What is PoTS?

PoTS is an abnormal response by the autonomic nervous system to becoming upright. There is an abnormally high increase in heart rate and altered blood supply to the brain on standing. It results in a large number of symptoms, the commonest of which are dizziness, fainting, tiredness and palpitations. There are many causes and, in some cases, no cause can be found. Although patients can be very disabled by their symptoms, many get better without treatment. However, some patients will have problems over many years. Fortunately changes in lifestyle and medicines will help and the majority of patients will respond to treatment.

Many doctors and nurses do not know that PoTS exists and so may not think of the diagnosis. If you think you may have this condition, it may help to take a printout of information from this website to your appointment.

The Autonomic Nervous System

The Postural Tachycardia Syndrome (PoTS) is an abnormality of the autonomic nervous system (sometimes called ‘dysautonomia’). The autonomic nervous system (ANS) is in charge of all bodily functions that we don’t have to think about, such as:

- Heart rate and blood pressure
- Digestion
- Bladder control
- Sweating
- Stress response

The sympathetic nervous system is part of the autonomic nervous system. It produces the ‘fight or flight’ or ‘stress’ response. When activated, a chemical called noradrenaline is released. Amongst other things, this causes sweating and increases heart rate and blood pressure.
What should happen to my blood when I stand up?

When lying down, 25% of our blood lies in our chest cavity. Under normal circumstances, when we stand, up to 800 millilitres of blood will be pulled down by gravity from our chest to the abdomen and legs. To maintain blood supply to our brain, the sympathetic nervous system will react by immediately narrowing blood vessels. Heart rate increases by 10-15 beats per minute and there is a very slight increase in blood pressure.

What happens when I have PoTS and stand up?

In some people, these mechanisms fail, altering the return of blood to the heart and brain. Extra noradrenaline can be produced to compensate and the heart races. Within 10 minutes of standing up, patients experience an increase in heart rate of 30 beats per minute or more (40 beats per minute in teenagers) and this is associated with symptoms of PoTS.

- **Postural** position of the body
- **Tachycardia** increased heart rate
- **Syndrome** a combination of symptoms
**Symptoms**

**Orthostatic intolerance** is a term that is sometimes used to describe symptoms of PoTS. It means symptoms that occur on standing and are relieved by lying down. In addition to heart rate and blood pressure, other bodily functions that are regulated by the autonomic nervous system can be affected. Symptoms can fluctuate from day to day and include:

- **Dizziness or light-headedness or presyncope** (almost fainting) - These symptoms usually occur when standing up, but can occur with prolonged sitting.
- **Syncope** (fainting or blackouts) - Approximately 30% of people with PoTS experience syncope.
- **Tiredness or weakness** - These are common symptoms of PoTS and can last for a considerable time after a bout of symptoms of PoTS.
- **Brain fog** - People with PoTS often complain of ‘brain fog’ which is difficulty in thinking or concentrating.
- **Palpitations** are a sensation of your heart pounding in your chest.
- **Shakiness**
- **Shortness of breath** - Patients can feel breathless when standing up or with slight exertion.
- **Chest pain**
- **Excessive or reduced sweating**
- **Gut problems** - Nausea is common. Other symptoms include diarrhoea, constipation, bloating, abdominal pain and vomiting. Many people with PoTS are told that they have irritable bowel syndrome.
- **Poor sleep**
- **Headaches** - These can be orthostatic headaches which means they occur as a result of being upright and may be caused by reduced blood supply to the brain. People with PoTS also commonly experience migraines and migraine-like headaches which are worse when the patient is upright.
- **Visual problems** - This can be described as excessive glare, blurred or tunnel vision.
- **Bladder problems**
**Symptom Triggers**

- Excess heat
- After eating – especially refined carbohydrate eg sugar, white flour
- Speed of positional change – don’t stand up quickly
- Dehydration
- Time of day (may be worse in the morning, especially rising after wakening)
- Menstrual period
- Deconditioning or prolonged bed rest
- Alcohol which dilates blood vessels
- Exercise

See “Living with PoTS” section of the website for further information.

PoTS symptoms can be:

- Mild
- Moderate
- Severe - with marked limitation of daily activities
How is PoTS Diagnosed?

