

Postural Orthostatic Tachycardia Syndrome -An Invisible Disability

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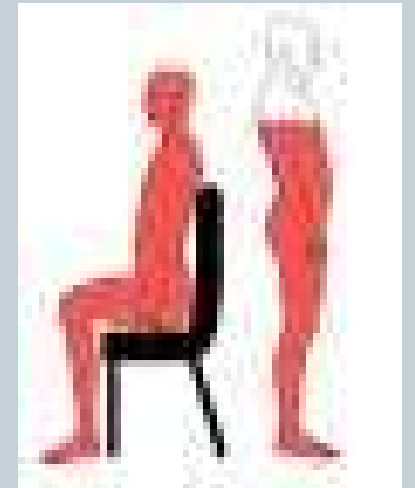


Definition



- POTS is defined as the presence of chronic symptoms of orthostatic intolerance (at least 6 month) accompanied by an increased Heart Rate ≥ 30 bpm within 10 minutes of assuming an upright posture and the absence of orthostatic hypotension.

- * Postural vital signs should be performed in morning.
- * Orthostatic tachycardia diagnosis need exclusion of other causes:
 - Prolonged bed rest
 - Drugs: Vasodilator, Diuretic, Antidepressant
 - Dehydration , Anemia , Hypothyroidism



Epidemiology



- POTS is the most prevalent form of orthostatic intolerance
- Patients present at a relatively young age (14 to 45 years)
- Predominantly effects woman with a female to male ratio of 4-5: 1
- In USA: 500,000 suffer from this disorder. It is the most common syndrome of young people seen in autonomic dysfunction clinics.



Fast Facts on POTS

(Postural Orthostatic Tachycardia Syndrome)



25% of POTS patients are so **DISABLED** they can't work.



85% of POTS patients are *female*.

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ZERO DRUGS approved by the FDA to treat POTS.

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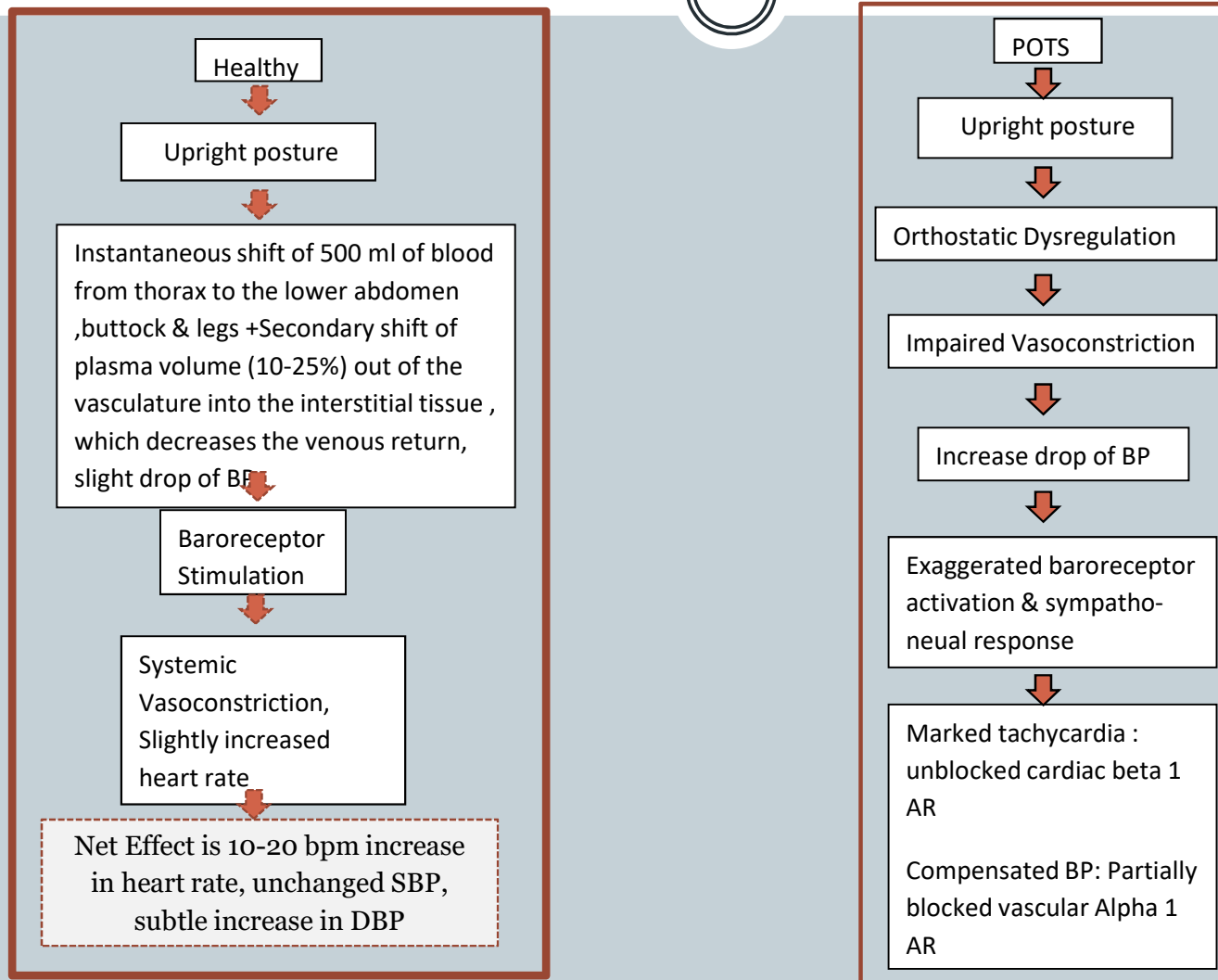
The many prescriptions used to treat POTS are all "off-label."

