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# PROGRESSION AND SURVIVAL OF SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE IN A MULTIETHNIC COHORT

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## INTRODUCTION

Systemic sclerosis (SSc) is a rare autoimmune disease that is characterised by vasculopathy and progressive fibrosis of the skin and internal organs. Interstitial lung disease (ILD) is a frequent complication of SSc and the leading cause of death. Serial decline in forced vital capacity (FVC) is a strong predictor of mortality. Our objective is to assess the progression and survival of patients with systemic sclerosis-associated ILD (SSc-ILD).

## METHODS

This retrospective cohort study was conducted at Kuala Lumpur Hospital. SSc patients who had high-resolution computed tomography (HRCT) thorax between 2008 to 2024 were identified. ILD was diagnosed based on radiologists' interpretation of HRCT thorax. Progressive ILD was determined from serial HRCT thorax and/or absolute decline in FVC ≥10% predicted. Cumulative survival rates following the diagnosis of ILD were estimated using the Kaplan-Meier method and log-rank test was applied to evaluate statistical significance.

## RESULTS

In total, 59 patients with SSc who had HRCT thorax were included. ILD was present in 40 (67.8%) patients. Mean age at diagnosis of ILD was 42.6 (SD 12.8) years and mean duration of follow-up from diagnosis of ILD was 5.1 (SD 4.1) years. Table 1 depicts the characteristics of SSc patients.

Nonspecific interstitial pneumonia was the main pattern observed on HRCT thorax, occurring in 47.5% of patients. Twenty-two patients (56.4%) had limited lung involvement, of <20%. Of the 25 patients with ILD who had serial HRCT thorax, 20 (80%) showed progressive ILD. In terms of treatment, 29 patients with ILD received mycophenolate mofetil (MMF). Twenty-six of them had serial measurements of FVC, of whom 4 showed absolute decline in FVC ≥10% predicted. Three of the 4 patients had received MMF for >12 months (range: 18 to 73 months). Correspondingly, progressive ILD on HRCT thorax was evident.

Twenty-two (75.9%) patients received MMF within 12 months of the diagnosis of ILD. There were 8 (20%) deaths among patients with ILD, wherein all had never received MMF in view of various factors. The cumulative survival rates of patients with ILD who were not treated with MMF at 2, 5 and 10 years were 81.8%, 43.6% and 29.1%, respectively. In contrast, all patients with SSc-ILD who had received MMF survived throughout the follow-up period (p<0.001) (Figure 1).

Table 1. Demographics and characteristics of SSc patients

| | Patients with SSc (n=59) | SSc-ILD (n=40) | SSc without ILD (n=19) | p value |
|---|---|---|---|---|
| **Female gender, n (%)** | 57 (96.6) | 38 (95) | 19 (100) | |
| **Mean age at diagnosis of SSc (SD), years** | 39.5 (14.2) | 38.6 (13.8) | 41.8 (14.8) | 0.412 |
| **Median duration of SSc (IQR), years** | 7.4 (9.5) | 7.3 (10.5) | 7.4 (9.8) | 0.667 |
| **Median duration of follow-up (IQR), years** | 4.8 (7.3) | 4.6 (5.9) | 5.0 (8.1) | 0.974 |
| **Subtype** | | | | 0.057 |
| **Sine** | 6 (10.2) | 2 (5.0) | 4 (21.0) | |
| **Limited** | 34 (57.6) | 22 (55.0) | 12 (63.2) | |
| **Diffuse** | 19 (32.2) | 16 (40.0) | 3 (15.8) | |
| **Autoantibodies** | | | | |
| **Anti-topoisomerase I antibody (n=58)** | 34 (58.6) | 28 (70.0) | 6 (33.3) | 0.009* |
| **Anti-centromere antibody (n=58)** | 10 (17.2) | 3 (7.5) | 7 (38.9) | 0.003* |

*denotes significant *p* value of <0.05

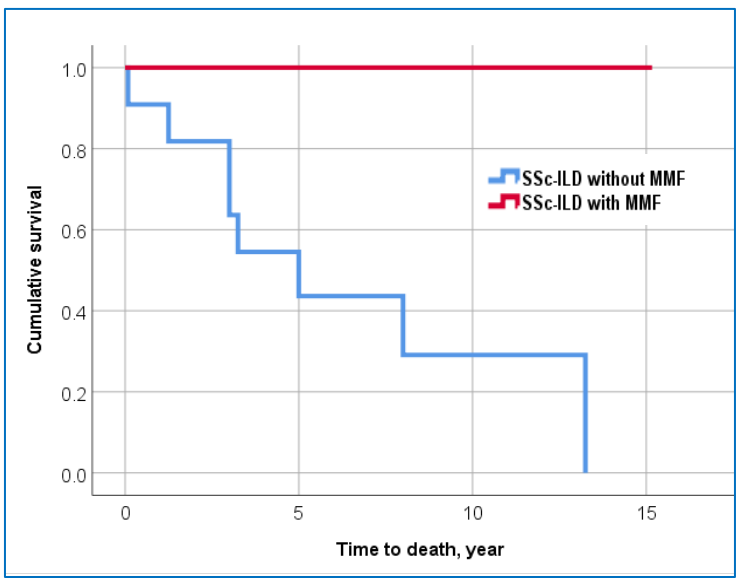


Figure 1. Cumulative survival rates at 2, 5 and 10 years following diagnosis of interstitial lung disease in SSc as shown by Kaplan-Meier curves (p<0.001).

## CONCLUSION

The majority of patients with SSc-ILD developed progressive lung disease. This study provides real-world data suggesting the effectiveness of MMF in the treatment of SSc-ILD and its influence in patient survival. Therefore, it would be prudent to initiate early treatment with MMF in patients with ILD, regardless of the extent of lung involvement. Nevertheless, studies with greater number of subjects and longer follow-up are essential to confirm the observations.

Disclosure of interests: None declared