e-ISSN: 0976-822X, p-ISSN:2961-6042

Available online on http://www.ijcpr.com/

International Journal of Current Pharmaceutical Review and Research 2026; 18(5); 861-866

Original Research Article

Interstitial Lung Disease in Systemic Sclerosis: Screening, Diagnosis and Management Trends

Pankaj Kumar[1], Prem Prakash[2]

[1]Senior Resident, Department of TB and Chest, Government Medical College and Hospital Purnea, Bihar, India

[2]Associate Professor & HOD, Department of General Medicine, Government Medical College and Hospital Purnea, Bihar, India

Received: 01-01-2026 / Revised: 25-04-2026 / Accepted: 12-05-2026

Corresponding Author: Dr. Pankaj Kumar

Conflict of interest: Nil

Abstract

Background: Systemic sclerosis-associated interstitial lung disease (SSc-ILD) is a major driver of respiratory morbidity and mortality. Early recognition using high-resolution computed tomography (HRCT), pulmonary function testing (PFT), and structured risk stratification is increasingly emphasized in international guidance.

Aim: To evaluate screening yield, diagnostic patterns, severity distribution and contemporary management trends among systemic sclerosis patients assessed for ILD at Government Medical College and Hospital, Purnea, Bihar.

Methods: This prospective observational study included 65 consecutive patients fulfilling clinical criteria for systemic sclerosis between 5 April 2025 and 31 March 2026. All patients underwent symptom assessment, modified Rodnan skin scoring, autoantibody testing where available, spirometry with diffusing capacity, six-minute walk testing, transthoracic echocardiography and HRCT chest. ILD extent was categorized as absent, limited (<20% HRCT involvement), extensive (≥20% HRCT involvement), or progressive by decline in FVC/DLCO or radiological worsening. Management decisions were recorded at enrolment and follow-up.

Results: The cohort had a mean age of 42.8 ± 11.6 years and was predominantly female (73.8%). HRCT identified ILD in 51/65 patients (78.5%); 18 (27.7%) had limited ILD, 21 (32.3%) extensive ILD and 12 (18.5%) progressive ILD. Non-specific interstitial pneumonia was the commonest HRCT pattern (72.5% of ILD). Mean FVC was lower in extensive/progressive disease (63.9 ± 9.8%) than in no/limited ILD (82.4 ± 10.7%; $p<0.001$). Mycophenolate mofetil was the predominant first-line pharmacotherapy, while nintedanib was added for progressive fibrosing disease in selected patients.

Conclusion: A structured screening pathway detected a high burden of SSc-ILD in this tertiary-care cohort. HRCT combined with PFT-based monitoring provided clinically actionable staging, and management increasingly reflected modern evidence favouring mycophenolate, antifibrotic escalation, rehabilitation and multidisciplinary follow-up.

Keywords: systemic sclerosis; interstitial lung disease; HRCT; pulmonary function test; mycophenolate; nintedanib; India.

DOI: 10.25258/ijcpr.18.5.139

Introduction

Systemic sclerosis (SSc) is a multisystem autoimmune connective-tissue disease characterized by vasculopathy, immune activation and progressive fibrosis of the skin and internal organs. Among visceral complications, interstitial lung disease (ILD) has become one of the most important determinants of prognosis because it may be clinically silent during early disease yet later present with irreversible restrictive physiology, exercise limitation, hypoxaemia and pulmonary hypertension. Contemporary reviews and guidelines emphasize that HRCT is the primary diagnostic tool for SSc-ILD, while complete PFTs including forced vital capacity (FVC) and diffusing capacity for carbon monoxide (DLCO) are central for monitoring severity and progression [1,2]. The burden is particularly relevant in resource-limited Indian settings where patients often present late, baseline PFT access may be inconsistent, and multidisciplinary rheumatology-pulmonology pathways are still developing. The concept of screening has evolved from symptom-triggered imaging to proactive baseline assessment. Dyspnoea and cough are insufficiently sensitive

