

Evaluation of Suspected Pulmonary Hypertension in Systemic Sclerosis

PH Risk Factors

- Limited cutaneous SSC
- Disease duration >3 years
- Age > 50 years
- Female sex
- Interstitial lung disease

Symptoms

- Fatigue
- Dyspnea on exertion
- Palpitations
- Chest pain
- Lower extremity edema
- Ascites
- Right upper quadrant pain
- Cough
- Lightheadedness/near syncope

Pulmonary Function

- Diffusion (DLCO) < 60% predicted
- FVC/DLCO >1.6
- Isolated drop in diffusion by 15%

Six Minute Walk Test

- Poor distance
- Oxygen desaturation
- 1 minute heart rate recovery <13 beats/min

Electrocardiogram

- Right atrial enlargement
- Right ventricular hypertrophy
- Right ventricular strain
- Right bundle branch block

Imaging

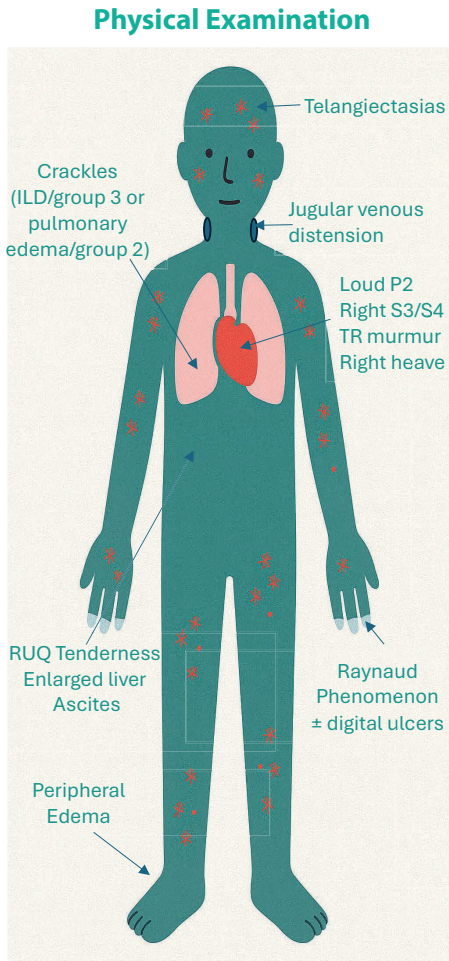
- PA:Ao ratio >1
- RV/LV ratio >1
- PA >32 mm
- Peripheral pruning
- Interstitial lung disease
- PVOD (centrilobular ground glass, reticulation, adenopathy)

Echocardiography

- RVSP >35 mmHg
- TRV >2.8 m/s
- Abnormal rightsided morphology
- Abnormal rightsided function
- Pericardial effusion

Biomarkers

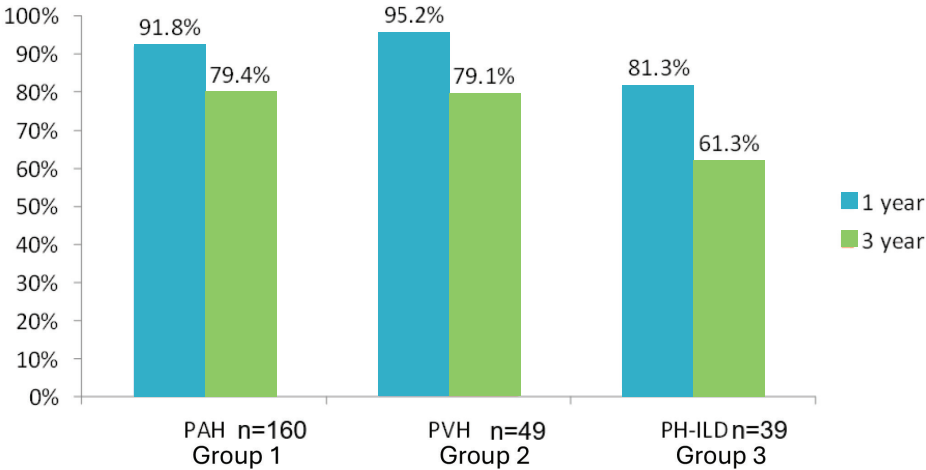
- Elevated nt-pro BNP or BNP
- Anticentromere antibody (PAH risk)
- Anti-Scl-70 (ILD risk)
- Nucleolar ANA



Refer Early!

