

# Postural Orthostatic Tachycardia Syndrome



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## Case Presentation



- ✎ A 29 year-old woman had been well until 7 months previously when, after a viral syndrome, she developed palpitations, fatigue, and frequent episodes of light headedness and near syncope.
- ✎ On further questioning she notes exercise intolerance and dyspnea on exertion. She has stopped working as cashier.
- ✎ Her mother thinks she is having panic attacks and needs "something to calm her nerves." ECG, echocardiogram, and endocrine evaluation are all normal.
- ✎ On physical examination, she displayed a postural heart rate increase of 35 beats per minute on standing, along with a 15mmHg fall in diastolic blood pressure.

# INTRODUCTION



- ❧ Much of the action of the body in maintaining, cardiovascular, gastrointestinal and thermal homeostasis occurs through the autonomic nervous system (ANS).
- ❧ The ANS is our primary defense against challenges, to maintain homeostasis. It provides involuntary control and organization of both maintenance and stress responses.

3



## What is POTS?

## Introduction



- ❧ Postural Orthostatic Tachycardia Syndrome (POTS) is characterized by an excessive increase in heart rate when a person is in a prolonged upright position and presents with symptoms of orthostatic intolerance.

## History



- ❧ Not widely known, recognized about 15 years ago.
- ❧ Often misdiagnosed as depression, anxiety or patient is told that it's all in their head.

## Demographics of Patients with POTS



- ❧ POTS commonly affects females.
- ❧ POTS patients tend to be younger and often show signs after the onset of puberty.
- ❧ The cause of POTS has a big impact on the demographic and psychographic profile of the individual affected with POTS

## Postural Tachycardia Syndrome(POTS)



A clinical syndrome that is usually characterized by

- ❧ (1) frequent symptoms that occur with standing such as lightheadedness , palpitations, generalized weakness ,blurred vision , exercise intolerance , and fatigue.
- ❧ (2) Increase in heart rate of 30 bpm when moving from a recumbent to a standing position held for more than 30 seconds (or 40 bpm in individuals 12 to 19years of age)
- ❧ (3) Absence of orthostatic hypotension (>20 mmHg drop in systolic blood pressure).

Heart Rhythm, Vol 12, No 6, June 2015



## Physiology

### Peripheral Autonomic Denervation



- ❧ 50% of patients with POTS have a **restricted** autonomic neuropathy of small and distal postganglionic sudomotor fibers, predominantly of the **feet and toes**.
- ❧ Impaired sympathetic tone reduces veno constriction, leading to venous pooling in the **lower limbs and splanchnic beds**.
- ❧ This requires a high cardiac out put to compensate for reduced splanchnic and peripheral Resistance and venous pooling.
- ❧ - The autonomic denervation might be due to an autoimmune disease in some patients.

Heart Rhythm, Vol 12, No 6, June 2015

# Hypovolemia



- ☞ The renin-angiotensin-aldosterone system plays a key role in the neurohormonal regulation of plasma volume in humans.
- ☞ Blood volume is reduced in up to 70% of patients with POTS. Paradoxically, some of these patients **have low plasma renin activity and aldosterone levels compared with healthy subjects.**

(*Circulation*. 2013;127:2336-2342.)

# Hyperadrenergic POTS



- ☞ Systolic blood pressure **increases of 10 mm Hg** while standing upright **for 10 minutes** and plasma norepinephrine levels 600 pg/mL while standing.
- ☞ Patients have heart rate increases similar to those of other patients with POTS .
- ☞ Prominent sympathetic activation symptoms, such as **palpitations, anxiety, tachycardia, and tremors.**
- ☞ **Hypersensitive to isoproterenol** and have marked tachycardic responses to dosages that don't induce hemodynamic changes in healthy

# Deconditioning Patients

- ⌘ POTS patients often have poor exercise tolerance and deconditioning.
- ⌘ Deconditioned patients with POTS have reduced left ventricular mass, stroke volume, and blood volume, which improve with exercise training.
- ⌘ Stroke volume decreases in POTS, with impaired cardiac filling when standing.
- ⌘ This situation suggests that orthostatic symptoms induced by an initial illness can, in some patients, lead to over interpretation of the symptoms due to hypervigilance, which in turn leads to reduced physical activity and deconditioning.

