

Dysautonomia / POTS

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Internal Medicine
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1

Disclosures

- Financial: None
- There are no approved medications for POTS. All discussed medications are off-label.

2

Objectives

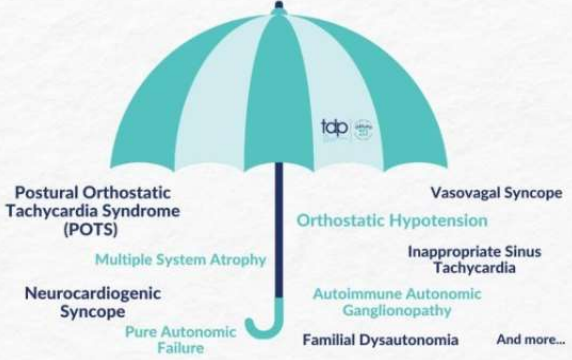
1. What is POTS?
2. How do we diagnose POTS?
3. How do we treat POTS?

3

What is Dysautonomia?

When there is a dysfunction or failure of the autonomic nervous system, the result is a disorder classified as a type of dysautonomia.

Dysautonomia is not a diagnosis. It's an umbrella term to describe autonomic disorders.



tcb

Postural Orthostatic Tachycardia Syndrome (POTS)

Multiple System Atrophy

Neurocardiogenic Syncope

Pure Autonomic Failure

Orthostatic Hypotension

Inappropriate Sinus Tachycardia

Autoimmune Autonomic Ganglionopathy

Familial Dysautonomia

And more...

