Quick guide to postural orthostatic tachycardia syndrome (POTS)

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What is postural orthostatic tachycardia syndrome?

Postural orthostatic tachycardia syndrome (POTS) is a condition characterised by orthostatic tachycardia with accompanying orthostatic symptoms. It is a common post-acute-infection and post-vaccination sequelae, but can be from other things too. POTS typically falls under the umbrella of dysautonomia, though it may also be due to other conditions, such as May-Thurner syndrome. Growing evidence suggests POTS is likely an autoimmune, inflammatory, and/or coagulopathic disease.

What are the symptoms of postural orthostatic tachycardia syndrome?

Symptoms are often systemic and all-consuming, with worsening upon an orthostatic challenge:

Common orthostatic symptoms:
Blood pressure dysregulation
Chest pain
Discolouration of legs
Dizziness
Dyspnoea
Excessive sweating
Fainting
Lower leg oedema
Palpitations
Tremoring

General:
Adrenaline surges
"Coat-hanger" pain
Exercise intolerance
Fatigue
Insomnia
Mast cell activation syndrome (MCAS)
Polydipsia
"Pretzel legs" with prolonged standing
Thermodysregulation

Sensory:
(Pulsatile) Tinnitus
Vision problems

Genitourinary:
Polyuria

Skin:
(Facial) Flushing

Neurological:
Cognitive dysfunction
Headaches
Myoclonus
Small fibre neuropathy
Weakness

Respiratory:
Hyperventilation syndrome

Endocrine:
Low aldosterone
Low arginine vasopressin (AVP)

Gut:
Constipation
Bloating
Diarrhoea
Nausea

How is postural orthostatic tachycardia syndrome diagnosed?

There are several validated ways to test for POTS. Typically, a tilt table test is done, but this can take time to organise, thus delaying treatment. An easier test is the active stand test, and requires a blood pressure cuff to conduct:

1. **Patient lies down for 10 minutes:** Take blood pressure and pulse rate at 5 and 10 minutes
2. **Patient stands up for 10 minutes:** Take blood pressure and pulse rate every minute. *It is important the patient remains relaxed in the standing position, trying to avoid subconscious muscle tension*

If the patient feels faint, the test should be stopped immediately

A diagnosis is made if heart rate increases and sustains by roughly ≥ 30 bpm for adults, or ≥ 40 bpm for children/adolescents (these are not hard cut-offs, and clinical judgement is required), with accompanying symptoms (usually those outlined above, but please be aware that symptoms vary widely).

Blood pressure should remain roughly stable in POTS. However, if blood pressure increases by ≥ 10 mmHg, the patient likely has hyperadrenergic POTS. If blood pressure drops and/or they (nearly) faint, they likely have POTS with orthostatic hypotension and/or vasovagal syncope.

What tests are there for postural orthostatic tachycardia syndrome?

Tests for orthostatic tachycardia include the tilt table test, NASA lean test, and the active stand test (described above). In addition to these, other tests can support the diagnosis and offer clues regarding the most appropriate treatment.

- **Norepinephrine:** ≥ 600 pg/mL on standing helps indicate hyperadrenergic POTS
- **24-hour urine sodium:** < 170 mmol/24-h can indicate salt as a potentially helpful treatment
- **Renin, aldosterone, AVP:** Low levels can indicate hypovolaemia

Other tests and diagnoses should also be considered to improve the clinical picture, rule-out other causes, and/or offer potential treatment options:

- ≥ 24-h ambulatory blood pressure monitoring
- Cardiac imaging
- Chest x-ray
- Electrocardiogram and/or ≥ 24-h Holter monitor
- Full blood count
- May-Thurner syndrome (or similar venous disease)
- MCAS
- Nutrition screen (particularly for anaemia, and B12/folate deficiency)
- Physical examination
- Signs of hypermobility
- Small fibre neuropathy

What treatments are there for postural orthostatic tachycardia syndrome?

There are many available treatments for POTS. Often multi-targeted treatment is most appropriate, depending on the clinical presentation, test results, co-pathologies, and patient preference, for example:

Target	Treatment	Example clinical observation
Lower heart rate	Ivabradine	
	Beta blockers[1] Clonidine Alpha-methyldopa	Hyperadrenergic POTS
Vasoconstriction	Midodrine	Orthostatic hypotension
Increase blood volume	Desmopressin[2]	Low AVP
	Fludrocortisone[2,3]	Low aldosterone
	Salt (can titrate up to 12 g/d)[2]	Urine sodium < 170 mmol/24-h
	I.V. saline infusion	
Neurological support	Pyridostigmine	Prominent symptoms related to the cholinergic system (e.g. dry eyes, xerostomia, bradykinesia)

[1] Beta blockers can be a trigger in patients with MCAS
[2] Electrolytes should be checked regularly
[3] Some recommend potassium and magnesium supplementation with fludrocortisone

In addition to the above, patients are often advised to try:

- Consuming 2-3 L/d fluid (ideally electrolyte drinks, being mindful that some contain sugar which can aggravate POTS, and may cause other problems)
- Wearing compression stockings
- Eating small (more frequent) meals, with low refined carbohydrates

Exercise is often recommended, but this should be based on the patient. For example, exercise is contraindicated in those with myalgic encephalomyelitis (ME), which is a common co-pathology. For other patients, exercise might be a more viable option once POTS is better controlled, rather than as a direct treatment.

Resources

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