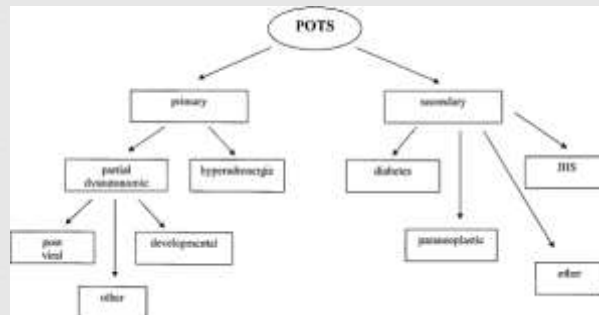
	NORMAL PHYSIOLOGY	"VASOVAGAL" SYNCOPE	POTS
Venous return	↓	↓	↓
Stroke volume	↓	↓	↓
Pulse pressure	↓	↓	↓
Sympathetic tone	↑		↑↑↑
Vagal tone	↓	↑	
Heart rate	↑ (10 - 15 bpm)	↓	↑↑↑ (>30 bpm)
Systolic pressure	Stable	↓↓	↓
Diastolic pressure	↑ (~10 mm)	↓	↓ <sup>14</sup>

# What causes POTS?



- ❧ Adrenal Disorders (such as Addison's disease)
- ❧ Anemia
- ❧ Autoimmune Disorders
- ❧ Cardiac Disease
- ❧ Lesions
- ❧ Liver Disease
- ❧ Neuropathy
- ❧ Tumors
- ❧ Viruses

## *Subtypes of postural orthostatic tachycardia syndrome*



[BJMP 2012;5\(4\):a540](#)

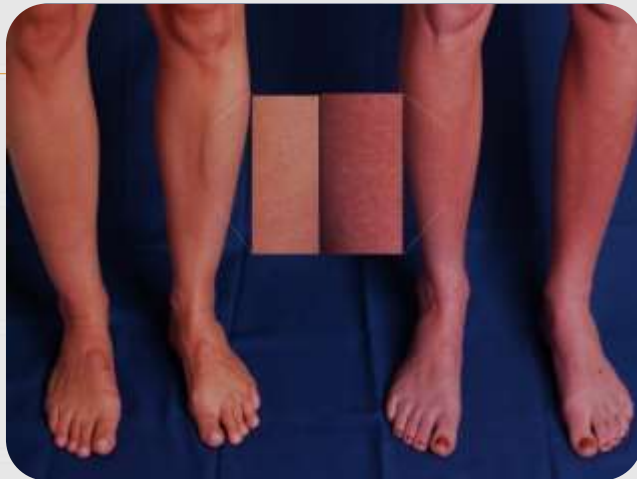


# Symptoms

- ☞ Lightheadedness
- ☞ Exercise intolerance
- ☞ Extreme fatigue
- ☞ Excessive thirst
- ☞ Fainting
- ☞ Cold extremities
- ☞ Chest pain and discomfort
- ☞ Disorientation
- ☞ Ringing in ears
- ☞ Shortness of breath
- ☞ Headache
- ☞ Muscle weakness
- ☞ Visual disturbances
- ☞ Bloating
- ☞ Constant nausea
- ☞ Irritability
- ☞ Concentration problems
- ☞ Mental clouding

While pre-syncope is common in these patients, only a minority (~30%) actually pass out.

# Got that Purple



## How does POTS Affect one's life?



- ⌘ Makes everyday activities very difficult
- ⌘ Patients lack motivation
- ⌘ Exercise is extremely difficult
- ⌘ Some patients find difficulty going to work or school based on severity of symptoms.

## Grading of Orthostatic Intolerance

GRADE 0	Normal orthostatic tolerance
GRADE I	Orthostatic symptoms infrequent or occur only under increased orthostatic stress Patient can stand > 15 minutes most occasions ADL typically unrestricted
GRADE II	Orthostatic symptoms frequent (weekly or more often) and commonly develop with orthostatic stress Patient can stand > 5 minutes on most occasions Typically some limitation in ADL
GRADE III	Orthostatic symptoms develop on most occasions and are regularly unmasked by orthostatic stresses Patient can stand > 1 minute on most occasions Patient is seriously incapacitated (bed- or wheelchair-bound) Syncope/presyncope common



## Do all POTS patients faint?

### Recommended Investigations of POTS

	Class	Level
A complete history and physical exam with orthostatic vital signs and 12-lead ECG should be performed on patients being assessed for POTS.	I	E
Complete blood count and thyroid function studies can be useful for selected patients being assessed for POTS.	IIa	E
A 24-hour Holter monitor may be considered for selected patients being assessed for POTS, although its clinical efficacy is uncertain.	IIIb	E
Detailed autonomic testing, transthoracic echocardiogram, tilt-table testing, and exercise stress testing may be considered for selected patients being assessed for POTS.	IIIb	E

