Did you know that if you have Sickle Cell Disease you are at risk for Pulmonary Hypertension?
PULMONARY HYPERTENSION, OR PH, is a complex and often misunderstood disease. Pulmonary hypertension means high blood pressure that is located in the lungs. Pulmonary hypertension is different from regular hypertension. In regular hypertension (also known as high blood pressure), a person’s arteries throughout the body are constricted. In pulmonary hypertension, the blood vessels in the lungs become narrowed and the heart has to work harder to pump blood through them. PH can also be caused by multiple blood clots, diseases that damage the lung, or heart failure. Early symptoms are often non-specific and diagnosis is often delayed.

Pulmonary hypertension 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 problems, diagnosing a case of pulmonary hypertension can be difficult and may require a specialist. Once pulmonary hypertension is diagnosed, however, treatment can begin immediately.

One form of PH is called pulmonary arterial hypertension (PAH). In PAH, the blood vessels that carry blood to the lungs (known as the pulmonary arteries) are narrowed, thickened and stiff. Because of this, the pressure in those arteries is abnormally high. This means that the right side of the heart has to pump much harder to move blood. There are several types of PAH. In what is known as idiopathic PAH (IPAH), the cause of the PAH is never known. In what is known as familial PAH (FPAH), the disease is inherited, meaning another member or members of the patient’s family was diagnosed with pulmonary hypertension. PAH can also be associated with other medical conditions such as connective tissue diseases (scleroderma and lupus, for example), chronic liver disease, congenital heart disease,
sickle cell disease and HIV infection. Finally, PAH can be associated with past or present drug use, such as methamphetamines or certain diet pills.

Some patients who are diagnosed with PH also suffer from other specific problems as well. These other problems might include left-sided heart disease (for example, people who have had heart attacks, have hypertension or heart valve disease), chronic obstructive pulmonary disease (COPD) and emphysema, and chronic thromboembolic disease (chronic blood clots in the lungs).

PAH is a serious condition, and without treatment, symptoms can only become worse, leading to heart failure and even death. Therefore, it is essential that patients follow the advice of their doctor. PAH, like PH itself, is rare and its signs and symptoms are vague and often confused with other disorders. Proper diagnosis and therapy are essential. Increasing public awareness of PAH (and PH) will help patients seek treatment earlier, which can lead to better long-term health. While no cure has yet been found for PH, increased research has resulted in new and better medications and therapies that allow PH patients to live longer, fuller lives with far less interference from the disease. Even more promising research is being conducted every day, and advances are made every year.
I have sickle cell disease. My doctor wants to screen me for pulmonary hypertension. Why is this?

Sickle cell disease (SCD), a type of anemia (shortage of red blood cells), is characterized by abnormally shaped red blood cells. Sickle cells (named for the sickle, or oblong, slender shape of the malformed blood cells) have decreased flexibility, meaning they break easily. This results in a number of life-long complications. PH is an increasingly recognized complication of SCD. Studies show that approximately 30% of screened adult patients with SCD develop mild PH in adulthood. In 10% of patients, the PH is more severe. Recent autopsy studies also suggest that up to 75% of sickle cell patients show changes in the lung tissue at the time of death, indicating the existence of pulmonary arterial hypertension (PAH).

One possible reason sickle cell patients seem to be at risk for developing PH is that increased breakdown of red blood cells within the blood vessels causes inflammation and a decrease of nitric oxide (a substance that causes relaxation of the blood vessels). This causes constriction of the small arteries of the lungs which may lead to PH.

What are some of the symptoms of pulmonary hypertension?

Because they are somewhat general symptoms, the characteristics of PH felt by the patient often are mistakenly attributed to other problems. These symptoms include shortness of breath, feeling fatigued, being unable to tolerate increased levels of exercise, unexplained swelling of the ankles, legs, abdomen or arms, chest discomfort or pain, light-headedness and fainting. Your doctor wants to make sure you do not have symptoms that are being overlooked.

Why is it so important to screen for PH now?

Individuals who suffer from SCD are more likely to develop PH, and those who suffer from PH in addition to SCD are at a higher risk of death than that of SCD patients without PH. Therefore, your doctor wants to test your pulmonary pressure to make sure you’re not currently affected by PH. If PH is present, he or she will want to begin treatment immediately to control both your SCD and your PH. Studies from the National Heart, Lung and Blood Institute suggest that PH is the greatest risk factor for death in the aging population of patients with SCD. An echocardiogram is an important screening tool to check for the development of PH in patients with problems like SCD. The echocardiogram can also help to determine the cause of the high pulmonary pressures.
If a doctor determines that I have PH, what's the next step?

Because SCD is a complex disorder with many complications, no specific clinical guidelines have been written for patients who have both SCD and PH. Researchers do not know exactly what role PH plays in the worsening of disease.

The National Institute of Health recommends that each case of PH be evaluated by a specialist, and assigned a value, from mild to severe. The therapy used to treat various gradations of the disease will likewise increase or decrease, depending on the severity. A number of different treatments are available, and have been shown to be very effective. Your doctor will help determine the best treatment for you, and will continue to monitor your progress throughout your treatment. Specialists at a PH center can also help guide you through the process and answer your questions.
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. 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 PH care.

**Pulmonary Hypertension: A Patient’s Survival Guide:**
This extraordinary 300+ page book was written by a patient and medically reviewed. It presents the illness in a very human and readable way. It is available from PHA at minimal cost, at a discount for PHA members, and one complimentary copy is provided to members of PHA’s professional membership bodies. *The Survival Guide* is available for purchase online at [www.PHAssociation.org/SurvivalGuide](http://www.PHAssociation.org/SurvivalGuide).

**Online information about PH:**
For information on PH diagnosis, symptoms, treatments and more, visit [www.PHAssociation.org/Patients/AboutPH](http://www.PHAssociation.org/Patients/AboutPH).

**Support Groups:**
From the first support group started in 1990 around a kitchen table in Florida, PHA grew to 45 in 2001, and to over 215 in 2010. 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 find ways to prevent and cure pulmonary hypertension, and to provide hope for the pulmonary hypertension community through support, education, advocacy and awareness. PHA’s members stand as part of a community that is fighting back against this terrible illness.

**PHA fulfills its mission through:**

- Funding for research
- 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 Resource Network—for nurses and allied health professionals
- Educational conferences and materials for medical professionals and patients
- 300+ page patient’s survival guide
- PH patient support groups
- Quarterly newsletter *Pathlight*
- Advocacy and awareness campaigns
- Toll-free Patient-to-Patient Helpline (1-800-748-7274)
- PHA website with PH discussion boards, email groups and online support chats ([www.PHAssociation.org/ConnectOnline](http://www.PHAssociation.org/ConnectOnline))

**More Information on Sickle Cell Disease**

Sickle Cell Disease Association of America: [www.sicklecelldisease.org](http://www.sicklecelldisease.org)

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