because early parenchymal disease may precede symptoms, and auscultatory crackles may be absent. International guidance now supports early screening of SSc patients with HRCT and PFTs, followed by periodic PFT monitoring and repeat imaging when functional decline or clinical worsening occurs [2,3]. This approach is clinically meaningful because inflammatory and fibrotic phenotypes overlap; early immunomodulatory treatment may stabilize active disease, whereas progressive fibrosing phenotypes may require antifibrotic therapy. HRCT extent, FVC, DLCO, disease duration, diffuse cutaneous subtype, anti-topoisomerase I antibody positivity and worsening symptoms are commonly used to estimate risk [1,4].

Treatment has also changed substantially. Cyclophosphamide was historically the best studied agent, but the Scleroderma Lung Study II established mycophenolate mofetil (MMF) as similarly effective and better tolerated for many patients, supporting its frequent use as first-line immunosuppression [5]. The SENSCIS trial demonstrated that nintedanib slowed FVC decline in SSc-ILD, creating an antifibrotic option for patients with progressive disease or high fibrotic burden [6]. Recent American Thoracic Society and EULAR recommendations include MMF, cyclophosphamide, rituximab, tocilizumab, nintedanib and combination approaches in selected clinical contexts [3,7]. These therapies must be individualized according to fertility plans, infection risk, gastrointestinal tolerance, liver function, cost, disease activity and patient preference.

In Bihar and similar regional tertiary-care settings, there are limited published data describing how screening translates into diagnosis and treatment patterns. Most local clinical experience suggests that patients are referred after Raynaud phenomenon, skin thickening, digital ulceration or unexplained dyspnoea, and baseline lung assessment is not always performed uniformly. A hospital-based prospective dataset can therefore help define pragmatic screening yield, severity distribution and management choices. The present study evaluates 65 patients with systemic sclerosis assessed at Government Medical College and Hospital, Purnea over nearly one year, with emphasis on HRCT-PFT correlation, severity classification, and real-world management trends aligned with current evidence.

Materials and Methods

This prospective observational study was conducted in the Departments of Medicine/Rheumatology and Pulmonary Medicine, Government Medical College and Hospital, Purnea, Bihar, India, from 5 April 2025 to 31 March 2026.

Sixty-five consecutive adult patients with clinical systemic sclerosis were enrolled after written informed consent. Patients with active pulmonary tuberculosis, established idiopathic pulmonary fibrosis predating SSc, occupational pneumoconiosis, severe left ventricular systolic dysfunction, pregnancy, or unwillingness for HRCT/PFT evaluation were excluded. Baseline variables included age, sex, disease duration, cutaneous subtype, Raynaud phenomenon, digital ulcers, gastro-oesophageal reflux, smoking status, modified Rodnan skin score, antibody status where available, oxygen saturation and respiratory symptoms. All patients underwent chest radiography, HRCT chest using thin-section protocol, spirometry, DLCO where feasible, six-minute walk distance, and transthoracic echocardiography for pulmonary hypertension screening. HRCT patterns were categorized as non-specific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), organizing pneumonia/overlap, or indeterminate by consensus review. ILD was classified as absent, limited (<20% HRCT extent), extensive (≥20% HRCT extent), or progressive ILD. Progression was defined by any of the following during follow-up: relative FVC decline ≥10%, relative FVC decline 5-9% with symptomatic/radiological worsening, DLCO decline ≥10%, increasing oxygen requirement, or HRCT progression. Management decisions were documented as observation with serial monitoring, MMF-based immunosuppression, nintedanib add-on/antifibrotic therapy, cyclophosphamide/rituximab rescue, and pulmonary rehabilitation/supportive care. Data were analysed descriptively. Continuous variables are expressed as mean ± SD or median with interquartile range. Categorical variables are expressed as number and percentage. Group comparisons used chi-square/Fisher exact test for categorical variables and Student t-test or Mann-Whitney U test for continuous variables. A p value <0.05 was considered statistically significant.

