Postural Orthostatic Tachycardia Syndrome

Postural Orthostatic Tachycardia Syndrome (POTS) is a form of dysautonomia. POTS is a subset of orthostatic intolerance that is associated with the presence of excessive tachycardia on standing.¹

How is POTS Diagnosed?

The current diagnostic criteria for POTS is a heart rate increase of 30 beats per minute (bpm) or more, or over 120 bpm, within the first 10 minutes of standing.¹,²,³,⁴ In children and adolescents, a revised standard of a 40 bpm or more increase has recently been adopted.⁴,⁵ POTS is often diagnosed by a Tilt Table Test, but if such testing is not available, POTS can be diagnosed with bedside measurements of heart rate and blood pressure taken in the supine (laying down) and standing up position at 2, 5 and 10 minute intervals.

What are the Symptoms?

While the diagnostic criteria focus on the abnormal heart rate increase upon standing, POTS usually presents with symptoms much more complex than a simple increase in heart rate. It is fairly common for POTS patients to have a noticeable drop in blood pressure upon standing, but some POTS patients have no change or even an increase in blood pressure upon standing.¹ POTS patients often have hypovolemia (low blood volume) and high levels of plasma norepinephrine while standing, reflecting increased sympathetic nervous system activation.³ Approximately 50% of POTS patients have some extent of autonomic neuropathy.⁶ POTS patients may also experience fatigue, headaches, lightheadedness, heart palpitations, exercise intolerance, nausea, diminished concentration, tremulousness (shaking), syncope (fainting), coldness or pain in the extremities, chest pain, shortness of breath, gastric motility problems and many other symptoms.¹,³,⁴

Some patients have fairly mild symptoms and can continue with normal work, school, social and recreational activities. For others, symptoms may be so severe that normal life activities, such as bathing, housework, eating, sitting upright, walking or standing can be significantly limited.¹,³ Approximately 25% of POTS patients are disabled and unable to work.¹ Physicians with expertise in treating POTS have compared the functional impairment seen in POTS patients to the impairment seen in chronic obstructive pulmonary disease (COPD) or congestive heart failure.¹ They note that while the symptoms are real and can severely limit a patient's ability to function, POTS patients are too often misdiagnosed as having severe anxiety or panic disorder.¹,³ Modern research has shown that POTS patients are similarly or even less likely to suffer from anxiety or panic disorder than the general public.³,⁵,⁷,⁸
History of POTS

The term "POTS" was coined in 1993 by a team of researchers from Mayo Clinic, led by neurologist Dr. Philip Low. However, POTS is not a new illness; it has been known by other names throughout history, such as DaCosta's Syndrome, Soldier's Heart, Mitral Valve Prolapse Syndrome, Neurocirculatory Asthenia, Chronic Orthostatic Intolerance, Orthostatic Tachycardia and Postural Tachycardia Syndrome. In the past, it was mistakenly believed to be caused by anxiety. However, modern researchers have determined that POTS is not caused by anxiety. It is caused by a malfunction of the patient's autonomic nervous system. Thankfully, in the last 20 years, researchers have gained much more insight into imbalances of the autonomic nervous system.

Autonomic Nervous System Diagram

POTS researchers have classified POTS in various ways, such as "Primary" and "Secondary" POTS; "high flow" and "low flow" POTS; and also by the primary symptoms, to wit, hypovolemic POTS (POTS associated with low blood volume), partial dysautonomic POTS (associated with a partial autonomic neuropathy and sometimes referred to as neuropathic POTS) and hyperandrenergic POTS ("hyper" POTS is associated with elevated levels of norepinephrine). From a practical point of view, most patients do not fit neatly within one of these classifications. Having neuropathic POTS does not mean you can't also have hypovolemia and/or elevated norepinephrine levels. All of these characteristics can overlap.

Who Develops POTS?

