**Referral Support Service**

**Cardiology**

**C**

**Postural Tachycardia Syndrome**

**Definition and Symptoms**

Disorder of the autonomic nervous system caused by standing up and reduced with sitting or lying down. Can be defined as:

* Sustained increase in heart rate of 30 beats per minute within 10 minutes of standing or upright tilt (during tilt table testing) or
* An increase in heart rate exceeding 120 bpm on prolonged standing.
* A heart rate increase of greater than 40 bpm is required for those aged 12-19.
* Findings should be associated with symptoms of orthostatic intolerance, such as dizziness, fatigue, sweating, nausea and palpitations.

**PoTS can be secondary to the following conditions:**

* Joint hypermobility syndrome / Ehlers Danlos Hypermobility type (most common)
* Chronic Fatigue syndrome/ME
* Lupus
* Fibromyalgia
* Diabetes
* Sarcoidosis
* Amyloidosis
* Alcoholism
* Chemotherapy (esp. with vinca alkaloids)
* Sjorgens syndrome
* Heavy metal poisoning
* Lyme disease

**Differential diagnosis**

Obtain a detailed history, examination and general medical evaluation, and evaluation of joint hyper mobility using either the Beighton Hypermobility score or the Brighton criteria.

Identify conditions that may produce orthostatic intolerance - dehydration, anaemia, Addison disease or any other endocrinopathy including pituitary disease.

Measure lying and standing heart rate and BP (lying for 3 mins, standing at 2, 5 and 10 minutes). Observe for venous pooling in both the hands and feet whilst standing, which appears as a purplish discolouration.

**Investigations**

The following tests should usually be undertaken:

* **ECG** - to rule out presence of an accessory pathway or any other abnormality of cardiac conduction
* **U&E’s, TFT’s, FBC**
* **Ferritin** – as often low
* **Vit B12 / folate** - rule out deficiency
* **Basic Endocrine tests** including Thyroid function testing
* **Short synacthen testing** with ATCH to assess the adrenal axis
* **Echocardiography** – to exclude structural or functional heart disease
* **24 hr Holter monitor** – In order to ascertain that the palpitation symptom of PoTS are mediated by sinus tachycardia and not an arrhythmia; to determine mean heart rate and heart rate fluctuation due to any activity, and correlate any symptoms to rate and rhythm

Some patients may need additional heart rate monitoring with event recorders.

* **CXR**
* **24hr urinary catecholamines and free metanephrines** - Used to exclude pheochromocytoma, this can be confused with PoTS especially in patients with Hyperadrenergic PoTS. Patients with pheochromocytoma are more likely to have symptoms when lying down.
* **24hr urinary sodium** - This will provide documentation that the patient is taking sufficient fluids and sodium. The goal is a volume of 1,500–2,500 mL and sodium excretion of 170 mmol/24 hours. The latter indicates that the patient is taking adequate sodium and probably has a normal plasma volume.
* **Head up tilt table test (HUTT) and Active Stand Test**

**Management**

No therapy is successful in all patients with PoTS, and large-scale prospective controlled trial data is unavailable.

* Initially efforts should be made to identify and treat any reversible causes.
* Withdraw if possible, any medications which may contributing to symptoms.
* If the patient has been immobile or bed-bound their symptoms may gradually improve with reconditioning to the upright posture.
* Optimise treatment for any chronic condition.
* If there is any evidence of re-entrant tachycardia this must be treated
* Radiofrequency of the SA node is not recommended.
* Educate patient about nature of disorder.
* Avoid aggravating factors.

**Non-pharmacological treatments**

* **Water** - at least 2-3 litres per day
* **Salt** at least 150-250 mEq daily
* **Compression stockings** - to deliver at least 30 mmHg of compression at the ankles
* **Sleeping with head of the bed elevated**
* **Countermanoevures**. Pumping calves before rising, using countermnanouvres if feeling lightheaded or dizzy
* **Rising slowly** from a lying down to sitting or standing position
* **Exercise** - both aerobic and resistance training should be encouraged and has shown to be beneficial. It is important that patients start slowly and build up exercise tolerance
* **Avoiding** – alcohol, recreational drugs
* **Keep cool**
* **Eat regular meals**

**Pharmacological treatments (Amber SI drugs)**

* **Fludrocortisone - (50-200 mcg once daily Max dose can be up to 400 mcg daily)**
* Used with patients with partial dysautonomic PoTS and patients in whom hypovoleamia is known or strongly suspected.
* Side effects: worsening headaches, depression, hypokalaemia, hypomagnesemia, acne and fluid retention. Numerous symptoms of sympathetic over activity can be enhanced.
* **Ivabradine - 2.5 – 5 mg twice daily**
* A sinus node blocker has reportedly helped some PoTS patients experience less symptoms. Ivabradine is sometimes used as an alternative to beta-blockers because it results in heart rate reduction without vasodilatation, sexual disturbances, or negative inotropic effects.
* Side effects: muscle cramps

**Referral Information**

* **Secondary care referral should only be made when there is:**
* diagnostic uncertainty,
* complexity due to comorbidities or
* difficulty with management
* **Information to include in referral letter**
* Please detail what treatments have been tried and their response
* Relevant past medical/surgical history
* Current regular medication

**Patient information leaflets/PDAs** *(these may not represent local commissioning guidance)- from the British Association of Dermatology*

* [Download PoTS Leaflets - PoTS UK](https://www.potsuk.org/about-pots/download-pots-leaflets/) – January 2025

**References**

* [Guidelines - PoTS UK](https://www.potsuk.org/pots-for-medics/guidelines/)
* [GP Guide: PoTS on a Page - PoTS UK](https://www.potsuk.org/pots-for-medics/gp-guide/)