



Supplementary Figure 1. Forced vital capacity (FVC) predicted % changes among MCTD-ILD patients. We observed a FVC decline $\geq 10\%$ in 33 patients, a FVC decline comprised between 5% and 10% in 14 patients and a stabilization/improvement in 52 patients.

Characteristics	MCTD-ILD with follow-up (n=109)	MCTD-ILD without follow-up (n=41)	P-value
Demographics			
Age, mean (SD)	41 (16)	45 (16)	0.21
Male, n (%)	38 (35)	14 (34)	0.93
Smoking, n (%)	31 (28)	4 (10)	0.02
Clinical features			
Dyspnea, n (%)	92 (87)	29 (81)	0.36
FVC%, mean (SD)	80 (20)	80 (22)	0.97
DLCO%, mean (SD)	62 (17)	62 (21)	0.99

Supplementary Table 1. Characteristics of MCTD-ILD patients with and without at least on follow-up study on lung function.

Abbreviations: MCTD, mixed connective tissue disease; ILD, interstitial lung disease; FVC, forced vital capacity; DLCO, diffusing capacity for carbon monoxide.

Supplementary Table 2. Predictors of ILD progression defined as a FVC decline $\geq 10\%$ in MCTD-ILD patients analyzed by cox regression

Clinical predictors	Univariable		Multivariable	
	HR (95% CI)	P-value	HR (95% CI)	P-value
Digital ulcers	6.75 (1.72, 26.41)	0.006	16.7 (2.80, 100)	0.002
Rheumatoid factor	0.32 (0.10, 1.00)	0.05		

Abbreviations: ILD, interstitial lung disease; FVC, forced vital capacity; MCTD, mixed connective tissue disease; HR, hazard ratio.