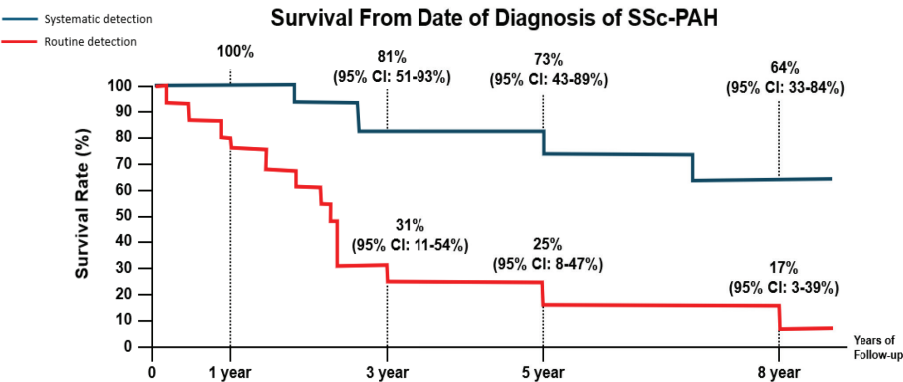


Survival in Scleroderma Patients by Type of PH¹⁰



10. Gordon JK et al. ACR/AHRP Annual Meeting. Nov 9-14, 2012. Washington DC. Abstract 1465

Potential Survival Benefit with Screening¹¹

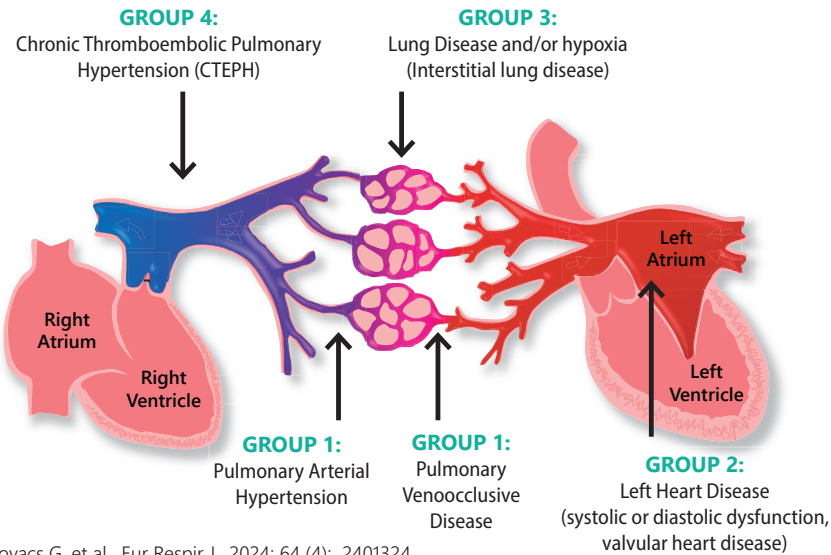


N=16 patients diagnosed with SSc-PAH via systematic detection program compared with 16 matched controls diagnosed through routine clinical practice. P=0.0037, HR=4.15.

11. Humbert M, et al. Arthritis Rheum 2011; 63: 3522-3530

Screening for Pulmonary Hypertension in Patients with Systemic Sclerosis (Scleroderma)

Clinical Classification of Pulmonary Hypertension¹ in Scleroderma



1. Kovacs G, et al. Eur Respir J. 2024; 64 (4): 2401324

Hemodynamic Definition of Pulmonary Hypertension²

Definition	Hemodynamic characteristics	Group
PH	mPAP >20 mmHg	
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU	1, 3, 4
lpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU	2
CpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU	Mixed

2. Humbert M, et al. Eur Respir J. 2023; 61 (1): 2200879

This educational resource was developed with support from United Therapeutics



A Thorough Evaluation of the Right Heart is Necessary When Using Echo to Screen for PH

DETECT⁵



Australian Scleroderma Interest Group Screening Algorithm^{7,8}

Tricuspid regurgitant jet velocity (TRV) is used with other ECHO signs of PH to determine ESC/ERS probability of PH³

Low probability	Intermediate probability	High probability
TRV ≤2.8 m/s or unmeasurable, Echo signs of PH from ≤1 category	TRV 2.9-3.4 m/s, Echo signs of PH from ≤1 category OR TRV ≤2.8 m/s plus signs from >1 Echo category	TRV >3.4 m/s OR TRV 2.9-3.4 m/s plus signs from >1 Echo category

Probability of PH: Echo signs by category		
Ventricles	Pulmonary artery	IVC and RA
• RV/LV diameter/area ratio >1.0 • TAPSE/sPAP ratio <0.55 mm/mm Hg • Septal flattening (LVEI >1.1 in systole and/or diastole)	• RVOT AT <105 ms and/or mid-systolic notching • PA diameter >AR diameter • PA diameter >25 mm • Early diastolic pulmonary regurgitation velocity >2.2 m/s	• RA area (end-systole) >18 cm ² • IVC diameter >21 mm with decreased inspiratory collapse*

