

Case Report

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## Postural Tachycardia Syndrome

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### Abstract

We report a case diagnosed with Postural Tachycardia Syndrome (PoTS), 8 years after onset of symptoms. The patient, a 47-year old female, presented with recurrent episodes of light-headedness, presyncope, palpitations and tremulousness. There were no abnormalities on clinical examination. On two earlier occasions, the patient was evaluated by cardiologist, neurologist, pulmonologist and the ear nose and throat physician for her symptoms. She underwent several diagnostic tests, which showed no abnormalities. No diagnosis was established and the symptoms spontaneously disappeared twice. On the current occasion of recurrence of symptoms, the diagnosis of PoTS was highly suspected and proved by a positive tilt table test. She was successfully managed accordingly.

**Keywords:** Dizziness, Palpitation; Presyncope; Postural tachycardia syndrome; Tilt table test.

### Introduction

The postural tachycardia syndrome (PoTS) is an autonomic disorder, characterized by a sustained heart rate increment of  $\geq 30$  beats/min ( $\geq 40$  beats/min in individuals  $< 19$  years of age) within 10 min of standing or head-up tilt in the absence of orthostatic hypotension [1]. Although the specific pathophysiology is still unclear, PoTS is often classified into different subtypes, including hypovolemia, neuropathic, hyperadrenergic, autoimmune and/or mast cell activation disorder. These different subtypes are not separate causes of PoTS, but features that are often observed in patients with PoTS [2;3]. The true incidence is unknown. Usually the symptoms are non-specific and diagnosis is challenging. Symptoms may encounter neurologic manifestations of cerebral hypoperfusion and cardiac presentations.

The diagnosis is challenging and it may take years to be

established. We present a female patient who had non-specific symptoms for years, initially misdiagnosed, finally diagnosed with PoTS following positive Tilt Table Test.

### CASE REPORT

A 47-year-old female presented at the outpatient clinic of our hospital with history of recurrent episodes of light-headedness, presyncope, palpitations and tremulousness for 6 months. These symptoms occurred spontaneously while sitting, standing or walking but never when patient was lying down, accompanied with sweating, tunneled vision, shortness of breath and anxiety (Figure 1A). Spontaneously disappearance of symptoms occurred within one to three minutes. The patient was evaluated by cardiologist, neurologist, pulmonologist and the ear nose and throat physician during her previous episodes. The patient underwent diagnostic

tests including routine electrocardiogram (ECG), 48-hour ambulatory ECG recording, exercise tolerance testing, laboratory testing, chest X-ray, echocardiography, audiometric testing, electroencephalogram and a magnetic resonance imaging scan of the brain. However, despite all the tests a cause of her symptoms was not ascertained. Since spring 2015, the symptoms reemerged hampering the patient's daily activities both professionally and personally.

During presentation and on clinical examination she had body mass index (BMI) of 30.1 kg/m<sup>2</sup>, blood pressure of 143/90 mmHg and a heart rate of 102 beats per minute (bpm). Ambulatory ECG recording was performed for 48 hours, revealed a normal variation in heart rhythm, from 50 to 148 bpm. With the exercise tolerance testing, the patient showed physical deconditioning, reaching only 69% of her expected maximum exercise capacity. Echocardiogram showed no structural heart disease. Since other causes of presyncope and palpitation were excluded, tilt table test (TTT) was performed which revealed blood pressure of 135/86 mmHg with a heart rate of 90 bpm in supine position. When tilted, the blood pressure remained stable, while the heart rate increased to a maximum of 147 bpm (Figure 1B). The heart rate was still around 120 bpm after 10 minutes of the tilting. With these results the patient fulfilled the criteria of PoTS. Diagnosis of PoTS was favoured because of persistence of presyncope, light-headedness and palpitation after exclusion of other causes and in association with positive results of tilt table test. She was advised to keep well hydrated, take in enough sodium (kitchen salt) and to increase her exercise tolerance (potentially in the cardiac rehabilitation program).

## DISCUSSION

Postural tachycardia syndrome (PoTS) is an autonomic disorder, characterized by sustained heart rate increment of  $\geq 30$  beats/min ( $\geq 40$  beats/min in individuals  $< 19$  years of age) within 10 minutes of standing in the absence of orthostatic hypotension [1]. PoTS is multifactorial in etiology i.e. hypovolemia, neuropathic, hyperadrenergic, deconditioning, autoimmune and/or mast cell activation disorder) but the specific pathophysiology is still unclear [2-4]. The true prevalence of PoTS is unknown, but it is estimated that the prevalence is at least 170/100.000 [5]. Symptoms involve the presentation of cerebral hypoperfusion and/or autonomic dysfunction. As the symptoms are often nonspecific, the diagnosis is especially challenging. When PoTS is suspected, it is subsequently confirmed by the tilt table test.

