A Thank You to Our Volunteers

By Michelle Sawicki

April 17-23rd was National Volunteer Week. I would like to express my sincerest gratitude to the following people for regularly volunteering with DINET:
Jane Bennett
Leader of the Meet Others Program, Jane has spent countless hours connecting dysautonomia patients and caregivers to others in their geographical region.

Linda Bentley
Serving as DINET’s Web site editor, as well as on DINET’s Board of Directors, Linda Bentley is always just a phone call away when we need her expertise.

Valerie Fenston
The person who did all the “behind the scenes” work to build DINET’s Physician List, Valerie has also dedicated her time to updating DINET’s Meet Others Program and support group list. Valerie will soon be mailing brochures containing information on POTS to physicians around the country.

Katherine Munson
Creator of the “Faces of DINET” Web Page, Katherine has helped hundreds of forum members put faces to the names of fellow members.

Judith Pettibone
Voluntarily adding the Dysautonomia News for years, Judith’s service is invaluable.

Marie Kelly-Warden
Assisting in the obtaining of research articles, Marie has helped keep DINET’s website up-to-date.

Pam Reicher
Designer of the Dysautonomia News, Pam devotes her skills to make DINET’s newsletters look great.

Nina Wilde
Forum moderator and support leader, Nina Wilde directly helps DINET’s members every day. Nina solely conducted DINET’s first fundraiser, selling hundreds of bracelets to aid in DINET’s expenses.
THE BROCHURES ARE COMING!
Visit DINET’s Web site in the near future to learn how to send educational brochures to friends, family and physicians.

MY, HOW WE HAVE GROWN....
DINET currently has 675 members, 532 of which participate in the Meet Others Program.

Dysautonomia News is a quarterly publication of the Dysautonomia Information Network. Subscribe to Dysautonomia News at www.dinet.org/join.htm
Drinking water could be beneficial to patients with low blood pressure

Ordinary tap or bottled water could help people suffering from low blood pressure who faint while standing, claim researchers from Imperial College London and St Mary's Hospital.

According to research published in the latest issue of Journal of Neurology, Neurosurgery and Psychiatry, drinking two glasses of water can raise blood pressure, potentially providing a solution for patients with low blood pressure while standing, caused by autonomic failure. Autonomic failure is where parts of the nervous system, responsible for the control of bodily functions not consciously directed, such as blood pressure, heart rate and sweating, do not function properly.

Professor Christopher Mathias, from Imperial College London and St Mary's Hospital, and the senior author of the research, comments: "This surprising discovery that water can have such an effect on blood pressure could help us to treat both sufferers of autonomic failure, and many people who suffer from low blood pressure generally, especially those who faint, such as with vasovagal syncope."

The researchers looked at 14 patients with autonomic failure, and measured their blood pressure while lying and standing, before, and 15 and 35 minutes after drinking 480ml of distilled water. When asked to stand, before drinking water, this caused a fall in blood pressure.

The 14 patients were divided into two groups, seven of whom had multiple system atrophy (MSA), while the other seven had pure autonomic failure (PAF). MSA is a neurodegenerative disease marked by a combination of symptoms affecting movement, blood pressure, and other body functions. PAF is a disorder affecting only the autonomic nervous system. They both often present in middle to late life.

The patients then drank water causing a significant rise in blood pressure. For the patients with PAF it took five minutes for a significant rise in blood pressure to be recorded, and for patients with MSA it took 13 minutes. In both the fall in blood pressure and symptoms of low blood pressure, was reduced while standing.

Professor Mathias adds: "While autonomic failure itself is generally not life threatening, it can have a significant impact on an individual's quality of life. People with low blood pressure caused by autonomic failure are at a greater risk of fainting when standing upright, after food or even after mild exertion. This can affect their life in many ways, stopping them from driving, or in extreme cases, from being able to work. This discovery could be of considerable use in helping these patients to understand why this happens. It may also benefit the many without autonomic failure who faint as a result of low blood pressure."

The research was supported by a grant from the Sarah Matheson Trust Autonomic Disorders Association.
Research in Review


Treatment of pediatric chronic pain with tramadol hydrochloride: siblings with Ehlers-Danlos syndrome - Hypermobility type.

Brown SC, Stinson J.
Department of Anaesthesia, The Hospital for Sick Children, Toronto, Ontario.
stephen.brown@sickkids.ca

OBJECTIVE: To evaluate the effectiveness of tramadol hydrochloride for the treatment of chronic pain refractory to previous treatment in two pediatric patients.

METHODS: Tramadol hydrochloride was administered (50 mg/day to 150 mg/day) to two siblings with Ehlers-Danlos syndrome - Hypermobility type refractory to previous pharmacological treatments, and changes in pain intensity and physical activity were assessed.

RESULTS: Pain intensity decreased and physical activity improved within days of starting therapy. Positive results have been maintained for 30 months.

