

Supplementary Table S1. Basic characteristics of pSS and SLE patients with PAH.

Variable	pSS-PAH (n=33)	SLE-PAH (n=33)	p-value
Age, years	42.7 ± 12.6	38.3 ± 11	0.267
Female gender	33 (100%)	31 (93.9%)	0.492
Body mass index, kg/m ²	22.8 ± 3.2	23.1 ± 4.7	0.744
Body surface area, m ²	1.6 ± 0.1	1.6 ± 0.2	0.369
Disease duration, months	8 (6, 24)	12 (5.5, 30)	0.517
Respiratory impairment			
Interstitial lung disease	1 (3.0%)	0	1.000
Emphysema	0	0	-
Pulmonary infection	0	2 (6.1%)	0.492
chronic bronchitis	0	1 (3.0%)	1.000
Cardiac impairment			
tricuspid insufficiency	5 (15.2%)	2 (6.1%)	0.427
left ventricular dysfunction	2 (6.1%)	0	0.492
Pulmonary function test			
FVC, % predicted	84.3 ± 13.5	84.3 ± 14.5	0.386
FEV1, % predicted	79.4 ± 13.3	78 ± 12.4	0.665
FEV1/FVC ratio	0.8 ± 0.1	0.8 ± 0.1	0.547
WHO functional class			0.215
I–II	12 (36.4%)	17 (51.5%)	
III–IV	21 (63.6%)	16 (48.5%)	
6MWD, m	400.5 ± 107.7	406.3 ± 115.8	0.846
NT-pro-BNP, pg/ml	1255.7 (432.8, 2518.3)	1097.8(380, 2346.5)	0.783
Co-morbidities			
Systemic hypertension	1 (3.0%)	4 (12.1%)	0.355
Diabetes mellitus	5 (15.2%)	1 (3%)	0.197
Hyperlipidaemia	5 (15.2%)	2 (6.1%)	0.427
Smoking	2 (6.1%)	4 (12.1%)	0.672
Alcohol intake	1 (3%)	3 (9.1%)	0.613
Drug therapy [†]			
None	2 (6.1%)	7 (21.2%)	0.149
Mono therapy	23 (69.7%)	17 (51.5%)	0.207
Combination therapy	8 (24.2%)	9 (27.3%)	1.000

Data are presented as mean±standard deviation, median (range) or number (percentage).

BMI: body mass index; BSA: body surface area; FEV1: forced expiratory volume in 1 second; FVC: forced vital capacity; NT-proBNP: N-terminal pro-brain natriuretic peptide; PAH: pulmonary arterial hypertension; pSS: primary Sjögren's syndrome; SLE: systemic lupus erythematosus; WHO-FC: World Health Organisation functional class; 6MWD: 6-min walk distance.

[†]Including endothelin receptor antagonists, phosphodiesterase type-5 inhibitors, prostacyclins and calcium channel blockers.

Supplementary Table S2. Echocardiographic and haemodynamic parameters of pSS and SLE patients with PAH.

Variable	pSS-PAH (n=33)	SLE-PAH (n=33)	p-value
Echocardiography			
LVEF, %	65.5 ± 9.2	65 ± 6.4	0.464
LA, mm	30.3 ± 4.3	30.2 ± 4.1	0.907
LVED, mm	36.9 ± 6.8	36.9 ± 5.2	1.000
RVED, mm	33.4 ± 7.3	31.2 ± 5.9	0.272
sPAP, mm Hg	92.2 ± 19.9	82.4 ± 23.7	0.079
Pericardial effusion	9 (27.3%)	9 (27.3%)	1.000
RHC			
SvO ₂ , %	68.6 ± 6.7	69.5 ± 5.4	0.608
mRAP, mm Hg	2 (1, 7)	6 (3, 8)	0.067
mPAP, mm Hg	47.5 ± 11.3	46 ± 9.6	0.604
CI, L/min/m ²	2.9 ± 0.7	3.1 ± 0.7	0.464
PVR, dyn•s/cm ⁵	876.1 ± 384.6	701.7 ± 263.1	0.077
PCWP, mm Hg	4.5 (3, 9)	7 (4, 9.5)	0.255

Data are presented as mean ± standard deviation or median (range).

