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Review

The patient perspective: What postural orthostatic tachycardia syndrome patients want physicians to know

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ABSTRACT

Diagnosing and treating postural orthostatic tachycardia syndrome (POTS) can be a frustrating experience for patients and physicians alike. Experienced patient leaders solicited input from the large online POTS community to identify patient suggestions and concerns, with the goal of improving the patient-physician relationship and outcomes in POTS. This review article offers practical tips to improve POTS patient care and links to credible resources for your patients. The authors emphasize the urgent need for improved physician education, a tailored treatment approach, and expanded research efforts.

1. Introduction

Diagnosing and treating postural orthostatic tachycardia syndrome (POTS) can be a frustrating experience for patients and physicians alike. Physicians may not feel comfortable managing POTS due to a lack of training on the topic. Diagnostic delays, frequent misdiagnosis, being blamed for their symptoms and a plethora of misleading information on the web leaves many POTS patients feeling scared and confused, and this can impact the way patients interact with physicians.

The authors of this article serve on the Board of Directors (LS) and Patient Advisory Board (JC and IB) of Dysautonomia International, a non-profit organization that advocates for individuals living with POTS and other autonomic disorders. To prepare for this article, we asked patients and caregivers to list five things they wanted physicians to know about POTS. We posed the question on Dysautonomia International's Facebook page, which has over 44,000 followers. Many individuals responded with thoughtful answers, which we summarize below.

2. Understanding the POTS patient experience

2.1. POTS is common, POTS experts are rare

POTS is estimated to impact 500,000 to 3,000,000 Americans (Mar and Raj, 2014), making it more common than well-known medical conditions like multiple sclerosis, which impacts 400,000 Americans (Dilokthornsakul et al., 2016), yet few medical professionals receive formal training on POTS. As of this publication, there are only five academic centers that offer a fellowship in Autonomic Disorders (UCNS, 2018a), and there are only 37 physicians Board Certified in Autonomic Disorders (UCNS, 2018b).

As a result, POTS patients experience an average diagnostic delay of over four years, and see an average of seven physicians prior to POTS diagnosis, with one-quarter (23%) seeing 10 or more physicians (Raj et al., 2016). Prior to POTS diagnosis, 76% of patients are misdiagnosed (Stiles et al., 2017a).

After diagnosis, the majority (73%) of POTS patients encounter physicians who have not heard of POTS (Stiles et al., 2017a). Approximately 50% of POTS patients have to travel more than 100 miles from home to receive POTS related specialty care, and 21% have traveled over 500 miles (Stiles and Ross, 2014).

2.2. A typical patient narrative

I was a happy and healthy teenager until one day, at age 13, I woke up not feeling well. Every time I stood up I felt lightheaded. I also had joint pain and terrible stomach pain, especially when I would eat. I was told I had Lyme and I'd be fine after I took the normal course of antibiotics. It only got worse from there. Suddenly, I went from being a volleyball player with great grades, to barely being able to get out of bed.

I saw 10 doctors over the next few years trying to figure out what was wrong. I was told I had an eating disorder. I didn't. I was told I was just anxious. I wasn't. Doctors gave me numerous medications to try. Nothing worked. Some doctors gave up on me. I began to feel hopeless.

I kept declining, but no one was taking me seriously. After four years I traveled across the country to Mayo Clinic to find answers. I was diagnosed with POTS and all of my symptoms finally made sense. I tried all of the standard POTS treatments, but this only helped a little bit. I

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completed the Mayo Clinic Pain Rehabilitation program, but I was still very sick. A few years later, another Mayo Clinic doctor diagnosed me with Sjögren's syndrome, which we think may be the root cause of my POTS.

At 21 I still struggle with symptoms every day, but I am thankful for the tailored treatment plan and resources available now that I have an accurate diagnosis. I want physicians to learn about POTS and related disorders so other people don't have to suffer for years without a diagnosis like I did.

This may seem like an extreme example of prolonged misdiagnosis, but this is the typical diagnostic journey that POTS patients endure, both in adolescent and adult onset patients.

2.3. "Post-traumatic misdiagnosis disorder"

For many POTS patients and their families, the diagnostic process is a traumatic experience, fraught with misdiagnoses, unnecessary tests, inappropriate treatments, accusations made against the patient or their parents, large medical bills, long periods of missed school or work, and prolonged suffering because the patient's actual diagnosis is not being treated.

