Q: I am a scleroderma patient and my doctor has recently also diagnosed me with pulmonary hypertension (PH). As I have looked for more information on this topic, I have seen that it is sometimes referred to as pulmonary arterial hypertension (PAH). Is PAH different from PH?

A: Yes, there is a difference between pulmonary hypertension (PH) and pulmonary arterial hypertension (PAH).

PAH is a subgroup of PH, meaning that PAH is a more specific term, implying that the disease is due to changes in the arteries and arterioles of the lung, as oppose to, for example, disease of the left side of the heart with back pressure into the lungs. The classification of PH includes five groups, of which PAH is the first. PAH occurring in association with scleroderma is generally considered a subgroup of this first diagnostic group, and it may be referred to as “APAH” (associated pulmonary arterial hypertension), occurring in association with scleroderma.

The updated classification of PH is fully described in “Updated Clinical Classification of Pulmonary Hypertension,” Journal of the American College of Cardiology, Volume 54, No. 1, Supplement S; 2009.

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