CI: cardiac index; LA: left atrium dimension; LVED: left ventricular end-diastolic diameter; LVEF: left ventricular ejection fraction; mPAP: mean pulmonary arterial pressure; mRAP: mean right atrial pressure; PAH: pulmonary arterial hypertension; PCWP: pulmonary capillary wedge pressure; pSS: primary Sjögren's syndrome; PVR: pulmonary vascular resistance; RHC: right-sided heart catheterisation; RVED: right ventricular end-diastolic diameter; SLE: systemic lupus erythematosus; sPAP: systolic pulmonary arterial pressure; SvO₂: mixed venous oxygen saturation.

Supplementary Table S3. CPET parameters of pSS-PAH and SLE-PAH patients.

Variable	pSS-PAH (n=33)	SLE-PAH (n=33)	p-value
Power@AT, W	32.6 ± 12	38 ± 17.9	0.174
Power@Peak, W	59.4 ± 21.5	65.6 ± 30.6	0.354
VO ₂ @Rest, mL/min/kg	4.7 ± 0.7	4.9 ± 1	0.487
VO ₂ @AT, mL/min/kg	9.1 ± 1.6	9.8 ± 4.2	0.798
VO ₂ @Peak, mL/min/kg	11.6 ± 3.2	12.8 ± 4.7	0.211
METS@Rest	1.4 ± 0.2	1.4 ± 0.3	0.542
METS@AT	2.6 ± 0.5	2.8 ± 1.2	0.832
METS@Peak	3.4 ± 0.9	3.7 ± 1.4	0.248
RER@Rest	0.8 ± 0.1	0.9 ± 0.1	0.491
RER@AT	0.9 ± 0.1	0.9 ± 0.1	0.593
RER@Peak	1.1 ± 0.1	1.1 ± 0.1	0.681
VE@Rest, L/min	9.8 ± 2.4	10.6 ± 2.2	0.182
VE@AT, L/min	19.6 ± 4.6	21.8 ± 9	0.465
VE@Peak, L/min	33.5 ± 10.7	38.3 ± 15.7	0.233
VT@Rest, L	0.5 ± 0.1	0.6 ± 0.2	0.408
VT@AT, L	0.8 ± 0.2	0.9 ± 0.3	0.383
VT@Peak, L	1.1 ± 0.3	1.2 ± 0.4	0.384
RF@Rest, l/min	18.5 ± 4.7	18.6 ± 4.2	0.929
RF@AT, l/min	24.5 ± 5.3	24.5 ± 6.0	1.000
RF@Peak, l/min	30.5 ± 5.9	32.3 ± 6.5	0.270
HR@Rest, beat/min	84.7 ± 12	87.5 ± 11.9	0.356
HR@AT, beat/min	106.8 ± 13.3	108.2 ± 18.9	0.725
HR@Peak, beat/min	131.2 ± 21	133.4 ± 25.1	0.703
VO ₂ /HR@Rest, mL/beat	3.2 ± 0.6	3.3 ± 0.6	0.367
VO ₂ /HR@AT, mL/beat	4.8 ± 0.9	5.2 ± 1.8	0.464
VO ₂ /HR@Peak, mL/beat	5.1 ± 1.3	5.6 ± 1.6	0.106
SBP@Rest, mm Hg	103.8 ± 16.8	108.1 ± 14	0.353
SBP@AT, mm Hg	114.5 ± 25	123.9 ± 26.3	0.062
SBP@Peak, mm Hg	124.1 ± 31.5	135 ± 25.5	0.043
DBP@Rest, mm Hg	73 ± 12.8	73.4 ± 9	0.574
DBP@AT, mm Hg	77.4 ± 16.5	74.6 ± 19.7	0.659
DBP@Peak, mm Hg	78.8 ± 15.6	80.8 ± 14.8	0.291
P _{ET} CO ₂ @Rest, mm Hg	28.4 ± 4.4	27.9 ± 3.8	0.617
P _{ET} CO ₂ @AT, mmHg	28.4 ± 5.4	27.7 ± 5.3	0.613
P _{ET} CO ₂ @Peak, mmHg	25.6 ± 5.8	24.9 ± 5.3	0.629
P _{ET} O ₂ @Rest, mmHg	113.4 ± 6.3	115.2 ± 5.3	0.146
P _{ET} O ₂ @AT, mmHg	116.2 ± 5.7	117.2 ± 6	0.516
P _{ET} O ₂ @Peak, mmHg	122.8 ± 5.5	123.6 ± 4.6	0.513
VE/VO ₂ @Rest	35.8 ± 6.5	36 ± 4.6	0.930
VE/VO ₂ @AT	38.5 ± 8.1	39.2 ± 9.3	0.739
VE/VO ₂ @Peak	51.1 ± 10.3	50.8 ± 10.2	0.925
VE/VCO ₂ @Rest	42.5 ± 7	42 ± 4.2	0.870
VE/VCO ₂ @AT	42.4 ± 8.9	43.6 ± 12.2	0.923
VE/VCO ₂ @Peak	47.3 ± 10.3	48.1 ± 12.4	0.939
SpO ₂ @Rest, %	94.6 ± 4.5	96 ± 2.8	0.282
SpO ₂ @AT, %	93.8 ± 5.7	94.4 ± 3.9	0.959
SpO ₂ @Peak, %	93.8 ± 4.7	92.5 ± 6.2	0.607
VE/VCO ₂ slope, L/min/L/min	43.1 ± 10.6	45.7 ± 17.8	0.891
VO ₂ /WR slope, mL/(min•W)	3.6 (-0.3, 5.2)	4.8 (0, 7.6)	0.261
Lowest VE/VCO ₂ , L/min	39.9 ± 8	38.8 ± 6.5	0.919
OUES, L/mL/log(1/min)	778.2 ± 301.4	945 ± 361.6	0.065

