

Supplementary Table S1. Frequency and features of subgroups in patients with IPAF.

Subgroups	Features	n (%)
Scleroderma-like	<i>Antibodies:</i> Nucleolar ANA (n=6), SCL-70 (n=7) or Anti-centromere Ab (n=1) <i>Clinical findings:</i> skin thickening (n=7), palmar telangiectasia (n=4), Raynaud's phenomenon (n=17), digital ulcers (n=1), unexplained digital oedema (n=6), or positive nailfold capilloscopy (n=6).	21 (35.6)
Myositis-like	<i>Antibodies:</i> AS (n=2) or MDM5 (n=0) or PM-SCL Ab (n=1). <i>Clinical findings:</i> Gottron's papules (n=2), mechanic hands (n=1), arthritis (n=2); Raynaud's phenomenon (n=2), or muscle weakness (n=0).	4 (6.8)
Sjögren's syndrome-like	<i>Antibodies:</i> SSA (n=3) or SSB Ab (n=2). <i>Clinical findings:</i> Sicca symptoms (n=4) or parotid swelling (n=1).	4 (6.8)
Rheumatoid arthritis-like	<i>Antibodies:</i> CCP (n=3) or RF (n=5). <i>Clinical finding:</i> arthritis (n=5), or morning stiffness (n=3)	5 (8.5)
Systemic lupus erythematosus -like	<i>Antibodies:</i> ds-DNA (n=0) or Anti-Sm Ab (n=0) <i>Clinical Findings:</i> SLE rash (n=0), oral ulcers (n=0), photosensitivity (n=1), arthritis (n=1) or cytopenias (n=0).	1 (1.7)
Unclassifiable	<i>Antibodies:</i> ANA (n=11), dsDNA (n=4), RF (n=4), CCP (n=5), RNP (n=3), SSA (n=6), SSB (n=2), Scl-70 (n=2), and Anti-centromere (n=1) <i>Clinical findings:</i> Arthritis (n=8), Raynaud's phenomenon (n=6), morning stiffness (n=4), sicca symptoms (n=2), digital oedema (n=2), photosensitivity (n=1), skin thickening (n=1), and telangiectasias (n=1).	24 (40.7)

Ab: antibodies; AS: anti-synthetase.

Supplementary Table S2. Radiological diagnosis according to IPAF subgroups.

Subgroup/Pattern	NSIP	IncUIP	OP	pUIP	UIP	IND	Total (%)
SLE-like	0	0	0	0	1	0	1 (1.7)
Myositis-like	1	0	0	2	0	1	4 (6.8)
Sjögren's syndrome-like	0	1	1	1	0	1	4 (6.8)
RA-like	1	2	0	1	1	0	5 (8.5)
Scleroderma-like	1	3	0	6	5	6	21 (35.6)
Unclassifiable	6	3	3	8	1	3	24 (40.7)
Total (%)	9 (15.3)	9 (15.3)	4 (6.8)	18 (30.5)	8 (13.6)	11 (18.6)	59 (100)

NSIP: non-specific interstitial pneumonia; UIP: usual interstitial pneumonia; inc: inconsistent with; OP: organising pneumonia; p: probable; IND: indeterminate; SLE: systemic lupus erythematosus; RA: rheumatoid arthritis.

Supplementary Table S3. Baseline pulmonary function testing according to IPAF subgroups.

Subgroup	FVC % mean (SD)	n	FEV1/FVC mean (SD)	n	DLCO % mean (SD)	n	TLC % mean (SD)	n	SMWD mean (SD)	n
None	69.65 (22.14)	23	72.05 (19.82)	21	51.35 (19.82)	20	71.10 (17.13)	20	378.59 (98.94)	16
Myositis-like	73.50 (29.24)	4	86.75 (26.78)	4	51.67 (3.06)	3	64.00 (17.61)	4	515.00 (83.22)	3
Scleroderma-like	65.63 (15.73)	19	64.86 (17.31)	14	45.94 (21.91)	17	71.29 (20.23)	14	408.20 (42.77)	12
SLE-like	84.00	1	71.00	1	73.00	1	74.00	1	429.00	1
RA-like	64.20 (14.72)	5	64.50 (14.48)	4	43.75 (10.34)	4	62.00 (11.46)	4	274.40 (125.10)	5
Sjögren's syndrome-like	62.00 (13.00)	3	69.67 (20.82)	3	38.00 (2.83)	2	63.50 (6.36)	2	456.00 (50.27)	3

FVC%: percent-predicted forced vital capacity; FEV1: forced expiratory volume in one second; TLC%: percent-predicted total lung capacity; DLCO%: percent-predicted diffusion capacity of the lung for carbon monoxide; SMWD: six-minute walk distance; SLE: systemic lupus erythematosus; RA: rheumatoid arthritis; SD: standard deviation.

