Postural Orthostatic Tachycardia Syndrome (POTS): A Frequently Missed Diagnosis

Aubrey R. George
Brigham Young University - Provo, aubrey.r.george17@gmail.com

Blaine A. Winters
Brigham Young University - Provo, Blaine-Winters@byu.edu

Follow this and additional works at: https://scholarsarchive.byu.edu/studentpub

Part of the Nursing Commons

BYU ScholarsArchive Citation
https://scholarsarchive.byu.edu/studentpub/319

This Peer-Reviewed Article is brought to you for free and open access by BYU ScholarsArchive. It has been accepted for inclusion in Student Works by an authorized administrator of BYU ScholarsArchive. For more information, please contact scholarsarchive@byu.edu, ellen_amatangelo@byu.edu.
Postural Orthostatic Tachycardia Syndrome (POTS):
A Frequently Missed Diagnosis

Aubrey R. George

A scholarly paper submitted to the faculty of
Brigham Young University
in partial fulfillment of the requirements for the degree of

Master of Science

Blaine Winters, Chair

College of Nursing
Brigham Young University

Copyright © 2021 Aubrey R. George
All Rights Reserved
ABSTRACT

Postural Orthostatic Tachycardia Syndrome (POTS): A Frequently Missed Diagnosis

Aubrey R. George
College of Nursing, BYU
Master of Science

Postural Orthostatic Tachycardia Syndrome (POTS) is a debilitating chronic illness that involves a sustained tachycardia upon standing and affects millions of patients in the United States alone. POTS is most commonly seen in adolescent females and substantially reduces overall quality of life. In 2019, less than a third of all providers had heard of POTS and patients may wait over a decade for a proper diagnosis and treatment. With the intermittent and variable nature of symptoms, 83% of patients reported being misdiagnosed with a psychological condition before receiving a diagnosis of POTS. Understanding the relationship between autonomic compensation and hemodynamic stability can help a provider recognize and manage POTS. Nurse practitioners are especially suited to manage POTS because of their emphasis on creating a relationship with patients and emphasizing the importance of self-care. The purpose of this article is to provide nurse practitioners with a practical guide to recognizing, diagnosing, managing and educating patients with POTS.

Keywords: Postural orthostatic tachycardia syndrome (POTS), dysautonomia, orthostatic intolerance, adolescent, dizziness, fatigue, headache, presyncope, intermittent symptoms, self-care strategies, patient education, resources, primary care providers (PCP)
ACKNOWLEDGEMENTS

I would like to thank everyone who has helped make this paper possible. None of this would have been possible without my POTS provider Dr. Craig Coleby. He has taught me everything I understand about POTS and has always believed in me and been willing to answer my questions, share resources, be my preceptor and help me with this paper. I am also grateful for Chris Harper and everyone at Active Lifestyles Physical Therapy who have helped me and taught me so much. To all the nurses at the infusion center, thank you for dealing with the worst veins on earth and helping me stay healthy enough to finish school! Thank you to my amazing preceptors Chris Renfro and Dr. Wynn who have taught me so much! I am grateful for all my amazing professors who truly care about their students and consistently went out of their way to help us succeed, even during a worldwide pandemic. Thank you to my amazing cohort for being the best group to go through nurse practitioner school with! I am also grateful to the College of Nursing and the opportunity to learn the Healer’s Art. I also would not have been able to do this without the support of my family, they have been encouraging and supporting me since before I even knew what POTS was. My amazing husband Sean has helped me focus on finishing this paper and has believed in me and supported me when I did not think I could keep going. He has reminded me that I am so much more than a POTS patient and I honestly would not have been able to do any of this without him. And to Blaine, the chair of my paper who has probably spent hundreds of hours on this paper! Thank you for being patient and taking so much time and energy to help me write about something so personal! I think there were times when we both thought this paper would never be finished, so thank you for not giving up on me! I am also grateful for my Savior because His grace has enabled me to do so much more than I ever thought I could.
TABLE OF CONTENTS

Abstract ........................................................................................................................................... ii

Postural Orthostatic Tachycardia Syndrome (POTS): A Frequently Missed Diagnosis ........... 1

Introduction ....................................................................................................................................... 1

Clinical Presentation .......................................................................................................................... 2

Clinical Vignette.................................................................................................................................. 2

History of Present Illness ...................................................................................................................... 2

Dizziness ............................................................................................................................................... 2

Headache ............................................................................................................................................ 3

Fatigue ................................................................................................................................................ 3

Pathophysiology .................................................................................................................................. 3

Normal Physiology .............................................................................................................................. 4

Normal Compensation in Standing ...................................................................................................... 4

Pathophysiology of POTS .................................................................................................................... 5

Abnormal Compensation in Standing .................................................................................................. 5

The Final Common Pathway ................................................................................................................ 6

Diagnosis ............................................................................................................................................. 7

Prognosis ............................................................................................................................................. 8

Management ........................................................................................................................................ 8

Patient Education ............................................................................................................................... 9

Orem’s Theory on Self-Care ................................................................................................................ 9

Self-Care Strategies ............................................................................................................................ 10

