

Supplement to: Prevalence, imaging patterns, and risk factors of interstitial lung disease in connective tissue disease: a systematic review and meta-analysis

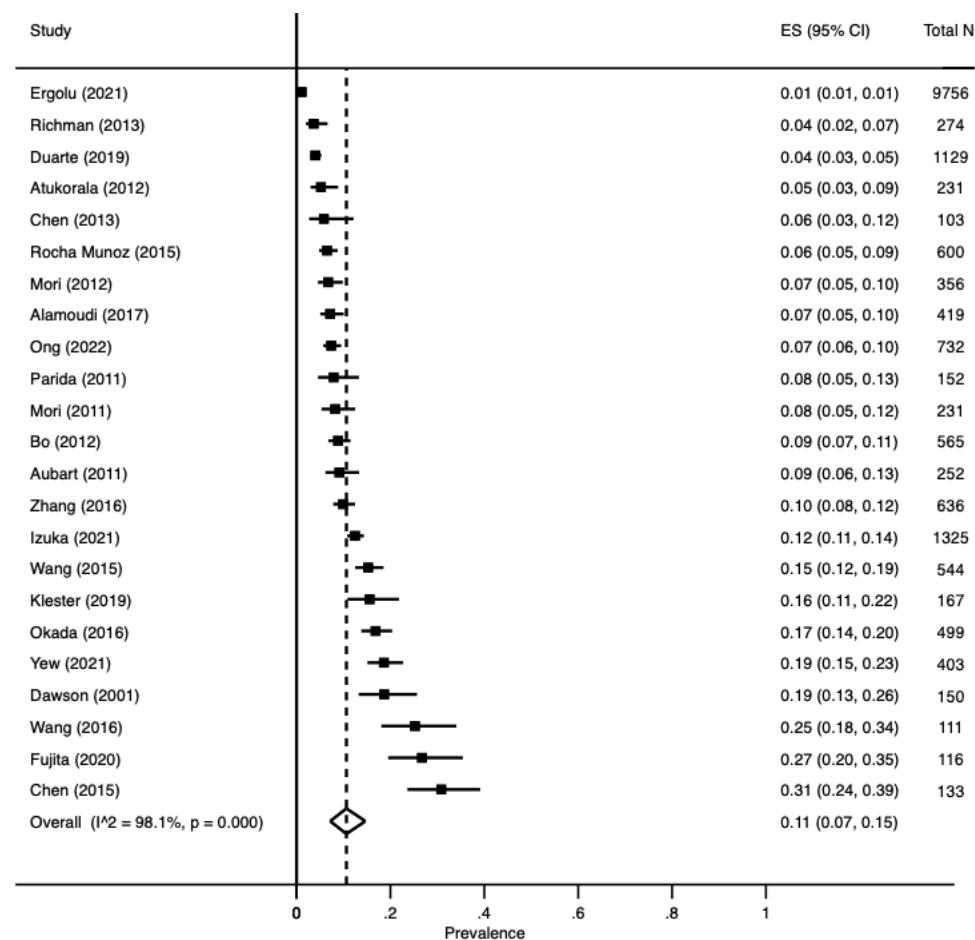
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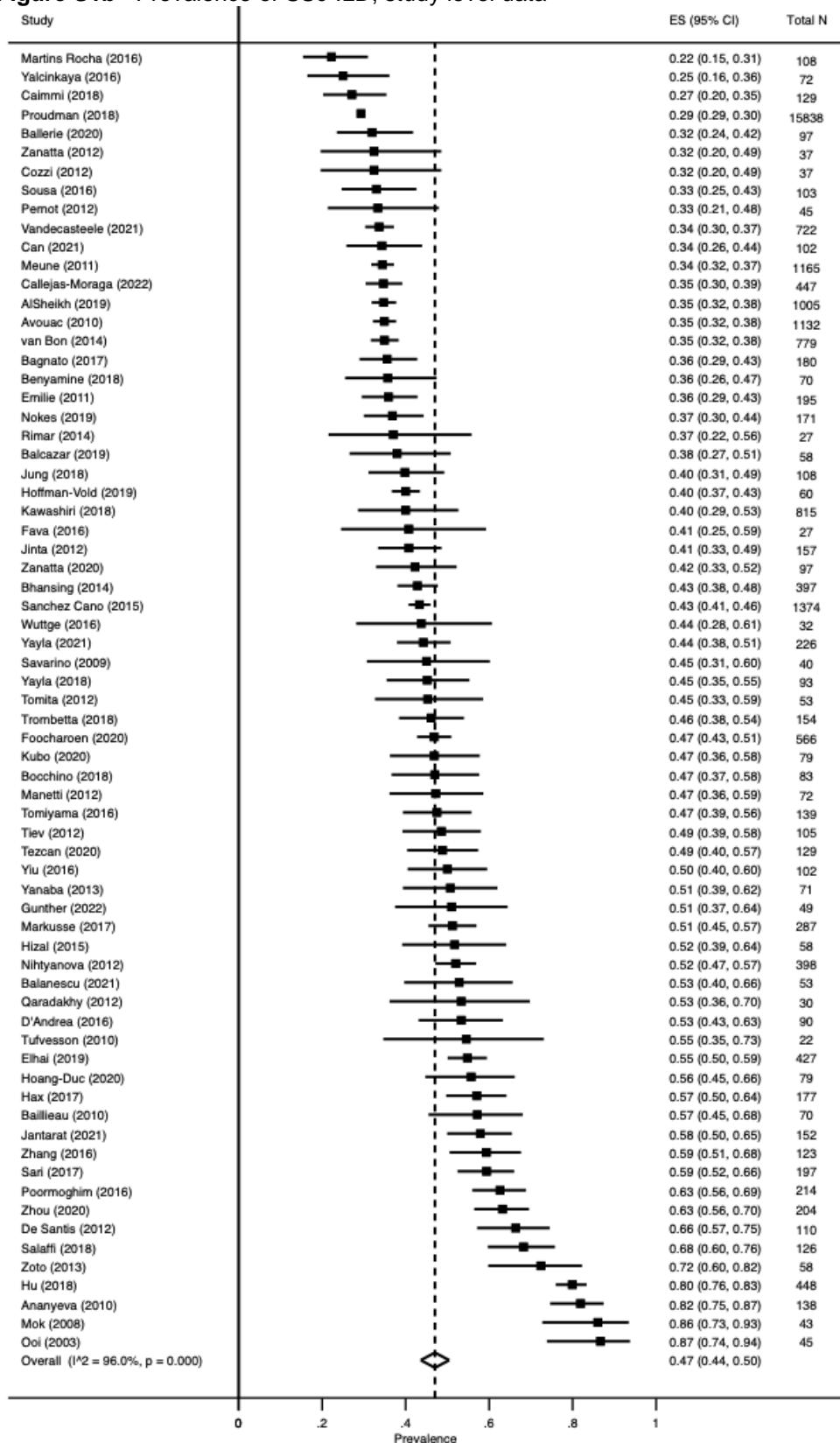
SUPPLEMENTAL FIGURES