CONCLUSIONS: Tramadol hydrochloride was a safe and effective treatment for relieving chronic pain in two pediatric patients suffering from the hypermobility type of Ehlers-Danlos syndrome. No morbidity or side effects were noted during the 30-month follow-up.

PMID: 15605135

Gastrointestinal symptoms associated with orthostatic intolerance.

Sullivan SD, Hanauer J, Rowe PC, Barron DF, Darbari A, Oliva-Hemker M.
Departments of Pediatric Gastroenterology and Nutrition and Pediatrics, The Johns Hopkins University School of Medicine, The Johns Hopkins University, Baltimore, Maryland.

BACKGROUND: The term orthostatic intolerance is used to describe symptoms of hemodynamic instability such as lightheadedness, fatigue, impaired cognition and syncope that develop on assuming an upright posture. Common forms of orthostatic intolerance in childhood include postural tachycardia syndrome and neurally mediated hypotension.

OBJECTIVE: A descriptive report of the clinical characteristics of patients presenting with gastrointestinal symptoms who are ultimately found to have orthostatic intolerance.

METHODS: A medical record review of all patients referred to the pediatric gastroenterology service at the Johns Hopkins Children’s Center who had an abnormal tilt table test between June 1996 and December 2000.

RESULTS: Of 24 eligible subjects aged 9-17 years (mean, 14.3 years), four had postural tachycardia syndrome, eight had both postural tachycardia and neurally mediated hypotension, and 12 had neurally mediated hypotension alone. The most common presenting gastrointestinal symptoms were abdominal pain, nausea and vomiting. Median number of gastrointestinal symptoms per patient was 3 (range, 1-7), and 87% of the patients experienced gastrointestinal symptoms for more than 1 year and 48% experienced gastrointestinal symptoms for more than 3 years. Follow-up information was available on 18 patients. Seventy-eight percent of patients (14 of 18) had complete resolution of symptoms with treatment of...
CONCLUSION: Pediatric patients with chronic upper gastrointestinal symptoms may have underlying orthostatic intolerance. In patients with upper gastrointestinal symptoms and orthostatic intolerance, treatment of orthostatic intolerance may result in resolution of gastrointestinal symptoms.

PMID: 15795588


Renin-aldosterone paradox and perturbed blood volume regulation underlying postural tachycardia syndrome.

Raj SR, Biaggioni I, Yamhure PC, Black BK, Paranjape SY, Byrne DW, Robertson D.

Division of Clinical Pharmacology, Vanderbilt University, Nashville, Tenn 37232-2195, USA. satish.raj@vanderbilt.edu

BACKGROUND: Patients with postural tachycardia syndrome (POTS) experience considerable disability, but in most, the pathophysiology remains obscure. Plasma volume disturbances have been implicated in some patients. We prospectively tested the hypothesis that patients with POTS are hypovolemic compared with healthy controls and explored the role of plasma renin activity and aldosterone in the regulation of plasma volume.

METHODS AND RESULTS: Patients with POTS (n=15) and healthy controls (n=14) underwent investigation. Heart rate (HR), blood pressure (BP), plasma renin activity, and aldosterone were measured with patients both supine and upright. Blood volumes were measured with 131I-labeled albumin and hematocrit. Patients with POTS had a higher orthostatic increase in HR than controls (51+/-18 versus 16+/-10 bpm, P<0.001). Patients with POTS had a greater deficit in plasma volume (334+/-187 versus 10+/-250 mL, P<0.001), red blood cell volume (356+/-128 versus 218+/-140 mL, P=0.010), and total blood volume (689+/-270 versus 228+/-353 mL, P<0.001) than controls. Despite the lower plasma volume in patients with POTS, there was not a compensatory increase in plasma renin activity (0.79+/-0.58 versus 0.79+/-0.74 ng x mL(-1) x h(-1), P=0.996). There was a paradoxically low level of aldosterone in the patients with POTS (190+/-140 pmol/L versus 380+/-230 pmol/L; P=0.017).

CONCLUSIONS: Patients with POTS have paradoxically unchanged plasma renin activity and low aldosterone given their marked reduction in plasma volume. These patients also have a significant red blood cell volume deficit, which is regulated by the renal hormone erythropoietin. These abnormalities suggest that the kidney may play a key role in the pathophysiology of POTS.

PMID: 15781744


Hyperadrenergic postural tachycardia syndrome in mast cell activation disorders.

Shibao C, Arzubiaga C, Roberts LJ 2nd, Raj S, Black B, Harris P, Biaggioni I.

Division of Clinical Pharmacology, Department of Medicine and Pharmacology, and the Autonomic Dysfunction Center, Vanderbilt University School of Medicine, Nashville, Tenn 37212, USA.