Data are presented as mean ± standard deviation or median (range).

AT: anaerobic threshold; CPET: cardiopulmonary exercise testing; DBP: diastolic blood pressure; HR: heart rate; METS: metabolic equivalents; OUES: oxygen uptake efficiency slope; PAH: pulmonary arterial hypertension; P_{ET}CO₂: end-tidal partial pressure of carbon dioxide output; P_{ET}O₂: end-tidal partial pressure of oxygen; pSS: primary Sjögren's syndrome; RER: respiratory exchange rate; RF: respiratory frequency; SBP: systolic blood pressure; SLE: systemic lupus erythematosus; SpO₂: oxygen saturation; VCO₂: carbon dioxide output; VE: minute ventilation; VO₂: oxygen uptake; VT: tidal volume; WR: work rate.

Supplementary Table S4. Results of multiple linear regression analysis between CTD-PAH and IPAH.

Dependent variable	Independent variable	B	SE	β	<i>p</i> -value [†]	Adjusted R ²
Power@Peak	Constant	90.864	22.069		<0.001	0.219
	Aetiology of PAH	-12.684	3.917	-0.259	0.001	
	FVC	0.313	0.141	0.168	0.028	
	Smoking	12.255	7.903	0.118	0.123	
	mPAP	-0.610	0.143	-0.323	<0.001	
	Pericardial effusion	-12.593	5.089	-0.189	0.014	
	Interstitial lung disease	-12.265	11.147	-0.081	0.273	
VO ₂ @Peak	Constant	20.725	3.305		<0.001	0.204
	Aetiology of PAH	-1.766	0.587	-0.243	0.003	
	FVC	0.025	0.021	0.090	0.241	
	Smoking	-0.673	1.184	-0.044	0.570	
	mPAP	-0.099	0.021	-0.353	<0.001	
	Pericardial effusion	-2.028	0.762	-0.205	0.009	
	Interstitial lung disease	-1.008	1.669	-0.045	0.547	
METS@Peak	Constant	5.755	0.958		<0.001	0.189
	Aetiology of PAH	-0.492	0.170	-0.236	0.004	
	FVC	0.008	0.006	0.100	0.199	
	Smoking	-0.150	0.343	-0.034	0.663	
	mPAP	-0.028	0.006	-0.345	<0.001	
	Pericardial effusion	-0.531	0.221	-0.187	0.017	
	Interstitial lung disease	-0.321	0.484	-0.050	0.508	
VE@Peak	Constant	27.012	10.907		0.014	0.141
	Aetiology of PAH	-4.711	1.936	-0.214	0.016	
	FVC	0.203	0.070	0.231	0.004	
	Smoking	10.130	3.906	0.207	0.010	
	mPAP	-0.144	0.071	-0.162	0.043	
	Pericardial effusion	-3.379	2.515	-0.108	0.181	
	Interstitial lung disease	-4.546	5.509	-0.064	0.411	
VT@Peak	Constant	0.644	0.268		0.017	0.298
	Aetiology of PAH	-0.165	0.048	-0.263	0.001	
	FVC	0.010	0.002	0.400	<0.001	
	Smoking	0.257	0.096	0.193	0.008	
	mPAP	-0.005	0.002	-0.207	0.004	
	Pericardial effusion	-0.077	0.062	-0.090	0.217	
	Interstitial lung disease	-0.119	0.135	-0.061	0.380	
HR@Rest	Constant	74.536	12.702		<0.001	0.096
	Aetiology of PAH	8.311	2.254	0.318	<0.001	
	FVC	0.023	0.081	0.023	0.778	
	Smoking	-10.466	4.549	-0.189	0.023	
	mPAP	0.151	0.082	0.150	0.068	
	Pericardial effusion	3.469	2.929	0.098	0.238	
	Interstitial lung disease	0.415	6.416	0.005	0.948	
HR@Peak	Constant	148.726	22.197		<0.001	0.036
	Aetiology of PAH	-8.287	3.940	-0.187	0.037	
	FVC	0.177	0.142	0.105	0.213	
	Smoking	-0.795	7.949	-0.008	0.920	
	mPAP	-0.182	0.144	-0.107	0.207	
	Pericardial effusion	-1.865	5.118	-0.031	0.716	
	Interstitial lung disease	-11.773	11.212	-0.086	0.295	
VO ₂ /HR@Rest	Constant	3.852	0.747		<0.001	0.098
	Aetiology of PAH	-0.451	0.133	-0.293	0.001	
	FVC	0.001	0.005	0.013	0.871	
	Smoking	0.541	0.268	0.166	0.045	
	mPAP	-0.016	0.005	-0.262	0.002	
	Pericardial effusion	-0.157	0.172	-0.075	0.363	
	Interstitial lung disease	0.157	0.378	0.033	0.679	