When POTS patients ask physicians for help with their symptoms and are repeatedly told nothing is wrong, or some version of "it's all in your head," this can lead to patients feeling rejected or disbelieved by physicians. As a result, some patients develop a mistrust of or even a hostile attitude towards the entire medical profession. Some POTS patients even report falling into depression because physicians have treated them so poorly. While you will not find it in the *Diagnostic and Statistical Manual of Mental Disorders*, we have coined the term "post-traumatic misdiagnosis disorder" to refer to this phenomenon.

2.4. POTS symptoms can be frightening

It is important for physicians to understand that POTS symptoms can be truly frightening. POTS patients frequently experience shortness of breath, chest pains, and palpitations. Lay people are taught, if you're having chest pain and shortness of breath, it might be a heart attack – call the ambulance! Thus, when POTS patients experience these symptoms, it is natural for them to worry. Is this a heart attack? Should I go to the emergency room? Patient education from a trusted physician is needed to provide reassurance with compassion and understanding. Please do not assume your patient is catastrophizing or attention seeking. They may be genuinely afraid of the bodily sensations they are experiencing, as anyone would be.

In our experience, it takes about two years after diagnosis for a POTS patient to get a solid understanding of how POTS impacts their body, and how to best manage their symptoms. This timeframe can be accelerated when a trusted physician takes the time to explain the basic physiology of POTS, and ensures the patient has access to high-quality information on the many non-pharmacological management approaches they will have to incorporate into their daily routines.

2.5. Living with an invisible illness

POTS patients tend to be young people with a normal physical appearance, although, a skilled physician will know where to look for visible signs associated with POTS. Swollen lower limbs after upright activity, acrocyanotic legs, prolonged capillary refill when pressure is applied to the skin, livedo reticularis, Bier spots, and flushing which tends to occur on the upper half of the body may be present in some patients (Huang et al., 2016; Huang and Hohler, 2015; Bessis et al., 2016). See Fig. 1.

The mostly invisible nature of POTS is both a blessing and a curse. Looking "fine" may help patients avoid the judgement of those who discriminate against individuals with visible disabilities. But looking "fine" when a patient cannot participate in all of the activities they would like to, can result in judgement, disbelief and shaming from peers or family members who cast doubt on their POTS diagnosis. This judgement can be especially difficult for adolescent patients to cope with. See Fig. 2.

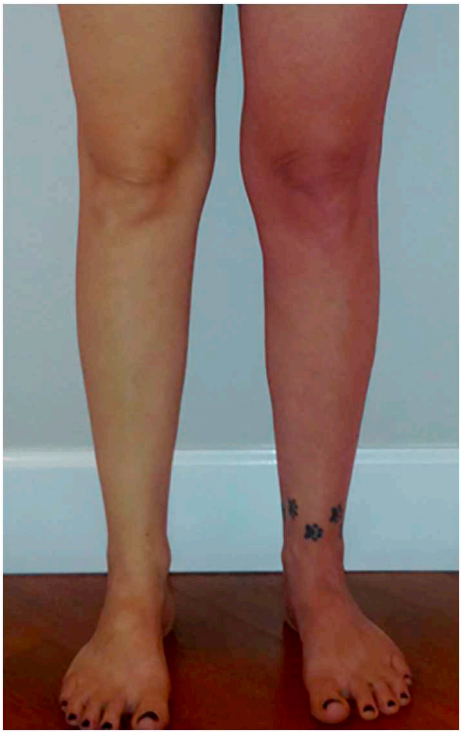

Fig. 1. Both of this POTS patient's legs become acrocyanotic within 5 min of standing. The acrocyanosis dissipated in the right leg after elevating it on a counter for 3 min immediately prior to this photo.

Fig. 2. Looking fine doesn't mean a patient feels fine. This 14-year-old POTS patient's symptoms wax and wane. Some days he can play football and some days he can barely stand. It is hard for his peers to understand this.

3. How to help your POTS patients

In addition to the medical guidance contained throughout this issue, there are simple practical things you can do to help your patients.

3.1. Validate your patient

Validating the patient after they have been repeatedly let down by other doctors is the best way to build trust. “I believe you” or “POTS is real, it's not in your head” are simple statements that can have a powerful positive impact on your patient's life.

3.2. Set realistic expectations

In most cases finding the ideal treatment plan will require trial and error. Acknowledge this at the first office visit to set realistic expectations for the patient and their family.

Do not oversell the benefits of exercise by telling patients it will cure them. For most patients, exercise is not a cure, it is an important treatment tool that can help manage symptoms. It is also helpful to explain to your patient that beginning a new exercise plan may exacerbate their symptoms in the short term, but it is important to stick with it at a pace they can tolerate.

