

Dysautonomia and Postural Orthostatic Tachycardia Syndrome (POTS)

What is Dysautonomia?

Dysautonomia is a disorder of the autonomic nervous system – the part of the nervous system that is responsible for control of the bodily functions not consciously directed, such as breathing, heart rate and blood pressure control. Dysautonomia results in an imbalance of the sympathetic nervous system (our “fight or flight” responses) and the parasympathetic nervous system (our “rest and digest” responses). Dysfunction of the autonomic nervous system causes many different symptoms in people and may be called different names depending on those symptoms: orthostatic intolerance, POTS, neuro-cardiogenic syncope, or dysautonomia.

What is POTS?

Postural Orthostatic Tachycardia Syndrome (POTS) is a disorder characterized by orthostatic intolerance, a condition that often causes multiple symptoms, especially when standing for a long time. When a person with POTS stands up, there is a significant increase in the heart rate above the normal rate, and the heart rate stays elevated longer than usual. This heart rate response is key to diagnosing POTS. With position changes, blood pressure may fall, stay the same, or increase.

These symptoms are caused by an exaggerated response in the autonomic nervous system. POTS may appear after an illness or vaccination (for example, “mono”, flu vaccine), after trauma (for example, a head concussion), after surgery, or during or after a growth spurt. Other family members may have POTS. Many people develop POTS without any known cause.

What are the symptoms of dysautonomia and POTS?

Patients often have a variety of symptoms, which may include any of the following:

- Cardiovascular: dizziness or lightheadedness (particularly when standing up quickly, or standing for a long period of time), near-fainting, fainting, chest pain, palpitations (for example “skipped beats,” feelings of heart racing or pounding)
- Nervous System: headaches, mental fog, anxiety, depression, fatigue/tiredness, difficulty with sleep, tingling of extremities
- Gastrointestinal: stomach pain, diarrhea, constipation, nausea, vomiting
- Musculoskeletal: muscle or joint pain, very flexible joints, discoloration of hands and feet

How is dysautonomia or POTS diagnosed?

Patients who are suspected to have POTS or related symptoms are often referred to a cardiologist. Here’s what to expect at the initial cardiology appointment:

- Health history
- Electrocardiogram (EKG or ECG) to record the electrical activity of the heart
- Examination by the cardiologist

Dysautonomia and Postural Orthostatic Tachycardia Syndrome (POTS)

Depending on the symptoms and the outcome of the examination, the cardiologist may order additional tests such as:

- **Bedside standing test** (also called a “bedside tilt”): We will measure your resting blood pressure and heart rate while you lie flat on the exam table. You will then sit up and then stand up for 10 minutes. We will check your blood pressure and heart rate at regular intervals throughout and monitor your symptoms.
- **Echocardiogram**: We will use ultrasound to look at the structure and function of your heart.
- **Formal tilt table testing**: We may recommend a formal tilt table test for patients who have frequent fainting (syncope) episodes, or who have symptoms suspicious of POTS and a normal bedside standing test. The test will show how the heart rate and blood pressure respond to a change in position. The formal tilt table test will be scheduled for another day/time in the cardiology clinic at the main hospital. You will need to fast overnight (no eating or drinking after midnight) before coming to clinic the morning of the tilt test. You will change into a hospital gown and then lie on a special bed. We will place several monitors on you so that we can monitor blood pressure and heart rate while you are lying flat and then after the bed is placed in the upright position.

What is the treatment for POTS or dysautonomia?

There is not one specific treatment or one medication to treat all of the symptoms of POTS or dysautonomia. Depending on an individual’s symptoms, someone with POTS may need to see multiple specialists for treatment of symptoms (cardiology, neurology, gastroenterology, pain medicine, etc.). The goal of treatment is to decrease symptoms and the impact of the symptoms on day-to-day life. This may be managed by your primary care provider and/or specialists. After meeting with you, discussing your particular symptoms and performing some or all of the tests above, your cardiologist will likely recommend some or all of the following treatments:

- *Dietary adjustments*
 - **Increased oral fluid intake** of 2-3 liters (60-100 ounces) of water or non-caffeinated drinks per day. Increasing fluid intake helps to increase and stabilize blood pressure. It is important to plan and track your daily fluid intake. Drinking the amount of extra fluid you need daily is HARD and can only be accomplished by drinking throughout the day, not only at meals.
 - **Increased salt intake** either by adding table salt to your diet or, if your doctor feels it’s necessary, by taking over-the-counter salt supplements. Increasing salt helps the body “hold onto” the extra fluid you are drinking. Eating salty food, adding salt to foods or taking salt supplements may be a change in your normal eating pattern but may be an important part of the treatment in conjunction with drinking adequate water.
- *Lifestyle adjustments*
 - **Exercise**. Studies show that regular exercise is one of the **most important** treatments for POTS. Starting slowly and steadily increasing activity will gradually help you to feel better, even if you feel too tired or don’t want to exercise. You should exercise 5-7 days per week. If unable to exercise on your own, sometimes we recommend physical therapy or a personal trainer.

Dysautonomia and Postural Orthostatic Tachycardia Syndrome (POTS)

- **Adequate sleep.** Going to bed at the same time every night and unplugging from ALL electronics (games, TV, phones, tablets, computers) at least 1.5 hours before bedtime are very important. Please ask for more information on sleep hygiene if you need it.
- **Healthy eating, without skipping meals.** Skipping breakfast or other meals leads to increased mental fog, dizziness and nausea.
- **Maintaining a normal routine as much as possible.** Decreased activity can make symptoms worse. Try not to miss school or skip social activities, even on “bad” days.
- *Medications*
 - If the above measures do not work or don’t provide enough improvement in symptoms, there are several types of medicines that can be trialed to help reduce symptoms.
- *Cognitive Behavioral Therapy (CBT)*
 - This counseling is done with a psychologist trained in the technique. CBT helps patients work to adapt to the POTS symptoms and be more functional in their daily lives. Mindfulness can also be helpful.

What is the outlook for a person with POTS?

POTS can cause significant symptoms, but it is not a life-threatening condition. Symptoms may fluctuate. Medication alone cannot ease the symptoms of POTS. Recovery requires a combination of many different strategies. It takes work on your part to help with your recovery. Keeping up with the recommendations given to you by your healthcare team is key to success. Consistent exercise, increased fluid intake, increasing salt in your diet, taking medications (if necessary) as prescribed, good sleep patterns, and not skipping meals are **important and necessary** steps to improved functioning and recovery. As you and your doctor work to minimize symptoms, it is important to remain active and maintain your normal routine as much as possible, including going to school and socializing with your friends.

The vast majority of patients recover and symptoms improve or resolve. We can’t predict when that may happen. Recovery may be over the course of several months to several years. We will work with you to help you be as functional as possible as you travel along this journey.

For more information, visit a well-established resource such as Dysautonomia International (www.dysautonomiainternational.org) or Dysautonomia Youth Network of America, Inc. (www.dyninc.org). To speak to a member of the Children’s National Team, please call 202-476-2020.