85%

of patients developed POTS after an **infection.**



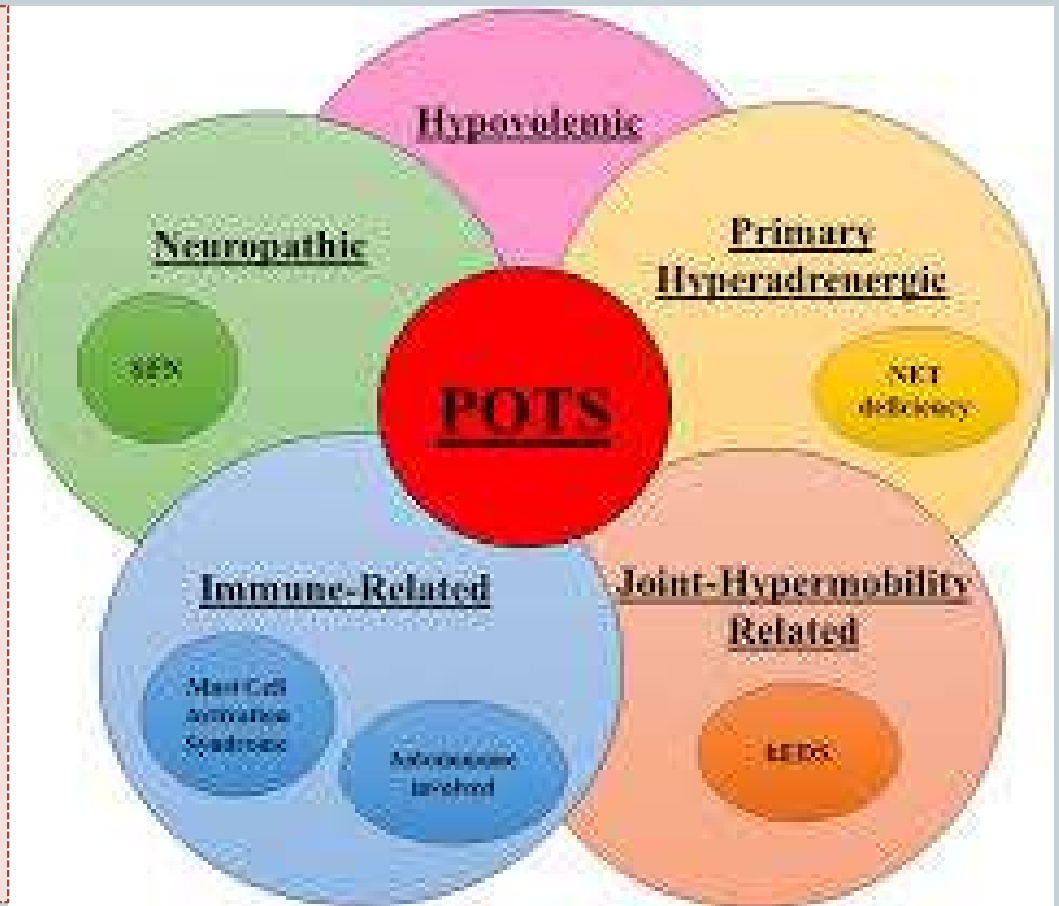
Pathophysiology



Types



- ❑ Neuropathic POTS
- ❑ Hyperadrenergic POTS
- ❑ Norepinephrine transporter deficiency



