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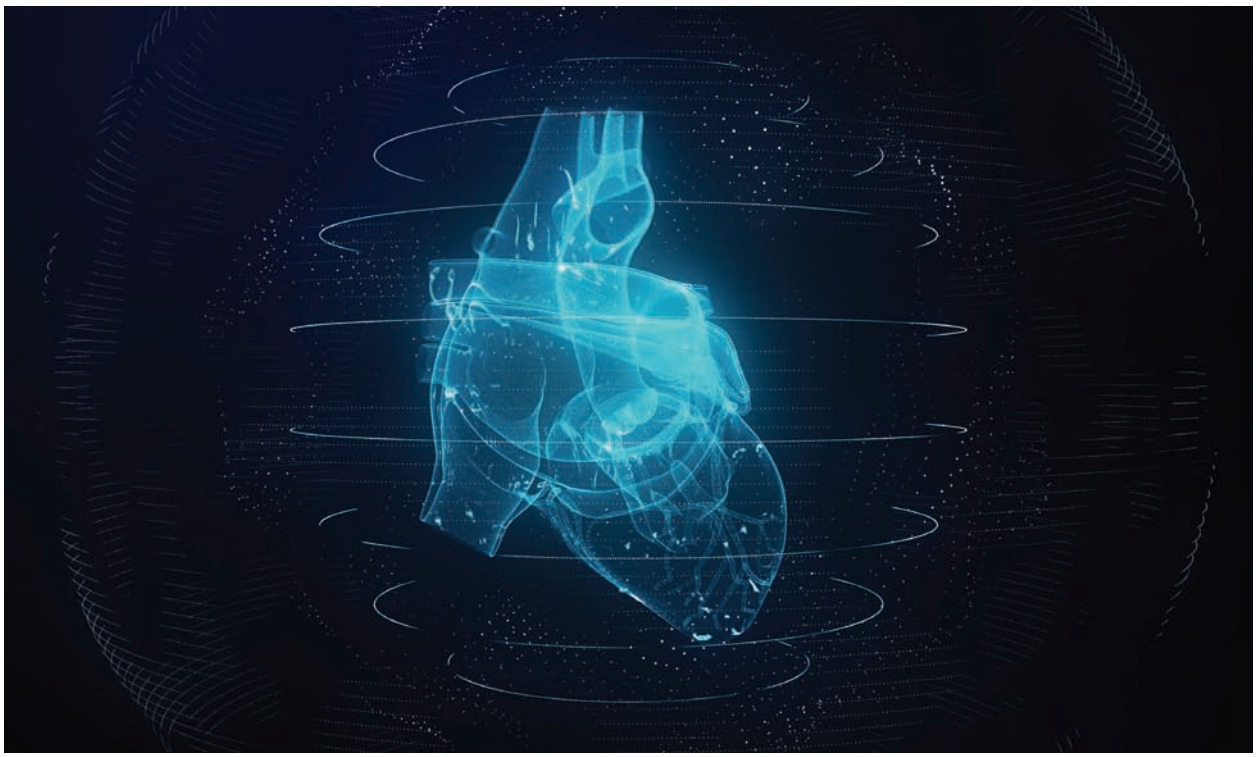


SHUTTERSTOCK/RELIGHT MOTION

Postural orthostatic tachycardia syndrome: Recognition and treatment

BY LAUREN CZOSEK, BSN, RN

Abstract: Postural orthostatic tachycardia syndrome (POTS) affects 3 million in the US and 11 million globally. Signs and symptoms can vary and greatly impact a patient's quality of life. This article focuses on the prevalence, clinical manifestations, diagnosis, treatment, and patient education surrounding POTS.

Keywords: orthostatic intolerance, postural orthostatic tachycardia syndrome, postural tachycardia syndrome, POTS, tilt-table testing

Postural orthostatic tachycardia syndrome (POTS), also known as postural tachycardia syndrome, is characterized by orthostatic intolerance. An abnormal autonomic response causes a specific group of signs and symptoms that only occurs while standing upright from supine positioning and is not associated with hypotension.[1] The patient's heart rate increases by at least 30 beats/minute or greater than 120 beats/minute while assuming an upright position.[2]

Pathophysiology

The exact pathophysiology of POTS remains relatively unknown. However, many theories include hypovolemia, autonomic dysfunction, a hyperadrenergic state, adrenergic receptor deficiency, mast cell activation, renin-aldosterone

