

Issues in Diagnostic Medicine and Legal Implications for Patients with Rarely Heard of Disorders: Dysautonomia—An Overview

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Introduction

Dysautonomia is an umbrella term used to describe a malfunction of the autonomic nervous system.¹ Disorders of the autonomic nervous system refer to “disorders of the part of the nervous system that regulates involuntary action, as the intestines, heart, and the glands.”² In other words, the autonomic nervous system controls everything we do not think about. Despite the number of individuals with dysautonomia, the condition is not widely understood or recognized in the medical, legal, and payer communities. This article provides an overview of dysautonomia and its emergence as a recognizable category of disease. It also considers some of the medical and legal issues individuals with dysautonomia face because of the persistent lack of awareness of the condition.

What’s in a Name?

If you have never heard of dysautonomia, you are not alone, which is somewhat surprising considering how many patients nationally and worldwide are affected by one or more conditions on the dysautonomia spectrum. While some forms of dysautonomia are considered rare, other forms are quite common.

In 1993, a team of neurologists first coined the term “Postural Orthostatic Tachycardia Syndrome” (POTS),³ which is one of the most statistically prevalent syndromes under the dysautonomia umbrella. POTS is not a new ailment. Several different names have been used to describe the same syndrome for over 150 years,⁴ including DaCosta’s Syndrome, Soldier’s Heart, Irritable Heart, and Mitral Valve Prolapse Syndrome. POTS is not rare, but is rarely heard of. Estimates indicate that about 500,000 to one million individuals in the United States have POTS.⁵ Given the general lack of awareness of this syndrome, however, these numbers are likely much higher. The Mayo Clinic estimates that approximately one out of every 100 teenagers develops POTS before adulthood.⁶ POTS is most often seen in females between the ages of 15 and 50. Males can also develop POTS, but approximately 75-80% of individuals with POTS are female.⁷

The syndrome itself is characterized by excessive tachycardia upon standing.⁸ Approximately 25% of those diagnosed with POTS are unable to work.⁹ Physicians familiar with treating POTS have compared the impairment seen in POTS patients to the impairment seen in “chronic obstructive pulmonary disease and congestive heart failure . . .”¹⁰

What’s in a Number?

The ICD-9 is the tool providers must use to categorize a patient’s diagnosis or diagnoses. This system of classification aids in the government’s ability to gather statistics related to disease and mortality. This uniform system of categorizing patients also provides the grounds for provider reimbursement from commercial and governmental payers.¹¹

Under “Disorders of the autonomic nervous system,” at the end of a rather short list, comes diagnostic code 337.9—“Unspecified disorder of the autonomic nervous system.”¹² This is the catch-all category where tens of thousands of dysautonomia patients land, presumably, if at all. While, for example, Epilepsy is broken down into extremely detailed sets and subsets in the ICD-9, autonomic disorders are not. Within the POTS diagnosis alone are several distinct categories: Neuropathic POTS, Central Hyperadrenergic POTS, Norepinephrine Transporter Deficiency, Mast Cell Activation, Hypovolemia and Blood Regulation type.¹³

A one-size-fits-all diagnostic category for such a significant number of patients is inefficient and leads to inaccuracies in accounting for those afflicted. If patient A is a 62-year-old male who has been diagnosed with Pure Autonomic Failure (a rarer subset of dysautonomia), patient B is a 17-year-old teenage female diagnosed with Neurocardiogenic Syncope (a more common form of dysautonomia), and patient C is a 35-year-old female diagnosed with POTS, all three patients should not be categorized under the same diagnostic code. However, no code exists at the present time to better differentiate and accurately count the dysautonomia population.

Reasonable Accommodations

Patients with dysautonomia may have trouble receiving accommodations legally mandated by the Americans with Disability Act in both educational and employment settings because of the lack of recognition of the condition in the general community. “New” or rarely heard of” diagnoses, particularly for syndromes where pathology is highly complex like dysautonomia, also present challenges for employers or academic institutions in crafting reasonable accommodations. How does a company create accommodations for its employees, and an educational institution create accommodations for its students, when a disease or syndrome is not understood well—even by patients, providers, and attorneys?



Advances in diagnostic medicine have increased the ability of medical professionals to determine the presence of previously inchoate conditions. Diagnostic tools are important for employers and schools to better understand what reasonable accommodations should be made for an employee or student with dysautonomia. Human resource professionals and academic support staff should take affirmative steps to learn more about dysautonomia, as statistics indicate a great deal of patients with dysautonomia work, or go to school, or both.

Once an employer or academic institution recognizes a person's disability as a subset of dysautonomia, it should be easier to craft reasonable accommodations. Regardless of whether dysautonomias are objectively verifiable, which most are, care must be given to ensure accommodations are followed based on the physician's recommendation.