3. Humbert M, et al. Eur Heart J. 2022; 43 (38): 3618-3731

When to Consider Right Heart Catheterization⁴

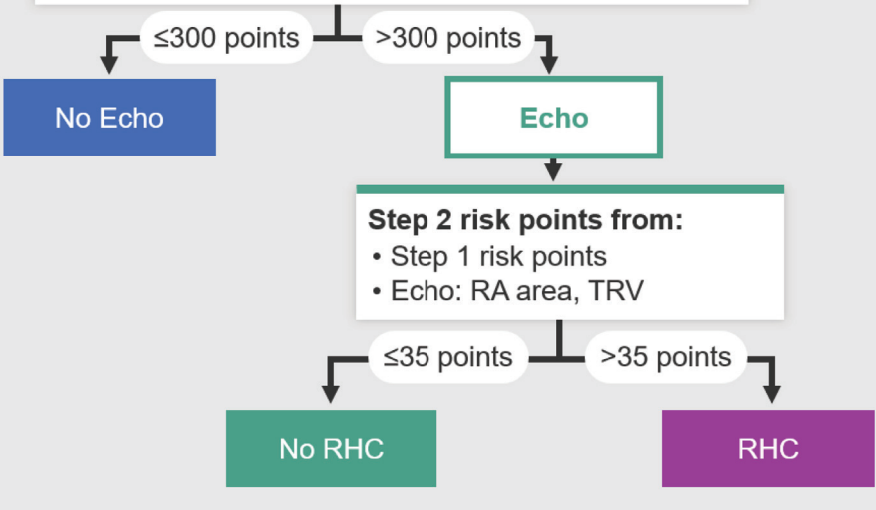
Clinical suspicion of PH	Low	Moderate	High
Echocardiogram	Low	Moderate	High
	No RHC Observe/ Re-evaluate	RHC* (If results impact management)	

A right heart catheterization is required to confirm the diagnosis of pulmonary hypertension

4. Shoblin OA et al. Eur Respir J. 2024; 64 (4): 2401200

Step 1 risk points determine the need for Echo:

- FVC/D_{LCO} ratio
- Telangiectasia Hx
- Anti-centromere Ab
- NT-proBNP
- Urate
- ECG: right axis deviation



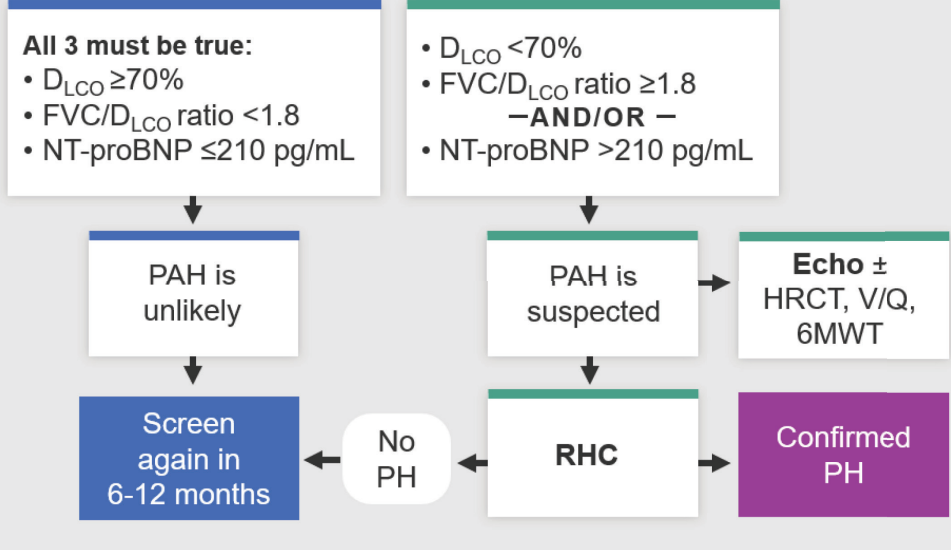
5. Coghlan JG, et al. Ann Rheum Dis 2014; 73 (7): 1340-1349
*FVC ≥ 40% predicted, DLCO <60% predicted, and SSc > 3 years

2024 WSPH⁶ RECOMMENDATIONS:

- ANNUAL ECHO, ECG, AND NT-PRO BNP ARE USEFUL TOOLS
- DETECT CAN BE USED FOR PATIENTS MEETING ITS KEY ENROLLMENT CRITERIA*

6. Kovacs G, et al. Eur Respir J. 2024 64(4): 2401324

PFT and NT-proBNP results determine the need for RHC



7. Thakkar V, et al. Arthritis Res Ther. 2012; 14(3): R143
8. Thakkar V, et al. Arthritis Res Ther 2013; 15 (6): R193

2022 ESC/ERS Guidelines⁹ recommend yearly multimodal screening for patients with scleroderma spectrum disorders

9. Humbert M, et al. Eur Heart J 2022; 43 (38):3618-3713