Figure S1a Prevalence of RA-ILD, study level data



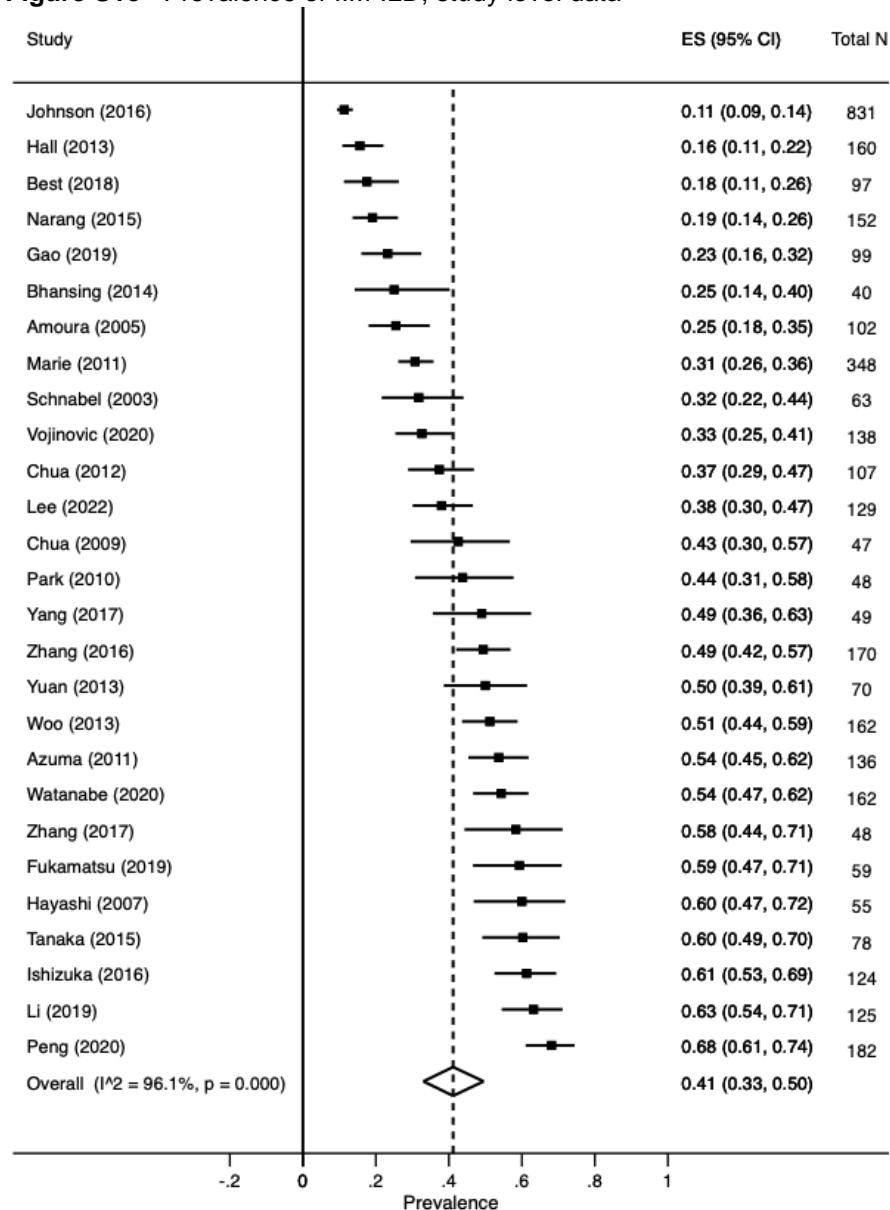
Abbreviations: ILD: interstitial lung disease, RA: rheumatoid arthritis

Figure S1b Prevalence of SSc-ILD, study level data



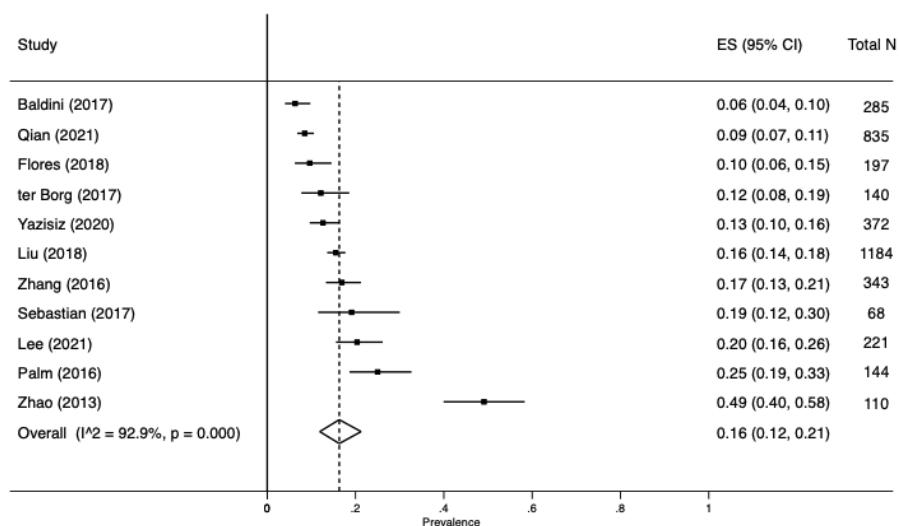
Abbreviations: ILD: interstitial lung disease, SSc: systemic sclerosis

Figure S1c Prevalence of IIM-ILD, study level data



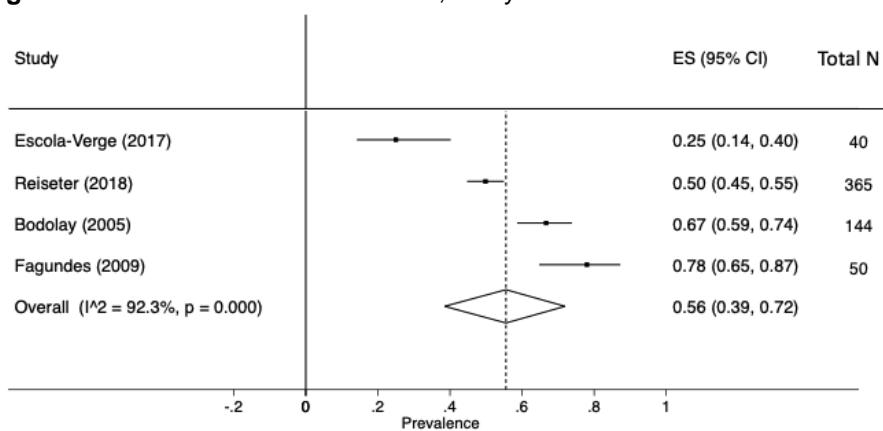
Abbreviations: IIM: idiopathic inflammatory myositis, ILD: interstitial lung disease

Figure S1d Prevalence of pSS-ILD, study level data



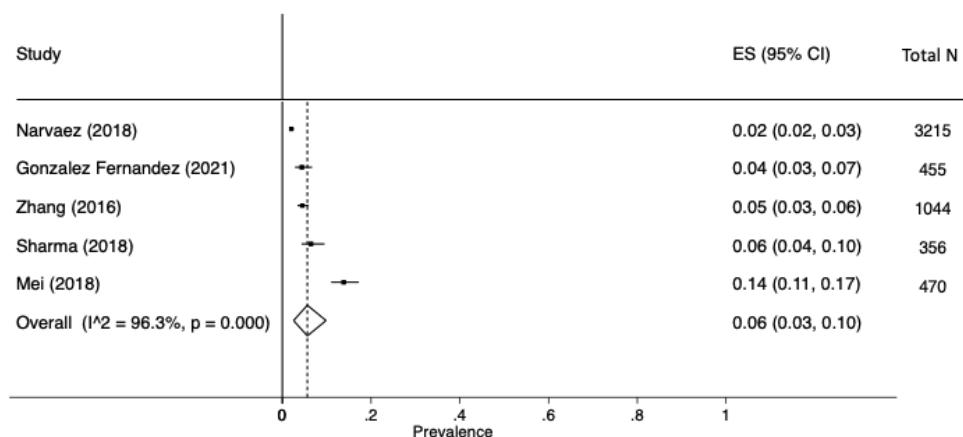
Abbreviations: ILD: interstitial lung disease, pSS: primary Sjögren syndrome

