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POTS Syndrome

This handout is written by Dr. Marc Le Gras. The intent of this handout is to enhance the information provided to you at the consultation and on follow-up visits. It is provided for education and help with your condition. The more you know about POTS, the better you will do.

What is POTS?

POTS is an abbreviation for Postural Orthostatic Tachycardia Syndrome. In plain English, this means an abnormal rise in heart rate along with symptoms mainly consisting of dizziness or feeling weak and lightheaded when going from lying down or sitting down to standing up or walking. People with this chronic illness may be unable to stand or walk for long periods of time, or even briefly. There can be a drop in blood pressure with these symptoms, called hypotension. At times, people can lose consciousness or faint – this is called "**syncope**." The medical definition of POTS in pediatrics is a persistent rise in heart rate of more than 30-40 beats per minute when standing, or a standing heart rate of greater than 120 beats per minute. There may be a drop in blood pressure as well. The medical definition sometimes includes a duration of at least six months of symptoms, but we don't need to wait that long to start making you feel better. POTS is not a deadly or lethal condition.

But doesn't everyone get dizzy at times when they stand up?

Yes, normal healthy people can get brief symptoms occasionally when they stand up. When we stand, there is a normal shift in blood distribution from our chest to our abdomen and legs of up to 1 liter and this can cause brief symptoms of lightheadedness and an increase in heart rate. Usually the cardiovascular system adapts to this quickly and symptoms of an increased heart rate will resolve within 30 seconds. These brief episodes are called **orthostatic hypotension**.

So why do I faint?

There are times where the blood pressure drops so much that there isn't enough blood going to the brain. The blood pressure drop alone can be enough to cause fainting, but it also triggers an increased heart rate – there are areas in the brain that receive the signal of a fast heart rate and cause an abnormal response in which a brain nerve (called the vagal nerve) slows or stops the heart. This type of fainting is called vasovagal or neurocardiogenic syncope.

Why do I have POTS?

The exact scientific mechanism and cause of POTS is not fully understood and there are different types of POTS. In pediatrics, most POTS cases are the neurogenic type where abnormal pooling of blood occurs, which causes an abnormal rise in heart rate. The ability of the blood vessels to tighten or vasoconstrict and the brain's blood pressure and heart rate regulation may also be abnormal. Usually in pediatric patients, the heart is normal. Some pediatric patients may have a condition called "joint hypermobility syndrome," also known as "Ehlers-Danlos syndrome, Type III" – these people have very distensible or stretchy veins which cause abnormal blood pooling. Other situations which can cause POTS include certain medications, hyperthyroidism, neuropathies, low adrenal function, and eating disorders. In adults, POTS can be seen with certain cancers, diabetes, multiple sclerosis, amyloidosis, and there is a rare "hyperadrenergic" type with abnormal elevation of adrenaline with standing.

We see POTS mainly in females (four times more commonly than in males). In pediatrics, it can start with a rapid growth spurt, a severe illness, surgery or an injury.

POTS is a chronic illness. In some patients there may be a cure and symptoms go away completely, but in many it is very difficult to treat this condition and it may go on well into the adult years. Having a chronic illness like POTS can be devastating. People may feel isolated and in many cases they may be labeled as having a purely psychiatric or psychological problem. Many patients are unable to do sports, exercise or even attend school. Many people end up feeling helpless, sad or depressed, angry, and misunderstood. You need to remember that this condition is real; you have an illness and it is not your fault or anyone's fault. Remember that you can and will get better but you may have good days and bad days. Symptoms such as fatigue, poor sleep, headaches, abdominal pain, nausea and diarrhea, along with chest pain, trouble breathing, palpitations (fast or irregular heartbeats), and inability to tolerate heat or standing are part of this chronic illness.

How will I get better?

Dr. Le Gras will check the boxes on the therapies that are listed in this handout that will be helpful to you. Over time, the therapies may change as you feel better and we have a better understanding of your condition. This is a team effort – you, your family, and Dr. Le Gras.

THERAPIES

1. **We need to increase your circulating blood volume.** No, this is not a blood transfusion. We need to have more fluid in your body – that way, the blood pressure will be more stable and the fluid shifts that occur with standing will not bother you as much.

