Postural Tachycardia Syndrome and Reflex Syncope: Similarities and Differences

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As awareness of postural tachycardia syndrome (POTS) has increased in recent years,[1-3] our laboratory has received numerous patient referrals for "POTS going to faint." These comprise patients who, during tilt table testing, have development of excessive tachycardia, sometimes gradually decreasing blood pressure and symptoms of orthostatic intolerance, followed by a simple postural faint (current designation "reflex neurocardiogenic syncope"[4]). Consider 2 representative teenage patients placed on a tilt table for a 20- to 30-minute supine equilibration period and then tilted upright to 70 degrees. Both have development of excessive increases in heart rate within 5 minutes of the onset of upright tilt (Figure 1). Both become lightheaded and nauseated and have development of progressive neurocognitive impairment. Respirations deepen, and end tidal carbon dioxide is significantly decreased. They are both pale and somewhat diaphoretic. Blood pressure may slowly decrease. Just before pronouncing the diagnosis of POTS, 1 patient abruptly loses consciousness in association with a rapid decrease in blood pressure and heart rate characteristic of the "vasovagal faint" variant of reflex syncope because of the decrease in blood pressure (the vaso or vascular part) followed immediately by a sudden drop in heart rate (the vagal part).[5] This causes an equally abrupt change in the diagnosis from POTS to simple faint. The other patient continues to have an excessively increased heart rate and symptoms of orthostatic intolerance but has no decrease in blood pressure or heart rate, or loss of consciousness. Blood pressure and heart rate data for the representative tilt subjects are shown in Figure 1. These scenarios have fostered concerns about the clinical and physiological similarities and differences between simple faint and POTS. The answer appears to be a qualified no to clinical similarity and a qualified yes to physiological similarity. One could easily reverse this question and ask whether there are clinical and physiological differences between simple faint and POTS. The answer appears to be a qualified yes to clinical difference and a qualified no to physiological difference. The most notable differences between POTS and simple fainting are found in their clinical presentations, and therefore history taking is paramount in diagnoses. POTS is a chronic day-to-day form of orthostatic intolerance, and simple faint is most often episodic and associated with long periods of "wellness."

ACUTE AND CHRONIC ORTHOSTATIC INTOLERANCE

Orthostatic intolerance (OI) is defined by the presence of symptoms and signs while upright, relieved by recumbency. Symptoms include dizziness, impending loss of consciousness, headache, fatigue, neurocognitive or sleep disturbance, exercise intolerance, nausea/abdominal pain, heat, and sweating.[6] These roughly divide into symptoms of reduced cerebral and regional circulatory blood flow[7,8] and symptoms of sympathetic activation.[9] Acute orthostatic intolerance during adolescence usually presents as episodic simple postural faint (vasovagal syncope), and chronic orthostatic intolerance presents with POTS in which symptoms are associated with excessive upright tachycardia.[10] Both vasovagal syncope and POTS are defined clinically; there is no other reference standard test for their diagnosis. Syncope (fainting) is defined by a sudden transient loss of consciousness and postural tone caused by cerebral hypoperfusion.[11] Cerebral blood flow needs to decrease to approximately 50% of normal before consciousness is seriously impaired.[12] Symptoms and signs of impending faint are the signs and symptoms of orthostatic intolerance: commonly pallor, nausea or stomach discomfort, headache, cold or warm feelings, and dizziness, which is often initially misinterpreted as a more ill-defined lightheadedness or cognitive loss. Post faint recovery may be protracted with residual pallor and weakness.

Generally, syncope can result from serious cardiovascular and cerebrovascular pathology.[13] This is beyond the scope of the present review, which focuses exclusively on orthostatic intolerance. POTS is identified with chronic orthostatic intolerance[14-16] and

OI	Orthostatic intolerance	POTS	Postural tachycardia syndrome

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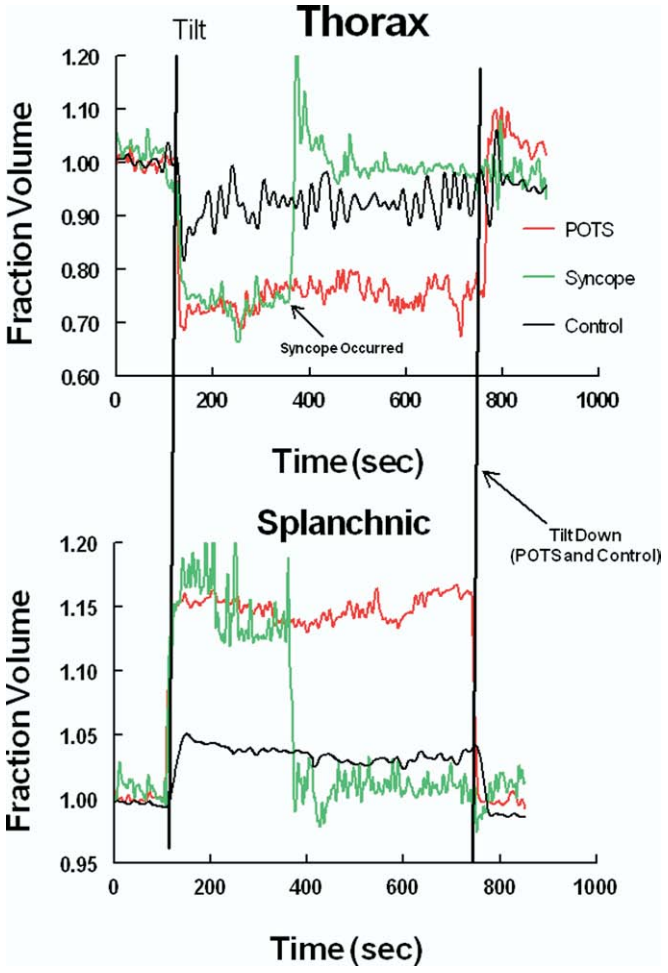

