

POTS UK
POSTURAL TACHYCARDIA SYNDROME

POSTURAL TACHYCARDIA SYNDROME (PoTS)

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GLOSSARY

Antibody is a protein produced to counteract foreign substances in the body.

Antiphospholipid syndrome (previously called Hughes Syndrome) is an autoimmune condition that can cause sticky blood resulting in stroke, vein thrombosis (blood clot) or miscarriage.

Autoimmune disorders occur when our immune system produces autoantibodies – antibodies against our body's own tissues.

Autonomic nervous system is the involuntary nervous system that controls bodily functions that we do not have to think about. It is important in 'fight and flight' and 'rest and digest' responses.

EEG is an electroencephalogram, a test that records electrical activity of the brain.

Electrophysiologist is a cardiology doctor with special expertise in managing heart rhythm problems.

Epinephrine and norepinephrine are also called adrenaline and noradrenaline and are **catecholamines**, chemicals that are released into the bloodstream in stressful, 'flight or fight' situations.

MRI scan is magnetic resonance imaging.

MSA is multisystem atrophy, a neurological disorder that severely impairs the body's involuntary (autonomic) functions and other parts of the brain.

Orthostatic intolerance means symptoms that develop in the upright position that are relieved by lying down.

Phaeochromocytoma is a catecholamine-producing tumour that is usually not cancerous.

Pooling is dropping of blood with gravity into abdominal cavity and limbs.

Pure autonomic failure is a neurological disorder that causes impairment of the involuntary nervous system.

Sjögren's syndrome is an autoimmune condition that causes a number of symptoms including dry mouth and eyes, joint pains and fatigue.

Syncope is brief loss of consciousness due to altered blood supply to the brain.

Tilt table test is an autonomic test used to induce an attack or episode whilst connected to heart and blood pressure monitors.

* The star symbol indicates that there is no evidence to support a statement in this booklet. The statement is based on patient experience or agreed by experts.

WHAT IS POSTURAL TACHYCARDIA SYNDROME (PoTS)?

Postural tachycardia syndrome is an abnormal response by the autonomic (involuntary) nervous system to standing up. To be diagnosed with PoTS, an individual must experience the following:

1. A group of symptoms that occur frequently in an upright position (usually standing) and are relieved by lying down.
2. These symptoms should be associated with an abnormally high and persistent increase in heart rate of 30 beats per minute (40 beats per minute if under 19 years of age).

Some symptoms of PoTS are similar to those experienced by people with abnormally low blood pressure (BP), but blood pressure does not necessarily drop in PoTS.

PoTS can develop in association with a number of different diseases or situations. The reason for this is not properly understood but it is possible that there is a malfunction of the nervous system that controls autonomic functions in the body.

When a healthy person stands up, to avoid blood dropping down into the limbs and abdominal cavity, blood vessels contract immediately and heart rate increases slightly to maintain blood supply to the heart and brain. In PoTS, this automatic adjustment to standing does not work correctly, resulting in an excessive rise in heart rate, increased epinephrine in the blood and altered blood flow to the brain. PoTS is more common in women, and between the ages of 13 and 50 years.

WHAT ARE THE SYMPTOMS OF PoTS?

Individuals with PoTS experience 'orthostatic intolerance' which means that they experience symptoms on moving to an upright position. Symptoms can be debilitating, ranging from mild to severe, and varying from day to day.

PoTS symptoms

- Dizziness or pre-syncope (almost fainting)
- Palpitation (awareness of heart beat)
- Fatigue
- Headaches - orthostatic headaches (due to upright posture)/migraine
- Brain fog (difficulty in thinking)
- Sense of anxiety
- Syncope (fainting)
- Shakiness
- Visual problems (greying, tunnel or glare)
- Gut problems (including nausea, diarrhoea, pain)
- Sweating
- Chest pain
- Poor sleep
- Purplish discolouration of skin due to blood pooling in hands and feet
- Bladder problems

Triggers that can worsen PoTS

- Excess heat
- Eating - especially refined carbohydrate (e.g. sugar, white flour)
- Standing up quickly
- Dehydration
- Time of day (especially rising after wakening)
- Menstrual period
- Deconditioning (lack of fitness) or prolonged bed rest
- Alcohol (as it dilates blood vessels)
- Inappropriately excessive exercise
- Temporarily during illness such as viral infections or after operations*

HOW IS PoTS DIAGNOSED?

