

Online Supplemental Material

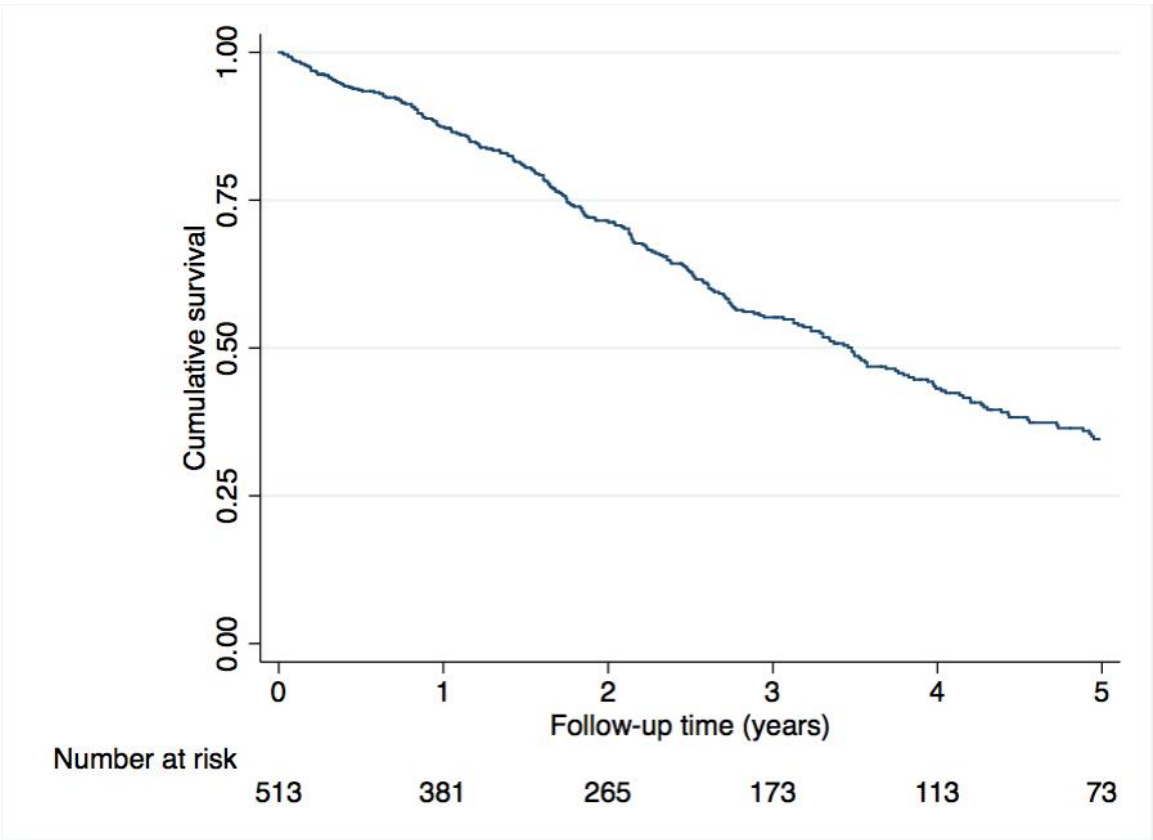
Haemodynamics and risk assessment in systemic-sclerosis associated pulmonary arterial hypertension

Jason Weatherald, Athénaïs Boucly, David Launay, Vincent Cottin, Grégoire Prévôt,
Delphine Bourlier, Claire Dauphin, Ari Chaouat, Laurent Savale, Xavier Jaïs, Mitja
Jevnikar, Julie Traclet, Pascal De Groote, Gérald Simonneau, Eric Hachulla, Luc
Mouthon, David Montani, Marc Humbert, Olivier Sitbon