Figure 1. The figure shows heart rate in the **top panel** and mean arterial pressure in the **bottom panel** during upright tilt to 70 degrees. A representative patient with POTS is shown in *red* and a representative patient with syncope is shown in *green*. Excessive tachycardia and symptoms occur in both patients during tilt. Tilt is terminated in the patient with syncope at the time of faint, and the table is placed in the supine position.

is defined operationally by symptoms of OI in association with excessive tachycardia. In adults excessive postural tachycardia comprises an increase of heart rate by 30 or more beats/min during upright tilt or to a heart rate exceeding 120 beats/min within 10 minutes of being placed upright.[1] Hypotension has been until recently an exclusionary criterion during the 10 minute testing period but can occur later during tilt table testing in some patients with typical chronic OI (Phillip Low, personal communication, April 2008) in which it is associated with severe cerebral hypoperfusion.[17,18] Some patients have waxing and waning symptoms, although usually not at regular intervals. In healthy young people, especially those with a low supine resting heart rate, the normal postural increase in heart rate is often much larger than in adults. The reason for this is straightforward: tonic vagal activity is maximum during the teenage years[19] and low heart rates are common, particularly in young athletes.[20] Baroreflex-mediated vagal withdrawal (maximum vagal withdrawal is typically associated with a heart rate in the range of 100 beats/min in orthotopic transplants[21]) occurs during the standing process[22] and a relatively large increase in heart rate is common. A diagnosis of POTS in adolescent patients requires chronic symptoms of OI, as well as "excessive tachycardia" that appear with standing and are relieved by recumbency. During laboratory tilt testing symptoms of OI are typically duplicated and are associated with an excessive increase in heart rate that is usually much in excess of 30 beats/min. Symptoms may worsen with prolonged standing. Hypotension and fainting may occur during long stand times, and the patients are trained to sit or lie down once symptoms occur. However, other patients with POTS will not faint throughout 30 or more minutes of tilt. In addition, many healthy volunteers can have symptoms and signs of POTS during laboratory testing but not during daily life where they are generally untroubled by chronic OI.[23]

DEFINING ILLNESS IN OI

How then to define illness? The 2 general ways that patients are evaluated for medical ailments are by history/physical examination and by laboratory tests. There are no evidence-based studies indicating the clinical utility of brain imaging studies. For orthostatic intolerance, tilt table testing, a kind of orthostatic stress test, has been used as a primary laboratory tool. Tilt testing was first used in the 19th century,[24] and later by NASA and aerospace scientists as a test for physiological changes with change of posture.[25] The first research reports with tilt testing used to evaluate simple faint were published in the 1980s.[26,27] Although tilt testing can be made reasonably specific and sensitive in the evaluation of fainting in adults, debate continues concerning excessive false-positive and false-negative results in children and adolescents.[23,28] Thus on tilt table tests "fainters" do not always faint, and non-fainters sometimes do. Interestingly, evidence suggests that so-called false-positive results may indeed have a propensity to faint masked by movement-induced skeletal muscle pump activity.[29] This may be consistent with the idea that fainting is a normal response in the pediatric age range, but that the threshold for fainting varies widely from person to person.[30] Indeed, fainting is exceedingly common with onset typically in the pediatric age range.[31] However, just because something is "normal" does not make it desirable. Thus it is reasonable to speak of illness when orthostatic intolerance occurs with frequency or severity sufficient to impair quality of life.

Under the laboratory conditions that use tilt table stress combined with lower body negative pressure (which further forces blood to fill lower body veins and produces interstitial transudation), virtually all healthy volunteers can be made to experience a form of faint, vasovagal or otherwise.[32] So is tilt table testing useful? Perhaps it does not provide an accurate diagnostic tool for children. However, as shown below, it has considerable value in physiological assessment. Data from prior experiments indicate detectable differences of episodic fainters from healthy control subjects during the measurement of regional blood flows even in so-called "false-negative" results.