Pulmonary Hypertension

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Many people affected by pulmonary hypertension are able to lead productive and satisfying lives, in part because of advances in medical care and treatments. It is important that those affected by this illness keep themselves informed about the problem and understand the available treatments to improve their health and quality of life. They need to know that, without treatment, pulmonary hypertension can be a life-threatening illness (Figure).

What Is Pulmonary Hypertension?

Pulmonary hypertension is an abnormal elevation of the pressure in the blood vessels of the lungs. In fact, it could be called the "high blood pressure" of the lungs. In normal lungs, the pressure in the blood vessels is about one-quarter of the pressure in the arteries of the body and can temporarily adapt to increased pressures that occur during exercise. In pulmonary hypertension, the small arteries in the lungs are too narrow, so the pressure rises in these vessels. As a result, the right side of the heart, which pumps blood into the lungs, has to pump against a higher resistance to blood flow. This makes it more difficult to pump the blood through the lungs, particularly when increased flow is needed, as when a patient exercises.

What Causes Pulmonary Hypertension?

Many things can obstruct the passage of blood through the vessels in the lungs and lead to pulmonary hypertension. Some of the more common causes are:

- congenital heart defects,
- connective tissue disease (for example, scleroderma),
- medication (for example, the diet pill fen-phen),
- HIV infection,
- blood clots,
- liver disease, and
- primary pulmonary hypertension (no associated cause).

Because not everyone who experiences one of the above also develops pulmonary hypertension, in most cases there is probably also a genetic vulnerability to the disease.

Is All Pulmonary Hypertension the Same?

There are different forms of pulmonary hypertension. The term primary pulmonary hypertension is often used when the underlying cause or trigger has not yet been found. Secondary pulmonary hypertension includes those forms with an associated underlying cause or trigger that has been identified.

How Is Pulmonary Hypertension Diagnosed?

Symptoms are often the first clue to pulmonary hypertension. Unfortunately, when symptoms appear, the disease has already progressed quite far. When the disease is present, several tests can be done to confirm the diagnosis and determine the type of pulmonary hypertension that is present.

SYMPTOMS CAUSED BY PULMONARY HYPERTENSION*

- Unusual fatigue
- Shortness of breath
- Chest pain
- Loss of consciousness (or near-fainting episodes)
- Ankle swelling

*These symptoms are common to many medical problems and may not indicate pulmonary hypertension. A detailed medical examination should be performed by your physician to determine the cause.

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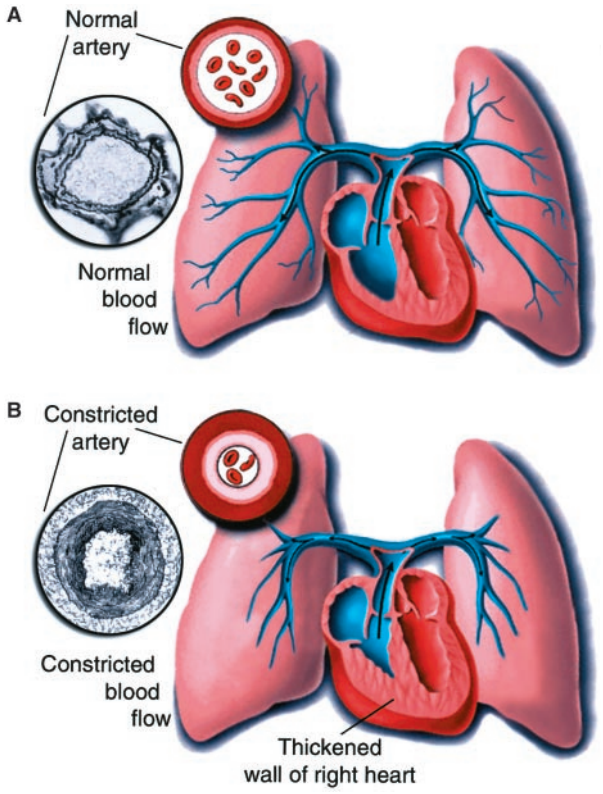

A, Diagram of the heart and lungs. This illustration shows a normal heart and lung vessels. B, Diagram of an abnormal heart and lungs as would occur in pulmonary hypertension. Note the loss of vessels in the abnormal lungs and the thickening of the right heart to counteract the rise in pressure in the lung vessels.

One or more of the following tests may be done if pulmonary hypertension is suspected:

- A *chest x-ray* may show heart enlargement and abnormal lung vessels.
- *Autoantibody blood tests* may be done to look for autoimmune diseases like lupus and scleroderma.
- *Liver function tests* may be done to look for cirrhosis or other forms of liver disease.
- *Echocardiograms* use sound waves to create a sonar-like picture of the heart and measure the heart size, function, and blood flow, and can indirectly estimate the pressure in the lung vessels.
- An *ECG* may be done to record the electrical activity of the heart and show changes in heart rhythm and wall thickness.
- *Heart catheterization*, involving long, thin tubes called catheters inserted into the heart and lung vessels, may be performed to measure pressures and the flow of blood. Sometimes, an x-ray dye may be injected into the lung vessels to look at the type and extent of obstruction.
- *Pulmonary function tests* may be done to look for other lung conditions.
- *Ventilation-perfusion scans* may be done with the use of radioactive tracers to look for certain causes of pulmonary hypertension like blood clots.
- A *CAT scan of the chest* may be done to look for abnormal lung vessels, blood clots, and lung disease outside the blood vessels.
- *HIV tests* may be done to look for HIV infection.