3.3. Provide clear advice to your patient

Patients are easily overwhelmed by the influx of new information they will receive when they are newly diagnosed. Provide recommendations in writing that patients can review again at home. Provide specific recommendations on salt, fluids, and exercise. See Table 1. Advise your patient on the importance of consuming a *healthy* high-salt diet and avoid recommending sugary sports drinks. Provide clear guidance on when to go to the emergency room, when to stay home, and when to call the doctor's office. Provide prescriptions for compression stockings, as many insurers will cover them as a durable medical good.

4. Addressing misconceptions about POTS

There are a number of misconceptions about POTS that impede patient care.

4.1. POTS is more than tachycardia

POTS patients experience symptoms in all domains of autonomic function evaluated by the COMPASS-31 questionnaire (Rea et al., 2017), as well as symptoms suggestive of sensory dysfunction (Stiles et al., 2017b), and cognitive impairment, both orthostatic and non-orthostatic (Ross et al., 2013). Looking at the whole patient is essential to improving your patient's health. See Fig. 3.

4.2. Deconditioning is usually a consequence of POTS, not a cause

While the role of deconditioning in POTS has been the subject of debate amongst researchers (Fu et al., 2010; Pianosi et al., 2016; Blitshteyn and Fries, 2016; Oldham et al., 2016; Parsaik et al., 2012), what is lost in this debate is the fact that most POTS patients were active young people before developing POTS. Notable cases include an Olympic athlete who suddenly developed POTS after a concussion (Bowe, 2018), and a Paralympic athlete who acutely developed POTS after an infection (Stiles, 2018). Recent data suggests that 65% of POTS patients regularly exercised (defined as 3–4 days per week) prior to being diagnosed with POTS, thus it seems unlikely that deconditioning is the cause of POTS in these patients (Stiles et al., 2018). In our experience managing large online support groups, deconditioning is more likely to play a significant role when the patient experiences a prolonged diagnostic delay, without proper treatment for their orthostatic intolerance (Table 2).

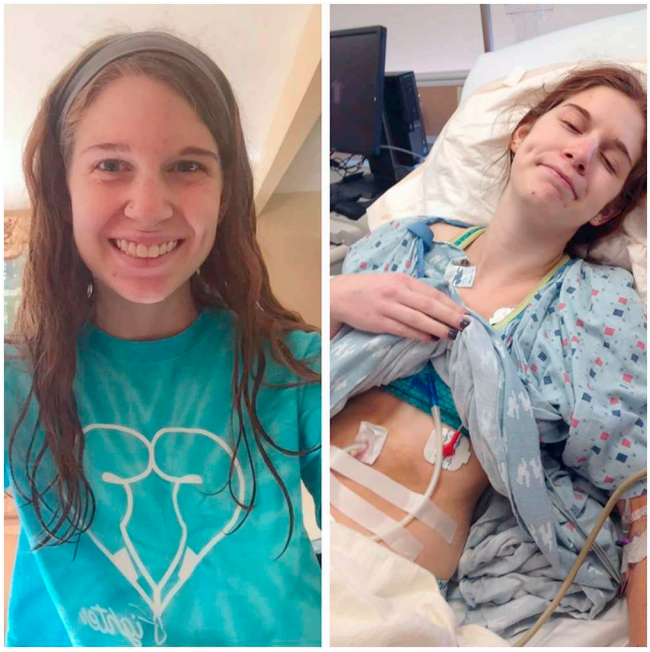

Fig. 3. The autonomic dysfunction seen in POTS goes beyond orthostatic intolerance, and some patients can experience significant gastrointestinal dysmotility. This is the same patient on a good day and a bad day.

4.3. Everyone doesn't grow out of it

Some physicians confidently assure their adolescent POTS patients that they will “grow out of it.” The longest longitudinal study to date found that only 19% of adolescent onset POTS patients self-reported “symptoms completely resolved” with a mean of 5.4 years after diagnosis, and a mean age at the time of survey completion of 21.8 years (Bhatia et al., 2016). While there is reason to be hopeful, because 86% of adolescent onset POTS patients reported *some* degree of improvement over time (Bhatia et al., 2016), there is insufficient evidence to state that all or even the majority of adolescent onset POTS patients will “grow out of it.” Longer longitudinal studies with clinical evaluation on follow-up are needed to provide a better understanding of the natural history of POTS.

