

Results

Validation of sample size

The sample size was calculated to provide 80% power to detect a between-group difference of survival time. Based on a past study comparing survival time of patients with a UIP pattern and those with a probable UIP pattern [1], median 3-year survival time was assumed to be 40% in UIP pattern group and 55% in possible UIP pattern (not probable UIP pattern) group. Based on these assumptions, when the ratio of sample number of UIP pattern and probable UIP pattern was estimated to be 1:2, the sample size was calculated as 155 patients in UIP pattern group and 233 patients in probable UIP pattern group for a Kaplan-Meier analysis at a significance level of $< 5\%$. The actual sample size of this study was 160 patients in UIP pattern group and 242 patients in probable UIP pattern group.

Validation of the diagnostic approach

To confirm the validity of the diagnoses we made for our cohort, we made a specific provisional diagnosis when the likelihood of the disease was $> 70\%$, the same as a high confidence provisional diagnosis proposed by Ryerson et al [2]. In terms of those who we originally made a diagnosis of unclassifiable interstitial lung disease (ILD) with a probable usual interstitial pneumonia (UIP) pattern on chest computed tomography (CT) and without histopathology, only 3 out of 48 patients were recognized as having a high likelihood of a specific disease: 1 idiopathic pulmonary fibrosis (IPF), 1 pleuroparenchymal fibroelastosis and 1 smoking-related ILD. All IPF diagnoses did not change, as in our clinical practice, we usually make a diagnosis of IPF when the likelihood is suggested to be $>70\%$. These changes did not affect the results of our analyses.

Annual rate of change in forced vital capacity

The adjusted annual rate of change in forced vital capacity (FVC) was not significantly different between UIP pattern and probable UIP pattern. The rate was -150 ml/year in UIP pattern as compared with -120 ml/year in probable UIP pattern (difference, -30 ml/year [95% confidence interval (95%CI), -10-40]; $p = 0.414$). Similarly, it was not significantly different between the two CT patterns in subjects with a diagnosis of IPF (-160 ml/year in UIP pattern vs. -190 ml/year in probable UIP pattern; difference, 30 ml/year [95%CI, -50-110]; $p = 0.515$). In subjects with a probable UIP pattern, it was significantly larger in those with a diagnosis of IPF (-200 ml/year) than of non-IPF (10 ml/year) (difference, -210 ml/year [95%CI, -290-110]; $p < 0.001$).

References

1. Lee JW, Shehu E, Gjonbrataj J, Bahn YE, Rho BH, Lee MY, Choi WI. Clinical findings and outcomes in patients with possible usual interstitial pneumonia. *Respir Med* 2015; 109: 510-516.
2. Ryerson CJ, Corte TJ, Lee JS, Richeldi L, Walsh SLF, Myers JL, Behr J, Cottin V, Danoff SK, Flaherty KR, Lederer DJ, Lynch DA, Martinez FJ, Raghu G, Travis WD, Udwadia Z, Wells AU, Collard HR. A standardized diagnostic ontology for fibrotic interstitial lung disease: An international working group perspective. *Am J Respir Crit Care Med* 2017; 196: 1249-1254.

TABLE S1 Cox proportional hazard analysis for survival time		
Parameters	Adjusted HR [95%CI]	P-value for HR
All patients		
UIP pattern on CT	Ref.	
Probable UIP pattern on CT	0.749 [0.557-1.008]	0.056
IPF only		
UIP pattern on CT	Ref.	
Probable UIP pattern on CT	0.885 [0.638-1.227]	0.464
Probable UIP pattern only		
Final diagnosis of non-IPF	Ref.	
Final diagnosis of IPF	1.791 [1.079-2.974]	0.024
Based on the non-imputed data. Hazard ratios were adjusted by age, sex, baseline forced vital capacity (% predicted), baseline diffusing capacity for the lung of carbon monoxide (% predicted) and use of antifibrotics. HR: hazard ratio; 95% CI: 95% confidence interval; UIP: usual interstitial pneumonia; CT: computed tomography; IPF: idiopathic pulmonary fibrosis.		

TABLE S2 Competing risk analysis for time to first acute exacerbation		
Parameters	Adjusted HR [95%CI]	P-value for HR
All patients		
UIP pattern on CT	Ref.	
Probable UIP pattern on CT	0.704 [0.462-1.074]	0.103
IPF only		
UIP pattern on CT	Ref.	
Probable UIP pattern on CT	0.881 [0.563-1.377]	0.577
Probable UIP pattern only		
Final diagnosis of non-IPF	Ref.	
Final diagnosis of IPF	2.212 [0.960-5.097]	0.062
Based on the non-imputed data. Hazard ratios were adjusted by baseline forced vital capacity (% predicted), baseline diffusing capacity for the lung of carbon monoxide (% predicted) and use of antifibrotics. AE: acute exacerbation; HR: hazard ratio; 95% CI: 95% confidence interval; UIP: usual interstitial pneumonia; CT: computed tomography; IPF: idiopathic pulmonary fibrosis.		