

Supplementary Table S1. Clinical characteristics of patients at baseline in RP-ILD group and no RP-ILD group.

	RP-ILD (n=36)	No RP-ILD (n=79)	p
Age, mean \pm SD, years	54.64 \pm 12.5	49.91 \pm 13.92	0.615
Course of the disease, median (range), months	1 (1, 2)	4 (2, 7)	<0.001*
Male, No. (%)	15 (41.67%)	30 (37.97%)	0.707
Triple-Combination Therapy, No. (%)	13 (36.11%)	11 (13.92%)	0.007*
Death, No. (%)	27 (75%)	8 (10.13%)	<0.001*
Myasthenia, No. (%)	15 (41.67%)	27 (34.18%)	0.439
Gottron papule, No. (%)	15 (41.67%)	45 (56.96%)	0.128
Heliotrope rash, No. (%)	12 (33.33%)	46 (58.23%)	0.013*
V sign, No. (%)	11 (30.56%)	27 (34.18%)	0.702
Shawl sign, No. (%)	5 (13.89%)	21 (26.58%)	0.131
Skin erythema, No. (%)	12 (33.33%)	30 (37.97%)	0.632
Raynaud's phenomenon	1 (2.78%)	3 (3.8%)	1.000
Periungual erythemasotus, No. (%)	8 (22.22%)	23 (29.11%)	0.440
Arthritis, no. (%)	9 (25%)	23 (29.11%)	0.648
Mechanic's hands, No. (%)	11 (30.56%)	27 (34.18%)	0.702
Superficial erosion and ulcer, No. (%)	3 (8.33%)	14 (17.72%)	0.188
Anti-MDA5 antibody			0.300
Low titre (+), No. (%)	6 (16.67%)	21 (26.58%)	
Moderate titre (++) , No. (%)	7 (19.44%)	17 (21.52%)	
High titre (+++), No. (%)	22 (61.11%)	41 (51.9%)	0.046*
Anti-Ro52 antibody			
Low titre (+), No. (%)	14 (38.89%)	14 (17.72%)	
Moderate titre (++) , No. (%)	4 (11.11%)	11 (13.92%)	
High titre (+++), No. (%)	11 (30.56%)	22 (27.85%)	
CTX, No. (%)	15 (41.67%)	15 (18.99%)	0.010*
TAC, No. (%)	30 (83.33%)	56 (70.89%)	0.154
MMF, No. (%)	1 (2.78%)	3 (3.8%)	0.782
JAK, No. (%)	4 (11.11%)	10 (12.66%)	0.814
LEF	0 (0%)	1 (1.27%)	0.498
Nintedanib, No. (%)	8 (22.22%)	4 (5.06%)	0.005*
Pirfenidone, No. (%)	3 (8.33%)	2 (2.53%)	0.157

*Values statistically significant at p<0.05.

Supplementary Table S2. Laboratory characteristics of patients at baseline in RP-ILD group and no RP-ILD group.

