

The postural tachycardia syndrome, or PoTS, was named and defined in 1993,<sup>1</sup> but is likely synonymous with earlier conditions such as 'The Soldier's Heart', a term coined by Sir James Mackenzie in 1916.<sup>2</sup> Affected patients experience an abnormal response to upright posture resulting in multiple symptoms.<sup>3</sup>

Autonomic dysregulation causes inadequate vasoconstriction when standing, resulting in blood pooling within the splanchnic vasculature and limbs, and consequent reduced venous return to the heart. An excessive compensatory tachycardia and increased plasma noradrenaline levels contribute to symptoms, the commonest of which are fatigue, palpitations, light-headedness, headache, and nausea.<sup>3,4</sup> See Table 1 for a list of symptoms. If not adequately treated, PoTS can become a debilitating disorder that can lead to significant disability and impaired quality of life.<sup>1</sup>

Forty-one per cent of patients with PoTS describe a viral infection preceding the onset of PoTS,<sup>5</sup> and therefore it was no surprise to clinicians with an interest in PoTS that it would also follow COVID-19 infection. The National Institute for Health and Care Excellence recommends testing for PoTS in patients with long COVID with symptoms of orthostatic intolerance.<sup>6</sup>

Parallels can be drawn between long COVID (with or without PoTS) and pre-pandemic PoTS. Both are complex, multisystem conditions with a preponderance in younger and middle-age women, although they can occur in any sex and age.<sup>4,7</sup> There is considerable crossover between symptoms of both conditions. Perhaps because symptoms are multiple and non-specific, both groups of patients commonly describe that their symptoms were not believed or recognised by healthcare professionals, and are often dismissed as anxiety.<sup>7-9</sup>

The precise pathophysiology of PoTS is not fully understood. It is not a unique disease entity. As with heart failure, it is a syndrome and there is evidence that several mechanisms are involved in PoTS including hypovolaemia, autonomic denervation, hyperadrenergic stimulation, and autoimmune pathology. More than one mechanism can occur in an individual.<sup>1</sup> It is not yet known whether the pathophysiologies of PoTS prior to the pandemic are the same as those found in patients with long COVID and PoTS; further research in this area is necessary. However, to date, the symptom profile of PoTS and response to treatments appear to mirror those of non-COVID PoTS.<sup>5</sup>

**Table 1. Common postural tachycardia syndrome symptoms**

Patients experiencing symptoms, %	Orthostatic intolerance symptoms <sup>a</sup>	Symptoms not necessarily associated with posture
>90	<ul style="list-style-type: none"> <li>• Light-headedness, presyncope</li> <li>• Palpitations (heart racing)</li> </ul>	<ul style="list-style-type: none"> <li>• Cognitive impairment (brain fog)</li> <li>• Frequent nausea</li> <li>• Headache</li> <li>• Fatigue</li> </ul>
>80	<ul style="list-style-type: none"> <li>• Breathlessness</li> </ul>	<ul style="list-style-type: none"> <li>• Abdominal pain</li> <li>• Muscle pain/weakness</li> <li>• Cold feet/hands</li> </ul>
>70	<ul style="list-style-type: none"> <li>• Atypical chest pain</li> <li>• Tremulousness</li> <li>• Blurred vision</li> </ul>	<ul style="list-style-type: none"> <li>• Bloating/constipation</li> <li>• Hand tingling</li> </ul>
Other common symptoms	<ul style="list-style-type: none"> <li>• Syncope (36%)</li> <li>• Sweating</li> </ul>	<ul style="list-style-type: none"> <li>• Diarrhoea (69%)</li> <li>• Poor sleep</li> <li>• Exercise intolerance</li> </ul>

<sup>a</sup>Symptoms that develop when upright and are relieved by reclining.

### ASSESSMENT

Following a detailed history and appropriate examination, if PoTS is suspected, GPs can make the diagnosis with an active stand test (also known as a NASA Lean Test). GPs should record the heart rate after resting the patient supine for a minimum of 5 mins, immediately on standing, and then at 2, 5, and 10 mins. Ask about symptoms and observe for acrocyanosis, a purple discolouration of dependent limbs that occurs in 50% of affected patients.<sup>1,2</sup> Practical tip: stand the patient against a wall, chair, or bed and observe carefully in case they faint. Diagnostic criteria are:

- a sustained increase in heart rate of  $\geq 30$  beats per minute ( $\geq 40$  in teenagers) from supine to an upright position;
- in association with symptoms of PoTS for at least 3 months; and
- in the absence of hypotension.

Although the definition of PoTS excludes hypotension, PoTS symptoms are very similar to those of postural hypotension. Patients with PoTS usually have a tendency to hypotension and it is common for vasovagal syncope to coexist.<sup>3</sup>

It may be necessary to request that patients return with a longer appointment to facilitate testing. Referral for tilt table testing may be appropriate if the diagnosis is unclear. Conditions with symptoms similar to PoTS need to be excluded; blood tests including electrolytes, renal function, FBC, ferritin, thyroid-stimulating hormone, and morning cortisol level, and an electrocardiogram are

recommended.<sup>1</sup> In the setting of long COVID, myocarditis is common and exhibits similar symptoms to PoTS; an echocardiogram or cardiac MRI may be required to exclude cardiac pathology.

