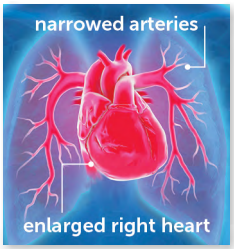
Pulmonary Hypertension and Scleroderma

PH&

PHA
Pulmonary Hypertension Association
Empowered by hope

ABOUT PULMONARY HYPERTENSION

Pulmonary hypertension is a complex and often misunderstood disease. PH sometimes is confused with systemic hypertension or "high blood pressure" that affects arteries throughout the body.



PH refers to pressure within the blood vessels of the lungs. The blood vessels can become stiff and narrow, which makes it more difficult for the right side of the heart to pump blood through them.

There are five types of PH based on different causes. Each form of PH is different, so it is important for newly diagnosed patients to find PH specialists who can pinpoint the cause of their PH. The specialist then develops a treatment plan specifically for the patient's type of PH.

Every person with PH is different, and scientists continually conduct new research to improve the outlook for people living with PH. With proper care and treatment, people with PH can live many years.

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 20 mmHg.

Scleroderma-Associated PH

Scleroderma is an autoimmune disease that affects connective tissues and blood vessels. Autoimmune disease means the body's immune system attacks its own organs and tissues. This condition causes inflammation and fibrosis (thickening) of the skin and other areas of the body. Hardening and tightening of the skin is the most common visible sign of scleroderma.

Connective tissue is mostly made of collagen, a protein that makes scar tissue to repair the body after injury. Connective tissue gives structure and support to organs, like scaffolding. With scleroderma, the body produces extra collagen, causing more scar tissue to form in the skin.

Derived from the Greek words sklerosis, meaning hardness, and derma, meaning skin, scleroderma literally translates to hard skin.

There are two major forms of scleroderma. Localized scleroderma is a hardening of the skin that does not affect the internal organs. Typically, it affects only the skin, but can also affect muscles.

Systemic scleroderma or systemic sclerosis is a more serious condition that affects the skin, lungs, kidneys, heart, blood vessels, joints, muscles and gastrointestinal tract. It also can damage blood vessels and internal organs.

The term systemic sclerosis refers to hardening that can occur in the internal organs. Most people with SSc also have Raynaud's phenomenon. Raynaud's causes decreased blood flow to fingers and toes. The decreased blood flow occurs because of spasms in fingers and toes when exposed to cold or stress. Some people can develop painful ulcers on their fingers or toes because of poor blood flow.

There are two types of SSc based on the degree of skin involvement: limited or diffuse.

Limited (sometimes called CREST Syndrome) is more common and usually progresses more slowly than than diffuse. Skin hardening typically affects lower arms and legs, as well as the face. Serious complications that affect the esophagus, other parts of the gastrointestinal track and lungs can occur.

Diffuse scleroderma (or diffuse SSc) progresses more rapidly and the hardening of the skin can occur throughout the body. People with diffuse scleroderma 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 person to person.

PH Related to SSc

If someone with SSc experiences shortness of breath and fatigue, they may have lung fibrosis or Group 1 PH (pulmonary arterial hypertension), a common complication of limited SSc. About 5% to 15% of people with SSc have PAH.

Other forms of pulmonary hypertension can develop with SSc, including PH related to left heart disease, interstitial lung disease, chronic thromboembolic disease and pulmonary venous occlusive disease. Those conditions further complicate diagnosis and management.

PH Symptoms

PAH causes the arteries to become stiff and narrow, making it difficult for the right side of the heart to pump blood through the lungs.

People with SSc typically have problems exercising because of inflammation and stiffness in the joints, which cause pain. Let your doctor know if you have unexplained symptoms that affect physical ability, including:

- Shortness of breath with physical activity.
- Increased fatigue.
- Swelling of the ankles, legs, arms or stomach.
- Chest discomfort or pain.
- Light headedness or fainting with physical activity.

PAH usually is treatable if diagnosed early enough. PAH is a serious and complex condition that requires immediate medical treatment and care. Although many people with Ssc lead long lives, pulmonary complications, including PAH and lung fibrosis, are the leading causes of death for people with SSc.

Screening for PAH is recommended for anyone with scleroderma regardless of symptoms.

Diagnosing PH

Annual screening for PAH will result in an earlier diagnosis. The sooner treatments begin, the better the outlook.

Even when people with scleroderma aren't being screened for PAH, they should always bring shortness of breath, swelling and other symptoms to a doctor's attention.

People with SSc should have these tests every year:

Echocardiogram: An ultrasound of the heart that provides a rough estimate of the pressure in the lungs. It assesses the function of how blood flows through the heart by examining the right and left ventricles, valves, etc.

Pulmonary function tests: 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).

Natriuretic peptide tests: Measure specific proteins in a blood sample and help screen for heart failure. Tests of the proteins NT-proBNP (N-terminal pro b-type natriuretic peptide) and BNP (brain natriuretic peptide) are two examples. Elevated levels of these proteins can signify heart failure.

Right heart catheterization is the most accurate diagnostic test for PH. It measures the pressures and flow directly in the pulmonary artery. The invasive procedure usually takes place on an outpatient basis in the hospital. Doctors guide a small tube (catheter) through the right side of the heart and into the pulmonary artery to directly measure the pulmonary artery pressure.

In addition to directly measuring pressures in the heart and lungs, a right heart catheterization can determine whether patients have problems with a stiff left side of the heart. Left heart disease also can

cause PH. In those cases, doctors refer patients to a cardiologist to treat heart disease.

The goal is to diagnose PAH as early as possible before any symptoms occur. However, because of other medical conditions, it may be hard for a doctor to determine the cause of symptoms. For example, if a patient has shortness of breath while physically active, the cause could be an underlying lung fibrosis or PAH.

Many people who reduce 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.

ADDITIONAL RESOURCES

National Scleroderma Foundation | Scleroderma.org

Scleroderma Research Foundation | Srfcure.org

REFERENCES

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About the Pulmonary Hypertension Association

Headquartered in Washington, D.C., the Pulmonary Hypertension Association is the oldest and largest nonprofit patient association dedicated to the pulmonary hypertension community. PHA's mission is to extend and improve the lives of those affected by PH.

PHA engages people with PH and their families, caregivers, health care providers and researchers, who work together to advocate for the PH community, provide support to patients, caregivers and families, offer up-to-date education and information on PH, improve quality patient care, and fund and promote research. For information, visit PHAssociation.org.

PHA's mission is to extend and improve the lives of those affected by pulmonary hypertension. PHA's vision is a world without PH, empowered by hope.

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