

Supplemental Table S1: Baseline characteristics of patients initiated with PAH-targeted therapy (PAH-TT) compared to those of patients who did not receive any PAH-TT.

	Patients not receiving any PAH-TT (n=18)	Patients initiated with PAH-TT (n=97)	P value*
WHO/NYHA (II-III : IV)	13 : 5	77 : 20	0.49
Radiologic stage (non IV : IV)	3 : 15	29 : 68	0.25
6MWD, m	357 ± 167	311 ± 137	0.06
RAP, mmHg	7 ± 5	7 ± 5	0.76
mPAP, mmHg	39 ± 10	47 ± 9	<0.001
Cardiac index, L/min/m ²	2.8 ± 0.8	2.6 ± 0.8	0.31
PVR, WU	6.4 ± 2.7	9.4 ± 4.4	<0.001
FVC, %	63 ± 21	64 ± 20	0.65
KCO, %	57 ± 23	53 ± 23	0.46

Abbreviations: PAH-TT: Pulmonary arterial hypertension-targeted therapy; WHO/NYHA FC: World Health Organisation/New York Heart Association functional class; 6-MWD: 6-minute walk distance; RAP: right atrial pressure; mPAP: mean pulmonary artery pressure; PVR: pulmonary vascular resistance; FVC: forced vital capacity; KCO: Transfer coefficient for carbon monoxide (corrected for lung volume).

*: Unpaired Student' t-test or Chi-square test as appropriate

Supplemental Table S2: Effect of long-term therapy on 6-minute walk distance according to baseline forced vital capacity

6-MWD, m (mean±SD)	n	Baseline	First follow up visit 4.5 months [4.0 – 6.0]	Last follow up visit 28.0 months [18.0 – 54.0]	p-value (ANOVA)
Overall	50	334±118	330±118	298±153	0.29
FVC ≤ 50% of predicted	13	300±117	311±131	275±156	0.39
FVC > 50% of predicted	37	344±118	361±126	323±139	0.09

Abbreviations: 6-MWD: 6-minute walk distance; FVC: Forced vital capacity