Symptom #1: Dizziness ....................................................................................................................... 10
Symptom #2: Headache ................................................................. 11
Symptom #3: Fatigue and Exercise Intolerance ............................ 11
Dysautonomia International ....................................................... 12
Conclusion .................................................................................. 12
References .................................................................................. 13
Appendix A ................................................................................... 17
   Table A1: Diagnostic Criterion for POTS .................................. 17
   Table A2: Multi-system Symptoms in POTS ............................ 18
   Table A3: Autonomic Responses and Symptoms ...................... 20
   Table A4: Recommended Medications for POTS ....................... 22
   Table A5: Common Comorbidities and Red Flag Symptoms ......... 23
   Table A6: General Recommendations for POTS Management ....... 24
Appendix B ................................................................................... 26
   Figure B1: Relevant Hemodynamic Equations and Components ...... 26
   Figure B2: Normal Physiology of Standing Up ............................ 27
   Figure B3: Sympathetic Compensation for Postural Hemodynamic Instability ............ 28
   Figure B4: Final Common Pathway for POTS ............................ 29
   Figure B5: Diagnostic Approach to POTS .................................. 30
Postural Orthostatic Tachycardia Syndrome (POTS): A Frequently Missed Diagnosis

Introduction

Postural orthostatic tachycardia syndrome (POTS) is a form of dysautonomia characterized by sustained tachycardia upon standing (Arnold et al., 2018). While some providers classify POTS as an uncommon diagnosis, it is more common than many well-known diagnoses, including multiple sclerosis and Parkinson’s disease (Dysautonomia International, n.d.). POTS occurs in about 1% of the population in developed countries, with more than 3 million cases in the United States alone (Zadourian et al., 2018). It is one of the most common causes of syncope and presyncope in primary care, cardiology, neurology, and the emergency department, and it is especially common in Caucasian adolescent and young adult females (Arnold et al., 2018; Shaw et al., 2019). However, despite the prevalence of POTS, only 28% of primary care providers have heard of this syndrome (Shaw et al., 2019). Currently, it is estimated that the average pediatrician has 20–30 undiagnosed POTS patients receiving care in their clinic (Coleby, 2019).

This lack of provider awareness has created significant delays in care, where the average POTS patient waits 6 years and sees seven providers before receiving an appropriate diagnosis (Goodman, 2018). Lack of awareness can also increase the risk of misdiagnosis. As a result of the presentation of vague and intermittent symptoms, 83% of patients with POTS were initially misdiagnosed with a psychiatric condition (Goodman, 2018). Even if a patient receives a proper diagnosis of POTS, a provider’s lack of understanding can also delay treatment (Stiles et al., 2018). POTS patients often wait an additional 3–36 months after diagnosis before receiving proper treatment, and half of all patients must travel more than 100 miles for this treatment, with one-fifth traveling at least 500 miles (Stiles et al., 2018). The purpose of this paper is to provide
nurse practitioners with a practical guide to recognizing, diagnosing, managing, and educating patients with POTS.

Clinical Presentation

Clinical Vignette

Jane, a 19-year-old female patient, presented to her primary care provider with a chief complaint of dizziness. Her chart reflects several years of visits, various testing, and failed treatment. She looks defeated when the provider walks in but begins telling her story about how she woke up one day feeling weak and dizzy. Initially, she thought it was a brief illness. However, over time, her dizziness worsened, and she developed headaches, fatigue, exercise intolerance, and numerous other symptoms. She finished her story and asked, “Can you please help me? I went from being a high school athlete to being too weak to stand up in just a few weeks! Nobody seems to believe me, and I don’t know what to do anymore.”

History of Present Illness

Dizziness

During her evaluation, Jane reports that her first symptom of dizziness started about 8 months ago, and it has occurred daily since that time. She reported having a concussion a few months prior to the onset of dizziness (Miranda et al., 2018). Jane described the dizziness as a feeling of unsteadiness, as if she might fall over or pass out. While Jane said the dizziness does occur daily, it is not constant throughout the day. The dizziness worsens when she has been standing for a long period of time, during exercise, and when she feels hot. Sitting down and lying down seems to ease the symptoms, but does not always alleviate them. She reports that her dizziness is significantly affecting her ability to function.
Headache

Jane reports that frequent headaches began shortly after the onset of her dizziness. She described them as a generalized dull ache, with the pain radiating from behind her eyes to the top of her neck. Ibuprofen provides moderate relief but does not completely alleviate the pain. Loud noise, bright lights, the act of standing up, lack of sleep, and dehydration all seem to make the headaches worse. Additionally, her headaches seem to be worse later in the day, and she would rate their severity, on average, as a 6/10.

Fatigue

Jane’s fatigue also began after her dizziness and has been constant for at least six months. She feels weak most of the time, and, in conjunction with her dizziness, she occasionally experiences difficulty walking. Sleep does not alleviate the fatigue, because she wakes up feeling as if she has not had adequate rest. The fatigue is especially noticeable at the end of the day or following even minimal exercise. The combination of her dizziness, headaches, and fatigue have significantly limited her daily activities and have decreased her quality of life.

Pathophysiology

Historically, subtypes of POTS were used to categorize various patient presentations (Thanavaro & Thanavaro, 2011). However, this approach fails to recognize POTS as a syndrome instead of a disease. One author described this concept as POTS being an “end-phenotype and not a disease in itself” with the example of a fever (Raj & Robertson, 2018). A fever is a diagnosable problem, but there is an underlying pathology such as a bacterial infection causing the fever. To treat the fever, the bacterial infection must be diagnosed (Raj & Robertson, 2018).

