

POTS Recommendations



What is POTS?

- Postural Orthostatic Tachycardia Syndrome
- Your heart speeds up when you stand up!
- Increased work of the heart when changing positions

Diet

- Fluids, fluids, fluids – Drink at least a GALLON of water daily (128 oz or 3.7 L)!
- Sodium rich diet (~10-12 g or ~2 tsp of salt daily)
- Potassium rich diet:
 - Younger than 8 years: 3800 mg daily (3/4 tsp daily)
 - Older than 8 years: 4500-4700 mg daily (approximately 1 tsp daily)
- Try some of these potassium rich foods: Bananas, cantaloupe, kiwis, oranges, strawberries, grapes, peaches, apples, pineapple, potatoes, acorn squash, sweet potatoes, dried apricots, dried peaches, dried figs, kidney beans, white beans, lima beans, soybeans, tomatoes, avocados, dark leafy greens, mushrooms, salmon, tuna, halibut, flounder, cod, lean beef, chicken, turkey, milk and yogurt
- Beverages with electrolytes: e.g. Pedialyte or other sports drinks without sugar
- NEVER skip breakfast – have some PROTEIN
- Avoid caffeine, energy drinks and decongestants



Exercise



- AEROBIC – such as brisk walking or swimming
- Helps to improve circulation
- When standing, move legs frequently to increase circulation; when sitting, elevate, flex and move feet
- Strengthen lower limbs & abdominal muscles – helps to avoid pooling of blood when standing
- Exercise regularly, 30 minutes every other day

Attitude

- Stay POSITIVE – you may have “good” and “bad” days
- Think of the glass half-FULL not half-empty
- Have PATIENCE – patient AND parents
- Counseling for life stressors
- Most POTS patients show improvement of their symptoms with treatment and become functional – returning to normal or near normal



Medications (for a little extra help)



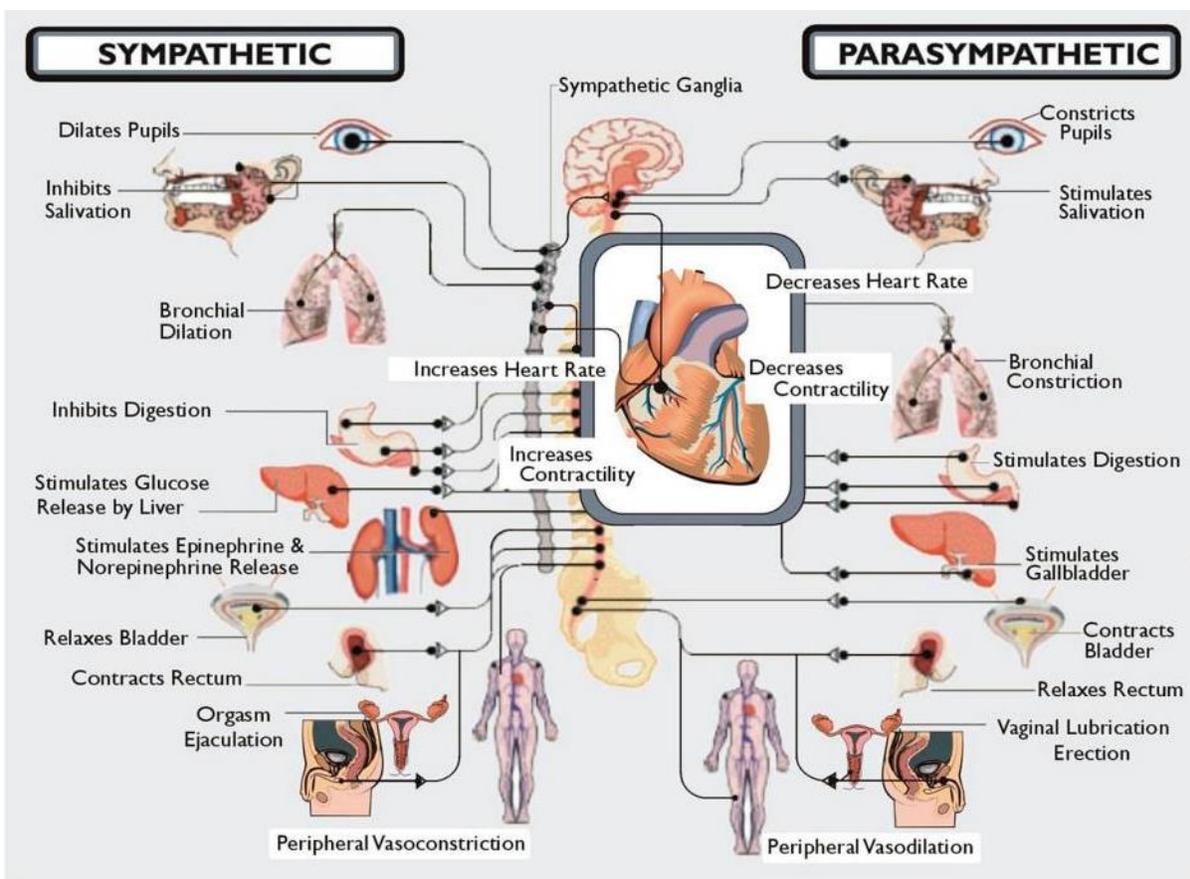
- Fludrocortisone (Florinef) – helps with volume distribution
- Midodrine (Alpha 1 agonist) – helps perfusion through constricting blood vessels
- Beta blocker medications – helps to slow the heart rate
- Clonidine – helps sleep disturbances
- Pyridostigmine (Mestinon) – helps treat GI symptoms & fatigue
- Vasopressin (DDAVP) – helps to retain free water but not sodium
- Selective serotonin re-uptake inhibitors (SSRIs)/Serotonin and norepinephrine re-uptake inhibitors (SNRIs) – medications (e.g. Prozac, Zoloft, Paxil, Celexa or Cymbalta)