GPs may recognise the condition, but a clear diagnosis would normally be made by an electrophysiologist (a heart rhythm expert), neurologist or other hospital physician. These specialists are often found in blackout or syncope clinics, although only a minority of people with PoTS experience blackouts.

The following tests may be used to confirm a diagnosis or exclude other conditions with similar symptoms:

- **Active stand test** – The patient rests flat for a few minutes and heart rate and BP are recorded whilst lying. After standing up, further recordings are taken over 10 minutes.
- **Tilt table test** – The patient rests flat on a special bed with a footplate whilst BP and heart rate recordings are made. The bed is then tilted (head end up) for up to 45 minutes while further recordings are taken.

(Stand and tilt tests are stopped earlier if the patient faints or if satisfactory recordings have been made).

- **12-lead electrocardiogram (ECG)** – Electrical activity of the heart is recorded through little stickers on the chest wall
- **24 hour ECG** – Similar to above, but the stickers are left on for 24 hours and results are recorded on a small portable machine as the patient undertakes normal daily activities.
- **24 hour blood pressure monitoring** – a small machine is worn continuously and takes intermittent recordings as the patient undertakes normal daily activities.
- **Blood tests** – these may include full blood count, kidney and liver function, thyroid function, calcium, diabetes tests, lying and standing norepinephrine levels.
- **Echocardiogram (cardiac echo)** – A simple test where the structure of the heart is visualised through jelly on the chest wall. It is quick and painless and rules out structural heart abnormalities.
- **Exercise Test** – The patient exercises in a laboratory and heart rate, blood pressure and ECG recordings may be made during and after the test.

The following tests are more likely to be used in autonomic or PoTS clinics:

- **Autonomic screening tests**
- **Thermoregulatory sweat test**
- **EEG**
- **MRI scan of brain**

Misdiagnosis

PoTS was first recognised by medical journals in 1993, but many medical professionals are still unaware of the condition today. Obtaining the right diagnosis can therefore be a lengthy and frustrating process. Meanwhile, patients are commonly misdiagnosed with other conditions such as anxiety and depression, reflex syncope (vasovagal syncope) and chronic fatigue syndrome.

If you think you could have PoTS, it may help to discuss this booklet with your doctor and ask for a referral to a specialist with an interest in PoTS. There is a list of such doctors on the PoTS UK website.

WHAT CAUSES PoTS?

There are a number of factors and disorders causing or associated with the condition but in some cases a cause is never identified. PoTS can follow a viral illness such as glandular fever, or be linked to pregnancy or a traumatic event. Sometimes teenagers are affected after a rapid growth spurt and most will improve within a few years. Some patients develop PoTS-like symptoms due to lack of fitness and the heart pumping inefficiently after being confined to bed for some time.

Hyperadrenergic PoTS is a type of PoTS that may in some cases have a genetic cause and therefore run in families. When upright, this group of patients may have an urge to pass urine frequently. They tend to complain of a severe sensation of anxiety and tremor, migraine headaches and clammy hands and feet. They may also present with high blood pressure and high blood norepinephrine levels. Norepinephrine levels are also elevated in pheochromocytoma, and it may be necessary to have tests to rule out this condition.

Conditions associated with PoTS

Hypermobile Spectrum Disorder and Hypermobile Ehlers-Danlos Syndrome

Hypermobile (excessively flexible) joints can be a normal finding in some people and even an advantage in sport and dance. However, in some people, they are part of a spectrum of inherited conditions that is associated with other problems including painful joints, dislocating joints, abnormal skin (stretchy soft skin, stretch marks and wide scars), allergies, gut problems, bladder problems, and abnormal functioning of the autonomic nervous system.

PoTS was found to be relatively common in conditions previously called Ehlers-Danlos Syndrome (EDS) - hypermobility type, EDS type III, joint hypermobility syndrome and benign joint hypermobility syndrome. Since 2017, these conditions are now called hypermobility spectrum disorder and hypermobile EDS.

