

SUPPLEMENTARY DATA

Table S1. Baseline Characteristics at start standardized PAH-specific treatment protocol

<i>Variables</i>	<i>All Patients</i>	<i>1 QoL questionnaire</i>	<i>>1 QoL questionnaire</i>	<i>p</i>
Number	61	22	39	
Age, mean \pm SD, y	49 \pm 13	52 \pm 15	47 \pm 12	0.047
Male, n (%)	26 (43)	9 (41)	17 (44)	0.839
Down syndrome, n (%)	23 (38)	4 (18)	19 (49)	0.018
Eisenmenger syndrome, n (%)	48 (79)	13 (59)	35 (90)	0.005
Body Mass index, mean \pm SD, kg	24 \pm 5	25 \pm 6	24 \pm 5	0.494
NYHA class III or IV, n (%)	31 (51)	16 (73)	15 (39)	0.025
SF36 PCS, mean \pm SD	35 \pm 9	35 \pm 11	35 \pm 8	0.930
SF36 MCS, mean \pm SD	49 \pm 12	48 \pm 13	50 \pm 11	0.426
Medication				
-bosentan, n (%)	61 (100)	22 (100)	39 (100)	1.000
-diuretics, n (%)	22 (39)	7 (39)	15 (39)	0.975
Cardiac surgery, n (%)	17 (28)	9 (41)	8 (21)	0.006
-Age surgery, median (range), y	4 (1-75)	4 (2-75)	6 (1-66)	0.867
Saturation in rest, median (IQR), %	87 (80-93)	88 (80-96)	85 (80-91)	0.256
NT-pro-BNP, median (IQR), ng/L	466 (218-1209)	424 (161-1623)	556 (244-1127)	0.500
6-MWD, mean \pm SD, m	377 \pm 124	401 \pm 125	364 \pm 123	0.279
TAPSE, median (IQR), mm	19 (16-23)	17 (15-22)	19 (16-23)	0.293
SPAP, median (IQR), mmHg	89 (71-98)	81 (49-91)	91 (76-103)	0.016
Shunt Type				
-pre tricuspid, n (%)	11 (18)	5 (23)	6 (15)	
-post tricuspid, n (%)	27 (44)	9 (41)	18 (46)	0.769
-complex*, n (%)	23 (38)	8 (36)	15 (39)	

Abbreviations: SF36 PCS, short form health survey of physical health; SF36 MCS, short form health survey of mental health; SD, standard deviation; IQR, interquartile range; QoL, quality of life; 6-MWD, six-minute walk distance; SPAP, systolic pulmonary artery pressure; TAPSE, tricuspid annular plane systolic excursion; NT-pro-BNP, N-terminal pro-brain natriuretic peptide; NYHA functional class, New York Heart Association functional class.

* Complex shunts included atrioventricular septal defect and univentricular heart.