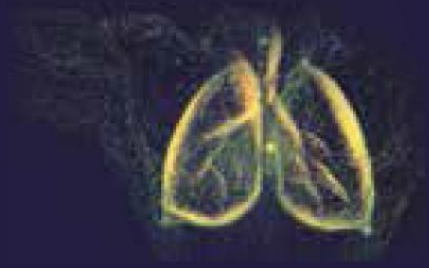
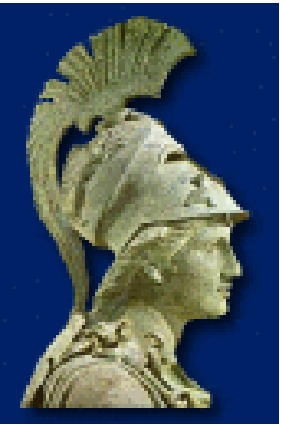


Θεραπεία Πνευμονικής Υπέρτασης - φαρμακευτική και μη



Ηρακλής Τσαγκάρης

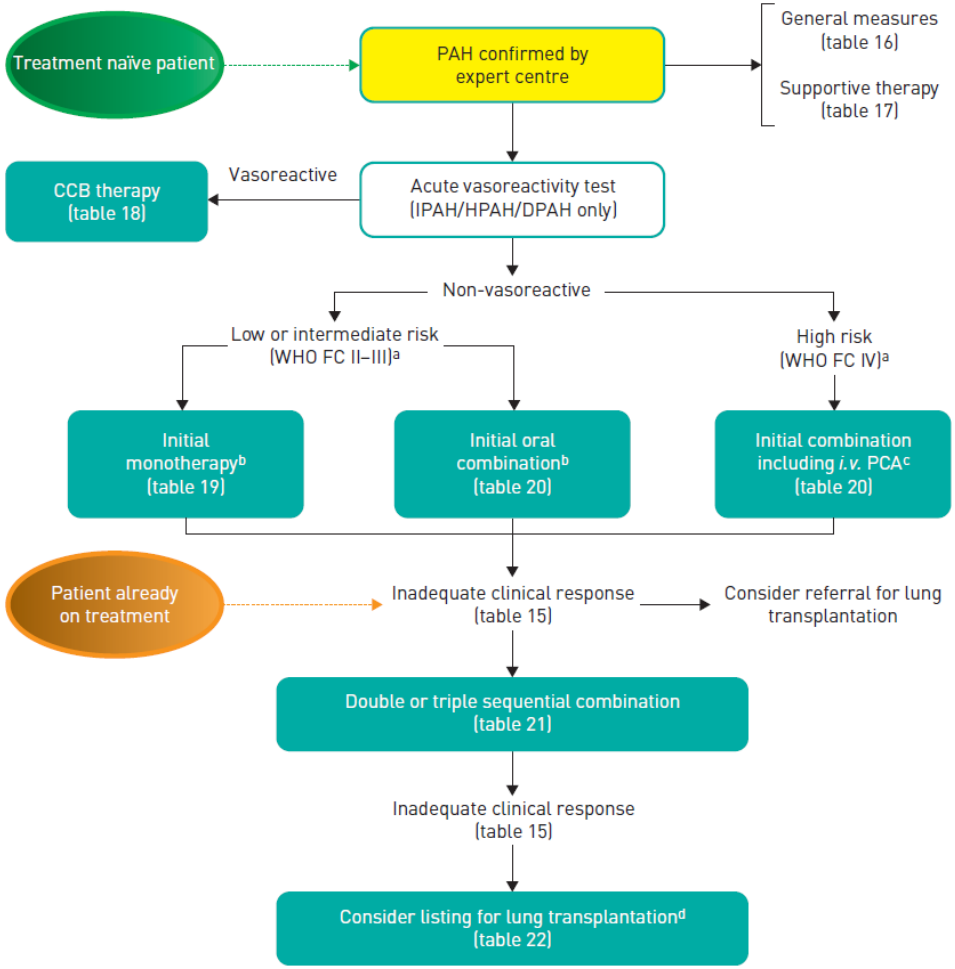
Αναπληρωτής Καθηγητής ΕΚΠΑ

Νοσοκομείο ΑΤΤΙΚΟΝ

Δήλωση συμφερόντων

- Επιδοτούμενη συμμετοχή σε συνέδρια, κλινικές μελέτες ή συμβουλευτικά των εταιρειών Actelion, Bayer, ELPEN, Galenica, Glaxo GSK, Lilly, MSD, Pfizer
- Υπότροφος Ιδρύματος Ωνάση

