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Take a Stand

A neurologic disorder known as POTS causes dizziness and fainting—and frustration, due to lack of awareness and inadequate treatment.

BY AMY PATUREL

When 20-year-old Jenna Bock left her home in Oak Park, CA, to attend college in Wisconsin, she had visions of freedom and fraternity parties. Those dreams evaporated in November of her freshman year when she was hospitalized with the flu. A few weeks later, after watching a movie in her friend's dorm room, she stood up and promptly passed out.

"I thought it was a fluke, but a couple of days later, it happened again. By January, I was fainting up to 10 times a day," says Bock, a busy college student who was active in campus government and worked as an intern for a counseling service.

Bock visited dozens of physicians and spent weeks hospitalized with various ailments, yet no one could identify the cause of her symptoms. Some doctors even claimed she was making it up. Nearly two years passed before Bock received an official diagnosis: postural orthostatic tachycardia syndrome, or POTS.

Bock's fainting spells are one of the hallmarks of POTS. Another is trouble standing up, primarily because not enough blood returns to the heart upon standing, which causes a rapid increase in heart rate called tachycardia. In fact, the diagnostic

criteria for this syndrome include an increase in heart rate of more than 30 beats per minute (40 beats per minute for children) or a heart rate of 120 beats per minute within 10 minutes of standing. Other symptoms include chronic fatigue, "brain fog," and pain. (To learn more about how POTS is diagnosed, see "Do You Have POTS?" on page 46.)

To confirm her diagnosis, doctors monitored Bock's heart rate and blood pressure while moving her from a horizontal to a vertical position on a special tilting table. Unfortunately, once Bock tested positive during this "tilt table test," doctors still didn't know how to help her. (For information about how POTS is treated, see "5 Ways to Alleviate POTS" on page 45.)

HIDING IN PLAIN SIGHT

Even though Mayo Clinic researchers coined the term "POTS" in 1993, many physicians have never seen the condition in their practice, or at least have not identified it accurately, in part because there are so many accompanying symptoms that can be mistaken for something else.

Despite the lack of awareness, POTS isn't rare, explains Svetlana Blitshteyn, MD, clinical assistant professor of neuro-

logy at the University at Buffalo School of Medicine and director of the Dysautonomia Clinic in Buffalo, NY. "It's estimated that between 1 and 3 million Americans are affected by the syndrome." POTS can strike at any age, but it primarily affects women between the ages of 15 and 50 who, like Bock, are in the prime of their lives. These active, vibrant, productive members of society are incapacitated, sometimes abruptly and for long periods of time.

Now, with a greater understanding of POTS and its many co-existing conditions, scientists are working to unravel this mysterious illness.

STANDING GETS COMPLICATED

For most of us, standing up is simple. Blood pools in the lower body and pelvic area as we rise to our feet and our heart rate increases slightly as part of a reflexive adjustment.

In people with POTS, however, this regulatory system is out of whack. Blood doesn't flow where it should, forcing the heart to work harder, explains Thomas C. Chelmsky, MD, a professor of neurology at the Medical College of Wisconsin in Milwaukee, who is a Fellow



“ There’s a lot of misconception and a lack of awareness about the diagnosis. POTS is a syndrome that we may not understand fully, but it is a real disorder. ”

—SVETLANA BLITSHEYN, MD

of the American Academy of Neurology (FAAN). Since the heart isn’t getting enough blood, patients feel light-headed, dizzy, and out of breath.

Debilitating fatigue is another common symptom of this syndrome, although its cause remains a mystery. In fact, POTS can be as disabling as congestive heart failure (CHF) and chronic obstructive pulmonary disease (COPD), according to a study from the Mayo Clinic.

But unlike people with CHF, COPD, or kidney disease, people with POTS often look healthy, baffling physicians

who examine and re-examine them but can’t find the culprit behind the symptoms. In many cases, POTS patients are misdiagnosed with a psychiatric illness—and get stuck chasing referrals from one mental health professional to another as a result.

“There’s a lot of misconception and a lack of awareness about the diagnosis, and for some physicians, POTS has become a wastebasket diagnosis,” says Dr. Blitshteyn. “It’s not. POTS is a syndrome that we may not understand fully, but it is a real disorder.”

