



Practical POTS for the Practical Pediatrician

Pointers to Prepare the Previously Pessimistic Provider to Provide Premium Proclamations to these Periodically Prickly Patients




Camden Hebson, M.D.
Pediatric Cardiology




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Blissfully unaware POTS existed Aware of POTS, dislikes it




Born: Oct 24, 1979
Gadsden, AL



Cam's POTS Journey




2002
Went to medical school




First job at CHOA
Seeing lots of POTS pts

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Alpharetta, GA




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Alpharetta Clinic

Time	Patient	Reason for Visit
8:00	Otterton, Emmitt	POTS 2 nd opinion
8:15	Fitzherbert, Eugene	Syncope
8:30	Hamada, Hiro	Dizziness
8:45	Moana of Motunui	Dizziness and headache
9:00	Darling, Michael	Dysautonomia eval, from Neuro
9:15	Merida of DunBroch	Near syncope, Aunt with POTS
9:30	Von Schweetz, Vanellope	Dizziness, syncope, palpitations
9:45	Callisto, Miles	Tetralogy, but worried about POTS
10:00	Kion the Lion	? POTS
10:30	Apatosaurus, Arlo	Syncope, palpitations
10:45	Turtle, Crush	Cousin with dysautonomia, worried it is contagious

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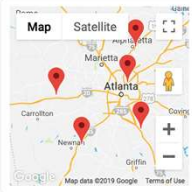


Michael E McConnell, MD
Pediatric Cardiologist

Gender: Male | Language: English

Locations

- Sibley Heart Center Cardiology**
60 Oak Hill Boulevard, Suite 102
Newnan, GA 30265
[Get Directions](#)
404-256-2593
- Sibley Heart Center Cardiology**
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404-256-2593



PRIMARY SPECIALTY
Cardiology

OTHER SPECIALTY
Pediatric Cardiology

EDUCATION
Board Certification:
Am Bd Pediatrics (Sub:
Pediatric Cardiology),
American Board of Pediatrics

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Dr. Mike McConnell

Blissfully unaware POTS existed

enjoys taking care of these patients!

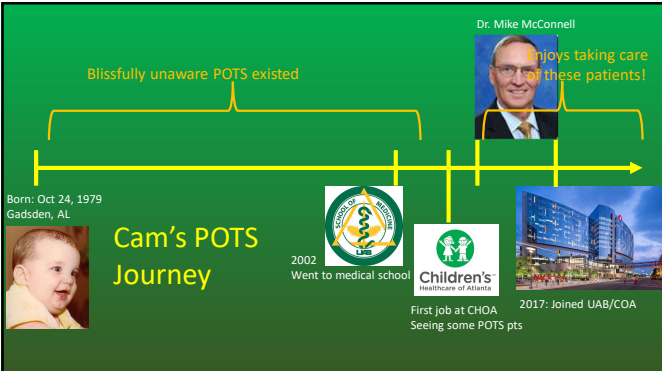
Born: Oct 24, 1979
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Cam's POTS Journey



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Today's Agenda - POTS

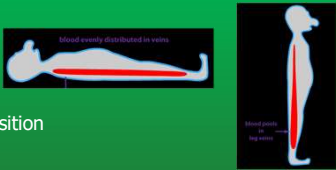
- Definitions and Pathophysiology
- Clinical Pearls
- Treatment Options
- Outcomes



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Definitions

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

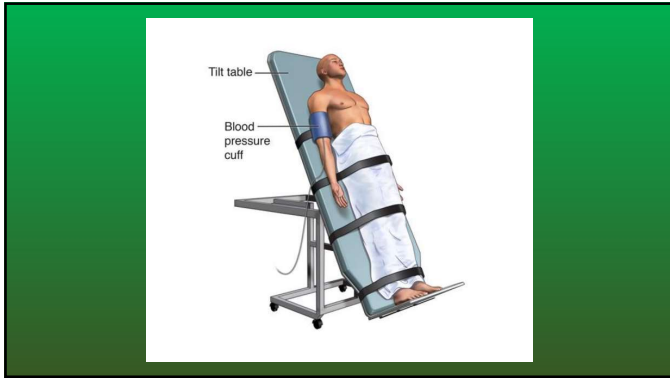


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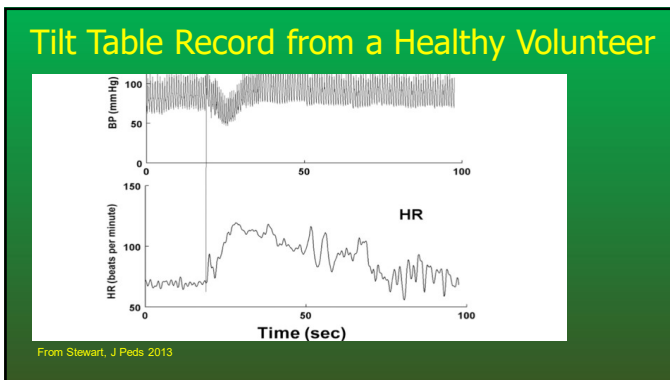
Definitions (2)

- **Orthostatic intolerance**
 - **Symptoms** of lightheadedness, dizziness, blurry vision, etc that occur with assuming upright posture
 - Symptoms resolve with resuming supine position
 - 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: hypovolemia, standing, pain, emotional startle