Similarly, POTS is a diagnosable sustained tachycardia due to a separate and often overlapping underlying pathology (Garland et al., 2015). To understand POTS, providers must
transition from a compartmentalized view of body systems to a systemic understanding of the autonomic nervous system (ANS) and its effect on hemodynamic stability (Goldstein, 2017).

**Normal Physiology**

The ANS consists of the sympathetic and parasympathetic divisions, which work together through opposite effects to maintain homeostasis (Goldstein, 2017). In a healthy individual, the ANS uses a number of mechanisms to maintain adequate cardiac output and hemodynamic stability (Koya & Paul, 2020). This relationship between autonomic compensation and hemodynamic stability is especially relevant when standing. Figure 1 includes relevant hemodynamic components.

**Normal Compensation in Standing**

When a healthy person stands, the force of gravity causes an average person to shift 500–1,000mL of blood from the chest to the lower extremities, and up to 25% of their vascular volume moves to the interstitial space (Arnold et al., 2018). These changes reduce the amount of venous blood return to the heart and affect preload, stroke volume, and cardiac output (Arnold et al., 2018). Thus, when a healthy person stands, a positional shift in blood occurs and causes hemodynamic instability (see Figure 2). Almost immediately, autonomic baroreceptors sense gravitational changes in hemodynamics and compensate by increasing sympathetic activity (Heyer, 2017). This sympathetic response leads to a temporary rise in heart rate of 10–30 beats per minute (bpm), increased cardiac contractility, increased systemic vascular resistance, and various other changes (Garland et al., 2015). These sympathetic changes stabilize blood pressure until the body is able to regain orthostatic hemodynamic stability (Arnold et al., 2018). These autonomic compensatory adjustments occur quickly and restore appropriate cardiac output within seconds (see Figure 3).
Pathophysiology of POTS

When a patient with POTS stands up, gravity causes the same hemodynamic shifts to occur, and cardiac output decreases; however, the normal autonomic compensation is not adequate to restore hemodynamic stability (Arnold et al., 2018). While some providers have correlated the postural tachycardia in POTS patients with a pathologic decrease of central venous pressure (CVP) to 1–2 mmHg upon standing, the underlying cause of this change is rarely known (Coleby, 2019). This is why POTS is considered a syndrome and not a disease (Calvo et al., 2003).

Abnormal Compensation in Standing

The name “postural orthostatic tachycardia syndrome” itself explains that the tachycardia and other symptoms experienced by patients with POTS are based on position, especially the standing position. Abnormal compensation can occur when the body is unable to maintain adequate blood pressure upon standing, as seen in patients with orthostatic hypotension (Nwazue & Raj, 2013). However, the diagnostic criteria for POTS cannot include a decrease in blood pressure upon standing (Fedorowski, 2019). Instead, the sympathetic compensation in a POTS patient is the ability to maintain enough hemodynamic stability to avoid a dramatic drop in blood pressure. While this compensation is temporary in healthy individuals, the compensatory response remains constant in POTS patients or continues to increase as long as the patient is standing (Arnold et al., 2018). Thus, the sustained postural tachycardia and symptoms of orthostatic intolerance come from the continual autonomic compensation and tachycardia required to combat the persistently low CVP and cardiac output (Oketa-Onyut Julu, 2020). Figure 4 reflects the increased need for continual ANS compensation to maintain homeostasis (Fedorowski, 2019). In fact, some POTS patients may even have an increase in blood pressure as
they stand as a result of the continuous sympathetic activation. Similar to patients with orthostatic hypotension or vasovagal syncope, approximately 30–50% of POTS patients can still experience syncope. Instead, most patients with POTS experience daily presyncope, feeling as if they are going to pass out and experiencing severe autonomic symptoms each time they stand (Oketa-Onyut Julu, 2020).

The Final Common Pathway

To summarize, the final common pathway of POTS is prolonged sympathetic compensatory response to decreased cardiac output upon standing (Arnold et al., 2018). While most patients have an unknown and multifactorial cause of their POTS, some studies have found a profound decrease of CVP by at least 50%, with an average CVP of 1–2 mmHg when standing (Coleby, 2019). Another recent study demonstrated that 90% of patients with POTS and chronic fatigue syndrome (CFS) had reduced cerebral blood flow during a head-up tilt test (Van Campen et al., 2020). Thus, when a POTS patient stands, they experience profound cerebral hypoperfusion, which can manifest as dizziness, presyncope, and headaches (Van Campen et al., 2020). However, the ANS compensates for this decreased cardiac output and cerebral hypoperfusion through sympathetic activation (Koya & Paul, 2020). With constant sympathetic compensation for low cardiac output, increased levels of norepinephrine produce a sustained postural tachycardia (Garland et al., 2015). Sustained tachycardia and prolonged sympathetic activity also lead to orthostatic intolerance symptoms, including dizziness, headaches, and fatigue, all of which are commonly experienced by patients with POTS (Wells & Tonkin, 2016). Understanding the pathophysiology of POTS as continual autonomic compensation for global postural hemodynamic instability, providers can begin connecting the dots of seemingly unrelated problems to create a clearer diagnostic picture (Fedorowski, 2019).
Diagnosis

Table 1 indicates the major diagnostic criteria for POTS, including a sustained tachycardia upon standing, lack of orthostatic hypotension, symptoms of orthostatic intolerance for at least 6 months, and the elimination of other known causes of tachycardia (Fedorowski, 2019). While postural tachycardia is the hallmark symptom, POTS is a multifactorial disorder with involvement in most body systems (Busmer, 2011). Each patient can have a unique presentation with a variety of intermittent symptoms (see Table 2). This can make recognition difficult; however, most patients report symptoms of orthostatic intolerance, including lightheadedness, headache, fatigue, palpitations, shortness of breath, chest pain, blurred vision, nausea, trembling, or acrocyanosis (a purple color in the legs) when standing (Arnold et al., 2018).