   ☐ **A dramatic increase in salt.** Yes, salt is not considered healthy for most people because it can cause high blood pressure. For now, in your case, we need a more stable blood pressure. The salt will help you retain fluids and prevent your blood pressure from dropping. You must aim for 8-10 grams of salt per day – that is a lot of salt. The best way is to eat salty foods such as pretzels, saltine crackers, and soups, and to add extra salt to your food. Some people prefer to take salt pills (sodium chloride, 1 to 2 pills three times a day), but salt pills can cause stomach upset, so I recommend the dietary approach first.

   ☐ **You need to dramatically increase your fluid intake**. I recommend that you drink 64 to 80 ounces of fluids per day. Start by drinking at least 20 ounces of those fluids within one hour of waking up. Very often it is hard to remember to drink that much fluid and the best way is to keep a record how much you are drinking as the day goes on, or drink from a container that you bring with you everywhere you go, which will permit you to total up your fluid intake at the end of the day. Find fluids that you like, such as Crystal Light, Gatorade or other sports drinks or fluids. Please avoid soft drinks or caffeine. Remember that the increased fluid therapy is the most important part of the treatment of your POTS.

2. **Regular physical exercise.** This piece of advice is the hardest to follow since many people with POTS feel very weak and may become exhausted just walking around the home. Why exercise? People who have POTS are similar in physiology to astronauts who have been in space for a long time. Their bodies are no longer adapting to the effects of gravity and the heart seems to pump smaller amounts of blood at faster heart rates. By exercising, the heart gets stronger, pumps more blood with each heartbeat, and the heart rates become slower. Also, stronger muscles help more of the venous blood return to the heart and help prevent pooling of blood in the legs. There are studies that have demonstrated that the effects of exercise can be even more effective than using medications and it may even permit me to eventually stop your medications.

   - Start with exercise in a pool. The water pressure on your veins will help you start building up your fitness and help prevent you from feeling dizzy while exercising. Try just walking in the pool, or even doing some water jogging or water aerobics.

   - Do exercises that do not require standing upright at first. An excellent example is using a rowing machine; you are unlikely to get lightheaded and over time it will improve your fitness and muscle strength. Another workout that doesn't involve standing is using a recumbent stationary bicycle. Some days you may not be able to do very much apart from some isometric contractions, which consist of squeezing and contracting your muscles very forcefully for a long time, such as your hands, feet, arms, legs, and abdominal muscles. Any exercise is better than none!

   - The goal of the exercise program is to build up to a point where you can exercise at least 30 minutes three times a week or even more. Eventually build up to walking and progress to other gravity dependant sports, such as jogging and weight lifting. Build up to several hours a week if possible, but be patient, all this could take many months.

   - A study by Fu (JACC 55, 2858-2868, 2010) showed that half of a group of POTS patients were cured after a 3 month exercise training program of increasing intensity. Remember it is important to listen to your body. You may have good days and bad days and what counts is that you keep on trying.

3. **Changing sleep habits.** Many patients with POTS are tired and some even have chronic fatigue syndrome. Many don't feel rested after a night's sleep and end up oversleeping and taking naps. Healthy teens usually need 8 to 9 hours of sleep and some people with POTS may need up to 10 hours, but sometimes oversleeping can make you feel worse.

   - Please go to bed and wake up at the same time every day. Our brains do much better with consistent sleep and wake times.

   - Make sure you have a comfortable bed and a quiet, dark sleep environment.

   - Don't drink caffeine or exercise for 4 to 6 hours before bedtime.

   - Don't nap. If this is not possible, limit your nap to once a day and no longer than 30 minutes.

   - Speak with Dr. Le Gras if you have persistent sleep issues. We can explore whether there may be special sleep issues that would benefit from further testing, such as a sleep study, or whether you might benefit from certain medications (modafinil, stimulants) or other solutions.

4. **Compression garments.** This advice may not help everyone, but you may consider trying this option to see if it helps you. The idea is to wear special compression leg wraps that extend all the way up your legs and over your waist up to your bellybutton, or even over your whole abdomen. Wearing compression garments prevents abnormal blood pooling and the resulting fast heart rate symptoms, but in order for the compression garments to work they must be very tight. Some people feel uncomfortable with very tight compression garments or have had skin problems such as a rash or feeling hot and sweaty, or even having skin irritation and skin breakdown. You may need to experiment with different brands of compression garments and be careful at first to check your skin for problems. Some brands include Sigvaris (*sigvarisusa.com*) or Skins (*skins.net*), or you can search for other brands on the internet under the heading medical compression garments. Please let Dr. Le Gras know if you have found a brand that works well for you, it may be helpful to other patients.