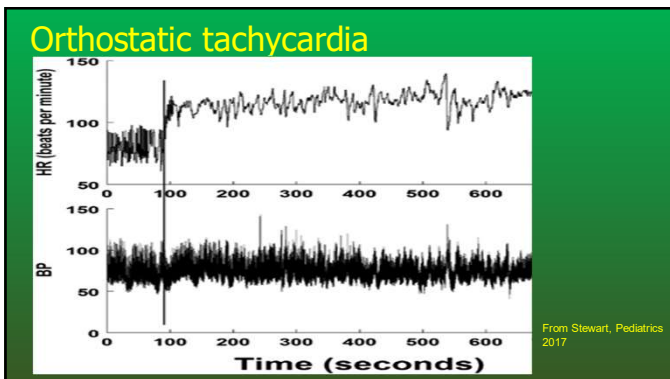
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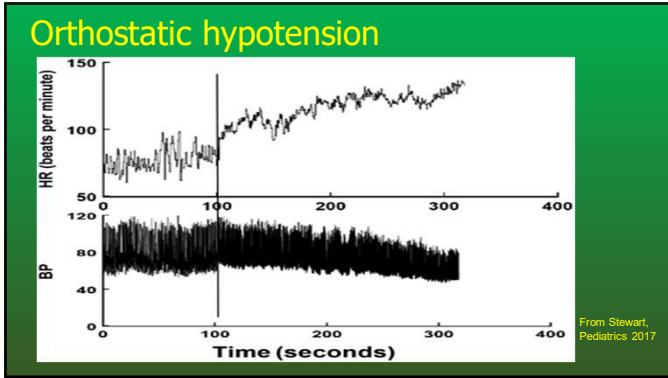
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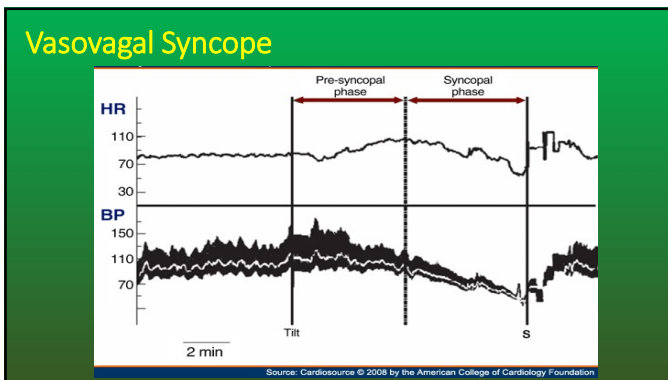
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Postural Orthostatic Tachycardia Syndrome

- 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 testing
 3. Symptoms are **chronic** (> 6 months) and relieved by recumbence
 4. Diagnosis of exclusion
- **Practical Additions**
 - Requiring treatment with medications
 - Symptoms are severe enough that the patient is missing school or work

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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!**

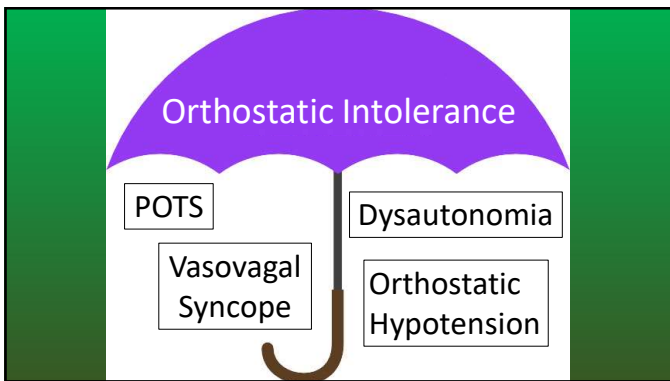
* Brinith L. Auto Neurosc 2018
 ** Sheldon. Heart Rhythm 2015

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Dysautonomia and POTS

- “Sister” diagnoses
- **Dysautonomia**
 - Significant orthostatic intolerance
 - Prominent autonomic symptoms
 - Abnormal sweating
 - Heat/cold intolerance
 - Sleep disturbance
 - Gastroparesis
 - Urinary retention

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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

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Compensatory Responses to Orthostasis

- Without these, we would pass out all the time!

1. Role of arterial baroreceptors
2. Autonomic nervous system
3. Skeletal muscle pump

Figure 1. Location and innervation of arterial baroreceptors.

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Arterial Baroreceptors

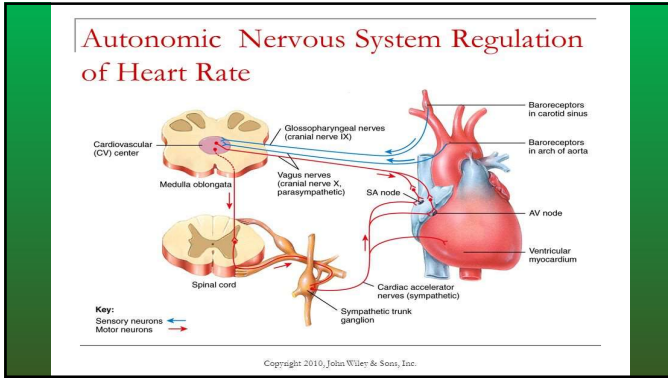
- Located in the aortic arch and carotid sinus
- These receptors respond to arterial wall stretch

→ when BP rises and the walls stretch, baroreceptor firing increases

→ if BP falls, baroreceptor firing decreases

Figure 1. Location and innervation of arterial baroreceptors.

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Autonomic Response to Decrease in BP

- Under **normal conditions**, baroreceptor firing exerts a tonic **inhibitory** influence on sympathetic outflow from the medulla
- Acute hypotension** → decreased baroreceptor firing → SNS activity from the medulla → vasoconstriction, tachycardia, etc. → restoration of BP
- Standing or upright posture actuates this response

BP = CO * SVR

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The Skeletal Muscle Pump

- Skeletal muscle aids the heart in circulating blood through the body by **"pumping" venous blood back to the heart** with muscular contractions
 - Particularly important in the legs
- Importance of exercise in patients with orthostatic intolerance

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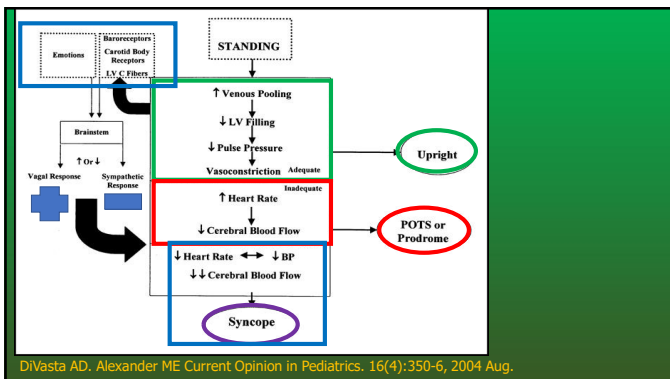
When It All Goes Wrong

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Pathophysiology of Vasovagal Syncope


- Venous pooling results in sudden reduction in blood return to the heart and compensatory SNS activity
- Sympathetic input to the heart can result in overly vigorous cardiac contractions
- These contractions result in stimulation of specialized myocardial C fibers, sending a stimulus BACK to the brainstem that is interpreted as "hypertension"
- Brain stem response (vagal nerve mediated) leads to hypotension and bradycardia, and thus syncope

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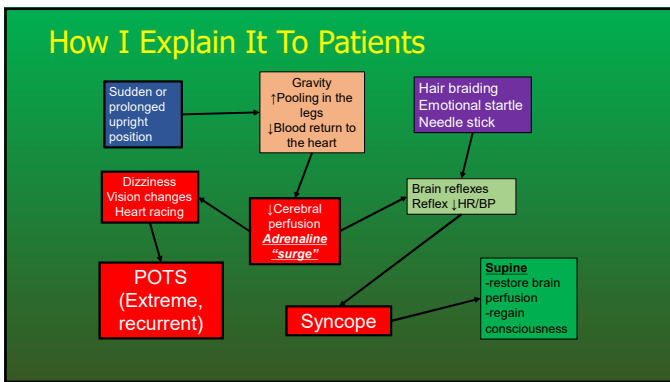


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Did you just say "LV C-fiber stimulation"?



