

## CASE REPORT

**Syncope and hypermobile joints: Not rare, but rarely diagnosed****Senkop ve hiper mobil eklemler: Nadir değildir fakat nadiren tanı konulur**

Elnur Tahirović, M.D., PhD.

Cardiovascular Surgery Clinic, University of Sarajevo Clinical Center, Sarajevo, Bosnia and Herzegovina

**Summary**– Postural orthostatic tachycardia syndrome (POTS) is a chronic, debilitating condition characterized by heterogeneous symptoms, such as lightheadedness, palpitations, pre-syncope, syncope, and weakness or heaviness, especially of the legs. It is frequently associated with hypermobile joints or conditions such as chronic fatigue syndrome, chronic abdominal pain, migraine headache, and diabetes mellitus. Described is a case of POTS, which though it is not rare, is rarely diagnosed. It can be diagnosed quickly with simple methods.

Postural orthostatic tachycardia syndrome (POTS) is an abnormal increase in heart rate that occurs after sitting up or standing, followed by symptoms such as presyncope and syncope, and is frequently associated with hypermobile joints.<sup>[1]</sup> The pathophysiology underlying POTS remains incompletely understood, but is likely to be multifactorial and varies in different subgroups of POTS patients.<sup>[2]</sup> POTS predominantly affects women, with a female: male ratio of 4.5:1 and with an age range from 15 to 50 years.<sup>[1,3]</sup> The condition can be quite disabling. The symptoms are heterogeneous, may range from mild to severe, and vary from day to day. That is why POTS is often underdiagnosed.

Presently described is a case of POTS, which though it is not rare, is rarely diagnosed. It may, however, be diagnosed quickly with the help of simple methods.

**CASE REPORT**

A 16-year-old female patient, accompanied by her mother, was admitted to the arrhythmia center because of unusual episodes of dizziness and syncope with spontaneous recovery. Most of these symptoms occurred when she stood up. She had symptoms of palpi-

**Özet**– Postural ortostatik taşikardi sendromu (POTS) baş dönmesi, çarpıntı, presenkop, senkop ve özellikle bacaklarda güçsüzlük veya ağırlık gibi heterojen semptomlarla karakterize edilen, kronik ve güçsüzleştirici bir durumdur. Sıklıkla hiper mobil eklemler veya kronik yorgunluk sendromu, kronik karın ağrısı, migren, baş ağrısı ve şeker hastalığı gibi durumlarla ilişkilidir. Tartışılan olgu, nadir olmamasına rağmen nadiren tanı konan bir POTS olgusudur. Basit yöntemlerle hızlı bir şekilde tanı konabilir.

tations, tiredness, nausea, and some gastrointestinal problems, and she did not tolerate heat. Most of these symptoms had started in puberty. Her anamnesis indicated problems with her knee joints as a result of snowboarding, with a diagnosis of instability of the patella and the patella alta. An initial Holter electrocardiogram (ECG) and echocardiography exam were normal. A physical examination revealed no abnormalities. Her pulse was about 65 bpm and her blood pressure was 120/80 mm Hg. Upon examination, it was noticed that her joints were hypermobile and her skin was hyperelastic (Fig. 1a, b). The Beighton score test<sup>[4]</sup> result was 7. An active stand test was performed, and it was observed that her pulse increased by more than 30 bpm during the 10-minute test when compared with the supine heart rate recorded before the test. Another Holter ECG was performed with the recommendation to perform similar test activities a few times, such as the active stand test and standing for 10 minutes after waking on the second day. A similar increase in heart rate was noticed on the Holter ECG report after she woke up in the morning, which was associated with

**Abbreviations:**

ECG	Electrocardiogram
POTS	Postural orthostatic tachycardia syndrome

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Correspondence: Dr. Elnur Tahirovic. Bolnicka 25 71000 Sarajevo - Bosnia and Herzegovina.

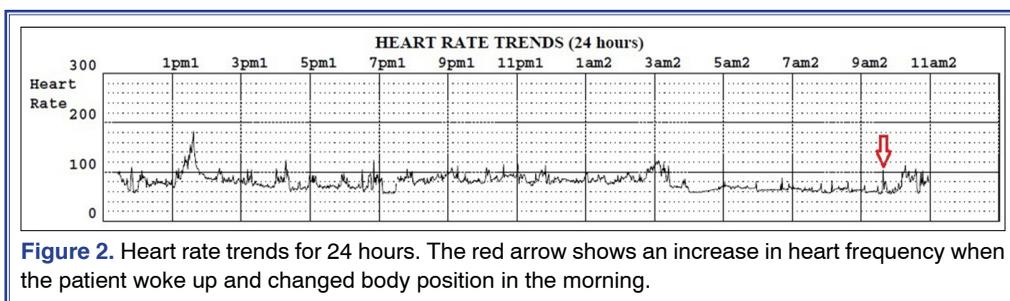
Tel: 0038761800269 e-mail: elnur.tahirovic@gmail.com

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**Figure 1.** (A) Passive dorsiflexion of the fifth metacarpal joint to  $>90^\circ$ . (B) Apposition of the thumb to the flexor aspect of the forearm.