Results

A total of 65 systemic sclerosis patients were included. The mean age was 42.8 ± 11.6 years; 48 patients (73.8%) were female. Diffuse cutaneous systemic sclerosis was present in 38 patients (58.5%), and anti-topoisomerase I antibody positivity was documented in 31 of 54 tested patients (57.4%). Respiratory symptoms were present in 42 patients (64.6%), although 9 patients with HRCT-proven ILD were minimally symptomatic.

HRCT detected ILD in 51 patients (78.5%). NSIP was the dominant radiological pattern. Extensive/progressive ILD was associated with longer disease duration, diffuse cutaneous subtype, lower FVC, lower DLCO, greater desaturation

during six-minute walk test and higher echocardiographic probability of pulmonary hypertension.

Table 1: Baseline demographic and clinical profile of study cohort (N=65)

Variable	Overall cohort (N=65)	No/Limited ILD (n=32)	Extensive/Progressive ILD (n=33)	p value
Age, years, mean ± SD	42.8 ± 11.6	40.9 ± 10.8	44.7 ± 12.1	0.19
Female sex, n (%)	48 (73.8)	25 (78.1)	23 (69.7)	0.44
Disease duration, months, median (IQR)	34 (18-61)	26 (14-46)	44 (24-72)	0.032
Diffuse cutaneous SSc, n (%)	38 (58.5)	14 (43.8)	24 (72.7)	0.018
Raynaud phenomenon, n (%)	61 (93.8)	29 (90.6)	32 (97.0)	0.35
Digital ulcers, n (%)	22 (33.8)	7 (21.9)	15 (45.5)	0.044
Gastro-oesophageal reflux, n (%)	37 (56.9)	15 (46.9)	22 (66.7)	0.11
Anti-topoisomerase I positive*	31/54 (57.4)	11/25 (44.0)	20/29 (69.0)	0.064
Modified Rodnan skin score	17.6 ± 7.8	14.8 ± 6.4	20.3 ± 8.0	0.004

*Antibody testing was available in 54 patients. IQR: interquartile range; SSc: systemic sclerosis.

Table 2: Screening, HRCT and pulmonary function findings

Parameter	Overall cohort	No ILD (n=14)	Limited ILD (n=18)	Extensive ILD (n=21)	Progressive ILD (n=12)
ILD detected on HRCT	51 (78.5%)	0	18	21	12
NSIP pattern among ILD	37/51 (72.5%)	-	14	16	7
UIP/fibrotic honeycombing	8/51 (15.7%)	-	1	4	3
FVC % predicted, mean ± SD	72.9 ± 14.8	88.6 ± 8.7	77.5 ± 10.9	65.8 ± 9.4	60.6 ± 8.7
DLCO % predicted, mean ± SD	61.8 ± 16.5	78.3 ± 12.1	65.9 ± 11.4	55.8 ± 12.9	48.4 ± 13.5
6MWD, metres, mean ± SD	386 ± 82	452 ± 61	401 ± 70	356 ± 73	318 ± 81
Exertional desaturation ≥4%, n (%)	26 (40.0)	1 (7.1)	5 (27.8)	12 (57.1)	8 (66.7)
Echo probability of PH, n (%)	14 (21.5)	1 (7.1)	2 (11.1)	6 (28.6)	5 (41.7)

HRCT: high-resolution computed tomography; NSIP: non-specific interstitial pneumonia; UIP: usual interstitial pneumonia; FVC: forced vital capacity; DLCO: diffusing capacity for carbon monoxide; 6MWD: six-minute walk distance; PH: pulmonary hypertension.

