ECRONICON OPEN ACCESS

EC PULMONOLOGY AND RESPIRATORY MEDICINE

Research Article

Profile of Patients with CTD-ILD Presenting to a Tertiary Care Hospital in South India

Shona Arlin Christopher[1], Barney Isaac[1], Balamugesh Thangakunam[1], Devasahayam J Christopher[1]* and Leena Robinson Vimala[2]

[1]*Department of Pulmonary Medicine, Christian Medical College, Vellore, India*

[2]*Department of Radiology, Christian Medical College, Vellore, India*

***Corresponding Author:** Devasahayam J Christopher, Department of Pulmonary Medicine, Christian Medical College, Vellore, India.

Received: May 08, 2025; **Published:** June 23, 2025

Abstract

Background: Connective tissue diseases associated Interstitial lung disease (CTD-ILD) is commonly observed in systemic sclerosis, polymyositis/dermatomyositis and rheumatoid arthritis patients and is a leading cause of morbidity and mortality. Very few studies have examined the demographics, clinico-radiologic and lung function features and the prevalence of various ILD patterns in patients with different connective tissue diseases (CTD).

Methods: In this prospective study, we recruited patients who were diagnosed with CTD-ILD in tertiary care teaching hospital in southern India, between January 2019 and March 2020. All clinical and laboratory data (including serology, radiology and pulmonary function tests) were obtained.

Results: A total of 138 patients with CTD-ILD were recruited, 73.9% of whom were females. The mean age was 50.4 (SD- 11.8) yrs. Most common symptoms were cough (88.4%) and breathlessness (85.5%). A total of 73.9% had joint pain, 95.1% of which was symmetrical, 20.9% had skin lesions characteristic of their CTD, 27.3 had finger clubbing and 26.8% had velcro crackles. The mean FVC and TLC were 1.64L (57.8%) and 2.70 (60.8%) and DLCO was 46.97% of predicted. The mean 6-minute walk distance was 349.3 m. Among patients with CTD-ILD, the most common CTD was UCTD (34.1%), followed by rheumatoid arthritis (30.4%). The common ILD patterns on HRCT were; NSIP (62.3%), UIP (23.9%) and OP (4.3%).

Conclusion: We have reported herewith the clinical and laboratory profile of the largest single centre Indian case series on CTD-ILD.

Keywords: *Interstitial Lung Disease (ILD); Connective Tissue Disease (CTD); Pulmonary Function Tests; Autoantibodies; Pulmonary Hypertension*

Introduction

Connective tissue diseases (CTDs) are a group of systemic autoimmune disorders and are associated with autoimmune-mediated organ damage. The lung is a frequent target organ in autoimmune disorders, although any component of the respiratory system could be involved [1]. Interstitial lung disease (ILD), a group of diffuse parenchymal lung injury patterns characterized by varying degrees of inflammation and fibrosis, is a common manifestation of CTD, particularly in rheumatoid arthritis, systemic sclerosis and polymyositis/dermatomyositis [2-4]. It is also a leading cause of morbidity and mortality in these patients. The lung injury pattern of CTD-associated

ILD (CTD-ILD) mimics that of idiopathic interstitial pneumonia and may manifest either during the initial presentation of the CTD or during the course of the disease or prior to the manifestation of CTD elsewhere [5]. There is a dearth of data on CTD-ILD in India. Here, we report our series of patients with CTD-ILD.

Materials and Methods

This study is part of a prospective ILD registry, which was approved by the Institutional review board (IRB Min. No. 11661 dated 28.11.2018), and recruited consecutive patients with ILD attending the pulmonary medicine outpatient clinic at the Christian Medical College, Vellore, a tertiary care hospital in southern India between January 2019 and March 2020. The hospital is located in the state of Tamil Nadu and draws a large number of referred patients from the rest of the country and the adjacent countries, particularly Bangladesh.

Study design

This study is a cross-sectional observational study aimed at evaluating clinical, laboratory and lung function characteristics in patients with CTD-related ILD.

Study population

Inclusion criteria:

- Consecutive adult patients (≥18 years) diagnosed with a connective tissue disease (e.g., rheumatoid arthritis, systemic sclerosis, dermatomyositis, etc.), who presented to the pulmonary medicine outpatient clinic.
- Features of ILD on high-resolution computed tomography (HRCT).
- Diagnosis of CTD-ILD by the MDT discussions.