Figure S1e Prevalence of MCTD-ILD, study level data



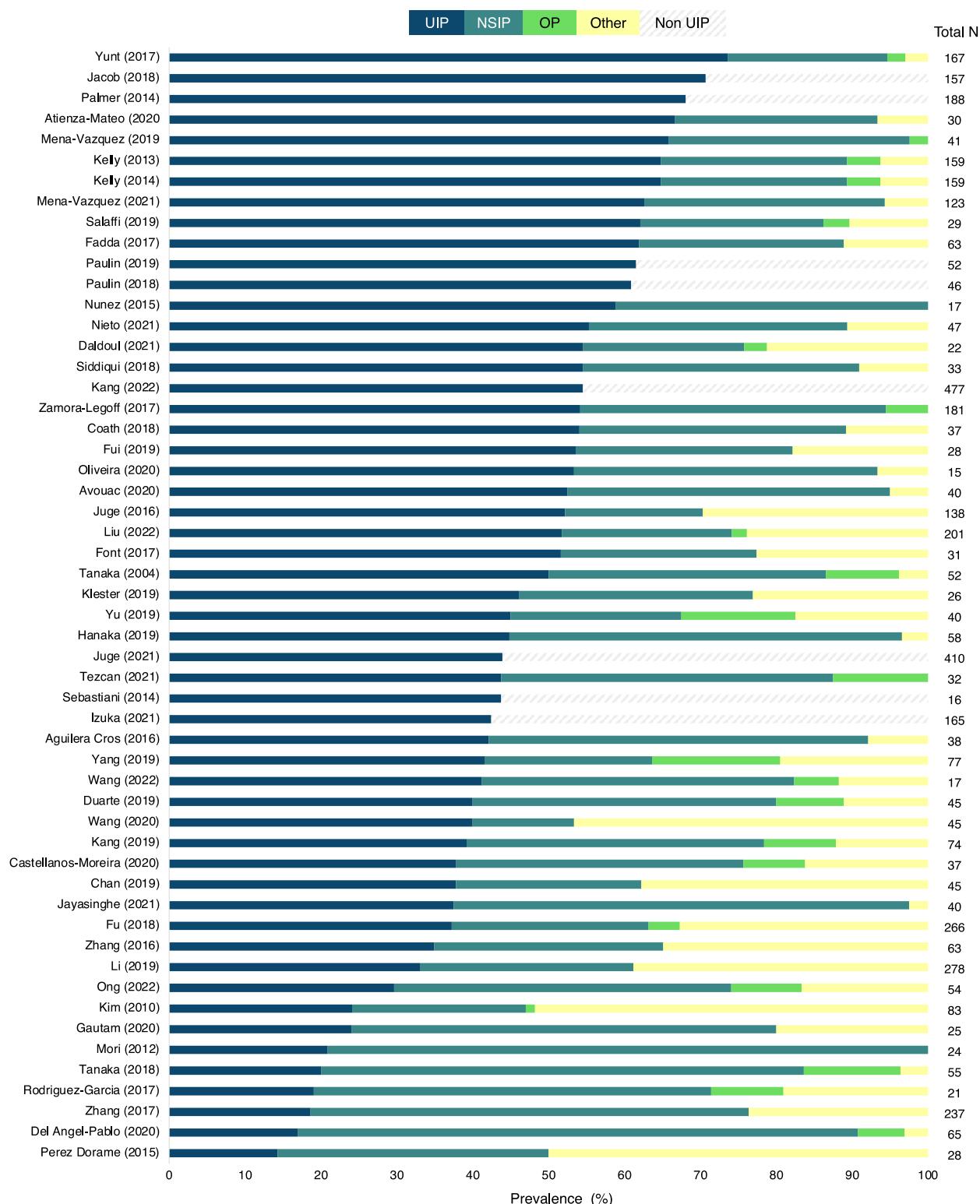
Abbreviations: ILD: interstitial lung disease, MCTD: mixed connective tissue disease

Figure S1f Prevalence of SLE-ILD, study level data



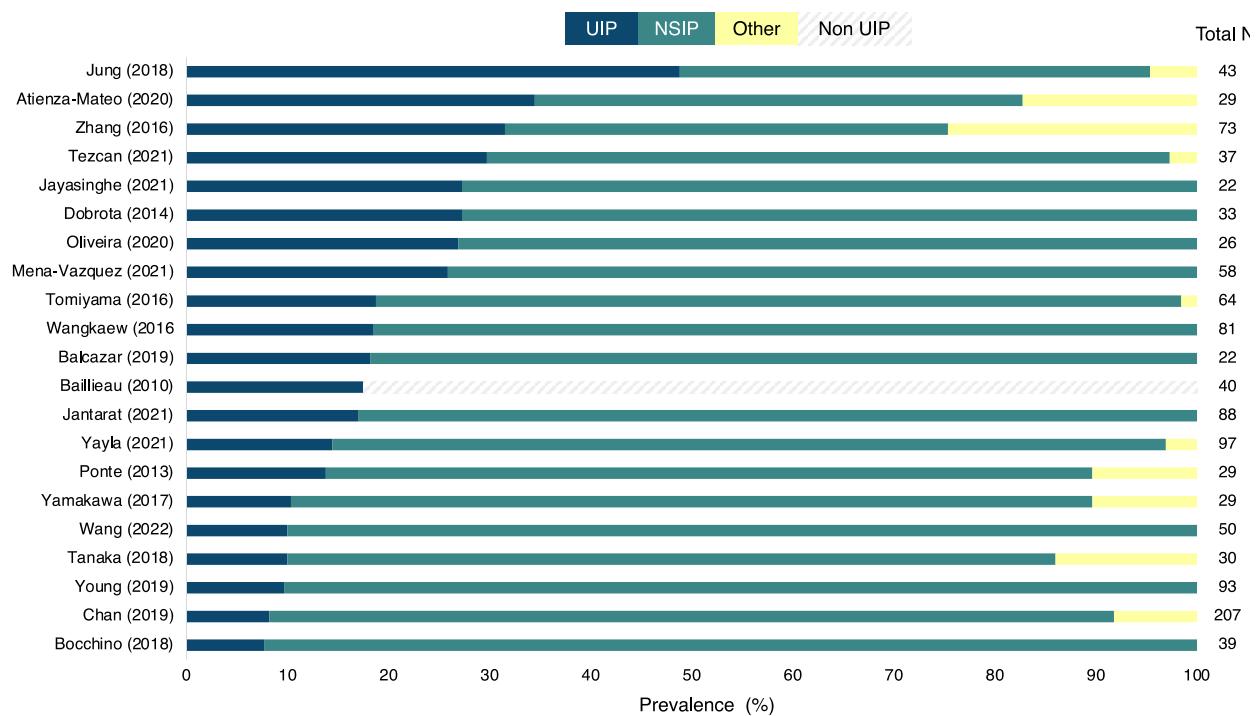
Abbreviations: ILD: interstitial lung disease, SLE: systemic lupus erythematosus

