Postural orthostatic tachycardia syndrome (POTS, also known as postural tachycardia syndrome) is a condition in which a change from the supine position to an upright position causes an abnormally large increase in heart rate, called tachycardia. Other symptoms of an orthostatic nature — occurring in response to upright posture — may accompany the tachycardia.

The causes of POTS are poorly understood, and it is likely that several distinct underlying problems can lead to the symptoms of POTS. A variety of treatments, including exercise and medications, can improve symptoms for the majority of people with POTS. Many experience "a reasonable recovery with recovery defined as the relative absence of orthostatic symptoms alone with the ability to perform the activities of daily living with minimal restriction", while in others the condition persists indefinitely. It has been estimated that there are between 500,000 and 3,000,000 people with POTS in the United States alone.

Signs and symptoms

The hallmark symptom of POTS is an increase in heart rate by at least 30 beats per minute within 10 minutes of assuming an upright position. For people aged between 12 and 19, the minimum increase for diagnosis is 40 beats per minute. This symptom is known as orthostatic (upright) tachycardia (fast heart rate). It occurs without any coinciding drop in blood pressure, as that would indicate orthostatic hypotension. It is accompanied by other features of orthostatic intolerance — symptoms which develop in an upright position and are relieved by reclining. These orthostatic symptoms include palpitations, light-headedness, chest discomfort, shortness of breath, weakness and blurred vision.

In up to one third of people with POTS, fainting occurs in response to postural changes or exercise. Migraine-like headaches are common, sometimes with symptoms worsening in an upright position (orthostatic headache). Some people with POTS develop blotchy, sometimes purplish skin when standing, especially over the feet (indicative of blood pooling). 48% of people with POTS report chronic fatigue and 32% report sleep disturbances. Others exhibit only the cardinal symptom of orthostatic tachycardia.

The hypermobility type of Ehlers–Danlos syndrome (EDS) sometimes co-occurs with POTS. This type of EDS is characterized by loose, clicking joints prone to subluxations and dislocations, skin that exhibits moderate or greater laxity, meeting the Beighton criteria for joint hypermobility, and other miscellaneous signs of laxity including the ability to perform a reverse Namaskar sign and the ability to touch the tip of the nose with the tongue. POTS is also often accompanied by vasovagal syncope, with a 25% overlap being reported.[6] There is significant overlap between POTS and chronic fatigue syndrome, with evidence of POTS in 25-50% of CFS cases. Fatigue and reduced exercise tolerance are prominent symptoms of both conditions, and dysautonomia may underlie both conditions.

Causes

The symptoms of POTS can be caused by several distinct pathophysiological mechanisms. These mechanisms are poorly understood, and can overlap, with many people showing features of multiple POTS types. Many people with POTS exhibit low blood volume (hypovolemia), which
can decrease the rate of blood flow to the heart. To compensate for this, the heart increases its cardiac output by beating faster, leading to the symptoms of presyncope and reflex tachycardia. In the 30 to 60% of cases classified as hyperadrenergic POTS, norepinephrine levels are elevated on standing, often due to hypovolemia or partial autonomic neuropathy. A smaller minority of people with POTS have (typically very high) standing norepinephrine levels that are elevated even in the absence of hypovolemia and autonomic neuropathy; this is classified as central hyperadrenergic POTS. The high norepinephrine levels contribute to symptoms of tachycardia. Another subtype, neuropathic POTS, is associated with denervation of sympathetic nerves in the lower limbs. In this subtype, it is thought that impaired constriction of the blood vessels causes blood to pool in the veins of the lower limbs. Heart rate increases to compensate for this blood pooling.

Genetics likely plays a role, with one study finding that 1 in 8 POTS patients reported a history of orthostatic intolerance in their family. In up to 50% of cases, POTS is associated with recent viral illness. It may also be associated with physical deconditioning or chronic fatigue syndrome. During viral illness or prolonged bed rest, the body may become conditioned to orthostatic intolerance and excitability of the central nervous system, resulting in a failure to re-adapt to the normal demands of standing or exercise.

If POTS is caused by another condition, it may be classified as secondary POTS. Chronic diabetes mellitus is one frequently seen primary cause. POTS can also be secondary to gastrointestinal disorders that are associated with low fluid intake due to nausea or fluid loss through diarrhea, leading to hypovolemia.

There are a subset of patients that present with both POTS and Mast cell activation syndrome (MCAS), and it is not yet clear whether MCAS is a secondary cause of POTS or simply comorbid, however treating MCAS for these patients can significantly improve POTS symptoms.

