ADVOCACY RESOURCE

This resource aims to support multidisciplinary stakeholders, including healthcare professionals, patient advocates, healthcare management and decision-makers, to support advocacy dialogue, assess opportunities for optimal diagnosis and treatment of pulmonary arterial hypertension (PAH), and instigate policy and organisational change initiatives.

EARLY DETECTION OF PULMONARY HYPERTENSION BY RHEUMATOLOGISTS

1 WHAT IS PULMONARY ARTERIAL HYPERTENSION (PAH)?

PAH is a rare and progressive – but treatable – form of pulmonary hypertension (PH) that arises when the pulmonary arteries narrow.[1,2] PH is defined by a mean pulmonary arterial pressure (mPAP) >20 mmHg at rest and pulmonary vascular resistance (PVR) >2 WU.[1,2] The diagnosis requires a right-heart catheterisation (RHC) performed in a PH expert centre.

2 WHAT IS THE BURDEN OF PAH ON PEOPLE WITH SYSTEMIC SCLEROSIS?

The prevalence of PAH is estimated at **8–12%** in people with systemic sclerosis.[3,4]

PAH is the **second leading cause of death** in people with systemic sclerosis.[5]

The presence of **PAH doubles the risk of death** in people with systemic sclerosis.[5]

TIME IS ESSENTIAL

Fewer than 50% of people with both sclerosis and PAH survive five years after diagnosis.[6,7]

3 PAH SCREENING IN SYSTEMIC SCLEROSIS

According to the 2022 PH clinical guidelines, the pathway to detection in people with systemic sclerosis includes:[1]

Suspicion by first-line rheumatologists

Annual PAH screening (pulmonary function testing and DETECT algorithm (see *Figure 1*))[1]

Referral to echocardiography

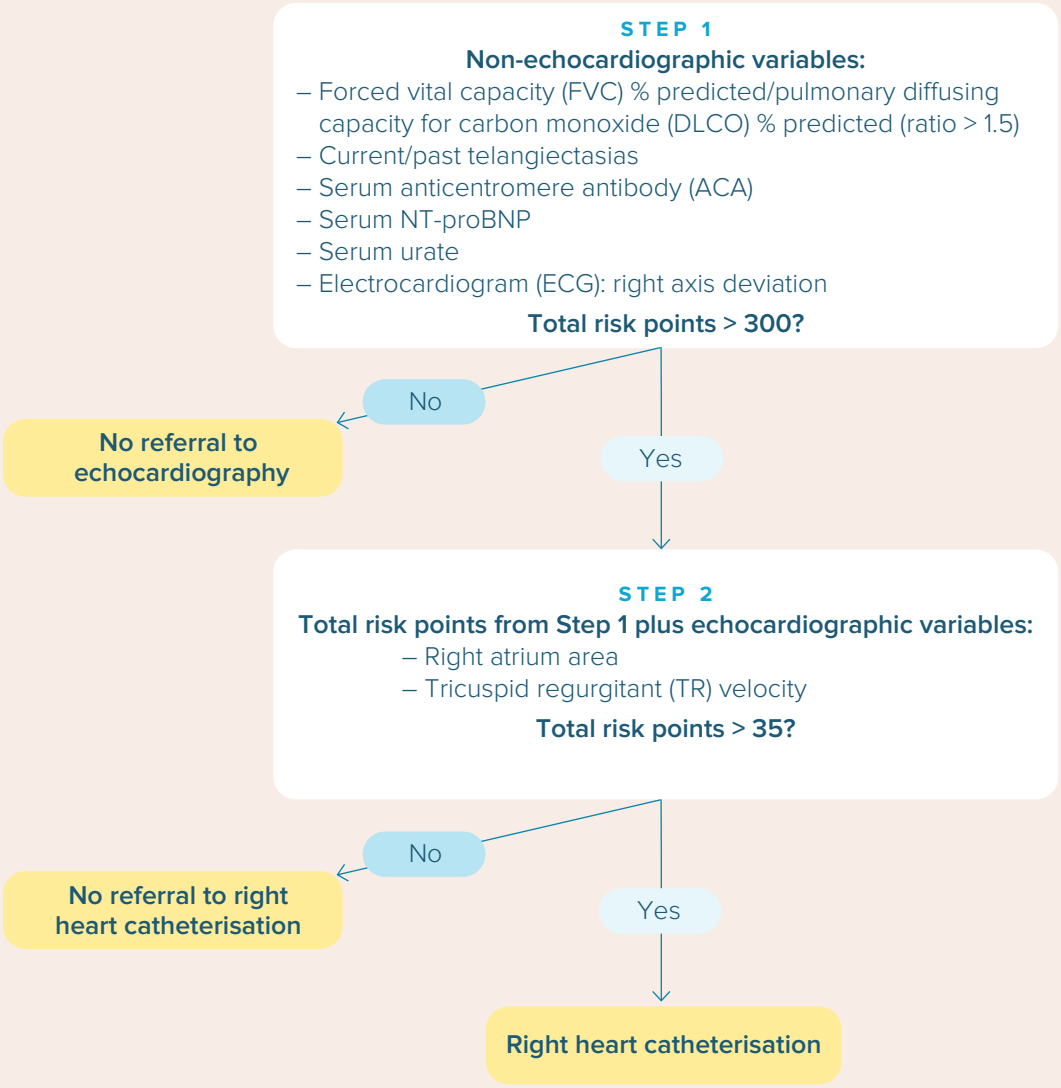
Confirmation with RHC in PH centres

What is the impact of screening?

PAH treatment in people with the condition and connective tissue disease (CTD) can **reduce risk of clinical worsening by almost 40%.**[8]

People with systemic sclerosis who are actively screened for PAH experience an **improved five-year survival rate to 63–73%.**[9,10]

Figure 1. Two-step decision flowchart for the detection of PAH in people with systemic sclerosis[11]



Not for use in clinical decision-making; please see disclaimer below.

4 ACTIONS FOR THE EARLY DETECTION OF PAH

Integrate PH screening in existing protocols for the treatment of people with systemic sclerosis.

Promote multidisciplinary collaboration between rheumatologists, cardiologists and pulmonologists to provide best-practice care for people with both systemic sclerosis and PAH.

The information in this resource is not intended for use in clinical decision-making. Please refer to the latest formal guidelines in your country or region. This document is not intended as educational material. The information is based on clinical guidelines to ensure accuracy, but it is for advocacy purposes only to assist multidisciplinary audiences in taking action on the topic.

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The Health Policy Partnership

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