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, even in the 6 minute walking test and were less frequently treated with low dose corticosteroids and non-smokers, had more frequently anticentromere antibodies and in the 6 minute walking test and were less frequently treated with low dose corticosteroids and non-smokers, had more frequently anticentromere antibodies and less frequently smokers, had more frequently anti-centromere 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. 57.57 [±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 [1.00-1.08], p=0.031), a 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) and less frequent prescription of glucocorticoids (OR: 0.036 [0.01-0.12], p=0.007) were independently associated with 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 [1.26-21.62], 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 for these patients for treatment.

REFERENCES:

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