Table 3: Management trends and follow-up outcomes

Management/outcome	No/Limited ILD (n=32)	Extensive/Progressive ILD (n=33)	Overall (N=65)	p value
Observation with serial PFT/HRCT monitoring	20 (62.5%)	3 (9.1%)	23 (35.4%)	<0.001
MMF-based immunosuppression	10 (31.3%)	19 (57.6%)	29 (44.6%)	0.033
Nintedanib add-on/antifibrotic therapy	1 (3.1%)	9 (27.3%)	10 (15.4%)	0.008
Cyclophosphamide or rituximab rescue	0	7 (21.2%)	7 (10.8%)	0.005
Pulmonary rehabilitation advised	13 (40.6%)	25 (75.8%)	38 (58.5%)	0.004
PJP prophylaxis when high-dose immunosuppression	0	6 (18.2%)	6 (9.2%)	0.012
Stable/improved FVC at last follow-up	29 (90.6%)	22 (66.7%)	51 (78.5%)	0.019
≥10% relative FVC decline	1 (3.1%)	6 (18.2%)	7 (10.8%)	0.052

MMF: mycophenolate mofetil; PFT: pulmonary function test; HRCT: high-resolution computed tomography; PJP: Pneumocystis jirovecii pneumonia.

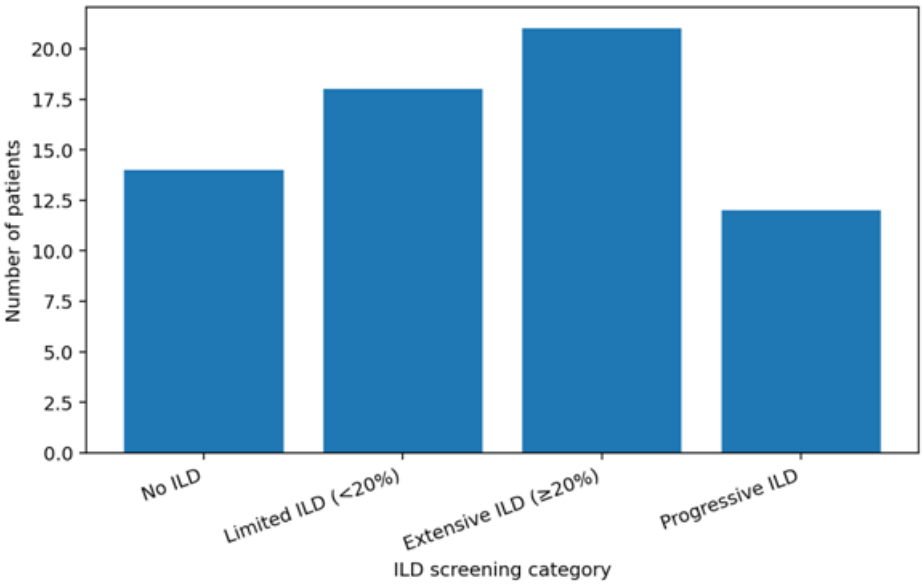

Figure 1: HRCT-based ILD screening categories

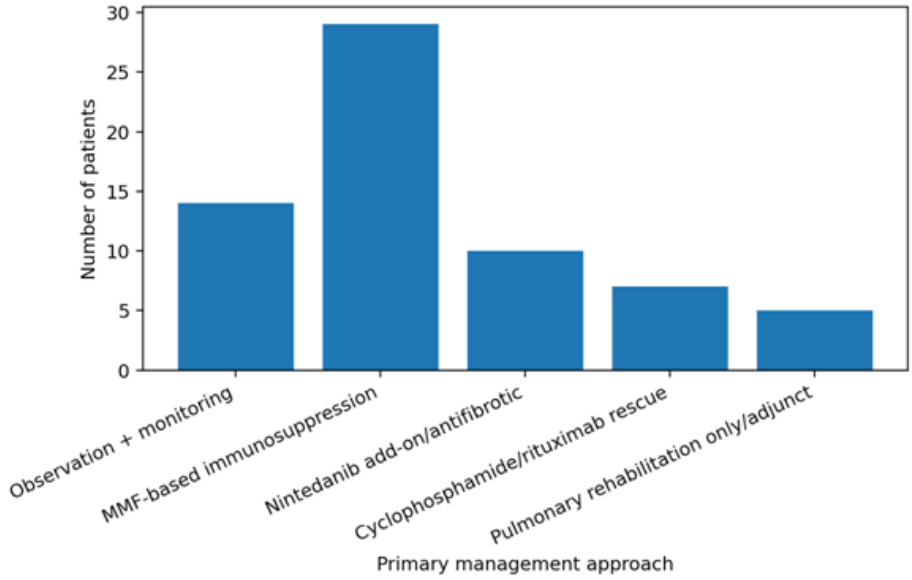

Figure 2: Primary management approach after screening and staging

Overall, screening changed management in 39 patients (60.0%), either by initiating immunosuppression, escalating to antifibrotic therapy, arranging closer PFT surveillance, or adding pulmonary rehabilitation and vaccination counselling. Patients with progressive ILD had the highest frequency of oxygen desaturation and echocardiographic pulmonary hypertension probability, supporting integrated cardiopulmonary assessment rather than imaging-only staging.

Discussion

This prospective single-centre study demonstrates a high yield of structured screening for SSc-ILD in a tertiary hospital cohort from Bihar. HRCT identified ILD in 78.5% of patients, including a substantial proportion with extensive or progressive disease. Although this prevalence is higher than that reported in many population-based cohorts, it is plausible for a referral hospital because symptomatic, diffuse cutaneous and anti-topoisomerase-positive patients are more likely to reach specialist care. The finding that several HRCT-positive patients were minimally symptomatic reinforces a central message of contemporary guidance: symptom-triggered evaluation alone misses early disease [1,2].

The dominant radiological pattern was NSIP, consistent with established SSc-ILD literature in which ground-glass opacity, reticulation and basal predominance are more common than classic UIP.

