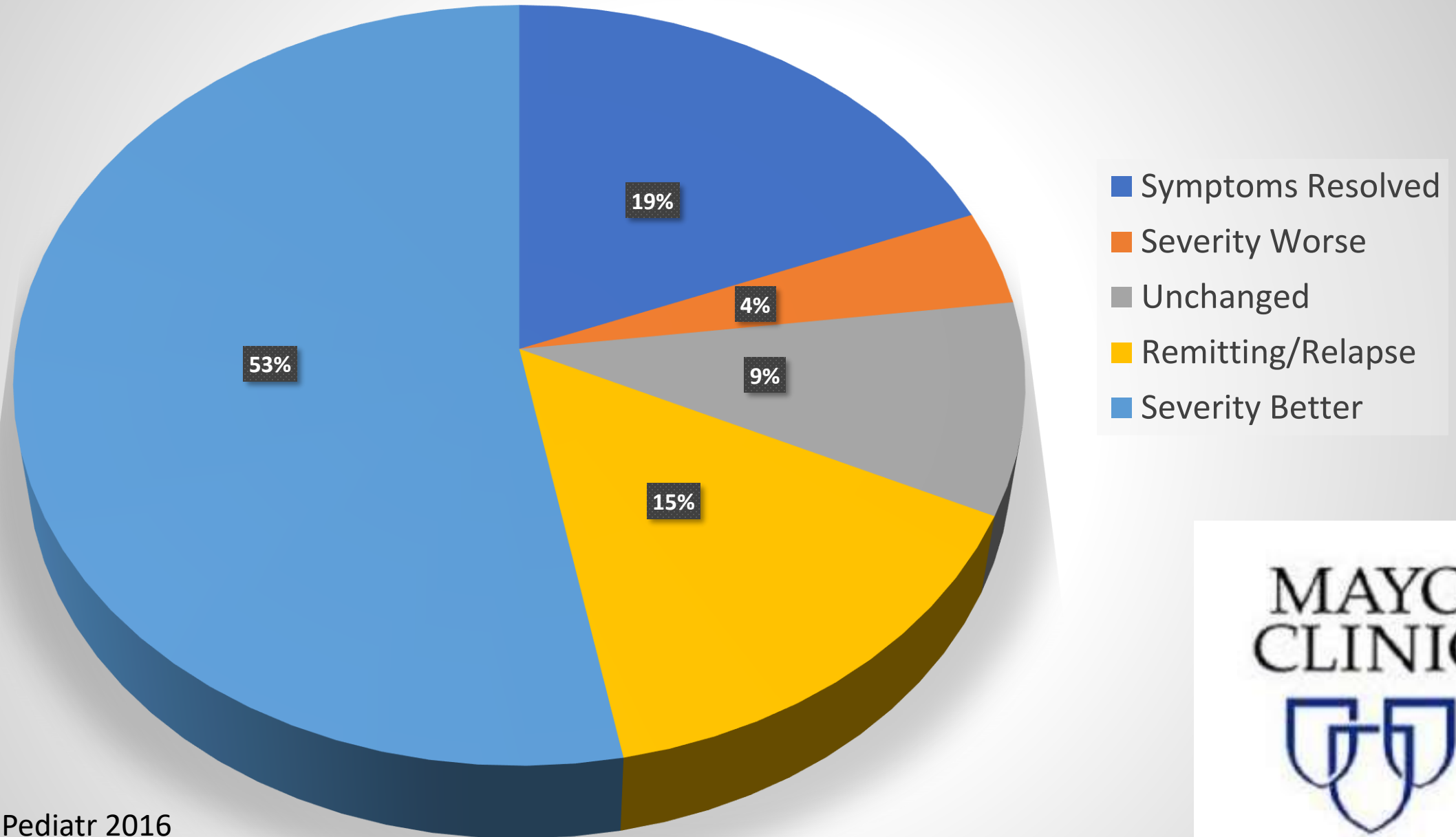


Mayo Clinic Multidisciplinary POTS Clinic

- **Clinic experience** – autonomic testing, PT, CBT, medication management, “pain rehab” program, life coach
- **Survey** of adolescents seen in clinic between 2003-2010 (N = 172)
 - Ages 13-18 at time of initial intake
 - Mean age at time of survey 21.8 years
 - Mean duration from clinic intake to survey = **5.4 years**
 - 84% female
- **Findings**
 - **13%** of patients report no improvement
 - **87%** report symptoms much improved, only intermittent symptoms, or symptoms resolved



Symptom Update at 5 Years - Adolescents with POTS at Mayo Clinic (N = 172)



Intake Description of Adolescent Patients with Significant Orthostatic Intolerance at UAB

