Patient & Family Guide
2021

Pulmonary Arterial Hypertension (PAH) Program

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Pulmonary Arterial Hypertension (PAH) Program

The Pulmonary Arterial Hypertension (PAH) Program coordinates the assessment, care, and treatment of people with suspected or diagnosed PAH.

What is PAH?

Pulmonary hypertension is a rare condition that causes high blood pressure in the blood vessels that connect the heart and lungs (pulmonary arteries).

If the high blood pressure is caused by a progressive (gets worse over time) disease of the pulmonary arteries, it is known as Pulmonary Arterial Hypertension (PAH).

PAH is a condition that progresses at different rates in different people. Symptoms may include:

- Shortness of breath
- Tiredness
- Dizziness and/or fainting
- Swelling of the legs and/or abdomen (stomach area)

What causes PAH?

In many cases, the cause of PAH is unknown. PAH may run in a family. It may also be related to:

- heart defects you were born with.
- autoimmune diseases like scleroderma or lupus.
- advanced liver disease.
- HIV/AIDS.

Your treatment will depend on the cause and level of your PAH and how you respond to treatment.

Why have I been referred to the PAH Program?

The doctor who referred you to the Program suspects that you may have PAH. We will find out whether or not you have PAH, and decide how best to help you.

How will the PAH Program help me?

When you visit the Clinic, you will meet with the PAH nurse coordinator and one of the PAH doctors. The team will review your lab test results and decide if you need more tests.

These tests will help us find out:

- if you have PAH.
- what may be causing it.
- how severe (bad) your condition is.
- what treatments may be best for you.

What tests will be done?

You **may** need the following tests:

☐ **Blood work:** Blood tests help us screen for possible causes of PAH.

☐ **Electrocardiogram (ECG):** This is a painless test that records the electrical impulses in your heart.

☐ **Echocardiogram ('Echo'):** This test uses an ultrasound to create pictures of your heart. It tells us how well your heart is working.

☐ **6-minute Walk Test:** You will be asked to walk in a hallway at your own pace while we monitor your symptoms and oxygen level. How far you can walk and the changes in your oxygen level tell us how much you are limited by PAH, and if your treatment is working.

☐ **Pulmonary Function Tests (PFTs):** These tests tell us how well your lungs are working.

☐ **CT Angiogram:** This special chest CT scan uses X-rays to look at the blood vessels and tissue in your lungs. It tells us if there are clots in the vessels and if the tissue has been scarred. An intravenous (IV) and an injection of dye are needed.

☐ **Ventilation/Perfusion Scan (V/Q):** This test measures ventilation (airflow) and perfusion (blood flow) in your lungs to find chronic (ongoing) clots in the blood vessels. This test has 2 parts:

- › You will be asked to breathe in oxygen that has been mixed with a radioisotope (radioactive material). Then we will take pictures of your lungs.
- › A radioisotope will then be injected into a vein in your arm using a needle. Then we will take more pictures of your lungs.

If your health care team needs more information, you may also need these tests:

☐ **Right Heart Catheterization:** This test measures the blood pressure in your lungs and heart. An IV in your neck or groin is needed. You may be asked to inhale (breathe in) oxygen and nitric oxide (a gas with no taste or smell) to see if this changes the pressure.

- ☐ **Sleep study:** This test checks for sleep apnea (a condition where you stop breathing for a few moments at a time while sleeping). It can be done in your home or in the sleep lab in the hospital.
- ☐ **Exercise Echocardiogram:** An ultrasound of your heart is done during an exercise test. The test may take up to 1 hour, including about 7 to 12 minutes of exercise.

How is PAH treated?

Treatment of PAH depends on how severe your condition is, and in some cases, the cause of the disease. After your tests are done, you will be asked to return to the PAH Clinic to talk about what medications or treatments are best for you. Oral (by mouth) or IV medications may help. Many people feel much better and are able to do more once they start treatment.

Surgery options

A lung transplant may be considered when medications no longer help. The PAH team may refer you for a lung transplant assessment. Not everyone is a candidate for lung transplant.

If your pulmonary hypertension is caused by chronic (ongoing) blood clots in the lungs (CTEPH), you may be a candidate for another surgery (pulmonary endarterectomy or PEA).

If you need a lung transplant or PEA, you will be referred out of the province, usually to Toronto.

How often do I have to visit the Clinic?

Once you start treatment, you will need regular tests in the Clinic. How often you are seen depends on your condition and how you respond to treatment.

For more information:

Pulmonary Hypertension Association of Canada

› www.phacanada.ca

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Notes:

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