**INTRODUCTION**

Prospective Registry of Early Diffuse Cutaneous Systemic Sclerosis

- Early diffuse cutaneous systemic sclerosis (dcSSc) carries a high morbidity and mortality, predominantly due to internal organ involvement.[1]
- Ongoing international cohorts have predominantly included prevalent patients with both limited cutaneous and dcSSc.
- The Prospective Registry of Early Systemic Sclerosis (PRESS) is an ongoing observational registry of early dcSSc that is being conducted at 12 different Scleroderma Centers.
- The purpose of the registry to understand the current management of dcSSc and trends in morbidity and mortality in current era.
- Data is entered in the RedCap and housed at the University of Michigan.

**METHOD**

- Inclusion criteria include a diagnosis of dcSSc and a disease duration of ≤ 5 years calculated from the date of onset of first non-Raynaud’s phenomenon.
- Organ involvement was defined as new involvement or worsening during follow up visits—
  1. absolute increase in mRSS of ≥ 10 units or ≥ 25%
  2. an absolute decline of FVC % of ≥ 10% or ≤ 83.3%
  3. Pulmonary Hypertension on right heart catheterization;
  4. LVEF of ≤ 45% on echo;
  5. Cardiac arrhythmia;
  6. all-cause mortality.
- We reported number of participants experiencing organ involvement during the course of the study via frequency tables and characterized the distribution of events through plots of the cumulative proportion of participants experiencing each event by time.
- Confidence intervals in the cumulative proportion plots were derived via bootstrap method.

**RESULTS**

**OBJECTIVE**

Time to Event Visceral Organ Involvement and All-Cause Mortality

- The purpose of this study was to investigate the time to event of new onset of visceral organ involvement and all-cause mortality in early dcSSc participants in an ongoing US multicenter registry (PRESS).

**RESULTS**

- The cohort consisted of 239 participants at baseline with median follow-up of 414 days (IQR=112-922 days).
- The baseline mean age was 50.1 ± 14.3; 71.1% were female; 76.2% were white; Mean baseline mRSS was 21.31 ± 9.62.
- Mean disease duration was 1.21 years; Mean HAO-DI score was 0.82 (Table 1).
- During the first year of the study, 82.0% were on immunomodulatory therapy.
- At any time during the course of the study, 83.3% were on immunomodulatory therapy.[2] (Table 2.)

**CONCLUSION**

- Despite 83.3% on immunomodulatory therapy during the course of the study, a high proportion of early dcSSc patients experience worsening of mRSS and lung function and mortality within 2 years of dcSSc diagnosis.
- There are ongoing vascular-predominant complications over the course of the disease, consistent with published literature.[3]
- This data supports an ongoing need to identify novel therapies for dcSSc.

**REFERENCES**