Vasovagal Syncope

The Human Nervous System

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graph TD
    HNS[The Human Nervous System] --> CNS[Central nervous system]
    HNS --> PNS[Peripheral nervous system]
    CNS --> Brain[Brain]
    CNS --> SpinalCord[Spinal cord]
    PNS --> Motor[Motor]
    PNS --> Sensory[Sensory]
    Motor --> Autonomic[Autonomic]
    Motor --> Somatic[Somatic]
    Autonomic --> Sympathetic[Sympathetic]
    Autonomic --> Parasympathetic[Parasympathetic]
  
```

(The Autonomic branch is circled in red in the original image.)

4

What is The Autonomic Nervous System?

The part of the nervous system that regulates functions that are automatic in nature such as heart rate, blood pressure, digestion, excretion, perspiration, temperature regulation, pupil dilation, circulation, and respiration among others.

Parasympathetic	Sympathetic
constricts pupils	dilates pupils
increases saliva production	inhibits saliva production
reduces the heart rate	dilates bronchia
constricts bronchia	raises the heart rate
stimulates the activity of the digestive organs	inhibits the activity of the digestive organs
stimulates the activity of the pancreas	inhibits the activity of the pancreas
stimulates the gall bladder	inhibits the gallbladder
constricts urinary bladder	stimulates the adrenal medulla to release adrenaline and noradrenaline
stimulates erection of genitals of man and woman	relaxes urinary bladder
	stimulates orgasm in genitals of man and woman

PUPILOMOTOR
impaired pupil response (uncomfortable in bright light)
difficulty with vision

SECRETOMOTOR
difficulty sweating, tearing and other fluid production (dry eyes, dry mouth, difficulty swallowing, dry skin)

GASTROINTESTINAL
nausea, vomiting, diarrhea, constipation, abdominal pain, reflux, heartburn, impaired motility

ORTHOSTATIC INTOLERANCE
difficulty standing still, fatigue, lightheadedness, increase in symptoms with upright posture, fainting (syncope) or near-fainting, pallor

NEUROLOGICAL
migraine, cognitive deficits, brain fog & mental clouding

PULMONARY
shortness of breath
easily winded
difficulty breathing

CARDIOVASCULAR
palpitations, chest discomfort
high heart rate (tachycardia)
low heart rate (bradycardia)
high or low blood pressure
abnormal blood vessel functioning
blood pooling

URINARY
difficulty with urine retention and/or excretion

Symptoms can be **SUDDEN** and **unpredictable** in onset

5

Idiopathic postural orthostatic tachycardia syndrome: An attenuated form of acute pandysautonomia?

Ronald Schondorf, PhD, MD, and Phillip A. Low, MD

- 16 Patients evaluated for OI with exaggerated orthostatic tachycardia in absence of OH underwent comprehensive autonomic testing
 - Sustained orthostatic HR increase >2 SD of mean HR of controls
- Predominantly young woman
- Acute onset
- Frequent triggering event
 - 7/16 postviral
- Mild form of dysautonomia

Table 1. Characteristics of patients with POTS

Pt no.	Sex	Age	Onset	Duration (mo)	Light-headedness	Fatigue	GI motility	Small fiber sensory
1	M	26	Unclear	?	Chronic	Chronic	N	No
2	F	33	Postviral	7	Chronic	Chronic	Abn	No
3	M	51	Postviral	3	Chronic	Chronic	N	No
4	F	42	Postviral	36	Chronic	Chronic	N	No
5	M	38	Postviral	16	Chronic	Chronic	N	No
6	F	28	Postviral	5	Occasional	Chronic	N	No
7	F	42	Postviral	26	Chronic	Chronic	Abn	Yes
8	F	48	Unclear	?	Occasional	?	N	Yes
9	F	35	Acute	3	Chronic	Chronic	N	No
10	F	35	Acute	3	Occasional	Chronic	N	No
11	F	26	Acute	24	Chronic	Chronic	N	No
12	F	51	Acute	2	Occasional	Chronic	N	No
13	F	20	Acute	3	Chronic	Chronic	N	No
14	F	25	Acute	18	No	No	Abn	No
15	F	32	Postviral	24	No	No	Abn	No
16	F	24	Acute	20	No	No	Abn	No

N Normal
Abn Abnormal.

Table 2. Tests of autonomic function in patients with POTS

Pt no.	HR supine (2.0 min)	HR upright (2.0 min)	HR supine (2.0 min)	HR upright (2.0 min)	TST % anhidrosis	QSBART	HR _{max} (bpm)	NE supine (pg/ml)	NE upright (pg/ml)
1	111	132	188/105	196/115	N	N	N	539	1300
2	114	140	141/95	85/50*	31%	Abn	Abn	61	224
3	51	89	132/44	112/72	N	Abn	N	57	224
4	94	131	128/86	126/84	28%	N	N	809	802
5	79	131	144/51	133/79	N	N	N	281	847
6	87	130	130/87	134/89	N	N	N	178	552
7	76	130	134/72	140/86	51%	Abn	Abn	123	123
8	92	138	126/70	96/58	91%	Abn	Abn	123	123
9	86	131	130/88	136/92	28%	N	N	330	1196
10	125	142	166/88	149/88	N	Abn	Abn	261	604
11	70	112	102/59	107/89	N	Abn	Abn	317	1021
12	60	105	126/74	70/53*	N	Abn	N	77	390
13	87	88	129/74	109/72	N	N	N	212	647
14	85	135	108/58	112/78	N	N	N	122	122
15	92	130	120/88	116/88	N	N	N	170	375
16	79	130	94/68	96/75	N	N	N	181	181