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Left Ventricular Geometry and Function Preceding Neurally Mediated Syncope

Liu, et al Circulation 2000; 101:777-783

	Supine			Upright		
	Negative Tilt	Positive Tilt	P	Negative Tilt	Positive Tilt	P
EDV, mL/m ²	83±12	83±14	NS	80±14	48±13	0.05
SV, mL	26±8	28±11	NS	24±7	23±11	0.70
SV, mL	88±19	83±18	NS	86±22	44±13	0.01
CI, mL/min per m ²	0.23±0.06	0.30±0.08	NS	0.22±0.06	0.25±0.09	0.26
MWS	0.18±0.03	0.17±0.03	NS	0.17±0.04	0.14±0.04	0.07
CI, mL/min per m ²	2174±395	2724±900	NS	2364±683	1964±483	0.14
EF	0.58±0.09	0.57±0.11	NS	0.58±0.09	0.54±0.14	0.36
HRSS, sphygmoman ²	94±38	101±53	NS	85±39	74±26	0.27
HRSS, sphygmoman ³	177±69	190±53	NS	163±50	142±44	0.33
SAR, dynes · s · cm ⁻⁴	1697±388	1596±484	NS	1819±456	1993±716	0.20
Stroke (LVEDV-corrected) FS, %	119±13	122±18	NS	117±23	94±19	0.05
Stroke (LVEDV-corrected) MWS, %	116±14	107±15	NS	109±29	94±17	0.03

EDV indicates end-diastolic volume indexed to body surface area; ESA, end-systolic volume indexed to body surface area; SV, stroke volume; MWS, mitral annular shortening; CI, cardiac index; EF, ejection fraction; and SAR, systemic vascular resistance.

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Left Ventricular Geometry and Function Preceding Neurally Mediated Syncope

Liu, et al Circulation 2000; 101:777-783

N = 21	Supine			Upright		
	Negative Tilt	Positive Tilt	P	Negative Tilt	Positive Tilt	P
Stroke Volume (mL)	68 ± 18	63 ± 18	NS	66 ± 22	44 ± 13	0.01

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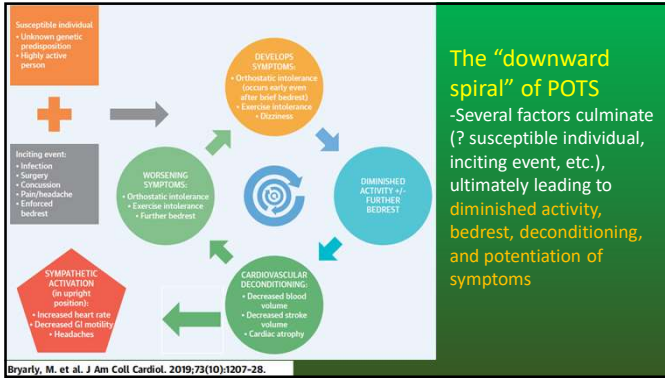
Left Ventricular Geometry and Function Preceding Neurally Mediated Syncope

Liu, et al Circulation 2000; 101:777-783

N = 21	Supine			Upright		
	Negative Tilt	Positive Tilt	P	Negative Tilt	Positive Tilt	P
Stroke Volume (mL)	68 ± 18	63 ± 18	NS	66 ± 22	44 ± 13	0.01

-Sustained decreased venous return to the heart → hypovolemia, excessive venous pooling in the legs, etc.
 -Sustained decreased stroke volume
 -Sustained decrease in cerebral perfusion → **symptoms**

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Today’s Agenda - POTS

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

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Assessment of the Patient with Orthostatic Intolerance

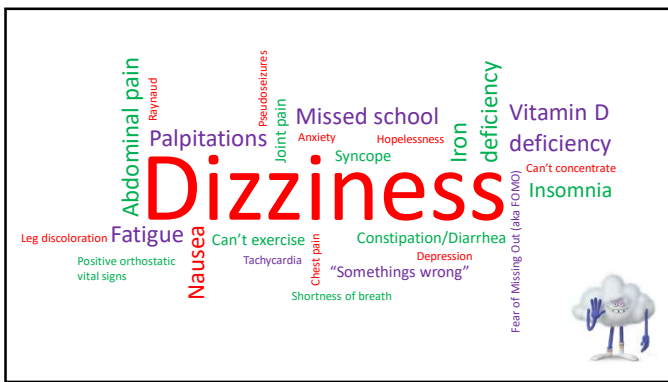
The best “test” is a detailed medical history

- 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?