Supplementary Table S4. Cox proportional hazard analysis influencing survival in patients with IPAF.

Parameter	Hazard ratio	95% HR CI	p-value
African American*	4.314	0.679 – 27.413	0.1213
Male	4.559	1.325 – 15.691	0.0161
Age	1.142	1.055 – 1.235	0.0010
Smoker [†]	0.141	0.025 – 0.813	0.0284
Prior treatment [‡]	3.003	0.780 – 11.562	0.1098
Scleroderma-like [^]	3.718	0.551 – 25.095	0.1777
Other ^{^&}	6.057	0.857 – 42.790	0.0710

*In compared to White patients.

[†]This category combined current and former smokers and compared survival to non-smokers.

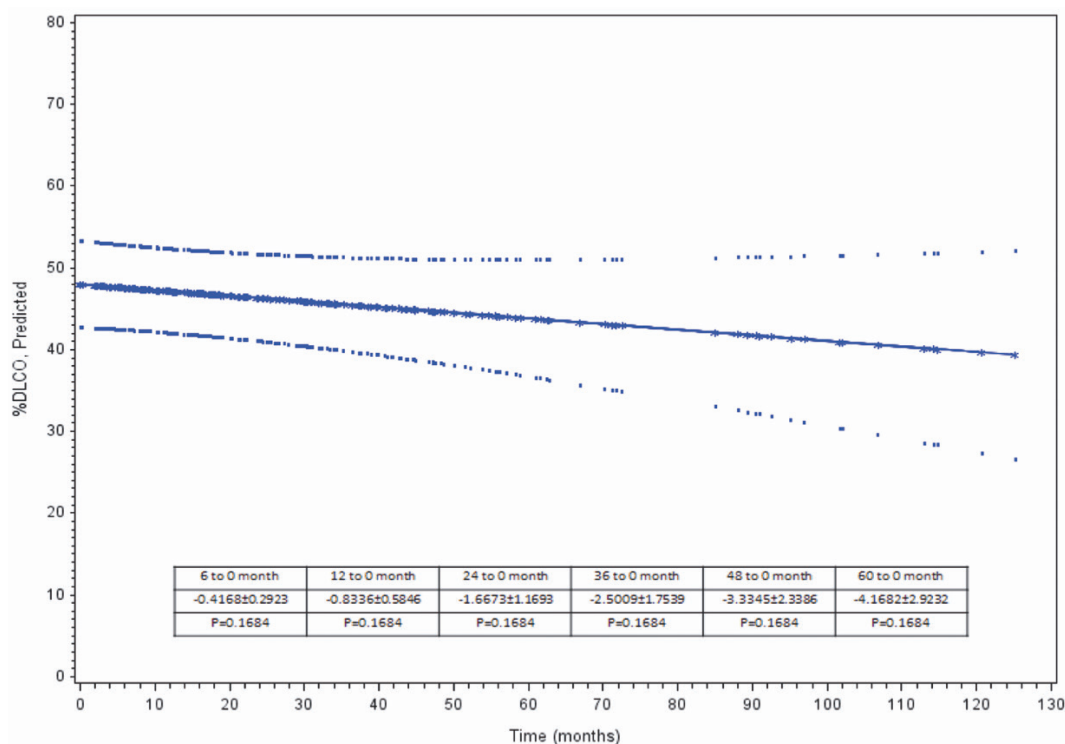
[‡]Defined as the use of steroids and/or immunosuppressive therapy.

[^]In compared to the unclassifiable subgroup.

[&]Other subgroup included RA-like, myositis-like, SLE-like and Sjögren's-like subgroups.

