# A Case Report and Review of Postural Orthostatic Syndrome in an Adolescent

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## ABSTRACT

Because there are many young women between the ages of 12 and 25 years who have been diagnosed with postural orthostatic syndrome (POTS), with the time to the diagnosis of about 5 years, it is important for health care providers to have an understanding of the clinical presentation of POTS to manage the diagnosis appropriately. The purpose of this article is to present a case study review of an adolescent woman who experienced POTS syndrome at age 16 years and to provide a clinical overview of POTS in the adolescent population. J Pediatr Health Care. (2017) ■, ■-■.

## KEY WORDS

Adolescent care, cardiac, gastrointestinal disorder, postural tachycardia syndrome, POTS

## POSTURAL TACHYCARDIA SYNDROME

Postural tachycardia syndrome (POTS) was first recognized in 1993 by neurologist Dr. Philip Low and his research team (Schondorf & Low, 1993). Other names associated with the term POTS are DaCosta's syndrome, soldier's heart, mitral valve prolapse syndrome, neurocirculatory asthenia, chronic orthostatic intolerance, and orthostatic tachycardia (Dysautonomia International, 2012). For children and adolescents, POTS is suspected in the presence of dizziness, weakness, and tachycardia (Kizilbash et al., 2014; Raj & Levine, 2013). Tachycardia associated with POTS is defined as a heart rate increase of more than 30 beats per minute (bpm) for those older than 19 years and an increase of more than 40 bpm for those younger than 19 years within the first minutes of standing upright (Kizilbash et al., 2014; Raj & Levine, 2013). There is no drop in blood pressure (BP) and no other obvious cause (such as active bleeding, medications, or acute dehydration) for the tachycardia, and the tachycardia persists longer than 6 months (Kizilbash et al., 2014; Raj & Levine, 2013). Other symptoms associated with POTS include sweating, temperature regulation issues, and altered bowel and bladder function (Raj, 2013). It is estimated that 500,000 Americans report being affected with POTS, most of them women between the ages of 12 and 25 years (Agarwal, Garg, Ritch, & Sarker, 2007).

POTS is thought to result from a malfunction in the autonomic nervous system, which regulates bodily functions not under conscious control such as heart rate, blood pressure, body temperature, breathing, and digestion (Benarroch, 2012). No single mechanism explains the clinical presentation of POTS. One theory suggests that outward symptoms are a result of a combination of processes that causes decreased blood return to the heart, producing the clinical symptoms seen (Benarroch, 2012). The dysfunctional process involves the messages sent to blood vessels, typically by the nerves, that tell the blood vessels when to expand and contract. For patients with POTS, the blood vessels become overdilated, and the blood pools in the lower part of the body, resulting in decreased blood flow to the head (Benarroch, 2012). The heart attempts to

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compensate for the reduced blood flow by increasing the heart rate, and thus patients report symptoms of dizziness and lightheadedness upon standing (Benarroch, 2012).

## CASE DESCRIPTION

Mandy, a 19-year-old woman says that up until her sophomore year in high school, she had been a good student, was involved in athletics, and had many friends. It was during her sophomore year that she began to experience severe abdominal pain, had frequent headaches, would faint on occasion, and experienced fatigue to the point that she had difficulty getting out of bed. Various health care providers, including her pediatrician and the gastroenterologist to whom she was referred, failed to find a physical cause for her symptoms. Unsure of a medical diagnosis, health care providers referred the girl to a psychiatrist for what was termed "manipulative malingering" behavior, and she was admitted to a mental health unit for treatment. She spent her sophomore year in this facility, where, as she tells the story, she was placed in a wheelchair and was not allowed out until she no longer fainted. All the while her fatigue, abdominal pain, and headaches continued in addition to her fainting. After a year of psychiatric treatment and no change in physical behavior, she was discharged. Within days of her hospital discharge, a neighbor had been watching a health news segment on the evening news talking about something called *postural tachycardia syndrome,* or *POTS* for short. As soon as the segment was over, the neighbor contacted Mandy's mother and relayed what she had heard, noting that much of what was said sounded like what she was experiencing. The neighbor encouraged her mother to contact her pediatrician and ask for a cardiology referral. Having no other options, her mother called the pediatrician and requested a cardiology consultation, where Mandy learned that she indeed had POTS.

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## POTS CLASSIFICATIONS

Classified by primary and secondary forms, *primary POTS* is considered idiopathic and not associated with any other disease, whereas *secondary POTS* occurs in conjunction with a known disease or disorder (Grubb, Kanjwal, & Kosinski, 2006). Within each classification are forms of POTS. Primary POTS forms include *partial dysautonomic* and *hyperadrenergic*. Within the partial dysautonomic subtype are two forms called developmental, which seems to affect adolescents, and postviral. Secondary POTS occurs in conjunction with diseases such as diabetes, joint hypermobility syndrome, and paraneoplastic syndrome (Grubb, Kanjwal, & Kosinski, 2006).

### Primary POTS

Patients with primary partial dysautonomic POTS present with a mild form of peripheral autonomic neuropathy in which the peripheral vasculature cannot maintain or initiate vascular resistance in the presence of gravitational pull; thus, blood pools in the lower extremities (Grubb, Kanjwal, & Kosinski, 2006). Of this type, there is a 5:1 female-to-male ratio. The developmental subtype of partial dysautonomic POTS seems to affect adolescents (Grubb, Row, & Calkins, 2005). Developmental POTS symptoms begin t or around age 14 years and worsen until about age 16 years, many times leaving the adolescent incapacitated (Grubb, 2008). Once symptoms have peaked, 80% of adolescents diagnosed with developmental dysautonomic POTS are asymptomatic by age 19 to 24 years (Grubb, 2008). Although the cause of developmental dysautonomic POTS is not known, it has been linked to the temporary autonomic imbalance that often occurs in the rapidly growing adolescent (Grubb, Kanjwal, & Kosinski, 2006).

Another form of partial dysautonomic POTS is triggered after an acute viral or bacterial infection such as the flu, mononucleosis, or gastroenteritis. After these illnesses, it appears that patients have difficulty recovering and continue to experience fatigue, flu-like illness, dizziness, nausea, and headaches (Grubb, Kanjwal, & Kosinski, 2006).

Hyperadrenergic POTS occurs when naturally occurring stimulants (adrenaline and noradrenaline) are present in higher levels, which can lead to an increase in heart rate and blood pressure; thus, patients present with both an increased blood pressure and an increased heart rate when standing (Grubb, Row, & Calkins, 2005). Patients experiencing this form of POTS often report a slower more progressive onset of symptoms over a long time (Grubb, Row, & Calkins, 2005).

### Secondary POTS

Secondary POTS refers to syndromes that occur as a result of another underlying condition that leads to damage of the nerves that control blood distribution. These conditions include diabetes, lupus, alcoholism, and chemotherapy (Grubb, Row, & Calkins, 2005).

Another form of secondary POTS is the connective tissue disorder known as joint hypermobility syndrome (Gazit, Nahir, Grahame, & Jacob, 2003). This syndrome