Strictly Clinical

Postural orthostatic tachycardia syndrome

By Marna M. Revlock, MSN, RN

Get your patients back on their feet with thorough assessments that lead to early and accurate diagnoses.

POSTURAL orthostatic tachycardia syndrome (POTS) affects over 1,000,000 Americans (80% are women of childbearing age). Since POTS was first characterized in 1993, diagnostic criteria and treatments have been established based new insights from research. However, the syndrome's diverse symptoms and practitioners' unfamiliarity with the illness leads to misdiagnosis and delays in appropriate treatment.

POTS, a dysfunction of the autonomic nervous system (ANS), is characterized by a group of disorders that are exacerbated upon standing (orthostatic intolerance). (See *POTS defined.*) Symptoms range from vague (lightheadedness, brain fog, and fatigue) to severe (complete autonomic failure and debilitation). A patient's quality of life varies depending on the degree of impairment. About 25% of all patients diagnosed with POTS (both primary and secondary) progress to complete disability, and many experience decreased quality of life as symptoms worsen.

Without an accurate diagnosis, nurses can't provide the education necessary to help patients cope with this syndrome. As POTS awareness grows, nurses will play a pivotal role in early diagnosis through accurate assessment, collaboration with providers and staff, and patient education.

ANS and POTS symptoms

The ANS is a network of neurofibers between the brain and spinal cord that extends to the organs and peripheral nerves. It's responsible for regulating automatic body functions, such as breathing, heartbeat, blood pressure, digestion, sexual function, and temperature control. Normally, when a person changes position from lying to standing, the body maintains blood pressure by constricting the blood vessels in the lower extremities and increasing the heart rate. With POTS, vasoconstriction and fluid regulation is abnormal, causing blood to pool in the lower extremities and reducing blood flow to crit-



A 2015 position statement from the Heart Rhythm Society says that postural orthostatic tachycardia syndrome (POTS) is a clinical syndrome with the following three characteristics:

- frequent symptoms that occur with standing such as lightheadedness, palpitations, tremulousness, generalized weakness, blurred vision, exercise intolerance, and fatigue
- an increase in heart rate of ≥30 beats per minute (bpm) when moving from a recumbent to a standing position held for more than 30 seconds (or ≥40 bpm in individuals 12 to 19 years old)
- absence of orthostatic hypotension (>20 mm Hg drop in systolic blood pressure).

ical organs. As a result, the patient experiences symptoms of tachycardia—heart palpitations, chest discomfort, weakness in the lower extremities, and fatigue. Patients may feel lightheaded or lose consciousness, but symptoms are typically relieved when the patient lies down.

Dysautonomia (ANS malfunction), extends beyond the cardiovascular system, creating other less common symptoms such as tremors, brain fog, headache, difficulty sleeping, shortness of breath, and blurred vision. Patients also may report problems regulating body temperature, bowel and bladder irregularities, poor sleep quality, and an inability to tolerate exercise. In fact, the severity and disability have been compared to a patient with heart failure or chronic obstructive pulmonary disease. Many individuals with POTS become wheelchair bound or bedbound, leaving them unable to care for their families, attend school, or work. Additionally, problems such as decreased cognitive function and memory further reduce functional abilities.

Forms of POTS

POTS is a heterogenous disorder with no single cause for the variety of symptoms that present, making identification particularly difficult; symptom manifestation and severity vary based on the etiology. POTS is categorized as primary or secondary.

Primary POTS

Primary POTS includes neuropathic, hyperadrenergic, and developmental forms. *Neuropathic* POTS is the most common, and some statistics indicate that up to 50% of patients with POTS have experienced a recent traumatic event such as an injury, viral illness, or pregnancy prior to onset. *Hyperadrenergic* POTS has an autoimmune connection or a genetic predisposition, which occurs in about 25% of those affected. *Developmental* POTS is seen during periods of rapid growth, such as adolescence.

Overall, primary forms of POTS are associated with disruptions in peripheral neurological connections, irregular sympathetic activity, and impaired volume control. Typically, those who are diagnosed younger have a better prognosis, and the symptoms may resolve with or without treatment. For others, the symptoms may become progressively worse.

Secondary POTS

Secondary POTS occurs as a manifestation of another illness such as Lyme disease, diabetes, or cancer, and management is aimed at treating the underlying illness. Regardless of the cause of illness, proper identification is key to determining the course of treatment because most symptoms can be effectively managed with medication, diet, and lifestyle management.

