

FALL 2011 DYSAUTONOMIA news

Coping with Dysautonomia

By DiAnna Fogle

Dealing with Dysautonomia can be challenging and often discouraging. Days will vary and life can be like a roller coaster of events. Along with the physical challenges, there might be emotional challenges. A chronic illness has many stages, as well as many emotions. How we feel physically drastically affects how we are emotionally. When we are diagnosed with an illness, we often feel that we have been shorted in life.

We may feel that our future has been taken away and there is no normal life left. We may be told that we need to accept our illness and resign ourselves to it. Some conclude that if they deny what is wrong, the symptoms will go away. However, acceptance can be a positive step towards a better life. We can have good, productive lives, even with this physical challenge.

It is certainly challenging when we can't know the outcome of a diagnosis. Who it will affect? Can you still work? What about the expenses? How can I take care of my family? All these questions and more are normal. Some might begin to isolate themselves from others. Perhaps it is embarrassing or perhaps the lack of independence (even though it might be temporary) is disheartening. On occasion, we can be too proud to ask for help and this may result in feelings of frustration.

There might be feelings of anger as you grieve a lost life and the feeling that everything must change. This anger can take over our emotions and strain relations with family and friends when those relationships are most needed. But hopefully, as time goes by, we learn new ways to survive the day and adapt to new ways of living. We may begin choosing our friends by how they react to our situation.

We begin accepting help from our families. Life is going back in the right direction. Friends and family begin to feel better about the situation, and we begin getting comfortable and celebrating the good days.

Then comes a difficult day, and life takes three steps back. Everyone's life is shaken up again. The depression comes back, but we aren't giving up. We accept that being depressed today is an acceptable feeling, and is not an act of self pity. It is the reality of what is wrong with us. We accept it, and set a better goal for tomorrow. Some things in our lives are manageable and within our reach. Having a chronic illness does not mean you can't have goals. Learn to set reasonable expectations. Changing how to something might help and all of a sudden it is accomplished.

Learn to be creative by thinking about something that is difficult to accomplish and try to find a unique way to accomplish it. Never, of course, to the point of exhaustion. Never give up hope. We are not alone. Turn the negative energy and anger into something positive and accomplish it. Energy is energy, so use it wisely. Try to get social again. Don't let this illness constrict and suffocate what is inside. We all had good qualities before we became ill and those have not disappeared. Restore those qualities and use them to help others.

Now is the time you can share with others positive ways to live life, and let others know they are not alone. Now is the time to think of others and focus on the encouragement we can give. When we bring others into our lives, we have less time to dwell on our struggles. It creates positive energy and a feeling of wholeness and well being. Begin to set short term goals and strive to reach them. No one knows what their future holds

[continued, click here](#)

in this issue...

Coping with Dysautonomia	1-2
Research in Review	2-4
Doctor's Corner Q&A	4-5

DINET
Dysautonomia Information Network

Dysautonomia News is a quarterly publication of the Dysautonomia Information Network. Subscribe to Dysautonomia News at www.dinet.org/join.php

Dysautonomia News exists to inform and educate. The content should not be used as a substitute for professional medical advice, diagnosis or treatment. Readers are encouraged to confirm all information with other sources and a physician. Please keep in mind that research is evolving and future discoveries may change or disprove some currently held beliefs.

and in that respect, we have the same opportunity as others. Make your goals attainable. Learn to live every day as fully as possible. share with others positive ways to live life, and let others know they are not alone. Now is the time to think of others and focus on the encouragement you can give to them. When you bring others into your life, you have less time to dwell on your struggles. It creates positive energy within yourself, and a feeling of wholeness and well being. Begin to set short term goals and strive to reach them. No one knows what their future holds and in that respect, we have the same opportunity as others. Make your goals attainable. Learn to live every minute of every day. Accept and enjoy each good moment you have. You never know when the next good one will come. Believe in yourself and your future. There is a future for you. Decide to share it with someone, it makes it special and memorable. Life has not shorted you in anyway, It gave you a little more opportunity in life, more challenges, more accomplishments and more goals to achieve. Enjoy, be positive, be productive and be happy! Life is good!

Meet the Member

Please tell us your story. It is amazing how one person's story can inspire or educate. Kristina, our "Meet The Member" volunteer is interested in hearing from you! We all are.

If you would be interested in being interviewed by Kristina, email her at kmgundersen@gmail.com

RESEARCH IN REVIEW

YOUR SOURCE FOR CURRENT DYSAUTONOMIA RESEARCH!

