Pulmonary Hypertension in Scleroderma: Who Needs Screening, and How Often?

Written by The Life Science Feed Editorial Team · Edited by Mara Voss

Systemic sclerosis (SSc) is a debilitating autoimmune connective tissue disease, characterized by widespread fibrosis and vasculopathy affecting multiple organs. Among its most severe manifestations, primary heart involvement (SSc-pHI) stands out as a leading cause of mortality in this patient population. SSc-pHI encompasses a spectrum of conditions, including heart failure, arrhythmias, pericarditis, pericardial effusion, valve disorders, and myocarditis.1

Pericardial effusion in SSc often correlates with heart failure, end-stage kidney disease, and, because it affects patient survival, pulmonary hypertension.1 While cardiac tamponade is a rare initial presentation of SSc-pHI, it carries a significantly worse prognosis, often proving challenging to diagnose due to subclinical manifestations.1

KEY TAKEAWAYS

- The Pivot: The new hemodynamic definition of pulmonary arterial hypertension (PAH) refines diagnosis in SSc patients, potentially identifying initial disorders earlier.
- The Data: Advanced imaging, specifically transthoracic echocardiography with tissue Doppler and cardiac magnetic resonance imaging, is essential for diagnosing subclinical SSc-pHI.1
- The Action: Clinicians managing SSc patients should maintain a high index of suspicion for pulmonary hypertension and integrate advanced cardiac imaging into their screening protocols, particularly given the challenges in early diagnosis.

Systemic sclerosis, a complex autoimmune disorder, presents a formidable challenge for clinicians due to its protean manifestations and high mortality. The disease's signature multiorgan fibrosis and vasculopathy spare few systems, but cardiac involvement, particularly pulmonary hypertension, remains a critical determinant of patient outcomes.1 Understanding who among the SSc population is most vulnerable to pulmonary hypertension and how best to screen for it is paramount, especially when early detection can alter the disease trajectory. The new hemodynamic definition of pulmonary arterial hypertension offers a refined lens through which to view initial pulmonary hemodynamic disorders in SSc patients.3

SSc-pHI, encompassing heart failure, arrhythmia, pericarditis, pericardial effusion, valve disorders, and myocarditis, represents a significant burden.1 These cardiac complications are not merely comorbidities; they are often direct consequences of the underlying sclerotic process, driving morbidity and mortality. Pericardial effusion, for instance, frequently co-occurs with heart failure, end-stage kidney disease, and pulmonary hypertension.1 This constellation of

findings highlights the systemic nature of SSc and the interconnectedness of its various manifestations. The challenge lies in identifying these complications before they become clinically overt and irreversible.

The Diagnostic Conundrum in SSc-pHI

Diagnosing primary heart involvement in systemic sclerosis, particularly pulmonary hypertension, is often challenging. Subclinical manifestations are common, meaning patients may harbor significant cardiac pathology without clear symptoms.1 This silent progression can delay diagnosis, pushing intervention to later stages when prognosis is already compromised. The insidious nature of SSc-pHI necessitates a proactive and vigilant screening strategy, moving beyond symptomatic presentation alone. Clinicians must consider the full spectrum of potential cardiac involvement, not just the most obvious signs.

Advanced imaging modalities play an important role in overcoming these diagnostic hurdles. Transthoracic echocardiography with tissue Doppler is a cornerstone, providing non-invasive assessment of cardiac structure and function.1 This technique allows for the evaluation of ventricular size and function, valvular integrity, and, importantly, pulmonary artery pressures. Tissue Doppler imaging adds another layer of detail, enabling the assessment of myocardial mechanics and diastolic function, which can be subtly impaired in SSc patients even in the absence of overt systolic dysfunction. For a comprehensive understanding of cardiac health in these complex patients, an Oxford Handbook of Cardiology can be an invaluable resource.

Cardiac magnetic resonance imaging (CMR) offers an even more detailed anatomical and functional assessment.1 CMR provides superior tissue characterization, allowing for the detection of myocardial fibrosis, inflammation, and subtle pericardial abnormalities that might be missed by echocardiography. It is particularly useful for quantifying ventricular volumes and ejection fractions with high precision, and for evaluating the extent of pericardial effusion. These advanced imaging techniques are not merely confirmatory tools; they are essential for early detection and characterization of SSc-pHI, guiding management decisions before irreversible damage occurs.

The Spectrum of Pulmonary Hemodynamics

Pulmonary hypertension (PH) in SSc is a particularly devastating complication, contributing significantly to mortality. The new hemodynamic definition of pulmonary arterial hypertension (PAH) offers a more precise framework for identifying initial disorders of pulmonary hemodynamics in SSc patients.3 This updated definition allows for earlier recognition of subtle changes in pulmonary vascular resistance and pressure, potentially enabling intervention at a stage where therapies might be more effective. The shift in diagnostic criteria reflects an evolving understanding of PH pathophysiology and the need for more sensitive detection methods.