Exclusion criteria:

- Other causes of ILD (e.g. infections, drug-induced lung disease, environmental exposures).
- Patients with insufficient data for analysis (e.g. incomplete clinical records).
- Unwilling to participate in the study.

Recruitment process

Patients were recruited during routine outpatient clinic visits. Informed consent was obtained from all participants before data collection. All the clinical, laboratory and lung function parameters were obtained at first contact with the patient.

The diagnosis of various connective tissue diseases (CTDs) was made by a rheumatologist based on the most current diagnostic criteria for each condition.

1. Rheumatoid Arthritis- ACR and EULAR classification criteria 2010 [6].
2. Systemic Lupus Erythematosus- ACR and EULAR classification criteria 2019 [7].
3. Sjogren's syndrome - ACR and EULAR classification criteria 2016 [8].
4. Systemic sclerosis- ACR and EULAR classification criteria 2013 [9].
5. Dermatomyositis/polymyositis - ACR and EULAR classification criteria 2017 [10].
6. MCTD - Alarcon Segovia criteria 1987 [19].
7. UCTD - Diagnostic criteria proposed on 1999 [11].

Data collection

Clinical data:

- Patient demographics (age, sex, duration of CTD etc.).
- Symptoms (e.g. cough, dyspnoea, fatigue).
- History of smoking and occupational exposures.
- Detailed clinical examination findings.

Laboratory data:

- Routine blood tests (CBC, liver and kidney function tests).
- Autoantibody panel (ANA, anti-SCL-70, anti-RNP etc.).
- Other relevant tests (e.g. CRP, ESR).

Lung function tests:

- Spirometry: FVC, FEV1, FEV1/FVC ratio.
- Lung volumes and diffusion capacity: TLC and DLCO measurements.

Imaging

HRCT chest scans to classify patterns of ILD (e.g., ground-glass opacities, reticular patterns etc.).

MDT meeting

All patients who were diagnosed with CTD-ILD based on the accepted criteria at the multidisciplinary team (MDT) meeting, which included senior pulmonologists and thoracic radiologists. Other specialists were invited to participate as needed.

Data analysis

The data was entered into Epidata version 3.1 and subsequently extracted into Microsoft Excel. The clinical, laboratory and pulmonary function variables were presented as the mean and standard deviation or median and interquartile range when 'continuous' and, frequency and percentages when 'categorical'.

Results

During the study period, 138 patients with CTD-ILD were recruited. The mean age of the cohort was 50.4 +/-11.8 years, and 102 (73.9%) of them were females. The highest proportion of the patients recruited were from the state of West Bengal (27.5%), followed by Tamil Nadu (15.9%).

Symptoms

The most common symptoms reported were cough and breathlessness in 88.4% and 85.5% of the patients, respectively. The mean (SD) score on the visual analogue scale (VAS) for cough severity was 47.5 (24.7%). Among those who experienced breathlessness, the severity of breathlessness was mMRC grade 2 for 45.4%. Constitutional symptoms such as fever, loss of weight and loss of appetite were present in 42.8%, 45.7% and 42.8% of patients, respectively.

The symptoms, including CTD symptoms and various physical findings are enumerated in table 1.

A. General and Respiratory symptoms		
S no.	**Symptom (N = 138)**	**N (%)**
1.	Cough VAS score (mean and standard deviation)	122 (88.4) 47.5 (24.7)
2.	Breathlessness	118 (85.5)
	mMRC grade (n = 118) • mMRC grade 0 • mMRC grade 1 • mMRC grade 2 • mMRC grade 3 • mMRC grade 4	 3 (2.5) 36 (30.2) 54 (45.4) 22 (18.5) 4 (3.4)
3.	Fever	59 (42.8)
4.	Loss of appetite	59 (42.8)
5.	Loss of weight	63 (45.7)
6.	Fatigue	62 (44.6)
B. CTD related symptoms (N = 138)		
1.	Joint pain • Arthralgia • Arthritis • Monoarticular • Oligoarticular • Polyarticular • Symmetrical • Asymmetrical	102 (73.9) 32 (31.4) 70 (68.6) 2 (1.96) 10 (9.8) 90 (88.2) 97 (95.1) 5 (4.9)
2.	Oral Ulcers	26 (18.8)
3.	Skin lesions	29 (20.9)
4.	Photosensitivity	11 (7.9)
5.	Alopecia	37 (26.6)
6.	Skin tightness	16 (11.5)
7.	Dysphagia	17 (12.2)
8.	Raynaud's phenomenon	24 (17.3)
9.	Gastro-esophageal reflux symptoms	46 (33.1)

