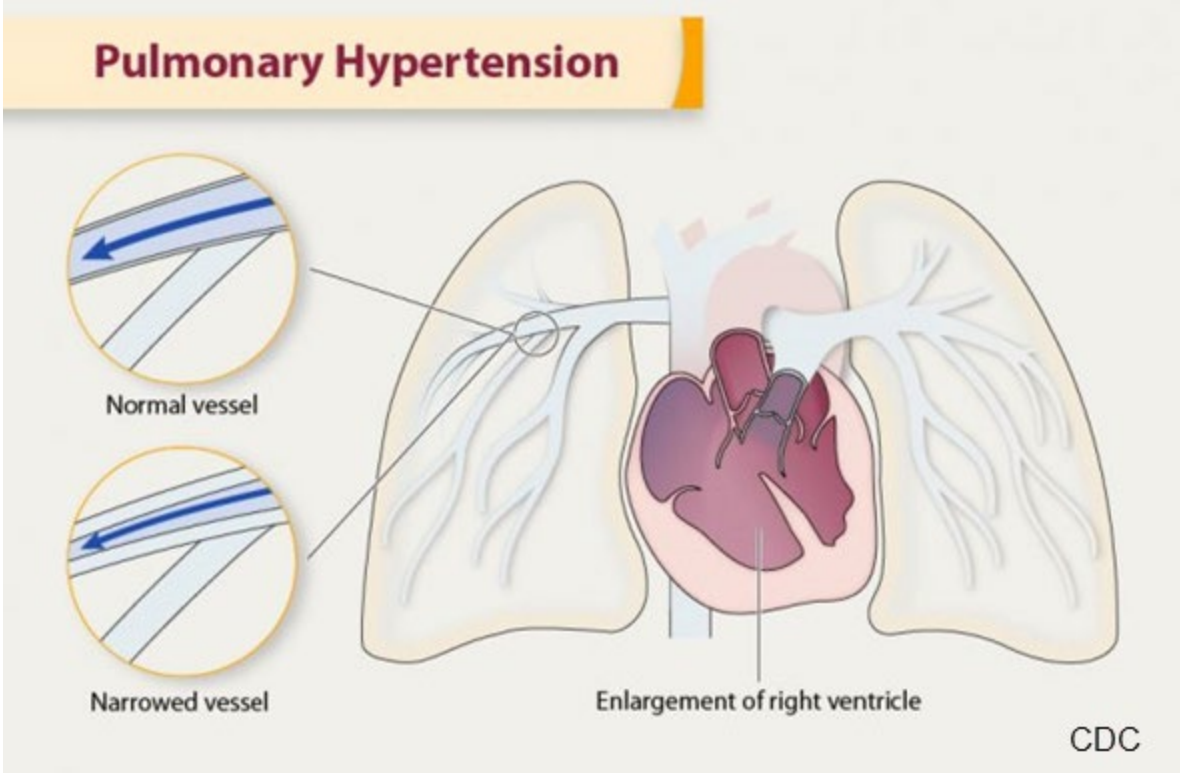
FRANKEL CARDIOVASCULAR CENTER
MICHIGAN MEDICINE

Pulmonary Hypertension

What is pulmonary hypertension?

Pulmonary hypertension (PH) is a general term used to describe high blood pressure in the **pulmonary arteries** (blood vessels of the lungs). PH is different than ordinary high blood pressure, which occurs throughout the body. In PH, the pulmonary arteries can become stiff and narrow, and the right side of the heart must work harder to pump blood through them.



During pulmonary hypertension, the arteries in the lungs can narrow and then the blood does not flow as well as it should, resulting in less oxygen in the blood.

What are the types of pulmonary hypertension?

Not all PH is the same. There are five different types of pulmonary hypertension based on different causes. Types of pulmonary hypertension include:

Group 1: Pulmonary arterial hypertension (PAH)

A disease in which the blood vessels of the lungs become thick and narrow, causing raised pressure.

There are several types of PAH:

- Idiopathic (no clear cause)
- Inherited (condition is genetically passed from parents to their children)
- Associated with other medical conditions including connective tissue diseases such as scleroderma and lupus, congenital heart disease, liver disease, HIV, and using drugs or toxins.

Group 2: Pulmonary hypertension caused by left heart disease

This can be caused by a weakness or stiffness of the left side of the heart, or problems with left sided heart valves.