Dysautonomia Disorder or Postural Orthostatic Tachycardia Syndrome "POTS"

In this disorder of dysautonomia, the usual antigravity reflexes (Orthostatic intolerance) are somewhat inadequate to maintain preload and systemic blood pressure with prolonged or sudden standing. While the hallmark of POTS is an excessive heart rate increment upon standing (Orthostatic Tachycardia), (i.e. >30 bpm above baseline or >120 bpm at any point), patients often exhibit numerous symptoms of autonomic nervous system dysregulation.

The autonomic nervous system regulates certain body functions that we cannot control, such as blood pressure and the rate of breathing. This system works automatically (autonomously), without a person's conscious effort.

Disorders of the autonomic nervous system can affect any body part or process. Autonomic disorders may result from other disorders that damage autonomic nerves or they may occur on their own. Autonomic disorders may be reversible or progressive.



Symptoms of POTS:

Patients with POTS may have variable symptoms ranging from simple lightheadedness to severe symptoms that may restrict daily activities making the patient disable and home-bound. The patients often complain of extreme fatigue, exercise intolerance, tachycardia (rapid heartbeat), palpitations (pounding chest), chest discomfort, shortness of breath, dizziness, headaches and mental clouding "brain fog", blurred or tunnel vision, nausea and abdominal pain.

POTS can sometimes arise a constitutional trait, and in some situations there is a family history of this condition. There are some reports suggesting an association between joint hypermobility, irritable bowel syndrome and POTS. There is an overlap between POTS and Anxiety, depression and panic attacks and sometime can be confused with these conditions.

Facts about POTS:

- Prevalence ~ 0.2%
- Presentation often between ages 15-25 years
- Female predominant (>75%). The female: male ratio is 4:1
- Clinical overlap with Chronic Fatigue Syndrome
- Markedly reduced Quality of Life
- **No reported Mortality.** In most cases (approximately 80%), an individual with POTS improves and becomes functional, although some residual symptoms are common.

Diagnosis of POTS:

There is no a specific test to diagnose POTS. It is usually a diagnosis by exclusion. The current diagnostic criteria for POTS is a heart rate increase of 30 beats per minute (bpm) or more, or over 120 bpm, within the first 10 minutes of standing, in the absence of orthostatic hypotension.

Generally, cardiac work up including obtaining blood pressure and heart response in different positions, EKG, echocardiogram and Stress test. Patients with suspected POTS may need to have autonomic function testing including Tilt table testing with heart rate variability, Valsalva maneuver, Quantitative Sudomotor Axon Reflex Test (QSART, sometimes called Q-Sweat), Thermoregulatory Sweat test (TST) and Gastric Motility Studies. In some cases, supine/upright catecholamine testing, blood volume analysis with a radio tracer and skin biopsies to screen for sudomotor and sensory small fiber neuropathy may be used. Extensive blood work may also be necessary to search for underlying or contributing conditions such as electrolytes imbalances, anemia, thyroid disease, liver or kidney issues, immunodeficiencies, autoimmunity, Ehlers-Danlos syndrome or mast cell disorders.

