

# Pulmonary Hypertension

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**Abstract:** As I got diagnosed in 2010 with Pulmonary Hypertension most of the time I use to think that if I have some information regarding this disease I would have taken precautions to prevent complications, so that my severity of disease will be controlled or may be delayed. But being a medical professional I was not much aware regarding this disease which leads me to suffer a lot. This is disease is very uncommon. Although Pulmonary Hypertension was discovered in 1891, there were no known treatments for the disease until 1994 when FLOLAN was introduced. Prior to the release of that medication, the prognosis and life expectancy for a patient with Pulmonary Hypertension was around 3 years and it took an average of 2 years to accurately diagnose a patient. Flolan was the first drug to increase the life expectancy by up to 5 years for pulmonary hypertension patients. While we do not currently have a cure for the disease, several more treatments have been approved for PAH thanks to research. Over the past 20 years, we have gone from no medication to treat PAH to over 10 medications. Even more medications are being studied. We are optimistic that our patients will continue to experience longer survival and better quality of life. These data were collected for the most part in the early 2000's as part of the reveal Registry and thus do not reflect today's best care. What this data showed us in that in this group of 2,635 patients, overall one year survival was 85%, three year survival was 68%, five year survival was 57% and seven year survival was 49%.

## 1. Pulmonary Hypertension

Pulmonary hypertension is a type of high blood pressure that affects the arteries in your lungs and the right side of your heart.

In one form of pulmonary hypertension, tiny arteries in your lungs, called pulmonary arterioles, and capillaries become narrowed, blocked or destroyed. This makes it harder for blood to flow through your lungs, and raises pressure within your lungs' arteries. As the pressure builds, your heart's lower right chamber (right ventricle) must work harder to pump blood through your lungs, eventually causing your heart muscle to weaken and fail.

Some forms of pulmonary hypertension are serious conditions that become progressively worse and are sometimes fatal. Although some forms of pulmonary hypertension aren't curable, treatment can help lessen symptoms and improve your quality of life.

## 2. Symptoms

The signs and symptoms of pulmonary hypertension in its early stages might not be noticeable for months or even years. As the disease progresses, symptoms become worse.

Pulmonary hypertension symptoms include:

- 1) Shortness of breath (dyspnea), initially while exercising and eventually while at rest.
- 2) Fatigue
- 3) Dizziness or fainting spells (syncope)
- 4) Chest pressure or pain
- 5) Swelling (edema) in your ankles, legs and eventually in your abdomen (ascites)
- 6) Bluish color to your lips and skin (cyanosis)
- 7) Racing pulse or heart palpitations

## 3. Causes

Your heart has two upper chambers (atria) and two lower chambers (ventricles). Each time blood passes through your heart, the lower right chamber (right ventricle) pumps blood

to your lungs through a large blood vessel (pulmonary artery).

### Pulmonary hypertension classification:

Pulmonary hypertension is classified into five groups, depending on the cause.

#### Group 1: Pulmonary arterial hypertension

- Cause unknown, known as idiopathic pulmonary arterial hypertension
- A specific gene mutation that can cause pulmonary hypertension to develop in families, also called heritable pulmonary arterial hypertension
- Certain drugs — such as certain prescription diet drugs or illegal drugs such as methamphetamines — or certain toxins
- Heart abnormalities present at birth (congenital heart disease)
- Other conditions, such as connective tissue disorders (scleroderma, lupus, others), HIV infection or chronic liver disease (cirrhosis)

#### Group 2: Pulmonary hypertension caused by left-sided heart disease

- Left-sided valvular heart disease, such as mitral valve or aortic valve disease
- Failure of the lower left heart chamber (left ventricle)

#### Group 3: Pulmonary hypertension caused by lung disease

- Chronic obstructive pulmonary disease, such as emphysema
- Lung disease such as pulmonary fibrosis, a condition that causes scarring in the tissue between the lungs' air sacs (interstitium)
- Sleep apnea and other sleep disorders
- Long-term exposure to high altitudes in people who may be at higher risk of pulmonary hypertension

#### Group 4: Pulmonary hypertension caused by chronic blood clots

- Chronic blood clots in the lungs (pulmonary emboli)

**Group 5: Pulmonary hypertension associated with other conditions that have unclear reasons why the pulmonary hypertension occurs**

- Blood disorders
- Disorders that affect several organs in the body, such as sarcoidosis
- Metabolic disorders, such as glycogen storage disease
- Tumors pressing against pulmonary arteries

**Eisenmenger syndrome and pulmonary hypertension**

Eisenmenger syndrome, a type of congenital heart disease, causes pulmonary hypertension. It's most commonly caused by a large hole in your heart between the two lower heart chambers (ventricles), called a ventricular septal defect. This hole in your heart causes blood to circulate abnormally in your heart. Oxygen-carrying blood (red blood) mixes with oxygen-poor blood (blue blood). The blood then returns to your lungs instead of going to the rest of your body, increasing the pressure in the pulmonary arteries and causing pulmonary hypertension.

