



MANAGEMENT FILE

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POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME (PoTS)

INTRODUCTION

A small but significant minority of people with ME/CFS also have a condition called postural orthostatic tachycardia syndrome (PoTS). But it often remains undiagnosed because of lack of knowledge amongst health professionals.

This Management File is based on a Workshop on PoTS that formed part of the 2016 CFS/ME Research Collaborative conference in Newcastle where up-to-date guidance on the diagnosis and management of PoTS was presented by Professor Julia Newton and representatives from the charity PoTS UK.

WHAT IS PoTS?

PoTS, or postural (= position of the body) orthostatic (= symptoms caused by being upright) tachycardia (= increase in heart rate) syndrome (= a collection of symptoms), is a condition caused by a failure of the autonomic nervous system (ANS) – which helps to control pulse rate and blood flow to vital organs – to respond appropriately when we stand up, or try to remain standing.

In terms of simple human plumbing, when we stand up around 500cc of blood rapidly descends from the chest into the abdomen and lower limbs.

If the ANS is functioning normally, messages are then sent from the brain to produce a rise in heart rate of about

10 to 20 beats per minute, along with a constriction of the blood vessels where this blood has accumulated. As a result, blood flow to the brain and heart is maintained and life goes on as normal.

In PoTS, there is a failure of this ANS response mechanism with inadequate constriction of the blood vessels and a larger rise in pulse rate.

Blood flow to the brain then falls, which is called cerebral hypoperfusion. This leads to the characteristic symptoms of PoTS: dizziness, feeling lightheaded or faint when moving from lying to standing, and sometimes even fainting.

Blood pressure usually remains fairly normal and does not fall on standing in PoTS. However, there is a sub-group of people with PoTS who have hypertension/raised blood pressure, which is associated with a hyperadrenergic (ie increased noradrenaline) state. They have profuse sweating, tremulousness, and a tachycardia (high pulse rate). And because there is an overlap with what is called neurally-mediated hypotension, there is another sub-group with PoTS who experience a drop in blood pressure on standing up.

PoTS tends to affect people between the ages of 15 to 50 and is four times more common in females. Teenagers are sometimes affected following the rapid adolescent growth spurt.

A common misconception is that PoTS

is caused by deconditioning. However, a survey by PoTS UK found that 66% of people were taking regular aerobic exercise before the onset of their PoTS.

WHY DOES PoTS SOMETIMES AFFECT PEOPLE WITH ME/CFS?

PoTS can affect people with ME/CFS, especially those in younger age groups and at the more severe end of the spectrum. This is because the ANS is commonly involved in ME/CFS where the malfunction can cause what is called orthostatic intolerance (difficulty in remaining upright – as in PoTS) and cold hands and feet.

The ANS also affects bowel and bladder control and may therefore cause irritable bowel type symptoms and 'irritable bladder' symptoms.

PRIMARY PoTS AND OVERLAP WITH OTHER CONDITIONS

Primary PoTS often has an abrupt onset and may follow acute infections, immunisations, surgery, pregnancy, or trauma – all of which can trigger ME/CFS. As with ME/CFS, there is evidence of an autoimmune component and there is a genetic component in some cases.

PoTS is also more common in people who have hypermobile joints (which can also overlap with ME/CFS), people

who are deconditioned after prolonged bed rest, diabetes, sarcoidosis, Sjögren's syndrome, SLE/lupus, antiphospholipid (Hughes) syndrome, and mast cell activation disorder – which should be checked for if flushing and allergies are present.

WHAT ARE THE DIAGNOSTIC SYMPTOMS AND SIGNS?

Characteristic symptoms, which occur on standing and are normally relieved by lying down, include:

- fatigue or weakness (91%)
- palpitations or awareness of the heart beating (86%)
- dizziness (90%)
- feeling light-headed, pre-syncope/feeling faint (90%)
- sometimes actually fainting/syncope (58%), or having a transient loss of consciousness

Percentages are taken from a PoTS UK patient survey.

Other common symptoms, which occur in up to 40% of cases, include:

- nausea and other irritable bowel-type symptoms
- headaches – which may be caused by lack of blood flow to the brain
- sleep disturbance
- shortness of breath and chest pain
- visual disturbances – including blurring
- 'brain fog'/cognitive dysfunction – as occurs in ME/CFS
- 'coat hanger pain' - which is thought to be due to poor blood supply to muscles around the neck and shoulder

Acrocyanosis – a puffy bluish discoloration of the hands and feet that is caused by blood pooling in the veins occurs in around 50% of cases.

Symptoms are often described as being more prominent on rising after waking up. They are frequently exacerbated by standing up too quickly, heat, some types of food or large meals,

alcohol and stress. They are often exacerbated during infections and may be worse during menstruation.

For some people, the disability caused by PoTS is considerable, especially when this is added to the health problems caused by ME/CFS. Patient evidence collected by PoTS UK indicates that 23% are wheelchair-users and 37% are unable to work. Although severely incapacitated, PoTS patients often look well.

HOW IS PoTS DIAGNOSED?

A diagnosis of PoTS is based on its characteristic symptoms and what is called a 10-minute active stand test and/or a tilt-table test.