N Normal
Abn Abnormal.

NEUROLOGY 1993;43:132-137

6

It's about time to know about POTS in the USA...
 by naughtylittlemastcells.com

1 MILLION have Chronic Fatigue Syndrome (ME/CFS)

1 MILLION have Parkinson's Disease

100,000 have Sickle Cell Anemia

30,000 have Cystic Fibrosis

15,000 have ALS (Lou Gehrig's Disease)

1.5 MILLION have Lupus

1.5 MILLION have Rheumatoid Arthritis

1.3 MILLION have Crohn's Disease or Ulcerative Colitis (IBD)

1.25 MILLION have Type I Diabetes

1 MILLION have Multiple Sclerosis (MS)

1 TO 3 MILLION have POTS
 POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME

POTS is a life altering illness commonly misdiagnosed as anxiety.

♀ = 100,000 people

POTS is Common!!

LEARN MORE dysautonomiainternational.org

PLEASE SHARE TO PROMOTE POTS AWARENESS!

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7

METHODS

Dysautonomia International developed a web-based 10 question survey. The survey was hosted on a secure commercial survey website. The survey was anonymous and no personally identifiable information was collected. The survey was designed so that each person could only take the survey once.

We announced the availability of the survey through our Facebook page on December 2, 2013 and the survey responses were collected until January 2, 2013.

Many of our Facebook followers shared the survey link on POTS related support groups. The survey announcement indicated that only people who had been diagnosed with POTS by a doctor should participate. The survey was open to POTS patients of any age, from any geographic location.

Age at Onset of POTS Symptoms

N = 696

34% of participants were teenagers when they first developed symptoms of POTS

Age Range (years)	Number of Participants
12 and under	~140
13-19	~240
20-29	~140
30-39	~120
40-49	~40
50-59	~10
60 and over	~5

Months from Onset of POTS Symptoms to POTS Diagnosis

75% of participants received a diagnosis of POTS 1 year or more after the onset of POTS symptoms

The average time to diagnosis was 5 years and 11 months

N = 696

Time (years)	Percentage of Participants (%)
less than 3 months	~15
3-6 months	~9
6-12 months	~10
1-2 years	~13
2-5 years	~18
5-10 years	~15
more than 10 years	~19

How many doctors did you see for your POTS symptoms before you were formally diagnosed with POTS?

N = 684

- 27% of subjects saw more than 10 doctors before being diagnosed with POTS
- 8% saw more than 20 doctors
- Only 6% diagnosed by the first doctor they saw for their POTS symptoms

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

N = 684

Diagnosis	Percentage
depression	~35%
"all in your head"	~30%
somatoform disorder	~10%
conversion disorder	~5%
factitious disorder	~2%
panic disorder	~15%
anxiety	~60%
stress from work/school/family	~65%
munchausen's syndrome	~2%
other mental/psychological	~20%
none of the above	~10%

What type of physician first diagnosed you with POTS?

Only 12% of participants were diagnosed with POTS by a pediatrician or family practitioner

N = 668

Physician Type	Percentage
Pediatrician	~12%
Family Practitioner	~12%
Cardiologist/Electrolysiologist	~60%
Neurologist	~25%
Gastroenterologist (GI doctor)	~5%
Gynecologist	~2%
Nephrologist (kidney doctor)	~1%

8

Quality of Life in Patients With Postural Tachycardia Syndrome

LISA M. BENRUD-LARSON, PhD; MELANIE S. DEWAR, BS; PAOLA SANDRONI, MD, PhD; TERESA A. RUMMANS, MD; JENNIFER A. HAYTHORNTHWAITE, PhD; AND PHILLIP A. LOW, MD

• Results: Ninety-four patients (89% female; mean age, 34.2 years) were enrolled in the study. Patients with POTS reported impairment across multiple domains on the SF-36.

Mayo Clin Proc. 2002;77:531-537

Postural orthostatic tachycardia syndrome is associated with significant employment and economic loss

Kate M. Bourne¹, Derek S. Chew^{1,2}, Lauren E. Stiles^{3,4}, Brett H. Shaw¹, Cyndya A. Shibus⁵, Luis E. Okamoto⁶, Emily M. Garland⁷, Alfredo Gamboa⁸, Amanda Peltier^{1,5}, André Diedrich⁵, Italo Biaggioni⁹, Robert S. Sheldon⁶, David Robertson⁶ & Satish R. Raj^{1,5}

Table 1. Summary of participant 'yes' responses to questions evaluating aspects of employment

	%	N (Yes)	N (Overall respondents)
Employed in previous 3 months	48.0	1,649	5,518
Would work more if not for health limitations	66.8	1,754	2,624
Lost a job due to POTS	20.9	547	2,616
Unable to work for > 1 week	74.0	4,033	5,448
Called in sick due to POTS	82.2	4,341	5,282
Lost income due to POTS	70.5	3,735	5,301
Spouse/Caregiver lost income due to POTS	28.5	1,551	5,442
