

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.^{1,2} **Because PAH can begin before you notice it, annual screening is an important tool that can help your doctor catch it early.**^{3,4}

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^{2,5-7} since your last appointment?

Check all that apply.

- Light-headedness
- Shortness of breath
- Fainting
- Constant tiredness
- Chest pain
- Swelling of the ankles, legs, or abdomen

Do you have any known risk factors for PAH^{1,8-10} 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: _____

QUESTIONS TO ASK YOUR DOCTOR ABOUT PAH (continued)

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: _____

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 September 28, 2021. <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.