The Active Stand Test: Under careful supervision, heart rate and blood pressure are measured after resting/lying down for ten minutes, then immediately upon standing still (unsupported with hands by sides) and after 2, 5 and 10 minutes. This test may bring on symptoms of PoTS and some people may faint. If the result is uncertain, the test can be repeated on another occasion.

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 five 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 beats per minute 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



Acrocyanosis in the toes can be a symptom of PoTS

(liquid meal challenge), exercise or heat.

Serum catecholamines (chemicals that transmit messages around the brain) should be checked in supine and upright positions, especially if a hyperadrenergic form of PoTS is suspected.

A heart rhythm test/electrocardiogram test should also be arranged to exclude heart problems that can cause a tachycardia as well (eg inappropriate sinus tachycardia) and thyroid function tests. Other cardiac/heart tests – eg 24-hour heart rate and blood pressure monitoring, echocardiogram, autonomic function screening tests - may be required in more complicated cases.

Unfortunately, many doctors still know little or almost nothing about how to diagnose PoTS and how to manage it. As a result, many people with PoTS are left undiagnosed, may even be misdiagnosed as having anxiety, panic attacks or hypochondria, or be labeled as 'heart-sink' patients.

HOW IS PoTS TREATED?

Treatment involves a combination of self-help measures and in more severe cases the use of drug treatments.

Activity management: Carefully planned and monitored activity management involving the lower limbs can be helpful. However, this obviously creates difficulties when PoTS is combined with ME/CFS. Doing so in a recumbent position is one option here.

Avoidance of exacerbating factors: Alcohol (which dilates blood vessels), dehydration, heat and hot baths, and large meals should be avoided.

Compression tights and support stockings may be helpful. These should be waist high and provide at least 30mmHg of pressure at the ankles (ie Grade 2 compression) to be of §1 maximum benefit. This helps to prevent the pooling of blood in the legs.

Support stockings and tights can be purchased from department stores and pharmacies. Where appropriate, doctors can also prescribe compression tights on the NHS. Detailed guidance can be found on the PoTS UK website.

Diet: Small frequent meals that are low in refined carbohydrate are recommended – as is eating plenty of unprocessed foods such as vegetables, fruits, beans and foods that contain whole grains. Avoiding rich sugary foods and those containing white flour can be helpful.

Symptoms will be worse after large meals because this diverts blood to the stomach.

Driving and flying: The law states 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.

Drink more fluid than normal if you are flying and wear compression tights or socks. Elevate legs where possible and carry any important medication in your hand luggage. Make sure you are covered for PoTS with travel insurance.

Fluids intake: This should be increased to at least two to three litres per day to boost blood volume. If symptoms are worst in the morning, it may help to boost fluids before getting out of bed.

In an emergency, two glasses of water can help to rapidly elevate blood pressure and lower heart rate. Intravenous fluids have been used but can cause serious complications if misused.

Caffeine and other caffeinated drinks (which can raise the pulse rate) often worsen symptoms but some people find them helpful.

Salt intake: This may need to be increased but should only be done under medical supervision because excessive intake of salt can be dangerous. Slow sodium tablets, which are coated to reduce nausea, are available on prescription from the doctor.

Posture and movement: Try to avoid sitting in one position for long periods. Move around as much as possible within your limitations.

To avoid fainting take notice of early warning signs – dizziness, feeling lightheaded. Lie down immediately and elevate the legs.

Working or studying with feet up may help to reduce brain fog. Try not to sit in one position for too long.

Temperature extremes: As excessive heat worsens symptoms, the condition is often worse in summer. During hot weather, dress in layers which can be easily removed to prevent overheating. Extra salt and fluids will be required if you sweat a lot.

There is some excellent and more detailed advice on all aspects of general management on the PoTS UK website: www.potsuk.org/general_advice

DRUGS

Existing drug treatments should be reviewed because some drugs can exacerbate PoTS. Examples include tricyclic antidepressants, diuretics (water-losing tablets) and drugs that lower blood pressure. Check with your pharmacist before using over-the-counter medications.

Drug treatment should be considered in more severe cases. As none of these drugs are licensed to treat PoTS, they should initially be prescribed and carefully monitored by a doctor with experience in treating this condition. Examples of drugs that are sometimes used are listed in the next column:

- To reduce tachycardia/palpitations: beta blockers
- To increase the blood volume: fludrocortisone – causing salt and water retention
- To constrict blood vessels: midodrine
- Improving central/brain blood flow: selective serotonin reuptake inhibitors stimulate the standing up vasoconstrictor reflex

FURTHER INFORMATION

PoTS UK is an excellent charity that provides information and support. They have a list of doctors that patients can be referred to for both diagnosis and management.

Website: www.potsuk.org

Information: info@potsuk.org

Tilt table test (short link to the Mayo Clinic website):

<https://tinyurl.com/y2d7farf>

Medical references:

Kavi L et al. (2012) Postural tachycardia syndrome – multiple symptoms but easily missed. *British Journal of General Practice*, 62, 286 – 287.

Kavi L et al. (2016) A profile of patients with postural orthostatic tachycardia syndrome and their experience of healthcare in the UK. *The British Journal of Cardiology*, 23, 33.

Medical information contained in this leaflet is not intended to be a substitute for medical advice or treatment from your doctor.

We say that you always consult your doctor or healthcare professional about a specific problem.

We also recommend that any medical information provided by The MEA is, where appropriate, shown to and discussed with your doctor.