CUTANEOUS CONSTITUTIVE NITRIC OXIDE SYNTHASE ACTIVATION IN POSTURAL TACHYCARDIA SYNDROME WITH SPLANCHNIC HYPEREMIA.

Stewart JM, Nafday A, Ocon AJ, Terilli C, Medow MS. Am J Physiol Heart Circ Physiol. 2011 Jun 3. [Epub ahead of print]

Models of microgravity are linked to excessive constitutive nitric oxide synthase (NOS), splanchnic vasodilation and orthostatic intolerance (OI). Normal flow postural tachycardia syndrome (POTS) is a form of chronic OI associated with splanchnic hyperemia. To test the hypothesis that there is excessive constitutive NOS in POTS, we determined whether cutaneous microvascular neuronal NO (nNO) and endothelial NO (eNO) are increased. We performed two sets of experiments in POTS and control subjects aged 21.4±2 years. We used laser Doppler flowmetry to measure the cutaneous response to local heating as an indicator of bioavailable nNO. To test for bioavailable eNO, we infused intradermal acetylcholine through intradermal microdialysis catheters and used the selective neuronal NOS inhibitor L-N()-Nitroarginine-2,4-L-diamino-butyric amide (N(), 10 mM), the selective inducible NOS inhibitor aminoguanidine (AG, 10 mM), the non-specific NOS inhibitor nitro-L-arginine (NLA 10mM), or Ringer's solution. The acetylcholine dose-response and the NO-dependent plateau of the local heating response were increased in POTS compared to control subjects. The local heating plateau was significantly higher, 98±1 %CVCmax in POTS compared to 88±2 %CVCmax in control, but decreased to the same level with N() (46±5 %CVCmax in POTS compared to 49±4 %CVCmax in control) or with NLA (45±3 %CVCmax in POTS compared to 47±4 %CVCmax in control). Only NLA blunted the acetylcholine dose-response indicating that NO produced by endothelial NOS was released by acetylcholine. AG was without effect. This is consistent with increased endothelial and neuronal NOS activity in normal flow POTS.
PMID: 21642500

ASCORBATE IMPROVES CIRCULATION IN POSTURAL TACHYCARDIA SYNDROME.

Stewart JM, Ocon AJ, Medow MS. Am J Physiol Heart Circ Physiol. 2011 May 27. [Epub ahead of print]

Low flow postural tachycardia syndrome (LFP) is associated with vasoconstriction, reduced cardiac output, increased plasma angiotensin-II, reduced bioavailable nitric oxide (NO), and oxidative stress. We tested whether ascorbate would improve cutaneous NO and reduce vasoconstriction when delivered systemically. We used local cutaneous heating to 42°C and laser Doppler Flowmetry to assess NO-dependent conductance (%CVCmax) to sodium ascorbate and the systemic hemodynamic response to ascorbic acid in 11 LFP patients and in 8 control subjects (aged 23±2 years). We perfused intradermal microdialysis catheters with sodium ascorbate 10mM, or Ringer solution. Pre-drug heat response was reduced in LFP, particularly the NO-dependent plateau phase (56±6 vs 88±7 %CVC(max)). Ascorbate increased baseline skin flow in LFP and control subjects and increased the LFP plateau response (82±6 vs 92±6 control). Systemic infusion experiments used Finometer and ModelFlow to estimate relative cardiac index (CI) along with forearm and calf venous occlusion plethysmography to estimate blood flows, peripheral arterial and venous resistances, and capacitance before and after infusing ascorbic acid. CI increased 40% after ascorbate as did peripheral flows. Peripheral resistances were increased (nearly double control) and decreased by nearly 50% after ascorbate. Calf capacitance and venous resistance were decreased compared to control but normalized with ascorbate. These data provide experimental support for the concept that oxidative stress and reduced NO possibly contribute to vasoconstriction and venoconstriction of LFP.
PMID: 21622825

ORTHOSTATIC HEADACHE WITH TACHYCARDIA.

Mathys J, Beiser I, Maurer G, Humm AM. Praxis (Bern 1994). 2011 May 11;100(10):613-616.

We report the case of a 17 year old male patient who presented with a history of orthostatic headache (present in the upright position only) for several months. The diagnostic investigations (MRI of the head and of the spine, lumbar puncture) revealed no signs of an intracranial hypotension or a CSF leak. In standing position, a significant raise of the heart rate (>40 bpm) without fall of the blood pressure occurred together with a bilateral, pressure-like headache. A diagnosis of postural tachycardia syndrome was made. Treatment with increase of fluid and salt intake, elastic compression stockings and regular exercise was successful.

