

Pulmonary Arterial Hypertension (PAH) Program

Halifax Infirmary, QE II

Notes:

This pamphlet is for educational purposes only. It is not intended to replace the advice or professional judgment of a health care provider. The information may not apply to all situations. If you have any questions, please ask your health care provider.

Find this pamphlet and all our patient resources here:
<https://library.nshealth.ca/Patients-Guides>

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Call 811 or visit: <https://811.novascotia.ca>

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Pulmonary Arterial Hypertension (PAH) Program

The Pulmonary Arterial Hypertension (PAH) Program assesses and treats people with possible or diagnosed PAH. You have been referred to the PAH Program because your primary health care provider (family doctor or nurse practitioner) thinks you may have PAH.

What is PAH?

- Pulmonary hypertension is a rare condition that causes high blood pressure (hypertension) in the blood vessels that connect your heart and lungs (pulmonary arteries).
- If the pulmonary hypertension is caused by a progressive (gets worse over time) disease, it is known as pulmonary arterial hypertension (PAH). PAH progresses at a different rate in each person.
- Symptoms may include:
 - › Trouble breathing
 - › Swollen legs
 - › Tiredness
 - › Swollen abdomen (stomach area)
 - › Dizziness
 - › Fainting

How often do I have to visit the Clinic?

- Once you start treatment, you will need to have regular tests at the Clinic. How often you are seen will depend on:
 - › Your condition
 - › How your body responds to treatment

PAH Program

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For more information:

Pulmonary Hypertension Association of Canada

- › www.phacanada.ca

- **Exercise echocardiogram:** This test uses an ultrasound to take pictures of your heart while you exercise. The test may take up to 1 hour, including about 7 to 12 minutes of exercise.

How is PAH treated?

After your tests, you will return to the PAH Clinic to talk about what treatment is best for you. Many people feel much better and are able to be more active once they start treatment.

Medication

- You may need to take medications by mouth or through an I.V.

Surgery

- If medications are no longer helping, the PAH team may refer you for a lung transplant assessment. Not everyone is a candidate for a lung transplant.
- If your pulmonary hypertension is caused by chronic blood clots in your lungs (CTEPH or chronic thromboembolic pulmonary hypertension), you may be a candidate for:
 - › Pulmonary endarterectomy (PEA)
 - › Balloon pulmonary angioplasty (BPA)
- If you need surgery, you will be referred out of Nova Scotia, usually to Toronto.

What causes PAH?

- In many cases, the cause of PAH is not known.
- PAH may be passed from a parent to a child. It may also be related to:
 - › Heart defects you were born with
 - › Autoimmune diseases (like scleroderma or lupus)
 - › Advanced liver disease
 - › HIV/AIDS
 - › Side effects of medication
- Your treatment will depend on:
 - › The cause of your PAH
 - › The level of your PAH
 - › How your body responds to treatment

How will the PAH Program help me?

- At your first visit, you will meet with a PAH nurse coordinator and a PAH doctor. They 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 your PAH.
 - › how severe (bad) your PAH is.
 - › what treatments may be best for you.

What tests will I have?

You may need the following tests:

- **Blood work:** Blood tests help us find possible causes of your PAH.
- **Electrocardiogram (ECG or EKG):** This is a painless test that measures your heart's reaction to activity.
- **Echocardiogram (also called an echo):** This test uses an ultrasound to take pictures of your heart. It tells us how well your heart is working.
- **6-minute walk test:** You will walk in a hallway at your own pace while we check your symptoms and oxygen level. How far you can walk and the changes in your oxygen level will tell us:
 - › how much you are affected by your PAH.
 - › if your treatment is working.
- **Pulmonary function tests (PFTs):** These tests tell us how well your lungs are working.
- **Computerized tomography (CT) angiogram:** This CT scan of your chest 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 is scarred. For this test, you will have dye injected into a vein in your arm through an intravenous (I.V.) tube.

- **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 – the health risks are low for small amounts of radiation). Then we will take pictures of your lungs.
 - › Then a radioisotope will be injected into a vein in your arm using a needle. 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.
 - › You will need an I.V. tube in a vein in your neck or groin.
 - › You may be asked to breathe in oxygen that has been mixed with 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 minutes at a time while sleeping). It can be done in your home or in the sleep lab at the QE II.