

# QUESTIONS TO ASK YOUR DOCTOR ABOUT PAH

Pulmonary arterial hypertension (PAH) is a disease of high blood pressure in the lungs. PAH can occur in people with different connective tissue diseases, but it is more common in people with systemic sclerosis, or scleroderma.<sup>1,2</sup> **Because PAH can begin before you notice it, annual screening is an important tool that can help your doctor catch it early.**<sup>3,4</sup>

Review these topics at your next appointment with the doctor who treats your scleroderma and find out when you are due for your next screening for PAH.

## Assessing Risk of PAH

When starting a conversation with your doctor, be sure to mention any PAH symptoms or risk factors you may have.

### Have you experienced any symptoms of PAH<sup>2,5-7</sup> since your last appointment?

*Check all that apply.*

- |   |  |   |
|---|--|---|
| <input type="checkbox"/> Light-headedness   | <input type="checkbox"/> Shortness of breath | <input type="checkbox"/> Fainting                                 |
| <input type="checkbox"/> Constant tiredness | <input type="checkbox"/> Chest pain          | <input type="checkbox"/> Swelling of the ankles, legs, or abdomen |

### Do you have any known risk factors for PAH<sup>1,8-10</sup> associated with scleroderma?

*Check all that apply. You may need to ask your doctor about some of these risk factors.*

- Was diagnosed with scleroderma at age 60 years or older
- Have had scleroderma for a long time
- Raynaud's phenomenon
- Limited cutaneous scleroderma
- Telangiectasias (spider veins on your skin but may develop anywhere within the body)
- Digital ischemia (painful pale, white, or blue fingers)
- Presence of certain antibodies
- Results in the low range on certain pulmonary function tests

## About Screening for PAH

### Have you ever been screened for PAH? If so, when and what were your results?

Date of last screening: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Tests and results: \_\_\_\_\_

\_\_\_\_\_

## Have you ever been screened for PAH? If so, when and what were your results? (continued)

Date of last screening: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Tests and results: \_\_\_\_\_  
\_\_\_\_\_

Date of last screening: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Tests and results: \_\_\_\_\_  
\_\_\_\_\_

Date of last screening: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Tests and results: \_\_\_\_\_  
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Date of last screening: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Tests and results: \_\_\_\_\_  
\_\_\_\_\_

Date of last screening: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Tests and results: \_\_\_\_\_  
\_\_\_\_\_

## When will you be screened again for PAH?

Date: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_ Time: \_\_\_\_\_ Location: \_\_\_\_\_

Date: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_ Time: \_\_\_\_\_ Location: \_\_\_\_\_

Date: \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_ Time: \_\_\_\_\_ Location: \_\_\_\_\_

## Potential Questions to Ask Your Medical Team About the Management of PAH

- How many patients with PAH associated with scleroderma have you had in your practice?
- Are there other doctors or members of your medical team that you work with if one of your patients has PAH associated with scleroderma?
- What happens if I do have PAH associated with scleroderma?

**References:** 1. Fischer A, Bull TM, Steen VD. Practical approach to screening for scleroderma-associated pulmonary arterial hypertension. *Arthritis Care Res (Hoboken)*. 2012;64(3):303-310. 2. Chaisson NF, Hassoun PM. Systemic sclerosis-associated pulmonary arterial hypertension. *CHEST*. 2013;144(4):1346-1356. 3. Coghlan JG, Denton CP, Grünig E, et al; DETECT study group. Evidence-based detection of pulmonary arterial hypertension in systemic sclerosis: the DETECT study. *Ann Rheum Dis*. 2014;73(7):1340-1349. 4. Khanna D, Gladue H, Channick R, et al. Recommendations for screening and detection of connective tissue disease-associated pulmonary arterial hypertension. *Arthritis Rheum*. 2013;65(12):3194-3201. 5. Hayes GB. *Pulmonary Hypertension: A Patient's Survival Guide*. 5th ed. Pulmonary Hypertension Association; 2012. 6. Mandras SA, Ventura HO, Corris PA. Breaking down the barriers: why the delay in referral for pulmonary arterial hypertension? *Ochsner J*. 2016;16(3):257-262. 7. Vachiéry J-L, Gaine S. Challenges in the diagnosis and treatment of pulmonary arterial hypertension. *Eur Respir Rev*. 2012;21(126):313-320. 8. Yaqub A, Chung L. Epidemiology and risk factors for pulmonary hypertension in systemic sclerosis. *Curr Rheumatol Rep*. 2013;15(1):302. 9. Penn Medicine. Telangiectasia (spider veins). Accessed November 18, 2024. <https://www.pennmedicine.org/for-patients-and-visitors/patient-information/conditions-treated-a-to-z/telangiectasia-spider-veins> 10. McMahan ZH, Wigley FM. Raynaud's phenomenon and digital ischemia: a practical approach to risk stratification, diagnosis and management. *Int J Clin Rheumatol*. 2010;5(3):355-370.