|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | Total  (n = 58) | mPAP ≥ 25 mmHg  (n = 24) | 25 mmHg > mPAP > 20 mmHg  (n = 9) | 20mmHg ≥ mPAP  (n = 25) |
| Organ involvement  　Interstitial lung disease, n (%)  　Raynaud`s phenomenon, n (%)  　GERD, n (%)  　Renal crisis, n (%) | 47 (81.0%)  53 (91.3%)  27 (46.5%)  4 (6.8%) | 20 (83.3%)  21 (87.5%)  0 (0.0%)  1 (4.1%) | 7 (77.7%)  8 (88.8%)  6 (66.6%)  0 (0.0%) | 20 (80.0%)  24 (96.0%)  21 (84.0%)  3 (12.0%) |
| Disease duration, median [min-max] | 8 [0-31] | 10 [1-27] | 10 [1-31] | 8 [0-31] |
| Disease subtype  Diffuse, n (%)  Limited, n (%)  Mixed/overlap, n (%) | 18 (31.0%)  18 (31.0%)  22 (37.9%) | 7 (29.1%)  6 (25.0%)  11 (45.8%) | 1 (11.1%)  3 (33.3%)  5 (55.5%) | 10 (40.0%)  9 (36.0%)  6 (24.0%) |
| Immunosuppressive therapy  PSL, n (%)  IVCY, n (%)  AZT, n (%) | 39 (67.2%)  16 (27.5%)  4 (6.8%) | 18 (75.0%)  8 (33.3%)  3 (12.5%) | 7 (77.7%)  3 (33.3%)  0 (0.0%) | 14 (56.0%)  5 (20.0%)  1 (4.0%) |

Supplementary Table 1. Patient characteristics

\*GERD, gastroesophageal reflux disease; PSL, prednisolone; IVCY, intra venous cyclophosphamide; AZT, azathioprine.