classification criteria, in annual follow-up (for a total of 165 patients/year) with Pulmonary Function Tests (PFTs), Health Assessment Questionnaire - Disability Index (HAQ-DI), Scleroderma Health Assessment Questionnaire (sHAQ) and Cochin Hand Function Score (CHFS). Hand disability index was assessed by CHFS and global disability index was assessed by HAQ and sHAQ. Patient reported arthritis activity was assessed by Visual Analog Scale for Arthritis Activity (VASA). Based on the median of VASA3, patients were classified in two groups and the evaluation of global and hand disability index was performed for each group. Furthermore, we assessed the correlation between the change of VAS and the modification of disability scores (sHAQ, sSHAQ, CHFS) over 12 months of follow-up. Following analysis of distribution, Spearman or Pearson Test were used to determine correlation coefficients, as appropriate (Prism 7).

Results: The median disease duration was 5 years (IQR 3-10). The median of VAS3 was 35 (IQR 2 - 66). In patients with VAS3 ≤35 the HAQ-DI median were 0.625 (IQR 0.25 - 1.5) and 1.75 (IQR 1.125-2.25) respectively, (p<0.0001); the sHAQ medians were 0.628 (IQR 0.255 - 1.114) and 1.701 (IQR 1.234-2.059), respectively, (p<0.0001); and the CHFS medians were 4 (IQR 0 – 19) and 28 (IQR 10 – 46) respectively, (p<0.0001). A significant correlation was observed between VAS3 and HAQ (r= 0.463, p<0.0001), sSHAQ (r=0.651, p<0.0001), CHFS (r=0.497, p<0.0001); between ∆VAS3 and ∆SHAQ (r=0.493, p<0.0001). Compared to SSc-PAH pts without ILD (n=128), pts with PAH + ILD (n=92) had worse functional status (NYHA FC III: 69.6% vs. 55.5% in SSc-PAH pts without ILD; P=0.036), lower mean FVC (79.5±21.9 vs. 86.2±18.6) and lower DLCO (39.4±17.0 vs. 49.1±17.9) both P<0.001. Five-year survival rate was 35% in SSc-PAH pts with ILD vs. 45% in SSc-PAH without ILD (P=0.444 [figure 1]).

Conclusion: PAH has a profound impact on functional status, pulmonary function and right ventricle function of SSc patients, independently of presence of ILD. Despite the deleterious effect of functional status and pulmonary function, in pts with SSc and PAH, presence of concomitant ILD has no impact on 5-year survival.