Scleroderma and the Risk of PAH
Learning about scleroderma

Scleroderma is a type of connective tissue disease
Connective tissue diseases are a group of diseases that affect blood vessels and connective tissues, such as:

- Muscles
- Cartilage
- Tendons
- Skin
- Ligaments

These diseases may appear in one or more areas of the body. Some examples of connective tissue diseases are:

- Rheumatoid arthritis
- Systemic sclerosis
- Vasculitis
- Systemic lupus erythematosus (lupus)

Systemic sclerosis
Scleroderma, also known as systemic sclerosis, affects the blood vessels and connective tissues of the body. This disease causes hardening of the skin, scar tissue buildup, and organ damage.

Pulmonary arterial hypertension (PAH) is a well-known complication in people living with connective tissue diseases

- Scleroderma is a type of connective tissue disease
- Scleroderma is also sometimes called systemic sclerosis

If you have scleroderma, you may be at risk for PAH

Between 8% and 12% of all scleroderma patients develop PAH

At Actelion, we believe that knowledge is power. When you understand your condition, you have the power to ask the right questions, make informed decisions, and get the most from your healthcare team and your therapeutic options. That’s our goal in PAH education and that’s KNOWLEDGE IN ACTION.
What you need to know about PAH

**Pulmonary arteries are a type of blood vessel**

Pulmonary arterial hypertension (PAH) affects the arteries that pump blood from the heart to the lungs (pulmonary arteries), causing them to become stiff and thick. As the disease worsens, the thickened artery walls form scar tissue.\(^6\)

These changes make it difficult for blood to flow. The result is PAH—a serious condition that causes continuous high blood pressure in the lungs.\(^7\)

**PAH and the right side of the heart**

- With the higher blood pressure caused by PAH, the right side of the heart must work harder to pump blood through the pulmonary arteries.\(^8\)
- The strain of this overload on the heart can lead to right heart failure.\(^8\)

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**Common symptoms of PAH**\(^9\)

- Unexplained shortness of breath
- Constant tiredness
- Chest pain or discomfort
- Light-headedness and fainting (also known as syncope)
- Swelling of the ankles, legs, abdomen, or arms (also known as edema)

These symptoms may be mild at first, but they might get worse over time. If you experience these symptoms and think you may have PAH, you should contact your doctor.

*Diagnosis is important. The sooner you are diagnosed, the sooner your doctor may be able to help manage your PAH.*
KNOWLEDGE IN ACTION: SCLERODERMA AND THE RISK OF PAH

Screening for PAH

Screening is a basic tool doctors use to check your health when you are at risk for a disease. There are a few simple tests your healthcare team may recommend.¹

Once symptoms are detected, these tests can help determine if you have PAH

<table>
<thead>
<tr>
<th>Diagnostic test</th>
<th>Purpose of test</th>
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<tr>
<td>Echocardiogram</td>
<td>This test uses sound waves to create images of the heart.² It enables your healthcare team to see how well your heart is beating and pumping blood.</td>
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<tr>
<td>Electrocardiogram (ECG/EKG)</td>
<td>This test records the heart’s electrical activity. It also shows whether your heart’s rhythm is steady or irregular and may show if your right ventricle is enlarged or strained.³</td>
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<tr>
<td>Pulmonary function test (PFT)</td>
<td>PFTs are breathing tests that measure how much air the lungs can hold, how well they move air, and how well they supply the body with oxygen.⁴</td>
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Right heart catheterization is the most accurate test for PAH and provides more information about the heart’s condition¹

This test involves passing a thin tube (catheter) into the right side of the heart in order to test heart function and measure blood pressure in the arteries of the lung. Right heart catheterization is required for a definitive diagnosis of PAH.¹²

Your healthcare team

Every situation is unique. Your doctor will work closely with you and your caregivers to create a plan that is best for you. Your healthcare team will work together to find out if you have PAH. Your team may include:

- Rheumatologist
- Heart doctor (cardiologist) or lung doctor (pulmonologist) to diagnose and treat your PAH
- Nurses to help manage your care and to answer any questions you may have
- Nutritionist, in case you need help managing a diet recommended for PAH patients
  - Fluid retention is problematic for patients with PAH, so you may need to reduce the amount of sodium in your diet¹³
- Social worker, to help address life challenges and support well-being