Pulmonary Hypertension & Sleep Apnea
PULMONARY HYPERTENSION, or PH, is complex and often misunderstood. PH means high blood pressure in the lungs and is different from regular hypertension. In PH, the blood vessels in the lungs become damaged and/or narrowed and the heart has to work harder to pump blood through them.

PH can be caused by many different problems such as heart failure, diseases that damage the lungs or multiple hardened blood clots. It is important to understand that not all PH is the same.

PH affects people of all ages and ethnic backgrounds. The most common symptoms are shortness of breath with physical activity, fatigue, lightheadedness and sometimes fainting. Because these symptoms can be caused by any number of other medical problems, diagnosis is often delayed. Identifying a case of PH can be difficult and may require a specialist. Once the type of PH is diagnosed, treatment can begin immediately.

One form of PH is called pulmonary arterial hypertension (PAH). PAH is a complex, progressive type of PH where the high blood pressure in the lungs occurs because tiny blood vessels that carry blood through the lungs (pulmonary arteries) are narrowed, thickened and stiff. As PAH advances, the heart may lose its ability to pump enough blood through the lungs to meet the needs of the body.

There are several types of PAH. Idiopathic PAH (IPAH) is PAH without any other obvious medical problem leading to high blood pressures in the lung. Heritable PAH (HPAH) comes from abnormal genes that cause PAH. Heritable PAH may be passed on to some members of your family. PAH can be associated with other medical conditions such as connective tissue diseases (scleroderma and lupus...
for example), chronic liver disease, congenital heart disease, or HIV infection. Finally PAH can be associated with past or present drug use, such as methamphetamines or certain diet pills. It is not known exactly how these medical problems or drugs cause PAH.

PAH is a serious condition, and without treatment, symptoms will become worse, leading to heart failure and even death. Proper diagnosis and treatment from a doctor who understands PAH is essential. Every patient is different. The choice of treatment is based on how sick a patient is and the risks and benefits of any particular therapy. Regardless of risk, is it important that patients and their health care providers engage in frequent follow ups with ongoing discussions about the management of their condition. Current guidelines suggest that changes in therapy should be considered for patients not reaching their treatment goals.

While no cure has yet been found for PAH, increased research has resulted in treatments that allow patients to live longer, fuller lives with far less interference from the disease. Even more promising research is being conducted every day that is not only advancing our understanding of the PAH disease, but also potentially identifying new treatment options for patients in the future.
Pulmonary Hypertension in Sleep Apnea

Sleep apnea is a potentially serious sleep disorder in which breathing repeatedly stops and starts while one is asleep. There are two types of sleep apnea: obstructive sleep apnea, in which the throat muscles relax during sleep causing a blockage in the airway, and central sleep apnea, where the brain does not send proper signals to the muscles that control breathing. Obstructive sleep apnea is more common of the two.

The signs and symptoms of both types are the same, so determining which type you may have could be difficult. The most common signs and symptoms of obstructive and central sleep apneas are loud snoring, brief episodes of when you stop breathing, abruptly waking up and waking up with a dry mouth or sore throat, morning headache, excessive daytime sleepiness, attention problems and feeling tired.

Pulmonary hypertension usually worsens during sleep; therefore, mild-to-moderate PH is a common complication of sleep apnea. Low levels of oxygen during sleep and associated stress is thought to be the cause of an increase in pulmonary artery pressure.

Patients with sleep apnea will often experience fatigue and possible limitations while exercising. However, unexplained shortness of breath or increasing fatigue; swelling of the ankles, legs, abdomen or arms; chest discomfort or pain; and light-headedness and fainting are all symptoms your doctor should know about since these can also be signs of PH associated with sleep apnea.

What can happen to patients with these problems, and why should I be concerned?

As a sleep apnea patient, it is important that you are aware of your condition and that you stay in touch with your doctor concerning your symptoms. While optimal treatment of sleep apnea can lead to improvement and resolution of pulmonary hypertension, ongoing or worsening symptoms despite treatment of sleep apnea requires further evaluation.

What tests can be done to see if I have PH?

As part of the tests used to determine what is causing PH in a patient with sleep apnea, or to determine if you have PH, your doctor may order pulmonary function tests, a chest x-ray or chest CT scan, echocardiogram and possibly a right heart catheterization.