Modified job due to POTS	72.4	3,715	5,129
Workplace modifications	55.4	2,001	3,609
Schedule reductions	74.1	2,692	3,633
Other modifications	67.2	2,392	3,559

Journal of Internal Medicine, 2021, 290, 203-212

9

POTS Triggers

41% of POTS patients report onset w/in 3 months of a specific event:

- infection (41%)
- surgery (12%)
- pregnancy (9%)
- accident (6%)
- concussion (4%)

Table 2 The different types of events that have been described as triggering PoTS

Triggering event
Infection (viral and bacterial) [36, 70, 72-77]
Vaccination [26, 31, 78, 79]
Trauma [13]
Pregnancy [36, 72]
Surgery [34]
EDS/JHS [71, 80]
Psychosocial stress [36]
Mast cell activation disorder [81]
Lyme disease [82]
Other autoimmune disease [83]
Medications e.g., antihypertensives, antipsychotics [12]

10

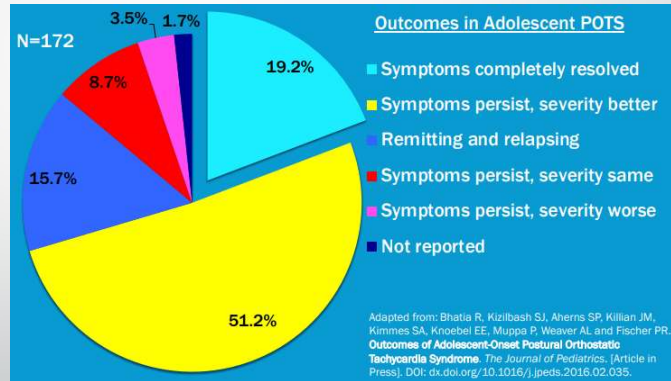
Outcomes of Adolescent-Onset Postural Orthostatic Tachycardia Syndrome

Roma Bhatia, BS¹, Sarah J. Kizilbash, MD^{1,2,*}, Shelley P. Ahrens, DNP^{1,2}, Jill M. Killian, BS^{1,3}, Stephanie A. Kimmes, CNP^{1,2}, Erin E. Knoebel, MD^{1,2}, Prasuna Muppa, MBBS^{1,2}, Amy L. Weaver, MS^{1,3}, and Philip R. Fischer, MD^{1,2}

Survey (n = 172)

Mean duration from diagnosis to survey completion was 5.4 years

Mean age of the respondents at the time of the survey was 21.8 years



(J Pediatr 2016;173:149-53)

11

For most, this is a Chronic Condition



Dysautonomia International
21h · 🌐

There are few studies documenting long term outcomes in #POTS, but a new study from Vanderbilt and University of Calgary gives us the longest follow up data to date. Researchers reached out to POTS patients who had participated in Vanderbilt research studies decades earlier to see how they were doing now. 45 POTS patients participated in the follow up study, with most patients being in their late 40s or older now. On follow up 20 years or more after their POTS symptoms started, only 2% of participants reported complete resolution of POTS symptoms, 46% noted some improvement, 11% experienced no changes in symptoms, 25% reported worsening symptoms, and 16% experienced variable symptoms. Patients who did not improve were more likely than those who did see some improvement to have dry eyes, mouth or skin (regulated by the secretomotor small fiber autonomic nerves) when they were first seen at Vanderbilt years earlier, and the non-improved patients were more likely to have neuropathy, gastroparesis, or overactive bladder symptoms at the time of the follow-up study. The findings emphasize the importance of screening for small fiber neuropathy at the time of POTS diagnosis, and screening for diseases associated with small fiber neuropathy and secretomotor deficits, such as Sjogren's syndrome and diabetes.

This data was presented as an abstract/poster at an academic conference recently. You can read the abstract at <https://link.springer.com/article/10.1007/s10286-024-01075-8> (Poster 106) .

12

Diagnostic Criteria for POTS

Clinical Feature	Diagnostic Criterion	Assessment
Excessive Orthostatic Tachycardia	HR increase > 30 bpm (>40 bpm teenagers)	Active stand or HUT
Symptoms of Orthostatic Intolerance	Orthostatic symptoms which improve with recumbence	Clinical evaluation and history
Absence of Orthostatic Hypotension	No decrease in BP > 20/10mmHg	Active stand or HUT
Chronicity	>3-6 months	Clinical evaluation and history
Absence of other causes	Other conditions causing orthostatic tachycardia and symptoms have been ruled out	Clinical evaluation and history

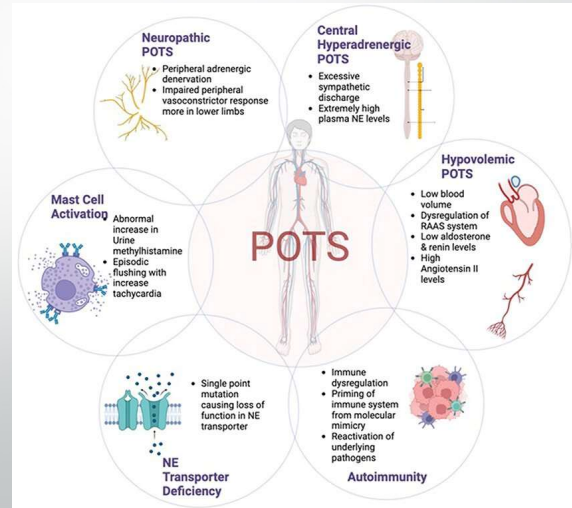
13

Pathophysiology

14

POTS as a Common Final Pathway

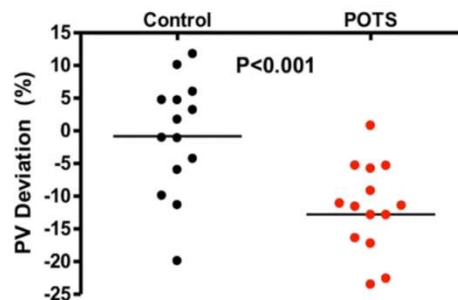
- POTS is a **syndrome** that may result from one or more different pathophysiological abnormalities
 - Volume dysregulation / Hypovolemia
 - Hyperadrenergic
 - Neuropathic
 - Deconditioning
 - + more



15

Volume Dysregulation

Plasma Volume is Low in POTS



Adapted from SR Raj et al., Circulation 2005;111:1574-1582

70% of POTS Patients

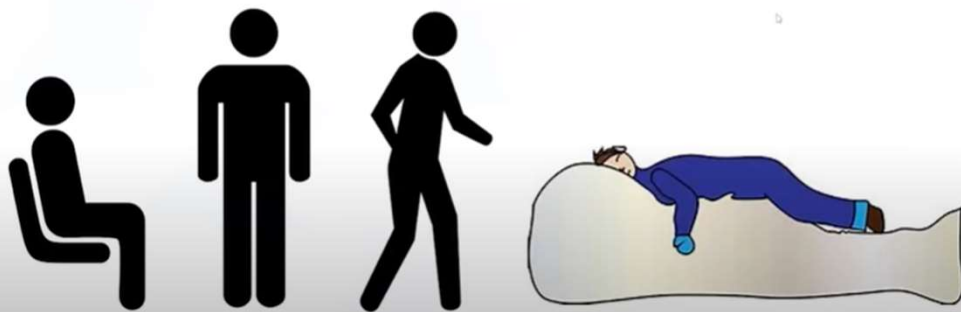
16

Clinical Presentation

17

What is Orthostatic Intolerance (OI)?


Orthostatic intolerance is the development of symptoms in **upright posture** that are relieved or partially relieved by **reclining** or **laying down**.



18

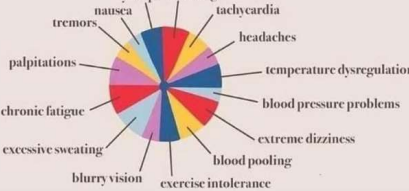
Symptoms

what people think POTS is like:



tachycardia

what POTS is actually like:



@chronically.kelsey


Table 1 Symptoms associated with POTS and their relative frequency in 152 patients

Symptom	Frequency (%)