**Diagnosis**

People with POTS will show a marked rise in heart rate within 10 minutes of standing or being tilted 60° head-up on a tilt table, without a corresponding decrease in blood pressure. A variety of autonomic tests are employed to exclude autonomic disorders that could underlie symptoms, while endocrine testing is used to exclude hyperthyroidism and rarer endocrine conditions. Electrocardiography is normally performed on all patients to exclude other possible causes of tachycardia. In cases where a particular associated condition or complicating factor are suspected, other non-autonomic tests may be used: echocardiography to exclude mitral valve prolapse, neuroimaging for suspected Chiari malformation, and thermal threshold tests for small-fiber neuropathy.

Testing the cardiovascular response to prolonged head-up tilting, exercise, eating, and heat stress may help determine the best strategy for managing symptoms. POTS has also been divided into several types (see § Causes), which may benefit from distinct treatments. People with neuropathic POTS show a loss of sweating in the feet during sweat tests, as well as impaired norepinephrine release in the leg, but not arm. This is believed to reflect peripheral sympathetic denervation in the lower limbs. People with hyperadrenergic POTS show a marked
increase of blood pressure and norepinephrine levels when standing, and are more likely to suffer from prominent palpitations, anxiety, and tachycardia.

Treatment
Varied treatments are needed to address the different features of POTS. For most patients, water intake should be increased, especially after waking, in order to expand blood volume (reducing hypovolemia). 8-10 cups of water daily are recommended. Increasing salt intake, by adding salt to food, taking salt tablets, or drinking sports drinks and other electrolyte solutions is an effective way to raise blood pressure by helping the body retain water. Different physicians recommend different amounts of sodium to their patients. Salt intake is not appropriate for people with high blood pressure. Combining these techniques with gradual physical training enhances their effect. In some cases, when increasing oral fluids and salt intake is not enough, intravenous saline or the drug desmopressin is used to help increase fluid retention.

Large meals worsen symptoms for some people. These people may benefit from eating small meals frequently throughout the day instead. Alcohol and food high in carbohydrates can also exacerbate symptoms of orthostatic hypotension. Excessive consumption of caffeine beverages should be avoided, because they can promote urine production (leading to fluid loss) and consequently hypovolemia. Exposure to extreme heat may also aggravate symptoms. Prolonged physical inactivity can worsen the symptoms of POTS. Techniques that increase a person’s capacity for exercise, such as endurance training or graded exercise therapy, can relieve symptoms for some patients. Aerobic exercise performed for 20 minutes a day, three times a week, is sometimes recommended for patients who can tolerate it. Exercise may have the immediate effect of worsening tachycardia, especially after a meal or on a hot day. In these cases, it may be easier to exercise in a semi-reclined position, such as riding a recumbent bicycle, rowing or swimming.

When changing to an upright posture, finishing a meal or concluding exercise, a sustained hand grip can briefly raise the blood pressure, possibly reducing symptoms. Compression garments can also be of benefit by constricting blood pressures with external body pressure.

Pharmacological management

Propranolol can reduce heart rate and treat migraines.

If nonpharmacological methods are ineffective, medication may be necessary. As of 2013, no medication has been approved by the U.S. Food and Drug Administration to treat POTS, but a variety are used off-label. Their efficacy has not yet been examined in long-term randomized controlled trials. These medications often target low blood pressure, even at levels that would not warrant treatment in a person without POTS. Fludrocortisone, considered a first-line drug, works by reducing salt loss, which increases blood volume. If that fails, midodrine (or another
vasoconstrictor, but excluding any that increase the heart rate) may be prescribed. Midodrine works by narrowing the blood vessels, preventing blood pooling, and raising blood pressure. If symptoms and tachycardia persist, a clinician may choose to add a cardioselective beta blocker such as bisoprolol, which prevents dilation of the blood vessels and tachycardia. Fludrocortisone and midodrine should not be used by people with normal or high supine blood pressure. These people may instead be prescribed a beta blocker, such as bisoprolol or propranolol, or another drug that lowers heart rate. Propranolol can also be used to treat associated migraine headaches. Preliminary evidence suggests that propranolol is most effective at low doses (10–20 mg). Pyridostigmine has been reported to improve chronic symptoms in about half of patients.

Prognosis

POTS has a favorable prognosis when managed appropriately. Symptoms improve within five years of diagnosis for many patients, and 60% return to their original level of functioning. About 90% of people with POTS respond to a combination of pharmacological and physical treatments. Those who develop POTS in their early to mid teens during a period of rapid growth will most likely see complete symptom resolution in two to five years. Outcomes are more guarded for adults newly diagnosed with POTS. Some people do not recover, and a few even worsen with time.[3] The hyperadrenergic type of POTS typically requires continuous therapy. If POTS is caused by another condition, outcomes depend on the prognosis of the underlying disorder.