Three important steps will aid in diagnosing POTS. First, it is essential to listen to the patient’s full story. In an article describing what patients with POTS want providers to know, patients urge health care professionals to look beyond their “invisible illness” and believe their stories (Stiles et al., 2018). Before providers assume patients are lying, they should consider that the wide variety of symptoms could be related to the ANS or to hemodynamics. Additionally, providers should remember that the intermittent nature of the symptoms could be related to the postural effect of POTS (Garland et al., 2015). Furthermore, providers should listen for common triggers before symptom onset, such as surgery, concussion, traumatic event, pregnancy, or viral illness (Garland et al., 2015). Lastly, providers should ask questions about autonomic symptoms or if the symptoms are related to standing, avoid discounting symptoms, and believe the patient.

Second, a provider should perform a cardiovascular assessment with the patient both supine and standing. POTS patients often have intermittent symptoms, so the assessment might
appear normal, especially if the patient is supine. As the standing cardiovascular assessment is performed, the provider should pay attention to differences in symptoms that could reflect hemodynamic changes, such as dependent acrocyanosis, tachycardia, sweating, eye dilation, or other autonomic findings (see Table 3). Third, the provider should perform orthostatic vital signs (Arnold et al., 2018). Figure 3 illustrates this diagnostic approach, which focuses on how to recognize intermittent and positional symptoms that could otherwise be missed.

**Prognosis**

POTS is a debilitating chronic illness that affects most body systems. One study revealed that half of all patients with POTS experienced at least 14 different symptoms, and 30% reported more than 26 symptoms (Thanavaro & Thanavaro, 2011). These symptoms vary from mild to severe, with 25% of patients progressing to complete disability (Revlock, 2018). Quality of life for POTS patients has been compared to that of patients with congestive heart failure and chronic obstructive pulmonary disease, which is especially troubling because most POTS patients are adolescents (McDonald et al., 2014). Increased rates of depression are also associated with the diminished quality of life that POTS patients experience (Moon et al., 2016). However, prognosis can change drastically with appropriate treatment. Approximately 70% of patients report symptomatic improvement following initial treatment (Coleby, 2019).

**Management**

Primary care providers, including nurse practitioners, are capable of managing the majority of POTS patients (Coleby, 2019). Research has found that most POTS patients find improvements through a combination of nonpharmacologic and pharmacologic interventions that combat hemodynamic instability and regulate autonomic imbalance (Thanavaro & Thanavaro, 2011). Pharmacotherapy is often individualized and focused on symptom management (Miller &
Raj, 2018). The four main goals of POTS pharmacotherapy are to increase blood volume, reduce heart rate, constrict blood vessels, and lower sympathetic activation (Miller & Raj, 2018). It is important to remember the final common pathway of POTS while initiating treatment, because if a provider does not increase blood volume before decreasing heart rate, symptoms may actually worsen, since the tachycardia is compensating to maintain cardiac output (see Figure 4). Table 4 includes a list of recommended medications. Interprofessional treatment and specialists can be used in the management of comorbidities, ruling out other diagnoses, managing patients with red-flag symptoms, or assisting those who do not respond to initial treatment (see Table 5).

**Orem’s Theory on Self-Care**

Patient education is an essential part of managing POTS. Since most treatment is individualized, providers and patients must work together to discuss symptoms and create treatment plans. Offering self-care, establishing a positive relationship with the patient, and providing education about recommendations are all crucial steps.

In the 20th century, Dorothea Orem developed the Orem Model of Nursing, or the Self-Care Deficit Nursing Theory, which focuses on the relationships between nursing, patients, and self-care (Petiprin, 2020). This theory has three parts: the theory of self-care, the theory of self-care deficit, and the theory of the nursing system. The theory of self-care illustrates the importance of individuals managing themselves (Denyes et al., 2001), which is especially important in patients with POTS.

Orem’s theory emphasizes the importance of a nurse’s relationship with patients at the times in their life when they are the most vulnerable. The patient Jane in the clinical vignette tearfully asked the provider to help her, because she was scared about how quickly her health had declined and because few providers had taken her seriously. When a patient develops POTS,
even the most basic fundamentals of life can be nearly impossible, and self-care is substantially impacted. Nurse practitioners are in a unique position to make a difference by developing a relationship with patients and collaborating on a plan of care. Educating patients and empowering them to be a part of their care can give them hope, and this is especially pertinent for patients with POTS (Raj & Robertson, 2018).

**Self-Care Strategies**

Non-pharmacologic interventions are a foundation of management for patients with POTS and are introduced as a first-line treatment, as seen in Table 6 (Miller & Raj, 2018). Jane’s most troubling symptoms were dizziness, headaches, fatigue, and exercise intolerance.

