FREQUENTLY ASKED QUESTIONS ABOUT PAH

Which parts of the body does scleroderma affect?

There are 2 main types of systemic sclerosis, or scleroderma. Localized scleroderma primarily affects the skin, while systemic sclerosis affects the skin, blood vessels, and major organs.[1]

Can scleroderma affect the lungs?

Scleroderma is not just a skin condition. It can also cause serious lung complications such as pulmonary arterial hypertension (PAH).[2,3] PAH is a disease of high blood pressure in the lungs.[4]

How does PAH affect the lungs?

Arteries that are healthy allow blood to flow easily through the vessels. In PAH, the blood pressure in the arteries of your lungs is high. This is because the arteries that carry blood from your heart to your lungs have become stiff and narrow, making it difficult for blood to flow through your lungs. This can force your heart to have to work harder than normal.[5]

Is PAH serious?

Yes, PAH can be life-threatening, especially for people with scleroderma.[6] PAH was the leading cause of death—ahead of cancer, infections, and other causes—in a registry[7]* of people with PAH associated with systemic sclerosis.[8]

While there is no cure for PAH, medications are available.[6,9] So, if you find out you also have PAH, discuss your options with your doctor.

How does scleroderma cause PAH?

Although scleroderma and PAH are 2 separate conditions, inflammation may play an important role in both.[10] Scleroderma can cause inflammation, which may then increase your chances of developing PAH.[5,10,11]

How common is PAH in people with scleroderma?

PAH can occur in people with different connective tissue diseases but is most common in people with scleroderma.[6] Five percent to 12% of people with scleroderma will also develop PAH.[12]

What are the symptoms of PAH?

Noticeable signs of PAH may start out mild but can get worse with time. They include light-headedness; fainting; constant tiredness; shortness of breath; chest pain; and swelling of the ankles, legs, or abdomen (also called edema).[4,5,7,13] But PAH can begin before you notice it, so screening every year is recommended by PAH experts and may help to detect it early.[12,14-16]

Which tests are used to screen for PAH?

Some signs of PAH are silent, so you may not see or feel them. Blood work, lung function tests, or an ultrasound of your heart can reveal signs of PAH that indicate the need for diagnostic testing. These screening tests can show if you might have PAH.[13,14]

What test confirms a diagnosis of PAH?

In order to confirm a PAH diagnosis, a test called right heart catheterization is required. Your doctor may order a right heart catheterization if other tests suggest you might have PAH.[6,13,14]

Is PAH curable in people with scleroderma?

No, there is no cure for PAH. However, regular screening may help to detect PAH early. PAH experts† recommend that you get screened every year. If you are diagnosed with PAH, medicines are available that may be able to help. Refer to your doctor.[6,12,14,15]

*A registry is a collection of standardized information about a group of patients who share a particular disease or condition.

†Recommendations for annual PAH screening of people with systemic sclerosis are included in the 2015 European Society of Cardiology (ESC)/European Respiratory Society (ERS) Guidelines, proceedings from the 6th World Symposium of Pulmonary Hypertension, and results of a consensus survey of experts in the field of PAH and scleroderma.[12,14,15]

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