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Commentary

Intensive rehabilitation can be tolerated by adolescents with Postural Orthostatic Tachycardia Syndrome (POTS)

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Introduction

Postural Orthostatic Tachycardia Syndrome (POTS) is a common comorbidity seen in patients who are also enrolled in chronic pain rehabilitation (Kizilbash et al., 2014), specifically those with chronic headaches (Mohr, 2017). It is more commonly, however, a secondary diagnosis. In our experience, although it may require some minor alterations clinically, functional rehabilitation can be tolerated by this population similarly to those with chronic pain diagnoses. This paper highlights the potential for adolescents with POTS to tolerate and participate in more rigorous, short-term rehabilitation programs. Gains made by these patients seem to be similar to those who have participated in treatment methods which take longer periods of time. Suggestions for physical and occupational therapy treatment and appropriate, supporting, outcomes from this specific diagnosis are included.

POTS is currently defined as an increase in heart rate by 30 beats per minute (bpm), or over 120 bpm within 10 minutes of standing, not due to orthostatic hypotension (Dysautonomia International, 2012). It has been suggested that diagnosis criteria for adults and children should be different and that criteria specifically for children and adolescents do not exist yet. Also, in using the current criteria of 30 bpm, it seems that there is a trend for over-diagnosis of POTS in children and adolescents (Singer et al., 2012; Stewart, 2000). There are many pharmacological interventions that may be recommended for individuals with POTS, in addition to physical and occupational therapies.

Potential barriers for these individuals during participation in physical activities include drops in blood pressure, dizziness, fatigue, heart palpitations, nausea, and tremulousness (Dysautonomia International, 2012). In more involved cases, syncope can occur with simple transitions, which may make it feel hazardous to participate in physical activities. When working with these patients, it is extremely important to have baseline information (e.g. resting heart rate and blood pressure), as well as information regarding anxiety prior to initiating physical activity. In addition, providing education to patients, their families, and other team members (e.g. physicians, nurses, teachers, and coaches) can facilitate more consistent physical activity and conditioning.

Exercise and increased hydration as a part of POTS treatment has been recommended in past research (Mtinangi & Hainsworth, 1998; Winker et al., 2005; Fu et al., 2011; Joyner, 2011; White et al., 2011; Nicholson, 2015), however, some additional supportive tips may be helpful for clinicians and teams as they begin working with this patient population. Previous research has documented treatment over a lengthy period of time (up to year) allowing for a very gradual increase in physical activity (Brilla et al., 1998; Winker et al., 2005; White et al., 2011; Fu & Levine, 2015). Although it is not documented in research, anecdotally we often hear that medical providers inform patients and families that they will not be able to participate in physical activity secondary to their symptoms. After being told this, we have often observed that patients' symptoms only seem to increase or become more frequent. Families then doubt the ability of the patient to tolerate or participate in a standard intensive rehabilitation program, being that the timeframe for treatment is often much shorter.

In the interdisciplinary pain rehabilitation program described herein, adolescents are seen intensively for 3 weeks, 5-6 days a week, with 3-4 hours of physical and occupational therapy each day. Within the program, adolescents participate in a combination of rehabilitation therapies, psychological services, medical subspecialty care, alternative therapies (aromatherapy, acupuncture, biofeedback, music, art, and Reiki), and school services. All sessions focus on improved functioning by increasing strength and endurance, facilitating return to daily life activities, and using appropriate self-directed coping and pain management skills despite symptoms and pain. These skills are reflected in each participant's evening recommendations, which are later put together as an official home program. This specific program does not focus on further diagnostics of chronic pain or endorse invasive medical procedures.

Areas that physical and occupational therapies focus on include: education and building body awareness and alignment, joint conservation skills, energy conservation skills, parent education about how slight increases in heart rate are safe and recommended, lower extremity and core strengthening, progression of endurance as tolerated, and higher level balance and higher impact skills (Brilla et al., 1998; Winker et al., 2005).

The recommended progression in rehabilitation therapies for adolescents with POTS can be broken into three phases, each with varying timelines based on the individual's tolerance and participation. The first phase focuses on background information, basic skill introduction, and foundational movement skills. The second phase incorporates more advanced education and activities, and the third phase focuses on endurance activities, return to life, and independence with skills. A brief summary of phases and treatment information can be viewed in Table 1.

The phase progression discussed is a general outline of the order in which we progress skills in the program. However, continual monitoring of how the patient is doing (using subjective observation to avoid over attending to objective measures, i.e. heart rate, blood pressure, or other symptoms) and how receptive or open they are to advancing to the next phase is very important. Patients with POTS can tolerate gradual exercise training as a way to manage syncope and orthostatic tolerance. Symptoms can improve without increasing resting blood pressures and causing additional orthostatic symptoms if completed gradually and properly (Masuki et al., 2007).