Assessment and diagnosis

Many patients wait years for an accurate diagnosis. Unless a provider is familiar with POTS, attention may fo-

Possible POTS symptoms

Postural orthostatic tachycardia syndrome (POTS) presents with diverse symptoms, which can lead to misdiagnosis. Thorough assessment can aid in early diagnosis and appropriate treatment.



cus on other diagnoses, preventing accurate testing and treatment. In fact, many patients are misdiagnosed with a psychiatric disorder or are identified as having anxiety, panic attacks, or chronic fatigue. A thorough assessment, including a family history, can help avoid misdiagnosis. It should include:

- A review of all systems. Patients won't recognize that other less prominent symptoms may be connected to neurologic dysfunction, so they may not mention them. (See *Possible POTS symptoms.*)
- Neurologic and cardiovascular assessment. Assess for palpitations, tachycardia, chest discomfort, pallor, syncope, shortness of breath, brain fog, or problems with memory. In peripheral extremities, assess bilaterally for pulse strength, sensation, discoloration, temperature, excessive or decreased sweating, edema or heaviness to the legs, weakness, and tremor. To best assess whether the patient has sustained, rather than transient, orthostatic tachycardia, take the first readings after he or she has been supine for at

Diagnosis

Diagnostic testing

Diagnostic testing for postural orthostatic tachycardia syndrome includes:

- Blood tests. Complete blood count, basic metabolic panel, thyroid function tests, liver function tests, and standing/lying down catecholamine levels are completed to rule out other causes of the patient's symptoms.
- Tilt table test. This test monitors changes in heart rate and blood pressure during positional changes. Diagnostic criteria during a tilt table test is a sustained heart rate > 120 beats per minute (bpm) or an increase > 30 bpm within 10 minutes. Blood pressure variables may or may not change, and those that fluctuate may be higher or lower than normal. Symptoms also may include lightheadedness, fainting, or overactive autonomic activity during the test.
- Heart monitoring. Electrocardiography and 24-hour blood pressure and heart rate monitoring help rule out arrhythmias.
- **24-hour urine collection.** The urine is tested for catecholamines and urine sodium levels.
- Autonomic function testing. This typically includes assessing cardiovagal and sudomotor function. Cardiovagal function testing assesses heart rate response to vagal stimulus. Sudomotor function testing assesses sweat response to acetylcholine and measures overactive autonomic activity.

least 5 minutes, and again after 1, 3, 5, and 10 minutes of standing.

• GI and urinary assessment. Assess for constipation, diarrhea, and bloating or cramps, as well as urinating difficulty or frequency.

Diagnostic testing includes blood tests, tilt table test, heart monitoring, 24-hour urine collection, and autonomic function testing. (See *Diagnostic testing*.)

Treatment

Individualized treatment is aimed at symptom control and improving vascular function with nonpharmacologic and pharmacologic strategies.

Nonpharmacologic interventions

Initial nonpharmacologic management includes improv-

ing volume expansion, preventing blood pooling, increasing endurance and orthostatic tolerance, avoiding triggers, and addressing psychological needs.

Improving volume expansion. Many patients with POTS have low urinary sodium levels indicative of low blood volume. Patients can expand volume by increasing their salt and fluid intake and elevating the head of the bed off the ground 4 to 6 inches.

Preventing blood pooling. Garments that, ideally, compress the calf, thigh, and lower abdomen are used to prevent blood from pooling in the lower extremities; leg compression alone tends to be less effective.

Increasing endurance and orthostatic tolerance. Strength training and aerobic exercise increase endurance and is recommended three times per week with gradual intensity. Because patients with POTS experience exercise intolerance, they'll need adequate rest periods before and after exercise. Initially, horizontal types of exercise, such as swimming, rowing, and riding a recumbent bike, are used to avoid POTS symptoms. As patients progress, upright activities can be added.

Physical countermeasure maneuvers can help improve the patient's orthostatic tolerance by raising arterial pressure. These maneuvers include squeezing a rubber ball, squatting, walking, or combined leg crossing and muscle tensing.

Avoiding triggers. Advise patients with POTS to avoid activities—such as prolonged standing, heat exposure, and consuming alcohol, caffeine, and large meals—that may trigger symptoms. And emphasize that if they experience symptoms, they should lie down or rest to prevent loss of consciousness or injury.

Addressing psychological needs. Although studies indicate that those with POTS typically have lower lifetime anxiety levels, many patients with POTS become hypervigilant in the presence of symptoms and experience increased stress and anxiety that affects their quality of life.