Orthostatic	
Light-headedness or dizziness	77.6
Pre-syncope	60.5
Weakness	50
Palpitations	75
Tremulousness	37.5
Shortness of breath	27.6
Chest pain	24.3
Loss of sweating	5.3
Hyperhidrosis	9.2
Non-orthostatic	
Bloating	23.7
Nausea	38.8
Vomiting	8.6
Abdominal pain	15.1
Constipation	15.1
Diarrhoea	17.8
Bladder dysfunction	9.2
Pupillary dysfunction	3.3
Generalized	
Fatigue	48
Sleep disturbance	31.6
Migraine headache	27.6
Myofascial pain	15.8
Neuropathic pain	2

Adapted from Thieben et al.⁴

Mayo Clinic Proceedings 82:308–313, 2007.

19



COAT HANGER PAIN

Suboccipital and paracervical pain that worsens in the upright position is common in orthostatic disorders and is believed to be caused by poor blood flow to the muscles of the upper back and neck.

www.dysautonomiainternational.org

20

Pretzel Legs



Water Bottle Sign



21

POTS Legs

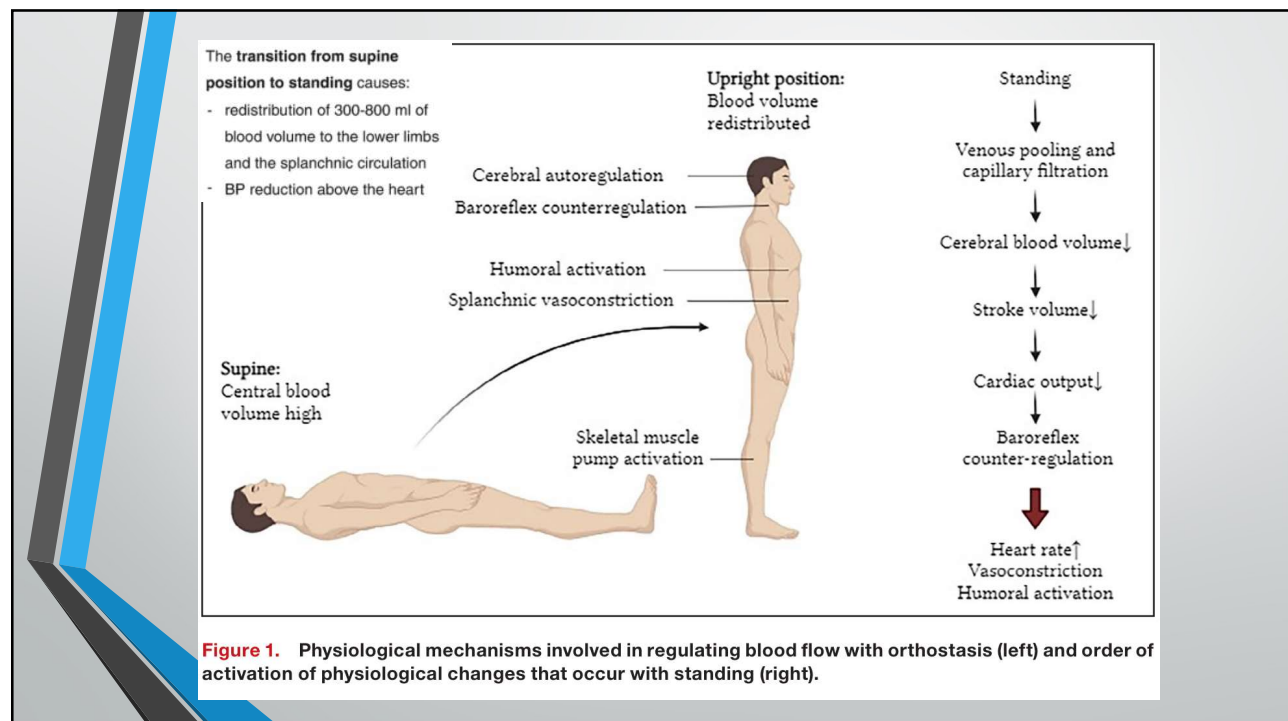


Fig. 1. Both of this POTS patient's legs become acrocyanotic within 5 min of standing. The acrocyanosis dissipated in the right leg after elevating it on a counter for 3 min immediately prior to this photo.

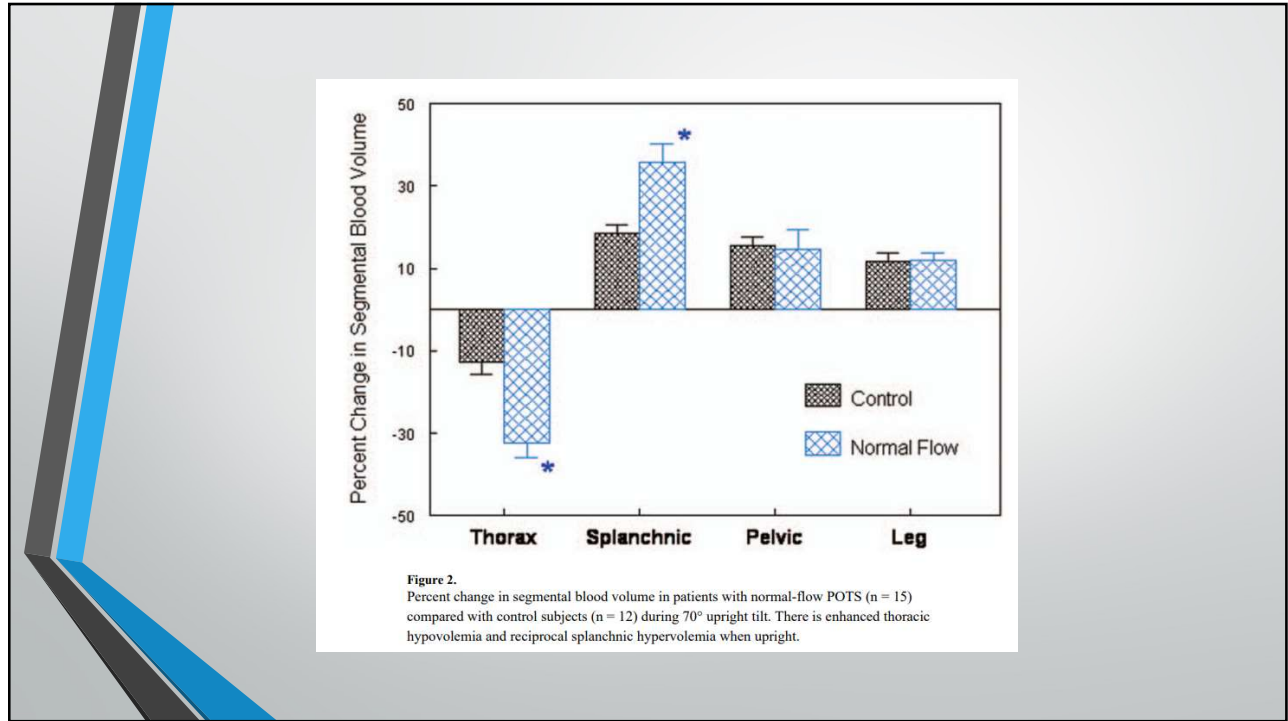
22

Assessing Orthostatic Tolerance

23



24



25

Step 1: Heart rate and blood pressure are documented after the patient lays down for 10+ minutes

Step 2: The patient is tilted. Heart rate and blood pressure are documented continuously for the next 10+ minutes

Diagnostic criteria:

- Heart rate change of 30+ bpm
- Or heart rate hits 120bpm

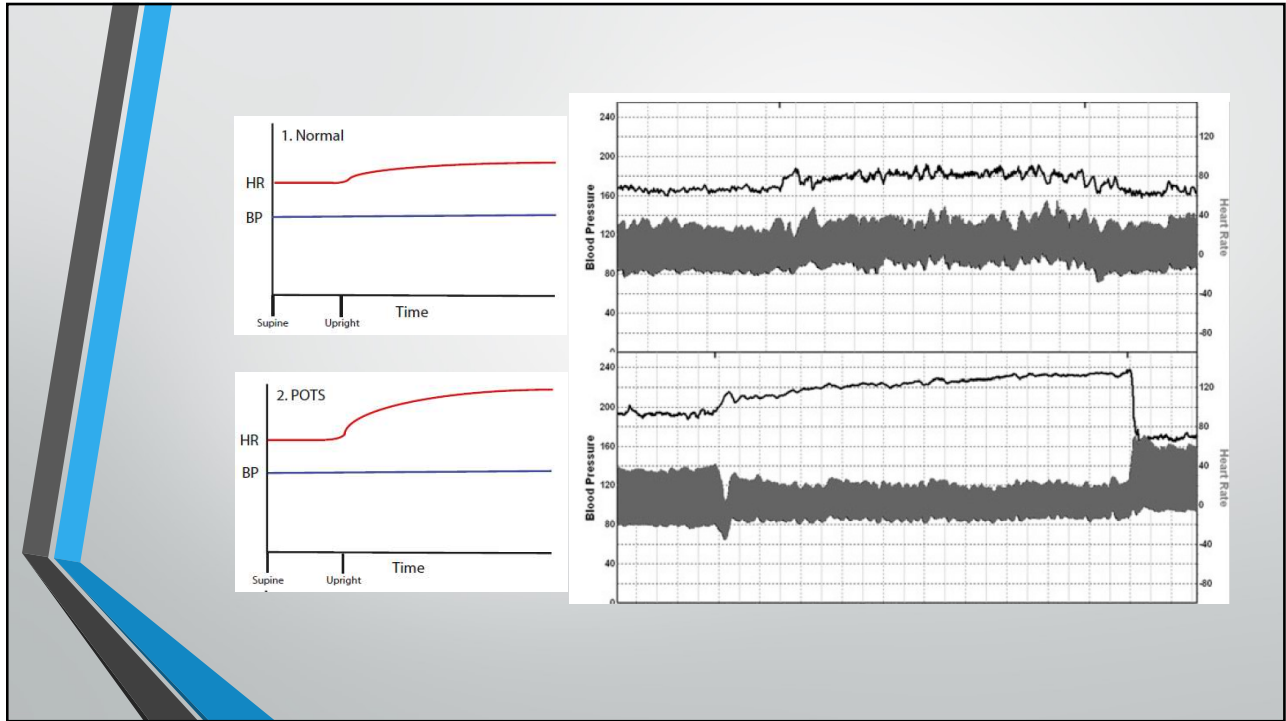
Diagnosis and Testing for the POTS

Tilt Table Test

tilt 0 to 75 degree

	Active stand testing	Head up tilt testing
Heart rate evaluation	First line diagnostic tool for PoTS	Elicits greater orthostatic tachycardia
Blood pressure evaluation	May elicit initial orthostatic hypotension	
Usage	May be performed in the physician's office	Requires specialized equipment
Sensitivity (%)	87	93
Specificity	67	40

26



27

Evaluate for Orthostatic Intolerance: 10-Minute NASA Lean Test

HR and BP after
10-15 minutes of
quiet supine rest

↓

HR and BP every
1-2 minutes for
10 minutes while
standing/leaning
in upright posture

RESEARCH | CLINICAL CARE | EDUCATION

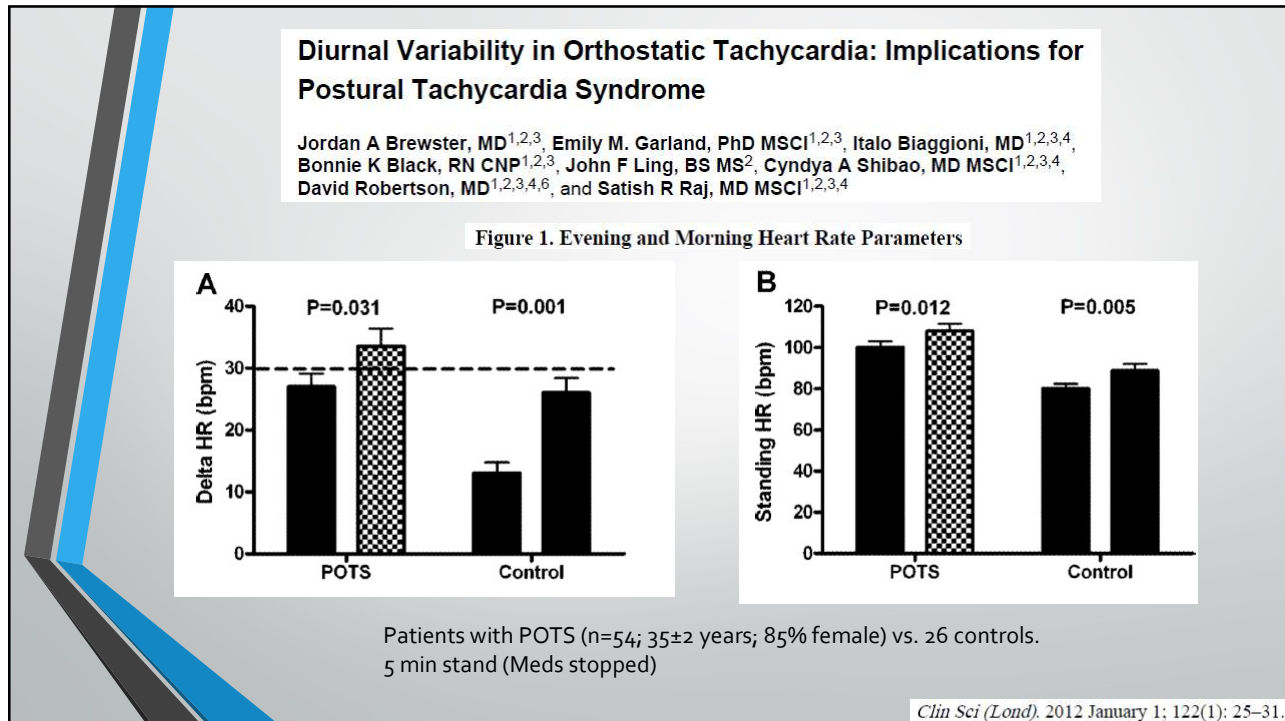
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28

	Blood Pressure (BP)		Pulse	Comments
	Systolic	Diastolic		
Supine 1 minute			68	
Supine 2 minute	124	87	72	
Standing 0 minute			116	LH1
Standing 1 minute			110	
Standing 2 minute	127	95	115	
Standing 3 minute			118	
Standing 4 minute			116	Hand swelling
Standing 5 minute	129	98	106	Dizziness
Standing 6 minute			112	Tugby Fever
Standing 7 minute			114	
Standing 8 minute			114	
Standing 9 minute	138	86	119	
Standing 10 minute			118	

HR Increase > 45 bpm

29



30

Diurnal Variability in Orthostatic Tachycardia: Implications for Postural Tachycardia Syndrome

Jordan A Brewster, MD^{1,2,3}, Emily M. Garland, PhD MSCI^{1,2,3}, Italo Biaggioni, MD^{1,2,3,4}, Bonnie K Black, RN CNP^{1,2,3}, John F Ling, BS MS², Cyndya A Shibao, MD MSCI^{1,2,3,4}, David Robertson, MD^{1,2,3,4,6}, and Satish R Raj, MD MSCI^{1,2,3,4}

Figure 1. Evening and Morning Heart Rate Parameters

Keep in mind active treatments and medications

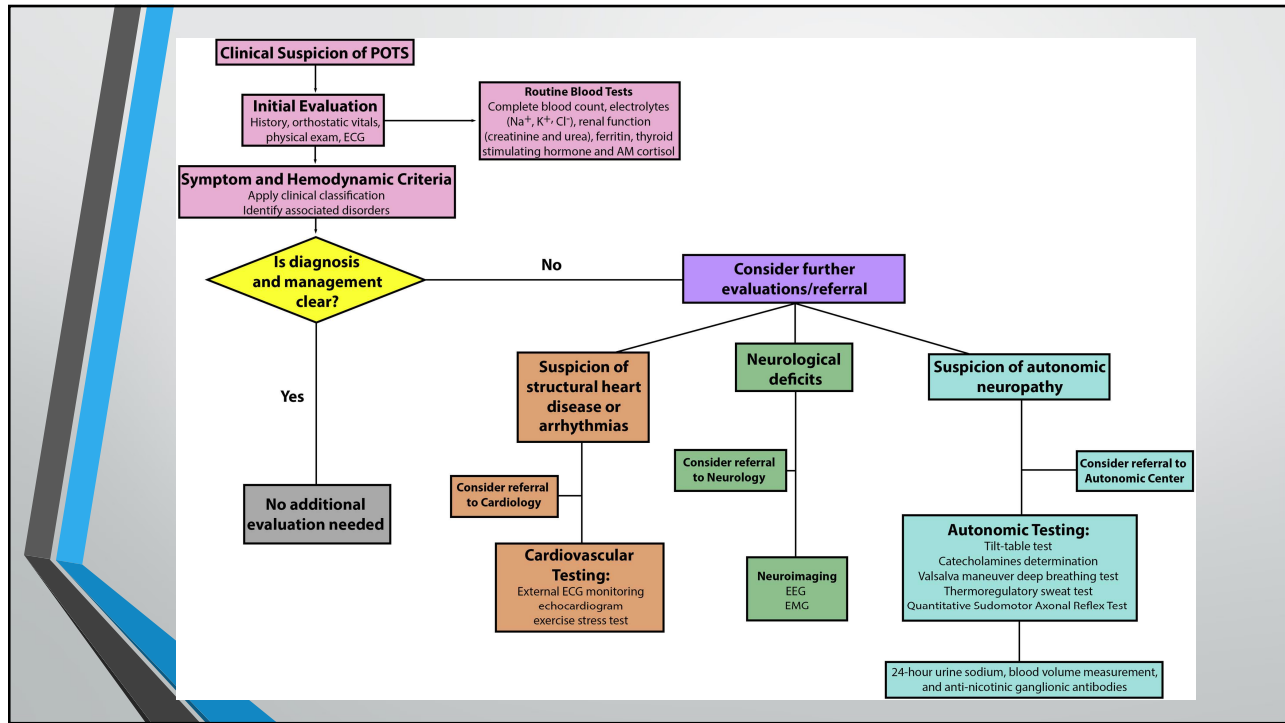
Patients with POTS (n=54; 35±2 years; 85% female) vs. 26 controls.
5 min stand (Meds stopped)

Clin Sci (Lond). 2012 January 1; 122(1): 25–31.

31

Making a Diagnosis

32



33

Box 3: Other conditions that could explain sinus tachycardia on standing⁵

- Acute hypovolemia (from dehydration or blood loss)
- Anemia
- Orthostatic hypotension
- Endocrinopathy
 - Adrenal insufficiency
 - Carcinoid tumour
 - Hyperthyroidism
 - Pheochromocytoma
- Adverse effects from medication
- Panic attacks and severe anxiety
- Prolonged or sustained bed rest
- Recreational drug effects

Absence of Other Causes

Table 4 Medications that can cause PoTS-like tachycardia

Medication type	Effect	Examples
Vasodilators	Vasodilation, reduced blood return	Nitroglycerin
Diuretics	Decreased fluid volume	Drosiprenone Oral Contraceptive
Norepinephrine transporter (NET) inhibitors	Reduced norepinephrine reuptake	Atomoxetine, reboxetine
Selective-norepinephrine reuptake inhibitors	Reduced norepinephrine reuptake	Duloxetine, venlafaxine
Tricyclic Antidepressants	Reduced norepinephrine reuptake	Nortriptyline
Attention Deficit Hyperactivity Disorder Medications	Sympathomimetic	Methylphenidate, Amphetamine
Beta-adrenergic Receptor Agonists	Direct inotropic effects. Vasodilation	Salbutamol

