

Supplemental Table II: Baseline Pulmonary Function Test, Imaging and Echocardiographic Characteristics of the Study

Cohort

| Characteristics | All PAH (N=576) | Subgroup[†] (N=282) |
|--|------------------------------|---|
| Pulmonary function test, % predicted | (N=466) | (N=233) |
| • Forced expiratory volume (n) | 76 ± 19 (457) | 80±18 (230) |
| • Forced vital capacity (n) | 78 ± 18 (455) | 84±17 (230) |
| • FEV1/FVC (n) | 89 ± 13 (311) | 89±13 (149) |
| • Total lung capacity (n) | 88 ± 20 (419) | 94±17 (206) |
| • Diffusion lung capacity of carbon monoxide (n) | 55 ± 23 (409) | 63±22 (197) |
| CT chest[‡] | (N=306) | (N=132) |
| • Normal, n (%) | 213 (70) | 111 (84) |
| • Interstitial lung disease, n (%) | 59 (19) | 7 (5) |
| • Thromboembolism, n (%) | 5 (2) | 3 (2) |
| • Other, n (%) | 29 (9) | 11 (9) |
| Ventilation/Perfusion scan | (N=419) | (N=221) |

| | | |
|--|----------------|----------------|
| • Normal, n (%) | 321 (77) | 164 (74) |
| • Mottled, n (%) | 85 (20) | 50 (23) |
| • Intermediate-high probability, n (%) | 13 (3) | 7 (3) |
| Echocardiography | (N=510) | (N=248) |
| • Ejection Fraction | 60±10 (284) | 60±9 (127) |
| • Left atrial enlargement, n (%) | 112 (22) | 36 (15) |
| • Right atrial enlargement, n (%) | 437 (86) | 222 (91) |
| • Right ventricular hypertrophy, n (%) | 232 (45) | 118 (48) |
| • Tricuspid regurgitation, m ² /sec (n) | 4.1±0.9 (246) | 4.2±0.8 (122) |

PAH = pulmonary arterial hypertension, FEV1 = forced expiratory volume in one second, and FVC = forced vital capacity

† = subgroup includes patients with idiopathic, familial, and anorexigen-associated pulmonary arterial hypertension

‡ = high resolution computerized tomographic scan of the chest