**Symptom #1: Dizziness**

Orthostatic intolerance is often described by patients as dizziness, lightheadedness, presyncope, or syncope, and it can be improved through compression (Zadourian et al., 2018). When POTS patients stand, excessive venous blood pools in their lower extremities and abdomen, which leads to lower cardiac output (Fu & Levine, 2018). Compression garments prevent blood pooling and help stabilize cardiac output (Fu & Levine, 2018). Class 2 waist-high compression stockings with at least 30mmHg, or 20mmHg–40mmHg, are recommended as the most effective treatment (Fedorowski, 2019). Levine emphasizes the importance of complete lower body compression, including legs and the lower abdomen. If this is not possible, abdominal compression is recommended over leg compression due to significant splanchnic blood pooling that occurs in POTS patients (Fu & Levine, 2018).

**Symptom #2: Headache**

Headache is one of the most common symptoms seen in POTS patients and can be improved through volume expansion (Van Campen et al., 2020). While some patients experience
relief with migraine treatment for primary headaches, most also have secondary headaches due to impaired blood flow (Cook & Sandroni, 2018). A recent study demonstrated that 90% of patients with POTS had reduced cerebral blood flow during a head-up tilt test (Van Campen et al., 2020). Understanding headaches due to cerebral hypoperfusion explains why volume expansion is an effective strategy to reduce the frequency and severity of headaches (Van Campen et al., 2020). While volume expansion can be addressed through medication or intravenous fluids, an increase of salt and fluid intake orally is crucial (Zadourian et al., 2018). Fu and Levine (2018) recommend intaking salt and water together throughout the day, with a combination of salt supplements, salting food, and drinking glucose-salt rehydration solutions to reach at least 10g of sodium and at least 3 liters of fluid a day. Thus, replacing water with sports or electrolyte drinks, salting food, supplementing additional sodium intake, and increasing fluid intake should help increase cardiac output and reduce headaches (Fu & Levine, 2018).

**Symptom #3: Fatigue and Exercise Intolerance**

Fatigue and exercise intolerance can be improved through recumbent exercise (Fedorowski, 2019). The orthostatic intolerance symptoms found in patients with POTS make it difficult at times to stand, let alone exercise; however, bed rest and cardiac deconditioning will worsen the symptoms associated with POTS (Fu & Levine, 2018). While symptoms are likely to be exacerbated initially, exercise is fundamental in the treatment of POTS (Fu & Levine, 2018). The benefits of exercise include decreasing baseline and orthostatic heart rate, increasing blood volume, improving baroreflex sensitivity, strengthening cardiac muscle, reducing symptoms, increasing exercise tolerance, and improving the overall quality of life (Arnold et al., 2018). Exercises such as swimming, biking, or rowing should be recommended (Zadourian et al., 2018). In addition to endurance training, strength training focused on the lower extremities and the core
POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME

is essential. The Levine protocol provides a 3-month program that includes interval training, strength training, and, eventually, upright exercise (Fu & Levine, 2018). While this protocol may be difficult for the patients at first, it is the cornerstone of treatment and self-care (Fu & Levine, 2018).

**Patient Resources**

Patients may want additional resources about POTS, to connect with others, or for strategies to manage their illness (see Table 6). Dysautonomia International (n.d.) is one helpful resource for both patients and providers that offers credible information about dysautonomic conditions, contains an example of the Levine Protocol, schedules educational conferences, and ways to network with other POTS patients.

**Conclusion**

POTS is a common but debilitating condition that affects millions of people, including many adolescent and young adult females (Goodman, 2018). Currently, a lack of understanding about POTS among health care providers has left many patients with a delayed diagnosis and inadequate treatment (Stiles et al., 2018). This article provides nurse practitioners with a practical guide to recognize, diagnose, and manage POTS. If providers understand POTS as a syndrome with an unknown cause but a common underlying problem of autonomic compensation for hemodynamic instability upon standing, they can make a difference. Nurse practitioners have a unique ability to create relationships with patients and collaborate with them to manage health challenges by providing education, including self-care strategies and resources for patients. Armed with compassion and a practical guide about POTS, one provider can connect the dots of seemingly disconnected symptoms and offer hope to countless POTS patients (Raj & Robertson, 2018).
References


Appendix A

Table A 1

*Diagnostic Criteria for POTS*

<table>
<thead>
<tr>
<th>Diagnostic Criteria for POTS Patients</th>
<th>Parameters &amp; Recommendations</th>
</tr>
</thead>
</table>
| Significant increase in heart rate upon standing that remains elevated or keeps increasing as long as the patient is standing | *Adults:* Heart rate increase >30bpm or above 120bpm within 10 minutes of standing and stays elevated or increases while standing  
*Children/Adolescents:* Heart rate increase >40bpm or above 120bpm within 10 minutes of standing and stays elevated or increases while standing |
| Lack of orthostatic hypotension, or significant drop in blood pressure, when patient is standing | Blood Pressure cannot drop more than 20mmHg systolic and 10mmHg Diastolic (20mmHg/10mmHg) |
| Chronic symptoms of orthostatic intolerance | Symptoms for at least 6 months |
| Absence of any known pathologies that cause sinus tachycardia | *Some recommended lab tests:* CBC, CMP, TSH & T4, Recumbent & standing plasma catecholamines, ANA & immune workup, Vitamin B12, Complement, ESR, 24-hour urine, etc.  
*Other recommended tests:* 24-hour Holter monitor, ECG, Echocardiogram, Autonomic testing or Tilt Table Test if needed, Brain MRI, etc. |

*Note:* POTS is a syndrome, thus the diagnostic criterion for POTS involves ruling out other known pathologies in addition to specific heart rate criteria (Arnold et al., 2018; Fedorowki, 2019).
### Table A 2

