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POTS Handout

This handout was originally written by Dr. Marc LeGras, retired pediatric cardiologist.
It is provided for education and to help with POTS and POTS-like symptoms.

What is POTS?

POTS is an abbreviation for **P**ostural **O**rthostatic **T**achycardia **S**yndrome. It is in a category of disorders known as "Autonomic dysfunction." In simple terms, it 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, which is called "hypotension." At times, people can lose consciousness or faint; this is called "**syncope**" (pronounced "SINK-oh-pee"). 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 (6) months of symptoms, but we don't need to wait that long to start helping 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 one (1) liter of blood 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 when 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 sometimes also triggers an increased heart rate from anxiety. There are then areas in the brain that receive this signal of a fast heart rate and cause an abnormal stress response where a cranial (brain) nerve called the "vagal" (or "vagus") nerve quickly lowers the heart rate. The fainting that is then caused by the sudden drop in heart rate combined with the sudden drop in blood pressure called **neurocardiogenic** or **vasovagal 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 the heart rate when changing from sitting or lying down to standing. There may also be an abnormality in the ability of the blood vessels to tighten (or vasoconstrict) and/or in the brain's blood pressure and heart rate regulation. Usually in pediatric patients, the heart is completely normal. Some pediatrics patients may have hypermobile joints or Ehlers-Danlos syndrome. These patients have very "stretchy" veins, which can cause the abnormal blood pooling. Other situations which can cause POTS or POTS-like symptoms include certain medications, hyperthyroidism, neuropathy, low adrenal function, and disordered or restricted eating. 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 (4) times more common 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. Some patients may find a cure and have their symptoms go away completely, but in many it is very difficult to treat, and it may go 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. May 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, and inability to tolerate heat or standing are sometimes part of this chronic illness.

How will I get better?

The therapies listed in this handout are the most helpful therapies to alleviate the symptoms of POTS. Over time, the therapies may change as you feel better and have a better understanding of your condition.

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 for teens and 3-5 grams of salt for younger children and smaller teens – that is a lot of salt. The best way is to eat salty foods such as pretzels, salty crackers, canned soups, and by adding extra salt to your food. Some people prefer to take salt pills or salt tablets (sodium chloride, 1000 mg (1 g), 1 to 2 pills three times a day), but salt pills can cause stomach upset, so we recommend a dietary approach first.
    - ♥ **You need to dramatically increase your fluid intake.** We 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 to do that is to keep a record of how much you are drinking as the day goes on, or drink from a water bottle with the volume printed on it that shows your progress. If you do not like to drink water, find fluids that you *do* like, such as Gatorade or other sports drinks. Please avoid soda/pop, caffeine, and energy drinks. Remember that the increased fluid therapy is the most important part of the treatment of your POTS symptoms.

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 house. 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 some people to eventually stop their medications.

- ♥ If you have access to a swimming pool, start with swimming exercises. 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 does not 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 consists 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. Eventually, you want to build up to walking and progress to other gravity-dependent sports, such as jogging and weightlifting. Work to build up to several hours of exercise a week if possible but be patient; 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 three-month exercise training program of increasing intensity. Remember it is important to listen to your body. You may have good days and bad days, but what counts is that you keep trying.

3. **Changing sleep habits.** Many patients with POTS are tired and some even have chronic fatigue. Many don't feel rested after a night's sleep and end up oversleeping and taking daytime 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.

   - ♥ Go to bed and wake up at the same time every day. Our brains do much better with consistent sleep and wake times.
   - ♥ Don't drink caffeine or exercise for 4 to 6 hours before bedtime.
   - ♥ Don't nap during the day. If this is not possible, limit your nap to once per day and no longer than 30 minutes.
   - ♥ Make sure you have a comfortable bed and a quiet, cool, dark sleep environment.
   - ♥ Put smartphones/tablets away one hour before bedtime. The blue light emitted by smartphones/tablets can decrease melatonin production and affect your internal body clock which makes it harder to fall asleep and stay asleep.
   - ♥ Talk to your primary care provider/pediatrician if you have persistent sleep issues. They can explore whether there may be special sleep issues that would benefit from further testing, such as a sleep study, or from medications 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 belly button, 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, they can cause you to feel hot or sweaty and can sometimes cause skin irritation or a rash. You may need to experiment with different brands and be careful at first to check your skin for problems. Some brands include Sigvaris (*sigvarisusa.com*) or Skins (*skinscompression.com*), or you can search for other brands on the internet under "medical compression garments."

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 or red. 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 an air conditioner.
- ♥ Dress in layers so that you can take some clothes off when you are overheated.
- ♥ Avoid hot tubs, saunas, and steam rooms.
- ♥ Avoid prolonged hot showers. Turn the water temperature down and use cooler water near the end of your showers. 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 after you have had breakfast and drank some fluids or in the evenings.
- ♥ 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.