	RP-ILD (n=36)	No RP-ILD (n=79)	p
White blood cell, median (range), 10 ⁹ /l	5.13 (3.77, 7.73)	4.88 (3.62, 6.86)	0.450
Lymphocyte, mean \pm SD, 10 ⁹ /l	0.75 \pm 0.41	0.96 \pm 0.46	0.969
Monocyte, median (range), 10 ⁹ /l	0.33 (0.26, 0.64)	0.48 (0.33, 0.61)	0.330
Neutrophile granulocyte, median (range), 10 ⁹ /l	4 (2.85, 6.65)	3.4 (2.36, 5.39)	0.233
PLT, mean \pm SD, 10 ⁹ /L	202.69 \pm 78.12	188.08 \pm 69.69	0.510
ALT, median (range), units/L	46.1 (24, 76.9)	42.05 (29.43, 78.73)	0.664
AST, median (range), units/L	51 (31.9, 84.1)	46.45 (30.18, 67.5)	0.399
LDH, median (range), units/L	344 (263, 440)	294 (238.5, 345.5)	0.015*
CK, median (range), units/L	39 (25, 112)	42 (29, 111)	0.649
ESR, mean \pm SD, mm/h	36.17 \pm 25.0	32.89 \pm 21.96	0.256
CRP, median (range), mg/L	9.66 (4.2, 23.5)	3.61 (2.05, 7.37)	<0.001*
Ferritin, median (range), ng/mL	941.6 (535.1, 1511.75)	497.8 (164.65, 872.35)	0.005
AFP, median (range), ng/mL	2.15 (0.79, 3.61)	2.31 (1.47, 4.07)	0.349
CEA, median (range), ng/mL	3.88 (0.58, 7.29)	3.39 (2.24, 5.82)	0.874
Ca-199, median (range), U/mL	9.17 (0.6, 17.83)	10.07 (5.02, 20.86)	0.403
Ca-724, median (range), U/mL	0.91 (0, 1.79)	1.17 (0.8, 3.3)	0.104
CYFRA21-1, median (range), ng/mL	3.62 (0, 5.84)	2.76 (2.12, 3.58)	0.140
NSE, median (range), ng/mL	17.84 (6.41, 23.9)	17.98 (13.71, 21.74)	0.913
CA125, median (range), U/mL	9.4 (0, 20.95)	6.85 (0, 10.78)	0.241
Fbg, mean \pm SD, g/L	3.38 \pm 1.13	2.96 \pm 0.97	0.081
D-Dimer, median (range), mg/L	1.13 (0.46, 2.16)	0.67 (0.48, 1.38)	0.283
IgG, median (range), g/L	14.2 (11, 16)	12.65 (10.65, 15.7)	0.313
IgA, mean \pm SD, g/L	2.95 \pm 0.98	2.73 \pm 1.35	0.059
IgM, median (range), g/L	1.23 (1, 2.05)	1.22 (0.84, 1.72)	0.228
C3, mean \pm SD, g/L	0.86 \pm 0.19	0.86 \pm 0.17	0.457
C4, median (range), g/L	0.26 (0.2, 0.3)	0.23 (0.19, 0.3)	0.759
LY%, mean \pm SD	11.09 \pm 7.45	17.49 \pm 10.79	0.302
CD3+, median (range)	64.4 (52.8, 73.75)	71.2 (60.43, 78.38)	0.102
CD4+, median (range)	44.55 (31.25, 50.95)	45.58 (36.1, 53.84)	0.538
CD8+ %, median (range)	14.8 (10.4, 24.9)	18.8 (12.83, 28.23)	0.311
CD19+, mean \pm SD	21.81 \pm 13.59	18.43 \pm 13.16	0.426
NK%, median (range)	7.3 (3.45, 9.4)	4.6 (2.1, 7.6)	0.175

* Values statistically significant at p<0.05.

Supplementary Table S3. Clinical characteristics of patients without RP-ILD at baseline in triple-combination therapy group and non-triple therapy group.