*Multi-system Symptoms in POTS*

<table>
<thead>
<tr>
<th>System</th>
<th>Subjective Symptom</th>
<th>Objective Sign</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiovascular</td>
<td>Chest pain, syncope (passing out) &amp; presyncope (feeling close to passing out), lightheadedness, palpitations, exercise intolerance, tunnel vision</td>
<td>Postural tachycardia (especially when standing), possible hypertension</td>
</tr>
<tr>
<td>Circulatory</td>
<td>Edema, mottling, venous pooling/extremities are purple or red, cold extremities,</td>
<td>Mottling, Dependent acrocyanosis, venous pooling, flash capillary refill on lower extremities &amp; delayed capillary refill in upper extremities</td>
</tr>
<tr>
<td>Respiratory</td>
<td>Dyspnea, cough, pressure on chest</td>
<td>Short of breath, difficulty talking &amp; hyperventilation</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Vertigo, paresthesia, headache, tremors, cognitive dysfunction, numbness, pain, hyperacusis, blurry vision, double vision, photophobia, dry eyes, dry mouth, hoarse voice, tinnitus</td>
<td>Dilated pupils, tremor, unsteady gait &amp; hoarse voice</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Nausea, dysphagia, early satiety, bloating, constipation, diarrhea, vomiting, abdominal pain, GI dysmotility, frequent throat clearing, acid reflux, hypoglycemia</td>
<td>Abdominal distension, painful to palpation, frequent throat clearing &amp; hypoactive or hyperactive bowel sounds</td>
</tr>
<tr>
<td>Renal</td>
<td>Nocturia, urinary retention, difficulty initiating urination,</td>
<td>N/A</td>
</tr>
<tr>
<td>System</td>
<td>Frequency, urgency, incontinence</td>
<td>Diaphoresis, flushing, weight gain, weight loss, fatigue, insomnia,</td>
</tr>
<tr>
<td>-----------------</td>
<td>----------------------------------</td>
<td>---------------------------------------------------------------------</td>
</tr>
<tr>
<td><strong>Endocrine</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Musculoskeletal</strong></td>
<td>Lower extremity weakness, heavy legs, generalized joint pain and swelling, generalized weakness &amp; restless leg pain</td>
<td></td>
</tr>
<tr>
<td><strong>Reproductive</strong></td>
<td>Impotence, dysmenorrhea, menorrhagia, metrorrhagia &amp; symptom severity increases around menstrual cycle</td>
<td></td>
</tr>
</tbody>
</table>

*Note: POTS is a complex dysautonomic syndrome that involves symptoms from most major body systems (Arnold et al., 2018; Busmer, 2011; Garland et al., 2018; Zadourian et al., 2018).*
Table A 3

Autonomic Responses and Symptoms

<table>
<thead>
<tr>
<th>Targeted Tissue</th>
<th>Sympathetic Response</th>
<th>Parasympathetic Response</th>
<th>Autonomic Signs &amp; Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Eyes</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>-Pupils</td>
<td><strong>Pupils: Dilation</strong></td>
<td><strong>Pupils: Constriction</strong></td>
<td>-Blurry vision or double vision</td>
</tr>
<tr>
<td>-Lacrimation</td>
<td><em>Lacrimation: ( \downarrow )</em></td>
<td><em>Lacrimation: ( \uparrow )</em></td>
<td>-Photophobia</td>
</tr>
<tr>
<td></td>
<td>secretions</td>
<td>secretions</td>
<td>-Dry Eyes</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Excessive tearing</td>
</tr>
<tr>
<td><strong>Oropharynx</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>-Secretions</td>
<td><strong>Secretions: Thick</strong></td>
<td><strong>Secretions: ↑</strong></td>
<td>-Dry Mouth</td>
</tr>
<tr>
<td>-Pharynx:</td>
<td><em>Pharynx: No change</em></td>
<td><em>Pharynx:</em></td>
<td>-Frequent Cough or clearing throat</td>
</tr>
<tr>
<td></td>
<td></td>
<td><em>Coordinate swallow</em></td>
<td>-Dysphagia</td>
</tr>
<tr>
<td></td>
<td></td>
<td><em>and speech</em></td>
<td>-Hoarse voice</td>
</tr>
<tr>
<td><strong>Heart</strong></td>
<td><strong>SA Node: ↑ Heart rate</strong></td>
<td><strong>SA Node: ↓ Heart rate</strong></td>
<td>-Chest Pain</td>
</tr>
<tr>
<td>-SA Node</td>
<td><em>Chambers: ↑ contractility</em></td>
<td><em>Chambers: ↓ contractility</em></td>
<td>-Dizziness</td>
</tr>
<tr>
<td>-Ventricles</td>
<td></td>
<td></td>
<td>-Syncope/Presyncope</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Palpitations</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Lightheaded</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Dizziness</td>
</tr>
<tr>
<td><strong>Blood Vessels</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td><strong>Vasoconstriction</strong></td>
<td><strong>Vasodilation</strong></td>
<td>-Productive cough</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Dyspnea</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Chest pain</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Hyperventilation</td>
</tr>
<tr>
<td><strong>Lungs</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>-Bronchi</td>
<td><strong>Bronchi: Dilate</strong></td>
<td><strong>Bronchi: Constrict</strong></td>
<td>-Nausea</td>
</tr>
<tr>
<td>-Secretions</td>
<td><em>Secretions: ↓ production</em></td>
<td><em>Secretions: ↑ production</em></td>
<td>-Vomiting</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Anorexia</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Abdominal pain</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Constipation</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Early satiety</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Bloating</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Acid reflux</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>-Diarrhea</td>
</tr>
<tr>
<td><strong>Gastrointestinal</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Tract</strong></td>
<td></td>
<td></td>
<td>-Urinary Retention</td>
</tr>
<tr>
<td>-Motility</td>
<td><strong>Motility: ↓</strong></td>
<td><strong>Motility: ↑</strong></td>
<td>-Neurogenic Bladder</td>
</tr>
<tr>
<td>-Sphincters</td>
<td><strong>Sphincters: Contract</strong></td>
<td><strong>Sphincters: Relax</strong></td>
<td></td>
</tr>
<tr>
<td>-Secretions</td>
<td><em>Sphincters: ↓ Perfusion:</em></td>
<td><em>Sphincters: ↑ Perfusion:</em></td>
<td></td>
</tr>
<tr>
<td>-Perfusion</td>
<td>*Liver: Glycogenolysis &amp;</td>
<td><em>Liver: Gallbladder</em></td>
<td></td>
</tr>
<tr>
<td>-Liver</td>
<td>Gluconeogenesis</td>
<td>and bile ducts dilate</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Kidneys: No change</strong></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
**Urinary Tract**  
- Kidneys  
  - Kidneys: ↓ Renin & urinary retention  
- Bladder  
  - Bladder: Detrusor relax & sphincter contract  
  - Bladder: Detrusor Contract & Sphincters relax  
  - Bladder Spasms  
  - Incontinence  
  - Frequency