5. **Avoid overheating.** When we are hot our bodies cool down by sending more blood to the skin – the blood vessels are vasodilated and we may look flushed. When this happens, there can be a drop in blood pressure and increase in heart rate along with other symptoms, so avoiding overheating is very important.

   - Avoid hot environments; use a fan or air conditioner.
   - Dress in layers so that you can take some clothes off when you are overheated.
   - Avoid hot tubs, saunas, or steam rooms.
   - Turn the water temperature down in the shower and use cool water near the end of the shower. Have a stool in the shower in case you need to sit down if feeling lightheaded or unwell. Since many people with POTS feel worse in the mornings, you may feel better showering in the evening or after you have had breakfast and lots of fluids.
   - Drink cool fluids. Stay in the shade. Wear a hat and dress in light colored clothing when in the sun.
   - Some people have used cooling vests in very warm weather and you may want to look into this. An example is *coolvest.com*.

6. **Having the right mindset and attitude:** Having a chronic disease like POTS is devastating and since many of your friends, neighbors, relatives, and teachers don't understand the illness, you may feel alone and misunderstood. So what can you do?

   - Talk to someone about your feelings – a parent, a friend, a teacher, or relative. Putting your feelings into words, even just writing them down, actually helps your brain process and cope better.
   - Maintain the fun in your life. Do things that you enjoy. Spend time with people that are important to you. I want you to make a list of things that are fun for you and make sure that you are doing those things on a regular basis. Don't let POTS define who you are.
   - Mobilize supports in your life: ask for help from people who you may know, or even don't know. Look to your religion or spiritual supports.

- Don't be hard on yourself. Don't blame yourself for having POTS. Go easy on yourself when you have a bad day and don't feel well – live life one day at a time. It may take time to have a new perspective on how things are different now with POTS. You will get better.

- Consider counseling. Many people with POTS feel anxiety, shame, self-doubt, hopeless, depressed and/or angry. Counseling can help you feel better and make some sense of what is happening to you. Counseling can help you sleep better and have the inner strength to fight POTS. Please talk to your parents if you feel like hurting yourself or using drugs or alcohol to cope. Your family and siblings may also feel better and have a better understanding of your point of view with counseling. Please speak with Dr. Le Gras or your PCP for assistance in setting up counseling if there are challenges in doing this.

7. **Make a school plan.** Many people with POTS are unable to attend school until they are feeling better. You will need to work out a plan that is best for you and which takes into account how you're feeling and how the school can accommodate your needs. Some people may need to have home schooling until they are feeling better, then return to school part time and increase school time depending on how they are doing.

8. **Avoid triggers.** The following is a list of situations that can make you feel worse or even faint:

   - Dehydration or inadequate fluids or fluid losses.
   - Inadequate food or skipping meals.
   - Too much or too little sleep.
   - Prolonged standing. Never stand motionless – this leads to blood pooling in your legs.
   - Prolonged sitting. Try to keep your legs elevated, otherwise there will be a lot of blood pooling in your legs.
   - Overheating.
   - Strong emotions, such as sudden terrible news.
   - Sudden pain, especially if unexpected.
   - Overexertion – this means doing too much exercise at once or working too hard.
   - Listen to your body – you need to adjust what you are doing when feeling unwell.

9. **Have an emergency plan.** When you are feeling very dizzy or unwell, lie down immediately or sit down with your head between your knees so that your head is lower than your heart. Ask someone to get you lots of water or other fluids. Do not stand up or try to walk outside for fresh air since this may cause you to faint or feel much worse. Your brain needs more blood flow and only gravity and rest will do that. You will also need to plan your day according to how you feel and some people do much worse in the mornings than in the afternoons and have to schedule their day accordingly.