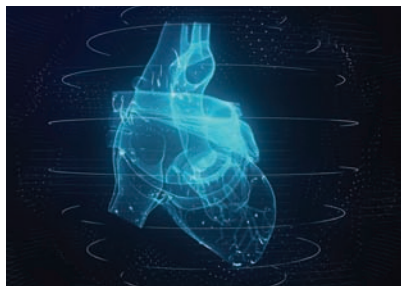
paradox, and physical deconditioning.[3] Some common causes of POTS include hypermobile Ehlers-Danlos syndrome; cancer; diabetes; amyloidosis, sarcoidosis; bacterial and viral infections; and autoimmune disorders, such as systemic lupus erythematosus.[4]

In one retrospective chart review including 20 patients diagnosed with POTS following a SARS-CoV-2 infection, 85% still had signs and symptoms of POTS 6 to 8 months after the initial infection. However, many of these patients improved with treatment.[5]

Subtypes of POTS include hypovolemic without hypotension, neuropathic, and hyperadrenergic. Hypovolemic POTS is due to a decrease in venous return to the heart that causes an increase in heart rate without the decrease in BP seen in orthostatic hypotension. Neuropathic POTS is due to patchy denervation of the sympathetic nerve fibers to the blood vessels in the extremities.[1] Hyperadrenergic POTS is characterized by a rise in systolic BP above 10 mm Hg within 10 minutes of standing or tilting upwards via a tilt-table test and an upright position plasma norepinephrine level of greater than or equal to 600 picograms (pg)/mL by blood specimens obtained during tilt-table testing.[1,2]

Epidemiology

Most patients diagnosed with POTS are female, with a 5:1 female-to-male diagnosis ratio.[3,6] About 170 out of every 100,000 people ages between 15 and 50 are diagnosed with POTS.[6] The median age of onset is 30 years old. In approximately 45% of patients, POTS symptoms began after 19 years.[6] POTS affects about 3 million persons in the US and 11 million globally, and is not associated with any particular race or ethnicity.[6] One study demonstrated a possible correlation between lower body mass index and POTS in patients with reduced peripheral blood flow and increased angiotensin II levels. Increased angiotensin II levels within the central nervous system can cause sympathoexcitation, a factor in hyperadrenergic POTS.[2]



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Signs and symptoms

Signs and symptoms of POTS can range from mild to severe, including fatigue, dizziness, palpitations, exercise intolerance, venous pooling, changes in cognitive function, headache, nausea, blurred vision, and tremors.[7] Other signs and symptoms include temperature dysregulation, sleep disturbances, chest pain, visual changes, diarrhea, migraines, constipation, or excessive sweating.[8] Signs and symptoms may impact patients' ability to work, interpersonal relationships, and activities of daily living.

Diagnosis

Begin by obtaining a thorough health history and basic labwork, including serum electrolytes, a complete blood cell count, and thyroid function tests. A medication history is important to identify other potential causes of tachycardia.[14] Medications that may increase heart rate may include sympathomimetics, serotonin-norepinephrine reuptake inhibitors, tricyclic antidepressants, atomoxetine, and anticholinergics. The clinician should also inquire about daily habits such as caffeine intake, weight loss, and decongestant use.[14]

After testing to rule out other possible causes of tachycardia, a tilt-table test is the gold standard for diagnosing POTS. The patient is secured, lying supine on a table. Then, the table is gradually tilted until the patient is upright. Vital signs are obtained throughout the process at specific intervals, and patient signs and symptoms are recorded throughout the test. A diagnosis is confirmed if the patient's heart rate increases by at least 30 beats/minute or is 120 beats/minute or more, without orthostatic hypotension. Blood specimens are obtained during the test to assess norepinephrine and epinephrine levels.[2]

If tilt-table testing is unavailable or the diagnosis is uncertain at an in-office appointment with nonspecific signs and symptoms, a screening 3-minute sit-to-stand or 10-minute lie-to-stand test should be considered. This test begins with the patient lying down for approximately 10 minutes, followed by standing up straight for 10 minutes. The patient's heart rate is checked periodically from the 10 minutes of standing and during the transition from lying to standing.[8]