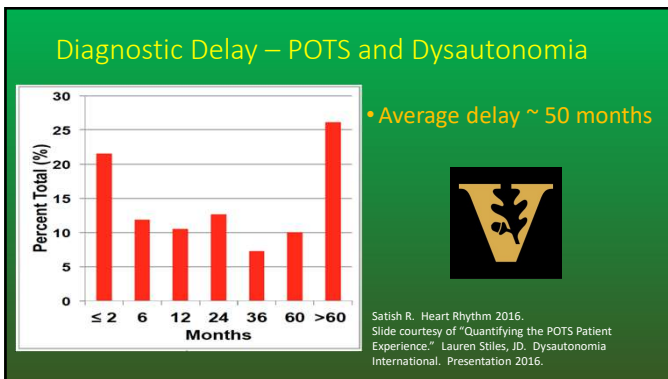
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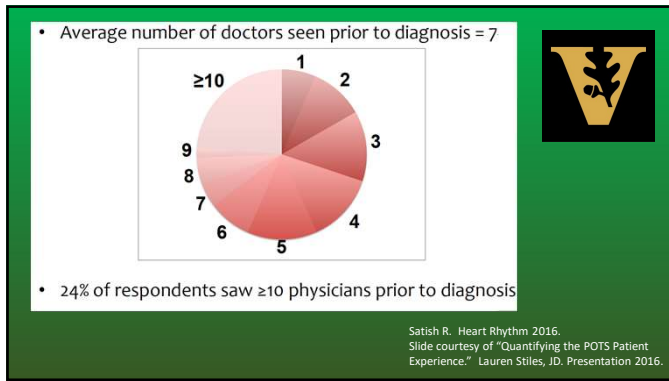
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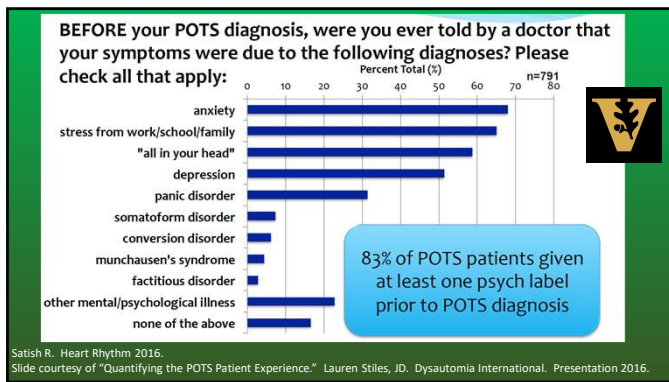
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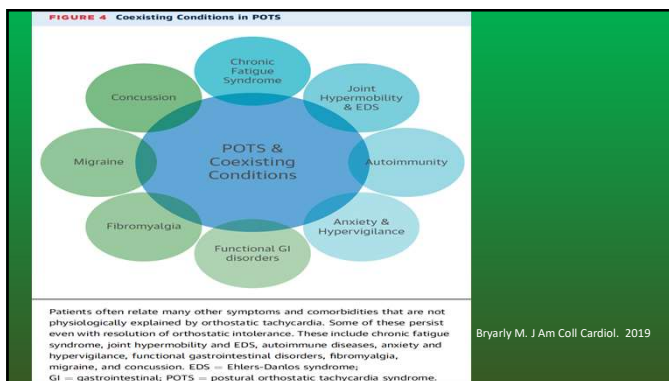
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Vasovagal syncope

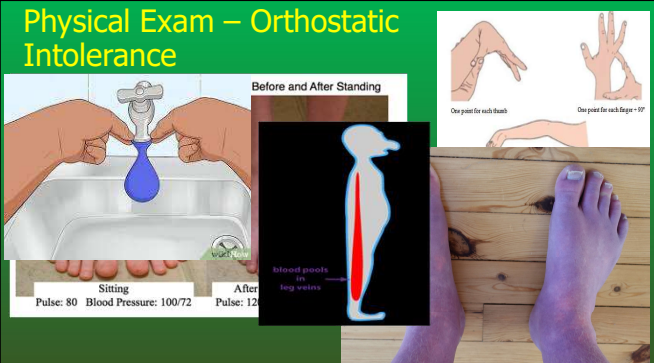
- NOT so prominent a symptom in POTS patients
- Only described in ~30% of patients with chronic symptoms related to POTS*
- Lots of syncope?



*Shen. JACC 2017

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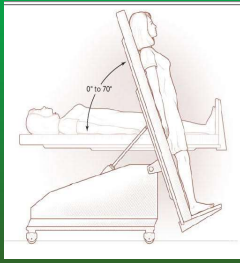
Physical Exam – Orthostatic Intolerance



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Tilt Table Testing

- De facto standard for orthostatic stress testing
- Patient is strapped snugly to the table → limits skeletal muscle pump, therefore more autonomic "stress"
- **Not routinely used**
 - Sensitivity and specificity less than perfect
 - Cumbersome
 - Orthostatic vital signs instead
- Can be useful if a **primary psychiatric cause** is being considered
 - Controlled setting
 - Syncope w/o hypotension (SBP < 60-65 mmHg) or bradycardia (HR < 40 bpm or asystole)



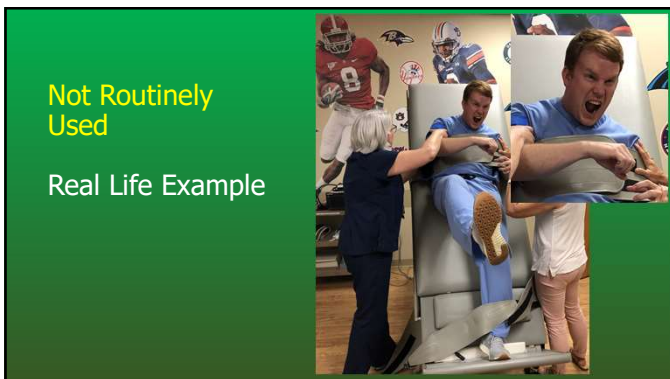
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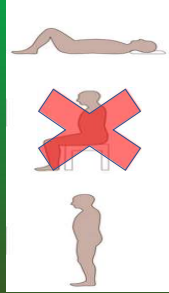


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Orthostatic Vitals – 3 Minute Standing Test

• 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



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3 Minute Standing Test

- Same unruly patient from earlier



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Lab Testing for POTS


- Vitamin D deficiency
- Iron deficiency



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Vitamin D deficiency

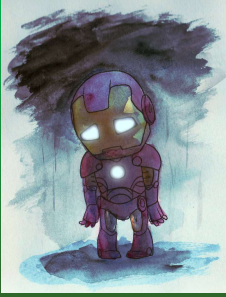
- Associated with **orthostatic hypotension** in adults^{1,2}
 - Association in children has not been well-defined
- Also associated with **depression and anxiety**³⁻¹⁰ → frequent cohabitants!
- Goal:** 25-Vitamin D > 40 ng/mL
- Dose:** Vitamin D3 4000 IU daily in adults, 1000 IU per 25 lbs in kids



- Anweiler C. J Intern Med 2014
- McCarroll K. Age Ageing 2012
- Parker G. J Affect Dis 2017
- Anglin R. J Psych 2013
- Balton C. Neurology 2012
- Wu C. Medicine 2016
- Huang J. J Women Health 2014
- Armstrong D. Clin Rheum 2007
- Kelley L. J Dev Orig Health 2016
- Bickova M. Phys Res 2015

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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

