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 mainly affects the skin, while systemic sclerosis affects the skin, blood vessels, and major organs.

Can scleroderma affect the lungs?

Scleroderma impacts your skin, but can also affect your lungs. People with scleroderma can develop pulmonary arterial hypertension (PAH). PAH is a disease of high blood pressure in the lungs.

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 hard for blood to flow through your lungs. As the muscles work harder to get blood flowing, the walls on the right side of your heart get thicker.

Is PAH serious?

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

While there is no cure for PAH, treatments are available. 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. Scleroderma can cause inflammation, which may then increase your chances of developing PAH.

How common is PAH in people with scleroderma?

PAH can happen in people with different connective tissue diseases, but is most common in people with scleroderma. In an international study of 466 adults with scleroderma at increased risk of PAH, 19% were confirmed to have PAH-scleroderma.

Black people with scleroderma may develop PAH more frequently than the overall population of people with scleroderma.

What are the symptoms of PAH?

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

Recommendations for yearly PAH screening of people with scleroderma are included in the 2022 European Society of Cardiology/European Respiratory Society Guidelines.

Which tests are used to screen for PAH?

Some signs of PAH may not be seen or felt. 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.

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.

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.

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

[†]Recommendations for yearly PAH screening of people with systemic sclerosis are included in the 2022 European Society of Cardiology/European Respiratory Society Guidelines.

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