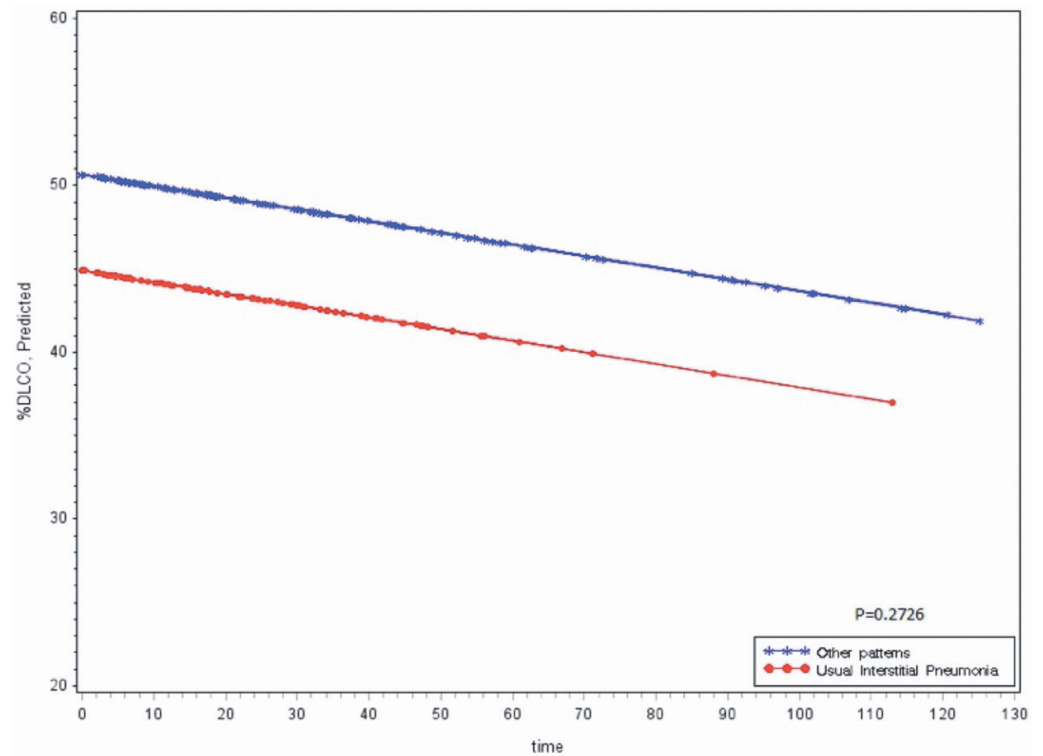
Supplementary Fig. S1.

Mixed-effects model estimates for predicted diffuse lung capacity over time in patients with IPAF.