Figure S2a Study level data on RA-ILD CT patterns



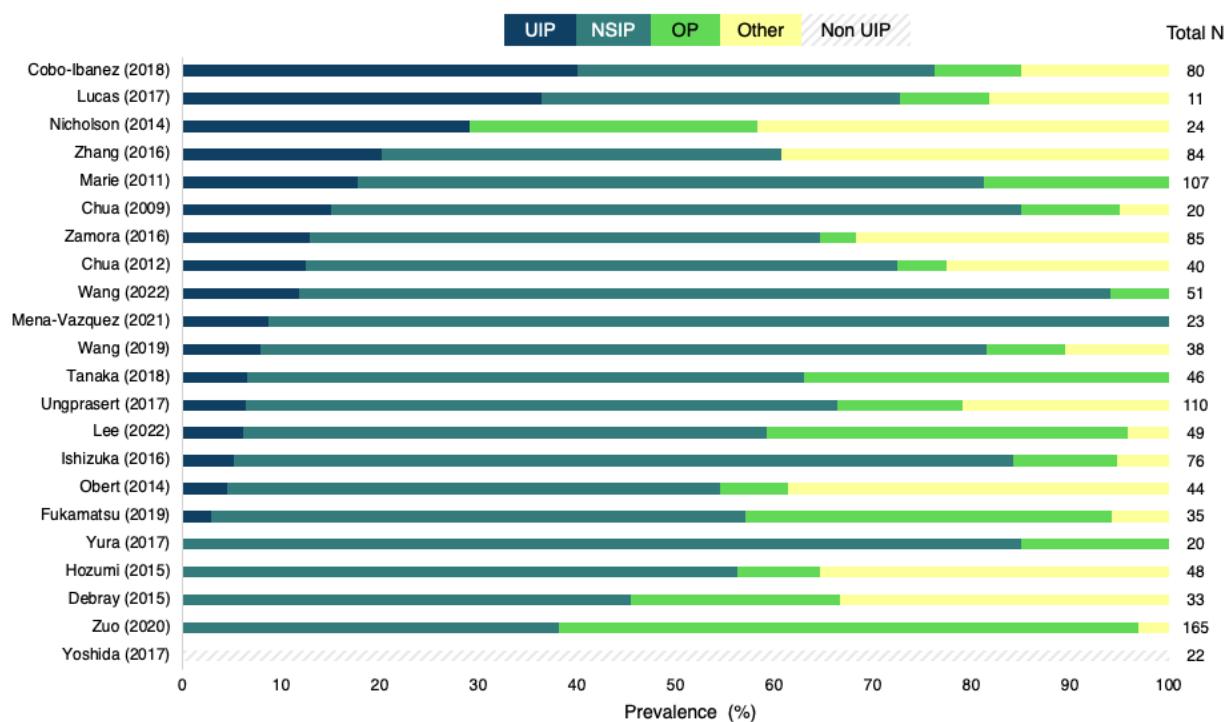
Abbreviations: CT: computed tomography, ILD: interstitial lung disease, NSIP: non-specific interstitial pneumonia, OP: organizing pneumonia, other: other and undifferentiated, RA: rheumatoid arthritis, UIP: usual interstitial pneumonia.

Figure S2b Study level data on SSc-ILD CT patterns



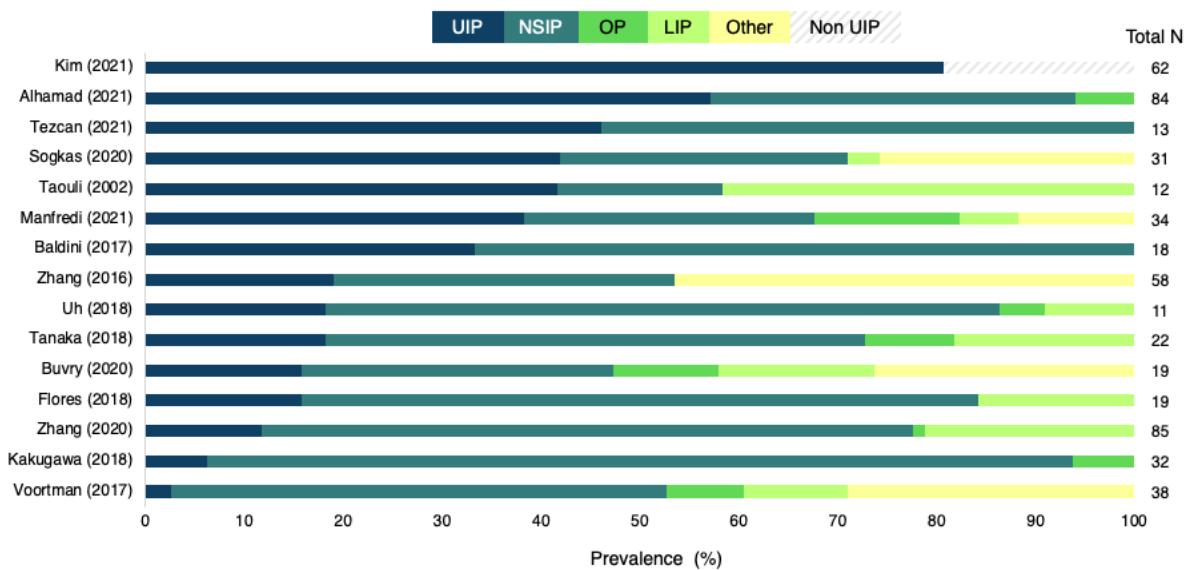
Abbreviations: CT: computed tomography, ILD: interstitial lung disease, NSIP: non-specific interstitial pneumonia, other: other and undifferentiated, SSc: systemic sclerosis, UIP: usual interstitial pneumonia.

Figure S2c Study level data on IIM-ILD CT patterns



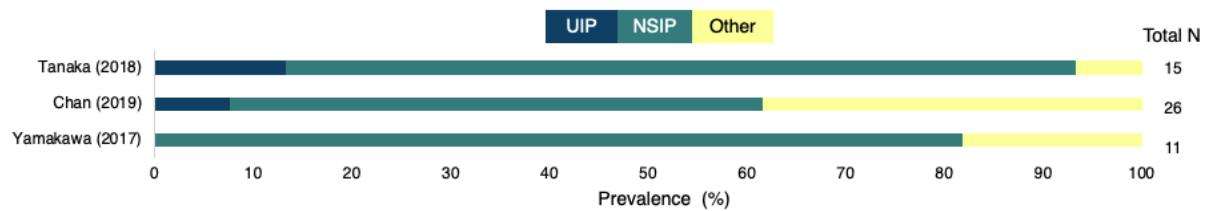
Abbreviations: CT: computed tomography, IIM: idiopathic inflammatory myositis, ILD: interstitial lung disease, NSIP: non-specific interstitial pneumonia, OP: organizing pneumonia, other: other and undifferentiated, UIP: usual interstitial pneumonia.