- Jarjour IT. Clin Auton Res, 2013
- Low PA. Curr Opin Neurol, 1994

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Today's Agenda - POTS

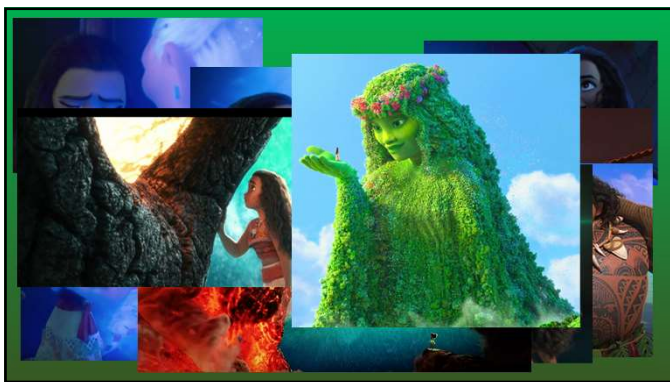
- Definitions and Pathophysiology
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POTS often can be managed by nonpharmacological measures alone. These strategies can help increase blood volume and minimize orthostatic symptoms. In addition to boosting blood volume, an exercise regimen geared toward 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

Avoid Situations That Can Exacerbate Symptoms 	Liberal Intake of Salt and Water 	Sleep With Head of Bed Elevated <p>Head posts should be elevated 4-6 inches</p>
Use of Compression Garments 	Physical Counter Maneuvers 	Drinking Water Before Getting Up in The Morning <p>Drinking a 16 oz glass of water daily before getting out of bed in the morning or mimicking standing to minimize orthostatic symptoms</p>
Strategies to Avoid Upright Exercise		
<p>Seated Rower</p>	<p>Swimming</p>	<p>Recumbent Bicycle</p>

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Treatment of Orthostatic Intolerance

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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	VERY GOOD
2	GOOD
3	FAIR
4	LIGHT DEHYDRATED
5	DEHYDRATED
6	VERY DEHYDRATED
7	SEVERE DEHYDRATED

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

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

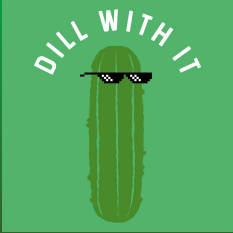
- Increasing salt intake leads to increased fluid retention
- Focus on Sodium
- 3-5 g Na/day if severe symptoms**
- Will not cause weight gain
- Hypertension?
- Salt tablets
- Literature support – symptom improvement*

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Na
Sodium
22,990

*Raj SR, Circulation, 2013

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High Salt Diet (3000-5000 mg Na/day)


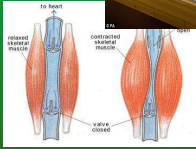


Food Item	Mg of Sodium (Google)
Bacon (3 slices)	390
Table salt (1 tsp)	2300
Pretzel snack	1400
Salted nuts (1/2 cup)	420
Hamburger	690
Salt tablet	250 mg / tablet
Dill pickle	1430
Soy sauce (1 tbsp.)	870
Beef Jerky	420

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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

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

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Article

Improvement in Functioning and Psychological Distress in Adolescents With Postural Orthostatic Tachycardia Syndrome Following Interdisciplinary Treatment

Clinical Pediatrics
2016, Vol 55(16) 1300-1304
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DOI: 10.1177/0279929816638663
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Postural Orthostatic Tachycardia Syndrome

Exercise Training Versus Propranolol in the Treatment of the Postural Orthostatic Tachycardia Syndrome

Qi Fu, Tiffany B. VanGundy, Shigeki Shibata, Richard J. Auchus, Gordon H. Williams, Benjamin D. Levine

See Editorial Commentary, pp 136-137

Abstract—We have found recently that exercise training is effective in the treatment of the postural orthostatic tachycardia syndrome (POTS). Whether this non-drug treatment is superior to “standard” drug therapies, such as β -blockade, is unknown. We tested the hypothesis that exercise training but not β -blockade treatment improves symptoms, hemodynamics, and renal-adrenal responses in POTS patients. Nineteen patients (18 women and 1 man) completed a double-blind drug trial (propranolol or placebo) for 4 weeks, followed by 3 months of exercise training. Fifteen

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Cardiovascular exercise as a treatment of postural orthostatic tachycardia syndrome: A pragmatic treatment trial

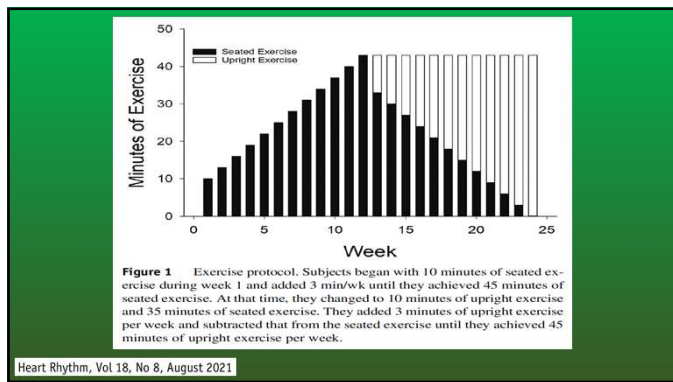
Heart Rhythm, Vol 18, No 8, August 2021

Christopher H. Gibbons, MD, MMSc, Gustavo Silva, MD, Roy Freeman, MD

From the Department of Neurology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, Massachusetts.

- Retrospective review of quality improvement project – **unsupervised home exercise training program** designed for program's POTS patients
- Gradual, 40 minute **seated** → **upright** physical activity.
 - Recumbent bike, rowing machine, swimming, floor exercise
- **N = 48 participants vs. N = 29 control patients** (with POTS who received program's "typical" care – fluids/salt/medicines).
 - No medicines for the exercise group!
- Orthostatic vital signs + patient survey (quality of life) results at **onset + 6 months** into training

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Table 1 Demographic characteristics of the study participants