C. General physical examination (N = 138)		
1.	Clubbing Grades of clubbing (n=38) • Grade 1 • Grade 2 • Grade 3 • Grade 4	38 (27.3) 11 (29) 8 (21) 18 (47.4) 1 (2.6)
2.	Cyanosis	5 (3.62)
3.	Adventitious lung sounds • Velcro crackles • Wheeze • Squeaks • Bronchial breath sounds	 37 (26.8) 13 (9.4) 2 (1.5) 1 (1.7)
4.	Loud P2	5 (3.6)
5.	Pedal oedema	10 (7.2)
6.	Elevated JVP	4 (2.9)
D. CTD related physical examinations findings		
1.	Oral ulcers	10 (7.3)
2.	Hand deformities	21 (15.2)
3.	Skin tightness	18 (13)
4.	Skin lesions	6 (4.4)

Table 1: *Symptomatology and physical findings of the study subjects.*

The most common symptoms specifically related to CTD were joint pain, followed by gastro-esophageal reflux symptoms and alopecia (73.9%, 33.1% and 26.6%, respectively). Among the patients who reported joint pain, 68.6% had arthritis, and the majority had symmetrical and poly-articular joint involvement (95.1% and 88.2%, respectively).

Among the 138 patients, 38 (27.3%) had clubbing, and more people had grade 3 clubbing (47.4%), compared to other grades. On auscultation, velcro crackles were heard in 26.8% of patients. Physical examination of the cardiovascular system revealed the following features of pulmonary hypertension: loud P2 (3.6%) and JVP elevation (2.9%). The two most common CTD-attributable physical findings were hand deformities (15.2%) and skin tightness (13%).

Pulmonary function and 6-minute walk test

The pulmonary function tests and 6-minute walk test exercise physiology data are tabulated in table 2.

S. no.	Variables	Mean	Standard deviation
1.	FEV1 in Litres (% predicted)	1.36 (58.1)	0.5 (17.6)
2.	FVC in Litres (% predicted)	1.64 (57.8)	0.6 (16.5)
3.	FEV1/FVC	82.76	7.5
4.	TLC in Litres (% predicted)	2.70 (60.8)	0.8 (13.4)
5.	DLCOc % predicted	46.97	24.8
6.	DLCOc/VA % predicted	99	35.6
7.	RV/TLC% predicted	104	33.8
8.	6 min walk distance in metres	349.3	95.7
9.	6 min walk distance % predicted	60	15.7
10.	Distance saturation product in meter %	316.5	98.1

Table 2: *Pulmonary function test and 6-minute walk test results.*

The mean FVC in the population was 1.64 (SD-0.6) litres and it was 57.8 (SD-16.5) % of the predicted FVC. The mean TLC was 2.70 (SD-0.8) litres it was 60.8 (SD-13.4) % of the predicted TLC. The mean DLCO percentage of predicted was 46.9 (SD-24.8) %. The mean 6 min walk distance was 349.3 (SD-95.7) m, and the mean distance saturation product was 316.5 (SD-98) m%. In all, 12.6% of the patients had features of pulmonary hypertension as diagnosed by characteristic echocardiogram features or dilated MPA on CT of the thorax. None of these patients were subjected to right heart catheterisation for confirmation.

CTDs and serology

In our CTD-ILD cohort, most patients were diagnosed with an underlying diagnosis of undifferentiated connective tissue disorder (UCTD), followed by rheumatoid arthritis and systemic sclerosis (34.1%, 30.4% and 12.3%, respectively). Nine patients in our study were classified as having CTD other than major connective tissue disorders. There were 3 patients with overlap syndrome: SLE-systemic sclerosis overlap, rheumatoid arthritis-systemic sclerosis overlap and myositis-scleroderma overlap. There were 5 patients with anti-synthetase syndrome and 1 patient with spondyloarthritis.

The auto-antibody profile of the study patients with various connective tissue disorders is shown in table 3.

