Table: Mean age, duration of disease and age and follow up.

Parameter	Mean	SD
Mean age at enrollment (yrs)	43.24	11.70
Mean duration of disease (yrs)	10.02	6.86
Mean duration of ILD (yrs)	6.02	2.94
Mean duration of prospective follow up (yrs)	3.80	2.28

Figure 1. Change in FVC of 59 patients of SSc with ILD after 5 years of follow up

Figure 2. Change in FVC of 46 patients of SSc with ILD taking MMF after 5 years of follow up

Conclusion: This prospective study was carried out with aim to determine progression of ILD in patients with systemic sclerosis with ILD over period of 5 years. In our study, there was no statistically significant change of FVC over period of time. Worsening of lung function with deterioration in FVC >10% was seen in only 13 patients (22%) ($P < 0.01$). Our findings are similar to study done by Le Gouellec et al in which 75 patients with scleroderma and ILD were followed over period of 72 months of which 26% patients showed significant worsening of FVC ($\geq 10\%$).¹ In our study there statistically significant decline in DLCO and increase in PAH over period of 5 years ($p > 0.001$). In study carried out by Le Gouellec et al studying Scleroderma/ILD, there was significant decline in DLCO over period of 72 months of follow up.¹ In study done by Stephen C. Mathai et al, there was progressive increase in PAH when patients with scleroderma and ILD with PAH were followed up for period of 4 years.²

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ALL-CAUSE AND CAUSE-SPECIFIC MORTALITY TREND IN PATIENTS WITH SYSTEMIC SCLEROSIS: A POPULATION-BASED STUDY

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Background: Systemic sclerosis (SSc) is a disease with one of the highest mortality rates among connective tissue diseases with 5-year survival ranging from 68-88%[1]. Recent research has shown an improvement in survival for some rheumatic diseases at the population level [2, 3]. Contemporary data on mortality trends in SSc is scarce, especially at the general population level.

Objectives: 1) To investigate all-cause and cause-specific mortality in patients with newly diagnosed systemic sclerosis (SSc) compared to the general population.

2) To assess mortality trends between 2 calendar time periods, 1997–2004 and 2005–2012

Methods: Using an administrative health database, we compared all patients with new diagnosis of SSc with non-SSc individuals randomly selected from the general population matched for sex, age, and time of entry into the study. The study cohorts were divided into 2 groups based on the year of diagnosis ("early cohort [1997–2004] and "late cohort" [2005–2012]). The outcome was death (all-cause and cause specific including cancer, cardiovascular disease [CVD], infections, renal disease, pulmonary disease and other causes) during the follow-up period. Hazard ratios (HR) were estimated using Cox proportional hazards models, first adjusted for age, sex, and entry cohort time and then adjusted for selected baseline covariates (health care resource utilization, medications, pre-existing comorbidities, and socioeconomic status), based on a purposeful selection algorithm.

Results: We included 874 patients with SSc (83.6% female and mean age of 54.19 in the early cohort, and 81.7% female and mean age of 55.32 in the late cohort) and 8,740 non-SSc individuals in this study, contributing 4,083.8 and 47,515.8 person-years of follow-up, respectively. We observed 168 deaths in the SSc cohort, and 504 deaths occurred in the non-SSc cohort during the follow-up. Overall, the age-, sex-, and entry time-adjusted all-cause mortality HR in the SSc cohort was 4.47 (95% CI 3.72–5.37), and the fully adjusted remained significant [HR=2.87 (95% CI 2.35–3.50)]. Moreover, there was excess mortality due to CVD-related causes in the age-, sex-, and entry time-adjusted model (HR=2.65 [95% CI 1.68 – 4.18]), but it did not persist in the fully adjusted model (HR= 1.52 [95% CI 0.93–2.48]). There was no significant difference in other cause-specific mortalities, including cancer mortality (HR=0.91 [95% CI 0.56–1.49]). As a consequence of the residual disclosure policy of Population Data BC to ensure patient confidentiality, we were not allowed to report on the deaths due to infection, renal or pulmonary disease, as <5 deaths were associated with each outcome. There was no improvement in all-cause mortality between the early and late cohort (fully adjusted HR= 3.73 and 95% CI 2.49–5.59; HR 2.51 and 95% CI 1.88–3.34, respectively).

Conclusion: This population-based study showed that people with SSc have a 3-fold increase risk of death when compared with the general population. The risk of death has not improved over time and this calls for further research to close this gap.

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