Clinical Features

- **Cardiac :**

- Palpitation
- Lightheadedness
- Chest discomfort
- Dyspnea
- Pre-syncope

- **Non Cardiac:**

- Mental Clouding(Brain Fog)
- Headache
- Nausea
- Tremulousness
- Slurred or Tunnel speech
- Poor Sleep
- Extreme Fatigue
- 30% Faint
- Un-refreshing Sleep
- Daytime Sleepiness

Psychiatric: Symptom-related anxiety, hypervigilance, depression

Cardiovascular: Increased heart rate, palpitations, chest pain or discomfort, high or low blood pressure

GI: Nausea, diarrhea, constipation, bloating, abdominal cramping

Genitourinary: Increased urination, difficulty urinating

Extremities: Increased sweating to extremities, cool and clammy feet and hands

Decreased cerebral perfusion: Brain fog, impaired memory, headache, lightheadedness, blurred vision, dizziness, fainting, poor sleep quality

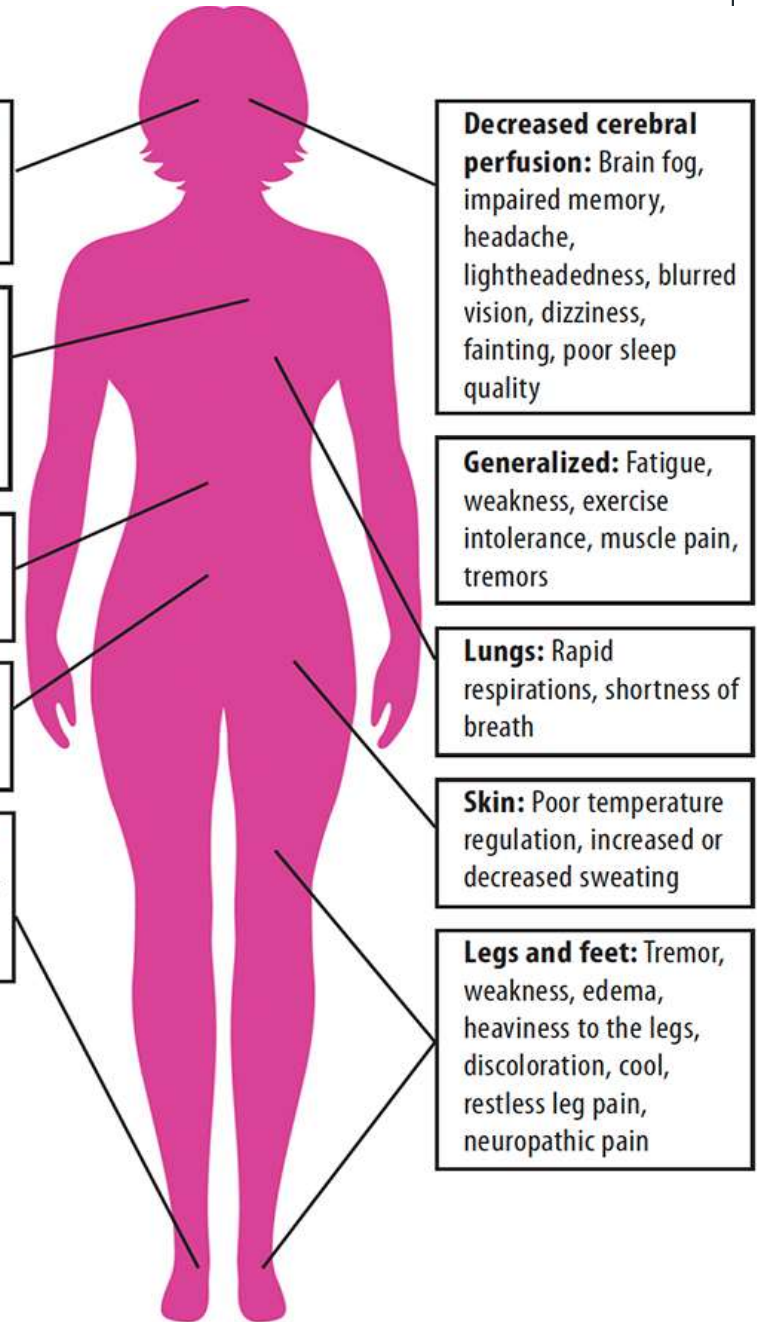
Generalized: Fatigue, weakness, exercise intolerance, muscle pain, tremors