PMID: 21563100

ATRIOVENTRICULAR NODE ABLATION AND PACEMAKER IMPLANTATION FOR RECURRENT SYNCOPE IN A PATIENT WITH POSTURAL TACHYCARDIA SYNDROME (POTS).

Nakatani Y, Mizumaki K, Nishida K, Inoue H. J Cardiovasc Electrophysiol. 2011 May 3. doi: 10.1111/j.1540-8167.2011.02078.x. [Epub ahead of print]

Ablate and Pace for POTS. A 42-year-old woman with postural tachycardia syndrome (POTS) was admitted to our hospital with severe palpitations, light-headedness, and syncope. Several drugs had been administered previously, but all had been discontinued due to intolerable adverse effects or limited efficacy. One of the drugs, the I(f) current inhibitor ivabradine, effectively slowed the patient's heart rate and relieved the symptoms, but was discontinued due to allergy. After unsuccessful sinus node ablation, atrioventricular node ablation and dual chamber pacemaker implantation was performed, which dramatically improved her symptoms and eliminated syncope. Atrioventricular node ablation could modify the cardiac autonomic balance and thereby suppressed the excessive orthostatic sympathetic activity. (J Cardiovasc Electrophysiol, Vol. pp. 1-4).

© 2011 Wiley Periodicals, Inc.

PMID: 21539639

BARORECEPTOR UNLOADING IN POSTURAL TACHYCARDIA SYNDROME AUGMENTS PERIPHERAL CHEMORECEPTOR SENSITIVITY AND DECREASES CENTRAL CHEMORECEPTOR SENSITIVITY.

Taneja I, Medow MS, Clarke DA, Ocon AJ, Stewart JM. Am J Physiol Heart Circ Physiol. 2011 May 2. [Epub ahead of print]

While orthostatic tachycardia is the hallmark of postural tachycardia syndrome (POTS), orthostasis also initiates increased minute ventilation (V(E)) and decreased end-tidal carbon dioxide (ETCO(2)) in many patients. We hypothesized that chemoreflex sensitivity would be increased in patients with POTS. We therefore measured chemoreceptor sensitivity in 20 POTS (16F, 4M) and 14 healthy controls (10F, 4M), 16-35 years old by exposing them to eucapnic hyperoxia (30% oxygen (O(2))), eucapnic hypoxia (10% O(2)) and hypercapnic hyperoxia (30% O(2)+5% carbon dioxide (CO(2))) while supine and during 70° upright tilt (HUT). Heart rate (HR), mean arterial pressure (MAP), O(2) saturation (SaO(2)), ETCO(2), and V(E) were measured. Peripheral chemoreflex sensitivity (PCS) was calculated as the difference in V(E) during hypoxia compared to room air

divided by the change in O(2) saturation. Central chemoreflex sensitivity (CCS) was determined by the difference in V(E) during hypercapnia divided by the change in CO(2). POTS subjects had an increased PCS (L/min/%) in response to hypoxia (0.42±0.38 vs 0.19±0.17) but a decreased CCS (L/min/Torr) CO2 response (0.49±0.38 vs 1.04 ±0.18) compared to controls. CO(2) sensitivity was also reduced in POTS subjects when supine. POTS patients are markedly sensitized to hypoxia when upright but desensitized to CO(2) while upright or supine. The interactions between orthostatic baroreflex unloading and altered chemoreflex sensitivities may explain hyperventilation in POTS.

PMID: 21536847

PYRIDOSTIGMINE IN THE TREATMENT OF POSTURAL ORTHOSTATIC TACHYCARDIA: A SINGLE-CENTER EXPERIENCE.

Kanjwal K, Karabin B, Sheikh M, Elmer L, Kanjwal Y, Saeed B, Grubb BP. Pacing Clin Electrophysiol. 2011 Mar 16. doi: 10.1111/j.1540-8159.2011.03047.x.