Group 3: Pulmonary hypertension caused by lung disease or low oxygen levels

This can be caused by:

- Diseases where the lung airways become narrow such as COPD or emphysema
- Diseases that make it hard for the lungs to expand such as pulmonary fibrosis
- Sleep apnea
- Living in an area of high altitude for a long period of time

Group 4: Chronic thromboembolic pulmonary hypertension (CTEPH):

Pulmonary hypertension caused by the body's inability to dissolve blood clots in the lungs.

Pulmonary hypertension due to unknown causes:

This type of PH is associated with other diseases in ways that are not well understood. These conditions include:

- Sarcoidosis
- Sickle cell anemia
- Chronic hemolytic anemia
- Spleen removal
- Certain metabolic disorders

What are the symptoms of pulmonary hypertension?

Symptoms are common across all types of pulmonary hypertension and may include:

- Shortness of breath
- Feeling tired or worn out (fatigue)
- Chest discomfort or pain
- Palpitations or a racing heart beat
- Lightheadedness or fainting
- Swollen ankles/legs (edema) or abdomen (ascites)

Once you've been diagnosed with pulmonary hypertension, your PH team will use a scale to classify the severity of your symptoms into one of several classes, including:

- Functional class I: no symptoms during ordinary physical activity
- Functional class II: ordinary physical activity is **somewhat** limited by breathlessness, chest pain, fatigue or dizziness
- Functional class III: physical activity is **greatly** limited by breathlessness, chest pain, fatigue or dizziness while doing routine things
- Functional class IV: may be breathless and tired **even while resting** and can't do any physical activity without symptoms. Anyone with fainting is considered to be in this class.

Because many people do not get diagnosed with PH in its early stages, many will be rated at a higher class when diagnosed. However, with treatment it is possible to be moved into a lower class.

How is pulmonary hypertension diagnosed?

PH can be difficult to diagnose as it has many symptoms that are similar to other heart and lung conditions. Your doctor may recommend several tests to confirm your diagnosis. The most common tests are listed below:

Blood tests – A small amount of blood is drawn and examined. These tests are used as a general health assessment and to look for markers of specific

diseases associated with Group 1 PAH such as kidney, liver and thyroid function; HIV, and connective tissue diseases.

Electrocardiogram (ECG) – Electrodes with wires are put on your chest, arms, and legs. Electrodes feel like sticky patches and connect you to the EKG machine, which graphs the electrical activity of your heart. Your doctor checks:

- How fast the heart is beating
- Whether the beats are regular
- If there is any previous heart damage
- If there are specific findings of ph.

Chest X-Ray – A chest x-ray is a picture of the heart and lungs; you are asked to sit or stand in different positions while holding your breath so that a picture can be taken. This test looks at the general shape of the heart to see if the chambers are larger than expected. It also checks if the pulmonary arteries (lung blood vessels) are more visible than normal and looks for signs of lung disease or fluid in the lungs.

Pulmonary Function Tests (PFTs) – PFTs measure how well your lungs take in air and exhale air and how efficiently they transfer oxygen into the blood. You breathe into a mouthpiece that is connected to a spirometer. A spirometer records how well your lungs work. The test usually takes 1 hour.

Computerized Tomography (CT) Scan – A chest CT scan is a test that uses an x-ray system to take detailed images of the structures in the chest, including the heart and the lungs. Some CT scans require the use of a contrast "dye" to make the images clearer. If you need contrast material for your exam, it will be injected directly into your vein through an I.V. in your arm. This test helps evaluate the size of the lungs, larger blood vessels in the lungs, and the lung tissue.

Ventilation/Perfusion (V/Q) Scan – This test is done to determine whether you have a blood clot in the blood vessels of the lungs. It uses two different scans: ventilation and perfusion. During the **ventilation scan**, you breathe in a medication mixed with oxygen through a mask while the scanner takes

pictures. During the **perfusion scan**, you receive a medication injected into a vein in the arm, so the scanner can see which sections of the lungs are getting enough blood flow.

Exercise Tolerance Test (6-Minute Walk Test) - The purpose of this test is to identify your exercise tolerance level. You are asked to walk at your normal pace for 6 minutes. The results of this test help your doctor evaluate your ability to perform physical activity.

Cardiopulmonary Exercise Test – During this test, you exercise on a stationary bike, while ECG, blood pressure and oxygen use are measured. This may help your doctor evaluate how well the heart and lungs respond to exercise.

Echocardiogram – An **echo** is an ultrasound of the heart that uses sound waves to measure:

- Your heart's size and shape
- How well it is squeezing
- How the right side of the heart looks
- How fast the blood is moving

This test gives an estimate of the blood pressure in the lungs. This tests usually takes 30 minutes. A gel is put on your chest and an ultrasounds probe is moved around your chest while pictures are taken.

