

Title

Pulmonary arterial hypertension associated with primary Sjögren's syndrome: a multi-centre cohort study from China

Authors and affiliations

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Online Data Supplement

Supplementary Table 1. Baseline data comparison between patients included in follow-up risk assessment and those not included.

	Risk assessment included n=62	Risk assessment not included n=41	<i>P</i>
Gender (F/M)	61/1	39/2	0.566
Age at recruitment, yr	38.8±9.1	49.5±14.5	<0.001
PAH duration, mth	12.0[6.0, 29.0]	12.5[2.8, 45.9]	0.789
pSS duration, mth	46.0[12.7, 96.7]	46.6[14.2, 110.3]	0.677
Onset interval of pSS and PAH, mth	0.0[0.0, 55.2]	2.1[0.0, 105.6]	0.743
ESSDAI, score	3.7±4.3	9.7±7.5	<0.001
SSDDI, score	2.0±1.3	2.3±1.6	0.217
WHO Fc I-II	34(54.8%)	25(60.5%)	0.567
6MWD, m	418.0±105.6	372.9±83.4	0.071
BNP, ng/L	239[106, 501]	275[77, 643]	0.854
NT-proBNP, pg/ml	846[292, 2114]	810[329, 2070]	0.957
RHC			
mPAP, mmHg	49.5±9.4	46.0±12.0	0.102
PAWP, mmHg	8.0±3.0	9.3±2.6	0.050
PVR, WU	11.6±5.1	11.3±5.8	0.795
CI, L/min/m ²	2.5±0.7	2.6±1.0	0.542
RAP, mmHg	6.6±4.8	6.6±4.3	0.972
TTE			
PASP, mmHg	83.7±17.9	74.8±22.1	0.027
LVEF, %	68.2±7.1	67.2±5.9	0.478
Pericardial effusion, no	16(27.1%)	21(51.2%)	0.014
Treatment regimen			
Glucocorticoids, no	56(90.3%)	35(85.4%)	0.276
Immunosuppressant, no	54(87.1%)	35(85.4%)	0.365
Target therapy for PAH, no	58(93.5%)	33(80.5%)	0.191
≥2	11(17.7%)	5(12.3%)	0.391
Binary therapy ^a	57(91.9%)	27(65.1%)	0.001

ESSDAI=EULAR primary Sjogren's syndrome disease activity index. SSDDI = Sjogren's syndrome disease damage index, WHO Fc = WHO functional class, 6MWD = 6 minutes walking distance, BNP = brain natriuretic peptide, NT-proBNP = N-terminal pro-brain natriuretic peptide, RHC = right heart catheterization, mPAP = mean pulmonary arterial pressure, PAWP = pulmonary arterial wedge pressure, PVR = pulmonary vascular resistance, CI = cardiac index, RAP=right atrial pressure, TTE=transthoracic echocardiography, PASP = pulmonary arterial systolic pressure, LVEF=left ventricle ejection fraction.

^a combination of immunosuppressant and PAH target therapy.