**Integumentary**  
- Sweat Glands  
- Hair Follicles  
  - Sweat glands: ↑ secretions  
  - Hair Follicles: Bristle  
  - No major change  
  - Hyperhidrosis  
  - Goosebumps

**Skeletal Muscles**  
- Increase blood flow  
- Rhythmic muscle contraction  
  - No major change  
  - Tremors  
  - Muscle spasms  
  - Teeth chattering

*Note:* Understanding the sympathetic and parasympathetic divisions of the ANS and recognizing autonomic symptoms can help a provider recognize and diagnosis POTS (Goldstein, 2017; Wells & Tonkin, 2016).
Table A 4

**Recommended Medications for POTS**

<table>
<thead>
<tr>
<th>Medication</th>
<th>Recommended Dose</th>
<th>Rationale</th>
<th>Important Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Fludrocortisone</strong></td>
<td>0.1mg-0.2mg daily</td>
<td>Volume Expander</td>
<td>Monitor Electrolytes</td>
</tr>
<tr>
<td><strong>Sodium Tablets</strong></td>
<td>1 gram 3-5 x daily</td>
<td>Volume Expander</td>
<td>Monitor Electrolytes &amp; increase fluid intake</td>
</tr>
<tr>
<td>IV Saline Infusions</td>
<td>2 Liters over 2-3 hours weekly or PRN</td>
<td>Volume Expander</td>
<td>Avoid central lines if possible, d/t risk of infection</td>
</tr>
<tr>
<td><strong>Low Dose Beta Blocker</strong></td>
<td>20mg Propranolol</td>
<td>Lower Heartrate</td>
<td>A normal dose will exacerbate symptoms d/t ↓ Blood Pressure</td>
</tr>
<tr>
<td>Ivabradine</td>
<td>2.5-7.5mg BID (start at 5mg BID)</td>
<td>Lower Heartrate</td>
<td>This reduces Heart rate WITHOUT affecting Blood Pressure</td>
</tr>
<tr>
<td><strong>Midodrine</strong></td>
<td>5-15mg TID (start at 2.5-5mg)</td>
<td>Vasoconstriction</td>
<td>This will increase Blood Pressure, do not use in Hypertension</td>
</tr>
<tr>
<td>Lower Dose Stimulants</td>
<td>Methylphenidate 10mg TID or 10-20mg Adderall daily</td>
<td>Vasoconstriction (↑ cerebral perfusion)</td>
<td>*Watch for Tachycardia, especially in higher doses</td>
</tr>
<tr>
<td>Clonidine</td>
<td>0.1mg-0.2mg at night 0.1-0.3mg transdermal patch</td>
<td>Sympatholytic (↓ sympathetic activation)</td>
<td>*This is especially helpful in patients with labs that show increased catecholamines upon standing</td>
</tr>
</tbody>
</table>

*Note: Aside from symptom management, in order to reduce symptoms while maintaining hemodynamic stability, most pharmacologic management for POTS includes volume expansion, reducing heart rate, vasoconstriction, and reducing sympathetic activation (Miller & Raj, 2018). The four bolded medications are considered the foundation to initial pharmacologic treatment (Coleby, 2019).*
### Table A 5

