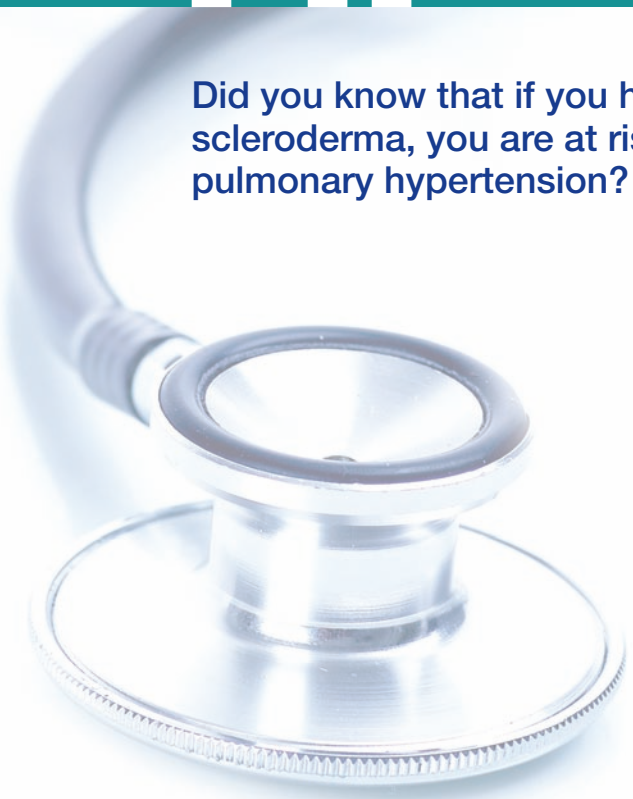
Scleroderma & PH Pulmonary Hypertension

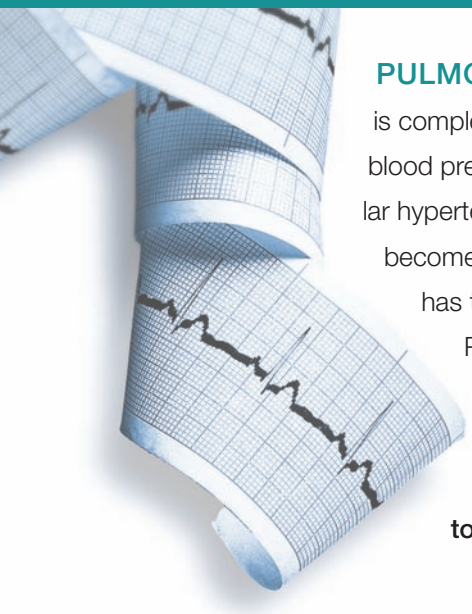
Did you know that if you have scleroderma, you are at risk for pulmonary hypertension?



PHA Pulmonary Hypertension Association
Empowered by hope

www.PHAssociation.org

About Pulmonary Hypertension



PULMONARY HYPERTENSION, OR PH, is complex and often misunderstood. PH means high blood pressure in the lungs. PH 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 lung or multiple hardened blood clots, to name a few. **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, however, 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) means that the patient develops 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 can only become worse, leading to heart failure and even death. Proper diagnosis and therapy 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 healthcare 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.

Scleroderma-Associated PH

I was just diagnosed with scleroderma, which I don't fully understand. My doctor also has asked me if I get short of breath, feel fatigued or like I'm going to faint. Why am I being asked this?

Scleroderma, which means "hard skin" in Latin, is a type of autoimmune disease that affects the connective tissues and blood vessels of the body. Connective tissue is mostly made of collagen and gives structure and support to the organs of the body. Collagen is a protein that the body uses to make scar tissue to repair itself. In scleroderma, the immune system attacks the body's organs and tissues. As a result, excess collagen is produced, and scar tissue forms.

The additional symptoms your doctor is mentioning (shortness of breath and fatigue) are those of pulmonary arterial hypertension (PAH), a disease of the small arteries of the lungs. PAH is a common complication of scleroderma. Between 8 and 12 percent of all scleroderma patients develop PAH. In patients suffering from PAH, arteries become damaged and narrowed, making it difficult for the right side of the heart to pump blood through the lungs. Your doctor, therefore, wants to make sure right away that you're not developing PAH.