POTS – Possible Mechanisms:

- Partial (peripheral) Autonomic Neuropathy.
- Abnormal Connective Tissue (e.g. Ehlers-Danlos Syndrome)
- Hypovolemia.
- Hyperadrenergic State.
- Deconditioning.
- Hypervigilance/Anxiety.

Triggering Factors of POTS:

The symptoms can be precipitated by any of these factors: viral illness, surgery, sleep deprivation, dehydration, climate changes, altitude changes, external stressors or even hormonal changes “during puberty and menstrual cycles”, suggestive of a possible autoimmune process. POTS may follow a relapsing-remitting course, in which symptoms come and go, for years. Symptoms of POTS may fluctuate and patients may have "good days" and "bad days", which can change periodically depending on precipitating factors, making the management sometimes very difficult and requiring lots of patience from the patient and caregiver.

Management of POTS:

Multidisciplinary approach is usually required including but not limited to a neurologist, cardiologist, psychologist/therapist, nutritionist, gastroenterologist, physical therapist, and personal trainer. Sleep study may also be needed in some patients to manage their sleep disturbances.

In general, therapies for POTS are targeted at relieving low blood volume or regulating circulatory problems that could be causing this condition. The mainstay therapy is volume expansion through voluntary intake of fluids and salt. Drinking 16 ounces of water (2 glassfuls) before getting up can also help raise blood pressure. Avoidance of aggravating factors such as dehydration, stresses, sleep deprivation and extreme heat exposure is also recommended. Having a positive feeling and biofeedback appears to be very helpful in the management of this condition. Think of the glass “half full not half empty”.

Recommendations for POTS: “Lifestyle Changes”

- Increase fluid consumption- at least a gallon of water a day (128 Oz or 3.7 L daily) - half of that amount should be with beverages that contain electrolytes (such as Pedialyte powder because of the high electrolyte content).
- Increase salt (~10-12 g “~ 2 teaspoons” daily of salt) such as salty soup. Consume extra salt while exercising (e.g. salty pretzels).
- Increase intake of potassium rich diet- helps retain fluids, redistribute the volume and prevents muscle cramping.
- Avoid energy drinks, caffeinated beverages and over-the-counter cough or cold medications with a decongestant (that overly stimulate the heart, such as sympathomimetic drugs). You may use allergy medications without decongestants (without the "D") and home remedies.
- Increase regular, aerobic exercise- to help mobilizing the circulation and enhancing vascular tone (i.e. Brisk walking and swimming, etc.).
- Establish exercises to strengthen lower limb and abdominal muscles so as to minimize lower limb and abdominal pooling of blood volume.
- Elevation of the head of the bed 4-6 inches.
- Consider the use of compression stockings and garments or abdominal binders may help reduce the pooling of blood in your legs and reduce the symptoms.
- While sitting and watching TV, elevate, flex and move feet.
- Avoid prolong standing/sitting and change positions slowly. Move your legs while standing to encourage blood to flow from your legs to your heart.
- When starting to feel dizzy or about to black out, lay on the ground and elevate your legs. This will increase blood flow to the brain which will prevent passing out/syncope.
- Incorporate positive feeling and biofeedback into lifestyle.
- Practice breathing relaxation techniques. (visit this website: www.Dawnbuse.com).
- Eliminate external stresses.
- Pursing enjoyable activities as this may help to keep you from focusing on your symptoms.
- Have enough sleep. Sleep deprivation may exacerbate POTS symptoms.
- Search the web for Sleep Hygiene for tips for better sleep.

Medications used to treat POTS:

There is no universally effective medication to treat this condition. Sometimes it is necessary to try different medications at different dosages, sometimes in conjunction with each other in order to treat this condition. We discussed the importance of lifestyle measures in the treatment of this condition. From a pharmacologic standpoint, there are some of the medications, which can be used in POTS including but not limited to:

