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Table E1. Baseline Characteristics of Patients according to Univariate Cox Regression

Characteristic	All Patients (*n* = 256)	Alive Patients (*n* = 163)	Dead Patients (*n* = 93)	*P* Value for Alive vs Dead Patients	Hazard Ratio	95% CI	*P* Value for Hazard Ratio
Age (y)	67 (52–75)	63 (48–71)	72 (65–78)	<.001	1.034	1.021, 1.047	<.001
Sex				.013	1.736	1.153, 2.614	.008
No. of women	143	101	42				
No. of men	113	62	51				
Body surface area (m^2)*	1.78 ± 0.21	1.87 ± 0.22	1.84 ± 0.24	.37	0.597	0.245, 1.454	.26
Race†				.168			
African	9	8	1		…	…	…
Asian	5	4	1		…	…	…
Caribbean	13	10	3		…	…	…
Caucasian	213	127	86		2.789	1.288, 6.042	.009
Subcontinental	15	12	3		…	…	…
Middle East	1	1	0		…	…	…
RVEDVI ($mL \cdot m^{-2}$)‡	105 (77–123)	96 (77–115)	110 (82–138)	.004	1.008	1.003, 1.013	.002
RVESVI ($mL \cdot m^{-2}$)‡	61 (40–84)	53 (37–75)	67 (46–98)	<.001	1.009	1.004, 1.014	<.001
RV ejection fraction (%)*	40 ± 14	42 ± 14	37 ± 12	<.001	0.975	0.961, 0.990	.001
SV/RVESV (%)+	0.65 (0.44–0.99)	0.74 (0.48–1.13)	0.58 (0.37–0.89)	.001	0.391	0.229, 0.669	<.001
6MWD (m)+§	258 (96–360)	302 (120–395)	180 (90–300)	<.001	0.997	0.995, 0.998	<.001
Hemodynamic parameters‡							
mPAP (mm Hg)	44 (32–54)	43 (31–54)	47 (36–55)	.09	1.009	0.995, 1.023	.20
Cardiac index	2.3 (1.9–2.9)	2.3 (1.9–2.9)	2.3 (1.9–2.9)	.78	0.963	0.775, 1.197	.73
PVR (U)	7.0 (3.9–11.2)	6.1 (3.2–11.0)	7.4 (5.2–11.6)	.08	1.023	0.988, 1.060	.20
mRAP	10 (6–14)	10 (6–13)	12 (8–16)	.003	1.065	1.026, 1.106	<.001
mRVEDP	10 (7–15)	10 (6–14)	12 (8–16)	.004	1.061	1.024, 1.100	.001
PCWP	12 (8–16)	11 (8–16)	13 (10–16)	.17	1.081	0.889, 1.314	.44
WHO functional class‖				<.001	2.105	1.505, 2.945	<.001
I	3	3	0				

II	43	36	7				
III	163	104	59				
IV	47	20	27				
Clinical classification				.422			
Group 1	88	54	34		1.038	0.810, 1.330	.77
Group 1.1	57	36	21		0.977	0.777, 1.230	.85
Group 1.2	3	1	2		1.187	1.018, 1.384	.03
Group 1.3	1	1	0		…	…	…
Group 1.4	25	16	9		0.981	0.791, 1.216	.76
Group 1′	2	0	2		1.009	0.999, 1.018	.07
Group 2	43	27	16		1.035	0.830, 1.291	.76
Group 3	15	2	13		1.423	1.239, 1.633	<.001
Group 4	106	78	28		0.681	0.512, 0.907	.009
Group 5	4	2	2		1.030	0.864, 1.229	.74

Note.—mPAP = mean pulmonary artery pressure, mRAP = mean right atrial pressure, mRVEDP = mean RV end-diastolic pressure, PCWP = pulmonary capillary wedge pressure, PVR = pulmonary vascular resistance, RVEDVI = indexed RV end-diastolic volume, RVESVI = indexed RV end-systolic volume, SV/RVESV = stroke volume divided by RV end-systolic volume, U = Woods units, WHO = World Health Organization, 6MWD = 6-minute walk distance.

* Values are means ± standard deviations (normal data).

† Hazard ratios were not calculated for groups with five or fewer deaths.

‡ Values are medians, with interquartile ranges in parentheses (nonnormal data).

§ Excluding 16 subjects who were unable to perform testing.

‖ Higher numbers indicate lower functional capacity. Group 1, pulmonary arterial hypertension; group 1.1, idiopathic; group 1.2, heritable; group 1.3, drug and toxin induced; group 1.4, associated with other conditions (except congenital heart disease); group 1′, peripheral veno-occlusive disease and/or pulmonary capillary hemangiomatosis; group 2, PH due to left-sided heart disease; group 3, PH due to lung diseases and/or hypoxia; group 4, chronic thromboembolic PH and other pulmonary artery obstructions; group 5, PH with unclear and/or multifactorial mechanisms.