	Triple therapy (n=11)	Non-triple therapy (n=68)	p
Age, mean±SD, years	44.64±14.52	50.76±13.74	0.873
Course of the disease, median (range), months	4 (2, 12)	4 (2, 6)	0.516
Male, No. (%)	4 (36.36%)	26 (38.24%)	0.906
Death, No. (%)	2 (18.18%)	6 (8.82%)	0.340
myasthenia, No. (%)	4 (36.36%)	23 (33.82%)	0.869
Gottron papule, No. (%)	7 (63.64%)	38 (55.88%)	0.630
Heliotrope rash, No. (%)	6 (54.55%)	40 (58.82%)	0.790
V sign, No. (%)	4 (36.36%)	23 (33.82%)	0.869
Shawl sign, No. (%)	2 (18.18%)	19 (27.94%)	0.497
Skin erythema, No. (%)	5 (45.45%)	25 (36.76%)	0.582
Raynaud's phenomenon	0 (0%)	3 (4.41%)	0.478
Periungual erythematous, No. (%)	7 (63.64%)	16 (23.53%)	0.007
Arthritis, No. (%)	3 (27.27%)	20 (29.41%)	0.885
Mechanic's hands, No. (%)	6 (54.55%)	21 (30.88%)	0.125
Superficial erosion and ulcer, No. (%)	3 (27.27%)	11 (16.18%)	0.371
Anti-MDA5 antibody			0.014 *
Low titre (+)	1 (9.09%)	20 (29.41%)	
Moderate titre (++)	6 (54.55%)	11 (16.18%)	
High titre (+++)	4 (36.36%)	37 (54.41%)	
Anti-Ro52 antibody			0.961
Low titre (+)	2 (18.18%)	12 (17.65%)	
Moderate titre (++)	1 (9.09%)	10 (14.71%)	
High titre (+++)	3 (27.27%)	19 (27.94%)	
CTX, No. (%)	11 (100%)	4 (5.88%)	<0.001*
TAC, No. (%)	10 (90.91%)	46 (67.65%)	0.115
MMF, No. (%)	1 (9.09%)	2 (2.94%)	0.322
JAK, No. (%)	1 (9.09%)	9 (13.24%)	0.701
LEF, No. (%)	0 (0%)	1 (1.47%)	0.686
Nintedanib, No. (%)	0 (0%)	4 (5.88%)	0.409
Pirfenidone, No. (%)	0 (0%)	2 (2.94%)	0.565

*Values statistically significant at p<0.05.

Supplementary Table S4. Laboratory characteristics of patients without RP-ILD at baseline in the triple-combination therapy group and non-triple therapy group.

	Triple therapy (n=11)	Non-triple therapy (n=68)	p
White blood cell, median (range), 10 ⁹ /l	5.67 (3.78, 7.13)	4.85 (3.53, 6.8)	0.492
Lymphocyte, mean±SD, 10 ⁹ /l	0.77 ± 0.25	1 ± 0.48	0.248
Monocyte, mean±SD, 10 ⁹ /l	0.42 ± 0.17	0.49 ± 0.23	0.422
Neutrophile granulocyte, median (range), 10 ⁹ /l	4.38 (2.49, 6.17)	3.33 (2.33, 5.14)	0.308
PLT, median (range), 10 ⁹ /L	190 (52.75, 287.5)	181 (138, 227.5)	0.895
ALT, median (range), units/L	40 (31, 63.5)	45.7 (28.3, 87.7)	0.626
AST, median (range), units/L	53.1 (38, 66)	45.8 (28.1, 67.9)	0.530
LDH, median (range), units/L	326 (245, 418)	287.5 (237.5, 344.25)	0.272
CK, median (range), units/L	86 (30.6, 275)	38.5 (27.75, 105)	0.079
ESR, median (range), mm/h	36 (14, 44)	28.5 (14.25, 45)	0.940
CRP, median (range), mg/L	4.81 (1.7, 6.82)	3.38 (2.17, 7.39)	0.669
Ferritin, median (range), ng/mL	828.6 (413.65, 1983.2)	451.6 (159.3, 870)	0.040*
AFP, median (range), ng/m	1.95 (1.32, 4.2)	2.43 (1.48, 4.04)	0.659
CEA, median (range), ng/mL	2.56 (1.63, 3.71)	3.77 (2.29, 5.97)	0.154
Ca-199, median (range), U/mL	10.96 (6.22, 21.36)	9.91 (4.66, 21.03)	0.795
Ca-724, median (range), U/mL	1.56 (1.1, 3.53)	1.14 (0.77, 3.51)	0.434
CYFRA21-1, median (range), ng/mL	2.82 (2.28, 3.55)	2.73 (2.03, 3.58)	0.861
NSE, median (range), ng/mL	14.78 (9.89, 28.48)	18.38 (14.3, 21.74)	0.522
CA125, median (range), U/mL	6.15 (0, 10.3)	6.85 (0, 11.03)	0.939
Fbg, mean±SD, g/L	3.08 ± 0.37	2.94± 1.02	0.043*
D-Dimer, median (range), mg/L	0.41 (0.2, 0.9)	0.76 (0.54, 1.39)	0.086
IgG, median (range), g/L	13.95 (11.45, 15.53)	12.3 (9.98, 15.83)	0.261
IgA, median (range), g/L	2.3 (1.77, 3.87)	2.38 (1.92, 3.49)	0.903
IgM, median (range), g/L	1.13 (0.68, 1.56)	1.22 (0.85, 1.73)	0.444
C4, median (range), g/L	0.28 (0.22, 0.33)	0.22 (0.19, 0.29)	0.111
LY%, mean±SD	17.16 ± 7.19	17.52 ± 11.13	0.426
CD3+, median (range)	76.4 (65.16, 87.89)	71.1 (56.55, 77.15)	0.097
CD4+, median (range)	50 (45.55, 63.43)	44.9 (34.15, 51.3)	0.092
CD8+, median (range)	25.8 (11.9, 37.29)	18.8 (12.85, 25.8)	0.229
CD19+, mean±SD	15.14 ± 10.25	19.11 ± 13.6	0.410
NK%, median (range)	6.2 (2.55, 11.84)	4.6 (2.05, 7.2)	0.441