Lungs: Rapid respirations, shortness of breath

Skin: Poor temperature regulation, increased or decreased sweating

Legs and feet: Tremor, weakness, edema, heaviness to the legs, discoloration, cool, restless leg pain, neuropathic pain

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Clinical feature (continued...)



- **Physical Sign :**
- ❑ Cardiac auscultation may reveal a murmur of mitral valve prolapse, but significant mitral regurgitation is unusual.
- ❑ A striking physical feature of POTS is the dependent acrocyanosis that occurs in 40-50% of patients with POTS .These patients experience a dark red-blue discoloration of their legs, which are cold to the touch. This can extend from the feet to above the level of the knees



Trigger Factor

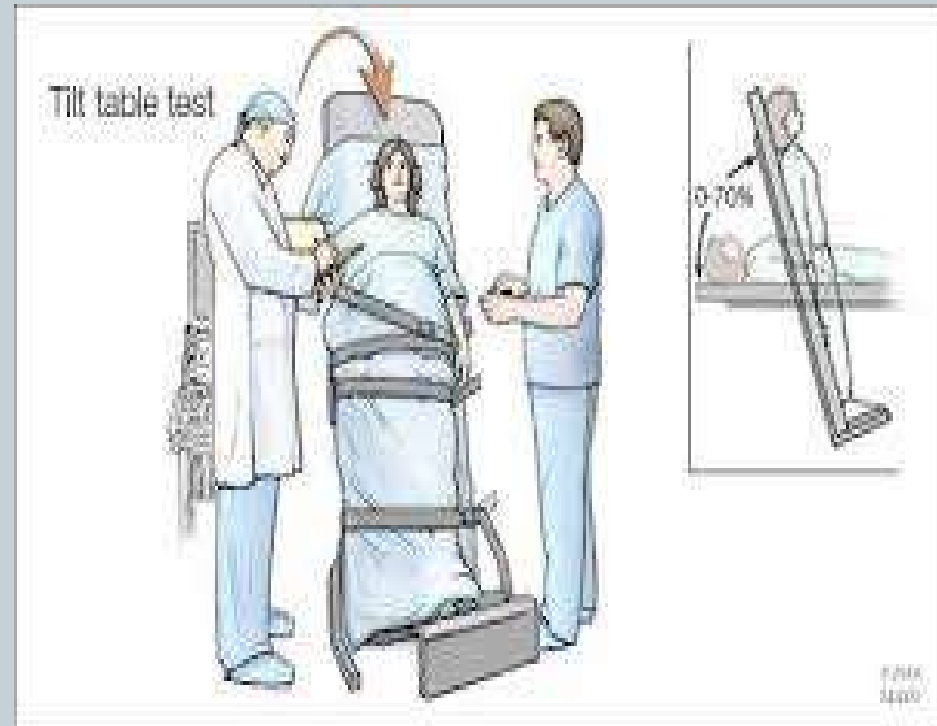


- **Symptoms often began following acute stressors such as**
 - Pregnancy
 - Major surgery,
 - Presumed viral illness,
 - About 80% of female patients report an exacerbation of symptoms in the pre-menstrual phase of their ovulatory cycle.
- **Secondary Triggers :**
 - Alcoholism
 - Autoimmune disease
 - Prolonged bed rest
 - Central hypotension (medication)
 - Chemotherapy
 - Diabetes
 - Heavy metal poisoning
 - Peripheral ANS Damage

Diagnosis



- POTS diagnosis can be complicated because the symptoms can affect a wide range of organ systems, and the symptoms for each patient may differ.
- POTS can be confirmed by performing physical exam such as a standing test or a head-up tilt table test.