Scleroderma patients tend to have exercise limitations, and it is important to report any changes in your current abilities. 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 will want to know about, since these can also be signs of pulmonary arterial hypertension.

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

It is absolutely essential that you as a scleroderma patient be aware of your condition and that you stay in touch with your doctor concerning your symptoms. While many scleroderma patients go on to lead long and enjoyable lives, PAH is a serious and complex condition that requires immediate treatment. Pulmonary complications are the leading cause of death in people with scleroderma.

In general, the medications that are used to treat regular PAH do not work as well when used on patients experiencing scleroderma-associated PAH. A possible explanation of this is that scleroderma-associated PAH occurs in older patients who might already have other forms of lung disease. It is also possible that patients with scleroderma have a more severe form of PAH in their pulmonary arterioles or that the right side of the heart is less able to handle the higher pressures caused by PAH. In all cases, the earlier a diagnosis is made, the sooner treatment can begin, and the better the outlook is for the patient.

What tests can be done?

Since patients with scleroderma are typically under a doctor's care for their scleroderma, annual screening for PAH and PH should be done. Screening should include a detailed history about the level of physical activity and if any problems are limiting those activities. The symptoms mentioned before (shortness of breath, swelling, etc.) should always be brought to a doctor's attention.

According to a panel of international experts on PH and scleroderma, physicians should run three specific tests on every patient diagnosed with scleroderma and other connective tissue diseases as a means of diagnosing PH earlier. The first test is an echocardiogram, or image of the workings of the heart. This image is created through a non-invasive ultrasound. The second test is a pulmonary function test to measure the capacity of the lung and the effectiveness of the lung in moving air in and out of the body. This test should include a diffusion capacity carbon monoxide (DLCO), measuring the ability of oxygen to move through the blood vessels in the lungs. The third test, known as an NT-Pro BNP (N-terminal pro b-type natriuretic peptide) is a blood test that monitors stress on the heart, checking for heart failure symptoms. Right-heart catheterization remains the most accurate diagnostic test and provides other information about the heart's condition as well.

If I have PAH as well as scleroderma, what can doctors do to help me?

All patients benefit from simple basic measures including attention to fluid balance and therapies that improve the efficiency of the heart. These may include supplemental oxygen or the medication digoxin, as well as anticoagulants (blood thinners). Once a doctor and patient have been able to establish an effective medical approach, many patients show significant improvement, often in conjunction with doctor-prescribed cardiopulmonary rehabilitation training (CPRT).

Through the years, a variety of medications have been shown to slow the damage and relieve the symptoms caused by PAH. These medications are very complex, and choosing the correct initial treatment requires the knowledge and expertise of a physician who specializes in PAH. Professionals at pulmonary hypertension centers can help identify pulmonary hypertension and choose the correct type of treatment.

PHA Resources

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**.

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**. 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 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 issues. PHA members receive a discount on this resource. The *Survival Guide* is available for purchase as a paperback and an e-book at: **www.PHAssociation.org/SurvivalGuide**.

Online information about PH:

For information on PH diagnosis, symptoms, treatments and more, visit **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 groups in 2001 and to more than 245 in 2013. 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**.

About the Pulmonary Hypertension Association

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, research, advocacy and awareness. PHA's members form 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)**
- Professional membership sections:
  - PH Clinicians and Researchers (PHCR)—for physicians and doctorate-level researchers
  - PH Professional Network—for nurses and allied health professionals
- Educational conferences and materials for medical professionals and patients
- A wealth of information in the *Survival Guide*
- PH patient support groups
- Quarterly newsletter *Pathlight*
- Advocacy and awareness campaigns
- Toll-free Patient-to-Patient Support Line (1-800-748-7274)
- PHA website with PH discussion boards, email groups and online support chats (**www.PHAssociation.org/ConnectOnline**)

More Information on Scleroderma

Scleroderma Foundation:
www.scleroderma.org

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