Background: The long-term efficacy of pyridostigmine, a reversible acetyl cholinesterase inhibitor, in the treatment of postural orthostatic tachycardia syndrome (POTS) patients remains unclear. We report our retrospective, single-center, long-term experience regarding the efficacy and adverse effect profile of pyridostigmine in the treatment of POTS patients. Methods: This retrospective study included an extensive review of electronic charts and data collection in regards to patient demographics, orthostatic parameters, side-effect profile, subjective response to therapy, as well as laboratory studies recorded at each follow-up visit to our institution's Syncope and Autonomic Disorders Center. The response to pyridostigmine therapy was considered successful if patient had both symptom relief in addition to an objective response in orthostatic hemodynamic parameters (heart rate [HR] and blood pressure). Three hundred patients with POTS were screened for evaluation in this study. Of these 300, 203 patients with POTS who received pyridostigmine therapy were reviewed. Of these 203 patients, 168 were able to tolerate the medication after careful dose titration. The mean follow-up duration in this group of patients was 12 ± 3 (9-15) months. Pyridostigmine improved symptoms of orthostatic intolerance in 88 of 203 (43%) of total patients or 88 of 172 (51%) who were able to tolerate the drug. The symptoms that improved the most included fatigue (55%), palpitations (60%), presyncope (60%), and syncope (48%). Symptom reduction correlated with a statistically significant improvement in upright HR and diastolic blood pressure after treatment with pyridostigmine as compared to their baseline hemodynamic parameters (standing HR 94 ± 19 vs 82 ± 16, P < 0.003, standing diastolic blood pressure 71 ± 11 vs 74 ± 12, P < 0.02). Gastrointestinal problems were the most common adverse effects (n = 39, 19%) reported. The overall efficacy of pyridostigmine in our study was seen in 42% of total patients or 52% of patients who could tolerate taking the drug. Conclusion: The subgroup of POTS patients who can tolerate oral pyridostigmine may demonstrate improvement in their standing HR, standing diastolic blood pressure, and clinical symptoms of orthostatic intolerance. (PACE 2011; 1-6). ©2011, The Authors. Journal compilation ©2011 Wiley Periodicals, Inc.

PMID: 21410722

POTS: An Overview

Click [here](#)

SLEEP DISTURBANCES AND DIMINISHED QUALITY OF LIFE IN POSTURAL TACHYCARDIA SYNDROME.

Bagai K, Song Y, Ling JF, Malow B, Black BK, Biaggioni I, Robertson D, Raj SR. *J Clin Sleep Med*. 2011 Apr 15;7(2):204-10.

STUDY OBJECTIVES:

Patients with postural tachycardia syndrome (POTS) commonly complain of fatigue, unrefreshing sleep, daytime sleepiness, and diminished quality of life. This study's objective was to assess the sleep quality and health-related quality of life in patients with POTS as compared with healthy control subjects.

METHODS:

Patients with POTS (n = 44) and healthy control subjects (n = 46) completed a battery of questionnaires including Medical Outcomes Study (MOS) Sleep Survey and the Epworth Sleepiness Scale to assess sleep, fatigue visual analogue scale (VAS) to assess fatigue, and the RAND-36 and EuroQol (EQ-5D) surveys to assess health-related quality of life.

RESULTS:

Compared with healthy control subjects, patients with POTS have more sleep problems (58 ± 18 vs. 20 ± 13 ; $p < 0.0001$) and excessive daytime sleepiness (10.2 ± 5.7 vs. 6.2 ± 3.2 ; $p < 0.0001$), higher fatigue levels (7.5 ± 2.0 vs. 2.8 ± 2.5 ; $p < 0.0001$), and poor health-related quality of life (EQ-5D health thermometer 53 ± 17 vs. 89 ± 7 ; $p < 0.0001$). There were strong correlations between MOS Sleep Survey index and the fatigue VAS ($R_s = 0.73$; $R(2) = 0.53$; $p < 0.0001$) and the RAND-36 physical health composite scores ($R_s = -0.70$; $R(2) = 0.53$; $p < 0.0001$)

CONCLUSIONS:

Patients with POTS have higher subjective daytime sleepiness, fatigue, and worse sleep and health related quality of life. The sleep problems contribute significantly to the diminished quality of life: 50% of the variability in HRQL can be explained by the variability in sleep problems. Further objective studies are needed to delineate the specific nature of the sleep problems in patients with POTS.

PMID: 21509337

A special thanks to our volunteers for making this newsletter possible:

Michelle Sawicki, **DINET President**

Staci Friedman, **Art Director**

Judith Pettibone, **Editor**

Janie, **Physicians's Q&A & Volunteer Coordinator**

Kristina Gundersen, **Writer**

"**Firewatcher**", **Research Volunteer**

Svetlana Blitshteyn, MD

Satish R Raj, MD, MSCI

Dr. Amer Suleman

Julian M. Stewart, MD, PhD

doctorscorner



Q: Dear Doctors,

Last week my 19 year old daughter had 2 episodes where she passed out and stayed "asleep" for 4-6 hours. She has been feeling very tired and her hands have been very shaky. Day 2, her hands were tingly and numb and legs tingling. When she fainted the second time, her arm continued to tremor. They could rouse her a bit, but she was so tired and lethargic and dead weight that she slept on people's laps (day 1 in NYC on a park bench!) and did not remember being carried to her dorm or to the Dr. She has a definite diagnosis of narcolepsy from a MLST, but takes an anti depressant so cataplexy was not considered. She was hospitalized and had every test possible EXCEPT A TABLE TEST. MRI, CAT, EEG, heart monitor, and blood tests were done and were all negative.