The echocardiogram is an image of the heart as it is working. This image is created through a non-invasive ultrasound.
A pulmonary function test measures the capacity of the lungs and its effectiveness of moving air in and out of the body. This test should include a diffusion capacity for carbon monoxide (DLCO), measuring the ability of oxygen to move from the air in the alveoli to the blood vessels in the lungs.

Another test, known as an NT-proBNP (N-terminal pro b-type natriuretic peptide) is a blood test that monitors stress on the heart, checking for heart failure symptoms.

A ventilation-perfusion (“V/Q”) scan can also be used to rule out old, organized clots in the lungs.

Right heart catheterization remains the most accurate PH diagnostic test measuring the pressures and flow directly in the heart and providing other information about the heart’s condition as well. It is an invasive procedure usually done on an outpatient basis in the hospital.

Once a doctor and patient have established an effective medical approach and treatment plan, many patients show significant improvement.

There is little information about the effectiveness of PH-targeted therapies in PH associated with sleep apnea. Treatment with current PH therapies based on echocardiogram findings is strongly discouraged. It is important to first treat sleep apnea to make sure that you are getting enough oxygen at night. If symptoms persistent, and the pulmonary artery pressures are still high, there may be a benefit from taking a PH medication. Please visit a PH care center for a detailed assessment and appropriate next steps.

If I have PH in addition to sleep apnea, what can doctors do to help me? All patients will benefit from following a few simple measures including:

- Paying special care and attention to fluid balance with a low sodium diet and water pills.
- Using supplemental oxygen as directed.
The Pulmonary Hypertension Association (PHA) was founded by and for PH patients. The organization has led the way in bringing pulmonary hypertension into the national and international consciousness. Additionally, PHA is constantly increasing its services to the medical community through educational programming, membership sections for medical professionals and much more:

Website:
PHA’s website is a comprehensive source of information for patients, caregivers and medical professionals. Please visit us at [www.PHAssociation.org](http://www.PHAssociation.org).

Find A Doctor:
The “Find A Doctor” section of PHA’s website allows patients and referring physicians to search for PH-treating physicians by state at: [www.PHAssociation.org/Patients/FindADoctor](http://www.PHAssociation.org/Patients/FindADoctor). While PHA does not endorse any of these physicians, PHA strongly recommends that all PH patients see a PH specialist who will be able to provide them with the best care.

**Pulmonary Hypertension: A Patient’s Survival Guide:**
This extraordinary 300+ page book was written by a patient and is medically reviewed and updated annually. It presents the illness in a very human and readable way, covering a wealth of topics like the mechanics of PH, the latest treatments, patient care and lifestyle tips. PHA members receive a discount on this resource. The *Survival Guide* is available for purchase at: [www.PHAssociation.org/SurvivalGuide](http://www.PHAssociation.org/SurvivalGuide).

Support Groups:
Knowledge, support, hope and empowerment: just a few of the things a PHA support group offers PH patients. No one should face this disease alone. In many places, patients have the opportunity to meet, learn from and find common understanding with others in similar circumstances. Find a support group in your area at [www.PHAssociation.org/LocalSupportGroups](http://www.PHAssociation.org/LocalSupportGroups).
The mission of the Pulmonary Hypertension Association is to extend and improve the lives of those affected by PH. Our vision is a world without PH, empowered by hope.

PHA fulfills its mission through:

• Providing funding for research
• A quarterly medical journal *Advances in Pulmonary Hypertension*
• *PHA Online University* offering free CME credits and the latest information on pulmonary hypertension: [www.PHAOnlineUniv.org](http://www.PHAOnlineUniv.org)
• Professional membership sections:
  - PH Clinicians and Researchers (PHCR) — for physicians and doctorate-level researchers
  - *PH Professional Network* — for allied health care professionals
• Educational conferences and materials for medical professionals and patients
• PH patient support groups
• A quarterly, patient focused magazine, *Pathlight*
• Advocacy and awareness campaigns
• Toll-free Patient-to-Patient Support Line (1-800-748-7274)
• myPHA, a social network connecting the PH community: [www.myPHAssociation.org](http://www.myPHAssociation.org)

More Information on Sleep Apnea: [www.sleepapnea.org](http://www.sleepapnea.org)
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