**Risk factors**

Your risk of developing pulmonary hypertension may be greater if:

- You're a young adult, as idiopathic pulmonary arterial hypertension is more common in younger adults
- overweight
- Family history of the disease
- You have one of various conditions that can increase your risk of developing pulmonary hypertension
- You use illegal drugs, such as cocaine
- taking certain appetite-suppressant medications
- You have an existing risk of developing pulmonary hypertension, such as a family history of the condition, and you live at a high altitude

**4. Complications**

Pulmonary hypertension can lead to a number of complications, including:

**Right-sided heart enlargement and heart failure (cor pulmonale):** In cor pulmonale, your heart's right ventricle becomes enlarged and has to pump harder than usual to move blood through narrowed or blocked pulmonary arteries.

**Blood clots:** Clots help stop bleeding after you've been injured. But sometimes clots form where they're not needed. A number of small clots or just a few large ones dislodge from these veins and travel to the lungs, leading to a form of pulmonary hypertension that can generally be reversible with time and treatment. Having pulmonary hypertension makes it more likely you'll develop clots in the small arteries in your lungs, which is dangerous if you already have narrowed or blocked blood vessels.

**Arrhythmia:** Irregular heartbeats (arrhythmias) from the upper or lower chambers of the heart are complications of pulmonary hypertension. These can lead to palpitations, dizziness or fainting and can be fatal.

**Bleeding:** Pulmonary hypertension can lead to bleeding into the lungs and coughing up blood (hemoptysis). This is another potentially fatal complication.

**Diagnosis**

Pulmonary hypertension is hard to diagnose early because it's not often detected in a routine physical exam. Even when the condition is more advanced, its signs and symptoms are similar to those of other heart and lung conditions.. Tests may include:

1. Echocardiogram.
2. Chest X-ray
3. Electrocardiogram (ECG).
4. Right heart catheterization.
5. Blood tests.
6. Computerized Tomography (CT) scans.
7. Magnetic resonance imaging (MRI).
8. Pulmonary function test.
9. Polysomnogram.
10. Ventilation/perfusion (V/Q) scan.
11. Open-lung biopsy.
12. Genetic tests

**5. Treatment****1. Medications**

- a) Blood vessel dilators (vasodilators). Vasodilators open narrowed blood vessels. One of the most commonly prescribed vasodilators for pulmonary hypertension is epoprostenol (Flolan, Veletri). The drawback to epoprostenol is that its effects last only a few minutes.
- b) iloprost (Ventavis), can be inhaled six to nine times a day through a nebulizer, a machine that vaporizes your medication. Because it's inhaled, it goes directly to the lungs. Side effects associated with iloprost include chest pain — often accompanied by a headache and nausea — and breathlessness.
- c) Treprostinil (Tyvaso, Remodulin, Orenitram), another form of the drug, can be given four times a day. It can be inhaled, taken as oral medication or administered by injection. It can cause side effects such as a headache, nausea and diarrhea.
- d) Endothelin receptor antagonists. These medications reverse the effect of endothelin, a substance in the walls of blood vessels that causes them to narrow. These drugs may improve your energy level and symptoms. However, these drugs shouldn't be taken if you're pregnant. Also, these drugs can damage your liver and you may need monthly liver monitoring. These medications include bosentan (Tracleer), macitentan (Opsumit), and ambrisentan (Letairis).
- e) Sildenafil and tadalafil. Sildenafil (Revatio, Viagra) and tadalafil (Cialis, Adcirca) are sometimes used to treat pulmonary hypertension. These drugs work by opening the blood vessels in the lungs to allow blood to flow through more easily. Side effects can include an upset stomach, headache and vision problems.
- f) High-dose calcium channel blockers. These drugs help relax the muscles in the walls of your blood vessels. They include medications such as amlodipine (Norvasc), diltiazem (Cardizem, Tiazac, others) and nifedipine (Procardia, others). Although calcium channel blockers

can be effective, only a small number of people with pulmonary hypertension respond to them.

- g) Soluble guanylate cyclase (SGC) stimulator. Soluble guanylate cyclase (SGC) stimulators (Adempas) interact with nitric oxide and help relax the pulmonary arteries and lower the pressure within the arteries. These medications should not be taken if you're pregnant. They can sometimes cause dizziness or nausea.
- h) Anticoagulants. warfarin (Coumadin, Jantoven) to help prevent the formation of blood clots within the small pulmonary arteries. Because anticoagulants prevent normal blood coagulation, they increase your risk of bleeding complications.
- i) Digoxin. Digoxin (Lanoxin) can help the heart beat stronger and pump more blood. It can help control the heart rate if you experience arrhythmias.
- j) Diuretics. Commonly known as water pills, these medications help eliminate excess fluid from your body. This reduces the amount of work your heart has to do. They may also be used to limit fluid buildup in your lungs.
- k) Oxygen. Your doctor might suggest that you sometimes breathe pure oxygen, a treatment known as oxygen therapy, to help treat pulmonary hypertension, especially if you live at a high altitude or have sleep apnea. Some people who have pulmonary hypertension eventually require continuous oxygen therapy.

## 2. Surgeries

- a) Atrial septostomy
- b) Transplantation. In some cases, a lung or heart-lung transplant might be an option, especially for younger people who have idiopathic pulmonary arterial hypertension.

Major risks of any type of transplantation include rejection of the transplanted organ and serious infection, and you must take immunosuppressant drugs for life to help reduce the chance of rejection.