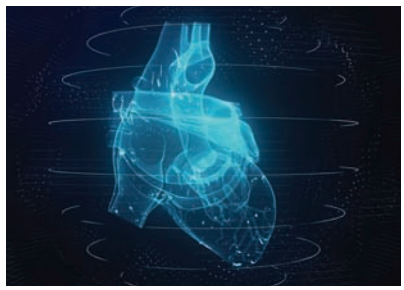
Patients often experience signs and symptoms years before diagnosis. In one study, most patients enrolled had, on average, 20 or more visits to their general practitioner and 4 to 5 visits to a specialist before being diagnosed with POTS.[8] The median time from initial presentation

to diagnosis may be 2 years, with about 15% of patients not receiving a diagnosis for over 10 years.[2] Time to diagnosis can take nearly 8 to 10 years in some cases.[6] In a large study including 500 patients, the average diagnostic delay from symptom onset was 5 years and 11 months, 15% of patients were diagnosed within their first year of symptoms, and 50% of patients traveled more than 100 miles from their home to receive medical care related to POTS.[9]

After the initial diagnosis, patients often require further testing, such as a cutaneous nerve biopsy, echocardiogram, or quantitative sudomotor axon reflex test (QSART). QSART involves a mild electrical stimulation on the skin called iontophoresis, which allows acetylcholine to stimulate sweat glands. QSART measures the volume of sweat produced by this stimulation to assess autonomic function.

Additional testing is completed based on the suspected POTS subtype and the professional recommendations of the patient's physician or advanced practice clinician (APC). For example, QSART may be performed if neuropathic abnormalities are suspected.[7]

Other studies to diagnose POTS may include blood volume analysis and hemodynamic studies.[7] Hemodynamic studies and monitoring to diagnose POTS include the injection of a radioactive isotope or tracer through an I.V. line during a scan to observe the pumping efficacy of the heart.[7] An ECG or ambulatory ECG monitoring and 24-hour urinary catecholamines should be completed.[2] Ambulatory ECG monitoring is completed to determine the extent of tachycardia over an extended period and if any underlying dysrhythmias are present.[2] Catecholamines should be tested to determine if the patient has reduced norepinephrine clearance and the POTS subtype. Twenty-four-hour urinary sodium excretion is completed if hypovolemia is suspected. Values less than 100 mEq/24 hours suggest hypovolemia.[14]



After testing and diagnosis, treatment can begin by focusing on symptom management and treating the specific POTS subtype.

Management

After testing and diagnosis, treatment can begin by focusing on symptom management and treating the specific POTS subtype. Management should involve a combination of nonpharmacologic and pharmacologic modalities. The following measures may improve the patient's quality of life; however, treatment results may vary significantly due to the unknown pathophysiology of POTS.[3]

Nonpharmacologic treatment options include exercise conditioning; compression garments; consuming 2 to 3 L of water or oral rehydrating solution per day; oral sodium increase with a goal of 8 to 12 g/day in those without hypertension; and avoiding potential triggers, such as extreme heat, caffeine, or alcohol.[10] Available sources of sodium include table salt, sports tablets, sports beverages, oral rehydration salts, and some soups.[14] Long-term effects on BP and increased sodium intake are unknown in those with POTS.[14]

Pharmacologic therapies used to treat patients with POTS should be individualized as the FDA has not approved any medications for POTS and some available medications may cause adverse reactions. Medications may include those that increase intravascular volume or lessen signs and symptoms. Beta-adrenergic receptor antagonists can blunt elevations in heart rate and are therefore used frequently in patients with POTS.[14] Low-dose propranolol is useful in symptom management and decreasing tachycardia; however, these medications may worsen fatigue.[2]

Patients with hypovolemic POTS may benefit from fludrocortisone, a synthetic mineralocorticoid aldosterone analogue that increases sodium retention and plasma volume.[2] Fludrocortisone should be avoided in patients with migraines, as it may exacerbate the disorder.[2] Midodrine, an alpha adrenoreceptor agonist, causes systemic vasoconstriction leading to an increase in venous return. Alpha$_2$-adrenergic agonists, such as clonidine, may be beneficial in hyperadrenergic POTS, in which hypertension is predominant.[2]

Medications to avoid include amphetamines and selective serotonin or norepinephrine reuptake inhibitors that may worsen tachycardia. Medications that may worsen orthostatic intolerance include calcium channel blockers, nitrates, opiates, and tricyclic antidepressants.[10] Treatment choices should be centered around the patient's presenting signs and symptoms.[10,11]