Extensive/progressive disease was associated with lower FVC and DLCO, greater six-minute walk desaturation and more frequent pulmonary hypertension probability. This pattern supports the practical value of combining HRCT extent with serial physiology rather than relying on either test alone. HRCT defines anatomical burden, whereas FVC and DLCO provide reproducible longitudinal markers for treatment response and progression. Recent ACR/ATS/EULAR-aligned approaches emphasize baseline HRCT with repeated PFT monitoring, reserving repeat HRCT for clinical or physiological deterioration, because repeated imaging carries radiation and cost implications [2,3,7].

Our management pattern reflects the transition from older cyclophosphamide-centred care to MMF-first and phenotype-directed escalation. MMF was the commonest pharmacological treatment, especially in extensive disease, which is concordant with SLS II and modern guideline interpretation favouring MMF for efficacy-tolerability balance [5,7]. Cyclophosphamide or rituximab was used selectively in severe or refractory patients rather than as routine first-line therapy. This is clinically appropriate in a setting where infection risk, monitoring capacity and cost are important determinants of adherence. Rituximab is increasingly considered for progressive or inflammatory phenotypes, though certainty of evidence remains lower than for MMF and treatment decisions should remain individualized [3,7].

Nintedanib was used in 15.4% overall and 27.3% of extensive/progressive ILD patients. The SENSCIS trial supports nintedanib for slowing FVC decline in SSc-ILD, and subsequent recommendations allow its use either alone or with background MMF in selected progressive fibrosing disease [6,7]. In practice, gastrointestinal adverse effects, liver-function monitoring, out-of-pocket cost and availability influence use in India. Therefore, a staged pathway--MMF for inflammatory/active disease, nintedanib for progressive fibrotic phenotype, and combination therapy when risk justifies toxicity--is pragmatic. Pulmonary rehabilitation was advised to more than half of the cohort, reflecting the growing appreciation that non-pharmacological care, vaccination, reflux management, smoking cessation, nutrition and oxygen assessment are part of comprehensive SSc-ILD management [8].