Social Security Disability Insurance

Depending on the degree of disability, some patients with dysautonomia may be unable to work. But applying for Social Security Disability Insurance (SSDI) can be difficult for these individuals because most patients with dysautonomia do not "look" sick and in fact appear young and healthy to the outside world.

The Social Security Administration, disability lawyers, and judges deal with recognizable illnesses every day: diabetes, bipolar disorder, depression. But likely few in the business of government benefits, including attorneys hired to help clients obtain them, have heard of disorders like "Postural Orthostatic Tachycardia Syndrome," "Neurocardiogenic Syncope," "Pure Autonomic Failure," or "Autoimmune Autonomic Ganglionopathy."

SSDI attorneys should familiarize themselves with the growing body of medical research about dysautonomias to ensure they can be effective advocates for their clients. While many patients already have extensive documentation of their diagnosis, a competent specialist, generally an autonomic neurologist and electrophysiologist or cardiologist, is imperative for obtaining a favorable benefits determination.

Defining a Standard of Care

Although dysautonomia awareness has increased incrementally in the past decade, case law involving this illness is sparse. One of the only medical malpractice cases ever published regarding dysautonomia involved a young law student who fell ill and was injured in 1993, the first year the disease was renamed. While procedural technicalities ultimately led to a decision in the provider's favor, the case history tells the story of a very sick law student who was misdiagnosed repeatedly.¹⁴ Without extensive case law, dysautonomia's standard of care must be based on guidelines set forth by medical researchers who specialize in dysautonomia.

Conclusion

While some types of dysautonomias are statistically rare, many are not, and are simply rarely heard of. Employers and academic institutions need a better understanding of dysautonomia so they are prepared to provide reasonable accommodations for employees and students with one of the conditions under the dysautonomia spectrum. SSDI attorneys should increase their awareness of dysautonomia so they can be more effective advocates for those clients whose dysautonomia has rendered them unable to work. Providers must put themselves in a position where they are prepared to properly recognize autonomic dysfunction, or refer the patient to a provider with expertise in this area. Raising awareness is key to helping patients with conditions like dysautonomia address the unique challenges they face.

The "Reasonable Accommodations" section of this article was written in conjunction with Steven Silva of Chapman Law Firm PC.

About the Author



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Endnotes

- 1 National Institutes of Health, National Institute of Neurological Disorders and Stroke, Dysautonomia Information Page, *available at* www.ninds.nih.gov/disorders/dysautonomia/dysautonomia.htm (last visited Sept. 3, 2013).
- 2 INTERNATIONAL CLASSIFICATION OF DISEASES, 9TH REVISION, CLINICAL MODIFICATION, SIXTH EDITION 339 (Office ed., Practice Management Information Corporation 2012).
- 3 Phillip A. Low & Ronald Schondorf, *Idiopathic postural orthostatic tachycardia syndrome: An attenuated form of acute pandysautonomia?*, 43 NEUROLOGY 132 (1993).
- 4 Charles F. Wooley, *Where are the diseases of yesteryear? DaCosta's Syndrome, Soldiers Heart, the Effort Syndrome, Neurocirculatory Asthenia—And the Mitral Valve Prolapse Syndrome*, 53 CIRCULATION 749, 749-750 (1976).
- 5 David Robertson, *The Epidemic of Orthostatic Tachycardia and Orthostatic Intolerance*, 317 AM. J. MED. SCIENCES 75, 75-77 (1999).
- 6 Postural Orthostatic Tachycardia Syndrome (Mayo Clinic Podcast Apr. 23, 2008) (on file with author).
- 7 National Institutes of Health, National Institute of Neurological Disorders and Stroke, Postural Tachycardia Syndrome Information Page, *available at* www.ninds.nih.gov/disorders/postural_tachycardia_syndrome/postural_tachycardia_syndrome.htm (last visited Sept. 3, 2013).
- 8 Blair P. Grubb, *Postural Tachycardia Syndrome*, 117 CIRCULATION 2814 (2008).
- 9 *Id.*
- 10 *Id.*
- 11 INTERNATIONAL CLASSIFICATION OF DISEASES, 9TH REVISION, CLINICAL MODIFICATION, SIXTH EDITION 13 (Office ed., Practice Management Information Corporation 2012).
- 12 *Id.* at 339-340.
- 13 Satish R. Raj, *Clinician Update: Postural Tachycardia Syndrome (POTS)*, 127 CIRCULATION 2336, 2338 (2013).
- 14 *Goldstein v. Reiskin*, 942 N.E. 2d 208 (Mass. App. Ct. 2011).