34

The face of postural tachycardia syndrome – insights from a large cross-sectional online community-based survey

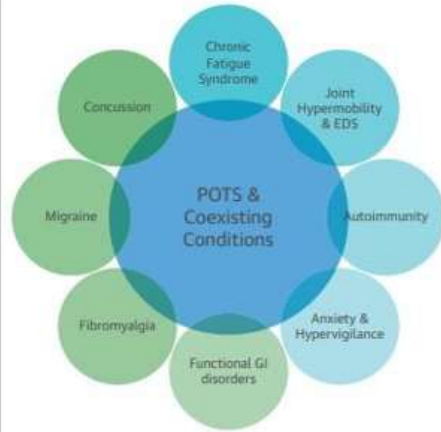
■ B. H. Shaw¹, L. E. Stiles^{2,3}, K. Bourne¹, E. A. Green⁴, C. A. Shihao⁴, L. E. Okamoto⁴, E. M. Garland⁴, A. Gamboa⁴, A. Diedrich⁴, V. Raj^{1,2}, R. S. Sheldon¹, I. Biaggioni⁴, D. Robertson⁴ & S. R. Raj^{1,4}

Table 2 Common comorbidities in POTS patients

Comorbidity	Number (%) (of 3933 respondents)
Migraine headaches	1557 (40%)
Irritable bowel syndrome	1192 (30%)
Ehlers-Danlos syndrome	994 (25%)
Chronic fatigue syndrome	809 (21%)
Asthma	798 (20%)
Fibromyalgia	786 (20%)
Raynaud's phenomena	610 (16%)
Iron deficiency anaemia	628 (16%)
Gastroparesis	548 (14%)
Vasovagal syncope	499 (13%)
Inappropriate sinus tachycardia	448 (11%)
Mast cell activation disorder	353 (9%)
Autoimmune disease	616 (16%)
Hashimoto's thyroiditis	228 (6%)
Celiac disease	133 (3%)
Sjögren's syndrome	112 (3%)
Rheumatoid arthritis	93 (2%)
Lupus	81 (2%)
Other	160 (4%)

Journal of Internal Medicine, 2019, 286; 438-448

FIGURE 4 Coexisting Conditions in POTS



Patients often relate many other symptoms and comorbidities that are not physiologically explained by orthostatic tachycardia. Some of these persist even with resolution of orthostatic intolerance. These include chronic fatigue syndrome, joint hypermobility and EDS, autoimmune diseases, anxiety and hypervigilance, functional gastrointestinal disorders, fibromyalgia, migraine, and concussion. EDS – Ehlers-Danlos syndrome; GI – gastrointestinal; POTS – postural orthostatic tachycardia syndrome.

35

TREATMENT

36

Education



37

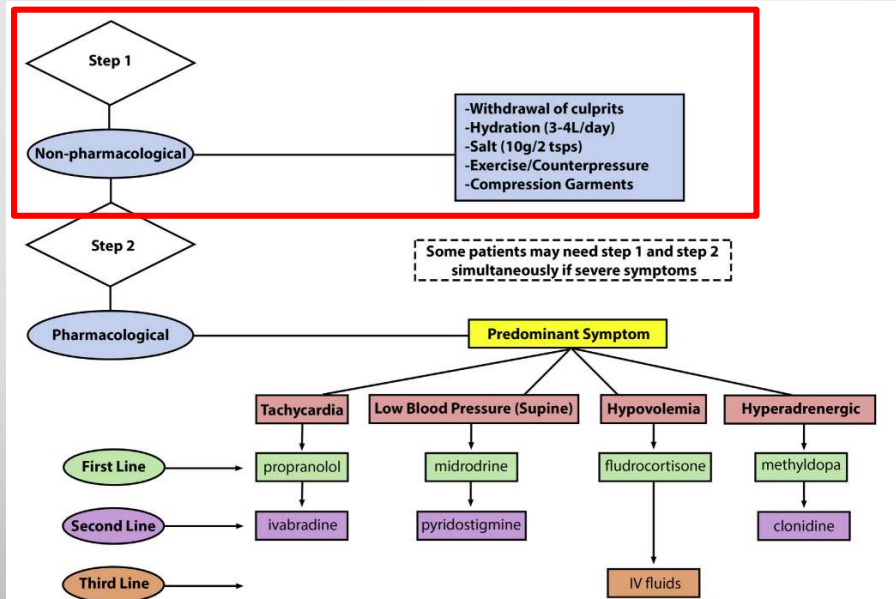


Figure 4. Postural orthostatic tachycardia syndrome treatment algorithm: a suggested treatment algorithm for patients with postural orthostatic tachycardia syndrome.

38

Non-Pharmacological Treatment

FIGURE 2 Nonpharmacological Interventions for Minimizing Orthostatic Intolerance Symptoms

<p>Avoid Situations That Can Exacerbate Symptoms</p>  <p>Large/Heavy Meals Heat Exposure Alcohol Intake</p>	<p>Liberal Intake of Salt and Water</p>  <p>Liberal Intake of Salt and Water</p>	<p>Sleep With Head of Bed Elevated</p>  <p>Head posts should be elevated 4-6 inches</p>
<p>Use of Compression Garments</p>  <p>Abdominal Binder</p>	<p>Physical Counter Maneuvers</p>  <p>Hose Leg Crossing Maneuver Squatting</p>	<p>Drinking Water Before Getting Up In The Morning</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>Recumbent Bicycle</p>

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.


39


Avoiding Triggers


@Vitassium


Potential POTS symptom triggers


according to the community


 Standing


 Eating or not eating enough


 Heat


 Certain foods


 Showering or bathing


 Consuming caffeine or alcohol


 Lack of sleep


 Dehydration


 Stress


 Menstrual cycle


 Physical activity or exercise

 Acute illness

 Movement like reaching or bending over

 Traveling

 Stairs

 Barometric pressure changes

40

Dietary Manipulation

Nutritional Factors that Improve POTS Symptoms

- 2-3 L water
- 10-12 g salt (as tolerated)

Factors that Stabilize POTS Symptoms

- Small frequent meals
- Start fluids early in the AM
- Reduce refined carbohydrates
- Manage digestive symptoms
- Refer for celiac screening, if appropriate
- Screen for eating disorders
- When appropriate, encourage paced recumbent activity
 - Refer to physical therapist if appropriate
- Coordinate with health care team

Nutritional Factors that Exacerbate POTS Symptoms

- Alcohol
- Caffeine
- Dehydration

Figure 3. Nutritional guidance for postural orthostatic tachycardia syndrome (POTS).^{11,14,17-19,21,23-26}

- Consume 2-3 L of water daily^{9,11}
 - Start fluids early in the morning, even before getting out of bed¹⁹
- Increase sodium as tolerated, up to 10-12 g/d⁹
 - Consult with medical team regarding sodium goal
 - If not provided, start with 6 g salt daily, divided throughout the day. Increase gradually²⁰
 - Patient can add salt to food. Or, if needed, patients can use slow-release salt tablets^{9,11}
 - Too much salt at one time can trigger nausea²¹
- Patients should avoid alcohol, which may increase symptoms of POTS¹²
- Patients should avoid caffeine, which may increase symptoms of POTS¹²
- Patients should avoid dehydration, which exacerbates the symptoms of POTS¹²
- Small frequent meals and fewer refined carbohydrates are recommended for glycemic balance¹⁹
- Coordinate all recommendations with interprofessional team