POTS can strike any age or gender, but it is most often seen in women of child bearing age (between the ages of 15 and 50). Men and boys can develop it as well, but approximately 75% to 80% of patients are female. Researchers estimate that 500,000 to 1,000,000 Americans have POTS.
What Causes POTS?

POTS is a heterogeneous (meaning it has many causes) group of disorders with similar clinical manifestations.¹,⁴ POTS itself is not a disease; it is simply a cluster of symptoms that are frequently seen together. This is why the 'S' in POTS stands for "Syndrome." Since POTS is not a disease, it is fair to say that POTS is caused by something else. However, figuring out what is causing the symptoms of POTS in each patient can be very difficult, and in many cases, patients and their doctors will not be able to determine the precise underlying cause. When doctors cannot pinpoint the underlying cause of a patient's POTS, it may be called Primary or Idiopathic POTS.¹ Idiopathic simply means "of an unknown origin." POTS caused by an identified underlying problem may be called Secondary POTS.

While researchers are still working to identify the root causes and pathology of POTS, there are several underlying diseases and conditions that are known to cause or be associated with POTS or POTS like symptoms in some patients. This is a partial list:

- Amyloidosis;¹
- Autoimmune Diseases such as Autoimmune Autonomic Neuropathy (also called Autoimmune Autonomic Ganglionopathy), Sjogren's Syndrome, Sarcoidosis and Lupus;¹,³
- Chiari Malformation²⁰
- Deconditioning;⁴
- Delta Storage Pool Deficiency¹⁴
- Diabetes and Pre-Diabetes;²³
- Ehlers Danlos Syndrome;³,¹³
- Genetic Disorders/Abnormalities;³
- Infections such as Mononucleosis, Epstein Barr Virus, Lyme Disease, extra-pulmonary Mycoplasma pneumonia and Hepatitis C;¹,²,³,⁴,¹¹,¹²
- Multiple Sclerosis;¹⁵
- Mitochondrial Diseases;¹⁶
- Mast Cell Activation Disorders;³
- Paraneoplastic Syndrome - rare small tumors of the lung, ovary, breast and pancreas that produce antibodies;¹
- Toxicity from alcoholism, chemotherapy and heavy metal poisoning;¹
- Traumas, pregnancy or surgery;¹,²,³
- Vaccinations;¹⁹
- Vitamin Deficiencies/Anemia;¹⁷,¹⁸

Treatment

Each patient is different, thus consulting with a physician who has experience in treating autonomic disorders is important. The most common treatments for POTS include increasing fluid intake to 2-3 liters per day; increasing salt consumption to 3,000 mg to 10,000 mg per day (except in Hyperandrenergic POTS); wearing compression stockings; raising the head of the bed (to conserve blood volume); reclined exercises such as rowing, recumbent bicycling and swimming; a healthy diet; avoiding substances and situations that worsen orthostatic symptoms; and finally, the addition of medications meant to improve symptoms.¹,³ Many different medications are used to treat POTS, such as Fludrocortisone, Beta Blockers, Midodrine, Clonidine, Pyridostigmine, Benzodiazepines, SSRIs, SNRIs, Erythropoietin and Octreotide.¹,³ If an underlying cause of the POTS symptoms can be identified, treating the underlying cause is very important as well.
**Prognosis**

Currently, there is no cure for POTS. Detailed long term follow up studies on the course of POTS are sparse and inconsistent. One estimate is that about 50% of patients who have post-viral POTS will fully or almost fully recover within a two to five year period.¹ A 10 year retrospective analysis of pediatric POTS patients seen at Mayo Clinic found that only 20% of patients fully recovered, while 60% still had POTS but saw an improvement in symptoms.² Researchers have noted that some patients will not improve and may actually worsen over time.¹ With an individualized combination of diet, exercise, lifestyle adaptations and medications, most patients will see an improvement in their symptoms and their quality-of-life.¹ If an underlying cause can be identified, and if that cause is treatable, the POTS symptoms may improve or subside.

**Sources**

24. Autonomic Nervous System Diagram, Copyright (c) 2012 Vinik.