The above-described case shows a deconditioned patient, suffering from several nonspecific symptoms associated with PoTS. After many years and several visits to different health care providers and various diagnostic tests over the years, she underwent the tilt table test, which confirmed the diagnosis of PoTS.

Symptoms of PoTS vary greatly, are often nonspecific and they can start and disappear suddenly. This was the case in our current patient, symptoms recurred twice and she underwent several diagnostic tests, which showed no abnormalities. On those two

earlier occasions, no TTT was performed. Orthostatic symptoms involves lightheadedness, dizziness, palpitations, tremulousness, weakness (mostly of the legs) and visual blurring. Other symptoms are fatigue, exercise intolerance, hyperventilation, dyspnea and anxiety. An overview of all possible symptoms, adopted from previous publications [6-8], is illustrated (Figure 1A). As the symptoms exacerbate by simple activities of daily life, such as eating or exercise [2-4], the impact of PoTS is debilitating. In patients suffering from the above-mentioned symptoms, it is important to rule out causes such as anemia, electrolyte disturbances, cardiac conditions and endocrine disorders. Subsequently, the diagnosis of PoTS can be confirmed by obtaining the tilt table test [2]. As this test is easy to perform, the most important part of the diagnostic process is the recognition of the symptoms by the health care professional. Until now, the awareness of PoTS is low, leading to frequent misdiagnosis of panic disorder or chronic anxiety [9]. In the above-described patient, this was also the case. The patient suffered from lightheadedness and palpitations for several years, in which she has visited the cardiologist (among others) several times and underwent various diagnostic tests. However, it was only in the last visit, 8 years after initiation of symptoms, in which the cardiologist associated the symptoms with PoTS.

