

Supplementary Table 1. Characteristics of the included studies (ILD)

Author	Year	Country	Study design	Population	Patients (n)	Mean age	Female (%)	Positive anti-Ro52 (n)	ILD prevalence (%)	Anti-Ro52 detection	ILD diagnosis
<i>Myositis</i>											
Cheng et al. (39)	2022	China	Cross-sectional	IIM	53	50	68	31	57		HAQ, 6MWT and HRCT
Bozzalla-Cassione et al. (40)	2022	Italy	Retrospective cohort	ASSD	60	65	65	34	92	Line blot	PFT and/or DLCO and/or HRCT
Vojinovic et al. (41)	2021	Italy	Retrospective cohort	IIM	92	61	75	30	25	Line blot	CXR, PFT, and HRCT
Chiang et al. (11)	2021	Taiwan	Cross-sectional	DM/PM	94	54	73			Line blot	PE, CXR, Sonography, CT, and biopsy if needed
Al Nokhatha et al. (22)	2021	Ireland	Retrospective cohort	IIM (MSA/MA A+)	87	58	61	46	33	Immunoblot	Diagnosed by a respiratory physician
Narain et al. (42)	2021	USA	Cross-sectional	IIM	60	54	72	25	42	Immunoassay	HRCT and PFT
Huang et al. (23)	2021	Taiwan	Retrospective cohort	IIM	97		73	26	47	Serology tests	HRCT and PFT
Bai et al. (43)	2021	China	Retrospective cohort	IIM	286	51	69	108	46	Immunoblotting	HRCT
Xing et al. (44)	2020	China	Retrospective cohort	DM	153	48	69	81	52	Immunoblotting	HRCT

Sánchez Romo et al. (45)	2020	Mexico	Cross-sectional	IIM	36	41	67	13	30		HRCT
Gan et al. (27)	2020	China	Cross-sectional	CADM	84	50	81	44	83	Immunoblotting	HRCT and PFT
Li et al. (46)	2019	China	Case-control	DM/PM	125	47	67	60	63	Line blot assay	HRCT
Temmoku et al. (47)	2019	Japan	Retrospective cohort	DM/PM	84		71	39	77	ELISA	HRCT
Srivastava et al. (24)	2016	India	Cohort	IIM	124	30	78	45	23	Line immunoblot	HRCT and PFT
Yamasaki et al. (48)	2015	Japan	Cross-sectional	IIM	97	53	72	35	68	ELISA	CXR and HRCT
Marie et al. (36)	2012	France	Retrospective cohort	ASS	89	59	64	36	74	ELISA	HRCT, PFT, and/or biopsy
Helmerts et al. (49)	2009	Sweden	Cross-sectional	DM/PM	29	50	76	8	38	Line blot assay	CXR or HRCT and/or PFT
Vancsa et al. (28)	2009	Hungary	Cross-sectional	ASS	27	40	93	12	70	ELISA	HRCT
Eloranta et al. (50)	2007	Sweden	Case-control	DM/PM/IBM	30	59	63	7	33	Inno-Lia assay and ELISA	CXR or HRCT and/or PFT
La Corte et al. (29)	2006	Italy	Cohort	ASS	21	55		11	76	Double Immunodiffusion	CXR, HRCT, and PFT
SLE											
Ruacho et al. (51)	2020	Sweden	Cohort	SLE	504	46	86	140	5	Multiplexd bead technology	Not mentioned
Lian et al. (52)	2016	China	Retrospective cohort	SLE	3948	32	78				HRCT and PFT