- **Fludrocortisone (Florinef)**; which helps volume expansion through enhanced sodium retention. Side effects may include hypokalemia, hypomagnesemia, worsening headaches, acne and fluid retention with edema.
- **Midodrine (Alpha 1 agonist)**; which helps perfusion through its vasoconstrictive effect. It is a short-acting agent, thus given 3-4 times daily during waking hours. Should not be taken 3-4 hours before going to bed to avoid supine hypertension. It causes tingling sensation or “goosebumps”.
- **Beta blockers (Atenolol, Propranolol, Nadolol)**; in low doses; which may help to slow heart rate by blunting the tachycardia associated with POTS and may provide symptomatic relief. Avoid higher dose, which may drop blood pressure.
- **Clonidine**; which in some situations can be helpful especially for associated sleep disturbances.
- **Pyridostigmine (Mestinon)**; which can be helpful in some patients to treat gastrointestinal symptoms and symptoms of fatigue.
- **Vasopressin (DDAVP)**; as a volume expanding agent to help in retention of free water but not sodium.
- **Selective serotonin re-uptake inhibitors (SSRIs)/Serotonin and norepinephrine re-uptake inhibitors (SNRIs)**; medications (e.g. Prozac, Zoloft, Paxil, Celexa or Cymbalta).

Non-pharmacological therapy of POTS:

- **Physical Therapy:** May help to mobilize the circulation by improving joints movements.

- **Importance of Exercise in POTS:** Short-term exercise training in POTS may help in the followings:

- increases fitness level
- increases blood volume
- enhances cardiac remodeling
- normalizes sympathetic activity
- decreases orthostatic tachycardia
- improves Quality of Life

- **Recommendations for Exercise in POTS:**

- Focus on Aerobic Activity: some resistance training focused on thighs
- Must be regular: every other day (4/week)
- Duration: 30 min/session
- Avoid Upright Exercises: use Rowing/Elliptical machines, Recumbent Cycles and Swimming

Keep in mind that it may take 4-5 weeks to start seeing benefits of regular exercise

- **Intravenous fluids (IV fluids):**

There is currently no consensus in the medical community on the utility of IV saline in people with dysautonomia disorder (POTS). This mode of therapy may help expanding the blood volume by flushing the veins directly. The main reason in using IV fluid therapy is accessing the bloodstream. When a person has low blood volume, their veins tend to be small and difficult to access. Some physicians will prescribe daily or weekly use of IV saline in treatment of POTS in some patients. Many people with POTS will use a PICC line or port a couple of times a week for a liter or two of saline. The decision to use regular IVs, PICC lines and ports must be made by the treating physician and depends many factors.

Majority of POTS patients are hypovolemic despite adequate hydration. Standard blood and urine tests may not detect this hypovolemia, as the patient is typically deficient in plasma and RBCs. Blood volume analysis with a radio-tracer can be used to evaluate a POTS patient for hypovolemia. ***In POTS, both the number of red blood cells and the plasma volume can be low (and thus the ratio appears normal on the CBC).*** This can be difficult for doctors to test because it is a ratio of red blood cells to plasma volume. For some with POTS, their blood levels can drop low enough to result in hypovolemic shock. Therefore, intravenous saline is used to treat significantly low blood volume, which can be a problem for some people with POTS.

In some cases, nausea and vomiting when drinking liquids defeats the purpose and can severely limit fluid intake. In others with rapid or slow gut motility, less water is absorbed into the body than normal as it is rushed through the intestines. In addition, if the blood volume is low, but the ratio between red blood cells and plasma is normal, you may not be able to absorb the fluids even if you are drinking several liters per day. Intravenous fluids bypass the digestive tract to eliminate issues with nausea/vomiting and gut motility. The saline goes directly into the bloodstream to increase the plasma volume. In fact, a liter of normal saline can reduce both heart rate and other symptoms like brain fog in a person with POTS if the blood volume is low.

- **Intravenous Immunoglobulin (IVIG):**

IVIG has been used to treat dysautonomia disorder (POTS) successfully. There are case reports and peer reviewed research studies in medical journals on this topic.

- **Sleep Hygiene:** Optimizing sleep is very helpful remedy to improve POTS symptoms. Referral for sleep study may be needed.

- **Nutritional Support:** Very important to maintain balanced diet and adequate calories.

- **Mental Counseling:** This may be necessary if the patient shows signs of depression or severe anxiety that may need further psychotropic medications.

- **Positive Biofeedback:** Several different relaxation exercises are used in biofeedback therapy, including:

- Deep breathing
- Progressive muscle relaxation – alternately tightening and then relaxing different muscle groups
- Guided imagery – concentrating on a specific image (such as the color and texture of an orange) to focus your mind and make you feel more relaxed
- Mindfulness meditation – focusing your thoughts and letting go of negative emotions; ALWAYS think positively “the glass is half full not half empty”
- Research websites for guided relaxation practices (i.e. dawnbuse.com)

Prognosis/Outcome of POTS:

In some patients, a cause for this condition is not apparent despite exhaustive diagnostic studies. Reassuringly, many if not most POTS patients (~80%) show improvement of their symptoms and become functional and back to normal or near normal. Therefore, recovery can take quite some time, up to several years.