10. ☐ **Increase your body weight – Dr. Le Gras will indicate if this applies to you.** For your height of __________, your ideal body weight is approximately __________. It is well established that people who are underweight can have more problems with low blood pressure, dizzy spells, and orthostatic hypotension or fainting and POTS. My goal is not to make you obese, but to get you to your ideal body weight or slightly above it – this can make a huge difference in how you feel. Ideally, you can increase your weight with a heart-healthy nutritious diet, but sometimes high-calorie, higher-fat content snacks (maybe even some "junk foods") are needed to gain weight, especially in people who have a poor appetite or a lot of nausea and gastrointestinal symptoms. If I ask you to gain weight, I will want you to weigh yourself on a weekly basis and chart the results and bring these along with you to your follow-up visit so we can view your progress.

THE INITIAL EVALUATION AND FOLLOW-UP VISITS

The initial visit with Dr. Le Gras focuses on the history of your symptoms and a complete medical history and physical examination. A follow-up visit may be needed to obtain more information to make the diagnosis and you may be asked to collect information from relatives, medical records, and test results if these could not be obtained at the initial encounter. Tests performed may include an ECG (evaluates the heart rhythm and certain disease markers) and/or echocardiogram (an ultrasound of the heart to evaluate the anatomy of the heart and confirm the function or how strong it is). Heart rhythm monitoring and other cardiac testing and blood work may be performed. Some tests may already have been performed – in some cases these tests may need to be repeated to ensure that no abnormality has been overlooked. Your PCP may have requested a "tilt test," but it is very unlikely that this test will be performed and Dr. Le Gras will make the final determination whether this test is required. A 10-minute orthostatic test may be performed. The initial consultation is done with a goal of determining the diagnosis, working out a treatment plan and educating you and your family on POTS.

Follow-up visits will be needed to assess how you are responding to treatments and to make changes if needed. When returning for follow-up visits, please bring a symptom diary using the monthly chart at the end of this handout (make extra blank copies please) where you record the following information:

- Record your total daily fluids, school attendance or absence, overall symptoms, and exercise and duration (example – walked 10 minutes).

- Indicate anything else that was important or relevant.

- Don't put too much detail in the diary because overall we are looking for trends in response to treatment. The charting also helps you stay accountable, actively involved in your care and actually making a difference in your outcome. Please don't forget to write down questions that you might have for me, ideas or thoughts that you would like to discuss or other observations that might be important.

- I will also indicate to you whether to chart your weight on a weekly basis if weight gain is desired.

MEDICATIONS

Most people with POTS will need to be on prescription medication. None of the medications are a cure, but it will help with symptoms. Medication doses may need to be adjusted depending on your response and sometimes are used on an as-needed basis, or in combination with other medicines. Please understand that any medication can have side effects and that Dr. Le Gras has considerable experience with these medications and will discuss them further with you. Remember that the perspective of a pediatric cardiologist with expertise in POTS will be much more realistic and tailored to your unique situation than what you will find on the internet.

☐ Fludrocortisone (Florinef)

This is the most frequently used medication in POTS. It is a steroid called a mineralocorticoid that will help increase your "circulating fluid volume." It is well tolerated and works well. It is taken in the morning and the dosage is usually 0.1 to 0.2 mg per day. A blood test to check your electrolytes will be done once you have been on this medication for two weeks; please remember to make sure that this blood work gets done. Call Dr. Le Gras' assistant for results two days after the blood test has been performed. Remember that if you are on fludrocortisone, you need to be seen in follow-up by Dr. Le Gras every six months in order to obtain refills for long term use.

☐ Midodrine (ProAmatine)

This medication tightens the blood vessels (this is called vasoconstriction), raises the blood pressure and starts to work within 15 minutes, but may only last two to four hours, so it may need to be prescribed three times daily. You may need to adjust the times of when you take it depending on how you feel when the medication is wearing off. The typical dose is 10 mg. It is best NOT to lie down to nap or sleep within four hours after taking this medication since the blood pressure can go up significantly when you are lying flat, but you can make an exception to this if you are feeling very dizzy or feeling faint. Dr. Le Gras may just have you take this medication as needed for days where you are feeling unwell or more lightheaded than usual. It can cause headaches, a feeling of goose-bumps, and sometimes this feeling of goose-bumps is a good indication that the medication is taking effect.