Cardiac Magnetic Resonance Imaging (MRI) – If images from an echocardiogram are unclear, your doctor may use a cardiac MRI to look at heart size, structure and function. This test uses a powerful magnetic field and radio waves to take pictures of the inside of the body with a special machine like an x-ray, but more detailed.

Sleep studies

Overnight oximetry is a test that measures oxygen levels in the blood during the night. During this test, you wear a plastic clip over the end of the finger throughout the night, which can be done at home.

Polysomnography is a recording of brain waves, used to find out if there are periods where breathing stops during sleep. This test is done at an overnight sleep center.

Genetic counseling and testing - In some cases, a **gene mutation** (any change in the DNA sequence of a cell) is inherited from family members. This increases the risk of PH. We'll help you identify these risks and discuss the option of genetic counseling and testing.

Additional tests may be recommended by your doctor and could include the following:

Right-Heart Catheterization – This is considered the best test for a definitive diagnosis of PH. This test directly measures the pressure and flow of blood inside the heart and blood vessels of the lungs. During the test, doctors insert a catheter (a thin rubber tube) through a large vein in the neck or groin and passes it to the heart to measure the blood pressure and fluid levels in your heart and lungs.

Pulmonary Angiogram – A pulmonary angiogram is a test that looks at blood vessel structures and branches in the lungs. During this test, a dye is injected into the lung blood vessels to make them appear more clearly on a screen. This test may be done to evaluate for blood clots in the lungs.

How is pulmonary hypertension treated?

Pulmonary hypertension should be evaluated and managed by a specialist, as there are numerous treatment options to help manage this rare condition. Treatments include overall healthy lifestyle behaviors and medications. A lung transplant may be an option depending on the severity of PH. Our treatments include **targeted treatments,** which are specific drugs used to treat the factors causing your condition.

Oral (by mouth) targeted treatments:		
Medication:	Purpose:	Brand names:
Phosphodiesterase type 5 inhibitors (PDE 5 Inhibitors)	Allow the lungs to produce more of its own natural vasodilators (widens narrowed blood vessels).	• Sildenafil (Revatio®) • Tadalafil (Adcirca®)
Endothelin receptor antagonists (ERAs)	Help prevent the blood vessels from narrowing.	• Ambrisentan (Letairis®) • Bosentan (Tracleer®) • Macitentan (Opsumit®)
Prostacyclin Analogue-	Allows the blood vessels in the lungs to relax	• Treprostinil (Orenitram®)
Selective IP Receptor Stimulator	Helps the blood vessels in the lungs relax	• Selexipag (Uptravi®)
Soluble Guanylate Cyclase Stimulators	Helps the blood vessels in the lungs relax.	• Riociguat (Adempas®)

Inhaled targeted-treatment options (nebulizer):		
Medication:	Purpose:	Brand name:
Prostacyclin Analogues	Relieves shortness of breath.	• Iloprost (Ventavi®s) • Inhaled Treprostinil (Tyvaso®)

Subcutaneous (into the skin) **and intravenous** (in the vein) **targeted-treatment options:**		
Medication:	Purpose:	Brand names:
Prostacyclin Analogues	Opens up the blood vessels and helps ease symptoms of PH.	• Epoprostenol (Flolan®) • Room Temperature Stable Epoprostenol (Veletri®) • Treprostinil (Remodulin®)

Three abnormal chemical imbalances are known to contribute to PH by causing the blood vessels in the lungs to tighten. PH-targeted medications help to change these chemical imbalances so the blood vessels in the lungs can relax, allowing blood to flow more easily through them. By lowering the blood pressure in the lungs, the right side of the heart can also pump more

effectively. All of the medications above are approved by the FDA (Food and Drug Administration).

Surgeries and procedures

If you have Chronic Thromboembolic Pulmonary Hypertension (CTEPH), your doctor may recommend the following procedures:

- **Balloon pulmonary angioplasty (BPA):** A procedure in which a deflated balloon is directed to the diseased lung vessels and inflated to break up blood clots and restore blood flow.
- **Pulmonary thromboendarterectomy (PEA):** A surgical procedure to remove blood clots from the lungs to restore blood flow.

The Pulmonary Hypertension Program at Frankel Cardiovascular Center is the largest and most experienced program in the state, and one of the largest in the country. We are a comprehensive resource for the care and treatment of patients who live with this challenging disease.

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