

## Postural Orthostatic Tachycardia Syndrome Associated with Neurocardiogenic Syncope: A Combined Form of Dysautonomia

Ibtissam Romani\*, Dounia Benzeroual, Saloua El Karimi, Mustapha El Hattouai

Cardiology and Vascular Disease Department of Mohamed VI University Hospital, Marrakech, Morocco

\*Corresponding author: Ibtissam Romani  
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### Abstract

### Case Report

The autonomic nervous system (ANS) controls all body functions. Dysregulation of this system may be responsible of Orthostatic intolerance (OI). OI forms may coexist in the same patients. So we report a 24-year-old female case who suffers from coexisting features of postural tachycardia syndrome (POTS) and neurocardiogenic syncope (NCS). Through this case we describe the clinical features and management of this autonomic disorder. The patient reported clinical symptoms consistent with both POTS and NCS and then demonstrated typical POTS (a rise in heart rate (HR) without change in blood pressure [BP]) within the first 10 minutes of upright posture followed by neurocardiogenic syncope (a sudden fall in HR and BP). The treatment (hygienodietetic measures, fludrocortisone) showed an improvement in the functional state of the patient. NCS may coexist with POTS in a subgroup of patients suffering from OI. Autonomic profile study helps to explain the causes of symptoms described by this patient with whose clinical and conventional paraclinical examinations are normal.

**Keywords:** orthostatic intolerance, postural tachycardia syndrome, neurocardiogenic syncope.

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## INTRODUCTION

The autonomic nervous system (ANS) controls all body functions and maintains internal homeostasis and stress responses. Dysregulation of ANS, called dysautonomia, may be responsible of Orthostatic intolerance(OI). Orthostatic intolerance syndromes refer to a heterogeneous group of disorders of hemodynamic regulation characterized by excessive pooling of blood in the dependent areas of the body during upright position, leading to insufficient cerebral perfusion causing a variety of symptoms.

OI can be divided into subgroups that include postural tachycardia syndrome (POTS), neurocardiogenic syncope (NCS) and dysautonomic (autonomic failure) syndromes.

One or more forms of these subgroups may coexist in the same patients. However, published data on the clinical features and management of patients who suffer from coexisting features of POTS and NCS is very poor. So we report our experience by this clinical case of a 24 years old patient who we found evidence of coexisting NCS and POTS.

## CASE REPORT

A 24-year-old female reports a 3 years' history of light-headedness on standing. This is often associated

with palpitations, fatigue and a feeling of intense anxiety. She has had much episodic syncope in the past 12 months. She had more than 6 black-outs per day causing several traumas. She is not taking any regular medications.

Her supine blood pressure (BP) was 97/61 mmHg with a heart rate (HR) of 66 bpm. The clinical cardiovascular and neurological has shown no abnormality. Para-clinical tests were also normal:

- Cardiovascular testing (Electrocardiogram, 24 hours ECG holer monitor, Echocardiography).
- Cranial CT scan and electroencephalogram.
- Blood tests including thyroid hormones.

The patient has then undergone an exploration in our ANS unit of the cardiology department. The results were as follows:

- ECG: Regular sinus rhythm at 65c/mn.
- Deep breathing test: The vagal response was 66% (N= 30%) associated with a decrease of blood pressure from 96/60mmHg to 88/60mmHg.
- Hand grip test:
  - The vagal response was 30% (N=10%).
  - The sympathetic peripheral alpha response was 20% (N=10%).

**Hyperventilation test**

- An increased heart rate from 70 BPM to 120 BPM.
- A decrease in the blood pressure from 95/60 mmhg to 89/56mmhg.

**Mental Stress**

- Central sympathetic response alpha: 8% (N=10%)
- Sympathetic beta response: 15% (N= 10%).

**Orthostatic Testing**

- An increased heart rate from 67 to 110 BPM, then to 132 BPM within the first 10 minutes.
- An increased in BP from 95/65 to 106/70 mmHg then maintained at around 108/ 70mmHg for 30min.
- Suddenly, in the 40th minute, we noted a concomitant fall of the HR at 50bpm and then at 35bpm, and of BP at 69 /50mmHg with a syncope imposing the stopping of the test and the return to the decubitus dorsal.

