Supplement Table 1

Variables reported to be associated with outcome in paediatric PAH

Parameters	References	Unique cohorts	Reported cut off values (number of publications)
Clinical variables			
*Growth (W/H z-scores)	(1, 2)	3	W/H Z-score< -2 (1)
Age	(3-5)	3	
Heart Rate (unadjusted for age)	(6-8)	2	
Heart Rate variability	(9)	2	
Transcutaneous SO2	(10, 11)	2	
Blood pressure	(5)	1	
ECG RV hypertrophy	(12)	1	
Hemoptysis	(13)	1	
Functional Variables			
*WHO-FC	(2, 3, 5, 6, 14-18)	>3	I-II vs III-IV (4) I-III vs IV (2)
6MWD (>6yr)	(9-11, 14)	3	< 352 m (1)
Pediatric FC (+/-)	(19)	1	
CPET (pVO2, VE/VCO2, systRR)	(20, 21)	1	
Physical Activity	(22)	1	
Hemodynamic variables			
*PVRi	(3, 14, 23-25)	>3	> 20 WU*m2 (1) > 11 WU*m2 (1) > 13 WU*m2 (1) > 6 WU*m2 (1)
*mRAP	(3, 4, 10, 14, 23, 26)	>3	>5 mmHg (1) >7 mmHg (1) > 10 mmHg (1)
*Cardiac Index	(14, 26)	>3	< 2,0 L/min/m2
*AVT response	(14, 27, 28)	>3	Sitbon criteria no
*mPAP/mSAP	(5, 18, 25, 27, 29)	3	0.76 (1)
*PACi	(8, 23, 24, 30)	3	< 0.7 ml/mmHg/m2 (1) < 0.85 ml/mmHg/m2 (1) < 0.9 ml/mmHg/m2 (1) < 1.27 ml/mmHg/m2 (1)
mPAP	(23, 25, 31)	3	
Stroke volume index	(8, 23)	2	
PVR/SVR	(27, 29)	2	
Systemic venous SO2	(3)	1	
Ventricular-vascular coupling	(25)	1	
Imaging variables			
ECHO			
*TAPSE	(6, 10, 14, 32, 33)	>3	<12 mm (1) < 14 mm (1) Z-score <-4.3 (1)S
*RVFAC	(33-35)	>3	<16% (1) <25% (1) <32% (1)
*RV-Longitudinal strain	(35-37)	3	-15% (s)/-17 (fw) (1) -19% (fw) (1) -14% (g) (1)
*LV-EI	(10, 33, 37)	3	Systolic > 1,89

			Diastolic >1,55
RV/LV dimension ratio	(6, 32, 38)	2	> 1 (1)
RA-area/volume	(10, 33)	2	
Pericardial effusion	(6, 32)	1	
RVEDD	(10, 33)	2	
RV ejection time	(32)	1	
LV diameter	(10, 32)	2	
RA-deformation	(39, 40)	1	
SV/ESV (3D)	(41)	1	
PAAT	(10)	1	
PV Diameter	(10)	1	
TV Diameter	(10)	1	
TV Velocity	(10)	1	
MV E- and A- wave velocity	(10)	1	
MRI			
*RVEF	(7, 42, 43)	3	<44% (1)
*RVmassi	(7, 42, 43)	3	>80 g/m2 (1)
RVEDVi	(7, 42)	2	
RVESVi	(7, 43)	2	
RVmass/LVmass ratio	(43)	1	
Mid-RV Diameter index	(7)	1	
LV-EI	(43)	1	
LVSVi	(7)	1	
RV Global functioning index	(42)	1	
Mechanical Dyssynchrony	(44)	1	
PWV	(45)	1	
CT-scan			
Fractal branching	(46)	1	
Serum biomarkers			
*NT-pro-BNP (BNP)	(6, 14, 18, 26, 31, 47-49)	>3	NTpBNP>1200 ng/L (1) NTpBNP > 300 ng/L (2) BNP < 300 ng/L (1)
Uric Acid	(18, 21, 47, 50)	2	
IL-6	(47, 51)	2	
Norepinephrine	(6, 49)	1	
SAA-4	(52)	1	
Apolipoproteine-A1	(52)	1	
ST2	(53)	1	
Insulin-like GF	(54)	1	
GDF15	(55)	1	
MPV	(17)	1	
Genetics	(56-58)	3	
"Risk scores"			
"Haarman" (number of low risk variables)	(59)	1	< 10-13 low risk variables
"Lammers" (echo: TAPSE, RA area, LVEI)	(10)	1	RA z-score >2.9 and LVEId >1,5 and TAPSE z-score < -2,85
"Reveal 2.0 +ST2"	(53)	1	
Qian	(26)	1	W z-score <-2 and WHO-FC III-IV and RV-enlargement

Overview of parameters reported to be associated with outcomes in pediatric pulmonary arterial hypertension with the appropriate references, the number of unique cohorts in which the association has been established, and the published cut off values that identify patients at high risk adverse outcomes including the number of studies that have established the cut off values (in brackets). Overview may not be fully exhaustive due to lack of a systematic review. *Prognostic parameters that were selected based on this table for the risk stratification tool.

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