|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | 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.