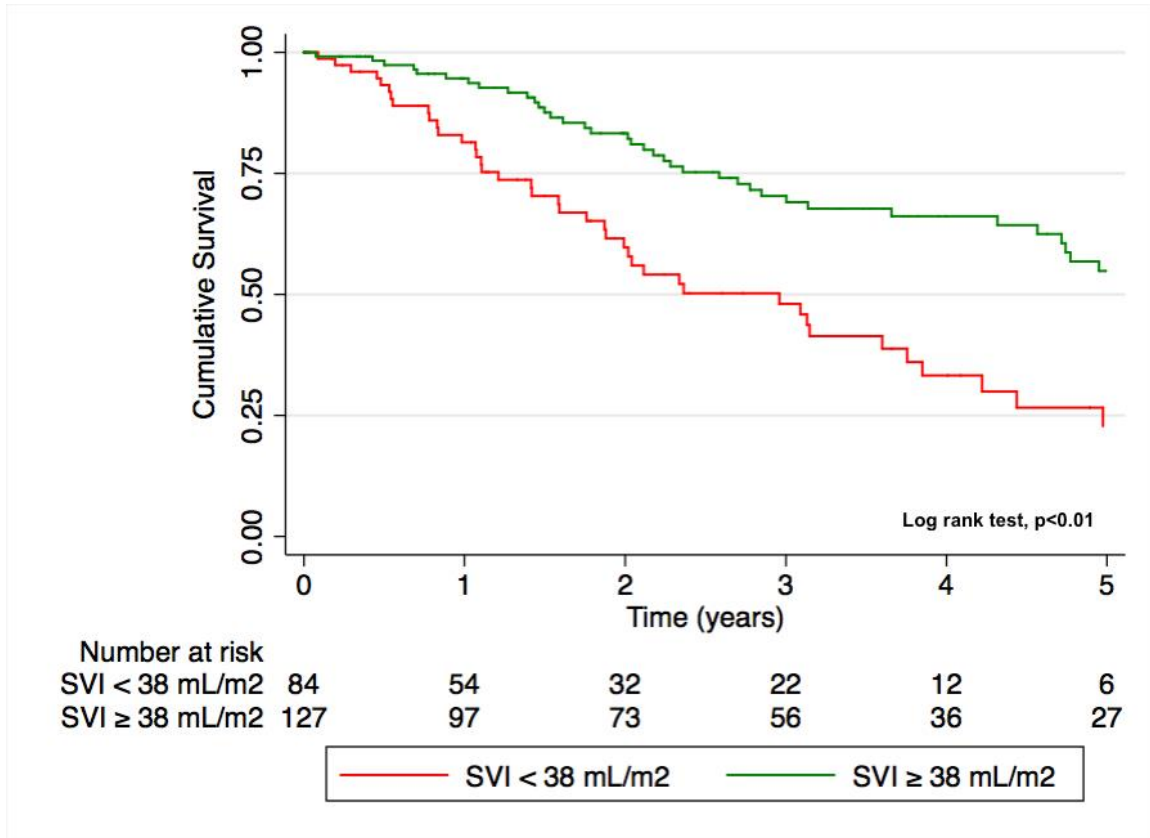
Table of Contents

Supplemental Figure S1	p.2
Supplemental Figure S2	p.3
Supplemental Table S1	p.4
Supplemental Table S2	p.5
Supplemental Table S3	p.6
Supplemental Table S4	p.7
Supplemental Table S5	p.8
Supplemental Table S6	p.9

Supplemental Figure S1 - Transplant-free survival



Supplemental Figure S2 - Transplant-free survival for patients with cardiac index ≥ 2.5 L/min/m² at first follow-up right heart catheterization according to stroke volume index (SVI).



Supplemental Table S1 - Cox proportional-hazards regression for the association
between number of low-risk criteria present at baseline and transplant-free survival

Number of Low-Risk Criteria	Hazard Ratio	(95% CI)	p
0	Reference		
1	0.81	(0.59-1.13)	0.22
2	0.69	(0.50-0.96)	0.028
3	0.51	(0.32-0.82)	0.006
4	0.21	(0.09-0.53)	0.001
Log-likelihood	-1403.1419		
p	0.0002		

Supplemental Table S2 - Discrimination of 1-year transplant-free survival by number of low-risk criteria present at baseline.

Low-risk criteria Cutpoint	Sensitivity	Specificity	+ Likelihood ratio	- Likelihood ratio
>= 0	1.0	0	1	
>= 1	0.81	0.32	1.19	0.60
>= 2	0.47	0.73	1.78	0.72
>= 3	0.16	0.95	3.18	0.89
>= 4	0	1.0		0.96

Area under the curve 0.63 (95%CI 0.56-0.69)

Supplemental Table S3 - Cox proportional-hazards regression for the association between number of low-risk criteria present at follow-up and transplant-free survival

Number of Low-risk			
Criteria	Hazard Ratio	(95% CI)	p
0	Reference		
1	0.57	(0.38-0.87)	0.009
2	0.32	(0.21-0.50)	<0.001
3	0.26	(0.16-0.43)	<0.001
4	0.08	(0.03-0.21)	<0.001
Log-likelihood	-865.678		
p	<0.001		

Supplemental Table S4 - Univariable and multivariable Cox proportional-hazards regression for the association between low-risk criteria at follow-up and transplant-free survival

Low-Risk Criteria	Univariable			Multivariable		
	Hazard Ratio	(95% CI)	p	Hazard Ratio	(95% CI)	p
NYHA I or II	0.42	(0.29-0.59)	<0.001	0.65	(0.44-0.99)	0.042
6MWD > 440 m	0.38	(0.21-0.68)	0.001	0.53	(0.28-0.98)	0.043
RAP < 8 mmHg	0.51	(0.37-0.70)	<0.001	0.66	(0.46-0.97)	0.03
CI \geq 2.5 L/m/m ²	0.46	(0.33-0.63)	<0.001	0.51	(0.35-0.75)	0.001
Log Likelihood	-560.369					
p	<0.001					

Abbreviations: NYHA - New York Heart Association; 6MWD - 6-minute walking distance; RAP - right atrial pressure; CI - cardiac index

Supplemental Table S5 - Discrimination of subsequent 1-year transplant-free survival by number of low-risk criteria present at first follow-up right heart catheterization.

Low-risk criteria Cutpoint	Sensitivity	Specificity	+ Likelihood ratio	- Likelihood ratio
>= 0	1.0	0	1	
>= 1	0.87	0.29	1.23	0.45
>= 2	0.63	0.76	2.58	0.49
>= 3	0.31	0.93	4.29	0.74
>= 4	0.07	0.97	2.76	0.96

Area under the curve 0.71 (95%CI 0.64-0.79)

Supplemental Table S6 - Multivariate analysis of low-risk criteria including NT-proBNP and BNP (n=160).

Low-Risk Criteria	Hazard Ratio	(95% CI)	p
NYHA I or II	0.31	(0.11-0.83)	0.02
6MWD > 440 m	0.71	(0.19-2.67)	0.617
RAP < 8 mmHg	0.63	(0.30-1.32)	0.224
CI \geq 2.5 L/m/m ²	0.62	(0.31-1.24)	0.176
NT-proBNP < 300 ng/L OR BNP < 50 ng/L	1.13	(0.37-3.42)	0.828