Connective tissue disorders	Rheumatoid arthritis (N = 42)	SLE (N = 6)	Sjogren's syndrome (N = 3)	Systemic sclerosis (N = 17)	DM/PM (N = 4)	MCTD (N = 10)
Serological tests	**No. (%)**	**No. (%)**	**No. (%)**	**No. (%)**	**No. (%)**	**No. (%)**
ANA	17/36 (47.2)	5/6 (83.3)	3/3 (100)	14/15 (93.3)	3/4 (75)	10/10 (100)
• Speckled	7/17 (41.2)	3/5 (60)	3/3 (100)	9/14 (60)	0	9/10 (90)
• Homogenous	7/17 (41.2)	2/5 (40)	0	1/14 (6.7)	3/3 (100)	1/10 (10)
• Nucleolar	1/17 (5.8)	0/5	0	3/14 (13.3)	0	0
• Centromere	2/17 (11.8)	0/5	0	0/14	0	0
Rheumatoid factor	32/39 (82.1)	1/4 (25)	2/3 (66.6)	5/11 (45.5)	1/4 (25)	4/8 (50)
Anti CCP	28/35 (80)	1/3 (33.3)	0	1/1 (100)	0/1	1/1 (100)
Anti Jo 1	-	0/1	-	1/4 (25)	1/1 (100)	0/3
Anti - SSA	1/8 (12.5)	1/4 (25)	3/3 (100)	0/5	0/3	3/7 (42.9)
Anti- SSB	0/6	0/1	1/2 (50)	0/4	0/2	0/4

SCL- 70	0/4	0/1	0/1	13/14 (92.9)	0/2	2/5 (40)
ds-DNA	0/3	2/4 (50)	0/1	0/2	0/1	1/3 (33.3)
CENP	0/3	0/1	-	1/5 (20)	0/1	0/5
U1RNP	0/7	2/4 (50)	1/2 (50)	1/5 (20)	0/2	9/10 (90)
C3	0/1	0/5	0/2	0/2	-	1/3 (33.3)
C4	0/1	0/5	0/2	0/2	-	2/3 (66.7)
Myositis profile	1/1 (100)	-	1/1 (100)	2/3 (66.7)	3/3 (100)	1/1 (100)

Table 3: *Underlying connective tissue disorders and their auto-antibody profile.*

DM/PM: Dermatomyositis/Polymyositis; MCTD: Mixed Connective Tissue Disorder; SLE: Systemic Lupus Erythematosus.

Among patients with rheumatoid arthritis, 82.1% had rheumatoid factor positivity, and 80% had anti-CCP positivity. Among those diagnosed with SLE, 83.3% showed ANA positivity, with speckling (60%) and homogenous (40%) pattern, and 50% showed the presence of ds-DNA antibodies.

All patients with Sjogren's syndrome showed ANA positivity (speckled pattern) and anti-SSA antibody positivity. Only 50% of the patients were positive for anti-SSB.

Among those diagnosed with systemic sclerosis, 93.3% had ANA positivity, 92.9% had Scl-70 positivity, among those diagnosed with dermatomyositis/polymyositis, 75% and 100% had ANA and myositis profile positivity, respectively and among those with mixed connective tissue disorder, 100% had ANA positivity and 90% anti-U1RNP positivity.

Radiological patterns in various CTDs

The ILD patterns of various CTDs are tabulated in table 4. Overall, 42 (30.4%) patients had rheumatoid arthritis. The majority of our patients with RA had a nonspecific interstitial pneumonitis (NSIP) pattern of ILD, followed by a usual interstitial pneumonitis (UIP) pattern (57.1% and 35.7%, respectively). This is contrary to the majority of the literature on RA-related ILD, where UIP is the most common ILD pattern. (4,12). Among the RA-ILD with NSIP pattern, 75% were of the fibrotic subtype and 25% of the cellular sub-type. There were 6 (4.4%) patients in the cohort with systemic lupus erythematosus, of whom 66.7% had an NSIP pattern, 16.7% had a UIP pattern and 16.6% had an OP (organizing pneumonia) pattern.

