Results: The cohort included 179 patients with SSc-ILD. Mean age at ILD diagnosis was 57.8 years. There was a female predominance (73%). The mean length of follow-up after ILD diagnosis was 4.3 years (SD 4.0). Based on high resolution chest CT (or biopsy when available), 147 (83%) were characterised as nonspecific interstitial pneumonia (NSIP), 31 (17%) as usual interstitial pneumonia (UIP), and 1 as unclassifiable ILD. A history of smoking was noted in 49%, and this was not associated with ILD pattern (former/current 46% in NSIP vs. 61% in UIP, p=0.11). Pulmonary hypertension (PHTN) was noted in 66 (49%) at baseline, and this did not differ between ILD subtypes. 84% had limited cutaneous SSc, 9% had diffuse cutaneous, and 7% SSc sine scleroderma. SSc specific serologies (i.e., SCL-70, centromere, and/or RNA Pol III) were positive in 73 (43%) patients, somewhat more common in NSIP than UIP (47% vs 26%; p=0.034). During a median of 3.2 years of follow-up, 65 patients died. SSc-ILD patients with UIP had a higher mortality than those with NSIP (hazard ratio: 2.27; 95% CI: 1.03–4.97). Other risk factors for progression included baseline DLCO (p<0.001), FVC (p<0.001) and PHTN (p=0.012). All 3 models had comparable discrimination (c=0.72, 0.72, and 0.70, respectively). Figure 1 shows the differential mortality based on the GAP and SADL staging systems. (Note the staging in the GAP and ILD-GAP models are identical.) Regarding calibration, the ILD-GAP model underestimated mortality (SMR: 1.5; 95% CI: 1.05–2.14). Calibration was acceptable for SADL (SMR: 0.77; 95% CI: 0.54–1.10) and GAP (SMR: 0.90; 95% CI: 0.63–1.29). The SADL model overestimated mortality in Stage III ILD.

Conclusions: The ILD-GAP model underestimated mortality, and the SADL model overestimated mortality in certain subgroups. However, the GAP model performed well in this cohort, providing the best prognostic information for SSc-ILD.

REFERENCES:

Disclosure of Interest: None declared