Figure S2d Study level data on pSS-ILD CT patterns



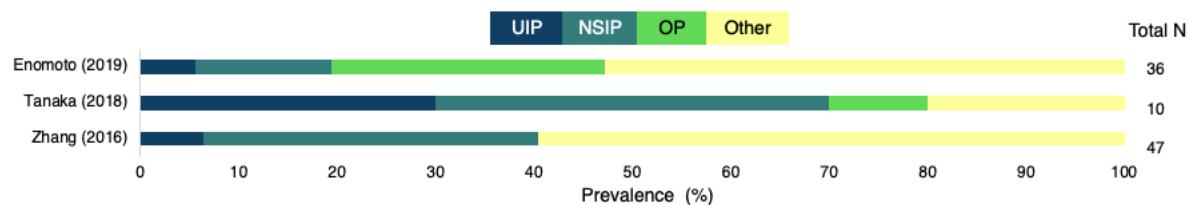
Abbreviations: CT: computed tomography, ILD: interstitial lung disease, LIP: lymphocytic interstitial pneumonia, NSIP: non-specific interstitial pneumonia, OP: organizing pneumonia, other: other and undifferentiated, pSS: primary Sjögren syndrome, UIP: usual interstitial pneumonia.

Figure S2e Study level data on MCTD-ILD CT patterns



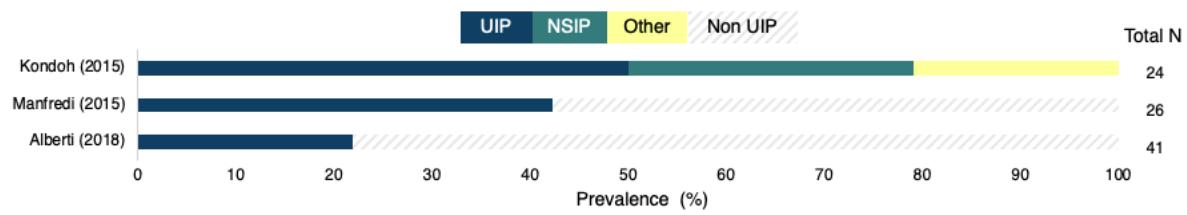
Abbreviations: CT: computed tomography, ILD: interstitial lung disease, MCTD: mixed connective tissue disease, NSIP: non-specific interstitial pneumonia, other: other and undifferentiated, UIP: usual interstitial pneumonia.

Figure S2f Study level data on SLE-ILD CT patterns



Abbreviations: CT: computed tomography, ILD: interstitial lung disease, NSIP: non-specific interstitial pneumonia, OP: organizing pneumonia, other: other and undifferentiated, SLE: systemic lupus erythematosus, UIP: usual interstitial pneumonia.

Figure S2f Study level data on UCTD-ILD CT patterns



Abbreviations: CT: computed tomography, ILD: interstitial lung disease, NSIP: non-specific interstitial pneumonia, other: other and undifferentiated, UCTD: undifferentiated connective tissue disease, UIP: usual interstitial pneumonia.

SUPPLEMENTAL TABLES

Table S1a Embase search strategy

1	Exp rheumatoid arthritis/
2	Rheumatoid.mp.
3	Exp scleroderma/
4	Scleroderma.mp.
5	"systemic sclero*".mp.
6	Exp Sjogren syndrome/
7	Sj#gren*.mp.
8	Myositis/
9	Antisynthetase syndrome/
10	Dermatomyositis/
11	Polymyositis/
12	Dermatomyositis.mp.
13	Polymyositis.mp.
14	"antisynthetase syndrome".mp.
15	"inflammatory myositis".mp.
16	Connective tissue disease/
17	"connective tissue dis*".mp.
18	"collagen vascular dis*".mp.
19	Exp mixed connective tissue disease/
20	"mixed ctd".mp.
21	"undifferentiated ctd".mp.
22	Exp systemic lupus erythematosus/
23	Lupus.mp.
24	or/1-23
25	Exp interstitial lung disease/
26	Exp lung fibrosis/
27	(interstitial\$ adj3 (lung\$ or pulmonary\$ or pneumon\$)).mp.
28	((pulmonary\$ or lung\$) adj3 (fibros\$ or fibrot\$)).mp.
29	ILD.mp.
30	UIP.mp.
31	NSIP.mp.
32	PPFE.mp.
33	"pleuroparenchymal fibroelastosis".mp.
34	Bronchiolitis.mp.
35	Alveolitis.mp.
36	"organ#ing pneumon*".mp.
37	or/25-36
38	Computer assisted tomography/ or computed tomographic angiography/ or high resolution computer tomography/ or multidetector computed tomography/ or spiral computer assisted tomography/ or x-ray computed tomography/
39	HRCT.mp.
40	"computed tomography".mp.
41	"ct-scan".mp.
42	Prevalence/
43	Prevalence.mp.
44	Risk factor/
45	"risk factor*".mp.
46	Biomarker*.mp
47	or/38-46
48	24 and 37 and 47
49	Limit 48 to yr="2000 – Current"

Table S1b Medline search strategy

1	exp Arthritis, Rheumatoid/
2	rheumatoid.mp.
3	exp Scleroderma, Systemic/ or exp Scleroderma, Localized/
4	scleroderma.mp.
5	"systemic sclero*" .mp.
6	exp Sjogren's Syndrome/
7	sj#gren*.mp.
8	exp Dermatomyositis/
9	dermatomyositis.mp.
10	exp Polymyositis/
11	polymyositis.mp.
12	"antisynthetase syndrome".mp.
13	"inflammatory myositis".mp.
14	Connective Tissue Diseases/
15	"connective tissue dis*" .mp.
16	"collagen vascular dis*" .mp.
17	exp Mixed Connective Tissue Disease/
18	"mixed ctd".mp.
19	exp Undifferentiated Connective Tissue Diseases/
20	"undifferentiated ctd".mp.
21	exp Lupus Erythematosus, Systemic/
22	lupus.mp.
23	or/1-22
24	exp Lung Diseases, Interstitial/
25	exp Pulmonary Fibrosis/
26	(interstitial\$ adj3 (lung\$ or pulmonary\$ or pneumon\$)).mp.
27	((pulmonary\$ or lung\$) adj3 (fibros\$ or fibrot\$)).mp.
28	ild.mp.
29	Uip.mp.
30	Nsip.mp.
31	Ppfe.mp.
32	"pleuroparenchymal fibroelastosis".mp.
33	bronchiolitis.mp.
34	alveolitis.mp.
35	"organ\$ing pneumon*".mp.
36	or/24-35
37	tomography, x-ray computed/ or computed tomography angiography/ or exp tomography, spiral computed/
38	HRCT.mp.
39	"computed tomography".mp.
40	"CT-scan".mp.
41	Prevalence/
42	Prevalence.mp.
43	protective factors/ or risk factors/
44	"risk factor*".mp.
45	biomarker*.mp.
46	or/37-45
47	23 and 36 and 46
48	limit 47 to yr="2020 -Current"

Table S2 Number of studies and total sample size addressing each objective. Data are reported as number of studies (included conference abstracts), N = total number of patients.

