

POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME

What is Postural Orthostatic Tachycardia Syndrome?

Postural Orthostatic Tachycardia Syndrome (POTS) is one of the more common forms of dysautonomia. The key features of POTS are a **sustained increased heart rate upon standing** and **orthostatic intolerance symptoms**. However, people with POTS also experience symptoms regardless of position.

The diagnostic criteria for POTS include:^{1,2}

- A *sustained* increase in heart rate of at least 30 beats per minute (bpm) in adults and 40 bpm in individuals aged 12-19 *within 10 minutes of standing*.
- No orthostatic hypotension (no drop in blood pressure of more than 20/10 mm Hg).
- Symptoms of orthostatic intolerance (lightheadedness, palpitation, tremors, chest discomfort) have been present for at least 3 months.
- Other conditions that cause increased heart rate have been ruled out (i.e. panic attacks, pain, exercise, caffeine, alcohol, medications, anemia, inappropriate sinus tachycardia, hyperthyroidism, and dehydration).

Current guidelines state that, generally, one person can not have orthostatic hypotension and POTS.^{3,4} Finally, people with POTS can pass out, but this is not a key symptom. In fact, only about 30% of those with POTS experience syncope.^{5,6}

Symptoms

Symptoms may include:^{2,7}

- Orthostatic tachycardia (increased heart rate when standing)
- Orthostatic intolerance (dizziness, lightheadedness, weakness, shortness of breath, chest tightness, chest pain, heart palpitations, and/or tremors)
- Fatigue
- Autonomic symptoms (ex: blood pooling, gastrointestinal symptoms, increased or decreased sweating)
- Headaches
- Cognitive dysfunction (brain fog)
- Pain
- Sleep disturbance
- Exercise intolerance

Incidence/Causes

It is estimated that between **500,000 - 3 million** Americans have POTS.^{2,8} More than 75% with POTS are female^{7,9} and the average age of onset is between 15-25 years of age.⁷

Not everyone is able to identify a cause, trigger, or precipitating event for their POTS. However, for some people, symptom onset has occurred after **viral illnesses/infection, a surgical procedure, concussion, or during or after pregnancy**.^{3,5,10} Researchers have found that POTS commonly occurs with other conditions such as **chronic fatigue syndrome, small fiber neuropathy, mast cell activation disorder, hypermobile Ehlers-Danlos syndrome, migraines, gastrointestinal dysfunction (e.g. irritable bowel syndrome), autoimmune disorders, and fibromyalgia**.⁹⁻¹¹



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Subtypes

There are **different pathophysiological processes** that lead to different subtypes within POTS. The subtypes of POTS can help in the understanding of the mechanisms that lead to the autonomic dysfunction and symptoms seen in the condition. *It is important to note that among the medical community there are no established medical definitions for each subtype that are uniformly accepted.*¹¹

Also, patients often do not fit into just one subtype. Some medical professionals will not give patients a subtype at their time of diagnosis. This is because treatment often needs to be individualized to each patient and sometimes people exhibit findings from several subtypes.^{10,12}

Hypovolemic POTS

Hypovolemic POTS refers to patients who have a **deficit in blood volume**.¹⁴ Researchers have found that some people with POTS can have approximately a 13% reduction in plasma volume compared to healthy individuals.¹⁵ It is believed that these individuals might have dysregulation of the renin-angiotensin-aldosterone (RAA) system and decreased angiotensin II metabolism contributing to low fluid volume and impaired sodium retention.^{14,16} The RAA system works to elevate blood volume and arterial tone by increasing sodium reabsorption, water reabsorption, and vascular tone.¹⁷ As noted above, hypovolemia can cause/appear like hyperadrenergic POTS. In these people, the body compensates for the decreased blood volume by increasing central sympathetic activation, such as *palpitations*.

Hyperadrenergic POTS

Hyperadrenergic POTS can be characterized by excessive tachycardia upon standing, an increase in systolic blood pressure of more than 10 mmHg, and elevated standing norepinephrine levels¹⁰ of 600 pg/mL or more.⁴ Patients may also experience symptoms of sympathetic activation such as *palpitations, sweating, nausea, and abdominal pain*.¹¹ Hyperadrenergic POTS may be secondary to another form of POTS like hypovolemic or neuropathic.¹⁰

Immune-mediated POTS

Some patients report symptom onset after an *acute viral illness or along with autoimmune conditions*, suggesting that autoimmune factors could play a role in the development of POTS.^{10,16} Patients with POTS do have a higher frequency of autoimmune disorders like Sjorgren syndrome, lupus, celiac disease, and rheumatoid arthritis.^{18,19} Studies have reported autoantibodies and non-specific autoimmune markers in people with POTS, however, more research is needed to understand the impact of those antibodies.¹⁹

Neuropathic POTS

Patients with neuropathic POTS often have some dysfunction with sympathetic denervation, usually a *small fiber neuropathy*.¹³ They can present with patchy areas of skin in the legs/feet where they do not sweat.^{3,10} Sympathetic denervation seen in neuropathic POTS can lead to *increased blood pooling in the lower extremities and in the abdomen*.¹⁰ The blood pooling results in less blood being able to make it back up to the heart, so sympathetic activation then leads to an increased heart rate.¹¹



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Diagnosis

The current “gold standard” for diagnosing POTS is the **Tile Table Test (TTT)** but some doctors also diagnose POTS with a poor man's tilt instead.¹⁰

Other tests are often also performed in route to diagnosing POTS in order to rule out other causes of orthostatic tachycardia (e.g. routine blood work, Holter monitor, etc.) but are not required for POTS diagnosis.² Also, some people with POTS will receive additional autonomic testing (e.g. QSART, plasma norepinephrine) but current guidelines do not list this as a routine recommendation.⁷



For references, please scan the QR code. (C) 2022

Treatment

POTS can be managed with both pharmacologic and nonpharmacologic methods.

Oftentimes treatment starts with **lifestyle adaptations** such as:^{11,20}

- Increasing salt and water intake
- Learning physical counter maneuvers to counteract symptoms
- Wearing compression garments
- Exercise training.

If non-pharmacologic lifestyle adaptations are not successful in reducing symptoms, then a **pharmacological approach** is considered through the use of medications.

Various medications can be prescribed to:^{4,7,20}

- Increase the fluid volume within the body (Fludrocortisone)
- Increase constriction of blood vessels (Midodrine), and/or
- Decrease heart rate (beta-blockers)

Please note, these are examples of medications used in POTS treatment and not an all-inclusive list.



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