Detailed questioning of the patient is the key to accurate diagnosis. Physical examination and appropriate investigations are also important. Potential underlying causes of PoTS needs to be considered and identified. Patients are usually diagnosed by a cardiologist, neurologist or medicine for the elderly consultant.

To be given a diagnosis of PoTS, a person needs to have:

- A sustained increase in heart rate of greater than 30 beats per minute within 10 minutes of standing
- The heart rate increase is often over 120 beats per minute when upright
- These criteria may not apply to those with a low heart rate when resting
- Those aged 12-19 years require an increase of at least 40 beats per minute
- There is usually no drop in blood pressure on standing

Investigations

It may be necessary for patients to have some or all of the tests below:

Electrocardiography (ECG)

An ECG is performed to rule out any heart problems that may cause symptoms similar to those found in PoTS.

The Active Stand Test

The active stand test can be used to diagnose PoTS. Under careful supervision, heart rate and blood pressure are measured after resting lying down, then immediately upon standing and after 2, 5 and 10 minutes. This test may bring on symptoms of PoTS and some people may faint.
Head-Up Tilt Table Test

This involves lying on a table that can be tilted to an angle of 60 to 70 degrees in a quiet, dimly lit, temperature controlled room. Blood pressure and heart rate are recorded in a continuous manner. After a period of 5 to 20 minutes of lying flat, the table is tilted. Although a diagnosis of PoTS should be made by an increase in heart rate of 30 bpm within the first 10 minutes, this upright position can last between 10 and 45 minutes. The patient will be asked how they are feeling during the test, so symptoms can be matched with heart rate and blood pressure. The test will end if your blood pressure becomes too low, satisfactory results have been obtained, or the maximum time has elapsed.

If facilities are available, some patients are tilted after a carbohydrate drink (liquid meal challenge), exercise or heat.

24 hour ambulatory blood pressure and heart rate monitor

Three stickers are applied to the chest and are linked to a little box which is attached by a belt your waist. It monitors heart rate over a 24 hour period. The patient is asked to go about their usual daily activities, trying to reproduce events that seem to cause the symptoms. A diary stating the time and activity performed at the moment when the symptoms started should be kept. Doctors look to see if there is a fast heart rate or drop in blood pressure at the time of symptoms.

Echocardiogram (Heart Ultrasound)

This test is to check if the heart’s structure is normal. In this test a technician will apply some jelly on the chest and will roll an ultrasound probe in many directions to create a 3 dimensional image of the heart. It is a painless and harmless test that similar to the ultrasound scan used routinely during pregnancy to look at the unborn baby.

24-hour Urine Collection

Individuals with PoTS often have low urinary sodium levels of less than 150 millimole per 24 hours which may indicate a low blood volume. Another 24 hour urine collection may be carried out to test for high levels of adrenaline to rule out pheochromocytoma (a growth on the adrenal gland) as a possible cause of symptoms.
Blood tests

Bloods are taken to rule out other conditions. These may include:

- Kidney function
- Blood count
- Liver tests
- Thyroid tests
- Calcium levels
- Glucose

Blood can also be taken for noradrenaline whilst lying and then standing (or when upright on the tilt table); if levels exceed 600 picogram/millilitre, it may suggest hyperadrenergic PoTS.

Other tests which may be carried out

- EEG
- Autonomic function screening tests
- Thermoregulatory sweat test
- MRI scan
Causes of PoTS

In primary PoTS there is no known cause. In secondary PoTS there is an underlying health condition. In some cases it is difficult to categorise the type of PoTS due to overlap between the subtypes.

**Primary PoTS**

**Partial Dysautonomia/Neuropathic**
This type often starts suddenly, and sufferers may have had a viral illness, pregnancy, immunisations, surgery or trauma prior to their symptoms starting. It is thought that this form of PoTS might be due to an abnormality of the body’s immune system.

**Developmental PoTS**
This type seems to afflict teenagers. It usually starts between the ages of 12-14 years old and may follow a very rapid growth spurt, with worsening symptoms until the age of 16. By the age of 19 to 24, about 80% of individuals will have no symptoms. The cause is unclear, but it seems to follow a rapid growth spurt.