Diagnostic Criteria for the Postural Orthostatic Tachycardia Syndrome

- Heart Rate increases ≥ 30 bpm from supine to standing (10 minutes).
- Symptoms worsen with standing and improve with recumbence
- Symptoms last ≥ 6 months
- Absence of other overt cause of orthostatic symptoms or tachycardia
- Absence of orthostatic hypotension (≥ 20 mmhg drop in systolic blood pressure)



Lab Investigations



Exclusion of Other Cause :

- CBC
- S. Electrolyte
- Iron Profile
- Vitamin B12
- Plasma & Urine Metanephrine
- ECG
- Holter Monitoring
- Echocardiogram

Supportive Investigations:

- ✓ Plasma Norepinephrine – Supine & Standing (10 min at each position)
- Supine norepinephrine is often within normal range
- Upright norepinephrine : > 600 pg/ml
- ✓ Blood Volume can be directly measured by nuclear medicine .
- ✓ QSART
- ✓ Anti gAChR antibody

Concurrent clinical syndromes



- Irritable bowel syndrome (IBS)
- Anxiety
- Chronic fatigue syndrome
- Depression
- Fibromyalgia
- Raynaud's phenomenon
- Mitral Valve Prolapse
- Hypermobility syndrome
- Vascular compression syndromes
- Mast Cell Activation Syndrome
- Ehlers-Danlos syndrome
- Joint hypermobility disorders
- Migraine
- Addison's disease
- Spontaneous intracranial hypotension

Differential Diagnosis



- These conditions can produce similar symptoms to POTS.....
- Anxiety
- Pheochromocytoma
- Autonomic neuropathies,
- Central dysautonomias,
- Bed rest deconditioning,
- Side effects of medications,
- Dehydration

Patients with POTS may be misdiagnosed to have panic, anxiety, somatization disorder, or chronic fatigue syndrome (CFS), also known as myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS), in part because of the vague nature of the symptoms.

Management



- No therapy is uniformly successful
- Combination approach is needed.

Nonpharmacologic measures :

- Exacerbating factors, medications, dehydration, and inactivity should be avoided.
- **Exercise program**
 - Primarily aerobic with some leg based resistance exercise.
 - Initially avoid upright exercise
 - Focus on rowing machines, swimming & recumbent cycling
- **Augment blood volume/venous return**
 - Increase water intake : 8-10 cups/day (2-2.5 L)
 - Increase NACL intake: 8-10 g /day
 - I/V Saline : 1 L over 1-2 hour (Short term emergency treatment)
 - Compression stockings :Panty hose style- 30-40 mm Hg Counter Pressure

Management (Continued...)



- **Medications:**

- Fludrocortisone

- DDAVP

- **Hemodynamic Agent**

- Propranolol

- Pyridostigmine

- Midodrine

- Modafinil

- **Central Sympatholytic agent**

- Clonidine

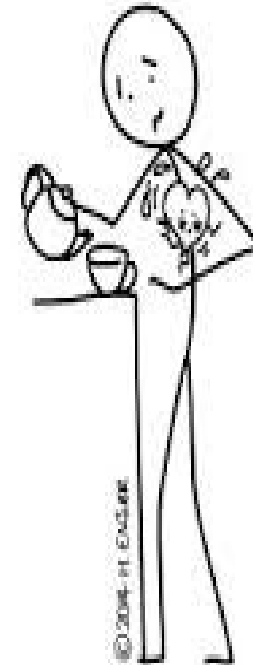
- Methyldopa

Take Home Message

POTS – An Invisible Illness

- People with postural orthostatic tachycardia syndrome have about the same level of disability as someone with chronic obstructive pulmonary disorder (COPD) or heart failure.
- Average time to diagnosis from symptom onset is 5 years and 11 months.

You know you have POTS when...



...you make a cup of tea and your heart thinks you've run a marathon.

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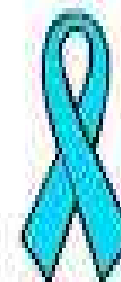
A patient's statement – I don't feel good



HI THERE!

POTS is an invisible illness – that means that you can't look and see my level of suffering. When most people get sick, they have a fever, swollen glands, or runny nose that allows others to see that they are not feeling well. That is not the case for people like me who are living with invisible illnesses. What does it look like if I am dizzy? Can you assess my level of fatigue without feeling it yourself? What does neuropathic pain look like to the outside observer? Please believe me when I tell you that I don't feel good!

THANK
YOU



strength & Courage
Faith

POTS

Hope & AWARENESS

LOVE

TLM Invisible Illness Artwork