**SUPPLEMENTARY DATA**

**Supplementary Table S1. Baseline characteristics of European Scleroderma Observational Study cohort (n=326)**

|  |  |  |
| --- | --- | --- |
| Characteristic |  | Missing atbaseline, no. (%) |
| Age, years, median (IQR) | 52.3 (43-60.8) | 0 (0) |
| Female, n (%) | 233 (71.5%) | 0 (0) |
| Months since onset of skin thickening, median (IQR) | 11.9 (7-21) | 18 (5.5) |
| Months since onset of first non-Raynaud's manifestation, median (IQR) | 14.7(9.5-24.4) | 6 (1.8) |
| Months since onset of Raynaud's phenomenon, median (IQR) | 20.5 (11.7-34.5) | 22 (6.7) |
| Previous immunosuppressant use, n (%)a | 26 (8) | 0 (0) |
| Current or previous steroid use, n (%) | 141 (43.8) | 4 (1.2) |
| Current smoker, n (%) | 57 (18.3) | 14 (4.3) |
| Previous history of cancer, n (%) | 20 (6.2) | 4 (1.2) |
| Caucasian, n (%) | 272 (83.4) | 0 (0) |
| mRSS, 0-51, median (IQR) | 21 (16-27) | 0 (0) |
| Haemoglobin, g/l, median (IQR) | 128 (118-137) | 13 (4) |
| White blood count, x109/l, median (IQR) | 8 (6.8-9.9) | 14 (4.3) |
| Platelets x109/l, median (IQR) | 298 (253-358) | 15 (4.6) |
| ESR, mm/hr, median (IQR) | 18 (8-34) | 77 (23.6) |
| CRP, mg/l, median (IQR) | 5 (2.1-11.8) | 90 (27.6) |
| Anti-topoisomerase (anti-Scl70), n (%) | 126 (39.5) | 7 (2.1) |
| Anti-RNA polymerase III, n (%) | 50 (19.1) | 64 (19.6) |
| Anticentromere, n (%) | 22 (7) | 12 (3.7) |
| *Organ involvement* |  |  |
| Pulmonary fibrosis, n (%) | 47 (14.4) | 0 (0) |
| FVC, % predicted, median (IQR) | 89 (75-102) | 19 (5.8) |
| DLCO, % predicted, median (IQR) | 64 (50-78) | 35 (10.7) |
| Pulmonary hypertension, n (%) | 26 (8) | 1 (0.3) |
| sPAP or RVSP, mmHg, median (IQR)b | 29 (21-34) | 124 (38) |
| Cardiac involvement, n (%) | 39 (12) | 2 (0.6) |
| Renal involvement, n (%) | 32 (9.8) | 0 (0) |
| eGFR, ml/min, median (IQR) | 85 (60-90) | 92 (28.2) |
| Renal crisis, n (%) | 16 (4.9) | 0 (0) |
| Plasma creatinine, μmol/l, median (IQR) | 65 (55-79.5) | 70 (21.5) |
| Any GI involvement, n (%) | 113 (34.7) | 0 (0) |
| Muscle involvement, n (%) | 31 (9.5) | 0 (0) |
| Current digital ulcers, n (%) | 55 (16.9) | 0 (0) |

Adapted with permission from BMJ Publishing Group Limited [1] Herrick AL, Pan X, Peytrignet S, et al. An observational study of treatment outcome in early diffuse cutaneous systemic sclerosis - European Scleroderma Observational Study (ESOS). Ann Rheum Dis doi: 10.1136/annrheumdis-2016-210503. [Epub ahead of print]. aOf the 26 patients who had previously received immunosuppressant therapy, in two patients this was for cancer. b86 patients had a sPAP/RVSP value assumed to be normal and thus not measured. If those cases are omitted, only 38 values of sPAP/RVSP are missing (11.7%). Median values are 'falsely' high because calculation omits unmeasured (normal) values. DLCO: Carbon monoxide diffusing capacity; eGFR: Estimated glomerular filtration rate; FVC: Forced vital capacity; GI: Gastrointestinal; HAQ-DI: Health Assessment Questionnaire - Disability Index; mRSS: modified Rodnan skin score (17 sites); RVSP: Right ventricular systolic pressure; sPAP: Systolic pulmonary artery pressure.