Direct interventions to both physical and psychological domains. Teach stress-reduction techniques and cognitive behavioral interventions to help patients manage symptoms and reduce fears about ongoing problems. Caregivers and family members also require education about how to support and care for their loved one. Direct patients and caregivers to agencies such as Dysautonomia International (dysautonomiainternational.org) or the Dysautonomia Foundation (familialdysautonomia.org), where they can find additional support, information about symptom management, and provider directories.

Pharmacologic management

Medications may be prescribed for severe symptoms or

if nonpharmacologic treatments aren't effective. Mineralocorticoids, antidiuretics, and hematopoietic agents help to increase blood volume; vasoconstrictors increase blood pressure; and beta-blockers decrease heart rate. Drugs such as selective serotonin reuptake inhibitors, selective norepinephrine reuptake inhibitors, clonidine, and pyridostigmine may be prescribed to facilitate cardiovascular function and synapse transmission within the neurologic system.

Early identification is key

Although POTS comes in many forms, researchers are learning more about the causes and treatments of this debilitating illness. You can help disseminate POTS awareness by educating your peers and patients. Early identification is key to treatment success because it will reduce the amount of time a patient spends seeking answers. Quicker diagnosis and treatment will increase patients' ability to get back on their feet and return to an optimal quality of life.

Marna M. Revlock is an assistant professor at Aultman College in Canton, Ohio.

Selected references

Anderson JW, Lambert EA, Sari CI, et al. Cognitive function, healthrelated quality of life, and symptoms of depression and anxiety sensitivity are impaired in patients with the postural orthostatic tachycardia syndrome (POTS). Front Physiol. 2014;5:230.

Arnold AC, Ng J, Raj SR. Postural tachycardia syndrome—Diagnosis, physiology, and prognosis [published online ahead of print, 2018]. *Auton Neurosci.* sciencedirect.com/science/article/pii/S1566070217 303545

Benarroch EE. Postural tachycardia syndrome: A heterogeneous and multifactorial disorder. *Mayo Clin Proc.* 2012;87(12):1214-25.

Busmer L. Diagnosis and management of postural tachycardia syndrome. *Nurs Stand.* 2013;27(20):44-8.

Fu Q, Levine BD. Exercise and non-pharmacological treatment of POTS [published online ahead of print, 2018]. *Auton Neurosci.* sciencedirect.com/science/article/pii/S1566070218300663

Kavi L, Gammage MD, Grubb BP, Karabin BL. Postural tachycardia syndrome: Multiple symptoms, but easily missed. *Br J Gen Pract.* 2012;62(599):286-7.

Lambert E, Lambert GW. Sympathetic dysfunction in vasovagal syncope and the postural orthostatic tachycardia syndrome. *Front Physiol.* 2014;5:280.

Miller AJ, Raj SR. Pharmacotherapy for postural tachycardia syndrome [published online ahead of print, 2018]. *Auton Neurosci*. sciencedirect.com/science/article/pii/S1566070218300250

Genetic and Rare Diseases (GARD) Information Center. Postural orthostatic tachycardia syndrome. Updated 2017. rarediseases.info .nih.gov/diseases/9597/postural-orthostatic-tachycardia-syndrome

Sheldon RS, Grubb BP, Olshansky B, et al. 2015 Heart Rhythm Society expert consensus statement on the diagnosis and treatment of postural tachycardia syndrome, inappropriate sinus tachycardia, and vasovagal syncope. *Heart Rhythm.* 2015;12(6):e41-63.



Online Adult/Adolescent or Pediatric/Adolescent Sexual Assault Nurse Examiner Training Program

40 Contact Hours

Attend Anytime, Anywhere

Simulation Training Resources Available After Online Course



Healthcare Designed For Every Sexual Assault Patient

Would you turn away a patient with chest pain? Of course not. So why turn away sexual assault patients? They require emergency care, too.

IAFN can provide live or online SANE training, or training for ED Staff, equipping you to provide the gold standard of care for sexual assault victims.

Online Adult/Adolescent or Pediatric/Adolescent Sexual Assault Nurse Examiner Training is now available through the International Association of Forensic Nurses.

Find out more about SANE Training today. www.forensicnurses.org/SANE365

The International Association of Forensic Nurses is accredited as a provider of continuing nursing education by the American Nurses Credentialing Center's Commission on Accreditation.