There were 3 (2.2%) patients with Sjogren's syndrome, one each with NSIP, LIP (lymphoid interstitial pneumonia) and NSIP-OP overlap patterns. There were 17 (12.3%) patients with Systemic sclerosis, 76.5% had NSIP pattern, and the remaining 23.5% had UIP pattern. Of the 4 (2.9%) patients who were diagnosed with dermatomyositis/polymyositis, 50% had an NSIP pattern, 25% UIP pattern and remaining 25% had NSIP/OP overlap pattern.

In all, 10 (7.2%) patients had mixed connective tissue disorder (MCTD), 60% had NSIP pattern, 20% had UIP pattern, and 10% each had LIP and NSIP-OP overlap patterns.

In our cohort, the majority (47; 34.1%) of the patients with CTD-ILD were classified as having undifferentiated connective tissue disorder (UCTD), 63.8% had NSIP, 17% UIP and 12.8% NSIP/OP overlap.

Previous studies have shown that there are a few specific signs that are predominantly observed in patients with UIP pattern of ILD, which may indicate the presence of underlying CTD [13]. They are:

1. Anterior upper lobe sign
2. Exuberant honey combing sign
3. Straight edge sign.

We performed a subgroup analysis on patients with a UIP pattern of ILD. A senior chest radiologist assessed the presence of these signs in this group.

Of the 33 patients with UIP pattern of ILD, 48.5% had anterior upper lobe sign, and 27.3% each had exuberant honey combing sign and straight edge sign. Only 2 patients had all three signs, 27% had at least 2 signs, and 36.4% did not have any of these signs.

Discussion

In this study, we recruited consecutive patients with CTD-ILD who presented to the Department of Pulmonary Medicine during the study period, and we endeavoured to study the clinical, pulmonary function, radiological and serological picture of the target population.

In our cohort of 138 patients the mean age was 50.5years, and 73.9% of them were females. In a study performed in Portugal [14], a total of 75 CTD-ILD patients were studied over a 6-year period, whose mean age was 56±15.5 years and they reported a female predominance. In the series on Canadian patients, published by Chan., *et al.* [15], 357 patients with CTD-ILD were studied, and the mean age of the population was 56 ± 13 years; again, the majority of the patients (73%) were female. It appears that our patients were slightly younger than the Caucasian population, while the female predominance was similar. In a study done in the Indian population across 12 rheumatology centres across India by Santhanam., *et al*. they recruited 620 patients of whom 505 patients were females (81.4%) [16]. Our study showed that cough and breathlessness were the predominant symptoms (88.4% and 85.5%, respectively); however, in a similar study performed by Agarwal., *et al.* [1] in Western India, these symptoms were present in a greater proportion of patients-breathlessness (97%) and cough (95%). The CTD-specific common symptoms reported in our study were, arthralgia - 23.1% and arthritis - 50.7%; however, Agarwal., *et al.* [1] reported arthralgia in 61% and arthritis in 34%.

We found velcro crackles in 26.8% of our subjects, but this was present in a much higher proportion (69%) in Agarwal., *et al.*'s series [1]. This difference is possibly due to the greater number of patients with the UIP pattern (69%) in their cohort [1]. CTD-specific signs were also different in our study compared with Agarwal., *et al.* Joint deformities, CTD-specific skin lesions and oral ulcers were detected in 15.2%, 17.3% and 7.3% of patients in our study and in 1%, 27% and 18% of patients, respectively, in Agarwal., *et al.* [1].

The mean FVC, DLCOc and 6 min walk distance in our population were 57.8%, 46.9% and 349 m, respectively. In the study by Chan., *et al.* [15], these values were higher -77%, 56% and 387m respectively. The reason for worse lung involvement in our cohort could be that they were recruited from the pulmonary department, whereas Chan., *et al.*'s [15] cohort was recruited from a rheumatology clinic. In Santhanam., *et al*.'s cohort the mean FVC was 61.9%, which was similar to ours [16].

Among the patients included in our study, 12.6% had pulmonary hypertension, either diagnosed by echocardiogram or CT of the thorax (showing dilated MPA). This was similar to the cohort of Oliveria., *et al.* [14] in which 12% of patients were diagnosed with pulmonary hypertension, either by echocardiogram or right heart catheterization. A significantly higher proportion (35.8%) in Santhanam., *et al.*'s cohort were reported to have pulmonary hypertension as assessed by echocardiogram [16].