**Common Comorbidities and Red Flag Symptoms**

<table>
<thead>
<tr>
<th>Red Flag Symptoms</th>
<th>Possible Comorbidity</th>
<th>Next Step</th>
</tr>
</thead>
<tbody>
<tr>
<td>Musculoskeletal symptoms with fatigue including joint hypermobility or instability, chronic arthralgia in at least 3 joints or widespread pain</td>
<td>Hypermobile Ehler-Danlos syndrome (hEDS)</td>
<td>-Beighton scale -Refer to Physical Therapist or Chiropractor that knows about hEDS</td>
</tr>
<tr>
<td>Allergic symptoms including flushing, anaphylaxis, recurrent crampy abdominal pain, diarrhea and itching</td>
<td>Mast cell activation syndrome (MCAS)</td>
<td>-Can refer to an Allergist -Can prescribe antihistamines and Montelukast</td>
</tr>
<tr>
<td>Epigastric pain with any food or water intake</td>
<td>Median Arcuate Ligament Syndrome (MALS)</td>
<td>-Order CT Angiogram -Celiac Plexus Block -Refer to General Surgeon</td>
</tr>
<tr>
<td>Daily persistent headache, Abdominal pain, back pain or flank pain, pelvic pain, dysuria and dyspareunia, hematuria</td>
<td>Nutcracker Syndrome</td>
<td>-Prescribe Aspirin and manage symptoms -Order CT Angiogram -Refer to Urology</td>
</tr>
<tr>
<td>Orthostatic headaches with interscapular pain, tinnitus and metallic taste</td>
<td>Spontaneous intracranial hypotension due to CSF leak</td>
<td>-MRI with and without contrast -Refer for epidural blood patch</td>
</tr>
</tbody>
</table>

*Note: Most POTS patients will respond to initial treatment, however common comorbidities can be recognized through treatment resistance or red flag symptoms (Coleby, 2019; Doherty & White, 2018; Roma et al., 2018; Scholbach, 2007; Stubberud et al., 2020).*
### Table A 6

*General Recommendations for POTS Management*

<table>
<thead>
<tr>
<th>Intervention Type</th>
<th>Recommendation</th>
<th>Rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Compression Stockings (waist high, at least30mmHg)</td>
<td>Improves venous return, stabilizes cardiac output and reduces symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>10 grams of Salt a Day</td>
<td>Increases blood volume and reduces symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>4+ Liters of Electrolyte Fluid a Day, not water</td>
<td>Increases blood volume and reduces symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Recumbent Exercise</td>
<td>Strengthens heart and improves quality of life</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Counter maneuvers (pumping leg muscles)</td>
<td>Combats blood pooling when standing to reduce symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Raise whole head of bed a few inches</td>
<td>Autonomic conditioning can help reduce symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Eat smaller meals more frequently</td>
<td>Eating large meals can exacerbate symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Avoid heat when possible (use fans and cooling cloths)</td>
<td>Heat can exacerbate symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Avoid other triggers (standing, dehydration, sleeping for too long, deconditioning, etc.)</td>
<td>These triggers can also exacerbate symptoms</td>
</tr>
<tr>
<td>Non-Pharmacological or Self Care</td>
<td>Visit Dysautonomia International Website</td>
<td>Helpful resources with exercise program, recommended self-care strategies and networking</td>
</tr>
<tr>
<td>Interdisciplinary Recommendation</td>
<td>Meet with Dietitian</td>
<td>Discuss salt intake, symptoms of gastroparesis and other specific concerns</td>
</tr>
<tr>
<td>Interdisciplinary Recommendation</td>
<td>Meet with Physical Therapist who knows about POTS</td>
<td>Can help strengthen lower extremities and start exercise program</td>
</tr>
<tr>
<td>Interdisciplinary Recommendation</td>
<td>Meet with a Pelvic Floor Physical Therapist</td>
<td>Can help with incontinence, dyspareunia, and other symptoms</td>
</tr>
<tr>
<td>Interdisciplinary Recommendation</td>
<td>Meet with a Cognitive Behavioral Therapist</td>
<td>Cope with life changing illness, limbic retraining and family dynamics</td>
</tr>
<tr>
<td>Interdisciplinary Recommendation</td>
<td>Go to Emergency Room or Urgent Care if Dehydrated or Acutely Ill</td>
<td>Intravenous Fluids can significantly improve POTS symptoms</td>
</tr>
</tbody>
</table>
Note: Non-pharmacologic, or self-care interventions, and interdisciplinary care are essential in POTS (Chelimsky & Chelimsky, 2018; Fu & Levine, 2018; Heyer, 2017; Miller & Raj, 2018; Raj et al., 2018).
Appendix B

Figure B 1

Relevant Hemodynamic Equations and Components

Note: There are many different components involved in hemodynamic stability and maintenance of homeostasis (King & Lowery, 2020).
Figure B 2

*Normal Physiology of Standing Up*

*Note:* When a health person stands, the force of gravity causes changes in blood flow and leads to hemodynamic instability (Arnold et al., 2018).
Figure B 3

*Sympathetic Compensation for Postural Hemodynamic Instability*

Note: The autonomic nervous system recognizes hemodynamic instability and the sympathetic division is activated to compensate for these changes and maintain homeostasis (Koya & Paul, 2020).
Figure B 4

*Final Common Pathway for POTS: Continual Autonomic Compensation for Hemodynamic Instability*

*Note:* In patients with POTS, normal sympathetic activation is *not* enough and the body must maintain the sympathetic compensation, including tachycardia, to maintain cardiac output (Arnold et al., 2018).
Figure B 5

Diagnostic Approach to POTS

Note: In order to diagnosis POTs, a provider must listen to a patient’s full history, perform a physical assessment while standing, and obtain orthostatic vital signs upon standing and at 2, 5 and 10 minutes as the patient stands (Arnold et al., 2018).