

# 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 Subgroup	Other	CTD-PAH	Heritable	PoPH	
	0	1	2	3	
Demographics - Male age > 60 years	No		Yes		
	0		2		
eGFR < 60 mL/min/1.73m <sup>2</sup> or renal insufficiency	No	Yes			
	0	1			
NYHA/WHO Functional Class	I	II	III	IV	
	-1	0	1	2	
Systolic BP (mm Hg)		SBP ≥110	SBP <110		
		0	1		
Heart Rate (BPM)		HR ≤96	HR >96		
		0	1		
All-Cause Hospitalizations ≤ 6 mo		No	Yes		
		0	1		
6-Minute Walk Test (m)	≥440	320 to 440	<320 to 165	<165	
	-2	-1	0	1	
BNP (pg/mL)	50	50 to <200	200 to <800	≥800	
	-2	0	1	2	
OR					
NT-proBNP (pg/mL)	<300	300 to <1100		≥1100	
	-2	0		2	
Pericardial Effusion on Echocardiogram		No	Yes		
		0	1		
% Predicted DLCO ≤ 40		No	Yes		
		0	1		
mRAP > 20 mm Hg Within 1 Year		No	Yes		
		0	1		
PVR < 5 Wood units on right heart catheterization		Yes	No		
		-1	0		
<b>Step 2</b> Sum of above (min. 7 variables)					
					+6
<b>Step 3</b> Risk score					



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 arterial 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

	<b>Score</b>			
BNP (pg/mL)**	<50 -2	50 to <200 0	200 to <800 1	≥800 2
OR				
NT-proBNP (pg/mL)**	<300 -2	300 to <1100 0		≥1100 2
6-Minute Walk Test (m)**	≥440 -2	320 to 440 -1	<320 to 165 0	<165 1
NYHA/WHO Functional Class**		I -1	II 0	III 1 IV 2

**Step 2** Select additional variables.

Systolic BP (mm Hg)	SBP ≥110 0	SBP <110 1	
Heart Rate (BPM)	HR ≤96 0	HR >96 1	
eGFR < 60 mL/min/1.73m <sup>2</sup> or renal insufficiency	No 0	Yes 1	
<b>Step 3</b> Sum of above (min. 3 variables)			
			<b>+6</b>
<b>Step 4</b> Risk score			

	<b>Low Risk</b>	<b>Intermediate Risk</b>	<b>High Risk</b>
<b>Risk Score</b>	<b>≤5</b>	<b>6-7</b>	<b>≥8</b>

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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

Reference: Benza RL, et al. *Chest*. 2021;159(1):337-346.

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# 2022 ESC/ERS 3-Strata Risk Score Calculator

The 2022 ESC/ERS Treatment Guidelines<sup>1</sup> 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	I, II	III	IV	
6MWD	>440 m	165–440 m	<165 m	
CPET	Peak VO <sub>2</sub> >15 mL/min/kg (>65% pred.) VE/VCO <sub>2</sub> slope <36	Peak VO <sub>2</sub> 11–15 mL/min/kg (35–65% pred.) VE/VCO <sub>2</sub> slope 36–44	Peak VO <sub>2</sub> <11 mL/min/kg (<35% pred.) VE/VCO <sub>2</sub> 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 cm <sup>2</sup> TAPSE/sPAP >0.32 mm/mmHg No pericardial effusion	RA area 18–26 cm <sup>2</sup> TAPSE/sPAP >0.32 mm/mmHg Minimal pericardial effusion	RA area >26 cm <sup>2</sup> TAPSE/sPAP >0.32 mm/mmHg Moderate–large pericardial effusion	
cMRI <sup>e</sup>	RVEF >54% SVI >40 mL/m <sup>2</sup> RVESVI <42 mL/m <sup>2</sup>	RVEF 37–54% SVI 26–40 mL/m <sup>2</sup> RVESVI 42–54 mL/m <sup>2</sup>	RVEF <37% SVI <26 mL/m <sup>2</sup> RVESVI >54 mL/m <sup>2</sup>	
Haemodynamics	RAP <8 mm Hg CI ≥2.5 L/min/m <sup>2</sup> SVI >38 mL/m <sup>2</sup> SvO <sub>2</sub> >65%	RAP 8–14 mm Hg CI 2–2.4 L/min/m <sup>2</sup> SVI >31–38 mL/m <sup>2</sup> SvO <sub>2</sub> 60%–65%	RAP >14 mm Hg CI <2 L/min/m <sup>2</sup> SVI <31 mL/m <sup>2</sup> SvO <sub>2</sub> <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	

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

**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. Hoepfer 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	I, II	–	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	
OR					
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	

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**References:** 1. Boucly A, Weatherald J, Salvale L, et al. External validation of a refined four-stratum risk assessment score from the French pulmonary hypertension registry. *Eur Respir J.* 2022; 59:2102419 [DOI: 10.1183/13993003.02419-2021].