Supplemental Tables

Supplemental Table 1. Patients' baseline characteristics

Case #	Sex	BSA (m^2)	Age at enrollment (years)	Medications at baseline	Etiology of PAH	Genetic test	REVEAL 2.0 risk score[1]	Risk group*
1	F	1.61	55	None	CHD-PAH	No mutations	6	Low
2	F	1.58	46	Macitentan 10 mg Tadalafil 40 mg Beraprost 120 μg	CTD-PAH	Not tested	6	Low
3	F	1.73	34	Bosentan 250 mg Tadalafil 40 mg Beraprost 360 μg	IPAH	No mutations	3	Low
4	F	1.54	42	Bosentan 250 mg Beraprost 120 μg	IPAH	No mutations	6	Low
5	F	1.39	48	Tadalafil 20 mg Beraprost 360 μg	CTD-PAH	Not tested	10	High
6	M	1.58	59	Beraprost 60 μg	HPAH	*BMPR2* mutation	14	High
7	F	1.64	54	None	IPAH	No mutations	9	High

8	M	1.79	52	Sildenafil 180 mg	CTD-PAH	Not tested	6	Low
9	F	1.62	39	None	CHD-PAH	No mutations	7	Intermediate
10	M	1.41	71	None	CTD-PAH	No mutations	8	Intermediate
11	M	1.97	21	Macitentan 10 mg Tadalafil 80 mg Beraprost 360 μg	IPAH	No mutations	1	Low
12	F	1.43	37	None	CTD-PAH	No mutations	10	High
13	F	1.64	17	None	HPAH	*BMPR2* mutation	11	High
14	F	1.65	22	None	CTD-PAH	No mutations	12	High
15	M	1.78	49	None	IPAH	No mutations	7	Intermediate
16	M	1.74	45	Bosentan 250 mg Tadalafil 20 mg Beraprost 120 μg	HPAH	*BMPR2* mutation	15	High
17	F	1.30	19	None	IPAH	*RNF213* mutation	9	High
18	F	1.35	20	None	HPAH	*BMPR2* mutation	11	High
19	F	1.48	40	Bosentan 125 mg	CHD-PAH	Not tested	5	Low
20	M	1.40	23	None	IPAH	No mutations	7	Intermediate
21	F	1.71	36	None	HPAH	*ACVRL1* mutation	11	High

						BMPR2 mutation		
22	F	1.53	23	None	CHD-PAH	No mutations	6	Low
23	F	1.36	44	None	IPAH	No mutations	3	Low
24	F	1.61	28	None	IPAH	No mutations	9	High
25	F	1.43	18	None	CTD-PAH	Not tested	10	High
26	F	1.36	23	None	IPAH	No mutations	5	Low

*Risk group was defined by three-category REVEAL 2.0 risk score.[1] The high-risk group was defined as patients with a predicted 1-year survival rate of <90% (REVEAL 2.0 risk score ≥9), the intermediate-risk group was defined as patients with a predicted 1-year survival rate of 90% to <95% (REVEAL 2.0 risk score =7 or 8), and the low-risk group was defined as patients with a predicted 1-year survival rate of ≥95% (REVEAL 2.0 risk score ≤6).

ACVRL1, activin A receptor-like kinase 1; *BMPR2*, bone morphogenetic protein receptor type 2; BSA, body surface area; CHD, congenital heart disease; CTD, connective tissue disease; F, female; HPAH, heritable pulmonary arterial hypertension; IPAH, idiopathic pulmonary arterial hypertension; M, male; PAH, pulmonary arterial hypertension; *RNF213*, ring finger protein 213.

Supplemental Table 2. Maximum tolerated dose of each drug

Drugs	Maximum tolerated dose (per day)	Patients, n (%)
Macitentan	10.0 mg	25 (96.2)
Riociguat	3.0 mg	2 (7.7)
	7.5 mg	24 (92.3)
Selexipag	0.4 mg	2 (7.7)
	0.8 mg	1 (3.8)
	1.6 mg	2 (7.7)
	2.0 mg	1 (3.8)
	3.2 mg	20 (76.9)

In one patient, macitentan could not be tolerated because 10 mg of macitentan caused suspected myelosuppression.

Supplemental Table 3. Clinical parameters in low/intermediate risk and high risk at baseline and follow-up

	Low-/Intermediate-risk group (n=9)		High-risk group (n=9)	
Variable	Baseline	Follow-up	Baseline	Follow-up
Mean RAP, mmHg	5.0 (4.0–7.0)	4.0 (3.0–4.0)	7.5 (6.5–13.3)	5.0 (4.0–-6.0)
Mean PAP, mmHg	52.0 (44.0–66.0)	35.0 (28.0–40.0)	63.0 (49.5–64.5)	41.0 (40.0–49.0)
PAWP, mmHg	7.0 (6.0–8.0)	9.0 (8.0–11.0)	8.0 (5.5–9.3)	9.0 (8.0–11.0)
CO, L/min	4.5 (2.9–5.2)	6.4 (5.8–7.5)	2.3 (2.1–3.5)	5.1 (4.7–6.0)
CI, L/min/m^2	2.7 (2.1–3.6)	4.3 (3.4–4.7)	1.7 (1.3–2.1)	3.6 (3.1–4.2)
PVR, WU	11.3 (7.8–13.4)	3.9 (2.5–4.8)	20.2 (14.5–25.5)	6.8 (3.8–8.8)

Data are expressed as median (interquartile range). CI, cardiac index; CO, cardiac output; PAP, pulmonary arterial pressure; PAWP, pulmonary arterial wedge pressure; PVR, pulmonary vascular resistance; RAP, right atrial pressure; WU, Wood units.

Supplemental Reference

1. Benza RL, Gomberg-Maitland M, Elliott CG, et al. Predicting survival in patients with pulmonary arterial hypertension: The REVEAL risk score calculator 2.0 and comparison with ESC/ERS-based risk assessment strategies. *Chest.* 2019; 156: 323-337.