Heart Rhythm, Vol 12, No 6, June 2015

## Expanded approach to the evaluation

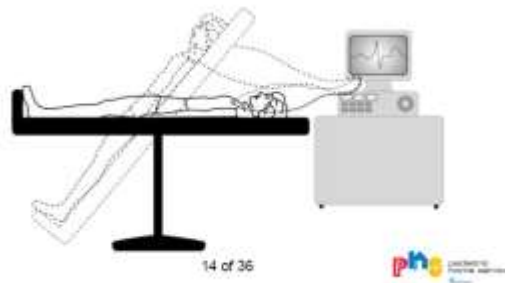


- Thermoregulatory sweat test to detect autonomic neuropathy (which manifests as abnormal patterns of body sweating).
- Supine and upright plasma epinephrine and norepinephrine level tests.
- 24-hour urine sample to assess sodium intake.
- Psychological assessment.

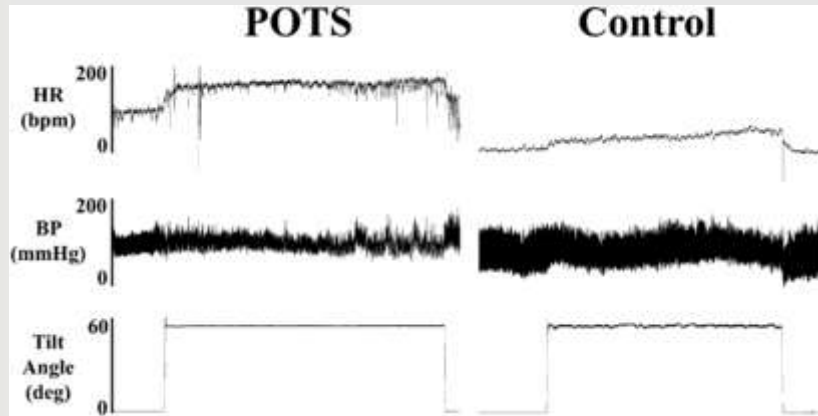


## diagnosis

- Tilt table



Heart rate (HR) and blood pressure (BP) with upright tilt in postural tachycardia syndrome (POTS).

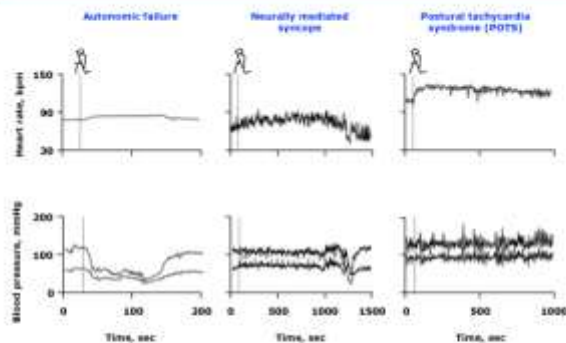


Satish R. Raj *Circulation*. 2013;127:2336-2342



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Heart rate and blood pressure patterns observed in head-up tilt table testing



Shown are the heart rate and blood pressure responses seen during tilt table testing in patients with various etiologies of syncope, including autonomic failure, neurally mediated syncope, and postural tachycardia syndrome (POTS). The absence of an appropriate reflex-induced increase in heart rate as the blood pressure falls is a useful clue to the presence of autonomic failure. During neurally mediated syncope, parasympathetic (vagal) activity increases as blood pressure declines, slowing the heart. With POTS, tilt table testing typically reproduces the clinical symptoms in association with a heart rate increase  $\geq 30$  beats/min or a maximum heart rate  $\geq 120$  beats/min within the first 10 minutes; these changes are not associated with hypotension.



## What sort of things should a POTS patient avoid?

- ⌘ Direct sunlight, tanning booths, any way to become dehydrated easily
- ⌘ Overexertion without proper cardiac rehab
- ⌘ Ablation of the sinus node
- ⌘ Alcohol
- ⌘ Bending up and down
- Blowing up Balloons
- Excessive stair climbing
- Energy Drinks
- Epinephrine
- Doctor's who don't know anything about POTS
- Giving Blood
- Travel by Airplane
- Singing and instruments
- (Thanks to Dinet.org!)





## Nonpharmacological Treatment of POTS

## Things that help POTS



☞ Exercise. This is one of the most difficult things for POTS patients cause their bodies constantly are telling them they are fatigued.

☞ Eating more salt.

☞ Drinking a lot more fluid then one normally would.



## Common Challenges Faced by Patients with POTS



- ❧ Starting an exercise regimen tends to be more difficult.
- ❧ Social anxiety from fear of fainting in social setting.
- ❧ Attending work & school is hard due to stronger symptoms in the morning.
- ❧ Frustration due to others misunderstanding the disease.