Decubitus dorsal: BP and HR returned progressively to normal (BP at 96 /70mmHg and HR at 68bpm).

The autonomic profile of the patient revealed significant peripheral and central beta hyperactivity, compatible with a severe POTS and central sympathetic alpha deficiency. The peripheral alpha sympathetic was very poor in orthostatic test with occurrence of severe hypotension within 30 minutes in favor of complicated POTS. The test showed also a major increased vagal tone.

The patient had received as treatment fluid volume filling, fludrocortisone, alpha mimetic, vitamins and venous contention. A regular follow-up of the patient showed a regression of the symptoms and an improvement of the test responses. Fludrocortisone was progressively reduced until arrest after 1 year. Beta blockers were intruded after improvement of BP values.

**DISCUSSION**

POTS was defined as ongoing symptoms of OI (of greater than 6 months' duration) accompanied by a heart rate increase of at least 30 beats/min (or a rate that exceeds 120 beats/min) observed during the first 10 minutes of upright posture or HUTT occurring in the absence of other chronic debilitating disorders [1,2].

Symptoms may include fatigue, orthostatic palpitations, exercise intolerance, lightheadedness, diminished concentration, headache, near syncope, and syncope [3, 4]. This syndrome can coexist with different clinical situations. NCS, most common cause of syncope, is characterized by inappropriate vasodilatation leading to transient neurally mediated systemic hypotension and bradycardia resulting in low cerebral blood flow and loss of consciousness.

Several potential mechanisms for pathophysiology of POTS are defined. This syndrome may have multiple etiologies and we now know that POTS can have multiple variants such as partial dysautonomia [5], centrally mediated hyperadrenergic stimulation [6, 7], norepinephrine transporter dysfunction [8] and POTS associated with deconditioning [9] and hypovolemia [10].

The patient had an increased vagal tone in all tests. Alshekhlee *et al.* describe a series of four POTS patients who had a surge of parasympathetic activity resulting in marked cardioinhibition and vasodepression [11]. They postulated that either a compensatory parasympathetic surge or a central aberration altering both sympathetic as well as parasympathetic output in a balanced fashion may account for increased parasympathetic activity in this group of patients.

Our patient presented in orthostatic test a NCS. The coexistence between POTS and NCS is frequent but unknown. This association is attributed to alpha sympathetic deficiency with hypovolemia. Furthermore, Ojha *et al.* postulate that an initial compensatory increase in sympathetic outflow that increases the inotropy as well as chronotropy of the heart may fatigue or norepinephrine stores may become exhausted, resulting in a state of relative sympathetic withdrawal causing a state of bradycardia and hypotension in this group of patients [12].

It is reasonable for all patients to receive advice and education on factors that influence systemic blood pressure, such as avoiding sudden head-up postural changes, standing still for a prolonged period of time, high environmental temperature (including hot baths, showers), severe exertion, large meals, alcohol and drugs with vasodepressor properties [13]. Additional treatment principles used alone or in combination, are appropriate for consideration on an individual patient basis: Chronic expansion of intravascular volume by encouraging a higher than normal salt intake and fluid intake of 2-2.5 liters per day. Additional options include use of fludrocortisone in low dose (0.1 to 0.2 mg per day), and counter pressure manoeuvres of the legs (leg crossing), or of the arms (hand grip and arm tensing) [14]. These measures were done to our patient with a spectacular improvement of all symptoms.

**CONCLUSION**

Dysautonomic syncope is frequent but unknown the delay diagnostic aggravates their recrudescence. The autonomic profile study helps to explain the causes of symptoms described by the patients whose clinical and conventional paraclinical examinations are normal. NCS may coexist with POTS in a subgroup of patients suffering from OI. This group of patients with mixed-form OI may be difficult to treat and may have syncope as a dominant symptom.

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