tomography (HRCT) and available data on pulmonary function tests and treatment. Longitudinal study included patients with at least one follow-up visit. Patients were classified as treated if they received a potential ILD modifying drug (immunosuppressive therapy or nintedanib). Treated and untreated patients were compared at baseline. Progression in the untreated group was defined as (i) forced vital capacity (FVC) decline from baseline of ≥10% or (ii) an FVC decline of 5-9% in association with a decline in diffusing capacity for carbon monoxide (DLCO) of ≥15%, or (iii) start of a ILD modifying treatment during follow-up. In the untreated group, patients who progressed at any time were compared with patients with stable disease during follow-up. Multivariable logistic regression was performed to identify (i) factors associated with non-prescription of a treatment in ILD patients at baseline and (ii) factors associated with progression in the untreated patients. Covariates were selected according to clinical experience and literature evidence.

**Results:** Among 496 patients included in our cohort, 209 (42%) patients had ILD on baseline HRCT: 48/209 (23%) were males, median disease duration 8 [IQ: 4-12] years, 67/209 (32%) of diffuse cutaneous subset and 86/209 (41%) had anti-ScI70 antibodies. Among them, 142/209 (68%) did not receive any potentially ILD modifying treatments at baseline. Untreated patients were older (59 vs. 54 years), had a longer disease duration, were less frequently smokers, had more frequently antinuclear antibodies and lower levels of CRP. They had more frequently a limited extent (<20%) of lung fibrosis on HRCT, higher FVC (97.02 [±19.76] % vs. 78.29 [±19.23] %) and DLCO (72.10 [±18.97] % vs. 57.37 [±20.81] %), better performances in the 6 minute walking test and were less frequently treated with low dose of glucocorticoids. In multivariable logistic regression, older age (OR: 1.04 [95% CI 1-1.00-1.09], p=0.032), a more extensive disease on HRCT (OR: 0.29 [0.09-0.90], p=0.037) and less frequent prescription of glucocorticoids (OR: 0.036 [0.12-0.92], p=0.037) were independent predictors of absence of ILD modifying treatment prescription in our cohort. From the 142 untreated patients, 96 were followed-up for 64 [39-96] months. Of these, 56 (58%) patients showed progression of ILD, of whom 43 progressed by lung function parameters. Of these 56 patients, 31 (56%) progressed in the first 18 months. Diffuse cutaneous subtype (OR: 5.26 [1.26-2.762], p=0.031), shorter disease duration (OR: 0.95 [0.90-0.99], p=0.035) and oesophageal symptoms (reflux, dysphagia) (OR: 3.51 [1.12-12.18], p=0.036) were independent predictors of progression during follow-up in untreated patients.

**Conclusion:** A considerable number of SSC patients with ILD are not treated in clinical practice, in particular patients with limited cutaneous SSc, older age and an overall less extensive ILD. However, during a follow-up of 5 years, contrary to the common belief, about 60% of the untreated patients showed ILD-progression. The diffuse cutaneous subtype, shorter disease duration and oesophageal symptoms at baseline characterized these patients. With the development of effective and safe therapies for SSC-ILD, our results support a change in practice.

**REFERENCES:**


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