PULMONARY ARTERIAL HYPERTENSION (PAH) GLOSSARY

Antibodies: Proteins made by your immune system that respond to and help protect your body from a specific substance. In people with scleroderma, the immune system may make antibodies that react to substances found in your own body.^{1,2}

Arteries: Vessels that carry blood from the heart to other parts of the body.3

Blood pressure: The force of your blood on artery walls as it moves from your heart through your body.4

Cardiologist: A doctor who takes care of your heart and PAH.

Complication: A medical problem that happens during the course of a disease. A complication may be caused by the disease, or it may be unrelated.⁵

Connective tissue disease: A disease that impacts the bones, cartilage, tendons, skin, ligaments, and internal organs. Examples of connective tissue disease include scleroderma, rheumatoid arthritis, mixed connective tissue disease, and lupus.⁶⁻⁸

Digital ischemia: Painful pale, white, or blue fingers due to poor blood flow. Digital ischemia can sometimes lead to sores on the fingers in people with scleroderma.⁹

Echocardiogram: A test that uses an ultrasound to show how well your heart is beating and pumping blood. It helps your doctor get a picture of your whole heart, including the right chambers.¹⁰

Edema: Swelling of your ankles, legs, and abdomen.¹¹

Limited cutaneous scleroderma: A type of scleroderma in which skin hardening and tightening occurs only on the fingers, hands, forearms, and face.⁶

Pulmonary arterial hypertension (PAH): A serious condition that can occur in people with scleroderma and in which the blood pressure in the arteries of the lungs is high.^{8,12}

Pulmonary function tests: Breathing tests that measure how much air your lungs can hold, and how well they exchange oxygen and carbon dioxide.^{8,10}

Pulmonologist: A doctor who takes care of your lungs and PAH.

Raynaud's phenomenon: A condition that can occur in people with scleroderma and in which the small blood vessels in the fingers and toes spasm in the cold. This can block blood flow and cause cold, blue fingers and painful sores.^{10,12}

Registry: A collection of standardized information about a group of patients who share a particular disease or condition.¹³

Rheumatologist: A doctor who takes care of your joints, muscles, and bones. You may already be seeing a rheumatologist for your scleroderma.

PULMONARY ARTERIAL HYPERTENSION (PAH) GLOSSARY (continued)

Right heart catheterization: A test that involves passing a thin tube, called a catheter, into the right side of the heart to test heart function and measure blood pressure in the arteries of the lungs. Right heart catheterization is needed to diagnose PAH.¹⁰

Screening: Checking for a disease when there are no symptoms to try to find it early.¹⁴

Systemic sclerosis: A condition also known as scleroderma that affects the skin, blood vessels, and major organs.⁶

Telangiectasias: Widened or broken blood vessels on the skin but may develop anywhere within the body. Also known as "spider veins." ¹⁵

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