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Journal

# *Pulmonary Hypertension*

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# UNDERSTANDING PULMONARY HYPERTENSION

Pulmonary hypertension (PH) is a complex disease that can be summed up as high blood pressure in the lungs. As many as 100,000 Americans are believed to have pulmonary hypertension, and - thousands of these people are living with the condition and don't know it.

In people with pulmonary hypertension, the small blood vessels of the lungs, known as the pulmonary arteries, become progressively narrowed over time. This makes it more difficult for blood to flow through the vessels. The result is a build-up of blood and pressure in the lungs and the right side of the heart.

As the build-up grows, the right side of the heart works harder to push the blood through the narrowed arteries, and the right ventricle gains muscle cells and starts to enlarge, a process known as remodeling. Eventually, the right ventricle enlarges and weakens and is no longer able to push enough blood through the lungs to pick up sufficient oxygen to deliver to the rest of the body. This can cause shortness of breath, fatigue and other debilitating symptoms.

Until about a decade ago, doctors had few options to treat patients. But the outlook for people with pulmonary hypertension is changing. Doctors are becoming better at recognizing the disease earlier, and new treatments are helping patients live longer with fewer symptoms.

## Risk Factors

Pulmonary hypertension can affect men or women of any ethnicity. Younger women appear to have the greatest risk, although researchers aren't sure why. Certain medical conditions may make a person more likely to develop pulmonary hypertension.

*These conditions include:*

- Blood Clots
- Liver Disease
- Connective tissue disease
- Scleroderma

When pulmonary hypertension is not related to an underlying condition, it is described as idiopathic or primary pulmonary hypertension. This means the cause is unknown, and it can affect anyone. Doctors have not identified specific risk factors for primary pulmonary hypertension.

## Symptoms

Pulmonary hypertension may have no obvious symptoms at first. Shortness of breath is usually the first sign of illness, and it can come on suddenly. As the body receives less and less oxygen, patients may experience fatigue, dizziness and other serious symptoms.

*The most common symptoms of pulmonary hypertension include:*

- Shortness of breath
- Fatigue
- Dizziness
- Fainting spells
- Swelling in the ankles or legs
- Chest pain
- Bluish tint to the lips and skin

Many patients with pulmonary hypertension often look healthy, so initial symptoms may be dismissed as nothing serious. By the time the more obvious symptoms develop, the condition is usually in a more advanced stage that may impact response to treatment. That's why it's critical that patients tell

# DIAGNOSIS & MANAGEMENT

their doctors about any early warning signs.

## Diagnosis

In the past, many patients with pulmonary hypertension never got an accurate diagnosis, or if they did, the condition was already in an advanced stage. The good news is that physicians are more informed about PH, the symptoms that may indicate PH and what other diseases portend a greater risk of developing PH.. Greater awareness and improved diagnostic tools are key to helping doctors identify patients sooner. However, there is no single, quick test to uncover pulmonary hypertension.

*Diagnosis usually requires many different tests, including:*

- Blood tests
- X-rays
- Lung scans
- Cardiac catheterization

Cardiac catheterization allows doctors to measure blood pressure in the heart and pulmonary arteries. It is considered the surest way to confirm a diagnosis of pulmonary hypertension.

Because pulmonary hypertension can be difficult to diagnosis, some patients may go months without learning the nature of their condition. Others may be told their symptoms are only in their head. But patients should never dismiss symptoms like shortness of breath. Those who don't get a diagnosis right away should keep searching until they find an answer. The sooner a patient receives an accurate diagnosis, the sooner treatment can begin.

## Treatment

Treatments for pulmonary hypertension have come a long way in recent years, giving patients hope for longer, healthier lives.

*While there is currently no cure, therapies aim to achieve three important goals:*

1. Prolong life
2. Reduce symptoms
3. Improve quality of life, including reducing hospitalizations

Most patients will receive some combination of background therapies including warfarin ( a drug that reduces the likelihood blood will clot), oxygen and/or diuretics ( drugs that help remove fluid from the body).