Postural Orthostatic Tachycardia Syndrome (POTS)

Philip Fischer, MD Mayo Clinic

Thousands of teenagers have a hard time getting moving in the morning. But for kids with POT Syndrome, or POTS, the fatigue is debilitating. It keeps these teens from normal activities like sports or social events. Many can't even go to school. How can you tell if you're teen's tiredness or other related symptoms are a sign of a real problem?

Triggers: Several different sorts of situations seem to be able to trigger an adolescent to develop POTS. Usually, the trigger is an infection, and "mono" is a common trigger in North America. Much of the "mono" we see is caused by the Epstein-Barr virus, but other viruses cause similar "mono-like illness." To receive a diagnosis of "mono" and to have negative "mono" tests usually means that the infection was by a non-Epstein-Barr virus germ that acted like the typical Epstein-Barr virus. Injury occasionally triggers POTS, but this is usually when the injury causes the teenager to be bedridden for several days. It would be very unusual for a minor blow to the back to trigger POTS unless it was so severe that the patient was stuck in bed recovering from the injury for several days.

Symptoms: There are lots of symptoms of POTS, and no single patient has all of them. Fatigue, dizziness, abdominal discomfort, and pains (headache or other) are common. Symptoms are typically, but not always, more obvious when standing still. Other conditions, however, can cause some similar symptoms. So, doctors evaluating teenagers with POTS need to be careful to make sure that an additional concurrent problem isn't present, and they need to remember that some symptoms in POTS patients might be due to something other than the POTS. This means that other causes of fast heart rates need to be ruled out; rarely hormone-secreting tumors can masquerade as POTS. Chest discomfort is seen in a fair number of adolescents with POTS - this can vary from true pain to a feeling of difficulty getting enough air in to pressure to what resembles a "panic attack" (without the panic). These chest symptoms probably relate to altered abdominal and chest blood flow and the chemicals that are associated with panic and depression and anxiety overlap with the chemicals that lead to the manifestations of POTS. At the same time, though, some POTS patients do develop depression and anxiety (very understandable when one has been feeling bad for so long) and might have actual panic attacks (instead of "POTS attacks") causing the chest symptoms. Migraines and chronic daily headache and POTS commonly occur at the same time in the same person - treating the POTS seems to help the headaches improve, too. Other conditions such as ovarian cysts and kidney stones and liver problems would likely be due to something separate from the POTS.

Diagnosis: There are different ways that people classify POTS. Some differentiate between post-infectious POTS and POTS that doesn't seem to follow a typical febrile or mono-like infection. Others separate high flow POTS from low flow POTS based on clever measurements of blood flow with tilting in a research setting. POTS can also be separated by whether there are anti-nerve antibodies in the circulation (seropositive versus seronegative), and these antibodies are found in about 15% of adults with POTS and in fewer adolescents. Others measure adrenaline-like chemicals in standing and lying positions and separate POTS into hyperadrenergic and normal forms. Practically speaking, though, we doctors don't understand POTS well enough yet to use these different categories of POTS in ways that make major changes in treatment.

Treatment: It sounds like several of you have good doctors using helpful medications as well as non-medical means to treat POTS. Increased intake of fluids and salt and having regular aerobic exercise clearly seem to help. Acupuncture and other therapies seem to be associated with improvement in some people but have not been studied well enough to know who all might respond favorably. "Bad days" happen, and the most important way to deal with them is to continue to maximize fluid and salt intake and exercise. Some people suggest avoiding caffeine except as a rare treatment to avert "bad days" - but I don't have experience with this plan to know if it helps - perhaps someone reading this note has tried this and can enlighten the rest of us. If someone hasn't responded well to initial treatment, it is important to continue with medical care and to customize that care to the individual's needs. Using "just" a beta blocker helps many POTS patients, but it is not unusual to need to add or change medications over time.

Prognosis: As best we know, adolescents with POTS usually do well and return to a fully productive life. It seems that about 80% recover completely. Optimism is appropriate!

Additional Patient Resources:

Dysautonomia Information Network

www.dinet.org