Postural tachycardia syndrome (POTS) is a disabling condition that commonly affects otherwise normal young females. Because these patients can present with a flushing disorder, we hypothesized that mast cell activation (MCA) can contribute to its pathogenesis. Here we describe POTS patients with MCA (MCA+POTS), diagnosed by episodes of flushing and abnormal increases in urine methylhistamine, and compared them to POTS patients with episodic flushing but normal urine methylhistamine and to normal healthy age-matched female controls. MCA+POTS patients were characterized by episodes of flushing, shortness of breath, headache, lightheadedness, excessive diuresis, and gastrointestinal symptoms such as diarrhea, nausea, and vomiting. Triggering
events include long-term standing, exercise, premenstrual cycle, meals, and sexual intercourse. In addition, patients were disabled by orthostatic intolerance and a characteristic hyperadrenergic response to posture, with orthostatic tachycardia (from 79+/−4 to 114+/−6 bpm), increased systolic blood pressure on standing (from 117+/−5 to 126+/−7 mm Hg versus no change in POTS controls), increased systolic blood pressure at the end of phase II of the Valsalva maneuver (157+/−12 versus 117+/−9 in normal controls and 119+/−7 mm Hg in POTS; P=0.048), and an exaggerated phase IV blood pressure overshoot (50+/−10 versus 17+/−3 mm Hg in normal controls; P<0.05). In conclusion, MCA should be considered in patients with POTS presenting with flushing. These patients often present with a typical hyperadrenergic response, but beta-blockers should be used with great caution, if at all, and treatment directed against mast cell mediators may be required.

PMID: 15710782

Research abstracts obtained from PubMed
The Patient's Voice

Views expressed in The Patient’s Voice are not necessarily those of the Dysautonomia Information Network or its members.

Share your experiences in The Patient's Voice!

The Patient’s Voice is a newsletter column where patients can express themselves and write about experiences relating to dysautonomia - both positive and negative. It is a place to share medical experiences, suggestions, short stories and poetry, etc. Send contributions to: staff@dinet.org

The top 10 reasons POTS sufferers don't date:

By Dan Jacoby

#10. Your date might think they're too good looking if he or she shows up and you faint after answering the door.
# 9. It's no fun having your blood pressure go down during your first goodnight kiss.
# 8. Your date thinks you always need to use the bathroom because you can't stop crossing your legs.
# 7. When you finally get the nerve to tell them you have POTS, your date says, "Let's light one up and party!"
# 6. You're tired of having one of your dates cry because when you say, "I need to sit down" he or she thinks you're going to dump 'em.
# 5. Florinef is not Viagra.
# 4. You're tired of telling them you have autonomic failure and they recommend a good transmission repairman.
# 3. You're afraid there will be one too many "standing ovations" at the show you're supposed to go to.
# 2. Your heart races even if they're ugly.
# 1. You can't take being stood up.

With Sincere Gratitude to Professor M. Esler

By Tracey

I have been diagnosed with autonomic dysfunction of the sympathetic nervous system. I have numerous symptoms which include very low blood pressure, severe tachycardia, tiredness, weakness, severe headaches, very high blood pressure at times and shakes. I also have lipomas all over my body and a series of other very annoying and painful symptoms, as well as a small pituitary adenoma.

I feel the worst when my blood pressure falls too low and the adrenaline kicks in. It’s a terrible feeling which gives me the sensation of severe anxiety. I have many other health problems, too many to list. However, there is hope and for anyone who reads this, after years of suffering and being treated like a neurotic, I was finally sent to see heart specialist Professor Murray Esler at the Heart Centre, in the Alfred Hospital in Melbourne.

I would recommend Professor Esler to everyone. He is one of the most caring, dedicated doctors I have ever had the pleasure of meeting. He believed in me. My case has been very difficult. I not only have numerous medical problems, but I am overly sensitive to medications. Despite the challenges, he has never failed me. He has looked at the bigger picture, referring me on to dermatologists and neurologists, as they all try to put the puzzle pieces together.

At the moment I am waiting to see if I have an extremely rare disease. Time will tell. I’m only 37 but this has gone on for years. I thank god for Professor Esler doing the tilt table test and finding dysfunction of the sympathetic nervous
Without Professor Esler and his ongoing help, I'd probably be in a nut house by now. He never gives up on you, even though he is an incredibly busy man. He has a very rare quality, which is compassion, and I can never thank him enough for his help, time and patience. I hope this helps people believe that there is hope when you think all hope is gone. There are incredible doctors, you just have to keep believing in yourself, listen to your body and never quit searching until you find someone like Professor M. Esler. Just look him up on the Internet and you will see what I mean.

This is with sincere gratitude to Professor M. Esler.

Good luck everyone,

Tracey
Hi all,
I hope you are OK. Things are really great here. I am getting stronger all the time and walking more and more... I can now take a few steps without any support and walk a short distance on crutches. My treatment is going good, but I’m still waiting for my new port. I hope it will be in soon.

I go sit skiing a lot now at Norfolk Ski Slope. It is great fun and a good way of meeting others. I also have started sailing, which is brill!!!

Emma