



CREATION OF A UNIQUE ICD-10-CM CODE FOR POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME (POTS)

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BACKGROUND

- POTS
 - Chronic autonomic nervous system disorder
 - Multiple systems affected
 - Neurologic, Cardiovascular, Gastrointestinal, Secretory
 - Causes severe disability and impaired QoL
 - Estimated to affect 500,000 - 3,000,000 people in the US
 - Estimates prior to SARS-CoV-2 pandemic

Shaw BH. J Intern Med 2019;286:438-48.

Garland EM. Curr Neuro Neurosci Rep 2015;15:60.

HISTORY

- First clinical description in medical literature: 1871
 - Civil War physician Jacob DaCosta
 - Post-infectious syndrome in soldiers
 - Severe lightheadedness, tachycardia, dyspnea, headache, fatigue, abdominal distension lasting months to years
 - Significant disability

HISTORY CONT.

- Known by multiple terms through history
 - Da Costa syndrome, Irritable Heart, Soldier's Heart, Civil War Syndrome, Effort Syndrome, Circulatory Neurasthenia
- Modern definition and name given in 1993
 - Schondorf and Low, Mayo Clinic

POTS CONSENSUS STATEMENTS

- American Autonomic Society, European Federation of Autonomic Societies, Autonomic Research Group of the World Federation of Neurology, Autonomic Disorders section of the American Academy of Neurology (2011)
- Heart Rhythm Society, American College of Cardiology, American Heart Association, Asia Pacific Heart Rhythm Society, European Heart Rhythm Association, Pediatric and Congenital Electrophysiology Society, Sociedad Latinoamericana de Estimulacion Cardiaca y Electrofisiologia (2015, 2020)
- Canadian Cardiovascular Society (2020)
- NIH POTS Research Workshop Expert Consensus (2021)

Freeman R Clin Auton Res 2011;21:69-72.
Sheldon R Heart Rhythm 2015; 12:e41.
Raj SR Can. J. Cardiol. 2020; 36: 357-372.
Vernino S Auton Neurosci 2021 (in press).

CURRENT DIAGNOSTIC CRITERIA

1. A sustained HR increment of not less than 30 beats/minute within 10 min of standing or head-up tilt. For individuals who are 12 to 19 years old, the required HR increment is at least 40 beats/minute; and
2. An absence of orthostatic hypotension (i.e. no sustained systolic blood pressure [BP] drop of 20 mmHg or more); and
3. Frequent symptoms of orthostatic intolerance during standing, with rapid improvement upon return to a supine position. Symptoms may include lightheadedness, palpitations, tremulousness, generalized weakness, blurred vision, and fatigue; and

CURRENT DIAGNOSTIC CRITERIA CONT.

4. Duration of symptoms for at least 3 months; and

5. Absence of other conditions explaining sinus tachycardia such as anorexia nervosa, primary anxiety disorders, hyperventilation, anemia, fever, pain, infection, dehydration, hyperthyroidism, pheochromocytoma, use of cardioactive drugs (e.g. sympathomimetics, anticholinergics) or severe deconditioning caused by prolonged bed rest.

PRESENT DAY

- Infection one of most common triggers of POTS
 - EBV, Lyme, URI, AGE, others
- Other triggers
 - Concussion, pregnancy, surgery, growth spurt, menarche
 - Some cases have no identifiable trigger

Shaw BH. J Intern Med 2019;286:438-48.

Boris JR. Cardiol Young 2018;28:668-74.

PRESENT ICD-10-CM CODING

- POTS often coded to I49.8: Other specified cardiac arrhythmias
 - Includes Brugada syndrome, coronary sinus rhythm disorder, ectopic rhythm disorder, nodal rhythm disorder
- Inaccurate placement and description
 - POTS is not a cardiac arrhythmia
 - Arrhythmias are not associated with POTS
 - Sinus tachycardia is noted upon standing (orthostasis), returns to NSR or lower rate of sinus tachycardia when seated/recumbent
- Others have used combination of R00.0 plus I95.1

IMPLICATIONS

- Limited ability to perform POTS research
 - Unique ICD-10 code necessary
 - Epidemiological, economic impact, medical utilization, other research
- Growing international interest
 - Academic medical centers publishing POTS research: US, Canada, Sweden, Germany, Austria, UK, Croatia, Italy, India, S. Korea, Japan, China, Australia
 - >1000 peer-reviewed articles

INCREASING GOVERNMENT FOCUS

2017: First US Congressional Briefing on POTS

2018: US Congress directs NIH to prepare report for Congress on state of POTS research/funding needs

2019: NIH held first international POTS Research Workshop

2020: NIH issued report on POTS to Congress

2021: NIH POTS Research Workshop panelists issued independent report – 2 peer-reviewed articles

2021: NIH issued Notice of Special Interest to stimulate research on diagnosis, treatment, mechanistic understanding of POTS

2021: NIH lists POTS funding in annual categorical spending report for the first time

<https://www.congress.gov/115/crpt/srpt289/CRPT-115srpt289.pdf>

<https://grants.nih.gov/grants/guide/notice-files/NOT-HL-21-008.html>

POTS IN ICD-10 RECOMMENDATIONS

- Placement in G90: Disorders of the autonomic nervous system
 - Definition: Postural Orthostatic Tachycardia Syndrome is a type of chronic orthostatic intolerance lasting three months or longer associated with excessive upright tachycardia in the absence of orthostatic hypotension, plus a constellation of typically daily symptoms which may include lightheadedness, dizziness, nausea, dyspnea, diaphoresis, headache, fatigue and other symptoms of autonomic dysfunction. Excessive tachycardia is defined by present consensus as a heart rate increase of at least 30 beats per minute in adults (40 beats per minute for adolescents), or a heart rate greater than 120 beats per minute, within 10 minutes of upright tilt table testing.
- Synonyms: Postural tachycardia syndrome, chronic orthostatic intolerance

IMPLEMENTATION

- If ICD-10-CM is amended to include a new POTS code, several organizations will assist in informing clinicians of the new code:
 - Dysautonomia International
 - American Autonomic Society
 - The above organizations will also reach out to a global network of professional societies and patient organizations with an interest in POTS.