6. **Having the right mindset and attitude.** Having a chronic illness like POTS is difficult 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 relative, a teacher, or a counselor. Putting your feelings into words, even just writing them down, helps your brain cope and process emotions better.
   - ♥ Do things that you enjoy. It's important to try to maintain the fun in your life. Spend time with people that are important to you. Make a list of things that are fun for you and do those things on a regular basis. Don't let POTS define who you are.
   - ♥ Mobilize supports in your life. Don't be afraid to ask for help when you need it. Look for support groups online or within your community or church.
   - ♥ 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 with POTS. Believe in yourself and that you will get better.
   - ♥ Consider mental health counseling. Many people with POTS feel anxiety, self-doubt, hopelessness, shame, anger, and/or depression. 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 members may also get a better understanding of your illness with counseling. Please talk to your cardiologist or primary care/pediatrician if you need help finding counseling.

7. **Make a school plan.** Many people with POTS are unable to attend school until they feel 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 homeschooling or online schooling until they are feeling better, then return to school part time and increase school time depending on how they are feeling.

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

   - ♥ Dehydration, inadequate fluid intake.
   - ♥ Skipping meals, inadequate nutrition
   - ♥ Too much or too little sleep
   - ♥ Prolonged standing. Never stand motionless – this leads to blood pooling in the legs.
   - ♥ Prolonged sitting. Try to keep your legs elevated, otherwise blood will pool in the legs.
   - ♥ Overheating.
   - ♥ Strong emotions.

- ♥ Sudden pain, especially if unexpected.
- ♥ Overexertion, doing too much exercise at once or straining to lift something heavy.
- ♥ 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 talk 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 plan their day accordingly.

10. **Increase your body weight (may not apply; your doctor will indicate if this applies to you.)** 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. The goal is not to make you overweight or 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 or other gastrointestinal symptoms.

MEDICATIONS

Many people with POTS will need to be on medication. None of the medications are a cure but they can 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.

- ♥ Fludrocortisone (Florinef)
  This is the most frequently used medication in treating POTS. It is a steroid called a "mineralocorticoid" that will help increase your "circulating fluid volume." It is usually well-tolerated and works well to stabilize blood pressure. It is typically 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 the medication for two weeks. It is important to not miss this blood work. If you are taking fludrocortisone, you will need to be seen in follow-up by pediatric cardiology (or whoever is prescribing the medication) every six (6) months to keep getting refills.

- ♥ Midodrine (ProAmatine)
  This medication tightens the blood vessels (called "vasoconstriction"), raises the blood pressure, and starts to work within 15 minutes but may only last two to four (2-4) hours, so it may need to be prescribed and taken three (3) times daily. You may need to adjust the times of when you take it depending on how you feel when the effects are wearing off. The typical dose is 10 mg. It is best to NOT lie down to nap or sleep within four (4) hours of taking the medication since your blood pressure can go up significantly when you are lying flat. You can make an exception to this if you are feeling dizzy or more lightheaded than usual. This medication can cause headaches and a feeling of goosebumps, but sometimes this feeling of goosebumps is a sign that the medication is taking effect.

- Propranolol (Inderal)
  This medication is known as a beta-blocker. Some people are very bothered by the fast heart rate episodes more than any other symptoms that are occurring. A low dose of a beta-blocker will blunt the feeling without lowering the blood pressure. It is usually taken once or twice a day at a dose of 10-20 mg depending on your symptoms and response to the medication. Beta-blockers cannot be used if a person has a history of depression or asthma.

OTHER POSSIBLE OPTIONS

- Neurology consultation
  Occasionally, seeing a neurologist (brain doctor) is helpful, especially if there are concerns regarding the diagnosis of recurrent migraines or other neurological issues. This can also be helpful if there is a question of possible vertigo or vestibular dysfunction. This would require a referral from your primary care provider or pediatrician.

- Endocrinology consultation
  A visit with an endocrinologist is sometimes helpful where there is possible abnormal adrenal gland function, thyroid disease, or diabetes. This would require a referral from your primary care provider or pediatrician.

- Gynecology consultation
  In situations where symptoms are made worse by menstrual cycles, some patients benefit from the suppression of their menses with medication such as oral contraceptives. This visit could be with either your pediatrician or a gynecologist. This would require a referral from your primary care provider or pediatrician.

- Eating Disorder consultation
  Patients who have issues with avoidant/restricted food intake disorder (ARFID) can often have low heart rates or very low blood pressure. In these cases, 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 at the Kartini Clinic. More information can be found at **KartiniClinic.com**. This would require a referral from your primary care provider or pediatrician.

- Mayo Clinic referral
  For families who can travel, the Mayo Clinic has a dedicated POTS clinic called the "Adolescent Autonomic Dysfunction Clinic." This service is currently only available at their Rochester, Minnesota, location. Our office can send them a referral after you have been seen in our clinic.

Please bring your observations and ideas to your office visit with the pediatric cardiologist. There may be patterns that you have noticed that are helpful in assessing or managing your symptoms.