Characteristic	Treated group: baseline (n = 48)	Treated group: 6-mo follow-up	Control group: baseline (n = 29)	Control group: follow-up
Age (y)	26.3 ± 6.6		26.0 ± 5.3	
Sex	43 F (90%), 5 M (10%)		26 F (90%), 3 M (10%)	
BMI (kg/m ²)	25.3 ± 1.9	24.6 ± 1.4	25.1 ± 1.8	25.2 ± 1.9
POTS duration (y)	4.4 ± 1.9		4.6 ± 2.0	
Supine heart rate (beats/min)	80 ± 11	68 ± 8*	78 ± 10	77 ± 10 [†]
Standing HR (beats/min)	116 ± 11	95 ± 11*	115 ± 10	115 ± 10 [†]
Postural change in HR (beats/min)	36 ± 9	27 ± 7*	37 ± 6	38 ± 7 [†]
Supine SBP (mm Hg)	110 ± 11	100 ± 12	110 ± 10	111 ± 11
Supine DBP (mm Hg)	68 ± 10	69 ± 10	68 ± 11	68 ± 12
Standing SBP (mm Hg)	109 ± 11	107 ± 12	110 ± 11	109 ± 11
Standing DBP (mm Hg)	71 ± 9	69 ± 8	72 ± 9	72 ± 10
HR variability to paced breathing (beats/min)	26 ± 7	26 ± 6	26 ± 6	27 ± 5
Valsalva ratio	2.15 ± 0.40	2.18 ± 0.39	2.13 ± 0.43	2.15 ± 0.42

Values are presented as mean ± SD or number (%).
 No significant differences were noted in baseline characteristics between the treated and control groups.
 BMI = body mass index; DBP = diastolic blood pressure; F = female; HR = heart rate; M = male; POTS = postural orthostatic tachycardia syndrome; SBP = systolic blood pressure.
 *P < .01 compared to baseline testing with the same group.
 †P < .001 treatment group vs control group. Within- and between-group comparisons using the paired tests (within group) and unpaired t tests (between group) with Bonferroni corrections for multiple comparisons. HR variability and Valsalva ratio included all individuals at baseline, 38 completers in the exercise group and 12 completers in the no-exercise group.

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Syncope

Symptom scores

The frequency of reported syncope or near-syncope was similar between groups at baseline. There was a significant decrease in the frequency of syncope in the treated group (median [interquartile range] 0 [0-2] events per 21 months in the treated group vs 3 [2-4] events per 21 months in the control group after 6 months; $P < .001$) but not in the control group (8 ± 2 vs 5 ± 2 after 6 months; $P < .001$) but not in the control group (8 ± 2 vs 8 ± 2 after 6 months; $P = .86$). The differences between groups were significant.

There was an improvement in the EuroQol perceived quality of life scale score in the exercise group (61 ± 15 vs 71 ± 12 after 6 months; $P < .001$) compared with the control group (64 ± 9 vs 66 ± 8 after 6 months; $P = .52$). There were

Heart Rhythm, Vol 18, No 8, August 2021

Figure 4 Symptom scores. The symptom scores for both groups are shown in a radial plot. The symptom scores are normalized to maximum and minimum values, with more severe symptoms/higher scores on the outside of the plot. The control group, shown in the blue plot, is largely unchanged from the baseline data. The exercise group, shown in the red plot, is significantly improved in all measures except the Krone Fatigue Severity Scale. * $P < .01$ between groups.

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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

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doi:10.1016/j.jacc.2006.06.019

Syncope

Effectiveness of Physical Counterpressure Maneuvers in Preventing Vasovagal Syncope

The Physical Counterpressure Manoeuvres Trial (PC-Trial)

Nyke van Dijk, MD,* Fabio Quartieri, MD,† Jean-Jaques Blanc, MD,‡ Roberto Garcia-Civera, MD,§ Michele Brignole, MD,¶ Angel Moya, MD,¶ Wooster Wieling, MD, PhD,* on behalf of the PC-Trial Investigators.
Amsterdam, the Netherlands; Reggio Emilia and Lavagna, Italy; Brest Cedex, France; and Valencia and Barcelona, Spain

OBJECTIVES In this study, we assessed the effectiveness of physical counterpressure maneuvers (PCM) in daily life.

BACKGROUND There is presently no evidence-based therapy for vasovagal syncope. Current treatment consists of explanation and life-style advice. Physical counterpressure maneuvers have been shown to raise blood pressure and to control or abort vasovagal episodes in laboratory.

CONCLUSIONS Physical counterpressure maneuvers are a risk-free, effective, and low-cost treatment method in patients with vasovagal syncope and recognizable prodromal symptoms, and should be advised as first-line treatment in patients presenting with vasovagal syncope with prodromal symptoms. (The PC-Trial; <http://www.controlled-trials.com/iscrt/trial/45146526/0/45146526.html>; ISRCTN45146526) (J Am Coll Cardiol 2006;48:1652-7) © 2006 by the American College of Cardiology Foundation

advised as first-line treatment in patients presenting with vasovagal syncope with prodromal symptoms. (The PC-Trial; <http://www.controlled-trials.com/iscrt/trial/45146526/0/45146526.html>; ISRCTN45146526) (J Am Coll Cardiol 2006;48:1652-7) © 2006 by the American College of Cardiology Foundation

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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



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Lab Testing - POTS

- Vitamin D deficiency
- Iron deficiency



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Medications to Avoid*

- Vasodilators
 - ACEI, CCB
- Many medications used for "migraines" or chronic pain
- Diuretics
- Opiates
- Antiepileptic medications
- Decongestants
- Stimulants?



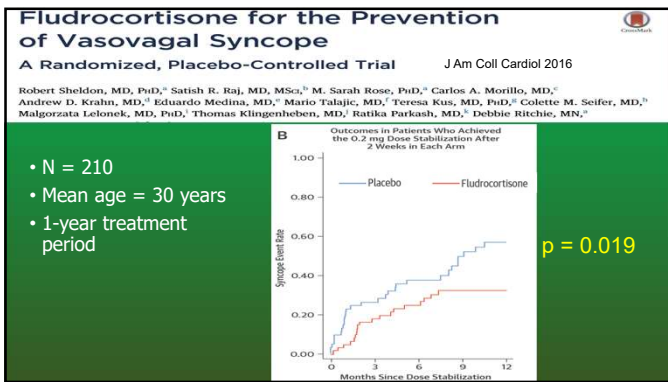
*Grubb & Karas. Pacing and Clin Electrophysiol, 1999

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TABLE 1 Neuropsychiatric medications commonly used in the treatment of dysautonomia-related symptoms with pertinent side effects