Li et al. (25)	2014	China	Cross-sectional	SLE	2024	33	91	481	4	Immunoblotting	CXR or HRCT
Mochizuki et al. (53)	1999	Japan	Cohort	SLE	137	42	92	63	8	ELISA	CXR and/or HRCT
<i>Sjogren</i>											
Shi et al. (2)	2022	China	Retrospective cohort	pSS	142	58	87	55	46		HRCT
Zhang at al. (54)	2020	China	Retrospective cohort	pSS	170		91	137	50	Immunoblotting	HRCT and PFT
Buvry et al. (37)	2020	France	Retrospective cohort	pSS	68	55	84	31	28		HRCT and PFT
Duarte et al. (55)	2019	Portugal	Retrospective cohort	pSS	137		95	48	12	Immunoassay	Imaging
Gao et al. (26)	2018	China	Case-control	pSS	249	57	93	139	66	Dot immunoblot and ELISA	HRCT and/or PFT
Liu et al. (56)	2018	China	Cohort	pSS	1015	52	95	685	16	Immunoblotting	HRCT
Solans-Laque et al. (57)	2018	Spain	Cross-sectional	pSS	160	61	96			ChemiLuminescent immune assay (CLIA) and immunoblot	Not mentioned
Zhao et al. (58)	2015	China	Cohort	pSS	471	49		363	30	ELISA	CT
Mekinian et al. (59)	2013	France	Retrospective cohort	Suspected pSS	84	60	58	18	54	Line immunoassay	HRCT and PFT
Ter Borg et al. (60)	2011	Netherlands	Retrospective cohort	pSS	65	61	89	48	6	ELISA and immunoblot	HRCT and/or histology

Tsuzaka et al. (61)	1993	Japan	Cross-sectional	pSS	84	47	52	5	Immunoprecipitation	Not mentioned	
Scleroderma											
Cozzani et al. (62)	2021	Italy	Cohort	SSc	52	62	75	18	65	ELISA	HRCT, PFT, and DLCO
Kawata et al. (63)	2018	Japan	Case-control	SSc	40	87	21	55	Immunoblot assay	HRCT	
Martins Rocha et al. (64)	2016	Portugal	Retrospective cohort	SSc	108	58	90	13	22	ELISA	HRCT
Wodkowski et al. (3)	2016	Canada/Australia/USA	Cohort	SSc	1250	55	85	103	35	Line immunoassay	HRCT or CXR and/or PE
Sánchez-Montalva et al. (65)	2014	Spain	Cross-sectional	SSc	132	57	87	47	33	Line immunoassay	PFT and HRCT or BAL
Ferreira et al. (66)	2012	Portugal	Retrospective cohort	Scleroderma	71	56	93	31	63	Immunoblotting	HRCT and/or BAL
Akiyama et al. (67)	2000	Japan	Cross-sectional	CREST	30	59	97	6	3	Double immunodiffusion	CXR and PFT
MCTD											
Reiseter et al. (68)	2018	Norway	Cohort	MCTD	119	51	75	29	41	Line immunoassay	HRCT
Gunnarsson et al. (69)	2016	Norway	Cross-sectional	MCTD	113	43	75	33	34	Line immunoassay	HRCT, PFT, and 6MWT
Other autoimmune diseases											
Cavagna et al. (70)	2022	Italy	Retrospective cohort	Anti-MDA5+	149	52	67	47	72	Line immunoblot	HRCT and PFT

Koulouri et al. (71)	2021	Greece	Cross-sectional	RA	67	63	75	9	39		HRCT
Decker et al. (72)	2021	France	Case-control	CTD	226	52	73	113	37	Line or dot immunoblot	HRCT and PFT
Kim et al. (73)	2019	Korea	Cohort	ARD (ANA+)	363	50	97	336	6		Not mentioned
Montoya et al. (74)	2017	Spain	Cohort	CTD	1432	59	18	241	10		Not mentioned
Irace et al. (75)	2016	Italy	Cross-sectional	UCTD	86	40	85	17	9		HRCT or PFT and DLCO
Ferreira et al. (76)	2012	Portugal	Cohort	CTD	41	50	78	35	63	Immunoblotting	HRCT

ILD = Interstitial Lung Disease; IIM = Idiopathic Inflammatory Myositis; HAQ = Health Assessment Questionnaire; 6MWT = The 6-minute Walking Test; HRCT/CT = High Resolution Computed Tomography; PFT = Pulmonary Function Test; ASS/ASSD = Antisynthetase syndrome; DLCO = Diffusing capacity of the lungs for carbon monoxide; CXR = Chest X Ray; DM = Dermatomyositis; PM = Polymyositis; IBM = Inclusion body myositis; SLE = Systemic lupus erythematosus; pSS = Primary Sjogren's Syndrome; SS = Sjogren's Syndrome; SSc = Systemic Sclerosis; BAL = Bronchoalveolar Lavage; CREST = Calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasia; RA = Rheumatoid arthritis; CTD = Connective tissue disease; ARD = Acute rheumatic disease; ANA = Anti-nuclear antibody; UCTD = Undifferentiated connective tissue disease; MCTD = Mixed connective tissue disease; PE = Physical examination; CADM = Clinically amyopathic dermatomyositis; ELISA = Enzyme-linked immunosorbent assay.

