DYSAUTONOMIA INTERNATIONAL

AWARENESS ADVOCACY ADVANCEMENT

Dysautonomia International
P.O. Box 596
East Moriches, NY 11940

September 14, 2021

ICD-10 Coordinating and Maintenance Committee
c/o David Berglund, MD, MPH
Medical Officer / Classification and Public Health Data Standards
National Center for Health Statistics, Mailstop P08
Metro IV, 2nd floor, Rm. 2534
3311 Toledo Rd.
Hyattsville, MD 20782

Re: Application for Unique ICD-10 Code for Postural Orthostatic Tachycardia Syndrome (POTS)

Dear Committee Members,

Dysautonomia International is the largest 501(c)(3) non-profit that advocates for over 70 million individuals worldwide who live with autonomic nervous system disorders. We are based in the US, but we provide support groups, clinician and patient education activities, and research grants around the world.

We write to you today in support of the creation of a unique ICD-10 code for postural orthostatic tachycardia syndrome (POTS). POTS is one of the most common autonomic nervous system disorders. It has been described in the medical literature since the 1800s under various terms, but was given its modern definition and objective diagnostic criteria in 1993 by the experienced autonomic neurologists at Mayo Clinic.

POTS is estimated to impact 500,000 to 3,000,000 individuals in the US, and millions of other individuals around the world. Performing more precise epidemiological estimates is hampered by the fact that POTS does not have a unique ICD-10 code.

As you may know, ICD-10 currently lists POTS under “I49.8 – Other specified cardiac arrhythmias.” ICD-10 notes that this code also “applicable to” Brugada syndrome, coronary sinus rhythm disorder, ectopic rhythm disorder, and nodal rhythm disorder. ICD-10 lists POTS as one of several “approximate synonyms” for this code: atrioventricular (av) tachycardia (fast heart beat), atrioventricular junctional heart rhythm, atrioventricular nodal re-entrant tachycardia, av junctional rhythm, junctional escape beats, postural orthostatic tachycardia syndrome, re-entrant atrioventricular node tachycardia, and re-entrant atrioventricular tachycardia.

Importantly, POTS is not a cardiac arrhythmia. The tachycardia seen in POTS is normal sinus rhythm tachycardia, which is exaggerated by upright posture as a response to several physiological mechanisms present in POTS patients.

POTS is a distinct clinical entity that is diagnosed using objective diagnostic criteria, confirmed by a physician. Research on POTS over the past 30 years has led to an understanding of the unique physiology of POTS, which includes hemodynamic, cardiovascular, neurologic, endocrine and immune findings. High-quality research on POTS is taking place at dozens of universities around the world, and hundreds of clinicians are offering POTS specialty care. Interest in POTS research and clinical care is growing rapidly, as some SARS-CoV-2 patients are developing a post-viral POTS. It has long been recognized that a subset of people with POTS develop POTS after viral infections. However, POTS may also be proceeded by concussions, pregnancy, surgery, bodily injury and other triggering events that stimulate the immune and autonomic nervous system. For some patients, there is no identifiable trigger. About 10-20% of POTS patients have a family history of POTS, suggesting genetic susceptibility may play a role.

The lack of unique ICD-10 code for POTS has made some forms of research difficult. For example, researchers at Walter Reed National Military Medical Center sought to determine the cost of POTS related healthcare claims for the military's Tricare health insurance program, which insures 10% of Americans. They evaluated all claims associated with ICD-10 code "I49.8 – Other specified cardiac arrhythmias," where POTS is currently listed as a synonym. Since so many other diagnoses are included in this code, they were unable to determine the cost of POTS related healthcare claims for the Tricare program. Confounding the problem, many clinicians do not use the I49.8 code for POTS, because POTS is not an arrhythmia.

The lack of a POTS specific code may also impede good healthcare for patients. In our modern healthcare system, patient records are often shared between hospital systems. Since there is no unique code for POTS, clinicians tend to use any code they think will get them paid. Reliance on imprecise coding can result in confusion for both the patient, other clinicians, and third-party payers. Having a unique code for POTS will result in more accurate medical records for patients, which will improve patient care.

If the Committee does grant this application, Dysautonomia International would be happy to assist with the dissemination of new POTS code information for clinicians. We maintain a mailing list of over 80,000 clinicians and patients interested in autonomic disorders that we can send any updated ICD-10 information to. We work closely with autonomic lab directors around the world (many of them signed a letter in support of this application, attached hereto), and they stand ready to educate their colleagues on any new POTS code. We have productive working relationships with several professional societies relevant to autonomic, cardiovascular, and neurologic disorders, and can work with them to educate their clinician members on any POTS code updates.

On behalf of millions of individuals living with POTS, we urge the Committee to support this application.

Respectfully submitted,

Lauren Stiles, JD
President, Dysautonomia International
Research Assistant Professor of Neurology, Stony Brook University School of Medicine

Enc.

September 14, 2021

ICD-10 Coordinating and Maintenance Committee
c/o David Berglund, MD, MPH
Medical Officer / Classification and Public Health Data Standards
National Center for Health Statistics, Mailstop P08
Metro IV, 2nd floor, Rm. 2534
3311 Toledo Rd.
Hyattsville, MD 20782

Dear Committee Members,

We are clinicians and researchers writing to offer comments in support of the creation of a unique code within ICD-10 for postural orthostatic tachycardia syndrome (POTS), a fairly common disorder of the autonomic nervous system, estimated to impact 500,000-3,000,000 Americans, and many others around the world.