Connective Tissue Disease	Prevalence	Risk Factors for ILD	ILD Pattern	Risk Factors for ILD Pattern
Rheumatoid Arthritis	23 (6) N = 18,884	41 (10) N = 11,806	54 (13) N = 4,897	8 (1) N = 1,543
Systemic Sclerosis	69 (13) N = 31,096	33 (3) N = 24,281	22 (4) N = 1,263	
Idiopathic Inflammatory Myositis	27 (1) N = 3,781	22 (2) N = 3,349	23 (2) N = 1,305	1 (0) N = 48
Primary Sjögren Syndrome	11 (4) N = 3,899	8 (3) N = 3,716	16 (5) N = 722	1 (0) N = 170
Mixed Connective Tissue Disease	4 (1) N = 599	3 (2) N = 537	3 (0) N = 52	
Systemic Lupus Erythematosus	5 (2) N = 6,749	1 (1) N = 289	3 (0) N = 93	
Undifferentiated Connective Tissue Disease			3 (1) N = 91	1 (0) N = 66

Abbreviations:
 CT: computed tomography,
 ILD: interstitial lung disease

Table S3a Characteristics of studies addressing prevalence of RA-ILD, distribution of CT patterns of RA-ILD, and risk factors for development of RA-ILD.

Study Characteristic	Prevalence Studies	Risk Factor ILD Studies	CT Pattern Studies	Risk Factor CT Pattern Studies
Total Studies <i>Conference Abstracts</i>	23 6	41 10	54 13	8 1
Total N	18,884	11,806	-	1,543
N ILD	1,055	3,548	4,897	1,206
Demographics				
Age (years, mean)	54.3 (N = 2,845)	57.1 (N = 3,309)	61.3 (N = 1,667)	61.7 (N = 610)
Disease Duration (years, mean)	7.3 (N = 2,372)	7.7 (N = 2,530)	9.6 (N = 455)	10.2 (N = 88)
Sex (male)	841/4,153 (20.3%)	1,788/6,901 (25.9%)	1,577/3,960 (39.8%)	353/848 (41.6%)
Smoking History (ever)	677/3,097 (21.9%)	1,935/6,012 (32.2%)	1,695/3,604 (47.0%)	115/269 (42.6%)
Serology				
RF	1,742/2,608 (66.8%)	2,992/4,146 (72.2%)	2,786/3,294 (84.6%)	-
Anti-CCP	1,608/2,158 (74.5%)	2,579/3,570 (72.2%)	2,427/2,862 (84.8%)	-
Lung Function				
FVC (mean % predicted)	-	-	80.1 (N = 2,359)	-
TLC (mean % predicted)	-	-	76.9 (N = 783)	-
DLCO (mean % predicted)	-	-	60.2 (N = 2,117)	-

Abbreviations: ANA: antinuclear antibody, anti-CCP: anti-cyclic citrullinated peptide, CT: computed tomography, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, ILD: interstitial lung disease, RA: rheumatoid arthritis, RF: rheumatoid factor, TLC: total lung capacity.

Table S3b Characteristics of studies addressing prevalence of SSc-ILD, distribution of CT patterns of SSc-ILD, and risk factors for development of SSc-ILD. No studies address risk factors for specific CT patterns.

Study Characteristic	Prevalence Studies	Risk Factor ILD Studies	CT Pattern Studies
Total Studies <i>Conference Abstracts</i>	69 13	33 3	22 4
Total N	31,096	24,281	-
Total N ILD	11,330	7,499	1,263
Demographics			
Age (years, mean)	55.4 (N = 26,273)	55.3 (N = 22,625)	56.0 (N = 659)
Disease Duration (years, mean)	9.1 (N = 23,874)	8.8 (N = 21,314)	-
Sex (male)	4,747/31,250 (15.2%)	3,519/24,041 (14.6%)	127/837 (15.2%)
Smoking History (ever)	950/2,705 (35.1%)	482/1,549 (31.1%)	178/570 (31.2%)
CTD Subtype			
Limited SSc	17,541/30,139 (58.2%)	13,034/23,298 (55.9%)	194/378 (51.3%)
Diffuse SSc	9,503/30,734 (30.9%)	7,340/23,696 (31.0%)	183/378 (48.4%)
Serology			
ANA	21,837/22,892 (95.4%)	20,536/21,498 (95.5%)	319/345 (92.0%)
Anti-Scl70	8,280/28,103 (29.5%)	7,007/22,876 (30.6%)	260/486 (53.5%)
Anti-Centromere	10,562/28,237 (37.4%)	8,713/23,207 (37.5%)	69/476 (14.4%)
Anti-RNP	1668/21,598 (7.7%)	1,536/20,839 (7.4%)	-
Lung Function			
FVC	90.2 (N = 1,720)	93.3 (N = 1,257)	80.8 (N = 595)
DLCO (mean % predicted)	66.0 (N = 1,598)	63.7 (N = 1,257)	57.2 (N = 595)

Abbreviations: ANA: antinuclear antibody, CT: computed tomography, CTD: connective tissue disease, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, ILD: interstitial lung disease, SSc: systemic sclerosis.

Table S3c Characteristics of studies addressing prevalence of IIM-ILD, distribution of CT patterns of IIM-ILD, and risk factors for development of IIM-ILD.

Study Characteristic	Prevalence Studies	Risk Factor ILD Studies	CT Pattern Studies	Risk Factor CT Pattern Studies
Total Studies <i>Conference Abstracts</i>	27 1	22 2	23 2	1 0
Total N	3,781	3,349	-	-
Total N ILD	1,335	948	1,305	48
Demographics				
Age (years, mean)	49.5 (N = 722)	49.4 (N = 682)	52.1 (N = 602)	-
Sex (male)	912/2,980 (30.6%)	933/3,011 (31.0%)	366/1,110 (33.0%)	17/48 (35.4%)
Smoking History (ever)	-	-	137/392 (34.9%)	19/48 (39.6%)
CTD Subtype				
Dermatomyositis	1,997/3,447 (57.9%)	1,674/3,075 (54.4%)	414/866 (47.8%)	18/48 (37.5%)
Polymyositis	1,206/2,729 (44.2%)	871/1,892 (46.0%)	323/831 (38.9%)	6/48 (12.5%)
Serology				
ANA	280/589 (47.5%)	-	2	-
Anti-Jo-1	105/841 (12.5%)	63/599 (10.4%)	2	6/48 (12.5%)
Lung Function				
FVC	-	-	72.2 (N = 586)	-
DLCO (mean % predicted)	-	-	60.4 (N = 522)	-

Abbreviations: ANA: antinuclear antibody, CT: computed tomography, CTD: connective tissue disease, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, IIM: idiopathic inflammatory myositis, ILD: interstitial lung disease.

Table S3d Characteristics of studies addressing prevalence of pSS-ILD, distribution of CT patterns of pSS-ILD, and risk factors for development of pSS-ILD.