Emergency management

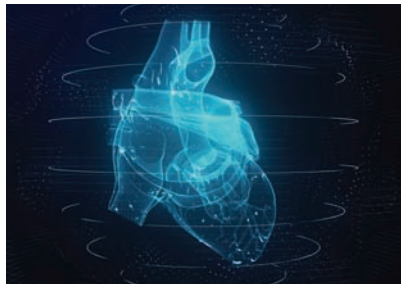
Patients with POTS being managed as outpatients and without any acute exacerbation of signs

and symptoms can generally avoid the ED. However, an acute flare may warrant a visit to the ED if it includes shortness of breath after 15 minutes of rest, a heart rate higher than one's baseline that does not resolve with lying down, or any episode of chest discomfort, especially if associated with shortness of breath, diaphoresis, or radiation to the neck, jaw, or arm.

Upon arrival to the ED, immediately assess patients with POTS for any syncopal episodes, especially if they are taking anticoagulants. Patients taking anticoagulants are at increased risk of adverse events due to syncope, such as hemorrhage. Some patients with POTS may arrive with acute dehydration and require I.V. hydration with 1 to 2 L of 0.9% sodium chloride solution administered over 1 to 2 hours.[2] Regular use of I.V. 0.9% sodium chloride solution is typically not utilized unless a patient cannot tolerate fluids by mouth or is dehydrated.[2]

Impact on quality of life

Research on the symptom severity of patients with POTS and its impact on daily living demonstrates a reduced quality of life compared with healthy individuals.[12] Inconclusive and inconsistent evidence exists among patients with POTS regarding prognosis. There is no decreased life expectancy for those diagnosed with POTS, but periods of acute exacerbations can negatively impact day-to-day life. POTS signs and symptoms are worsened by extreme temperature changes or stress.[10] Therapy is typically lifelong and requires a combination of nonpharmacologic interventions, lifestyle management, medications, and follow-up appointments.



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Nursing implications

Early recognition of signs and symptoms is key to diagnosis, treatment, and improved quality of life.[13] Patient education should include signs and symptoms of POTS, its causes, and what to do if a patient experiences signs and symptoms. For example, teach patients to sit or lie down immediately if they feel faint, dizzy, or lightheaded.

Educate patients on diet to avoid acute exacerbations and decrease signs and symptoms. As previously mentioned, patients may increase daily sodium intake to 8 to 12 g per day if hypertension is not present, drink 2 to 3 L of water or oral rehydrating solution per day, or eat smaller and more frequent meals since they are generally better tolerated.[7] Other dietary suggestions include avoiding overly processed foods, which typically contain less nutritional value, and eating a diet high in fiber and complex carbohydrates.[7] Teach patients about prescribed medications including action, dosage, and adverse reactions. Follow-up appointments should be patient-specific and can range from weekly to annually, depending on the patient's clinical status. Patients should also be assessed for increasing signs and symptoms during illness or stress. Advise patients to notify their physician or APC for any worsening signs or symptoms.

Educational resources and online support communities are available for patients with POTS (see *Patient resources*). Nurses can share these resources during patient appointments. These resources may decrease patients' feelings of isolation by connecting them with persons with similar health issues.[9] This is especially true for patients who cannot travel or meet with others in person.

Patient resources

Cleveland Clinic: https://my.clevelandclinic.org/health/diseases/16560-postural-orthostatic-tachycardia-syndrome-pots#living-with

Dysautonomia International: Finding a support group: www.dysautonomiainternational.org/page.php?ID=24
Education materials: www.dysautonomiainternational.org/page.php?ID=95
POTS Summary: www.dysautonomiainternational.org/pdf/SummaryOfPosturalOrthostaticTachycardiaSyndrome.pdf
10 Facts about POTS: www.dysautonomiainternational.org/pdf/10FactsAboutPOTS.pdf

Information for Shared Medical Appointments for those with POTS: https://my.clevelandclinic.org/departments/neurological/outcomes/882-postural-orthostatic-tachycardia-syndrome-pots

Conclusion

Nurses are critical to the early recognition and treatment of patients with POTS signs and symptoms. If clinicians recognize POTS in more patients who are currently being misdiagnosed or have unanswered questions regarding their symptoms, thousands or even millions of patients could have decreased symptom burden and time to proper treatment. ■

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Lauren Czosek is a family NP graduate of SUNY Brockport and an RN at the Gates Vascular Institute.

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