

Supplementary

Supplementary table S1

Comparison of baseline characteristics between survivors and non-survivors

Characteristics	Non-survivors (n = 42)	Survivors (n = 240)	<i>P</i> value
Age at recruitment, yr	36.2 ± 12.0	34.9 ± 10.0	0.439
Female sex, %	100.0	99.2	1.000
Lupus duration, yr	3.3 ± 4.3	4.5 ± 5.5	0.176
Clinical features			
WHO FC I-II, %	29.7	53.8	0.007**
6MWD, m	347.1 ± 81.3	414.4 ± 98.2	0.001**
Raynaud's phenomenon, %	59.5	50.4	0.276
Acute rash, %	35.7	32.9	0.723
Serositis, %	40.5	36.3	0.600
Lupus nephritis, %	38.1	34.6	0.660
Neuropsychiatric lupus, %	2.4	4.2	0.905
Thrombocytopenia, %	40.5	46.3	0.488
Hypocomplementemia, %	59.5	64.2	0.564
Anti-dsDNA, %	35.7	35.3	0.961
Anti-Sm,%	31.0	36.7	0.476
Anti-U1 RNP, %	69.0	59.4	0.238
APL, %	12.8	13.1	0.961
SLEDAI	5.2 ± 3.8	6.5 ± 5.9	0.067
RHC			
mPAP, mmHg	52.6 ± 11.8	45.4 ± 11.9	0.001**
PVR, WU	13.9 ± 5.8	10.4 ± 5.2	0.001**
CI < 2.5 L/min×m ² , %	64.9	38.0	0.003**
RAP mmHg, %	5.9 ± 4.5	5.6 ± 5.8	0.849
Treatment			
Glucocorticoids	100.0	99.2	1.000

Immunosuppressants	95.2	92.5	0.524
PAH-targeted medications	76.2	69.2	0.358

WHO FC = WHO functional class, 6MWD = 6-minute walking distance, APL = antiphospholipid, SLEDAI = systemic lupus erythematosus disease activity index, RHC = right heart catheterization, mPAP = mean pulmonary arterial pressure, PVR = pulmonary vascular resistance, CI = cardiac index, RAP = right atrial pressure

Supplementary table S2

Comparison of baseline characteristics between patients who achieved or did not achieve their treatment goals

Characteristics	Achieved treatment goal (n = 115)	Did not achieve treatment goal (n = 148)	<i>P</i> value
Age at recruitment, yr	34.7 ± 9.6	35.2 ± 10.8	0.676
Female sex, %	99.1	99.3	1.000
Lupus duration, yr	4.9 ± 5.8	3.7 ± 4.6	0.082
Clinical features			
WHO FC I-II, %	53.6	45.5	0.217
6MWD, m	428.2 ± 93.0	376.0 ± 99.4	< 0.001**
Raynaud's phenomenon, %	56.5	47.3	0.138
Acute rash, %	33.0	33.1	0.991
Serositis, %	46.1	31.1	0.013*
Lupus nephritis, %	40.9	33.1	0.195
Neuropsychiatric lupus, %	6.1	2.7	0.174
Thrombocytopenia, %	53.0	43.9	0.142
Hypocomplementemia, %	70.4	60.1	0.083
Anti-dsDNA, %	37.2	31.7	0.360
Anti-Sm,%	36.5	35.1	0.816
Anti-U1 RNP, %	67.0	56.9	0.105

APL, %	13.1	13.0	0.973
SLEDAI	6.5 ± 6.7	6.0 ± 4.8	0.484
RHC			
mPAP, mmHg	43.7 ± 11.3	49.5 ± 12.6	< 0.001**
PVR, WU	9.7 ± 4.3	12.5 ± 6.0	< 0.001**
CI < 2.5 L/min×m ² , %	27.8	55.5	0.001**
RAP, mmHg	5.0 ± 6.3	6.2 ± 5.0	0.182
Treatment			
Glucocorticoids	99.1	99.3	1.000
Immunosuppressants	92.2	92.6	0.905
PAH-targeted medications	60.0	77.0	0.003**

WHO FC = WHO functional class, 6MWD = 6-minute walking distance, APL = antiphospholipid, SLEDAI = systemic lupus erythematosus disease activity index, RHC = right heart catheterization, mPAP = mean pulmonary arterial pressure, PVR = pulmonary vascular resistance, CI = cardiac index, RAP = right atrial pressure

Supplementary table S3

Baseline characteristics between intensive immunosuppressant (IST) group and the HCQ only group stratified by baseline serositis

	Baseline serositis			Without baseline serositis		
	IST	HCQ	P	IST	HCQ only	P
	n = 64	only	value	n = 113	n = 31	value
	n = 20					
Age at recruitment, yr	33.4 ±	35.7 ±	0.292	35.8 ±	36.1 ±	0.874
	8.2	9.6		10.4	12.8	
Female sex, %	100	100	-	98.2	100	0.456
WHO FC I-II, %	43.5	42.1	0.912	53.8	65.4	0.295
6 MWD > 380 m, %	59.6	42.9	0.400	76.1	59.1	0.121
CI ≥ 2.5	35.9	60.0	0.057	35.5	48.4	0.191
L/min×m ² , %						

WHO FC = WHO functional class, 6MWD = 6-minute walking distance, CI = cardiac index