She DID have postural hypotension in High School. She also has night sweats pretty much every night. She weighs about 110 lbs. Takes Nuvigil for the narcolepsy and Zoloft.

Would you recommend having her checked for POTS or any other type of dysautonomia?

Jenny from Pennsylvania

A: This would be unlikely for POTS, you can check her heart rate lying down and standing just to confirm.

Dr. Amer Suleman

Q: Dr. Stewart,

In your research you've found all types of POTS have regional hypovolemia. Does this regional (thoracic and cerebral) hypovolemia cause our bodies to react like we have total low blood volume? What percentage of POTS patients do have total low blood volume?

Todd from New York

A: POTS patients have low central (heart, lungs, central blood vessels) blood volume when they are upright. This can occur because they have low blood volume even when lying down. I think we have found about 20-25% of patients in whom this is true. This can also occur when blood is redistributed excessively to the legs or to the abdominal organs. With low total blood volume or low upright central blood volume patients can develop a reflex tachycardia similar to what happens when you are dehydrated.

Still other patients may have problems controlling heart rate per se. Many of these have reflex tachycardia superimposed on problems controlling the nerves that set heart rate too. Still others with tachycardia while supine may have a similar illness called inappropriate sinus tachycardia.

In all of these problems secondary causes of fast heart rate such as anemia, thyroid disease, pheochromocytoma, Addison's disease and others should be ruled out.

Sincerely,

Julian M. Stewart, MD, PhD
New York Medical College

Q: I have POTS and NCS. The last two viruses I had caused my kidneys to empty out so much of my fluid that I had to hospitalized for several days each time. Both of these episodes happened within 4 months of each other. Can POTS cause the kidneys to expel a large amount of fluid through urine during a virus, but then stop once the virus passes?

Sarah from Texas

A: I do not know if the kidneys truly expel more fluid. What is known is that the body's response to the infection causes an increase in fluid requirements and probably causes you to drink more fluid. Whether you are sick or not, you will void the overwhelming majority of the fluid that you take in.

I think that the most likely scenario is that your fluid requirements went up with your viral infections, and thus left you relatively more

volume depleted. This happens to everyone (with or without POTS), but given that many POTS patients have a low blood volume even at baseline (research is ongoing as to the reasons why this might be so), the relatively increased volume depletion might be less well tolerated. I suspect that you put out a lot of urine whether sick or not, but you noticed it while more ill.

Satish R Raj, MD, MSCI
Vanderbilt University

Q: For so many of us dizziness/cognitive problems is the most disabling symptom. There is some talk in the literature about reduced blood flow to the brain (while standing) in POTS patients. Has this been proven as the cause for our dizziness/brain fog and if so, what can be done about it? Or are there any other explanations for this particular symptom?

Thank you,

Naomi from New Jersey

Cerebral hypoperfusion—i.e. reduced blood flow to the brain - has been demonstrated to result in various symptoms of pre-syncope, such as lightheadedness, dizziness, headache, nausea and lack of concentration. As you point out, cerebral hypoperfusion occurs in an upright position in patients with POTS and other autonomic disorders.

Regarding dizziness and cognitive problems that are experienced by patients while sitting or supine, I am not aware of any specific study addressing the underlying mechanism of these symptoms. Some patients feel significant improvement in their cognitive function in supine vs. sitting position, and therefore are able to do homework, read books and do other cognitively-demanding tasks in a recliner rather than sitting at a desk.

Non-pharmacologic measures, such as elevating your legs while sitting, utilizing compression stockings and using small amounts of caffeine, if tolerated, may be helpful for some patients, while others may benefit from medications, such as Provigil, Nuvigil, Wellbutrin, and others to alleviate cognitive complaints. In general, measures that are used to treat POTS in general are often helpful for diminishing cognitive complaints.

Svetlana Blitshteyn, MD
State University of New York at Buffalo School of Medicine and Biomedical Sciences