☐ Stimulant medications

These are the same medications that can be used for individuals who have ADD or ADHD, also known as Attention Deficit Disorder. They are also used in people who are having significant fatigue symptoms and they have the added benefit of raising the blood pressure by vasoconstriction. This medication will actually be prescribed by your PCP rather than Dr. Le Gras, but they will have a discussion regarding the use and dosage. Since these medications are "controlled substances," refills are not permitted without a new prescription.

☐ Modafinil (Provigil)

This medication is not a stimulant, but is helpful in people who have a lot of lethargy and fatigue symptoms and is also a vasoconstrictor. It has been helpful in patients who have had headaches and do not tolerate midodrine, and in people who have joint hyper-mobility syndrome. The usual dose is 50 mg daily in the morning or 25 mg on awakening and 25 mg at lunch.

☐ **Desmopressin (DDAVP)**

This medication was originally used for bedwetting and acts by causing fluid retention. At night when you are flat, the kidneys will eliminate a lot of excess fluid in your body, which is why people with POTS often feel worse in the morning. By retaining some of the fluid at night, the blood pressure is higher in the morning and people feel better. The usual dose is adjusted between 0.2 to 0.6 mg at bedtime. This medication also needs to be combined with a high salt diet. It is very important not to exceed the prescribed dose with this medication.

☐ **Propranolol (Inderal)**

This is a beta-blocker. Some people are very bothered by the fast heart rate episodes more than the other symptoms that are occurring with POTS. A low dose of propranolol blunts the pounding fast heart rate feeling without lowering the blood pressure. It is used once or twice a day at a dose of 10-20 mg depending on symptoms and response. The medication may not be used if a person has a history of depression or asthma.

☐ **Antidepressant medications: bupropion (Wellbutrin), paroxetine (Paxil), or sertraline (Zoloft)**

These medications act in the brain and there is solid medical literature showing that they can help in cases of fainting and POTS by raising the blood pressure and modifying the brain's response to low blood pressure signals. They are also beneficial in patients who have depression or anxiety issues. These would be prescribed by your PCP after discussion with Dr. Le Gras.

☐ **Pyridostigmine (Mestinon)**

This medication is a cholinesterase inhibitor. It is used in situations where other medications have not worked well enough. The typical dosage is 30-90 mg two to three times a day. It is very effective but there can be problems with stomach upset and it can cause diarrhea in some people.

☐ Other rarely used medications include epoetin, also known as EPO. This medication is rarely used since it is extremely expensive, needs to be monitored very closely and is given by injection. Another medication is octreotide, but its use is limited also by the fact that it has to be given by injection.

OTHER POSSIBLE OPTIONS

☐ **Neurology consultation**

Occasionally, seeing a neurologist is helpful, especially if there are concerns regarding the diagnosis or recurrent migraines, or other neurological issues. This can also be helpful if there is a question regarding problems with balance or vestibular function.

☐ **Endocrinology consultation**

A consultation with an endocrinologist is sometimes used in situations where there is possible hypoadrenalism (low or abnormal adrenal function), thyroid disease, or diabetes.

☐ **Eating Disorder consultation**

Patients who are initially noted to have very low heart rates or very low blood pressure and issues with fainting and dizziness may sometimes actually have an eating disorder rather than POTS syndrome. In these situations, the most effective therapy is treating the underlying eating disorder. We are fortunate to have an extremely experienced and dedicated eating disorder team in the Portland area and more information is available at *kartiniclinic.com*.

☐ **Gynecology consultation**

In situations where POTS symptoms are aggravated by menstrual periods, some women benefit from suppression of their menses with medications such as birth control pills. If we take this approach, it could either be organized with a gynecology consultation or with your PCP.

☐ Referral to another POTS specialist is sometimes helpful if you or I, or both of us would like a fresh look at the situation. This could be a one-time second opinion, combined care with that person with occasional visits to them, or a transfer of care if desired – what matters most is improving your situation.

☐ Please bring your ideas and observations to the next office visit. There may be patterns that you have noticed that are helpful in managing or assessing your POTS. On the other hand, you may have some misperceptions or habits that may be aggravating some of your symptoms and benefit from further discussion.

Name: Month: Year: ☺ Felt good 😐 Mild symptoms ☹ Unwell

Monday	**Tuesday**	**Wednesday**	**Thursday**	**Friday**	**Saturday**	**Sunday**
Fluids School Y N Exercise Date						