2022 ESC/ERS 3-Strata Risk Score Calculator

The 2022 ESC/ERS Treatment Guidelines¹ recommend using 3-strata risk stratification at baseline. A clinician's best judgment should be used when scoring the multi-parameter variables. *At least 3 variables are required to generate a risk score*.

Variables (Est. 1 Yr Mortality)	Low Risk (<5%)	Intermediate Risk (5%–20%)	High Risk (>20%)	Score		
Point Value Per Variable	1	2	3			
Signs of right HF	Absent	Absent	Present			
Progression of symptoms	No	Slow	Rapid			
Syncope	No	Occasional syncope	Repeated syncope			
WHO-FC	Ι, ΙΙ	Ш	IV			
6MWD	>440 m	165–440 m	<165 m			
СРЕТ	Peak VO ₂ >15 mL/min/kg (>65% pred.) VE/VCO ₂ slope <36	Peak VO ₂ 11–15 mL/min/kg (35–65% pred.) VE/VCO ₂ slope 36–44	Peak VO ₂ <11 mL/min/kg (<35% pred.) VE/VCO ₂ slope >44			
Biomarkers: BNP or NT-proBNP	BNP <50 ng/L NT-proBNP <300 ng/L	BNP 50–800 ng/L NT-proBNP 300–1100 ng/L	BNP >800 ng/L NT-proBNP >1100 ng/L			
Echocardiography	RA area <18 cm² TAPSE/sPAP >0.32 mm/mmHg No pericardial effusion	RA area 18–26 cm² TAPSE/sPAP >0.32 mm/mmHg Minimal pericardial effusion	RA area >26 cm² TAPSE/sPAP >0.32 mm/mmHg Moderate—large pericardial effusion			
cMRI°	RVEF >54% SVI >40 mL/m ² RVESVI <42 mL/m ²	RVEF 37–54% SVI 26–40 mL/m ² RVESVI 42–54 mL/m ²	RVEF <37% SVI <26 mL/m ² RVESVI >54 mL/m ²			
Haemodynamics	RAP <8 mm Hg CI ≥2.5 L/min/m² SVI >38 mL/m² SvO ₂ >65%	RAP 8–14 mm Hg CI 2–2.4 L/min/m ² SVI >31–38 mL/m ² SvO ₂ 60%–65%	RAP >14 mm Hg CI <2 L/min/m ² SVI <31 mL/m ² SvO ₂ <60%			
Total Risk Score	Divide the sum of all variable scores by the number of variables entered and round to the nearest decimal					

	Low Risk	Intermediate Risk	High Risk	
Risk Score	1 to <1.5	1.5 to <2.5	2.5 to 3	

This PAH risk calculation tool does not replace professional or medical advice. HCPs must exercise their own independent clinical judgment when using the tool.

The parameters and thresholds in the above table are taken from the 2022 ESC/ERS Guidelines for PAH.¹ The scoring methodology (1=low risk, 2=intermediate risk, 3=high risk) was adapted from the Swedish PAH Registry.².³

References: 1. Humbert M, et al. *Eur Heart J.* 2022;43(38):3618-3731. **2.** Kylhammar D, et al. *Eur Heart J.* 2018;39(47):4175-4181. **3.** Hoeper MM, et al. *Eur Respir J.* 2017;50(2). pii:1700740



COMPERA 2.0: 4-Strata Risk Assessment

The 2022 ESC/ERS Treatment Guidelines recommend using 4-strata risk stratification in follow-up visits every 3–6 months. All 3 values are required to calculate score. 1 year transplant-free mortality risk is based on average points assigned.

Variables (1 Yr Survival Rates)	Low Risk (98.5%)	Intermediate — Low Risk (97.2%)	Intermediate — High Risk (91.3%)	High Risk (78%)	Score	
Point Value Per Variable	1	2	3	4		
WHO FC	1, 11	_	III	IV		
6MWD	>440 m	320–440 m	165–319 m	<165 m		
BNP	<50 ng/L	50–199 ng/L	200–800 ng/L	>800 ng/L		
NT-proBNP	<300 ng/L	300–649 ng/L	650–1100 ng/L	>1100 ng/L		
Total Risk Score	Divide the sum of all variable scores by 3 and round to the nearest integer					
	Low Risk	Intermediate–Low Risk	Intermediate-High Risk	High Risk		
Risk Score	1 to <1.5	1.5 to <2.5	2.5 to <3.5	3.5 to 4		

This PAH risk calculation tool does not replace professional or medical advice. HCPs must exercise their own independent clinical judgment when using the tool.