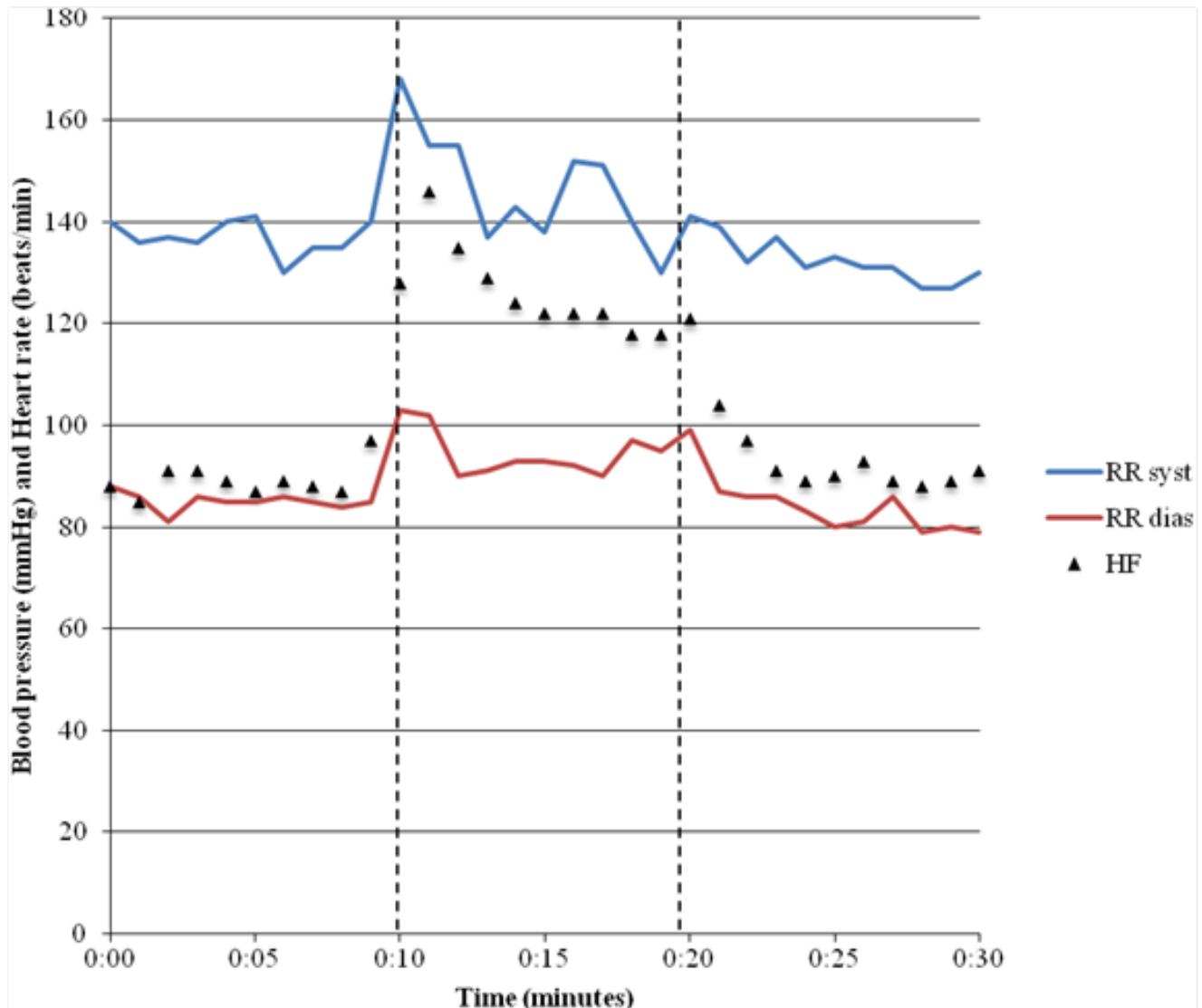
The recognition of PoTS is mainly important because there are several non-pharmacological and pharmacological treatment strategies available. Initially the focus should be on treating the reversible causes such as deconditioning, optimizing the treatment of chronic diseases, elastic support hoses to minimize peripheral venous pooling, increasing the water and salt intake and education about avoiding triggers such as dehydration, alcohol abuse and extreme heat [10;11]. As patients with PoTS also have exercise intolerance, it is suggested that a non-pharmacological approach of treatment may include endurance exercise training. Endurance training induces cardiac remodeling and an increase in blood volume. This can lead to an improvement in symptoms and quality of life and decrease in the standing heart rate [12;13]. Recently, it has been reported that there was a large group (about 50%) who did not meet the criteria of PoTS anymore after three months of endurance training [12;13]. Pharmacological treatment of PoTS patients involves fludrocortisone (volume expansion), midodrine (vasoconstriction), beta-adrenergic blockers (reduce sympathetic influence on sinus node) and/or pyridostigmine (increases synaptic acetylcholine) [10;14]. Even though there are small studies describing positive effects of these drugs [10], the Food and Drug Administration (FDA) hasn't approved any of these drugs for the treatment of PoTS because of the lack of large randomized clinical studies [10]. The current case reflects the challenge clinicians may face for timely diagnosis of PoTS.

PoTS is a challenging diagnosis and presumably under diagnosed because of the nonspecific presentation. Delay may occur until diagnosis is established. PoTS may be included in the differential diagnosis of palpitation, presyncope, shortness of breath and lightheadedness. Tilt table test is of a pivotal value in establishing and confirming the diagnosis of PoTS. This case report represents the difficulty of diagnosis and the value of the tilt table test.

**Figure 1A:** An intake list used for assessment of symptoms in suspected PoTS patients, adopted from [6-8]. Intake list regarding the symptoms of PoTS, filled in by the patient. - = none, + = discrete, ++ = moderate and +++ = severe.

Symptom	Score			
	-	+	++	+++
Sweating			moderate	
Weakness/extreme fatigue	none			
Sleep disorders	none			
Palpitations/tachycardia				severe
Exercise intolerance	none			
Light-headedness			moderate	
Headache	none			
Mental clouding	none			
Blurred or tunneled vision			moderate	
Shortness of breath			moderate	
Hyperventilation	none			
Tremulousness (shaking)	none			
Chest discomfort	none			
Vomiting	none			
Presyncope			moderate	
Syncope/fainting	none			
Anxiety				severe
Memory difficulty and concentration disturbances	none			
Facial flushing		discrete		
Acrocyanosis	none			
Mood changes	none			
Attention disturbances	none			

**Figure 1B:** Tilt table test . Positive Tilt Table Test of our described patient. The first 10 minutes is in supine position. At 10 minutes the table was tilted, with an increase in heart rate up to 147 beats/min. After 10 minutes in upright position, the heart rate was still 120 beats/min. blue line = RR syst = systolic blood pressure; red line = RR dias = diastolic blood pressure; ▲ = HF = heart frequency.



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