The proportion of various CTDs in our CTD-ILD population was as follows: Rheumatoid arthritis- 30.4%, Systemic sclerosis-12.3%, Sjogren's syndrome- 2.2%, SLE - 4.3%, PM/DM-2.9% and MCTD-7.2%. In Oliveria., *et al.*'s [14] cohort the distribution was as follows: Rheumatoid arthritis- 20%, Systemic sclerosis- 34.7%, Sjogren's syndrome- 2.7%, SLE - 0%, autoimmune myopathy - 6.7% and MCTD -

9.3%. However, in our cohort, a large number of patients were classified as having undifferentiated connective tissue disease (34.1%), but in theirs, the number was much smaller (5.3%). In Santhanam., *et al.*'s cohort, the distribution was: Rheumatoid arthritis - 40.3%, Systemic sclerosis - 25.7% and Mixed connective tissue disease - 7.3% [16].

The autoantibody profiles of our cohort were compared with that of Oliveria., *et al.* [14]. Among the patients who were diagnosed with RA, 82.1% were rheumatoid factor positive and 80% were anti-CCP positive in our study, which is marginally higher than the percentages reported by Oliveria., *et al.* [14], 73.3% positivity for both. Similarly, among those who were diagnosed with systemic sclerosis, the percentage of Scl 70 positivity was much higher in our cohort (92.9% vs 54%) compared to Oliveria., *et al.* All patients in Oliveria., *et al's* cohort [14] who were diagnosed with MCTD showed anti-RNP positivity, as was also the case in our cohort.

CTD-ILD can present with various radiologic patterns of ILD, such as NSIP, UIP, LIP, OP etc. We compared the patterns of ILD in our cohorts with those reported by Agarwal., *et al.* [1]. In our study, the majority of patients with RA had an NSIP pattern of ILD (57.1%), followed by a UIP pattern (35.7%), in contrast to Agarwal., *et al.* [1] where UIP was the most common ILD pattern (38.5%), followed by NSIP (30.8%). However, we found 3 other studies in which NSIP was the predominant pattern, similar to our study [16-18]. In the RA patients in Santhanam., *et al.*'s cohort also, the NSIP pattern (49.6%) was slightly higher than the UIP pattern (46.3%) [16]. In the Japanese cohort reported by Mori., *et al.* [17], the higher prevalence of the NSIP compared to the UIP pattern was explained by the prevalence of ground glass attenuation in their population. In another cohort from Japan reported by Tanaka., *et al.* the higher prevalence of NSIP compared to UIP was believed to be due to the possibility of RA overlapping with other CTDs or transformation to other CTDS over time [18]. Since 2 studies from Japan and our study from India reported higher prevalence of NSIP compared to UIP in the RA patients, we wonder whether the Asian population has a greater predilection for presentation with NSIP pattern.

Among those with systemic sclerosis, NSIP was more common than UIP in our series (76.5% and 23.5%, respectively). In the series of Agarwal., *et al.* [1], NSIP was the most common pattern (66.6%), followed by OP and UIP (19.1% and 14.3%, respectively). Among those with MCTD-ILD, the proportion of NSIP and UIP in our cohort was similar to theirs: NSIP (60% vs 52.6%) and UIP (20% vs 15.8%).

Chung., *et al.* [13] reported that the specific CT signs 'anterior upper lobe sign', 'exuberant honey combing sign' and 'straight edge sign' can help differentiate the CTD-related UIP pattern of ILD from that of idiopathic pulmonary fibrosis (IPF). In our study, among the patients with the UIP pattern of CTD-ILD, 48.5% had the 'anterior upper lobe sign', and 27.3% each had the 'exuberant honey combing sign' and 'straight edge sign'. In the cohort of Chung., *et al.* [13], a similar number of patients had the 'anterior upper lobe sign' (36%) and 'straight edge sign' (30%); however, the 'exuberant honey combing sign' was observed in fewer patients (2%).

Limitation of the Study

Our patient population was mainly drawn from the states of Tamil Nadu, Andhra Pradesh and West Bengal, with a smaller proportion from other states, and is therefore not representative of the spectrum of CTD-ILD in the country. All clinical, physiological and radiological data were collected at the first visit to the department of Pulmonary Medicine, and variations over time were not captured.