REVEAL 2.0 Risk Calculator

Directions: Select all variables that apply. **A minimum of 7 variables** are required to generate a score. Calculation accuracy increases with more selections.

Step 1 Select at least 7	variables.						Score
WHO Group 1			Other	CTD-PAH	Heritable	PoPH	
Subgroup			0	1	2	3	
Demographics -			No		Yes		
Male age > 60 years			0		2		
eGFR < 60 mL/min/1.73m ²			No	Yes			
or renal insufficiency			0	1			
NYHA/WHO		I	II	III	IV		
Functional Class		-1	0	1	2		
Secretalis DD (man Ha)			SBP ≥110	SBP <110			
Systolic BP (mm Hg)			0	1			
List Date (DDM)			HR ≤96	HR >96			
Heart Rate (BPM)			0	1			
All-Cause			No	Yes			
Hospitalizations ≤ 6 mo			0	1			
	≥440	320 to 440	<320 to 165	<165			
6-Minute Walk Test (m)	–2	-1	0	1			
	50		50 to <200	200 to <800	≥800		
BNP (pg/mL)	–2		0	1	2		
OR	<300		300 to <1100		≥1100		
NT-proBNP (pg/mL)	–2		0		2		
Pericardial Effusion			No	Yes			
on Echocardiogram			0	1			
			No	Yes			
% Predicted DLCO ≤ 40			0	1			
mRAP > 20 mm Hg			No	Yes			
Within 1 Year			0	1			
PVR < 5 Wood units on		Yes	No				
right heart catheterization		-1	0				
				Step 2 St	um of above in	nin. 7 variables)	
				31GP 2	uni oi above (ii	iii. / variables/	
							+6
					Step 3	Risk score	

	Low Risk	Intermediate Risk	High Risk	
Risk Score	0–6	7–8	≥9	

This PAH risk calculation tool does not replace professional or medical advice. HCPs must exercise their own independent clinical judgment when using the tool.

APAH=associated pulmonary arterial hypertension; BNP=B-type natriuretic peptide; BPM=beats per minute; CTD=connective tissue disease; DLCO=diffusing capacity of the lung for carbon monoxide; eGFR=estimated glomerular filtration rate; FPAH=familial pulmonary atrial hypertension; HR=heart rate; mRAP=mean right arterial pressure; NT-proBNP=N-terminal pro-B-type natriuretic peptide; NYHA=New York Heart Association; PAH=pulmonary arterial hypertension; PoPH=portopulmonary hypertension; PVR=pulmonary vascular resistance; REVEAL=Registry to EValuate Early And Long-term pulmonary arterial hypertension disease management; SBP=systolic blood pressure; WHO=World Health Organization

Reference: Benza RL, et al. Chest. 2019;156(2):323-337.

REVEAL Lite 2 Risk Calculator

Directions: Select all variables that apply. **A minimum of 3 variables** are required to generate a score, where at least 2 are from most predictive values denoted **.

Step 1 Select at least 2 of the most predictive variables						Score
	<50		50 to <200	200 to <800	≥800	
BNP (pg/mL)**	-2		0	1	2	
OR	<300		300 to <1100		≥1100	
NT-proBNP (pg/mL)**	-2		0		2	
	≥440	320 to 440	<320 to 165	<165		
6-Minute Walk Test (m)**	-2	-1	0	1		
NYHA/WHO			II	III	IV	
Functional Class**		-1	0	1	2	
Step 2 Select additiona	l variables.		SBP ≥110	SBP<110		
Systolic BP (mm Hg)			0	1		
			HR ≤96	HR >96		
Heart Rate (BPM)			0	1		
eGFR < 60 mL/min/1.73m ²			No	Yes		
or renal insufficiency			0	1		
				Step 3 Su	um of above (min. 3 variables)	
						+6
					Step 4 Risk score	

	Low Risk	Intermediate Risk	High Risk	
Risk Score	≤5	6–7	≥8	

This PAH risk calculation tool does not replace professional or medical advice. HCPs must exercise their own independent clinical judgment when using the tool.

BNP=B-type natriuretic peptide; BPM=beats per minute; eGFR=estimated glomerular filtration rate;HR=heart rate;NT-proBNP=N-terminal pro-B-type natriuretic peptide; NYHA=New York Heart Association; PAH=pulmonary arterial hypertension; REVEAL=Registry to EValuate Early And Long-term pulmonary arterial hypertension disease management; SBP=systolic blood pressure; WHO=World Health Organization



PAH Initiative
Sponsored by United Therapeutics
Committed to Improving the Lives of Patients