Generic (brand) name	Pertinent side effects
Amisulpride (Eli Lilly)	Hypotension, syncope, drowsiness, dizziness, tachycardia, insomnia, anxiety, and weight gain
Aripiprazole (Astellis)	Hypotension, syncope, dizziness, and headache
Atomoxetine (Eli Lilly)	Nausea, orthostatic hypotension, syncope, tachycardia, headaches, abdominal pain, anorexia, dizziness, constipation, and palpitations
Bupropion (Wellbutrin)	Headache, dizziness, anxiety, and tachycardia
Bupropion (Bupropion)	Dizziness, headaches, abdominal pain, and fatigue
Citalopram (Celexa)	Fatigue, anorexia, anxiety, abdominal pain, and dizziness
Clonazepam (Klonopin)	Tachycardia, syncope, drowsiness, dizziness, fatigue, constipation, incontinence, and hypotension
Clonidine (Catapres)	Hypotension, syncope, headache, abdominal pain, and fatigue
Desmethylphenidate (Focalin)	Anorexia, headache, anxiety, dizziness, weight loss, and blood pressure changes
Desoxyn (amphetamine/amphetamine) (Adderall)	Anorexia, abdominal pain, weight loss, anxiety, dizziness, nervousness, diarrhea, fatigue, tachycardia, and palpitations
Duloxetine (Cymbalta)	Orthostatic hypotension, syncope, headache, dizziness, and anxiety
Escitalopram (Lexapro)	Headaches, fatigue, dizziness, and abdominal pain
Fluoxetine (Prozac)	Nausea, headache, anxiety, dizziness, constipation, and vomiting
Flvoxamine (Luvox)	Nausea, headache, insomnia, dizziness, nervousness, and constipation
Gabapentin (Neurontin)	Dizziness, fatigue, nausea, vomiting, diarrhea, constipation, headache, weight gain, and depression
Guafacine (Tenesse)	Dizziness, constipation, fatigue, headache, syncope, and bradycardia
Lexapro (Escitalopram)	Dizziness, headache, and anxiety
Lisdexamfetamine (Vyvanse)	Abdominal pain, nausea and vomiting, diarrhea, tachycardia, anxiety, and dizziness
Lithium	Bradycardia, syncope, vomiting, diarrhea, drowsiness, and fatigue
Methylphenidate (Concerta)	Tachycardia, headache, and dizziness
Oxcarbazepine (Trileptal)	Dizziness, headache, nausea, vomiting, somnolence, diarrhea, constipation, and nervousness
Risperidone (Risperdal)	Hypotension, syncope, somnolence, fatigue, nausea, vomiting, constipation, abdominal pain, anxiety, dizziness, and headache
Sertraline (Zoloft)	Nausea, fatigue, dizziness, palpitations, abdominal pain, and headache
Venlafaxine (Effexor)	Arrhythmias, headache, dizziness, anorexia, nervousness, anxiety, and agitation
Ziprasidone (Geodon)	Somnolence, headache, dizziness, anxiety, tachycardia, and orthostatic hypotension

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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>

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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

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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 ^{10,126}
Midodrine	2.5-10 mg TID q4h	Tingling, goosebumps, headache, hypertension	Check supine BP 30-60 min after a dose ^{10,126}
Dramopressin	0.1-0.4 mg BID	Hypnatremia, headache ¹⁴¹	
Oxetropide	25-100 µg subcutaneously BID	Injection site discomfort, diarrhea, thyroid derangement	Decreased gastrointestinal transit time may be beneficial for some patients ^{128,142,143}
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 _s 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 ^{129,149}
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 escitalopram	—
Clonidine	0.1-0.5 mg transdermal every 7 d	Contact dermatitis with adhesive, fatigue, dry mouth, headache	Centrally acting α ₂ -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

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Why do some patients not get better (yet)?

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

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Advocacy / Empowerment → Disease Identity

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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

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POTS UK
POSTURAL TACHYCARDIA SYNDROME

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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Today's Agenda - POTS


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



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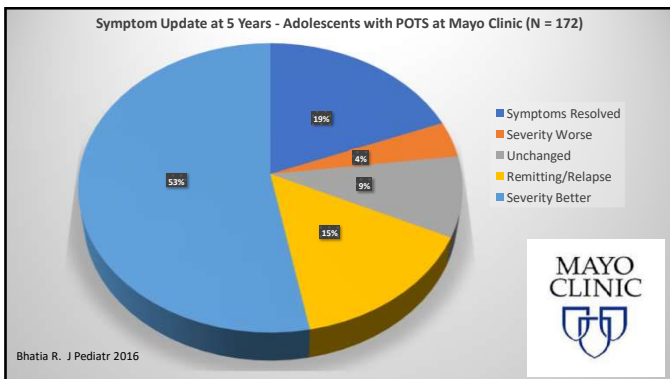
Mayo Clinic Multidisciplinary POTS Clinic

- **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



Bhatia R. J Pediatr 2016

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COA/UAB Pediatric Cardiology "Syncope Clinic"

- Patients referred to UAB Pediatric Cardiology Clinic
- Specific patients identified based on (1) pediatrician's concerns for POTS/dysautonomia, (2) seen by other cardiologist or subspecialist, or (3) specific family concern for the diagnosis
- Clinic set-up
 - 45 minute initial clinic slot
 - Intake and Return surveys
 - Orthostatic vital signs – 3 minute standing test



Children's of Alabama



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Children's of Alabama LUM MEDICINE PEDIATRICS		
POTS/Syncope Clinic (Dr. Hebson) - Initial Visit		
Name/Nickname: _____	Date of birth: _____	
Today's Date: _____		
Describe your main concern today: _____		
Have you previously seen a cardiologist (for this or other issues)? Who and when? _____		
Past medical history (Birth history, illnesses, surgeries): _____		
Medications: _____		
Any family history of heart disease in young people? _____		
Who all lives at home with the patient? _____		
List sports, hobbies, and interests or activities: _____		
Parents names and occupations: _____		
Review of Systems (Check mark if positive, otherwise leave blank)		
Other cardiac concerns: _____	Heart murmur: _____	Easy fatigue: _____
Muscle problems: _____	Spoken/hoarse throat: _____	Hearing loss: _____
Nausea/vomiting: _____	Stomach/constipation: _____	Asthma: _____
Frequent dizziness: _____	Frequent rashes: _____	Diabetes: _____
Easy bruising/bleeding: _____	Thyroid disease: _____	Seizures: _____
Anxiety/Depression: _____	Weight loss: _____	Swelling: _____
Fever/Chills: _____	Poor growth: _____	Joint pain: _____
Genetic disorder: _____	Muscle pain: _____	Other: _____

20 These questions help direct the clinic visit - please answer as accurately as able. Skip any that do not apply.

1	How often do you get dizzy? How often compared to previous?
2	How many times have you passed out in the last month 2 weeks?
3	How often do you have palpitations? Is this new?
4	How much weight have you gained or lost in the last month 2 weeks?
5	How often do you have any of the following symptoms? - Heart racing - Lightheadedness - Head spinning - Blurred vision - Sweating - Nausea - Dizziness
6	How much water do you drink a day? (Use cups, half or full)
7	Do you eat breakfast? (Specify) What is your typical meal?
8	Do you drink alcohol/caffeine? How often?
9	What is your current exercise regimen?
10	Are you taking any medications? How much?
11	Are you taking any supplements? How much?
12	Do you have any other symptoms?
13	Do you have any other concerns?