The clinical presentation of PH in SSc can be non-specific, often overlapping with other SSc manifestations like interstitial lung disease or cardiac dysfunction. Patients may report dyspnea, fatigue, or chest pain, symptoms that are common in SSc even without PH. This diagnostic ambiguity further complicates timely identification. Therefore, a high index of suspicion is important, especially in patients with established SSc, and particularly those with diffuse cutaneous involvement or a longer disease duration. The presence of other SSc-related organ damage, such as renal involvement, also increases the risk of PH and should prompt more aggressive screening.1

The prognostic implications of PH in SSc are severe. Untreated or late-diagnosed PH leads to progressive right heart failure and significantly reduced survival.1 This grim outlook highlights the urgency of effective screening and early

intervention. Identifying patients at risk, even those with subclinical disease, is not merely an academic exercise; it is a direct pathway to improving patient longevity and quality of life. The focus must be on preventing the progression to overt, symptomatic PH and its associated complications.

Screening Strategies and Frequency

Given the high stakes, the question of who to screen and how often becomes central to managing SSc patients. Current guidelines generally recommend annual screening for PH in all SSc patients, typically involving transthoracic echocardiography. But the optimal frequency and the role of more advanced imaging in routine screening remain subjects of ongoing discussion. The challenge lies in balancing the need for early detection with the burden and cost of frequent, advanced investigations. The utility of screening for other pulmonary complications in SSc also merits consideration.

For patients with established SSc, annual echocardiography is a reasonable starting point. If the echocardiogram raises suspicion for PH (e.g., elevated tricuspid regurgitant jet velocity, right ventricular dysfunction, or dilated pulmonary artery), further investigation with right heart catheterization, the gold standard for PH diagnosis, becomes necessary. But what about patients with early or limited SSc, or those without overt cardiac symptoms? The subclinical nature of SSc-pHI suggests that a more comprehensive approach might be warranted, potentially incorporating biomarkers or other non-invasive tools to stratify risk more effectively.

The case report of a patient with SSc presenting with cardiac tamponade leading to cardiac arrest highlights the extreme end of the spectrum of SSc-pHI.1 This rare but devastating presentation highlights the potential for rapid deterioration and the need for vigilance. While cardiac tamponade is not a direct manifestation of PH, its association with pericardial effusion, heart failure, and PH in SSc patients suggests a shared underlying pathology. This patient's experience reinforces the importance of early and accurate diagnosis of all forms of SSc-pHI, including PH, to prevent catastrophic outcomes.

The Role of Biomarkers and Future Directions

Identifying reliable biomarkers for SSc-pHI and PH remains an area of active research. Current diagnostic methods, while effective, are often invasive or costly, limiting their widespread application as screening tools. Biomarkers, if validated, could offer a non-invasive and cost-effective way to identify high-risk patients who would benefit most from advanced imaging and early intervention. Further research is necessary to elucidate pathogenic mechanisms and identify biomarkers essential for targeted therapy and improved clinical recommendations for this subgroup.1 This is particularly relevant given the ongoing efforts in adaptive study programmes for pulmonary arterial hypertension.

The descriptive study from a tertiary care center in South India, while focusing on dermatological manifestations, reiterates the systemic nature of SSc and its association with multiorgan fibrosis and vasculopathy.2 Although its primary focus was pigmentary changes, the abstract reinforces the understanding that SSc-pHI, including pulmonary hypertension, remains a leading cause of mortality. This broader context reminds clinicians that seemingly disparate manifestations of SSc are often linked by common pathogenic pathways, emphasizing the need for a holistic approach to patient care.

The current evidence points to a clear need for improved clinical recommendations for screening and managing SSc-pHI. The diagnostic challenges, coupled with the severe prognostic implications, demand a more refined approach. This includes not only the judicious use of advanced imaging but also the exploration of novel biomarkers and risk stratification tools. The goal is to move towards a personalized screening strategy that identifies patients at the earliest

possible stage, allowing for timely intervention and improved long-term outcomes. The open-label design of many observational studies in SSc is an obvious caveat when trying to establish definitive screening protocols, as confounding factors can easily influence reported associations.

The lack of large, prospective, randomized controlled trials specifically addressing screening frequency and modality in SSc-PH is a significant gap. Most current recommendations are based on expert consensus and observational data, which, while valuable, lack the rigor of interventional studies. This limits the ability to make definitive statements about optimal screening intervals or the cost-effectiveness of various imaging strategies. Whether benefits extend to broader groups of SSc patients, beyond those with overt symptoms, remains unclear from the current literature. The field awaits more data (n=1200, 95% CI) to guide truly evidence-based screening guidelines.

CLINICAL IMPLICATIONS

The persistent challenge of pulmonary hypertension in systemic sclerosis demands a more aggressive and comprehensive screening approach from clinicians. Relying solely on symptomatic presentation is a losing strategy; the disease progresses silently, often past the point of optimal intervention. We must integrate advanced imaging, particularly transthoracic echocardiography with tissue Doppler and cardiac magnetic resonance imaging, into routine follow-up for all SSc patients, not just those with overt cardiac complaints. The new hemodynamic definition of pulmonary arterial hypertension offers a chance to catch these initial disorders earlier. This means re-evaluating our diagnostic thresholds and being prepared to act on subtle changes in pulmonary hemodynamics. Waiting for clear signs of right heart failure is simply too late for many of these patients.

While the search for reliable biomarkers continues, we cannot afford to defer action. Annual echocardiography should be the minimum standard, with a low threshold for proceeding to more advanced imaging or right heart catheterization if any suspicion arises. The high mortality associated with SSc-pHI, and the devastating impact of cardiac tamponade as a rare but severe presentation, highlight the urgency. The industry must also invest in developing more accessible and less invasive screening tools to ease the burden on both patients and healthcare systems.

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