A SYNDROME WITH LOTS OF SYMPTOMS

To complicate matters further, patients with POTS often also experience migraine, fibromyalgia, and chronic fatigue. About 50 percent of patients with POTS have small fiber neuropathy, a condition that affects the sensory nerves, causing numbness or burning sensations. In addition, researchers have used advanced magnetic resonance imaging technology to uncover structural abnormalities in the brains of some POTS patients in areas associated with control of the autonomic nervous system. However, says David S. Goldstein, MD, PhD, founder

5 Ways to Alleviate POTS

Because so many symptoms are associated with POTS, treatment is tricky. Doctors have to determine which symptoms are due to POTS and which are pre- or co-existing. But many cases respond to simple lifestyle changes, says Paola Sandroni, MD, PhD, a professor of neurology at the Mayo Clinic in Rochester, NY.

1 Say Yes to Sodium. Salt helps the body retain water, which in turn increases blood volume. “With salt, we’re trying to expand blood volume and simultaneously constrict blood vessels, so we can get more blood up to the heart and brain,” explains Thomas C. Chelimsky, MD, a professor of neurology at the Medical College of Wisconsin in Milwaukee.

This may not work for all POTS patients, says David S. Goldstein, MD, PhD, founder and director of the clinical neurocardiology section in the division of intramural research of the National Institute of Neurological Disorders and Stroke, who believes another potential area of research is whether the kidneys of people with POTS are less efficient in retaining water and salt than those of healthy people.

2 Fill Up on Fluids. Drinking more fluids, especially in combination with salt, helps expand blood volume and increase blood flow. Most doctors recommend two to three liters per day of hydrating fluids such as vegetable or tomato juice, coconut water, decaffeinated tea with salt, or chicken broth. “POTS patients often have to drink water very frequently during the day or they feel worse,” says Dr. Goldstein.

3 Move Your body. Exercise helps POTS patients stay on their feet by maintaining muscle strength and preventing blood from pooling in the lower extremities. For those who can’t tolerate upright exercise, activities such as recumbent rowing, cycling, or swimming may help with reconditioning until they’re ready for more intense aerobic exercise.

4 Slip on Some Stockings. Compression garments, such as abdominal binders and waist-high compression stockings with extra firm pressure (look for “30-40 mm Hg” on the label) help enhance blood flow to the heart and can reduce the amount of blood that pools in the lower extremities upon standing.

5 Consider Medication. The US Food and Drug Administration has not approved any drugs for POTS, so prescriptions are based on trial and error—plus there are side effects, which can limit their use, Mayo researchers report. Commonly prescribed drugs include fludrocortisone to expand blood volume; beta-blockers to slow rapid heart rate; midodrine to constrict the blood vessels and help coax blood flow back to the heart; antidepressants to re-regulate the autonomic nervous system and increase blood pressure; stimulants such as methylphenidate and amphetamine/dextroamphetamine to reduce fatigue, constrict blood vessels, and increase blood pressure; pyridostigmine to reduce muscle weakness by inhibiting the breakdown of acetylcholine, a chemical that helps nerves and muscles communicate; and octreotide to constrict the blood vessels.

“Something in the brain is making people with POTS feel like they don't have adequate blood supply when they stand up.”

—THOMAS C. CHELIMSKY, MD

and director of the clinical neurocardiology section in the division of intramural research of the National Institute of Neurological Disorders and Stroke (NINDS) at the National Institutes of Health, there's no evidence that POTS causes these structural changes, or vice versa. “It could simply be guilt by association,” he says.

These various symptoms suggest that POTS may also be a central nervous system problem rather than a physiologic one in some patients, says Dr. Chelimsky. “Something in the brain is making people feel like they don't have adequate blood supply when they stand up. That feeds back into the body and produces this feedback loop where the brain constantly senses inadequate supply,” he says. “So even if we find an abnormality in the body, it could still be driven by the brain.”

Still, there's significant disagreement in the scientific community about what drives POTS. Some experts believe it's a disorder of both the mind and the body. Other researchers cite a neuropathic process. Still others claim it's a transient disorder that stems from deconditioning (physiological changes that occur after a period of inactivity or bed rest). Even simple genetics may contribute to the syndrome. A 2007 study in the *Mayo Clinic Proceedings* reported that 12.5 percent of 152 people with POTS had a family history of difficulty upon standing.

IT COULD BE AN AUTOIMMUNE DISORDER

Another theory gaining traction is that POTS triggers an immune response in which the body begins attacking the pieces of its machinery that make blood vessels constrict. POTS may co-exist with autoimmune disorders such as celiac disease, rheumatoid arthritis, and Sjögren's syndrome, which affects the glands that produce tears and saliva, or POTS may itself have an autoimmune basis, researchers suggest.

DO YOU HAVE POTS?