Study Characteristic	Prevalence Studies	Risk Factor ILD Studies	CT Pattern Studies	Risk Factor CT Pattern Studies
Total Studies <i>Conference Abstracts</i>	11 4	8 3	16 5	1 0
Total N	3,899	3,715	-	170
Total N ILD	562	641	722	85
Demographics				
Age (years, mean)	50.4 (N = 1,460)	-	58.8 (N = 457)	-
Disease Duration (years, mean)	8.7 (N = 482)	-	-	-
Sex (male)	202/3,555 (5.7%)	201/3,447 (5.8%)	90/522 (17.2%)	16/170 (9.4%)
Smoking History (ever)	-	-	59/309 (19.1%)	-
Serology				
ANA	1,080/1,677 (64.4%)	1,473/1,683 (87.5%)	378/475 (79.5%)	163/170 (95.9%)
Anti-SSA/Ro	1,004/1,821 (55.1%)	1,507/1,851 (81.4%)	317/478 (66.3%)	141/170 (82.9%)
Anti-SSB/La	510/1,821 (28.2%)	791/1,832 (43.2%)	144/475 (30.4%)	78/170 (45.9%)
Lung Function				
FVC (mean % predicted)	-	-	66.53 (N = 350)	-
DLCO (mean % predicted)	-	-	58.6 (N = 350)	-

Abbreviations: ANA: antinuclear antibody, CT: computed tomography, CTD: connective tissue disease, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, ILD: interstitial lung disease, pSS: primary Sjögren syndrome.

Table S3e Characteristics of studies addressing prevalence of SLE-ILD, distribution of CT patterns of SLE-ILD, and risk factors for development of SLE-ILD. No studies addressed CT pattern risk factors.

Study Characteristic	Prevalence Studies	Risk Factor ILD Studies	CT Pattern Studies
Total Studies <i>Conference Abstracts</i>	5 2	1 1	3 0
Total N	6,749	289	-
Total N ILD	498	23	93
Demographics			
Age (years, mean)	46.4 (N = 3,685)	-	55.4 (N = 10)
Disease Duration (years, mean)	9.1 (N = 3,685)	-	-
Sex (male)	360/3,974 (9.1%)	42/289 (14.5%)	15/65 (23.1%)
Smoking History (ever)	1,351/3,215 (42.0%)	-	16/55 (29.1%)
Serology			
ANA	-	-	-
Lung Function			
FVC (median % predicted)	-	-	81.8 (N = 55)
DLCO (median % predicted)	-	-	57.4 (N = 55)

Abbreviations: ANA: antinuclear antibody, CT: computed tomography, CTD: connective tissue disease, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, ILD: interstitial lung disease, SLE: systemic lupus erythematosus.

Table S3f Characteristics of studies addressing prevalence of MCTD-ILD, distribution of CT patterns of MCTD-ILD, and risk factors for development of MCTD-ILD. No studies addressed risk factors for specific CT patterns.

Study Characteristic	Prevalence Studies	Risk Factor ILD Studies	CT Pattern Studies
Total Studies	4	3	3
<i>Conference Abstracts</i>	1	2	0
Total N	599	537	-
Total N ILD	327	292	52
Demographics			
Age (years, mean)	47.5 (N = 194)	49.1 (N = 144)	49.8 (N = 52)
Disease Duration (years, mean)	12.0 (N = 194)	13.4 (N = 144)	-
Sex (male)	15/234 (6.4%)	11/144 (7.6%)	6/52 (11.5%)
Smoking History (ever)	8/50 (16.0%)	-	13/ 37 (35.1%)
Serology			
RF	43/144 (30.0%)	43/144 (30.0%)	-
Anti-CCP	-	-	-
ANA	144/144 (100.5%)	144/144 (100.0%)	-
Lung Function			
FVC (mean % predicted)	82.0 (N = 50)	-	77.6 (N = 37)
TLC (mean % predicted)	87.0 (N = 50)	-	-
DLCO (mean % predicted)	87.0 (N = 50)	-	58.3 (N = 37)

Abbreviations: ANA: antinuclear antibody, anti-CCP: anti-cyclic citrullinated peptide, CT: computed tomography, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, ILD: interstitial lung disease, MCTD: mixed connective tissue disease, RF: rheumatoid factor, TLC: total lung capacity.

Table S3g Characteristics of studies addressing prevalence of UCTD-ILD, distribution of CT patterns of UCTD-ILD, and risk factors for development of UCTD-ILD. No studies addressed prevalence of ILD or risk factors for ILD in UCTD.

Study Characteristic	CT Pattern Studies	Risk Factor CT Pattern Studies
Total Studies <i>Conference Abstracts</i>	3 1	1 0
Total N	-	66
Total N ILD	91	41
Demographics		
Age (years, mean)	62.4 (N = 24)	57.7 (N = 66)
Sex (male)	21/50 (42.0%)	21/66 (31.8%)
Smoking History (ever)	13/24 (54.2%)	29/66 (44.0%)
Serology		
RF	8/50 (15.2%)	29/62 (46.8%)
ANA	34/50 (67.3%)	57/65 (87.7%)
Lung Function		
FVC (mean % predicted)	79.0 (N = 24)	-
DLCO (mean % predicted)	59.7 (N = 24)	-

Abbreviations: ANA: antinuclear antibody, CT: computed tomography, DLCO: diffusion capacity for carbon monoxide, FVC: forced vital capacity, ILD: interstitial lung disease, RF: rheumatoid factor, UCTD: undifferentiated connective tissue disease.

Table S4 List of references, study characteristics, and quality of evidence.

Reference	Year	Country	Connective Tissue Disease	Total N	ILD N	Newcastle Ottawa Scale
			<p>1 = Rheumatoid arthritis 2 = Systemic sclerosis 3 = Idiopathic inflammatory myositis 4 = Primary Sjögren syndrome 5 = Mixed connective tissue disease 6 = Systemic lupus erythematosus 7 = Undifferentiated connective tissue disease</p>			<p>0-3 = Low 4-6 = Medium 7-9 = High</p>
Aguilera Cros C, Ruiz Roman A, Lisbona Munoz M, Luque Leon M, Leon Rubio P, Sanchez Serrano JP, Povedano Gomez J, Rodriguez Portal JA. Prevalence and significance of rheumatoid factor and anti-CCP in patients with interstitial lung disease and rheumatoid arthritis. In; 2016 2016: Annual European Congress of Rheumatology of the European League Against Rheumatism, EULAR 2016. United Kingdom. 75 (Supplement 2) (pp 956); BMJ Publishing Group; 2016.	2016	Spain	1	45	45	7
Alamoudi OSB, Attar SM. Pleuropulmonary manifestation in patients with rheumatoid arthritis in Saudi Arabia. <i>Annals of Thoracic Medicine</i> 2017; 12(4): 266-271.	2017	Saudi Arabia	1	419	30	8
Alberti ML, Paulin F, Toledo HM, Fernandez ME, Caro FM, Rojas-Serrano J, Mejia ME. Undifferentiated connective tissue disease and interstitial lung disease: Trying to define patterns. <i>Reumatología Clínica</i> 2018; 14(2): 75-80.	2018	Mexico, Argentina	7	66	41	7
Alemao E, Doyle T, Sparks J, Rao A, Saini Y, Iannaccone C, Weinblatt ME, Shadick N. Evaluation of rheumatoid arthritis treatments and joint outcomes in rheumatoid arthritis-associated interstitial lung disease. In; 2019 2019: Annual European Congress of Rheumatology, EULAR 2019. Spain. 78 (Supplement 2) (pp 1099-1100); BMJ Publishing Group; 2019.	2019	USA	1	75	46	8