Office Use Only - I have reviewed the information provided above.
Signature: _____ Date: _____ 1/18

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Children's of Alabama LUM MEDICINE PEDIATRICS	
POTS/Syncope Clinic (Dr. Hebson) - Return Visit	
Name/Nickname: _____	Date of birth: _____
Today's Date: _____	
Describe your main concern today: _____	
How are you compared to previous? (Give some detail) _____	
Is there anything you want to discuss specifically today? _____	
Medications: _____	
Any side effects with medications? _____	
Are any recommended consults, appointments, or treatments pending? Which ones? _____	

21 These questions help direct the clinic visit - please answer as accurately as able. Skip any that do not apply.

1	How often do you get dizzy? How often compared to previous?
2	How many times have you passed out in the last month 2 weeks?
3	How often do you have palpitations? Is this new?
4	How much weight have you gained or lost in the last month 2 weeks?
5	How often do you have any of the following symptoms? - Heart racing - Lightheadedness - Head spinning - Blurred vision - Sweating - Nausea - Dizziness
6	How much water do you drink a day? (Use cups, half or full)
7	Do you eat breakfast? (Specify) What is your typical meal?
8	Do you drink alcohol/caffeine? How often?
9	What is your current exercise regimen?
10	Are you taking any medications? How much?
11	Are you taking any supplements? How much?
12	Do you have any other symptoms?
13	Do you have any other concerns?

Office Use Only - I have reviewed the information provided above.
Signature: _____ Date: _____ 1/18

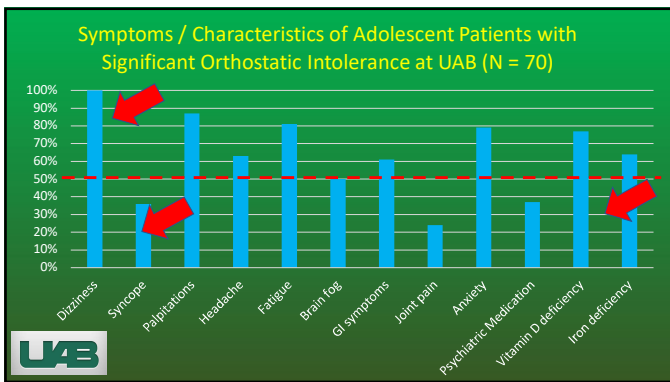
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Intake Description of Adolescent Patients with Significant Orthostatic Intolerance at UAB

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

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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

} P < 0.01

Medications	
Beta-blocker	64%
Fludrocortisone	41%
Midodrine	8%
Salt tablets	67%

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

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Today's Agenda - POTS

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



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JACC FOCUS SEMINAR

Received: 9 October 2018 | Accepted: 31 October 2018
DOI: 10.1111/ahaj12720

Postural Orthostatic Tachycardia Syndrome

JACC Focus Seminar

Meredith Bryarly, MD,^{1,2} Lauren T. Phillips, MD,^{1,2} Benjamin D. Levine, MD^{3,4}

Pediatric dysautonomia: Much-maligned, often overmedicated, but not as complex as you think

Camden L. Hebson MD¹ | Michael E. McConnell MD² | David W. Hannon MD³

ABSTRACT

Postural orthostatic tachycardia syndrome (POTS), the affects approximately 500,000 people in the United States and the beginning of their working lives. This is a hetero are not well understood. There are multiple contributing most current views on the potential causes, comorbid and treatment of POTS from cardiological and neurolog © 2019 by the American College of Cardiology Founda

Abstract

Dysautonomia is an increasingly recognized yet still poorly understood disease within the field of pediatrics. Symptoms, including dizziness, headaches, fatigue, joint pain, anxiety, and intolerance of heat or cold, are often significant and difficult to sort, especially in terms of their relation to each other. This often leads to referral to multiple subspecialists, who then proceed to treat seemingly familiar symptoms in kind. In the authors' experience, this leads to more frustration on the part of the patients and their physicians when symptom improvement does not follow for can even

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Thank you!

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Extra Slides

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Evidence Report:

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Risk of Orthostatic Hypotension in Patients, Bed Rest Subjects, and Astronauts

The Journal of Clinical Pharmacology
Official Publication of the American College of Clinical Pharmacology

Orthostatic Hypotension in Patients, Bed Rest Subjects, and Astronauts

Dr. Claire M. Lathers PhD, FCP, Dr. John B. Charles PhD

First published: May 1994 | <https://doi.org/10.1002/j.1552-4604.1994.tb04980.x> | Cited by: 19

Full Text provided by University of Alabama at Birmingham

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Original Article Mayo Clin Proc. 2002;77:531-537

Quality of Life in Patients With Postural Tachycardia Syndrome

LESA M. BENRUD-LARSON, PhD; MELANIE S. DEWAR, BS; PAGLA SANDRONI, MD, PhD; TERESA A. ROMIGANS, MD; JENNIFER A. HAYTHORNTWATTE, PhD; AND PHILLIP A. LOW, MD

Scale	Healthy	POTS	OEF
PF	~85	~55	~75
BP	~80	~55	~65
GH	~75	~50	~60
VIT	~65	~45	~55
SF	~75	~65	~70

- N = 94 (mean age = 34 years)
- Significant perceived decrease in:
 - Physical functioning (PF)
 - Bodily pain (BP)
 - General health (GH)
 - Vitality (VIT)
 - Social functioning (SF)
- **Similarly poor to known chronic disabling conditions**


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Clin Auton Res (2012) 22:151–153
DOI 10.1007/s10286-011-0155-1

RESEARCH LETTER

Long-term follow-up of patients with postural tachycardia syndrome

Alexandra Sousa · Ana Lebreiro · João Freitas ·
M^a Júlia Maciel



- Young adults with “severe” POTS
- Mean follow-up ~ 7.5 years
- Mean age 24 years
- 94% on at least one medication for POTS
- N = 34
- 31% symptom free
- 56% with significant improvement
- 13% not improved
- Only 62% still on a medication (mostly beta blockers)