Supplementary Table 2. Characteristics of the included studies (RP-ILD)

Author	Year	Country	Study design	Population	Patients (n)	Mean age	Female (%)	Positive anti-Ro52 (n)	ILD prevalence (%)	Anti-Ro52 detection	RP-ILD diagnosis
<i>Myositis</i>											
Bozzalla-Cassione et al. (40)	2022	Italy	Retrospective cohort	ASSD	55			34	27	Line blot	Rapid progression of ILD and acute dyspnea (within 4–6 weeks from symptom onset).
Wang et al. (12)	2022	China	Retrospective cohort	DM	284	53	72	120	16		One of the four conditions in one month: 1) Progressive worsening of dyspnea; 2) decrease in lung function, including FVC of more than 10% or in DLCO of more than 15%; 3) increase in the degree of interstitial pneumonia on chest HRCT; 4) decrease in partial pressure of oxygen more 10 mmHg
Truzzi et al. (77)	2022	Brazil	Cohort	DM	74			13	35	Immunoblotting	Not mentioned

You et al. (13)	2022	China	Cohort	Anti- MDA5+/D M	272	53	72	174	34		Progressive dyspnea with one of either of the conditions in one month: 1) Progressive worsening of dyspnea that required hospitalization; 2) decrease in lung function, including FVC of more than 10% or in DLCO of more than 15%; 3) increase in the degree of interstitial pneumonia on chest HRCT; 4) decrease in partial pressure of oxygen more than 10 mmHg
Gui et al. (78)	2022	China	Retrospective cohort	IIM/ILD	267	56	61	148	17	Immunoblotting	Severe dyspnea and new interstitial pneumonia on HRCT in one month
Xu et al. (35)	2021	China	Cohort	Anti- MDA5+/C ADM/ILD	83	52	67	39	75	Immunoblotting	One of either three conditions in one month: 1) Progressive worsening of dyspnea; 2) increase in the degree of interstitial pneumonia on chest HRCT; 3) decrease in partial pressure of oxygen more than 10 mmHg with no other apparent etiology

Li et al. (16)	2021	China	Retrospective cohort	DM/CADM	326	51	75			Immunoblotting	Progressive worsening of ILD in 3 months with simultaneous rapidly progressive dyspnea and hypoxemia, that required ventilation or oxygen therapy
Trallero-Araguas et al. (79)	2021	Spain	Retrospective cohort	Anti-MDA5+DM	90	47	56	38	31	Immunoblotting	Not mentioned
Li et al. (34)	2021	China	Cross-sectional	Myositis	20	49	75	11	55		progressive worsening of ILD in three months
Gan et al. (27)	2020	China	Retrospective cohort	CADM	70	51	83	34	57	Immunoblotting	(AIP: deterioration within one month, or SIP: deterioration within three months but more than one month). Deterioration is defined as one or more of 1) a worsening of symptoms; 2) an increase in the degree of interstitial pneumonia on chest HRCT, or 3) more than 10% decrease in vital capacity or more than 1.33 kPa decrease in PaO ₂ .
Temmoku et al. (47)	2019	Japan	Retrospective cohort	DM/PM	84		71	5	46	ELISA	Progressive dyspnea, progressive hypoxemia, and a worsening of interstitial pneumonia on chest radiography in one month.

AIP = Acute interstitial pneumonia; ASSD = Antisynthetase syndrome; CADM = Clinically amyopathic dermatomyositis; DLCO = Diffusing capacity of the lungs for carbon monoxide; DM = Dermatomyositis; FVC = Forced vital capacity; HRCT/CT = High resolution computed tomography; IIM = Idiopathic Inflammatory Myositis; ILD = Interstitial lung diseases; PaO₂ = Partial Pressure of Oxygen; PE = Physical examination; PFT = Pulmonary function test; PM = Polymyositis; SIP = Subacute interstitial pneumonia; ELISA = Enzyme-linked immunosorbent assay.