There is more information about hypermobility and EDS on the websites below:

www.ehlers-danlos.org
www.hypermobility.org

Low blood pressure

Low blood pressure can be healthy, but if it drops very low on standing it is called **orthostatic hypotension** and can cause symptoms similar to PoTS. Reflex syncope occurs when a drop in blood pressure results in fainting.

PoTS, low blood pressure and reflex syncope can exist together in the same patient and this combination can be seen in hypermobility spectrum disorder/hypermobile EDS and chronic fatigue syndrome. When blood pressure drops there is sometimes an increase in heart rate. This is called a **reactive tachycardia** and can look like PoTS. Treatments for PoTS and low blood pressure are similar.

Chronic Fatigue Syndrome (CFS)

PoTS may be underdiagnosed in those with chronic fatigue syndrome and is estimated to affect around one third of people with CFS.

Inappropriate Sinus Tachycardia (IST)

IST has similar symptoms to hyperadrenergic PoTS, but the high heart rate may be higher than in PoTS when lying down (around 90–100bpm) and rapidly accelerates with exertion or stress.

Mast Cell Activation Disorder

This needs to be considered if flushing or allergies are prominent.

Autoimmune conditions

Recent research has shown that autoantibodies affecting the autonomic nervous system are more common in people with PoTS. Autoimmune conditions that have been associated with PoTS include Sjögren's syndrome and antiphospholipid syndrome. Treatment of underlying autoimmune conditions can improve symptoms of PoTS.

Abnormally functioning genes

There is new evidence emerging of an abnormality in the gene that regulates epinephrine in the body.

Other associated conditions

There are limited reports of PoTS being associated with immunisations, multiple sclerosis, diabetes, alcoholism, pure autonomic failure, MSA, Lyme disease and cancer.

WHAT CAN I DO TO IMPROVE MY CONDITION?

For some, lifestyle changes may be all that is needed to control symptoms.

Fluids

Patients with PoTS often have a low blood volume and this can be increased with extra fluids. At least 2–3 litres per day is usually advised in adults. As symptoms can be worse in the morning, it may help to boost fluids before getting out of bed. In an emergency, drinking 2 glasses of water quickly can rapidly elevate blood pressure and lower heart rate.

Intravenous fluids (fluids passed directly from an external bag into a vein) have been used, but can cause serious complications with regular use.

Alcohol dilates blood vessels and may make symptoms worse. Coffee and other caffeinated drinks may also worsen symptoms, although some find them helpful.

Food and salt

A high salt diet of up to 10g per day may be recommended. Extra salt can be dangerous in some conditions such as high blood pressure, kidney and heart disease, and therefore should only be taken if recommended by your doctor. Salt tablets can also be prescribed.

Slow Sodium tablets are available in the UK on prescription and are coated to reduce nausea. (6g salt = one teaspoon = 10 Slow Sodium tablets).

Eating small amounts and often can be helpful. Symptoms can worsen after a large meal as blood is diverted to the digestive tract and away from other areas. Some people may find refined carbohydrates aggravate symptoms. Avoid rich sugary foods and those containing white flour. Eat lots of unprocessed foods such as vegetables, fruit, beans and foods that contain whole grain.

Posture

To avoid fainting or near fainting:

- Take notice of early warning signs such as light-headedness, dizziness, and nausea
- Lie down immediately and, if possible, elevate your legs

If your circumstances make this difficult:

- Cross your legs while standing or rock up and down on your toes
- Clench your buttocks and abdominal muscles
- Clench your fists

The risk of fainting can also be minimised by:

- Standing up slowly if you have been sitting for a while
- Avoiding prolonged standing. Fidgeting can be helpful*
- Avoiding having your hands over your head for any length of time
- Avoiding prolonged sitting, which can cause symptoms in some people. Elevating legs when seated can be helpful.*

Compression tights

Compression tights should be waist high and give at least 30mmHg of pressure at the ankle (Grade II compression) in order to help reduce the amount of blood pooling in the legs. Sports compression clothing may also help.*

Temperature regulation

Heat worsens symptoms and so many people find their symptoms are worse during the summer.

Dress in layers of clothes, so layers can be removed to prevent overheating.