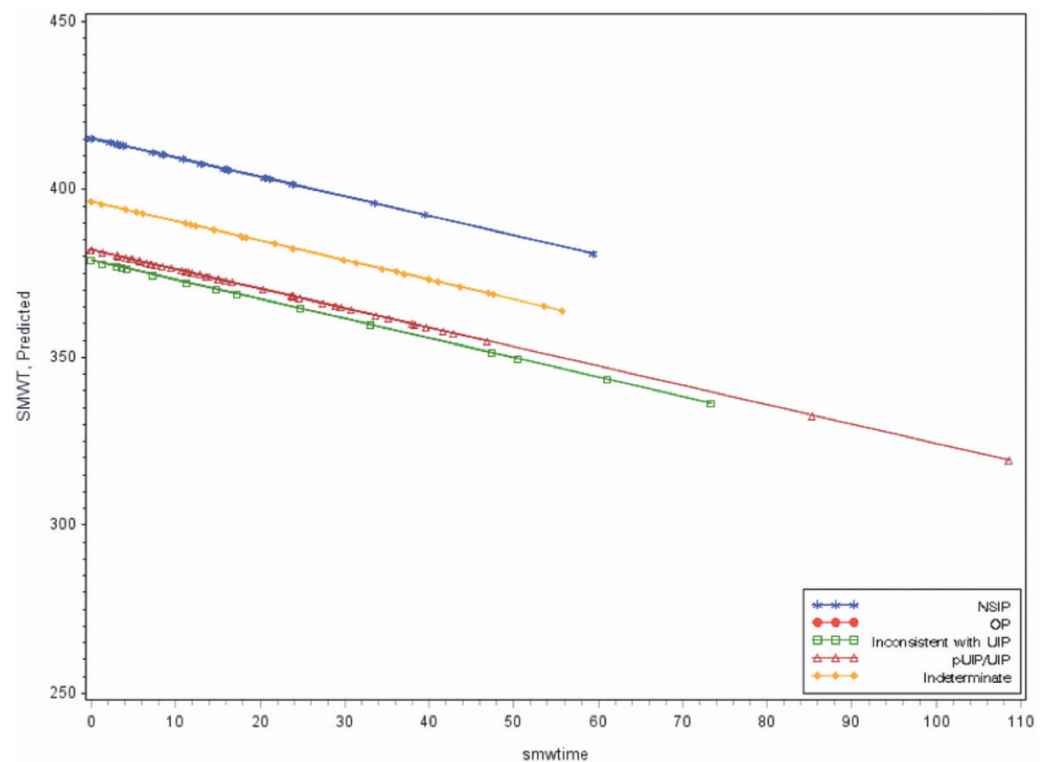


Supplementary Fig. S2.

Mixed-effects model estimates for predicted diffuse lung capacity over time in patients with IPAF according to the presence of radiological UIP.

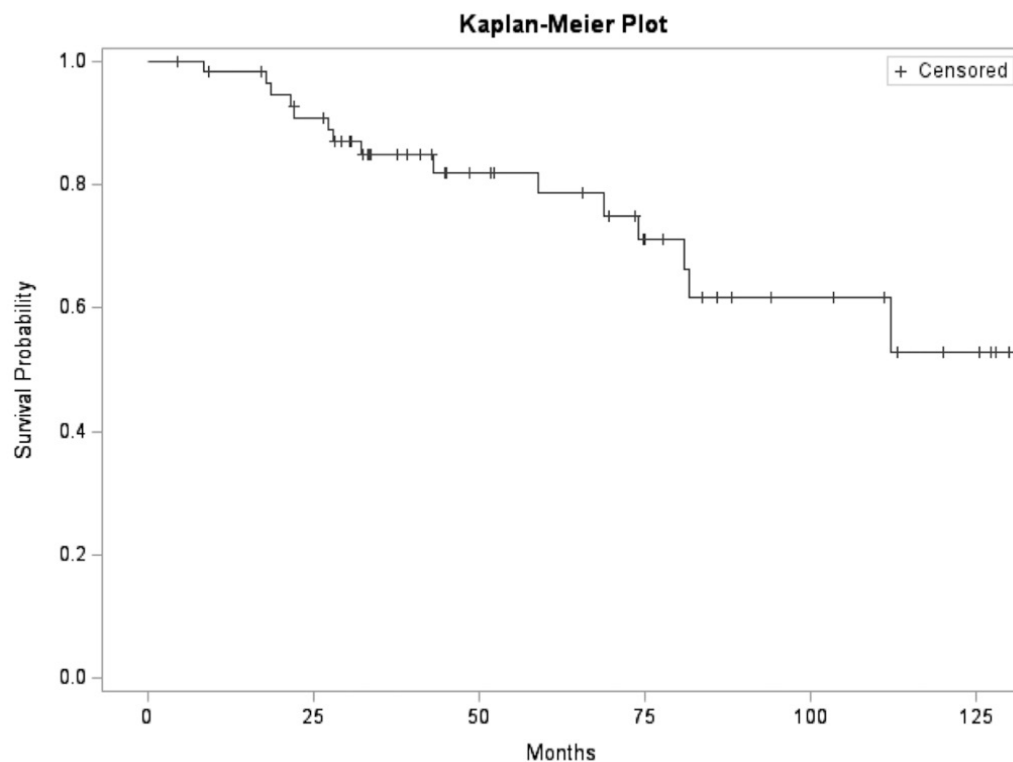
**Supplementary Fig. S3.**

Mixed-effects model estimates for six-minute walk distance (SMWD) over time in patients with IPAF according to radiological patterns.



Supplementary Fig. S4.

Overall Kaplan-Meier survival curve in patients with IPAF.



Supplementary Fig. S5.

Kaplan-Meier survival curve between UIP and non-UIP radiological pattern.