This study also highlights service-delivery issues. A tertiary-care screening pathway requires coordination between medicine, pulmonology, radiology and cardiology. Access to DLCO testing remains a limiting factor in many district and regional hospitals. The high observed frequency of desaturation and echocardiographic pulmonary hypertension probability suggests that SSc-ILD clinics should include six-minute walk testing and echocardiography as routine adjuncts, particularly for patients with disproportionate dyspnoea or low DLCO. Such integration is important because SSc-related pulmonary vascular disease can coexist with ILD and worsens outcomes [9]. The study has limitations. It is single-centre, non-randomized and relatively small, with management decisions reflecting real-world physician judgment. Autoantibody testing and DLCO were not available in every patient, and follow-up was limited to the study period. Mortality and long-term treatment persistence could not be robustly estimated. Nevertheless, the dataset provides a useful regional snapshot and supports a reproducible screening algorithm: baseline HRCT and PFT at SSc diagnosis, risk stratification by extent and physiology, three- to six-month PFT monitoring for active or extensive disease, and multidisciplinary escalation using MMF, antifibrotic therapy, rescue immunomodulation and rehabilitation as clinically indicated.

Conclusion

In this prospective cohort of 65 systemic sclerosis patients, structured HRCT-PFT screening identified a high burden of ILD and provided actionable severity staging. Extensive and progressive ILD were associated with reduced FVC/DLCO, exercise desaturation and greater pulmonary hypertension probability. Contemporary management at this tertiary centre showed increasing use of MMF-based immunosuppression, selective antifibrotic escalation with nintedanib, rescue therapy for refractory disease and broader pulmonary rehabilitation. Early screening and multidisciplinary follow-up should be prioritized in regional Indian SSc care pathways.

References

1. Bautista-Sanchez R, et al. Systemic sclerosis-associated interstitial lung disease. PubMed. 2024; PMID:39439972.
2. Denton CP, et al. The 2024 British Society for Rheumatology guideline for management of systemic sclerosis. Rheumatology. 2024;63(11):2956-3021.
3. American College of Rheumatology. ACR introduces new guidelines to screen, monitor and treat interstitial lung disease in patients with rheumatic conditions. 2023.
4. Fairley JL, et al. Systemic sclerosis-associated interstitial lung disease: screening, progression and monitoring. 2024.
5. Tashkin DP, et al. Mycophenolate mofetil versus oral cyclophosphamide in scleroderma-related interstitial lung disease (Scleroderma Lung Study II). Lancet Respir Med. 2016;4:708-719.

6. Distler O, et al. Nintedanib for systemic sclerosis-associated interstitial lung disease. N Engl J Med. 2019; 380:2518-2528.
7. Raghu G, Montesi SB, Silver RM, et al. Treatment of systemic sclerosis-associated interstitial lung disease: an official ATS clinical practice guideline. Am J Respir Crit Care Med/Ann Am Thorac Soc. 2024.
8. Del Galdo F, Lescoat A, Conaghan PG, et al. EULAR recommendations for the treatment of systemic sclerosis: 2023 update. Ann Rheum Dis. 2025;84(1):29-40. doi:10.1136/ard-2024-226430.
9. Parodis I, et al. EULAR recommendations for non-pharmacological management of systemic lupus erythematosus and systemic sclerosis. Ann Rheum Dis. 2024; 83:720-729.
10. Hoffmann-Vold AM, et al. Assessment of recent evidence for the management of systemic sclerosis-associated interstitial lung disease. ERJ Open Res. 2021; 7:00235-2020.
11. Khanna D, et al. Safety and efficacy of tocilizumab in early systemic sclerosis-associated ILD: focused trial analyses. Lancet Respir Med. 2020; 8:963-974.
12. Roofeh D, et al. Systemic sclerosis-associated interstitial lung disease: screening and management concepts. Curr Opin Rheumatol. 2019; 31:241-249.