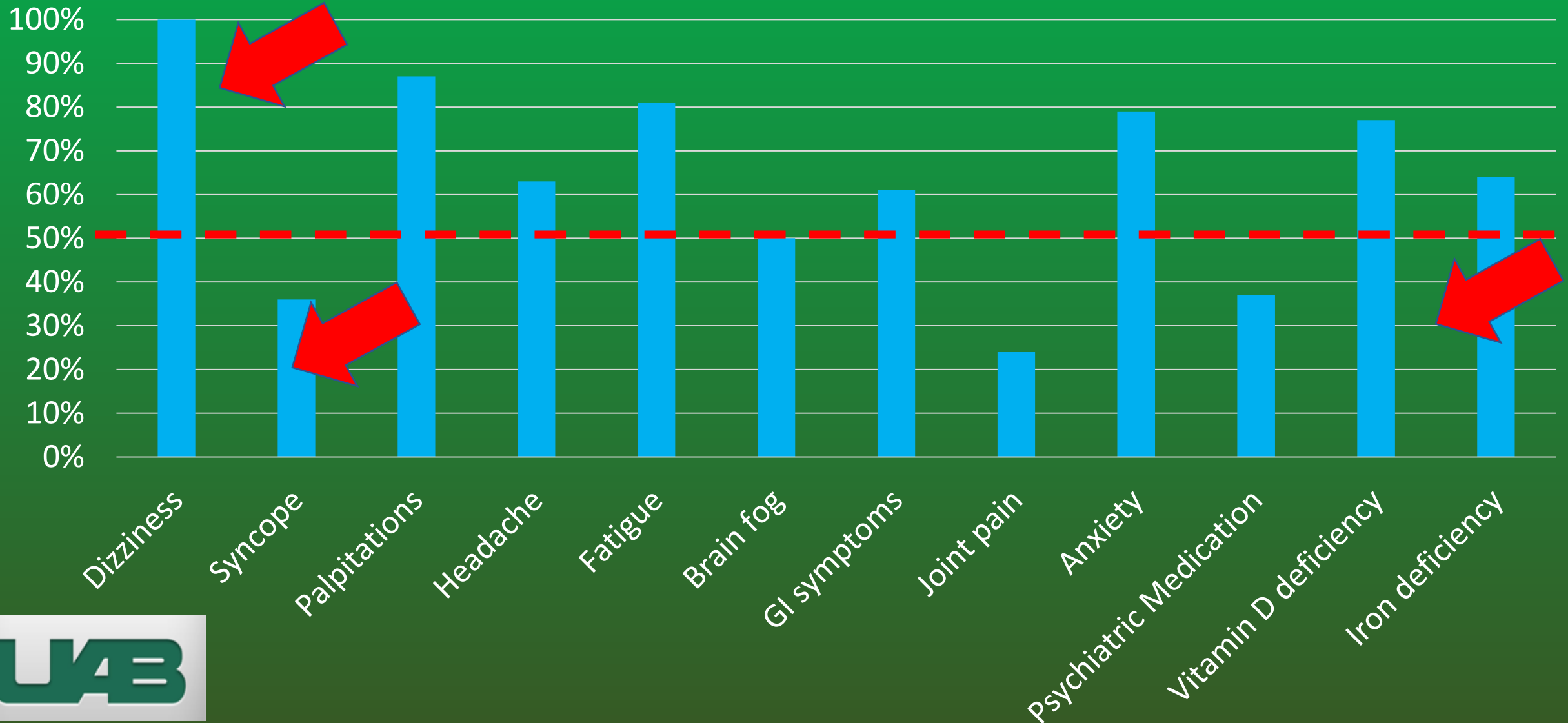


Patient Description (5-11/2019)	N = 70
Age	15.9 +/- 1.4 years
Female	91%
Orthostatic Tachycardia by Vital Signs*	67%
Missing Significant Amount of School / Work	60%
Seen by Other Pediatric Subspecialist for Same Complaints**	66%
Self-Report Quality of Life Rating (out of 10)	5.3 +/- 1.7

*HR increase by at least 30 bpm within 3 minutes of standing. No orthostatic hypotension

**GI, Rheum, Neuro, etc

Symptoms / Characteristics of Adolescent Patients with Significant Orthostatic Intolerance at UAB (N = 70)



Last Clinic Follow-Up: Adolescents With Significant Orthostatic Intolerance at UAB (5/19 – 11/19)

Symptoms Much Improved	64%
Symptoms Mildly Improved	21%
School Resumption	87%
Self-Report Quality of Life Rating (out of 10)	7.4 +/- 1.8
Intake Self-Report QOL Rating (out of 10)	5.3 +/- 1.7
Medications	
Beta-blocker	64%
Fludrocortisone	41%
Midodrine	8%
Salt tablets	67%



} **P < 0.01**

Average time from initial visit to last follow-up: 6.4 months

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Thank you!