Table E2. Baseline RV Volumetry and Hemodynamic and Functional Data in Patients with PH according to Diagnostic Subgroup

Parameter	Group 1						Group 2, LHD	Group 3, PH Lung	Group 4, CTEPH	Group 5, PH Miscellaneous
	Pulmonary Arterial Hypertension	IPAH	HPAH	DPAH	APAH	PVOD				
Age (y)	64 (44–73)	68 (46–74)	69 (61–77)	49	52 (42–70)	56 (48–64)	72 (60–77)	70 (69–80)	65 (51–72)	67 (63–72)

Sex										
No. of men	32	22	2	0	8	0	15	7	56	1
No. of women	54	35	1	1	17	2	28	4	50	3
6WMD (m)	243 (96–365)	215 (95–360)	120 (90–276)	309	308 (106–371)	146 (115–177)	180 (80–316)	276 (168–330)	297 (144–368)	45 (26–157)
WHO functional class*										
I	3	1	0	0	2	0	0	0	0	0
II	14	8	0	0	6	0	7	1	21	0
III	51	33	2	1	15	1	23	6	67	4
IV	16	13	1	0	2	1	5	4	18	0
Hemodynamic parameters										
mPAP (mm Hg)	49 (39–58)†	50 (42–59)‡	34 (34–38)	66	42 (31–50)	54 (54–55)	32 (27–43)	38 (33–44)	46 (35–54)†	49 (47–50)
mRAP (mm Hg)	9 (5–15)	10 (5–15)	7 (5–15)	28†	8 (6–15)	7 (5–8)	15 (12–17)‡	6 (5–9)‡	10 (6–13)‡	16 (14–19)
PCWP (mm Hg)	10 (7–14)†	10 (7–14)	8 (8–12)	15	10 (8–13)	9 (8–9)	17 (16–22)§	14 (11–17)	10 (8–13)†	19 (15–23)
PVR (U)	8.2 (5.3–12.2)‡	9.0 (5.6–13.9)§	9.1 (5.1–10.0)	20.4§	7.0 (2.8–10.7)	13.1 (11.0–15.2)	3.3 (2.0–5.9)	5.7 (3.2–6.6)	8.3 (4.5–12.1)†	7.8 (5.0–10.5)
RVEDP (mm Hg)	10 (6–16)	10 (6–16)	8 (7–10)	20	10 (6–14)	7 (6–8)	12 (8–16)	10 (8–12)	11 (7–14)	19 (14–23)‡
Cardiac index (L/min/m^2)	2.4 (2.0–3.0)§	2.4 (2.0–2.9)§	2.0 (1.9–5.4)§	1.5	2.8 (2.2–3.3)	2.4 (2.1–2.6)	2.4 (2.1–2.9)	2.5 (2.4–3.0)	2.2 (1.9–2.8)§	2.2 (1.7–2.6)
Heart rate (beats/min)	74 (66–85)	75 (68–86)	70 (58–72)	72	73 (63–84)‡	107 (98–115)	68 (60–80)	76 (66–79)	76 (67–87)§	75 (69–85)
Cardiovascular MR measurements										
RVEDVI (mL)	104 (77–124)	107 (75–129)	94 (78–175)	103	104 (88–120)	74 (71–76)	81 (74–109)	83 (73–110)	104 (80–131)+	87 (67–117)
RVESVI (mL)	63 (45–82)	67 (46–94)	45 (40–85)	51	61 (38–75)	47 (47–47)	44 (31–56)	49 (39–64)	70 (43–95)¶	41 (34–65)
RV ejection fraction (%)	38 (31–49)	34 (27–42)¶	51 (46–52)	50	48 (37–57)	36 (33–39)	50 (44–57)	44 (38–49)	34 (27–46)¶	49 (42–51)
SV/RVESV (%)	62 (44–94)	52 (37–72)¶	103 (88–107)	101	93 (58–132)	57 (49–64)	99 (80–132)	80 (60–96)	50 (37–86)¶	95 (78–104)
3D motion (PC)	9 (6–15)	9 (6–14)	11 (6–16)	15 (9–20)	9 (5–15)	9 (5–12)	11 (6–16)	10 (6–16)	10 (6–16)	11 (7–17)
Mortality rate (%)	39	37	67	0	36	100	28¶	92	26¶	50
No. of deaths	34/88	21/57	2/3	0/1	9/25	2/2	12/43	11/12	28/106	2/4

Note.—According to reference 1. Unless indicated otherwise, values are medians, with interquartile ranges in parentheses. Survival models were adjusted for RV ejection fraction. APAH = PH associated with another condition, CTEPH = chronic thromboembolic PH, DPAH = drug-and toxin-induced pulmonary arterial hypertension, HPAH = hereditary pulmonary arterial hypertension, IPAH = idiopathic PH, LHD = left-sided heart disease, PC = principal component units, PCWP = pulmonary capillary wedge pressure, PVOD = pulmonary veno-occlusive disease, PVR = pulmonary vascular resistance, mPAP = mean pulmonary artery pressure, mRAP = mean right atrial pressure, RVEDVI = indexed RV end-diastolic volume, RVEDP = RV end-diastolic pressure, RVESVI = indexed RV end-systolic volume, SV/RVESV = stroke volume divided by RV end-systolic volume, U = Woods units, WHO = World Health Organization, 6MWD = six-minute walk distance.