How Is Pulmonary Hypertension Treated?

Pulmonary hypertension can be treated with medications to lower the pressure in the lung vessels and to make the heart work more efficiently.

- *Prostacyclin* dilates the blood vessels in the lungs. It is also thought to have long-term beneficial effects on the structure of the blood vessels. Prostacyclin is usually given in its strongest form—by continuous intravenous pump—but similar forms (analogues) have been given by injection, by inhalation, or orally as a pill. Not all of these forms are available in the United States because they are pending approval by the Food and Drug Administration.
- *Calcium channel blockers* (such as nifedipine) can greatly improve the symptoms and survival of perhaps 20% of patients (those who respond well to vasodilators). To determine who will respond to this type of medication, a catheterization test is required.
- *Bosentan* blocks one of the proteins (endothelin) that constricts blood vessels in pulmonary hypertension. It has recently been approved by the Food and Drug Administration and is available as a pill taken orally.
- *Anticoagulants* "thin" the blood to prevent clot formation in the blood vessels.
- *Digoxin* can be used to make the heart pump more efficiently.
- *Diuretics* can help the kidney eliminate extra fluid.
- A *thromboendarterectomy* can remove big clots in the lungs of select patients with a form of the disease caused by clots in the large vessels of the lungs (chronic thromboembolic pulmonary hypertension). Thromboendarterectomy effectively cures such patients of pulmonary hypertension.
- *Transplantation* of lung(s) or of a heart and lung(s).
- *Supplemental oxygen* can help some patients feel better.

What Can I Expect?

Although pulmonary hypertension is a chronic disease, the outlook for patients with pulmonary hypertension

has improved dramatically in the past decade and continues to improve. New therapies and approaches are undergoing research and testing. The first step is for a patient to gather as much information as possible so he or she can make informed decisions. There are many centers of excellence that offer specialized care for patients with pulmonary hypertension. Given the life-threatening nature of this illness, affected patients should seriously consider visiting one of these centers for an evaluation.

Once the evaluation is completed, patients are likely to be offered therapy tailored to their particular condition. Patient support groups exist for pulmonary hypertension, and they offer an invaluable resource for information, support, and exchange of thoughts on how best to live with this condition.

What Can I Do to Take Care of Myself?

- Learn about pulmonary hypertension and its causes. Speak to your healthcare provider. Consult Internet web sites listed on pulmonary hypertension.
- You are not alone. Be open to help and join a patient support group.

Continued

- You may exercise, but do not overexert yourself. Any worrisome symptoms, including dizziness and chest pain, should prompt you to stop and rest. If these symptoms persist despite resting for 15 to 20 minutes, you should contact your physician promptly and get emergency care.
- Be careful to avoid dehydration and excessive heat.
- Learn about your medications, their side effects, and their interactions. Be careful to take your medication as prescribed and always discuss any changes with your healthcare provider. For some of the medications used for pulmonary hypertension, stopping medication, even for a short while, can be life threatening.
- Develop a plan with your healthcare provider. Make sure you know when to call with problems and what to do in case of an emergency.

Further Information

The Pulmonary Hypertension Association (PHA) is a nonprofit group started by patients that has grown to include healthcare professionals. PHA publishes a newsletter, a medical journal, and a patient's survival guide. It holds biannual conferences, sponsors support groups around the country, raises money for research, and raises awareness. PHA's web site can be accessed at http://www.phassociation.org. You can also call their help line at (800) 748-7274.

There are also several other excellent Internet web sites for patients that provide information on the disease and updates on research developments. Several of these sites have links to patient support groups, which have proved to be outstanding sources of social and emotional support for patients. The sites include:

- PHCentral. http://www.phcentral.org
- http://www.pha-uk.com
- The National Heart, Lung and Blood Institute. *Primary Pulmonary Hypertension.* NIH Publication No. 96-3291. http://www.nhlbi.nih.gov/health/public/lung/other/pph.htm
- http://www.phneighborhood.com
- PPH Cure Foundation. http://www.pphcure.org

Pulmonary hypertension is a chronic disease that one needs to learn how to manage with the right treatment and lifestyle. The lifestyle changes that need to be made include attention to diet (including salt and water intake), moderation in levels of effort and exercise with an awareness of symptoms that are worrisome (these include loss of consciousness or near-fainting and chest pain), and meticulous attention to medication doses and administration.