Many participants will agree to lower impact physical activities but struggle to tolerate prolonged endurance, circuit training, and high impact activities, which would facilitate quicker lower extremity strengthening and improved cardiovascular endurance and are functionally relevant. So, how do we promote tolerance of these more challenging tasks? During challenging physical activity, it is important to gradually progress skills to help facilitate self-confidence, allow for proper coping, and ensure safety. By teaching the individual to appropriately work into, and tolerate, increased heart rate and other symptoms, they can better elicit muscle fatigue, appropriately building strength and stamina. Providing examples and education about the importance of lower extremity strength training to encourage venous return (Goodman & Helgeson, 2011) to help maintain heart rate and blood pressure (Joyner, 2011) while teaching the patient to become more mindful to facilitate progression of skills. Clinicians should use caution because at times a patient becoming more in tune with their body can lead to further focus on symptoms. To this end, focusing on appropriate use of coping skills and normalizing symptoms to enhance confidence is important. Educating patients about appropriate rest breaks can help to minimize symptoms and more appropriately utilize energy conservation strategies. Specific feedback about individualized signs and symptoms that should or should not be utilized as warning signs (e.g. typical increase in heart rate vs. significant increase in heart rate causing difficulty breathing) or information regarding resting posture

Table 1
Typical treatment components

Phase 1:

- Stretching/flexibility training
- Basic breath training (to support activities and limit symptoms)
- Basic education about coping/relaxation skills
- Strengthening with focus on lower limb resistance training and joint stabilization
- Transition training (e.g. floor to stand, pivoting/turning, sit to stand)
- Lower impact exercise
- Body awareness
- Energy and joint conservation strategies
- Goal setting (i.e. identify specific activities are they motivated to return to and at what level)

Phase 2: (add to above)

- Initiate more advanced/challenging activities (i.e. focus on how to change or modify activities to either make basic activities more challenging or hard activities easier to tolerate)
- Focus on breathing technique and use of rest breaks
- Recognize muscle fatigue vs. increased heart rate or symptoms
- Continue discussion on pacing and appropriate modifications

Note: In this phase specific focus is placed on what symptoms are appropriate to acknowledge and to what extent, and then using previously mentioned coping skills to tolerate activities. Education focuses on the importance of recognizing muscle fatigue, alignment, and proper pace with patients paying less attention to elevated heard rate, dizziness, or other associated symptoms.

Phase 3: (add to above)

- Longer intervals of activities to build musculature and cardiovascular endurance
- Continue working on challenging and higher level skills
    - initiate jump training and jogging if appropriate
- Focus on specific goals and home going planning
- Work on independence with all skills and concepts learned so far
    - Can they independently modify activities to appropriately participate in gym class?
    - Are they able to take appropriate rest breaks without reminders?
    - Can they independently identify muscle fatigue to appropriately strength training without increasing symptoms?
    - Do they appropriately apply coping skills to work on more challenging physical activities?
    - Are they able to fully participate in preferred recreational, leisure, and social activities?

On request, appendix available from the author with more details on specific therapeutic activities and rationale.

and how it can exacerbate or limit symptoms (e.g. closing eyes or placing head in lap before returning to sitting position typically increases dizziness compared to sitting with eyes open and focused on a distant visual will help dizziness resolve) can also be helpful. Also, it can be useful to provide examples of how completion of daily activities, attending school, or participating in sporting practices using poor energy or joint conservation strategies will most likely result in working inefficiently, leading to exacerbated symptoms.

The ability of the patient to learn, understand, and utilize basic breath training, coping and relaxation skills can facilitate challenging tasks despite the presence of symptoms (e.g. using focused breathing to support transitions or progressive muscle relaxation to calm surrounding musculature prior to tasks to emphasize stability). With breath training, educate the patient how improper breathing can exacerbate symptoms and demonstrate how appropriate use of exhale and inhale can support activities and limit symptoms. Have patients use their ability to manage breathing as a way to continue or discontinue activities. Introduce basic coping skills that are helpful both during physical activities and at rest. Help the patient understand how to utilize these skills with specific practice. Some of the most helpful skills to teach include: breathing, distraction, problem solving, imagery, acupressure, and progressive muscle relaxation (Culbert & Kajander, 2007; Carter & Threlkeld, 2012; Bryant et al., 2014; Linehan, 2017; Allen et al., 2018).

To further improve the quality of treatment sessions, make sure to include education about all aspects of treatment, including the purpose of activities, what they accomplish, why they are important, and how each will translate to improve functioning. Stay focused on patient goals so that they stay motivated and are better able to cope and manage possible anxiety. Provide parent education often, encourage parents to observe the skills you are working on, discuss their rationale, and emphasize importance of prompting and reinforcing these skills at home.