POTS has many signs and symptoms, and the diagnostic criteria are not at all straightforward, says Gary S. Gronseth, MD, professor and vice chair of neurology at the University of Kansas, who is a Fellow of the American Academy of Neurology (FAAN) and a member of the *Neurology Now* editorial advisory board.

“Upon standing, many people have a rise in heart rate. In fact, up to 40 percent of 20-year-olds experience an increase in heart rate of 30 beats or more,” he says. So even if you have POTS-like symptoms, an increase in standing heart rate does not automatically mean that your symptoms are caused by POTS. “Without better criteria, it is difficult to be confident that POTS is the correct diagnosis.”

That said, many patients present with any or all of the following symptoms, usually after a viral illness, surgery, concussion, or other trauma.

Either way, there's significant evidence to suggest that a body affected by POTS attacks itself. For example, a 2014 Mayo Clinic study published in *Clinical Autonomic Research* found that antibodies—proteins that detect and defend against harmful antigens such as bacteria and viruses—targeting specific organs were more common among POTS patients than among those without the syndrome. Researchers from the Children's Heart Institute in Fairfax, VA, noted elevated infection-fighting cells among patients with POTS, suggesting that the immune systems of POTS patients are constantly overwhelmed.

Despite these findings, there's no published evidence that autoimmune treatments help POTS patients, cautions Dr. Goldstein.

Symptoms related to standing:

- ▶ Dizziness
- ▶ Lightheadedness
- ▶ Palpitations
- ▶ Near fainting (also called pre-syncope) when standing

Symptoms unrelated to standing:

- ▶ Headaches
- ▶ Fatigue
- ▶ Nausea
- ▶ Sleep disturbance
- ▶ Diarrhea
- ▶ Constipation
- ▶ Change in perspiration
- ▶ Temperature control problems
- ▶ Fibromyalgia
- ▶ Temporomandibular joint disorder
- ▶ Difficulty concentrating (“brain fog”)

PROGNOSIS IS UNPREDICTABLE

Statistics on long-term outcomes for POTS patients are evolving. A 2009 study published in the *Mayo Clinic Proceedings* suggested that about 80 percent of patients with POTS experience an improvement in symptoms within five years, and that symptoms almost completely resolve in around 60 percent of patients.

However, “clinically, we're not seeing those numbers,” says Dr. Blitshteyn, citing a larger study in 2013 from the Mayo Clinic, presented at the 24th International Symposium on the Autonomic Nervous System, with updated statistics. “In that study, only about 18.2 percent of patients reported complete resolution of symptoms, while 52.8 percent reported improved but persistent symptoms two



UPBEAT Jenna Bock at Froedtert Hospital in Milwaukee, WI, in July 2014 undergoing diagnostic testing for POTS.

to 10 years after the diagnosis." In that study, men were more likely to recover than women.

In general, the research points to a variable disease course and a prognosis that can change and evolve over time, depending on what triggered the syndrome. "Some patients will recover and will function at a normal or near-normal level; for others, POTS will remain a chronic or recurrent disorder," says Dr. Blitshteyn.

While Jenna Bock isn't back to normal, POTS hasn't derailed her college dreams. By scheduling her days with plenty of down time (she still faints up to five times each day), she even made the

Dean's List, she says. "Being sick has taught me to live in the present and take each day for what it is instead of what it should be." NN

Even though Mayo Clinic researchers coined the term "POTS" in 1993, many physicians have **never seen the condition** in their practice.

FOR MORE INFORMATION:

- ▶ Dysautonomia International: dysautonomiainternational.org/POTS
- ▶ Autonomic Disorders Video Library: vimeo.com/dysautonomia
- ▶ Vanderbilt University Autonomic Dysfunction Center: bit.ly/VanderbiltADC
- ▶ Children's Hospital of Philadelphia POTS Program: bit.ly/PhiladelphiaPOTS

WEB EXTRAS:

- ▶ For more on POTS, go to bit.ly/NN-POTS

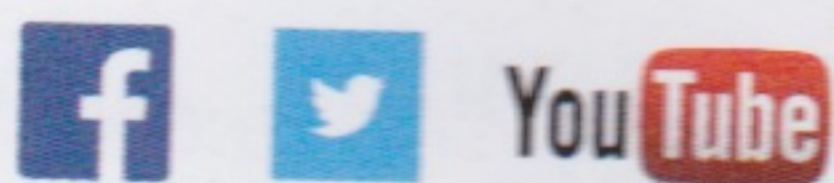
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