

	DISCOVERY COHORT						VALIDATION COHORT					
	SSc-PAH			SSc-no PAH			SSc-PAH			SSc-no PAH		
	N			N			N			N		
<b>Sex</b> (female), n (%)	15	14	(93%)	16	14	(88%)	24	17	(71%)	17	15	(88%)
<b>Age</b> (yo), mean ( $\pm$ SD)	15	65	( $\pm$ 7)	16	50	( $\pm$ 14)	24	68	( $\pm$ 9)	17	67	( $\pm$ 7)
<b>ANA</b> , n (%)	13	13	(100%)	14	14	(100%)	17	17	(100%)	14	14	(100%)
- ACA, n (%)	12	9	(75%)	9	4	(44%)	8	7	(88%)	14	10	(71%)
- ATA, n (%)	12	0	(0%)	9	1	(11%)	8	0	(0%)	14	4	(29%)
- Anti-U1RNP, n (%)	12	1	(8%)	9	2	(22%)	8	0	(0%)	14	0	(0%)
<b>Disease duration</b>												
- since RP onset (y), mean ( $\pm$ SD)	10	19	( $\pm$ 13)	11	13	( $\pm$ 13)	8	18	( $\pm$ 12)	15	24	( $\pm$ 17)
- since first non-RP symptom onset (y), median [Q1;Q3]	8	8	[2;13]	10	10	[3;12]	6	7	[0;17]	14	7	[4;10]
- since diagnosis (y), median [Q1;Q3]	15	1.25	[0.05;6]	16	7	[1;11]	9	0.5	[0;9]	6	7	[3;13]
<b>BMI</b> (kg/m <sup>2</sup> ), mean ( $\pm$ SD)	15	28	( $\pm$ 4)	15	24	( $\pm$ 5)	16	28	( $\pm$ 6)	14	25	( $\pm$ 4)
<b>mRSS</b> , median [Q1;Q3]	5	3	[2;5]	9	3	[2;5]	7	3	[3;5]	14	6	[3;8]
<b>NYHA functional class</b>	15			8			18			4		
- class I, n (%)		0	(0%)		8	(100%)		0	(0%)		2	(50%)
- class II, n (%)		7	(47%)		0	(0%)		6	(33%)		2	(50%)
- class III, n (%)		8	(53%)		0	(0%)		10	(56%)		0	(0%)
- class IV, n (%)		0	(0%)		0	(0%)		2	(11%)		0	(0%)
<b>BNP</b> (pg/mL), median [Q1;Q3]	12	147	[69;505]	2	25	[24;27]	14	84	[39;264]			
<b>Nt-pro-BNP</b> (pg/mL), median [Q1;Q3]							14	727	[319;1098]	5	126	[88;134]
<b>eGFR</b> (mL/min/1.73m <sup>2</sup> ), mean ( $\pm$ SD)	12	68	( $\pm$ 15)	7	78	( $\pm$ 21)	16	82	( $\pm$ 20)	15	74	( $\pm$ 16)
<b>LVEF</b> (%), mean ( $\pm$ SD)	12	60	( $\pm$ 9)	11	64	( $\pm$ 6)	14	65	( $\pm$ 9)	15	63	( $\pm$ 6)
<b>Estimated sPAP</b> (mmHg), mean ( $\pm$ SD)	12	72	( $\pm$ 19)	7	26	( $\pm$ 5)	15	59	( $\pm$ 16)	10	28	( $\pm$ 4)
<b>FVC</b> (% predicted), mean ( $\pm$ SD)	13	85	( $\pm$ 10)	16	110	( $\pm$ 15)	20	88	( $\pm$ 18)	13	111	( $\pm$ 14)
<b>DLCO</b> (% predicted), median [Q1;Q3]	15	35	[8;52]	16	92	[87;109]	10	41	[33;65]	12	65	[59;82]

<b>ILD on CT-scan, n (%)</b>	15	6 (40%)	16	0 (0%)	24	5 (21%)	17	6 (35%)		
<b>Treatments</b>										
- Prednisone, n (%)	15	0 (0%)	16	2 (13%)	16	1 (6%)	15	0 (0%)		
- Cyclophosphamide, n (%)	15	0 (0%)	16	0 (0%)	16	0 (0%)	15	0 (0%)		
- Mycophenolate mofetil, n (%)	15	0 (0%)	16	1 (6%)	16	0 (0%)	15	0 (0%)		
- D-penicillamine, n (%)	15	0 (0%)	16	0 (0%)	16	2 (13%)	15	0 (0%)		
- Azathioprine, n (%)	15	0 (0%)	16	0 (0%)	16	0 (0%)	15	0 (0%)		
- Methotrexate, n (%)	15	0 (0%)	16	1 (6%)	16	1 (6%)	15	0 (0%)		
<b>Right-heart catheterization</b>										
- mPAP (mmHg), mean ( $\pm$ SD)	15	44 ( $\pm$ 10)	/	/	/	24	37 ( $\pm$ 8)	/	/	/
- PAWP (mmHg), mean ( $\pm$ SD)	15	10 ( $\pm$ 2)	/	/	/	24	10 ( $\pm$ 3)	/	/	/
- CO (L/min), mean ( $\pm$ SD)	15	4.9 ( $\pm$ 1.6)	/	/	/	24	4.6 ( $\pm$ 1.2)	/	/	/
- CI (L/min/m <sup>2</sup> ), mean ( $\pm$ SD)	15	2.7 ( $\pm$ 1.0)	/	/	/	18	2.5 ( $\pm$ 0.5)	/	/	/
- PVR (WU), mean ( $\pm$ SD)	15	8.5 ( $\pm$ 5.0)	/	/	/	24	6.2 ( $\pm$ 2.3)	/	/	/

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**Supplementary Table 1. Detailed characteristics of patients from the discovery and validation cohorts**

ACA: anti-centromere antibodies; ANA: antinuclear antibodies; ATA: anti-topoisomerase I antibodies; Anti-U1RNP: anti-U1-ribonucleoprotein antibodies; BMI: body-mass index; BNP: brain natriuretic peptide; CI: cardiac index; CO: cardiac output; DLCO: diffusing capacity of the lung for carbon monoxide; eGFR: estimated glomerular filtration rate; FVC: forced vital capacity; ILD: interstitial lung disease; LVEF: left ventricular ejection fraction; mPAP: mean pulmonary arterial pressure; Nt-pro-BNP: N-terminal pro-brain natriuretic peptide; NYHA: New York Heart Association; PAH: pulmonary arterial hypertension; PAWP: pulmonary arterial wedge pressure; PVR: pulmonary vascular resistance; Q: quartile; RP: Raynaud phenomenon; SD: standard deviation; sPAP: systolic pulmonary arterial pressure; SSc: systemic sclerosis; WU: Wood unit; y: year; yo: year old.