Pulmonary Hypertension (PH)

What is Pulmonary Hypertension (PH)?

This is high blood pressure inside the blood vessels of the lungs. For different reasons, the walls of the blood vessels that supply the lungs may constrict or narrow, causing the pressure inside the blood vessels to increase. This can decrease the amount of blood that can move through the blood vessels of the lung to pick up oxygen that the body needs. The heart may need to work harder and harder to try and push more blood through the blood vessels to pick up oxygen with each breath. In time, the heart may not be able to keep up the increased work load, and you may become tired, dizzy, and short of breath. If the blood pressure in the blood vessels of the lung is not decreased, the heart may become damaged and perhaps even stop working.

Causes of Pulmonary Hypertension

Sometimes the cause is unknown, but there are some known causes:

- **Pulmonary arterial hypertension** means high blood pressure in the arteries of the lungs. The right side of your heart pumps the blood into the arteries of your lungs. With this type, the right side of your heart has to work too hard to force blood through the lung vessels. The right side of the heart is not made to work so hard.

- **Pulmonary venous hypertension** results in increased pressure in the veins of the lungs. A disease of the left side of the heart may cause increased pressure. Mitral valve disease or a damaged left ventricle may cause increased pressure in the veins, which leads to fluid build up in the lungs and high blood pressure.

- **Pulmonary hypertension from hypoxemia** (low oxygen levels in the body) causes the blood vessels in the lung to become blocked. This affects the amount blood and oxygen flowing through your body. Emphysema, sleep apnea, or being exposed to high altitudes for a long time are a few causes.

- **Pulmonary hypertension due to chronic thrombotic and/or embolic disease** is caused by blood clots or a blockage in the lungs. When blood clots in the lung form, blood flow is blocked and pressure builds up behind the blockage.

- **Other causes** may be due to an illness where there is a direct affect on the blood vessels in your lung such as in pulmonary capillary disease or an inflammatory disease like sarcoidosis.
Symptoms

Because the symptoms are the same as those from many other diseases, it is often found by looking for and ruling out those other diseases.

- Trouble breathing
- Chest pain
- Feeling dizzy
- Fainting
- Chronic fatigue
- Swollen ankles and legs
- Decreased ability to walk or exercise
- Dry cough
- Raynaud’s syndrome

If pressures inside the lungs increase, then symptoms get worse. Increased fatigue, increased ankle swelling, and fluid around the lungs and the abdomen are present with severe pulmonary hypertension.

Treatment

This is a long-term disease. Although it cannot be cured, it can be managed. Discuss the treatment plans with your doctor:

- **Manage the cause.** If you have a disease that caused the hypertension, that disease must be treated also.
- **Take medicines.** Today, there are many medicines to help manage most people with pulmonary hypertension. You must know your medicines and how they should be taken.
- **Use oxygen.** Low levels of oxygen in your blood will make the problem worse and increase the heart’s workload. Some patient’s need to wear oxygen in order to increase these levels.
- **Surgery** can help correct congenital heart problems (birth defects), improve blood flow when old, scarred blood clots are removed, and repair or replace damaged heart valves. Lung or heart/lung transplant may be an option if other treatments have failed.

Medicines

Not every person with Pulmonary Hypertension needs each of the medicines listed below. Since each person is different, their prescribed medicine plan may be different.

**Blood thinners**

Types: Coumadin® (warfarin)

Function: This medicine makes the blood less likely to clot or form a blockage.

**Calcium channel blockers**

Types: Nifedipine (Procardia®/Adalat®), Amlodipine Besylate (Norvasc®), Diltiazem Hydrochloride (Cardizem®, Cartia XT®)

Function: These work to decrease the pressure inside the lungs of some people with Pulmonary Arterial Hypertension.
Diuretics ("water pills")
Types: Lasix (Furosemide®), Bumex (Bumetanide®), Demadex (Torsemide®), Aldactone (Spironolactone®) or metolazone
Function: These work to get rid of extra body fluid.

Endothelin Receptor Antagonist
Types: Bosentan (Tracleer®), Ambrisentan (Letairis®)
Function: This works to reduce the high blood pressure in the blood vessels of the lungs and may allow the heart to work easier.

Phosphodiesterase Type 5 - Inhibitors
Types: Sildenafil (Viagra® or Revatio®), Tadalafil (Adcirca®)
Function: This works to reduce the high blood pressure in the blood vessels of the lungs and may allow the heart to work easier.

Prostacyclin Analogue
Types: Epoprostenol (Flolan®, Veletri®) Treprostinil (Remodulin®, Tyvaso®), Ventavis (Iloprost®)
Function: These drugs directly dilate lung blood vessels and reduce blood pressure in the lungs. Flolan® is given by a continuous intravenous (IV) infusion. Remodulin® is given through a constant subcutaneous (under the skin) or IV infusion. Iloprost® is inhaled (like a breathing treatment) every 2-3 hours and Tyvaso® is inhaled 4 times daily.

Non-Drug Therapies

Low Sodium Diet
Because ingestion of salt (sodium chloride) can increase water retention ("water weight gain"), and water retention can increase blood pressure, it is best if people with high blood pressure limit their total daily intake of salt (sodium chloride) to no more than 2,500mg. This may seem like a lot of salt (sodium chloride) to be able to use from a salt shaker and eat in a day, but salt is contained both inside the salt shaker, as well as hidden in foods we buy in the grocery store. So, to limit the total daily intake of salt (sodium chloride) to no more than 2,500mg, a person will need to add up the total amount of salt (sodium chloride) from all sources (salt shaker and within foods). The amount of salt (sodium chloride) per serving of food is often listed on the labels of foods that are purchased in a grocery store. Ask a doctor, dietician, pharmacist or nurse for more information about this if you have questions.

Supplemental Oxygen
If the doctor decides that adding supplemental oxygen to your daily routine would be helpful for you, using the way it is prescribed may help to provide enough oxygen for the best function of your body.

There is new research being done all the time and new treatments are being found.

Your health care team may have given you this information as part of your care. If so, please use it and call if you have any questions. If this information was not given to you as part of your care, please check with your doctor. This is not medical advice. This is not to be used for diagnosis or treatment of any medical condition. Because each person's health needs are different, you should talk with your doctor or others on your health care team when using this information. If you have an emergency, please call 911. Copyright © 5/2016. University of Wisconsin Hospitals and Clinics Authority. All rights reserved. Produced by the Department of Nursing HF#6719.