**Supplementary Table S2. Correlations between 12-month changes**

|  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
|  | mRSS | mRSS hand and fingers | FVC | DLCO | eGFR | HAQ-DI | CHFS | FACIT fatigue | SF36PCS | SF36 MCS | sHAQ Pain VAS |
| mRSS | 1.00 |  |  |  |  |  |  |  |  |  |  |
| mRSS hand and fingers | 0.66 [<0.0001] (226) | 1.00 |  |  |  |  |  |  |  |  |  |
| FVC | -0.16 [0.031] (184) | -0.24 [0.001] (180) | 1.00 |  |  |  |  |  |  |  |  |
| DLCO | -0.14 [0.071] (165) | -0.14 [0.080] (161) | 0.43 [<0.0001] (173) | 1.00 |  |  |  |  |  |  |  |
| eGFR | -0.06 [0.470] (143) | -0.09 [0.283] (140) | 0.09 [0.320] (117) | 0.11 [0.270] (108) | 1.00 |  |  |  |  |  |  |
| HAQ-DI | 0.40 [<0.0001] (175) | 0.27 [<0.0005] (173) | -0.21 [0.012] (145) | -0.21 [0.019] (127) | 0 [0.979] (121) | 1.00 |  |  |  |  |  |
| CHFS | 0.19 [0.035] (126) | 0.24 [0.006] (125) | -0.09 [0.370] (103) | 0 [0.964] (91) | -0.17 [0.099] (97) | 0.57 [<0.0001] (131) | 1.00 |  |  |  |  |
| FACIT fatigue | -0.17 [0.023] (171) | -0.14 [0.073] (170) | 0.11 [0.176] (144) | 0.09 [0.330] (125) | 0.12 [0.205] (119) | -0.53 [<0.0001] (176) | -0.50 [<0.0001] (129) | 1.00 |  |  |  |
| SF36 PCS | -0.27 [<0.0005] (177) | -0.10 [0.196] (175) | 0.05 [0.585] (147) | 0.03 [0.703] (129) | 0.19 [0.039] (122) | -0.48 [<0.0001] (182) | -0.33 [<0.0005] (132) | 0.38 [<0.0001] (178) | 1.00 |  |  |
| SF36 MCS | -0.01 [0.878] (177) | 0 [0.953] (175) | 0 [0.993] (147) | -0.07 [0.418] (129) | 0.02 [0.854] (122) | -0.13 [0.091] (182) | -0.19 [0.026] (132) | 0.30 [<0.0001] (178) | -0.26 [<0.0005] (184) | 1.00 |  |
| sHAQ Pain VAS | 0.13 [0.090] (175) | 0.09 [0.258] (173) | -0.02 [0.793] (145) | -0.18 [0.040] (128) | -0.15 [0.105] (121) | 0.32 [<0.0001] (180) | 0.31 [<0.0005] (132) | -0.39 [<0.0001] (176) | -0.45 [<0.0001] (182) | -0.09 [0.210] (182) | 1.00 |

For each pair of variables, Spearman’s ρ, its significance p-value [between brackets] and sample size for computation (n) are shown. DLCO: Carbon monoxide diffusing capacity; eGFR: Estimated glomerular filtration rate; FVC: Forced vital capacity; HAQ-DI: Health Assessment Questionnaire - Disability Index; mRSS: modified Rodnan skin score (17 sites)

**Supplementary Figure S1. 12-month changes in disability indicators**

 ****

**Supplementary Figure S2. 12-month changes in the disability and fatigue indicators with respect to their baseline values**

****

**Supplementary Figure S3. Values of disability indicators at baseline, 12 months and 24 months**

****

For each disability indicator, a ‘lasagna’ plot is constructed by stacking individual lines on top of each other, each line representing the evolution of disability for a patient from baseline to 24 months. Each line changes colour during this period as disability evolves, according to the colour scale shown at the right of each plot.

**References**

1. Herrick AL, Pan X, Peytrignet S, et al. An observational study of treatment outcome in early diffuse cutaneous systemic sclerosis - European Scleroderma Observational Study (ESOS). *Ann Rheum Dis* doi: 10.1136/annrheumdis-2016-210503. [Epub ahead of print].