--- ONLINE DATA SUPPLEMENT ---

Prevalence and Prognosis of Unclassifiable Interstitial Lung Disease

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Figure E1. Diagnoses of patients enrolled in the UCSF database during the study period.

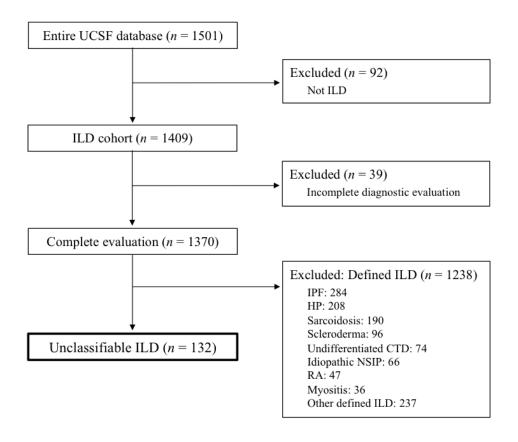


Table E1. Characteristics of patients excluded due to incomplete medical records.

Variable	Unclassifiable cohort (n=132)	Incomplete medical records (n=39)	p value	
Age, years	67.8 (12.9)	59.1 (13.3)	p = 0.0005	
Male sex, %	53.0%	43.6%	p = 0.34	
Smoking history				
Ever smoked, %	63.6%	64.1%	p = 0.98	
Pack-years	16.0 (23.9)	20.2 (27.7)	p = 0.39	
Measures of disease severity			-	
Long-term oxygen therapy, %	21.9%	12.5%	p = 0.21	
Pulmonary function			_	
FVC, % predicted	69.0 (22.1)	77.8 (20.9)	p = 0.03	
DLCO, % predicted	47.6 (19.7)	60.5 (21.2)	p = 0.0009	
HRCT findings				
Fibrosis score*	20.3 (13.1)	10.7 (7.2)	p = 0.002	
UIP pattern			p = 0.007	
Not UIP	33.0%	66.7%	_	
Possible UIP	49.5%	33.3%		
Definite UIP	17.4%	0%		
Honeycombing, %	49.5%	23.8%	p = 0.03	

Values are reported as mean (standard deviation) or percent. P values are reported for comparison between the stated subgroup and the remaining unclassifiable cohort, using Chisquared test, Fisher's Exact test, t-test, or Wilcoxon rank sum test as appropriate.

Abbreviations: DLCO, diffusion capacity of carbon monoxide; FVC, forced vital capacity; HRCT; high-resolution computed tomography; UIP, usual interstitial pneumonia.

Table E2. Differences in baseline characteristics comparing unclassifiable ILD subgroups.

Variable	Entire unclassifiable cohort (n=132)	Too old or frail for lung biopsy (n=68)	Conflicting CRP data (n=24)	Mild/Stable disease (n=12)	Insufficient tissue on lung biopsy (n=11)	Patient unwillingness for lung biopsy (n=10)
Age, years	67.8 (12.9)	73.0 (10.6)	60.2 (11.6)	60.9 (16.4)	62.0 (12.9)	71.2 (6.7)
		p < 0.00005	p = 0.001	p = 0.05	p = 0.12	p = 0.39
Male sex, %	53.0%	57.4%	41.7%	41.7%	45.5%	70.0%
		p = 0.31	p = 0.22	p = 0.41	p = 0.60	p = 0.33
Smoking history						
Ever smoked, %	63.6%	75.0%	50.0%	50.0%	54.6%	60.0%
		p = 0.005	p = 0.13	p = 0.30	p = 0.51	p = 1.00
Pack-years*	16.0 (23.9)	20.1 (27.6)	10.6 (16.9)	8.6 (14.3)	17.5 (28.2)	11.5 (16.4)
		p = 0.03	p = 0.20	p = 0.28	p = 0.83	p = 0.78
Measures of disease severity		•	•	•	•	•
Long-term oxygen therapy, %	21.9%	28.4%	17.4%	8.3%	36.4%	0%
		p = 0.06	p = 0.57	p = 0.69	p = 0.22	p = 0.12
Pulmonary function		1	1	1	1	1
FVC, % predicted	69.0 (22.1)	69.3 (18.8)	66.5 (24.6)	83.6 (26.6)	59.6%	69.1 (22.5)
	, ,	p = 0.86	p = 0.54	p = 0.02	p = 22.9	p = 0.99
DLCO, % predicted	47.6 (19.7)	39.4 (14.8)	55.1 (21.9)	65.1 (14.2)	54.6 (17.0)	52.8 (18.7)
	,	p < 0.00005	p = 0.47	p = 0.003	p = 0.27	p = 0.38
HRCT findings		r	r	1	1	r
Fibrosis score	20.3 (13.1)	23.0 (13.0)	19.4 (13.8)	9.6 (11.8)	20.4 (13.4)	18.5 (7.5)
	, ,	p = 0.02	p = 0.75	p = 0.01	p = 0.97	p = 0.68
UIP pattern		p = 0.08	0.48	p = 0.20	p = 0.47	p = 0.23
Inconsistent with UIP	33.0%	23.7%	44.4%	55.6%	50.0%	11.1%
Possible UIP	49.5%	55.9%	44.4%	44.4%	40.0%	55.6%
UIP	17.4%	20.3%	11.1%	0%	10.0%	33.3%
Honeycombing, %	49.5%	57.6%	33.3%	33.3%	50.0%	55.6%
		p = 0.07	p = 0.13	p = 0.49	p = 0.98	p = 0.74

Values are reported as mean (standard deviation) or percent. P values are reported for comparison between the stated subgroup and the remaining unclassifiable cohort, using Chi-squared test, Fisher's Exact test, t-test, or Wilcoxon rank sum test as appropriate.

Abbreviations: CRP, clinical, radiological and pathological; DLCO, diffusion capacity of carbon monoxide; FVC, forced vital capacity; HRCT; high-resolution computed tomography; UIP, usual interstitial pneumonia.