Figure 2. Current dietary recommendations for postural orthostatic tachycardia syndrome.

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41

Effect of High Dietary Sodium Intake in Patients With Postural Tachycardia Syndrome

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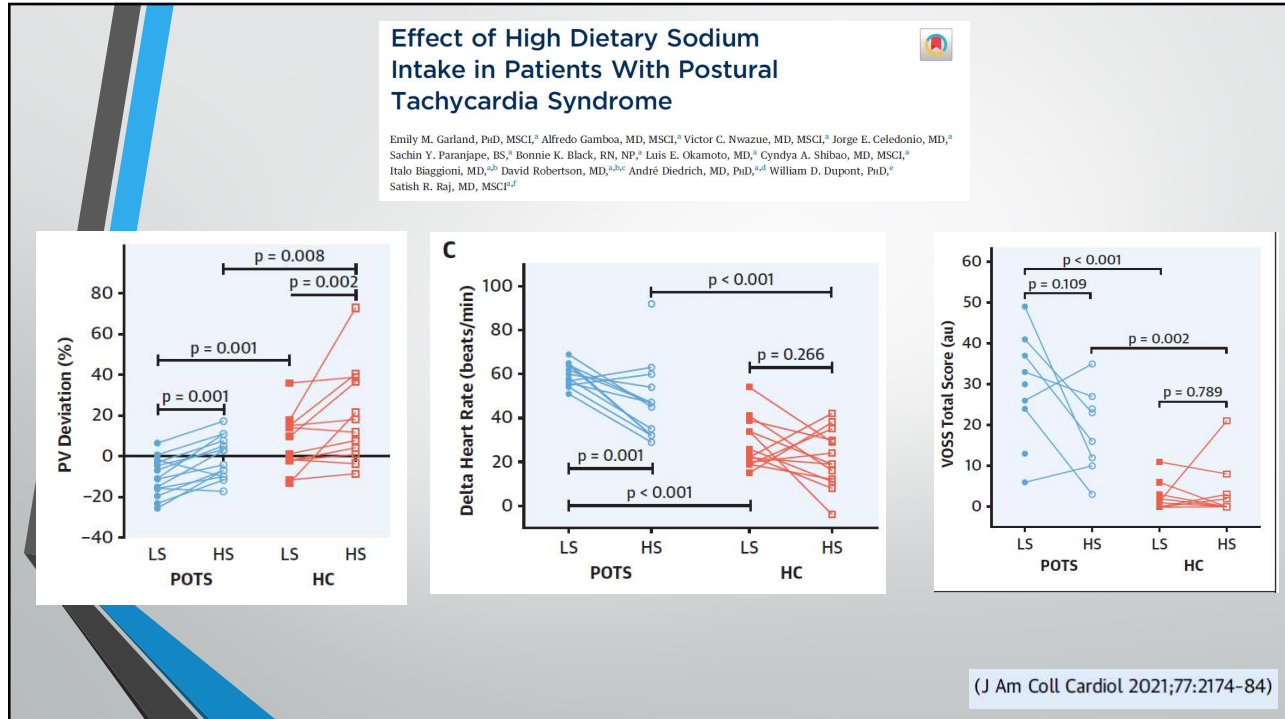
FIGURE 1 Crossover Design of Dietary Sodium Study in POTS

TABLE 3 Dietary Sodium and Responses to Orthostatic Stress

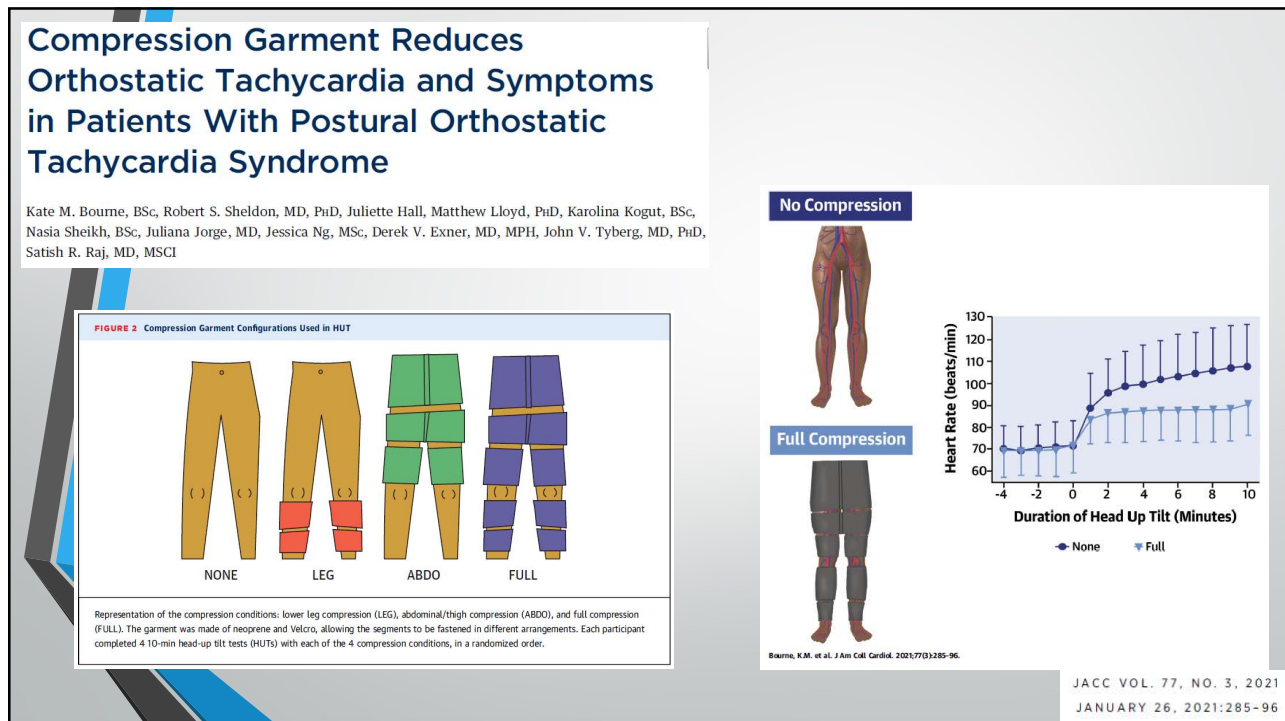
	Low Sodium vs. High Sodium Diets						POTS vs. Control Subjects	
	POTS			Controls			Diet	
	Low Sodium	High Sodium	p Value	Low Sodium	High Sodium	p Value	p Value for Low Sodium	p Value for High Sodium
Supine								
Heart rate, beats/min	73 (62-78)	69 (61-72)	0.055	70 (57-73)	62 (53-75)	0.133	0.157	0.525
Systolic blood pressure, mm Hg	105 (103-108)	104 (99-115)	0.891	105 (100-112)	108 (98-118)	0.529	0.746	0.510
Diastolic blood pressure, mm Hg	65 (63-71)	64 (58-74)	0.775	65 (58-70)	67 (63-70)	0.332	0.952	0.746
Norepinephrine, pg/ml	248 (162-332)	166 (90-332)	0.088	135 (104-225)	108 (84-162)*	0.018	0.030	0.193
Epinephrine, pg/ml	21 (12-31)	11 (5.7-20)	0.009	14 (6.6-19)	20 (6.7-28)*	0.060	0.143	0.163
Plasma renin activity, ng/ml/h	4.1 (0.9-4.7)†	0.7 (0.2-1.6)†	0.039	2.7 (1.5-5.0)†	0.3 (0.1-1.5)‡	0.029	0.606	0.734
Aldosterone, ng/dl	206 (157-249)	46 (34-58)	<0.001	131 (103-204)	41 (30-53)	<0.001	0.094	0.430
Upright								
Heart rate, beats/min	129 (124-141)‡	117 (98-121)§	0.002	96 (88-101)	85 (77-95)	0.091	<0.001	<0.001
Systolic blood pressure, mm Hg	112 (103-116)	116 (111-122)§	0.424	112 (105-117)	111 (107-118)	0.720	0.910	0.422
Diastolic blood pressure, mm Hg	68 (62-73)	71 (67-80)§	0.265	78 (69-80)	75 (74-80)	0.905	0.026	0.150
Norepinephrine, pg/ml	959 (736-1161)	753 (498-919)	0.017	520 (391-693)**	387 (312-433)**	0.014	<0.001	<0.001
Epinephrine, pg/ml	59 (33-86)	36 (13-68)	0.130	30 (24-46)**	24 (21-49)**	0.700	0.030	0.752
Plasma renin activity, ng/ml/h	24.6 (11.3-27.6)††	2.9 (1.5-5.0)††	0.002	7.3 (3.3-9.3)††	1.0 (0.8-1.9)‡‡	0.008	0.011	0.135
Aldosterone, ng/dl	455 (359-650)	64 (52-91)	<0.001	444 (277-515)	74 (52-129)	<0.001	0.173	0.302
Change from supine to upright								
Heart rate, beats/min	60 (55-64)	46 (32-55)	0.001	23 (19-36)	19 (11-32)	0.266	<0.001	<0.001
Systolic blood pressure, mm Hg	4 (0-12)	8 (1-12)	0.576	5 (0-8)	4 (1-11)	0.673	0.929	0.395
Diastolic blood pressure, mm Hg	2 (1-9)	6 (0-9)	0.416	10 (4-16)	11 (7-13)	0.553	0.019	0.077
Norepinephrine, pg/ml	692 (508-917)	535 (393-692)	0.035	295 (264-482)	304 (189-332)	0.019	<0.001	<0.001
Epinephrine, pg/ml	34 (17-56)	23 (5.4-57)	0.290	21 (13-29)	15 (2.5-23)	0.773	0.040	0.363
Plasma renin activity, ng/ml/h	21.6 (10.9-25.3)	1.4 (0.5-4.6)	0.008	4.4 (0.6-6.9)	0.8 (0.6-1.4)	0.078	0.023	0.285
Aldosterone, ng/dl	261 (164-431)	23 (8.1-40)	<0.001	200 (147-351)	23 (12-79)	0.002	0.488	0.239

(J Am Coll Cardiol 2021;77:2174-84)

42



43

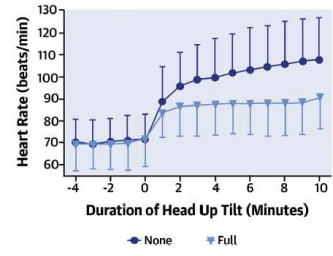
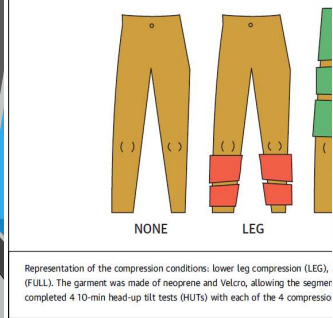


44

Compression Garment For Orthostatic Tachycardia in Patients With Postural Tachycardia Syndrome

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FIGURE 2 Compression Garment Configurations Used in HUT



2021:77(3):285-96

JACC VOL. 77, NO. 3, 2021
JANUARY 26, 2021:285-96

45

Exercise



- Many studies have demonstrated improvement symptoms and QOL with exercise
- Must be approached cautiously with emphasis on starting low and going slow – similar to cardiac rehabilitation
- Warn patients – initially may feel worse
- Improvement will take months not days / weeks
- CHOP / Dallas Protocol

46



Medications

47



General Comments

- Goal of pharmacological therapy is symptomatic improvement not cure
- Individualization
- Trial and error
- Go slow with conservative dosing

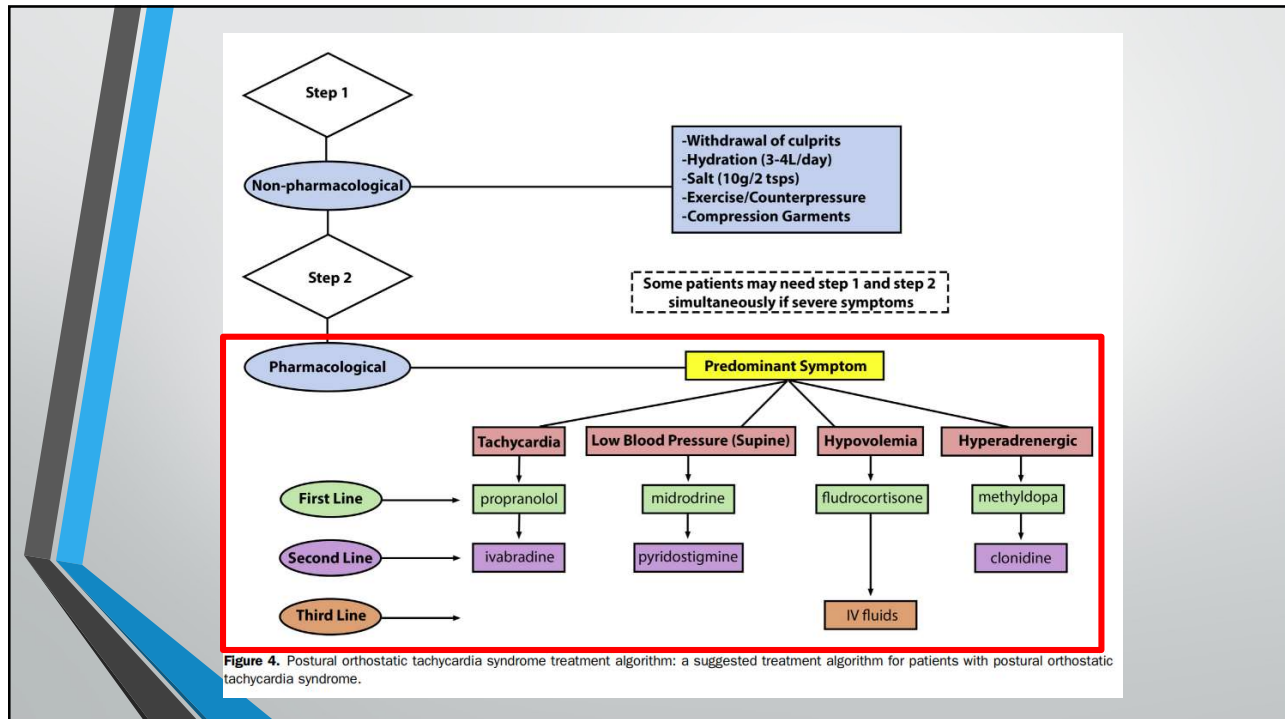
48

Table 2: Pharmacological treatments for postural orthostatic tachycardia syndrome				
Drug	Dosing	Quality of evidence*	Adverse effects	Other considerations
Heart rate inhibitors				
Propranolol	10–20 mg orally up to 4 times daily	Moderate	Hypotension, bradycardia, bronchospasm	Can worsen asthma
Ivabradine	2.5–7.5 mg orally twice daily	Moderate	Visual disturbances, bradycardia	Expensive
Pyridostigmine	30–60 mg orally up to 3 times daily	Low	Increased gastric motility and cramping	
Vasoconstrictors				
Midodrine	2.5–15 mg orally 3 times daily	Moderate	Headache, scalp tingling, supine hypertension	Avoid within 4 hr of bedtime to avoid supine hypertension
Sympatholytic drugs				