*Values statistically significant at p<0.05.

Supplementary Table S5. Clinical characteristics of patients with RP-ILD at baseline in triple-combination therapy group and non-triple therapy group.

	Triple therapy (n=13)	Non-triple therapy (n=23)	p
Age, mean±SD, years	50.15±15.44	57.17±10	0.090
Course of the disease, median (range), months	1 (1, 2)	2 (1, 3)	0.190
Male, No. (%)	6 (54.55%)	9 (13.24%)	0.681
Death, No. (%)	10 (90.91%)	17 (25%)	0.841
Myasthenia, No. (%)	7 (63.64%)	8 (11.76%)	0.265
Gotttron papule, No. (%)	4 (36.36%)	11 (16.18%)	0.319
Heliotrope rash, No. (%)	5 (45.45%)	7 (10.29%)	0.624
V sign, No. (%)	3 (27.27%)	8 (11.76%)	0.464
Shawl sign, No. (%)	2 (18.18%)	3 (4.41%)	0.845
Skin erythema, No. (%)	3 (27.27%)	9 (13.24%)	0.326
Raynaud's phenomenon	0 (0%)	1 (1.47%)	0.446
Periungual erythematous, No. (%)	4 (36.36%)	4 (5.88%)	0.354
Arthritis, No. (%)	3 (27.27%)	6 (8.82%)	0.841
Mechanic's hands, No. (%)	7 (63.64%)	4 (5.88%)	0.023*
Superficial erosion and ulcer, No. (%)	1 (9.09%)	2 (2.94%)	0.917
Anti-MDA5 antibody			0.409
Low titre (+)	1 (9.09%)	5 (7.35%)	
Moderate titre (++)	3 (27.27%)	4 (5.88%)	
High titre (+++)	8 (72.73%)	14 (20.59%)	
Anti-Ro52 antibody			0.421
Low titre (+)	5 (45.45%)	9 (13.24%)	
Moderate titre (++)	0 (0%)	4 (5.88%)	
High titre (+++)	5 (45.45%)	6 (8.82%)	
CTX, No. (%)	13 (118.18%)	2 (2.94%)	0.000
TAC, No. (%)	13 (118.18%)	17 (25%)	0.044 *
MMF, No. (%)	0 (0%)	1 (1.47%)	0.446
JAK, No. (%)	2 (18.18%)	2 (2.94%)	0.540
LEF, No. (%)	(0%)	(0%)	
Nintedanib, No. (%)	4 (36.36%)	4 (5.88%)	0.354
Pirfenidone, No. (%)	1 (9.09%)	2 (2.94%)	0.917

*Values statistically significant at p<0.05.