## Pharmacological approach



Pharmacological approach is to withdraw medications that might be predisposing to tachycardia



#### Drugs that can cause or worsen orthostatic intolerance

- α- Receptor blockers
- Angiotensin-converting-enzyme inhibitors
- β-Blockers
- Bromocriptine
- Calcium channel blockers
- Diuretics
- Ethanol
- Ganglionic blocking agents
- Hydralazine
- Monoamine oxidase inhibitors
- Nitrates
- Opiates
- Phenothiazines
- Sildenafil citrate (Viagra)
- Tricyclic antidepressants

[BJMP 2012;5\(4\):a540](#)

## β-Adrenergic blockers



- ☞ Commonly used in cardiology clinics to control tachycardia.
- ☞ Many patients with POTS, however, complain of excessive fatigue or intolerance to β-blockers.
- ☞ Reducing the HR in POTS would be counterproductive if the increase in HR were purely compensatory for another physiological shortfall (eg, low stroke volume) but could be useful if the tachycardia were overcompensation for the physiological stimuli.
- ☞ low-dose short-acting propranolol (10-20 mg orally) to be very effective at lowering standing HR and improving symptoms in POTS

# Midodrine



- ⌘ peripheral  $\alpha$ -1 agonist that serves as a vasoconstrictor. It might be most useful in patients with neuropathic POTS, which can be associated with a failure of vascular resistance.
- ⌘ midodrine reduces orthostatic tachycardia, but this effect is more modest than that of intravenous saline. Midodrine can cause goose bumps, scalp tingling, or headaches, which can limit its tolerability.

(*Circulation*. 2013;127:2336-2342.)

# Pyridostigmine



- ⌘ A peripheral acetylcholinesterase inhibitor that can increase the levels of synaptic acetylcholine at both the autonomic ganglia and peripheral muscarinic parasympathetic receptors.
- ⌘ Pyridostigmine 30 to 60 mg orally 3 times a day has been reported to result in long-term symptom improvement in  $\approx$ 50% of POTS patients.
- ⌘ Enhance bowel motility; thus, gastrointestinal adverse events are the most common reason for discontinuation of the drug.

(*Circulation*. 2013;127:2336-2342.)

## Recommended Treatment for POTS

	Class	Level
A regular, structured, and progressive exercise program for patients with POTS can be effective.	IIa	B-I
It is reasonable to treat patients with POTS who have short-term clinical decompensations with an	IIa	C
Drugs that block the norepinephrine reuptake transporter can worsen symptoms in patients with POTS and should not be administered.	III	B-I
Regular intravenous infusions of saline in patients with POTS are not recommended in the absence of evidence, and chronic or repeated intravenous cannulation is potentially harmful.	III	E
Radiofrequency sinus node modification, surgical correction of a Chiari malformation type I, and balloon dilation or stenting of the jugular veins are not recommended for routine use in patients with POTS and are potentially harmful.	III	B-III
It seems reasonable to treat patients with POTS who have prominent hyperadrenergic features with clonidine or alpha-methyldopa.	III	I

Heart Rhythm, Vol 12, No 6, June 2015

## What is a POTS emergency?



- ❧ Blood Pressure/Pulse out of control, erratic, or long episodes of tachycardia
- ❧ Outstanding pain or discomfort
- ❧ Dehydration (often leading to migraines, or other symptoms)

## What can be done at the ER to help a POTS patient?



- ❧ Patients can be given
- ❧ Correctional beta blockers
- ❧ IV saline.
- ❧ pain medication
- ❧ Muscle relaxants

## Prognosis



- ❧ There is limited data on the prognosis of POTS patients. Recent short term follow-up studies have shown better prognosis in patients with POTS. Roughly 50% of post-viral POTS patients make a meaningful recovery over about 2-5 years.
  
- ❧ Some patients experience a partial recovery and still others may demonstrate a progressive functional decline with time. As a general principle, a younger age of onset portends a better prognosis.

[BJMP 2012;5\(4\):a540](#)

# So,,,

☞ True or False?



POTS symptoms only occur while standing.

FALSE: Many patients report symptoms occurring while sitting or lying down. Standing does exacerbate symptoms.

Everyone with POTS faints.

FALSE: Many people with POTS have never fainted.

What do you do if you feel faint?

- A. Stand and continue to converse hoping the spell will pass
- B. Stand next to someone you think will catch you
- C. Run in hopes of increasing heart rate
- D. Lay down with legs bent at knees

## THANK YOU

