

Title: Progressive Interstitial Lung Disease in Patients with Systemic Sclerosis: A Ten-year Follow-up Multicenter Cohort Study

Research team

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Abstract

Interstitial lung disease (ILD) is a serious and relatively common manifestation and a leading cause of higher mortality in systemic sclerosis (SSc). An important subset of patients with SSc develops progressive ILD, but the overall disease course and patterns of SSc-ILD has been scarcely studied. In addition, current clinical practice emphasizes the initiation of treatment in SSc patients with progressive ILD. Therefore, there is an unmet need to specifically identify patients with SSc at risk of progressive ILD since the natural history of SSc-ILD is variable. However, at present, there is no validated biomarker that predicts the course of SSc-ILD.

This study aims to determine the prevalence of progressive ILD over 1-year periods; to analyse the changes in Forced Vital Capacity (FVC) patterns associated with ILD progression in SSc over 1-year periods and overall change during the 10-year follow-up period; and to identify factors predictive of progressive ILD in SSc-ILD patients.