Conclusion

In this study, we endeavoured to demonstrate the clinical, pulmonary function, radiological features and serological patterns of patients who presented with CTD-ILD. To our knowledge, this is the largest single-centre study of patients with CTD-related interstitial lung disease in India.

Funding Support

No funding was obtained for this project.

Competing Interests

None to declare.

Bibliography

1. Agarwal M., *et al*. "Clinico-radiological profile of connective tissue disease related-interstitial lung diseases from a tertiary care centre of India: a cross sectional study". *Monaldi Archives for Chest Disease* 91.4 (2021).

2. Schulte JJ and Husain AN. "Connective tissue disease related interstitial lung disease". *Surgical Pathology Clinics* 13.1 (2020): 165-188.

3. Antin-Ozerkis D., *et al*. "Interstitial lung disease in the connective tissue diseases". *Clinics in Chest Medicine* 33.1 (2012): 123-149.

4. Yoo H., *et al*. "Connective tissue disease-related interstitial lung disease (CTD-ILD) and interstitial lung abnormality (ILA): Evolving concept of CT findings, pathology and management". *European Journal of Radiology Open* 9 (2022): 100419.

5. Joshua J Solomon and Aryeh Fischer. "Connective Tissue Disease-Associated Interstitial Lung Disease: A Focused Review". *Journal of Intensive Care Medicine* 30.7 (2015): 392-400.

6. Aletaha D., *et al*. "2010 Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative". *Arthritis and Rheumatology* 62.9 (2010): 2569-2581.

7. Aringer M., *et al*. "2019 European league against rheumatism/American college of rheumatology classification criteria for systemic lupus erythematosus". *Arthritis and Rheumatology (Hoboken, N.J.)* 71.9 (2019): 1400-1412.

8. Shiboski CH., *et al*. "2016 American College of Rheumatology/European League Against Rheumatism Classification Criteria for Primary Sjögren's Syndrome: A Consensus and Data-Driven Methodology Involving Three International Patient Cohorts". *Arthritis and Rheumatology (Hoboken, N.J.)* 69.1 (2017): 35-45.

9. van den Hoogen F., *et al*. "2013 classification criteria for systemic sclerosis: an American college of rheumatology/European league against rheumatism collaborative initiative". *Annals of the Rheumatic Diseases* 72.11 (2013): 1747-1755.

10. Lundberg IE., *et al*. "EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups". *Annals of the Rheumatic Diseases* 76.12 (2017): 1955-1964.

11. Mosca M., *et al*. "Undifferentiated connective tissue diseases (UCTD): a review of the literature and a proposal for preliminary classification criteria". *Clinical and Experimental Rheumatology* 17.5 (1999): 615-620.

12. Assayag D., *et al*. "Rheumatoid arthritis associated interstitial lung disease: a review". *Medicina* 74.2 (2014): 158-165.

13. Chung JH., *et al*. "CT features of the usual interstitial pneumonia pattern: differentiating connective tissue disease-associated interstitial lung disease from idiopathic pulmonary fibrosis". *American Journal of Roentgenology* 210.2 (2018): 307-313.

14. Oliveira RP., *et al*. "Connective tissue disease-associated interstitial lung disease". *Pulmonology* (2020).

15. Chan C., *et al*. "Demographic and clinical predictors of progression and mortality in connective tissue disease-associated interstitial lung disease: a retrospective cohort study". *BMC Pulmonary Medicine* 19.1 (2019): 192.

16. Santhanam S., *et al*. "Connective tissue disease-associated interstitial lung disease - a multicenter observational study from India". *Indian Journal of Rheumatology* 16.1 (2021): S10.

17. Mori S., *et al*. "Comparison of pulmonary abnormalities on high-resolution computed tomography in patients with early versus longstanding rheumatoid arthritis". *Journal of Rheumatology* 35.8 (2008): 1513-1521.

18. Tanaka N., *et al*. "HRCT findings of collagen vascular disease-related interstitial pneumonia (CVD-IP): a comparative study among individual underlying diseases". *Clinical Radiology* 73.9 (2018): 833.e1-833.e10.

19. Alarcón-Segovia D and Villarreal-Alarcon Miguel. "Classification and diagnostic criteria for mixed connective tissue disease. Mixed connective tissue disease and anti-nuclear antibodies" (1987): 33-40.

Volume 14 Issue 7 July 2025