

PH&



Pulmonary Hypertension & **SCLERODERMA**

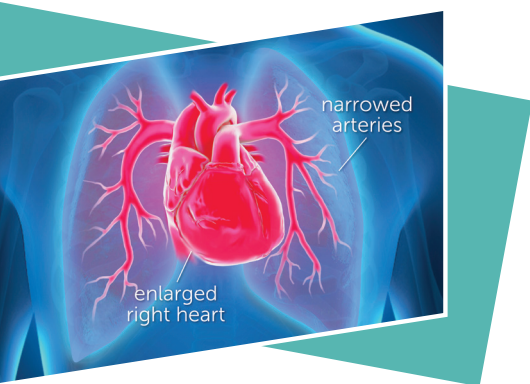


Pulmonary Hypertension Association
Empowered by hope

ABOUT PULMONARY HYPERTENSION

Pulmonary hypertension (PH) is a complex and often misunderstood disease. The term PH simply means high blood pressure in the arteries of the lungs. In regular hypertension — also known as high blood pressure or systemic hypertension, which you can check with a blood pressure cuff — the pressure in the arteries of the entire body is higher than it should be. In PH, the blood vessels specifically in the lungs are affected. They can become stiff and narrow, and the right side of the heart must work harder to pump blood through them. There are five groups of PH based on different causes.

Each form of PH is different, so it is important for newly diagnosed patients to find a PH specialist who can accurately pinpoint what is causing their PH. They then can develop a treatment plan for that specific type as soon as possible after a confirmed diagnosis. Every individual with PH is different, and new research with the potential to improve the outlook for people living with this disease is conducted on a continual basis. Once in the care of a PH-treating health care team and on appropriate therapy, individuals with PH can live for many years.



PH, or high blood pressure in the lungs, frequently results from a narrowing of the small blood vessels in the lungs, which leads to a larger right side of the heart.

*Normal mean pulmonary artery pressure is between **8-20 mmHg** at rest. Pulmonary hypertension is defined as a resting mean pulmonary artery pressure at or above **25 mmHg**.*



SCLERODERMA-ASSOCIATED PH

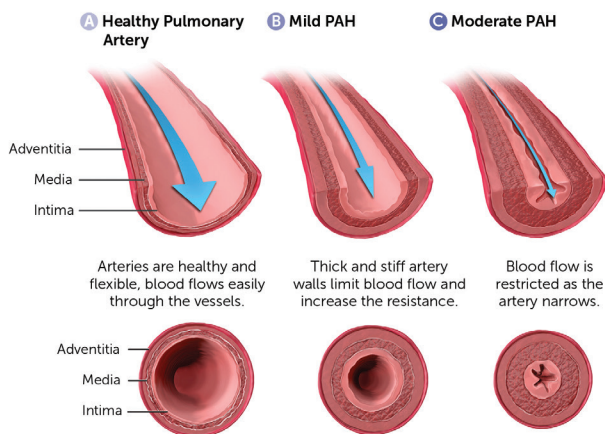
Derived from the Greek words sklerosis, meaning hardness, and derma, meaning skin, scleroderma literally translates to hard skin. Scleroderma, referred to as systemic sclerosis (SSc) if it involves more than just the skin, is a type of autoimmune disease. That means that the body's immune system attacks its own organs and tissues. With this condition, the connective tissues and blood vessels in the body are affected.

Connective tissue is mostly made of collagen — a protein that the body uses to make scar tissue to repair itself after injury. It gives structure and support to organs, like a scaffolding for the body. In people with scleroderma, the body produces extra collagen and more scar tissue forms in the skin. In SSc, the lungs, kidneys, heart, blood vessels, joints, muscles and gastrointestinal tract can be affected. Most patients with SSc suffer from Raynaud's phenomenon, in which small blood vessels in the fingers and toes spasm due to the cold, which blocks blood flow. Some patients can develop painful ulcers on their fingers or toes due to poor blood flow.

There are two major types of SSc: limited and diffuse. Limited SSc, previously called CREST syndrome, can be associated with skin hardening in the arms below the elbows and legs below the knees, as well as the face, but some patients just have puffy fingers. Diffuse SSc patients have skin hardening that can affect the entire body. People with diffuse SSc are more likely to have problems with their internal organs, including the digestive tract, kidneys, heart and lungs early in the disease, while their skin is tightening quickly. This internal organ involvement usually differs from patient to patient.

If an SSc patient is experiencing shortness of breath and tiredness, they might have lung fibrosis or World Health Organization Group 1 PH (pulmonary arterial hypertension, or PAH). PAH is a common complication of SSc; between 8 and 12 percent of all SSc patients develop PAH. In patients with PAH, the arteries become stiff and

narrow, making it difficult for the right side of the heart to pump blood through the lungs. Because this complication is so common in SSc, screening for PAH is recommended in all patients, even if they do not have any symptoms. The Food and Drug Administration (FDA) has approved multiple medications for PAH, and it is usually a treatable disease if diagnosed early enough. Therefore, it is vital that patients get examined to make sure they are not developing PAH.