**Deconditioning**
Deconditioning (being out of shape) is present in some individuals with PoTS. A traumatic event or illness can result in reduced activity or bedrest. Patients become unfit and the heart doesn’t pump as efficiently as before. This can trigger symptoms of orthostatic intolerance so patients avoid exercise which makes their symptoms worse, leading to more physical inactivity. There is a downward spiral of deconditioning which causes or worsens symptoms of PoTS.

**Hyperadrenergic PoTS**
Hyperadrenergic type PoTS appears to be less common. This can appear similar to pheochromocytoma (an adrenaline producing tumour) and tests may be needed to rule this out.
**Signs and Symptoms**
When upright, individuals experience severe anxiety, tremor, and cold sweaty hands and feet. Also, in a good portion of individuals, there is a significant urge to pass urine after even a short period of time upright. True migraine headaches are also common.

Blood noradrenaline levels are often high. High blood pressure may also occur on standing up.

**Causes**
This type of PoTS is suspected to have a strong genetic component as there is often a family history of this disorder. It is thought to be due to over activation of the sympathetic nervous system on standing. There may be an abnormality in a gene that leads to too much noradrenaline circulating in the body, hence the name ‘hyperadrenergic’.

**Mast Cell activation disorder**
Mast cell activation disorder needs to be considered in those who have episodes of flushing. Increased noradrenaline in the blood stimulates histamine release causing flushing.

**Overlapping Disorders**

**Inappropriate Sinus Tachycardia (IST)**
Inappropriate sinus tachycardia (IST) is another condition which has similar symptoms to hyperadrenergic PoTS. Patients with IST have a high heart rate when lying down (around 90-100bpm) which rapidly accelerates with slight exertion or emotional stress. The two conditions can overlap and may share the same underlying causes. Treatment options are similar and need to be tailored to the individual.

**Chronic Fatigue Syndrome (CFS)**
PoTS may be under diagnosed in those with chronic fatigue syndrome (CFS) and is estimated to affect around 25-50% of people with CFS. It has been suggested that these two conditions may be part of the same group of conditions with similar causes.
Reflex Syncope

30% of patients with PoTS will also have vasovagal syncope (also called the ‘simple faint’). On standing, after the early increase in heart rate, they have a drop in blood pressure. Sometimes their heart rate also drops for a short time and they lose consciousness. However, most patients with PoTS do not faint.

Secondary PoTS

PoTS may develop as a consequence of a number of medical conditions such as:

- Joint Hypermobility Syndrome (JHS)
- Diabetes
- Paraneoplastic syndrome
- Amyloidosis
- Sarcoidosis
- Systemic Lupus Erythematosus (SLE)
- Sjogren’s syndrome
- Multiple Sclerosis (MS)
- Lyme Disease

PoTS may be an early sign of other autonomic system abnormalities such as:

- Multiple System Atrophy (MSA)
- Primary Autonomic Failure (PAF)

Chemicals which may cause PoTS:

- Chemotherapy
- Alcohol
- Arsenic poisoning
Joint Hypermobility Syndrome/Ehlers-Danlos Syndrome (EDS) type III

JHS is an important cause of PoTS. It is an inherited disorder thought to be due to faulty proteins which are the main building block of the body, providing strength and support. As a result, joints can be hyper-mobile and more prone to injury and dislocation. Blood vessels may be more elastic causing pooling of blood in the lower body, tachycardia and symptoms of PoTS.

Patient with a combination of JHS and PoTS often display symptoms at an earlier age than patients with only PoTS and show an increased incidence of migraines and fainting.

Five questions can be used to check for hypermobile joints:

Answers to 2 or more questions suggest that your joints are hypermobile.

1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?
3. As a child did you amuse your friends by contorting your body into strange shapes OR could you do the splits?
4. As a child or teenager did your shoulder or kneecap dislocate on more than one occasion?
5. Do you consider yourself double-jointed?

Hakim & Grahame (2003)

Even if you have hypermobile joints, people with Joint Hypermobility Syndrome have additional problems and a diagnosis of JHS cannot be made unless a more detailed examination is carried by a specialist.