Conditions associated with PoTS should be sought. These include hypermobile Ehlers-Danlos syndrome and spectrum disorder, autoimmune conditions, and mast cell activation syndrome.<sup>4</sup>

### MANAGEMENT

Pre-COVID-19, management within a multidisciplinary team was recommended for PoTS, and long COVID clinics can provide an ideal setting to facilitate this. Initial treatment for PoTS involves supported self-management (Table 2).

Exercise intolerance resulting in prolonged post-exercise fatigue is a common symptom of both PoTS and long COVID, and may hinder ability to engage in an exercise regimen.<sup>5</sup> The role of physical activity including exercise in severely exercise-intolerant patients remains unclear and should be undertaken with great caution. Physiotherapists and occupational therapists treating patients with long COVID and PoTS should be familiar with managing these conditions.

Like long COVID, PoTS severity can fluctuate unpredictably, making rehabilitation and return to work challenging.

Medication that was recommended pre-pandemic for PoTS is being prescribed in long COVID PoTS when self-management measures are insufficient to control symptoms.<sup>5</sup> Although unlicensed for use in PoTS, many of these drugs such as propranolol, ivabradine,

**Table 2. Supported self-management for postural tachycardia syndrome<sup>1,3</sup>**

Method	Rationale	Comment
Avoid symptom triggers	To reduce vasodilation and venous pooling	Triggers include hot environments, prolonged standing, heavy meals, and alcohol
High fluid intake	Increase blood volume	3 l/day in adults
Additional salt (unless contraindicated)	Increase blood volume and pressure	10 g/day in adults, less in children
Lower body compression	Reduce venous pooling	Waist high, class 2. If not tolerated, sports compression clothing may help
Exercise	To strengthen skeletal muscle pump and prevent deconditioning	Start with low-level recumbent exercise
Counterpressure manoeuvres	Activating skeletal muscle pump to increase venous return/prevent syncope	Cross legs and squeeze thigh muscles, clench buttocks, and tightly fold arms
Sleep in a head-up tilt position (>10°)	To expand blood volume	Bricks under head end of bed
Pacing of activities	To regulate activity to achieve adaptive goals	To reduce severity of flares and fatigue
Psychotherapy	To help patient adjust to chronic illness and frightening symptoms	Cognitive behavioural therapy and mindfulness

midodrine, fludrocortisone, pyridostigmine, clonidine, and methyl dopa are familiar to GPs through use in other conditions and may be prescribed provided that the General Medical Council's guidance for good practice in prescribing unlicensed medicines is followed. Where possible, withdraw drugs that can exacerbate symptoms such as those that lower blood pressure or induce tachycardia.<sup>5</sup>

### PREVALENCE OF POTS IN LONG COVID

The prevalence of PoTS in the post-COVID-19 community has not yet been quantified. With COVID-19 emerging as a new condition, studies to date have often been retrospective, involved low participant numbers or selected cohorts, and are inconsistent around timing after initial COVID-19 infection, definitions of dysautonomia or PoTS, and testing protocols.

Long COVID services, where they ask patients about symptoms of orthostatic intolerance, describe finding PoTS in 15% to over 50% of patients with postural symptoms (unpublished data). Not all patients post-COVID-19 infection with orthostatic intolerance have PoTS. A smaller number have orthostatic hypotension, and some have reproducible postural symptoms but normal stand or tilt table test.<sup>5</sup>

### IMPACT OF POTS AND LONG COVID

The negative consequences of both PoTS and long COVID on patients, their families, healthcare services, and the wider socioeconomic implications have been recognised.<sup>4,10,11</sup> Symptoms are multiple, non-specific, and debilitating. The value of

validating symptoms and their impact on patients should not be underestimated.

Having a high index of suspicion in patients with orthostatic intolerance and testing for PoTS in appropriate patients will open therapeutic opportunities not otherwise available to them and can prevent physical and psychological deterioration. NICE recommends that long COVID services should be led by a doctor,<sup>6</sup> with the skills to diagnose PoTS and exclude conditions with similar symptoms.

Patients who develop PoTS following COVID-19 infection will benefit from the supported self-management offered by GPs and multidisciplinary teams in long COVID clinics. Training resources on PoTS and long COVID can be found on the RCGP and PoTS UK websites. More complex patients will require referral to PoTS specialists (often found within syncope, cardiology/electrophysiology, or autonomic clinics). Specialist PoTS clinics are reporting increases in referral numbers.<sup>5</sup> The number of patients with PoTS far exceeds the capacity of existing clinics and this is placing a strain on their already under-resourced services.<sup>5,12</sup>

There is a growing concern that, in addition to long COVID, we may now also be facing an epidemic of PoTS.<sup>12</sup> There has never been a greater need to invest in NHS services to serve the unmet needs of this population. Perhaps we can learn from the rapid development of multidisciplinary teams in long COVID clinics and utilise these new skills and resources in the long term to also provide long overdue equitable access to therapeutic pathways for all patients with PoTS.

### EMAIL FOR CORRESPONDENCE

Lesley Kavi  
Email: [lesley@potsuk.org](mailto:lesley@potsuk.org)

**Lesley Kavi,**  
Retired GP; Chair and Trustee, PoTS UK; Visiting Professor, Birmingham City University, Birmingham.

### Provenance

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### Competing interests

The author has declared no competing interests.

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