SSc patients also usually have problems exercising because their joints are inflamed and stiff, resulting in pain. Any changes in a person's current physical abilities should be reported to a doctor. Unexplained shortness of breath or increased fatigue, swelling of the ankles, legs, arms or stomach area, chest discomfort or pain, and light-headedness and fainting are all symptoms that a doctor should know about because these could be signs of PAH.

It is essential that SSc patients stay in frequent contact with their doctor. While many SSc patients go on to lead long lives, PAH is a serious and complex condition that requires immediate medical treatment and care. Pulmonary complications, including PAH and lung fibrosis, are the leading cause of death in people with SSc.

DIAGNOSING SYSTEMIC SCLEROSIS-ASSOCIATED PH

Careful screening for PAH on a regular basis will result in an earlier diagnosis. The sooner treatments can begin, the better the outlook is for the patient. Since people with SSc are usually already under a rheumatologist's care, they should be getting examined for PAH once a year. A study called the DETECT Study found that a non-invasive two-step screening process using screening test results to assess risk significantly reduced missed diagnoses of PAH in SSc patients compared to screening based on symptoms and abnormal echocardiography findings.¹

Even when not being screened for PAH, the symptoms mentioned earlier (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 annually on every patient:¹

1. An **echocardiogram**: An ultrasound of the heart that provides a rough estimate of the pressure in the lungs and assesses the function of the right ventricle.
2. **Pulmonary function tests (PFTs)**: Non-invasive tests that measure how much air the lungs can hold, how much and how quickly someone can blow air and how oxygen and carbon dioxide are exchanged between the air entering the lungs and the blood circulating through the lungs (diffusion capacity of carbon monoxide, or D_LCO).
3. **NT-proBNP (N-terminal pro b-type natriuretic peptide) or BNP (brain natriuretic peptide)**: A blood test that checks for stress on the heart and, when elevated, is a sign of heart failure.

¹ Coghlan JG, et al. Ann Rheum Dis. 2014;73(7):1340-9.

Right heart catheterization remains the most accurate PH diagnostic test as it directly measures the pressures and flow in the heart and lungs, as well as provides other information about the heart's condition. It is an invasive procedure usually done on an outpatient basis in the hospital.

The goal is to diagnose PAH as early as possible before any symptoms occur. However, because of other medical problems, it may be hard for a doctor to determine the cause of symptoms. For example, if a patient has shortness of breath while they are physically active, it could either be because of an underlying lung fibrosis or PAH. Also, many people who cut back on their day-to-day activities to cope with new and increasing symptoms may forget to tell their doctors. Regular screening is the only reliable way to diagnose PAH early.

TREATMENT OPTIONS FOR SYSTEMIC SCLEROSIS-ASSOCIATED PH

All patients benefit from following simple, basic measures, including paying attention to fluid balance and use of therapies that improve heart function. These may include supplemental oxygen, diuretics (water pills) and supervised cardiopulmonary rehabilitation programs.

A variety of medications have been shown to slow down the damage and relieve the symptoms caused by PAH. However, these medications are complex and choosing the right initial treatment requires the knowledge and expertise of a physician who specializes in PAH. Professionals at PH centers can help identify PAH and choose the correct course of treatment.

There also are several ongoing clinical trials that are testing the performance and safety of new drugs for treatment of PAH in SSc.

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ABOUT THE PULMONARY HYPERTENSION ASSOCIATION

The Pulmonary Hypertension Association (PHA) is a community of individuals with PH, caregivers, families, medical professionals and researchers who work together to empower people battling the disease, while supporting research toward improved treatments. PHA was the first organization in the world dedicated to providing comprehensive PH patient and caregiver support, medical education, research and services that improve patients' quality of life.

*PHA's mission is to extend
and improve the lives of
those affected by PH.*

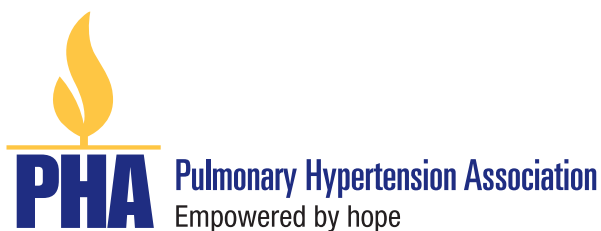
*PHA's vision is a
world without PH,
empowered by hope.*

PHA facilitates more than 200 active support groups around the country and serves as a model for more than 85 international PH organizations. In addition to in-person support, PHA offers a telephone support line, an email mentorship program, educational patient programs and a variety of print and online publications. Every two years, PHA hosts its International PH Conference and Scientific Sessions, which brings together nearly 1,500 individuals with PH, caregivers and medical professionals to share information on the latest research, treatments and lifestyle coping tips.

PHA also has provided more than \$8 million for PH research and offers continuing education programs for medical professionals through the *PHA Online University* and an in-person CME program.

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