Supplementary Table 3. Quality assessment table of the included studies based on the Newcastle-Ottawa scale for cross-sectional studies.

Study name	Selection				Comparability	Outcome assessment		Total
	1	2	3	4	5	6	7	
ILD								
Cheng et al. (39)	*		*		**	**	*	7
Cavagna et al. (70)	*	*	*		**	**		7
Bozzalla-Cassione et al. (40)	*		*	**	**	**	*	9
Vojinovic et al. (41)	*	*		**	**	**	*	9
Chiang et al. (11)	*		*	**	**	**	*	9
Al Nokhatha et al. (22)	*		*	**	**			6
Narain et al. (42)	*			**	**	**		7
Huang et al. (23)	*		*		**	**		6
Bai et al. (43)	*	*	*	**	**	**	*	10
Xing et al. (44)	*	*	*	**	**	**	*	10
Sánchez Romo et al. (45)	*		*	**		**		6
Gan et al. (27)	*	*	*	**	**	**	*	10
Li et al. (46)	*	*	*	**		**		7
Temmoku et al. (47)	*		*	**	**	**		8
Srivastava et al. (24)	*	*	*	**		**		7
Yamasaki et al. (48)	*		*	**	**	**		8
Marie et al. (36)	*		*	**	**	**		8
Helmerts et al. (49)			*	**	**			5
Vancsa et al. (28)	*		*	*		**		5
Eloranta et al. (50)			*	**	**	**		7

La Corte et al. (29)	*		*			**		4
Ruacho et al. (51)	*	*	*	**				5
Lian et al. (52)	*	*			**	**	*	7
Li et al. (25)	*	*	*	*		**		6
Mochizuki et al. (53)	*	*		*		**		5
Shi et al. (2)	*	*	*		**	**	*	8
Zhang et al. (54)	*	*	*	**	**	**		9
Buvry et al. (37)	*		*		**	**		6
Duarte et al. (55)	*	*	*		**	**		7
Gao et al. (26)	*	*	*	*	**	**		8
Liu et al. (56)	*	*			**	**	*	7
Solans-Laqué et al. (57)	*	*	*	**			*	6
Zhao et al. (58)	*	*	*	*	**	**		8
Mekinian et al. (59)	*		*	**	**	**		8
Ter Borg et al. (60)	*		*	*		**		5
Tsuzaka et al. (61)	*		*	*				3
Cozzani et al. (62)	*		*	*	**	**		7
Kawata et al. (63)	*		*	**		**		6
Martins Rocha et al. (64)	*	*	*	*	**	**		8
Wodkowski et al. (3)	*	*	*	**	**	**	*	10
Sánchez-Montalva et al. (65)	*	*	*	**	**	**	*	10
Ferreira et al. (66)	*		*	**		**		6
Akiyama et al. (67)	*		*	*	**	*		6
Kim et al. (73)	*	*	*		**			5

Koulouri et al. (71)			*	**	**			5
Decker et al. (72)	*	*	*	**	**	**		9
Reiseter et al. (68)	*	*	*	**	**	**	*	10
Montoya et al. (74)	*	*	*		**		*	6
Gunnarsson et al. (69)	*	*	*	**	**	**		9
Irace et al. (75)	*		*		**	**		6
Ferreira et al. (76)	*		*	**		**		6
RP-ILD								
Bozzalla-Cassione et al. (40)	*		*	**	**	*		7
Wang et al. (12)	*	*	*		**	**	*	8
Truzzi et al. (77)	*			**	**		*	8
You et al. (13)	*	*	*		**	**	*	8
Gui et al. (78)	*	*	*	**	**	**	*	10
Xu et al. (35)			*	**	**	**		7
Li et al. (16)	*	*	*	**	**	*	*	9
Trallero-Araguas et al. (79)			*					1
Li et al. (34)	*		*		**	*		5
Gan et al. (27)	*	*		**		*	*	6
Temmoku et al. (47)	*		*	**	**	**		8

1. Representativeness of sample 2. Sample size 3. Non-respondents 4. Ascertainment of the exposure 5. Comparability of subjects in different outcome groups based on design or analysis. 6. Assessment of the outcome 7. Statistical test; 9-10= Very good quality, 7-8= Good quality, 5-6= Satisfactory quality, 0-4= Unsatisfactory quality