Pulmonary Hypertension Information

What is pulmonary hypertension?

Blood pressure is the force (tension) that the blood exerts in our blood vessels. When blood vessels get narrower, blood pressure increases. In some people, the blood vessels in their lungs become narrowed, causing high blood pressure in the lungs. This is a complex health problem called pulmonary (PULL-muh-nair-ee) hypertension (hi-per-TEN-shun). It is called "PH" for short. **Pulmonary** refers to the lungs. **Hypertension** is the medical name for high blood pressure.

PH may also scar the blood vessels in the lungs. For the heart to push blood through smaller, scarred blood vessels, the heart's right side must work harder. Over time, the heart can enlarge and weaken. This may lead to congestive heart failure.

What Are the Types of Pulmonary Hypertension?

PH falls into the 4 main types listed below:

- **Pulmonary Arterial Hypertension** from thickening and narrowing of the arteries that carry blood from the heart's **right side** to the lungs. It may be primary or secondary. When the cause is not known, it is called idiopathic (id-ee-oh-PATH-ik) pulmonary hypertension (IPH). Another name is **primary** pulmonary hypertension (PPH).

  When another health problem is the cause, it is called **secondary** pulmonary hypertension. Some possible causes are:
- Collagen vascular disease, such as lupus, scleroderma, and rheumatoid arthritis
- Congenital heart defects, which are present at birth
- Severe liver disease
- HIV infection
- Certain weight loss medicines
- **Pulmonary Venous Hypertension** from diseases of the **left side** of the heart
- **Pulmonary hypertension** from **respiratory disease**, such as emphysema, sleep apnea, interstitial lung disease, and long-term high altitude exposure
- **Pulmonary hypertension** from **chronic blood clotting** in the lungs

What are the Symptoms of Pulmonary Hypertension?

The symptoms of PH may be similar to symptoms of other illnesses. This can make it difficult for doctors to identify PH. Common symptoms of PH include:

- Shortness of breath
- Extreme tiredness (fatigue)
- Fast heart beat (palpitations)
- Fainting
- Chest pain
- Swelling in the legs

How is Pulmonary Hypertension Detected?

There are many possible causes of PH. To decide on the best treatment for you, the doctor must find the exact cause of your PH. The doctor also needs to identify any other disorders you may have that are related to PH. You will need to have many tests, which your doctor will carefully review. Below is a list of tests that you may have:

- **Blood tests:** to detect other illnesses that can cause PH
- **Echocardiogram**: to look at your heart by making pictures of it with sound waves. This test checks the heart for size, ability to function, and blood flow.
- **Ventilation-perfusion scan:** to look at your lungs after injecting dye through an IV (intravenous line). This test checks how well air and blood go through your lungs and if there are blood clots in your lungs.
- **Pulmonary function test:** to measure how much air and how fast you can breathe into a machine. The test checks for lung diseases such as asthma or emphysema.
- **CT scan**: to look at your lungs by computerized x-rays of the chest. The test checks for lung tissue that is scarred or destroyed.

- **Heart catheterization** (KATH-it-er-iz-AY-shun): to measure exact blood pressure in your heart and arteries. A thin, flexible tube is inserted into an arm, leg, or neck vein, and then threaded into the heart and lung artery. A dye may be injected into the heart arteries to look for blockages. This test can show if you have PH, how severe it is, and what is causing it.
- **Sleep study**: to check oxygen levels in your blood while you sleep. The test shows if you have periods when you stop breathing for a few seconds during the night — called sleep apnea (AP-nee-uh).

How is Pulmonary Hypertension treated?

Several recent advances have been made in treatments for PH. This section tells you about the main types of treatment.

Medicines

The generic name of each medicine is given first; the brand name is second.

For blood pressure in the lungs:

- **Bosentan (Tracleer):** a pill taken twice a day to help reduce pressure in your lungs, enable you to exercise more, and slow down the disease. You must have blood tests every month to check your liver, because this medicine may harm the liver. If a liver problem develops, your liver should return to normal if you stop the medicine or take a lower dose. You should avoid pregnancy while on this medicine because it can harm an unborn child.
- **Epoprostenol (Flolan):** an IV (intravenous line) medicine that you receive non-stop. It requires a permanent IV catheter inserted into a vein and a portable pump. This medicine is usually given to patients with more serious illness. It can help to lessen your symptoms, enable you to exercise more, and reduce blood pressure in your lungs. It may prolong life. Some side effects may occur, such as headache, diarrhea, facial redness (flushing), jaw pain, muscle pain, and infection at the catheter site.
- **Treprostinil (Remodulin):** a shot (injection) into the fatty tissue on your belly. A pump the size of a pager is worn on the belt. The benefits of this medicine are similar to those of epoprostenol. The most common side effect is pain where the medicine goes into the skin. Infection is much less common with this medicine.
- **Calcium channel blockers**: a pill to help relax narrowed blood vessels in the lungs. This medicine works only for a small number of patients.

For heart failure and fluid build-up:

- **Digoxin:** a pill to help the right side of the heart work better
- **Diuretics:** a pill to help reduce fluid build-up by increasing urine flow. These medicines are also called "water pills."

Other therapies

- **Warfarin (Coumadin):** a pill taken once a day to prevent blood clots from forming
- **Oxygen Therapy**: inhaled through the nose by a tube that rests just under the nose. Oxygen is used if the levels of oxygen in your blood are low.

Transplantation

When medicines are not able to help enough, a lung transplant or a heart-and-lung transplant may be considered.