Most patients will also receive one or more of the following advanced therapies for PH which can be given through many different methods including by pill, inhalation, skin injection, or directly into the vein. These more advanced therapies fall into one of three classes: endothelin receptor antagonists, phosphodiesterase-5 inhibitors or prostacyclin agents.

Throughout treatment, patients can expect to undergo regular testing to evaluate their lung function. These exams may include exercise tests and many others.

## Pulmonary Hypertension & Pregnancy

Pregnancy can be life-threatening for women with pulmonary hypertension. For this reason, female patients are advised to avoid becoming pregnant. You should discuss this with your health care provider to fully understand the potential risks if this is a concern.

# FREQUENTLY ASKED QUESTIONS

## **Q** What is pulmonary hypertension?

**A** Pulmonary hypertension can be summed up as high blood pressure in the lungs. Over time, the condition can weaken the heart and lead to a shortage of oxygen in the bloodstream.

## **Q** Who gets pulmonary hypertension?

**A** Pulmonary hypertension can affect men or women of any ethnicity. Younger women appear to have a greater risk.

## **Q** What causes pulmonary hypertension?

**A** In some cases, pulmonary hypertension is associated with an underlying condition, such as scleroderma, blood clots or liver disease. In many cases a cause is unknown.

## **Q** What are the warning signs?

**A** Pulmonary hypertension often has no obvious early warning signs. As the disease progresses, common symptoms include shortness of breath, dizziness, fatigue, fainting and chest pain.

## **Q** How is pulmonary hypertension diagnosed?

**A** Pulmonary hypertension can be difficult to diagnose, particularly if a doctor does not suspect pulmonary hypertension. Among the tests that should be included, are lung scans, x-rays, blood tests and cardiac catheterization.

## **Q** What is cardiac catheterization?

**A** Cardiac catheterization allows doctors to measure blood pressure in the heart and pulmonary arteries. The doctor advances a tube into the vein and positions it in the heart and lungs. It is considered the standard for diagnosing pulmonary hypertension.

## **Q** How is pulmonary hypertension treated?

**A** Treatment for pulmonary hypertension usually involves a combination of medications and, in many cases, oxygen therapy.

## **Q** What is oxygen therapy?

**A** Oxygen therapy delivers supplemental oxygen to the body. Portable oxygen units are making it possible for oxygen-dependent patients to maintain activities outside.

# DATA, RESOURCES, HOTLINES, PUBLICATIONS and MEDIA CONTACTS

## **Pulmonary Hypertension Association**

801 Roeder Rd., Suite 400  
Silver Spring, MD 20910  
(800) 748-7274  
[www.phassociation.org](http://www.phassociation.org)

## **American College of Chest Physicians**

3300 Dundee Road  
Northbrook, IL 60062  
(800) 343-2227  
[www.chestnet.org](http://www.chestnet.org)

## **American Heart Association National Center**

7272 Greenville Avenue  
Dallas, TX 75231  
(800) 242-8721  
[www.americanheart.org](http://www.americanheart.org)

## **Medscape**

76 Ninth Avenue, Suite 719  
New York, NY 10011  
(888) 506-6098  
[www.medscape.com](http://www.medscape.com)

## **American Lung Association**

61 Broadway, 6th Floor  
NY, NY 10006  
(800) LUNGUSA / (800) 586-4872  
Lung HelpLine: (800) 548-8252  
[www.lungusa.org](http://www.lungusa.org)

## **National Heart, Lung, and Blood Institute**

**NHLBI Health Information Center**  
P.O. Box 30105  
Bethesda, MD 20824-0105  
(301) 592-8573  
[www.nhlbi.nih.gov](http://www.nhlbi.nih.gov)

## **American College of Rheumatology**

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For a copy of a 30 minute documentary  
style program "Breast Cancer" call 1-800-380-6500  
or visit [www.healthybodyhealthymind.com](http://www.healthybodyhealthymind.com)