* Higher numbers indicate lower functional capacity.

† Significant, $P < .001$.

‡ Significant, $P < .01$.

§ Significant $P < .05$.

Table E3. Baseline Characteristics of Group 4 Patients Categorized according to Subsequent Pulmonary Endarterectomy

Variable	Pulmonary Endarterectomy	No Pulmonary Endarterectomy	*P* Value
Age (y)	64.6 (50.7–69.2)	64.9 (51.2–73.2)	.31
Sex			.36
No. of men	22	34	
No. of women	24	25	
WHO functional class			.09
I	0	0	
II	6	15	
III	30	36	
IV	10	8	
Hemodynamic parameters			
mPAP (mm Hg)	48.5 (42.3–55.8)	43.0 (29.5–52.0)	.01
mRAP (mm Hg)	10.0 (7.0–13.8)	10 (6.0–12.0)	.55
PCWP (mm Hg)	11.0 (9.3–14.8)	10.0 (7.5–12.0)	.22
PVR (U)	9.5 (5.4–12.3)	7.0 (3.9–11.7)	.09
RVEDP (mm Hg)	10.0 (8.0–15.0)	11.0 (7.0–13.5)	.46
Cardiac index (L/min/m^2)	2.1 (1.9–2.8)	2.2 (1.9–2.8)	.50
Heart rate (beats/min)	79 (68–87)	74 (65–87)	.36
Cardiovascular MR measurements			
RVEDVI (mL/m^2)	102.9 (77.4–120.7)	108.8 (84.5–135.0)	.24
RVESVI (mL/m^2)	68.5 (42.9–88.6)	74.4 (44.1–97.5)	.47
RV ejection fraction (%)	35.0 (27.3–44.2)	33.4 (26.5–47.9)	>.99
SV/RVESV (%)	53.7 (37.6–79.4)	50.1 (36.0–92.0)	>.99
Mortality rate			.10
No. of deaths*	8/46 (17)	19/59 (32)	
Hazard ratio	0.511 (0.230–1.134)	1	

Note.—Unless indicated otherwise, values are medians, with interquartile ranges in parentheses. mPAP = mean pulmonary artery pressure, mRAP = mean right atrial pressure, PCWP = pulmonary capillary wedge pressure, PVR = pulmonary vascular resistance, RVEDP = RV end-diastolic pressure, RVEDVI = indexed RV end-diastolic volume, RVESVI = indexed RV end-systolic volume, SV/RVESV = stroke volume divided by RV end-systolic volume, U = Woods units, WHO = World Health Organization.

* Numbers in parentheses are percentages.

Table E4. Baseline Characteristics of Patients with PH Who Could and Who Could Not Perform the 6-Minute Walk Test

Variable	Performed Test	Unable to Perform Test	*P* Value
Age (y)	67 (51–74)	69 (55–72)	.59
Sex			.13
No. of men	109	4	
No. of women	131	12	
WHO functional class			.01
I	3	0	
II	41	2	
III	157	6	
IV	39	8	
Hemodynamic parameters*			
mPAP (mm Hg)	46 (33–54)	36 (27–42)	.02
mRAP (mm Hg)	10 (7–14)	11 (6–15)	.97
PCWP (mm Hg)	12 (8–15)	14 (10–17)	.29
PVR (U)	7.0 (4.0–11.4)	5.2 (2.7–9.9)	.13
RVEDP (mm Hg)	10 (7–15)	12 (8–14)	.72
Cardiac index (L/min/m^2)	2.3 (1.9–2.9)	2.3 (1.8–2.9)	.77
Heart rate (beats/min)	75 (65–85)	72 (68–81)	.76
Cardiovascular MR measurements*			
RVEDVI (mL/m^2)	102 (77–124)	84 (68–111)	.20
RVESVI (mL/m^2)	62 (41–84)	44 (30–68)	.08
RV ejection fraction (%)	39 (30–49)	50 (38–60)	.04
SV/RVESV (%)	0.64 (0.42–0.97)	0.99 (0.61–1.50)	.04
Mortality rate (%)	35	50	.05
No. of deaths	85/240	8/16	

Note.—mPAP = mean pulmonary artery pressure, mRAP = mean right atrial pressure, PCWP = pulmonary capillary wedge pressure, PVR = pulmonary vascular resistance, RVEDP = RV end-diastolic pressure, RVEDVI = indexed RV end-diastolic volume, RVESVI = indexed RV end-systolic volume, SV/RVESV = stroke volume/RV end-systolic volume, U = Woods units, WHO = World Health Organization.

* Values are medians, with interquartile ranges in parentheses.

Reference

1. Simonneau G, Gatzoulis MA, Adatia I, et al. Updated clinical classification of pulmonary hypertension. J Am Coll Cardiol 2013;62(25 Suppl):D34–D41.