Table 1
Quick links.

POTS-friendly exercise tips	www.dysautonomiainternational.org/exercise
Healthy high-salt diet tips	www.dysautonomiainternational.org/salt
Compression stocking tips	www.dysautonomiainternational.org/stockings
State and national support groups	www.dysautonomiainternational.org/support
Educational videos from experts	www.dysautonomiainternational.org/videos

Table 2
Tips for discussing exercise.

- Don't give the impression that you are blaming the patient for their exercise intolerance.
- Acknowledge that anyone who has an orthostatic disorder would have difficulty exercising.
- Suggest that you will work together to gradually improve the patient's exercise capacity.
- Explain the physiological benefits of regular exercise, particularly its ability to increase blood volume, which is important in POTS.

Setting unrealistic expectations for young POTS patients can be harmful. Patients may feel like failures if they go through their teens being told they will outgrow POTS, only to find themselves still sick in their late 20s. Learning to drive, dating, planning for college and other teenage milestones should not be postponed while patients wait to "grow out of it," since there is a chance they may not.

When POTS patients ask about their prognosis, let them know that POTS is not fatal, most patients see *some* improvement over time, and some fully recover. This is realistic and reflects the limited data we have on the long-term prognosis in POTS.

4.4. Doctor shopping isn't always a negative

"Doctor shopping" is a derogatory phrase used to describe individuals who surreptitiously pursue care from numerous physicians seeking addictive medications or engaging in attention-seeking behavior. However, not every patient who has seen an army of physicians is drug or attention seeking.

As noted above, patients see an average of seven physicians prior to POTS diagnosis (Raj et al., 2016). After diagnosis, most patients are referred to numerous specialists to evaluate and manage the various symptoms and comorbidities seen in POTS.

In our experience, when POTS patients seek multiple second opinions it is usually because the patient feels like their prior physician(s) did not do a thorough job looking for underlying conditions that may be contributing to their symptoms. It is no wonder patients feel this way, given the very long list of potentially contributing factors that have been described in the published POTS literature, including small fiber neuropathy (Peltier et al., 2010), Ehlers-Danlos syndrome (Wallman et al., 2014), a long list of autoimmune diseases (Vernino and Stiles, 2018), vitamin deficiencies (Oner et al., 2014; Blitshteyn, 2017), mitochondrial disorders (Kanjwal et al., 2010), Lyme disease (Kanjwal et al., 2011; Noyes and Kluger, 2015), vascular compression syndromes (Petrosyan et al., 2015; Kaymak et al., 2004), cerebral spinal fluid leaks (Aggarwal and Carroll, 2017), Chiari malformation (Prilipko et al., 2005; Pasupuleti and Vedre, 2005), and more. If a patient leaves your office thinking you have not diligently considered the possibility of contributing factors, patients who are motivated to get better and have the means to do so will often seek advice from another physician. Rather than viewing POTS patients who have seen "too many physicians" negatively, recognize that they are pursuing so many second opinions because they are desperate to feel better.

5. Working towards systemic policy change

5.1. Increasing physician education and access to care

Medical schools and professional societies must increase efforts to educate physicians on POTS and other autonomic disorders, with the goal of reducing diagnostic delays and improving the quality-of-care received once patients are diagnosed. This may also shorten wait times to see a POTS specialist, which currently range from 6 to 36 months.

5.2. Improving patient care

Patients need the POTS research community to develop an evidence-based diagnostic algorithm that will help physicians rule out the most common and most serious comorbidities seen in POTS patients. Dr. Brent Goodman's thoughtful article in this issue provides a good starting point (see Goodman, Evaluation of Postural Tachycardia Syndrome, in this issue).

5.3. Expanding research efforts

Patients need POTS research focused on understanding the mechanisms that lead to POTS and identifying more effective treatments. This requires a substantial investment in research. While POTS research had its origins in neurology and electrophysiology, it is clear that we also need to engage researchers from other disciplines to unravel the complexities of POTS.

6. Conclusion

POTS is a common autonomic disorder best managed by a physician who recognizes the heterogeneity of the syndrome and the need for a tailored treatment approach. The lack of medical training on POTS leads to difficulties in obtaining care and negative experiences for many patients, which can influence the way POTS patients interact with the medical profession. Expanded clinical education can likely reduce diagnostic delays, improve access to care, and improve outcomes in POTS. Additional research is urgently needed to provide physicians with clearer guidance on diagnostic and treatment approaches for this large, underserved patient population.

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