Each of us sees a large number of patients with autonomic nervous system disorders in our clinics and research programs, including POTS patients. POTS is one of the most common diagnoses seen within autonomic clinics. It is readily distinguished from other autonomic disorders through objective testing and clearly defined diagnostic criteria that have been in use, with slight modification and updates, since POTS was defined by Mayo Clinic researchers in 1993.

Having a unique ICD-10 code for POTS will result in improved epidemiological, healthcare utilization, and other research on POTS, and will improve patient care by allowing more precise diagnosis codes to appear in medical records shared between healthcare providers.

We urge the Committee to support this application.

Respectfully submitted,

Amy C. Arnold, PhD
Assistant Professor
Department of Neural & Behavioral Sciences
Penn State College of Medicine
Adjunct Assistant Professor
Division of Clinical Pharmacology
& Autonomic Dysfunction Center
Vanderbilt University Medical Center
Hershey, PA, USA

Jonas Axelsson, MD, PhD
Associate Professor
Department of Clinical Immunology
Medical Director, Center for Apheresis and Stem Cell Handling
Karolinska University Hospital
Stockholm, Sweden

Italo Biaggioni, MD
Professor of Medicine
Director, Autonomic Dysfunction Center
Vanderbilt University Medical Center
Nashville, TN, USA

Brent Goodman, MD
Assistant Professor of Neurology
Director, Autonomic Laboratory
Mayo Clinic
Scottsdale, AZ, USA

Blair P. Grubb, MD
Distinguished University Professor of Medicine and Pediatrics
Director, Clinical Cardiac Electrophysiology Program
Director, Clinical Autonomic Disorders Center
University of Toledo Medical Center
Toledo, OH, USA

Mitchell Miglis, MD
Clinical Assistant Professor of Neurology & Neurological Sciences
Clinical Assistant Professor of Psychiatry & Behavioral Sciences
Stanford University
Palo Alto, CA, USA

Svetlana Blitshteyn, MD
Clinical Assistant Professor
University at Buffalo School of Medicine & Biomedical Sciences
Director, Dysautonomia Clinic
Buffalo, NY, USA

Kamal Chémali, MD
Associate Professor of Neurology
Eastern Virginia Medical School
Director, Autonomic Laboratory
Case Western Reserve University
Cleveland, OH, USA

Tae Hwan Chung, MD
Assistant Professor of Physical Medicine & Rehabilitation and Neurology
Director, POTS Clinic
Johns Hopkins University
Baltimore, MD, USA

Glen Cook, MD
Assistant Professor of Neurology
Uniformed Services University F. Edward Hebert School of Medicine
Director, Autonomic Laboratory
Walter Reed National Military Medical Center
Bethesda, Maryland, USA

Melissa Cortez, DO
Assistant Professor of Neurology
Director, Autonomic Physiology Laboratory
University of Utah
Salt Lake City, UT, USA

Andre Diedrich, MD, PhD
Research Professor of Medicine and Biomedical Engineering
Vanderbilt University Medical Center
Nashville, TN, USA

Taylor Doherty, MD, FAAAAI
Associate Professor of Medicine
Section Chief, Allergy & Immunology
University of California, San Diego
Staff Physician, Allergy & Immunology
VA San Diego Health System
San Diego, CA, USA

Amanda J. Miller, PhD
Postdoctoral Fellow
Department of Neural and Behavioral Sciences
Pennsylvania State University College of Medicine
Hershey, PA, USA

Jeffrey P. Moak, MD
Director, Electrophysiology and Pacing
Children's National Health System
Washington, DC, USA

Laura A. Pace, MD, PhD
Assistant Professor of Medicine
Director, Neurogastroenterology Clinic
Co-Director, Multidisciplinary Ehlers-Danlos Syndromes Clinic
University of Utah Adult Co-Lead, NIH Undiagnosed Diseases Network
Center for Genomic Medicine
University of Utah
Salt Lake City, UT, USA

Satish R. Raj, MD, MSCI, FRCPC
Professor of Cardiovascular Sciences
Director, Autonomic Investigation & Management Center
University of Calgary
Calgary, Ontario, Canada

Julian Stewart, MD, PhD
Professor of Pediatrics, Physiology & Medicine
Director, Center for Hypotension
New York Medical College
Hawthorne, NY, USA

Paola Sandroni, MD, PhD
Professor of Neurology
Director, Autonomic Laboratory
Mayo Clinic
Rochester, MN, USA

David M. Systrom, MD, FRCPC
Assistant Professor of Medicine
Harvard Medical School
Boston, MA, USA

Cyndya Shibao, MD, MSCI, FAHA, FAAS
Associate Professor of Medicine
Division of Clinical Pharmacology
Vanderbilt Autonomic Dysfunction Center
Vanderbilt University Medical Center
Nashville, TN, USA

Artur Fedorowski, MD, PhD, FESC
Associate Professor
Dept. of Clinical Sciences
Lund University
Senior Consultant, Syncope Unit
Dept. of Cardiology
Skåne University Hospital
Karolinksa University Hospital
Stockholm, Sweden

Roy Freeman, MD
Professor of Neurology
Harvard Medical School
Director, Center for Autonomic and Peripheral Nerve Disorders
Beth Israel Deaconess Medical Center
Boston, MA, USA

Robert S. Sheldon, MD, PhD
Professor of Cardiac Sciences, Medicine and Medical Genetics
Libin Cardiovascular Institute of Alberta
University of Calgary
Calgary, Alberta, Canada

Steven Vernino, MD, PhD
Distinguished Teacher Professor & Vice Chair
Department of Neurology & Neurotherapeutics
Director, Autonomic Laboratory
University of Texas Southwestern
Dallas, TX, USA