**Figure 2.** Heart rate trends for 24 hours. The red arrow shows an increase in heart frequency when the patient woke up and changed body position in the morning.

upright posture dizziness, presyncope symptoms, and heavy arms and legs. It was impossible for the patient to stand and she had to lie in bed (Fig. 2). The patient was subsequently diagnosed with POTS, a form of dysautonomia, or an abnormality of the functioning of the autonomic nervous system. Three months after receiving suggestions for lifestyle adjustments and being advised to consume more water and salt, the patient became asymptomatic.

## DISCUSSION

POTS is defined as the presence of orthostatic intolerance associated with a heart rate that is increased by more than 30 bpm (for teenagers, an increase of 40 bpm) within 10 minutes of standing and in the absence of orthostatic hypotension.<sup>[2]</sup> It is frequently associated with hypermobile joints and conditions such as Ehlers-Danlos syndrome.<sup>[4,5]</sup> It is a disorder of the autonomic nervous system, characterized by heterogeneous symptoms of palpitations, exercise intolerance, fatigue, tremor, lightheadedness, migraine-like headaches, nausea, syncope, and near syncope, and impaired health-related quality of life. To make a diagnosis of POTS, in addition to a complete physical examination, it is also necessary to perform a head-up tilt table test, an active stand test and Holter ECG monitoring.<sup>[6]</sup> The head-up tilt table test is the gold standard

of POTS diagnosis, but when it is not available, it is possible to confirm the diagnosis of POTS based on Holter ECG results. The Holter ECG makes it possible to record heart rate and rhythm disorders during usual daily activities. It is necessary to explain to the patient that they should reproduce events that seem to cause the symptoms during the Holter ECG recording. Moon et al.<sup>[7]</sup> reported that orthostatic tachycardia was more prominent in the morning, but not always, and that among POTS patients, 82.6% met the diagnostic criteria for POTS in the morning and 52.2% in the afternoon.

In the present case, we asked the patient to stand for 10 minutes in the presence of an accompanying family member when she woke up in the morning of the second day of Holter ECG recording. The ECG documented an increase in heart rate of more than 40 beats per minute associated with other POTS symptoms noted for that period in the Holter diary. Similar results were reported by Brewster et al.<sup>[8]</sup> in their study. They reported that the key finding was that orthostatic tachycardia was significantly higher in the morning, with an absolute heart rate increase among patients with POTS. The increase in heart rate is a normal physiological phenomenon that is exaggerated in patients with POTS when standing in the morning. Their patients also reported that symptoms of palpita-

tions and lightheadedness were worse in the morning, while some other symptoms, such as fatigue, were worse later in the day.

In our case report, the use of the Holter ECG as a diagnostic tool was shown to be efficient in proving and confirming the diagnosis of POTS, with the addition of a physical examination, the active stand test, and the Beighton scale, but the full cooperation of the patient was necessary. The Holter ECG was helpful when the head-up tilt table test was not available.

The connection between POTS and hypermobile joints is not clearly understood. Gazit et al.<sup>[9]</sup> in their study found that a large percentage of patients with Ehler-Danlos syndrome, or hypermobile joints, also have some form of dysautonomia, such as POTS. These authors found that patients with hypermobile joints have a collagen defect, which is responsible for the overstretchy blood vessels and other symptoms of POTS.

The treatment of POTS can be pharmacological and nonpharmacological. In our case, we started with nonpharmacological treatment such as exercise, volume expansion, and increased salt and fluid intake. Many authors have found that patients with POTS have a small heart size and mass with reduced plasma and blood volume, which contributes significantly to a smaller stroke volume and reflex tachycardia during orthostasis.<sup>[10–12]</sup> In our case, this treatment was successful, but sometimes it requires a longer period of follow-up and the inclusion of drug therapy to achieve good symptom control and a good quality of life.

## Conclusion

This report describes a case of POTS, a form of dysautonomia associated with hypermobile joints. A Holter ECG monitoring may be very useful in confirming the diagnosis of POTS when a head-up tilt table test is inaccessible.

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**Keywords:** Holter electrocardiogram; hypermobile joints; postural orthostatic tachycardia syndrome; syncope.

**Anahtar sözcükler:** Holter elektrokardiyogram; hipermobil eklemler; postural ortostatik taşikardi sendromu; senkop.