Methyldopa	125–250 mg orally twice daily	Low	Hypotension, fatigue, brain fog	Start with a low dose
Clonidine	0.1–0.2 mg orally 2–3 times daily or long-acting patch	Low	Hypotension, fatigue, brain fog	Start with a low dose; withdrawal can lead to rebound tachycardia and hypertension
Blood volume expanders				
Fludrocortisone	0.1 to 0.2 mg orally per day	Low	Hypokalemia, edema, headache	Serum potassium should be monitored
Desmopressin	0.1 to 0.2 mg orally per day, as needed	Low	Hyponatremia, edema	Serum sodium should be monitored if used chronically

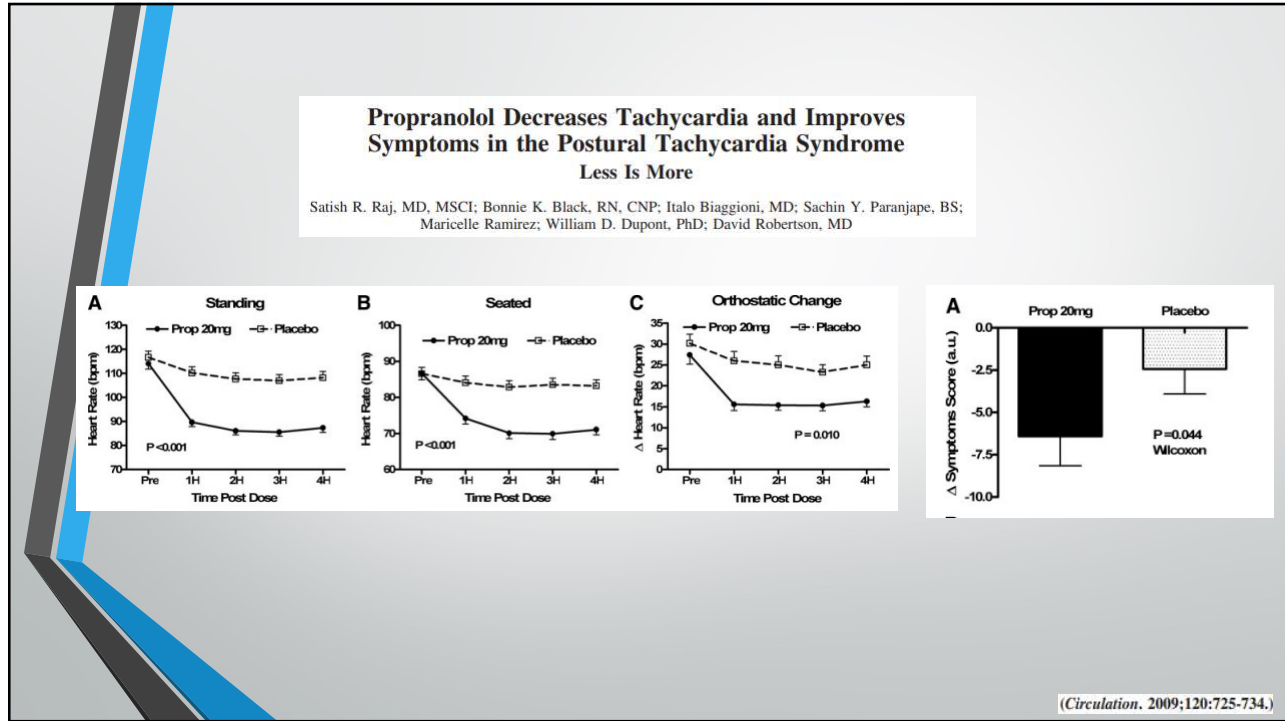
*We critically appraised the literature using the Grading of Recommendations, Assessment, Development and Evaluation (GRADE) methodology.⁴⁴ We rated the quality of the evidence as high, moderate, low or very low based on the likelihood that further research would change confidence in the estimate of effect.

Can J Cardiol 2020; 36(3):357-372

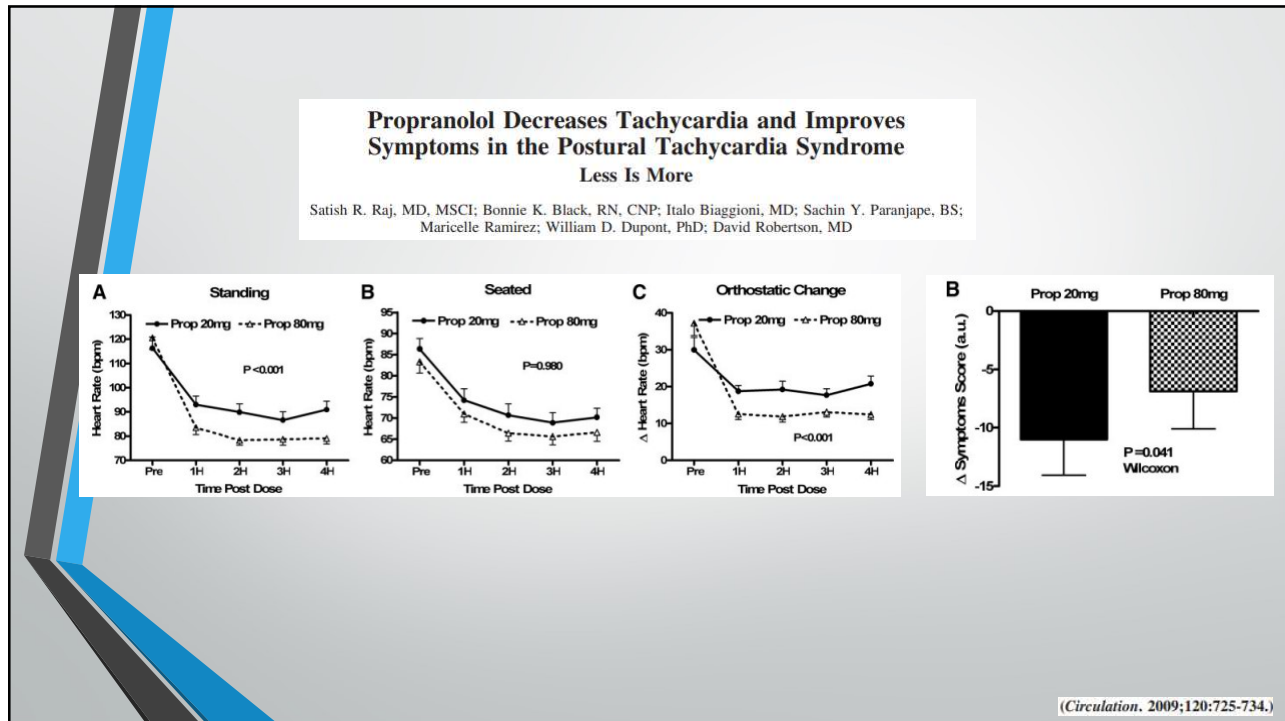
49



50



51



52

Randomized Trial of Ivabradine in Patients With Hyperadrenergic Postural Orthostatic Tachycardia Syndrome

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TABLE 2 The Effect of Ivabradine on Heart Rate

	Baseline	Ivabradine	Placebo	p Value	Cohen's D	95% CI
Supine heart rate, beats/min	73.6 ± 11.7	64.9 ± 6.5	77.5 ± 12.8	0.001*	1.26	0.706-1.820
Standing heart rate, beats/min	95.1 ± 16.8	77.9 ± 9.3	94.2 ± 16.2	0.001*	1.05	0.544-1.58
Delta heart rate (standing vs. supine), beats/min	21.4 ± 15.3	13.1 ± 8.6	17.0 ± 10.4	0.001*	0.753	0.300-1.250

TABLE 3 The Effect of Ivabradine on Quality of Life (SF-36)

	Baseline	Ivabradine	Placebo	p Value	Cohen's D	95% CI
Physical functioning	41.6 ± 21.9	53.4 ± 27.0	44.1 ± 22.4	0.008*	0.570	0.308-0.894
Physical health	13.1 ± 23.5	38.9 ± 40.0	26.1 ± 34.3	0.159	0.223	0.000-0.575
Emotional problems	53.4 ± 45.0	62.1 ± 43.2	57.2 ± 44.7	0.567	0.000	0.000-0.378
Vitality	19.0 ± 16.9	30.8 ± 24.4	23.8 ± 20.5	0.102	0.289	0.000-0.628
Emotional well-being	61.9 ± 16.5	63.9 ± 17.4	60.0 ± 18.8	0.104	0.286	0.000-0.626
Social functioning	43.2 ± 28.7	56.6 ± 31.1	43.8 ± 27.3	0.021*	0.477	0.228-0.800
Pain	48.7 ± 33.2	53.4 ± 30.9	49.2 ± 30.7	0.232	0.146	0.000-0.525
General health	33.5 ± 18.5	33.1 ± 19.3	32.7 ± 18.2	0.897	0.000	0.000-0.046

TABLE 4 The Effect of Ivabradine on NE Levels

	Baseline	Ivabradine	Placebo	p Value	Cohen's D	95% CI
Supine NE, pg/ml	459.43 ± 161.7	472.0 ± 219.3	523.4 ± 223.0	0.316	0.0493	0.000-0.625
Standing NE, pg/ml	1,046.0 ± 346.5	914.3 ± 358.5	1,055.4 ± 371.2	0.076	0.337	0.000-0.829
Delta NE (standing vs. supine), pg/ml	598.1 ± 316.2	442.3 ± 232.6	532.1 ± 259.0	0.056	0.377	0.000-0.866

CENTRAL ILLUSTRATION Ivabradine Improves Heart Rate, Quality of Life, and Norepinephrine Levels in Hyperadrenergic Postural Orthostatic Tachycardia Syndrome

Heart Rate (HR)

Ivabradine significantly lowered:

- HR compared to placebo
- Change in HR from supine to standing

Quality of Life (QOL)

Compared to placebo, ivabradine significantly improved:

- Physical functioning
- Social functioning

Norepinephrine (NE)

Ivabradine decreased:

- Change in NE from supine to standing

Taub, P.R. et al. J Am Coll Cardiol. 2021;77(7):861-71.

Patients with hyperadrenergic postural orthostatic tachycardia syndrome (POTS) experience severe tachycardia with symptoms of orthostatic intolerance upon standing and have a reduced QOL. After 1 month of ivabradine, patients exhibited significant improvements in heart rate and QOL. Our findings illustrate the clinical impact of ivabradine as a safe and effective treatment option for patients with hyperadrenergic POTS.

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FEBRUARY 23, 2021:861-71

53


Box 4: Suggested initial approach to treatment of patient with postural orthostatic tachycardia syndrome

- Nonpharmacological treatments
 - All started at initial visit
 - Water 3 L/d
 - Salt 5 mL/d (2 tsp/d)
 - Waist-high compression garments
- Pharmacological treatments
 - May start at initial visit if symptoms are severe
 - If standing heart rate very high: propranolol 10–20 mg, 4 times per day
 - If standing heart rate very high and β-blocker is contraindicated: ivabradine 5 mg 2 times per day
 - If standing heart rate is not too high and blood pressure is low: midodrine 5 mg orally every 4 hours, 3 times per day (8 am, noon, 4 pm)


Note: tsp = teaspoon.

54

SOME DISABILITIES LOOK LIKE THIS



SOME LOOK LIKE THIS



Questions ???

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