Dependent variable	Independent variable	B	SE	β	<i>p</i> -value [†]	Adjusted R ²
SBP@Peak	Constant	122.927	31.229		<0.001	-0.008
	Aetiology of PAH	-8.495	5.542	-0.140	0.127	
	FVC	0.189	0.199	0.082	0.345	
	Smoking	5.183	11.184	0.040	0.644	
	mPAP	-0.033	0.202	-0.014	0.871	
	Pericardial effusion	0.057	7.201	0.001	0.994	
	Interstitial lung disease	-7.430	15.774	-0.040	0.638	
DBP@Peak	Constant	64.289	22.203		0.004	0.059
	Aetiology of PAH	-13.131	3.940	-0.294	0.001	
	FVC	0.197	0.142	0.116	0.167	
	Smoking	2.068	7.951	0.022	0.795	
	mPAP	-0.072	0.144	-0.042	0.617	
	Pericardial effusion	2.762	5.120	0.046	0.590	
	Interstitial lung disease	9.846	11.214	0.071	0.381	

AT: anaerobic threshold; CTD-PAH: connective tissue disease-associated pulmonary arterial hypertension; DBP: diastolic blood pressure; FVC: forced vital capacity; HR: heart rate; IPAH: idiopathic pulmonary arterial hypertension; METS: metabolic equivalents; mPAP: mean pulmonary arterial pressure; SBP: systolic blood pressure; VE: minute ventilation; VO₂: oxygen uptake; VT: tidal volume.

[†]*p*<0.005 was considered statistically significant.

Supplementary Table S5. Results of multiple linear regression analysis between pSS-PAH and SLE-PAH patients.

Dependent variable	Independent variable	B	SE	β	<i>p</i> -value [†]	Adjusted R ²
SBP@AT	constant	116.911	15.369		<0.001	0.241
	subtype of CTD-PAH	-3.618	7.493	-0.071	0.632	
	sPAP	-0.221	0.185	-0.190	0.242	
	PVR	0.007	0.013	0.089	0.598	
	mRAP	3.557	1.049	0.490	0.002	
SBP@Peak	constant	125.535	17.456		<0.001	0.209
	subtype of CTD-PAH	-3.923	8.510	-0.068	0.647	
	sPAP	-0.169	0.210	-0.129	0.428	
	PVR	0.001	0.015	0.009	0.956	
	mRAP	3.989	1.191	0.488	0.002	
OUES	constant	1332.332	207.615		<0.001	0.177
	subtype of CTD-PAH	-158.698	101.214	-0.236	0.125	
	sPAP	-0.602	2.503	-0.040	0.811	
	PVR	-0.247	0.173	-0.244	0.162	
	mRAP	-29.763	14.166	-0.312	0.042	

CTD-PAH: connective tissue disease-associated pulmonary arterial hypertension; mRAP: mean right atrial pressure; OUES: oxygen uptake efficiency slope; PAH: pulmonary arterial hypertension; pSS: primary Sjögren's syndrome; PVR: pulmonary vascular resistance; SBP: systolic blood pressure; SLE: systemic lupus erythematosus; sPAP: systolic pulmonary arterial pressure.

[†]*p*<0.016 was considered statistically significant.