Supplementary Table S6. Laboratory characteristics of patients with RP-ILD at baseline in the triple-combination therapy group and non-triple therapy group.

	Triple therapy (n=13)	Non-triple therapy (n=23)	p
White blood cell, median (range), 10 ⁹ /l	5.43 ± 2.52	6.36 ± 3.06	0.733
Lymphocyte, mean±SD, 10 ⁹ /l	0.69 ± 0.29	0.78 ± 0.46	0.167
Monocyte, mean±SD, 10 ⁹ /l	0.4 (0.3, 0.85)	0.32 (0.25, 0.52)	0.151
Neutrophile granulocyte, median (range), 10 ⁹ /l	4.76 ± 2.33	4.32 ± 2.16	0.399
PLT, median (range), 10 ⁹ /L	191 ± 63.36	208.78 ± 85.51	0.244
ALT, median (range), units/L	50.85 (23.78, 116.65)	34.8 (24, 60)	0.719
AST, median (range), units/L	42 (24.3, 125.4)	51 (37.1, 79.7)	0.719
LDH, median (range), units/L	362.25 ± 161.37	379.35 ± 183.85	0.842
CK, median (range), units/L	209.25 ± 286.29	67.61 ± 68.25	<0.001*
ESR, median (range), mm/h	33.92 ± 23.59	37.35 ± 26.19	0.758
CRP, median (range), mg/L	13.73 (4.05, 40.3)	9.43 (4.2, 14.98)	0.420
Ferritin, median (range), ng/mL	1291.9 (447.03, 1656.18)	916.4 (535.1, 1118.78)	0.466
AFP, median (range), ng/mL	1.63 ± 1.51	2.75 ± 2.59	0.472
CEA, median (range), ng/mL	3.76 ± 4.82	5.02 ± 3.74	0.404
Ca-199, median (range), U/mL	12.49 ± 18.24	19.11 ± 30.31	0.516
Ca-724, median (range), U/mL	0 (0, 0.96)	1.22 (0.68, 2.35)	0.055
CYFRA21-1, median (range), ng/mL	4.79 ± 6.7	3.87 ± 2.53	0.019
NSE, median (range), ng/mL	13.93 ± 12.63	16.56 ± 9.2	0.164
CA125, median (range), U/mL	13.2 (0, 20.45)	0 (0, 21.3)	0.422
Fbg, mean±SD, g/L	3.41 ± 1.43	3.37 ± 0.99	0.123
D-Dimer, median (range), mg/L	1.67 (0.52, 2.48)	0.77 (0.37, 1.9)	0.214
IgG, median (range), g/L	14.87 ± 5.71	13.32 ± 2.87	0.050
IgA, median (range), g/L	2.64 ± 0.79	3.17 ± 1.06	0.404
IgM, median (range), g/L	1.15 ± 0.44	1.88 ± 0.93	0.006
C3, mean±SD, g/L	0.8 ± 0.15	0.9 ± 0.21	0.270
C4, median (range), g/L	0.25 ± 0.07	0.25 ± 0.07	0.984
LY%, mean±SD	13.53 ± 8.47	10.04 ± 7.04	0.448
CD3+, median (range)	65.1 (61.58, 76.9)	55.1 (49.73, 73.08)	0.136
CD4+, median (range)	43.93 ± 7.57	38.83 ± 19.5	0.015
CD8+, median (range)	22.66 ± 12.57	14.91 ± 9.29	0.310
CD19+, mean±SD	18.71 ± 11.93	23.55 ± 14.51	0.362
NK%, median (range)	6.63 ± 4.9	7.25 ± 4.95	0.879

*Values statistically significant at p<0.05.