Salt and fluid intake should be increased if you feel hot or sweat more.

A spray bottle of water for your face and neck will cool you down as the water evaporates from your skin.

A fan or air conditioning can be helpful.

Some people use cooling vests.

Fitness

Light to moderate exercise can help or even cure PoTS in some cases.

Increasing leg strength and core (central) muscles helps to pump blood back to the heart.

Physical exertion can initially worsen PoTS so factor in recovery time afterwards. Begin with recumbent (sitting or lying down) exercise if you are starting for the first time. Gradually increase duration and intensity. Upright exercise can be added after 2–3 months of recumbent exercise. Aim for twenty to thirty minutes of aerobic exercise, which should be undertaken at least three times a week in combination with resistance training.

Suitable exercise includes:

- Swimming
- Rowing
- Recumbent biking, progressing on to upright biking
- Pilates – mostly horizontal exercise focusing on core strength*
- Lower limb resistance training
- Walking
- Jogging

Sleep

PoTS patients often have poor quality sleep. Potential causes of sleep disturbance need to be identified such as underlying anxiety and depression. There may also be other physical causes such as low iron levels, which can cause restless legs.

Elevating the head of the bed has been recommended to boost blood volume and improve symptoms.

Pacing yourself

Symptoms tend to be worse in the morning so it may be best to plan activities for later in the day. Set yourself achievable goals and avoid rushing. Factor in time for rest.

Live within your body's limits – it will tell you if you have done too much.

Hygiene

Drink a glass of fluid before and after showering. Avoid taking a hot or long shower or bath as this dilates your blood vessels and can make symptoms worse. Finishing your shower with cool water may help. Sit on a stool by the basin and in the shower. Dry shampoo spray and wet wipes may be an option on a bad day!

Psychological support

Having PoTS can mean that a previously active person will need to make significant lifestyle changes. Psychological support can help you to find optimal ways to look after your mental and physical needs, getting you in the best shape to manage your condition most effectively. PoTS symptoms (e.g. racing heart, chest pain and dizziness) are very real and can be frightening; it can be helpful to learn to prevent any anxious responses that may cause these symptoms to worsen. Cognitive behavioural therapy (CBT) is a structured talking therapy that can help you work towards accepting your limitations, adjusting to the unpredictability of chronic illness, and helping you pace yourself to achieve the challenges of daily-living whilst still maintaining activities which most help you feel positive and fulfilled.

Travel

Driving – By law you must tell the DVLA of any health condition that may affect your ability to drive. Failure to do so could lead to prosecution and invalidate your insurance. Problems that you should consider reporting include tachycardia, syncope and dizziness. You should ask your GP or specialist if you need to inform the DVLA. It does not always mean that you will be stopped from driving. There is further information about medical fitness to drive and the Blue Badge Scheme on this website: <https://www.gov.uk/health-conditions-and-driving>

Flying – Drink more fluid than normal. You can take an empty drinks bottle through security and ask the flight attendants to fill it on the plane. Avoid alcohol and wear compression tights/socks. Elevate your legs when possible and keep mobile to prevent pooling. Remember to take medication in your hand luggage in case there are problems with lost luggage. Advise your travel insurer about your condition to ensure you are fully covered.

EDUCATION AND WORK

Some people with PoTS require additional support. In school, the Special Educational Needs Co-ordinator (SENCO) can help. Colleges and universities have a disability adviser whose team can provide extra support for students. If you are struggling to meet deadlines or are unfit to sit examinations, inform your tutor or head of department immediately. A medical report may be required, so see your doctor as soon as possible as they cannot write a report unless they have seen you at the time.

Larger employers have access to an occupational health department that will provide advice to your employer regarding adjustments that may be necessary to allow you to work to your full potential. Smaller employers will not usually have this facility. Provide as much relevant information to your managers as is necessary so they can understand any difficulties you may be facing. Further information is available on www.dwp.gov.uk

TOP TIPS FOR STUDYING OR WORKING*

Eat and drink regularly. Working or studying with your feet elevated may help avoid brain fog. Don't sit too long in one position. Move around or fidget. Keep cool – physically and mentally! Keep as fit as you can – consider swimming or pilates. If you cannot meet deadlines, tell your teachers or employer as soon as possible. Keep everyone informed – providing a copy of this booklet may be helpful.