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Mori S, Koga Y, Sugimoto M. Different risk factors between interstitial lung disease and airway disease in rheumatoid arthritis. <i>Respiratory medicine</i> 2012; 106(11): 1591-1599.	2012	Japan	1	356	24	7
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Wang N, Zhang Q, Jing X, Guo J, Huang H, Xu Z. The Association Between MUC5B Mutations and Clinical Outcome in Patients with Rheumatoid Arthritis-Associated Interstitial Lung Disease: A Retrospective Exploratory Study in China. <i>MedSciMonit</i> 2020; 26(Journal Article): e920137.	2020	China	1	96	45	8
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Yiu KH, Ninaber MK, Kroft LJ, Schouffoer AA, Stolk J, Scherer HU, Meijss J, De VriesBouwstra J, Tse HF, Delgado V, Bax JJ, Huizinga TWJ, Marsan NA. Impact of pulmonary fibrosis and elevated pulmonary pressures on right ventricular function in patients with systemic sclerosis. <i>Rheumatology (United Kingdom)</i> 2016; 55(3): 504-512.	2016	Netherlands	2	102	51	8
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Zhang G, Zhang X, Liu Q, Zhao Z, Lin H, Wu C. Correlation between imaging features of high-resolution computed tomography and histopathology of connective tissue diseasesassociated interstitial lung disease in Chinese Population. In; 2016 2016: 18th Asia Pacific League of Associations for Rheumatology Congress, APLAR. China. 19 (Supplement 2) (pp 94); Blackwell Publishing; 2016.	2016	China	1, 2, 3, 4, 5, 6	2320	325	8
Zhang R, Sun T, Song L, Zuo D, Xiao W. Increased levels of serum galectin-3 in patients with primary Sjogren's syndrome: associated with interstitial lung disease. <i>Cytokine</i> 2014; 69(2): 289-293.	2014	China	4	87	56	7
Zhang S, Shen H, Shu X, Peng Q, Wang G. Abnormally increased low-density granulocytes in peripheral blood mononuclear cells are associated with interstitial lung disease in dermatomyositis. <i>Modern Rheumatology</i> 2017; 27(1): 122-129.	2017	China	3	48	28	7
Zhang T, Yuan F, Xu L, Sun W, Liu L, Xue J. Characteristics of patients with primary Sjogren's syndrome associated interstitial lung disease and relevant features of disease progression. <i>Clinical rheumatology</i> 2020(Journal Article).	2020	China	4	170	85	8

Zhang T, Zhang J, Liu X, Zhang L, Zhao D, Wu X, Xu H. A clinical analysis of prognostic factors for dermatomyositis-associated interstitial lung disease. <i>International Journal of Clinical and Experimental Medicine</i> 2018; 11(6) (pp 5903-5911); Arte-Number: JEM0066897, ate of Pubaton: 0066830 Jun 0062018.	2018	China	3	103	103	8
Zhang Y, Li H, Wu N, Dong X, Zheng Y. Retrospective study of the clinical characteristics and risk factors of rheumatoid arthritis-associated interstitial lung disease. <i>Clinical rheumatology</i> 2017; 36(4): 817-823.	2017	China	1	550	237	7
Zhao J, Liu Z, Wang Q, Xu D, Li M, Zeng X. The risk factors and prognosis of primary Sjogren's syndrome-associated interstitial lung disease: A multi-centered cohort study. In; 2019 2019: 21st Asia Pacific League of Associations for Rheumatology Congress, APLAR 2019. Australia. 22 (Supplement 3) (pp 224-225); Blackwell Publishing; 2019.	2019	China	4	1501	258	6
Zhao L, Yao L, Yuan L, Xia L, Shen H, Lu J. Potential contribution of interleukin-33 to the development of interstitial lung disease in patients with primary Sjogren's Syndrome. <i>Cytokine</i> 2013; 64(1): 22-24.	2013	China	4	110	54	6
Zhou M, Jiang L, Nie L, Chen T, Zhang T, Sun W, Sutikno J, Du Y, Xue J. Myopathy is a Risk Factor for Poor Prognosis of Patients with Systemic Sclerosis: A retrospective cohort study. <i>Medicine (Baltimore)</i> 2020; 99(33): e21734.	2020	China	2	204	129	8
Zoto A, Hafizi H, Backa T, Petrela E, Zaimi M, Harxhi A, Osmenaj R, Ylli Z. Pulmonary involvement in scleroderma. In; 2013 2013: (var.pagings). 16 (SUPPL. 1) (pp 96); Blackwell Publishing; 2013.	2013	Albania	2	58	42	7
Zuo Y, Ye L, Liu M, Li S, Liu W, Chen F, Lu X, Gordon P, Wang G, Shu X. Clinical significance of radiological patterns of HRCT and their association with macrophage activation in dermatomyositis. <i>Rheumatology</i> 2020(Journal Article).	2020	China	3	165	165	8

Table S5 Pooled prevalence of ILD CT patterns, by connective tissue disease. Data shown are pooled percent (95% confidence interval). Heterogeneity is displayed as I^2 statistic and p value.

CTD	Total N	UIP	NSIP	OP	LIP	Other	Non UIP
RA	4897	46 (42-50) $I^2 = 88.7\%$ $p = 0.00$	35 (31-40) $I^2 = 84.1\%$ $p = 0.00$	4 (3-6) $I^2 = 60.9\%$ $p = 0.00$		17 (12-23) $I^2 = 95.3\%$ $p = 0.00$	
SSc	1263	19 (15-24) $I^2 = 72.3\%$ $p = 0.00$	76 (70-82) $I^2 = 80.6\%$ $p = 0.00$			4 (1-7) $I^2 = 81.1\%$ $p = 0.00$	
IIM	1305	8 (4-13) $I^2 = 86.6\%$ $p = 0.00$	59 (52-67) $I^2 = 83.3\%$ $p = 0.00$	16 (9-25) $I^2 = 91.2\%$ $p = 0.00$		13 (7-21) $I^2 = 91.3\%$ $p = 0.00$	
pSS	722	28 (15-43) $I^2 = 91.3\%$ $p = 0.00$	49 (39-59) $I^2 = 81.0\%$ $p = 0.00$	3 (1-6) $I^2 = 27.6\%$ $p = 0.17$	7 (2-15) $I^2 = 78.2\%$ $p = 0.00$	5 (0-15) $I^2 = 90.2\%$ $p = 0.00$	
MCTD	52	6 (1-16) $I^2 = 0.0\%$ $p = 0.41$	70 (50-87) $I^2 = 48.2\%$ $p = 0.15$			21 (5-44) $I^2 = 63.0\%$ $p = 0.07$	
SLE	93	9 (1-20) $I^2 = 51.1\%$ $p = 0.13$	27 (12-45) $I^2 = 45.2\%$ $p = 0.14$			49 (31-67) $I^2 = 60.7\%$ $p = 0.08$	
UCTD	91	37 (20-55) $I^2 = 67.0\%$ $p = 0.048$					63 (45-80) $I^2 = 61.4\%$ $p = 0.08$

Abbreviations: CT: computed tomography CTD: connective tissue disease, RA: rheumatoid arthritis, SSc: systemic sclerosis, IIM: idiopathic inflammatory myositis, ILD: interstitial lung disease, LIP: lymphocytic interstitial pneumonia, MCTD: mixed connective tissue disease, NSIP: non-specific interstitial pneumonia, OP: organizing pneumonia, Other: other and undifferentiated, pSS: primary Sjögren syndrome, SLE: systemic lupus erythematosus, UCTD: undifferentiated connective tissue disease, UIP: usual interstitial pneumonia.