In support of these recommendations, a retrospective review was completed on program participants with POTS as their primary diagnosis at admission (as clinically given by a referring provider). In total, outcome data from 15 patients at admission and discharge were reviewed. Participants consisted of adolescents ranging in age from 11 to 21 years of age, all with a primary diagnosis of POTS, along with other secondary chronic pain diagnoses (e.g. headaches, migraines, abdominal pain, pain amplification syndrome) and various chronicity of chronic pain (Table 2). Measures included self-report and objective measures, with positive change scores demonstrating improvements. Participants self-reported their perceived function using the Upper Extremity Functioning Index (UEFI; Stratford et al., 2001) and the Lower Extremity Functioning Scale (LEFS; Binkley et al., 1999). These assessment tools allow patients to rate their perceived ability to perform everyday tasks with upper and lower extremities by rating the difficulty or ease of completion. An improvement of 9 or more points for the LEFS and UEFI is considered clinically significant. In addition, a single-item measure evaluated pain severity rating using a numerical rating scale (NRS) from 0 (no pain) to 10 (worst pain; McGrath et al., 2008). These measures can clinically guide treatment as they represent a self-reflection of perceived abilities, physical gains, and improved mindfulness.

Table 2
Participant information

Demographics	
Age Range	11-21 years old
Female	10 (67%)
Male	5 (33%)
Length of Stay	
3 weeks	13 (87%)
4 weeks	2 (13%)
Chronicity	
12 months or less	0 (0%)
13-18 months	3 (20%)
19-24 months	4 (27%)
24+ months	8 (53%)

In terms of objective measures, participants also completed a number of physical exercises (kneeling push-ups, elbow planks [in seconds], box carry, and step ups) and recorded the number of repetitions they completed in 60 seconds. The participants were educated about appropriate energy and joint conservation techniques, as well as given demonstrations of how to complete each exercise with appropriate muscle activation. These measures objectively demonstrate changes in tolerance of physical activity, progression of strength and endurance, tolerance of prolonged or increased symptoms, and reflect a sense of self-confidence in that patients trialed and completed each interval to the best of their ability.

The results shown in Table 3 demonstrate that on average these participants demonstrated gains on all functional outcome measures. These improvements in physical and functional gains occurred despite a slight albeit non-significant increase in self-reported pain severity. It is important to point out that this increase in pain severity during treatment is an often anticipated short-term outcome, and prior studies have found that in a general chronic pain cohort, pain decreases longer term (Banez et al., 2014).

Table 3
Outcomes including means, standard deviations, and t-tests for outcome variables

	Admission Mean (*SD*)	**Discharge Mean (*SD*)**	***t*-test**	***p*-value**
Pain Score (0-10 scale) (Positive change score = increase in pain)	4 (2.4)	5.7 (2.6)	1.87	0.071
LEFS (0-80)	41.6 (15.9)	57.3 (15.8)	2.79	0.009***
UEFI (0-80)	52.2 (12.7)	67.6 (10.2)	3.66	0.001***
Objective Physical Activities				
Kneeling Push Ups (repetitions)	17 (9.3)	27.7 (12.6)	2.52	0.018**
Step Ups (repetitions)	21.3 (10.3)	33.3 (13.2)	2.77	0.01**
Box Carry (20' intervals)	9.4 (3.3)	12.4(3.1)	2.26	0.033**
Elbow Planks (seconds)	35.5 (23.1)	53.7 (22)	2.03	0.053

**$p \leq .01$, ***$p \leq .001$

Conclusion

It is important to recognize that patients with a diagnosis of POTS can tolerate and benefit from participation in an intensive rehabilitation program, but there are many elements of treatment that may facilitate or inhibit gains. An appropriate exercise progression, verbal education about key aspects of physical activity, and allowing the adolescent to help make progression decisions are important. In addition, looking at perception of physical gains and abilities, along with actual physical function, are important as both serve different purposes and are separate constructs in the rehabilitation process (Kempert et al., 2017). With the incorporation of educational activities, including concepts to help slowly increase activity level, tolerate increased symptoms while maintaining activity level, and better ability to pace, participants have demonstrated improved physical abilities after their intensive rehabilitation program participation. Future research efforts regarding POTS may benefit from the use of additional psychosocial measures, along with subjective and objective function-related data, to identify further characteristics or beliefs that impact participation and outcomes. In addition, long-term follow-up data (6+ months) to determine if participants are able to maintain gains, or if not, identify factors associated with a decline in functioning, may be a very helpful addition to existing literature.

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