DO MEDICINES HELP?

When these lifestyle changes are insufficient to control symptoms, medication may be recommended. All medicines used in PoTS are unlicensed, which means that they have not been officially approved for this use. For this reason, GPs usually prefer that they are recommended or prescribed by a specialist. For the more expensive drugs, special funding approval may be required.

Medicines more commonly used in PoTS

Midodrine
Fludrocortisone
Beta-blockers
Ivabradine
Desmopressin
SSRI antidepressants for their effect on the autonomic nervous system

SHOULD WE HAVE A BABY?

Pregnancy can be a rewarding experience, even for those severely affected by PoTS. Some women experience an improvement in symptoms during this time, but for others there can be a deterioration in early pregnancy, especially if nausea or vomiting cause dehydration.

To ensure the best possible outcome for mum and baby, speak to your doctor BEFORE becoming pregnant. The effects on the unborn baby of taking most medicines are unknown and some women choose to stop medication before conceiving – ask your doctor first.

The benefits of staying on medication versus the consequences of stopping need to be discussed with a specialist who is very knowledgeable about PoTS and its treatment. The majority of women have a normal delivery. Caesarean section and epidurals are also safe in women with PoTS. It is important to maintain fluid intake during labour and an intravenous infusion may be helpful. Medication containing epinephrine should be avoided.

It is helpful to ensure that your obstetrician and anaesthetist have some knowledge of PoTS and this booklet may be useful to them. If you feel too unwell or are taking large amounts of medicines, it may be sensible to delay becoming pregnant. Ask your GP or practice nurse about contraception. There is a more detailed leaflet and a list of medical journal papers on the PoTS UK website that may help your anaesthetist or obstetrician to obtain further information.

WHAT WILL THE FUTURE LOOK LIKE FOR ME?

For many, PoTS will improve with time and in some individuals it will disappear altogether. There are some forms of PoTS that are unlikely to go away but can improve through lifestyle changes and, if necessary, medication. The vast majority of patients learn to manage the condition and return to something close to their previous level of functioning. It is important to remember that while PoTS can be disabling, unlike some chronic illnesses, it will NOT shorten your life. Whilst many of the symptoms like a racing heart, dizziness or chest pain can be frightening before you understand their origin, they do NOT cause physical harm when caused by PoTS. Support from a good and well-informed healthcare professional and from patient support groups can help enormously.

It can be very hard work making all the necessary life-changes to live with PoTS. It can help to know that after making these changes and adjusting to what they cannot do, whilst focusing on actively engaging in what they can, many patients with PoTS report enjoying happy and fulfilling lives.

As with many long-term health conditions, although we would never wish them upon ourselves or others, a positive consequence can be a new appreciation for the things that really do matter to us, and an opportunity to re-establish priorities, living life thereafter to our full potential.

Medical Disclaimer

Please remember that this publication provides general information only, and should not be considered a substitute for medical advice from a healthcare professional. Individuals should always discuss their medical condition with their healthcare professional.

Sources of Evidence

Details regarding the sources of evidence used in the production of this booklet are available on the PoTS UK website.

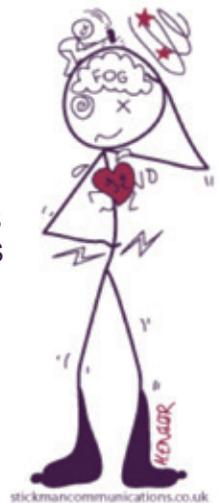
See www.potsuk.org/sources_of_evidence



PoTS UK is a charity that supports people with PoTS and their family and friends. It is run by volunteers from a medical background who are affected by PoTS.

Our aims are to:

- raise awareness about PoTS in the UK and beyond
- assist patients in achieving their correct diagnosis
- provide up to date evidence based information about PoTS
- raise awareness amongst health professionals about PoTS
- improve health services for patients with PoTS
- promote and participate in research
- raise funds to support these activities



PoTS UK aims to offer hope that life can be fulfilling whilst living with this health condition.

PoTS UK is a certified member of NHS England's certification scheme for social and healthcare information - the Information Standard.

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