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension



The goal of treatment is to achieve a low-risk status and to perform regular follow-ups to maintain the goal

Recommendations for evaluation of PAH severity and response to therapy	Class	Level
It is recommended to evaluate the severity of PAH patients with a panel of data derived from clinical assessment, exercise tests, biochemical markers and echocardiographic and haemodynamic evaluations	I	C
It is recommended to perform regular follow-up assessments every 3–6 months in stable patients	I	C
Achievement/maintenance of a low-risk profile is recommended as an adequate treatment response for patients with PAH	I	C
Achievement/maintenance of an intermediate-risk profile should be considered an inadequate treatment response for most patients with PAH	IIa	C

Treatment goal

Galiè N, *et al. Eur Respir J* 2015; 46:903-75;
Galiè N, *et al. Eur Heart J* 2016; 37:67-119

Why should we assess risk of disease progression?

- Stable clinical parameters can be present even in pts with deteriorating RV function
- With timely therapeutic intervention the progression of PAH can be altered
- Without risk assessment appropriate treatment decisions can be easily missed

Why is a multiparameter approach needed?

- No single variable (NYHA, 6MWT) provides sufficient diagnostic/prognostic information
- PAH is a multifaceted disease
- Risk assessment with several parameters provides a comprehensive view of the patient and is fundamental for the determination of optimal treatment strategy

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	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 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/ml	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
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Risk Assessment in PAH

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[b]Occasional syncope during brisk or heavy exercise, or occasional orthostatic syncope in an otherwise stable patient.
[c]Repeated episodes of syncope, even with little or regular physical activity.

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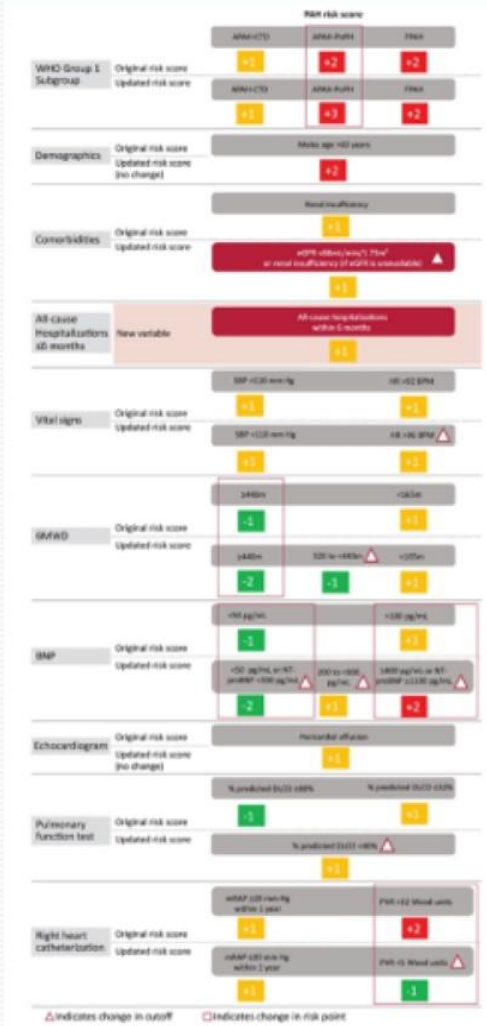
REVEAL Registry
Updated PAH Risk Score

- New variables added (hospitalization in past year, eGFR)
- New cut-off points for BNP, APAH-PoPH, HR, DLCO, NYHA FC, and PVR
- Change in score from 12 to 24 months adds predictive ability
- A consistently high score over time has a worse prognosis than having a higher score at 24 months than at 12 months



Reprinted from *The Journal of Heart and Lung Transplantation*, 36, 4, Raymond L. Benza, et al, Updated Risk Score Calculator for Pulmonary Arterial Hypertension Patients, S19, Copyright 2017, with permission from Elsevier

Benza RL, et al. ATS 2017. Poster 5447.

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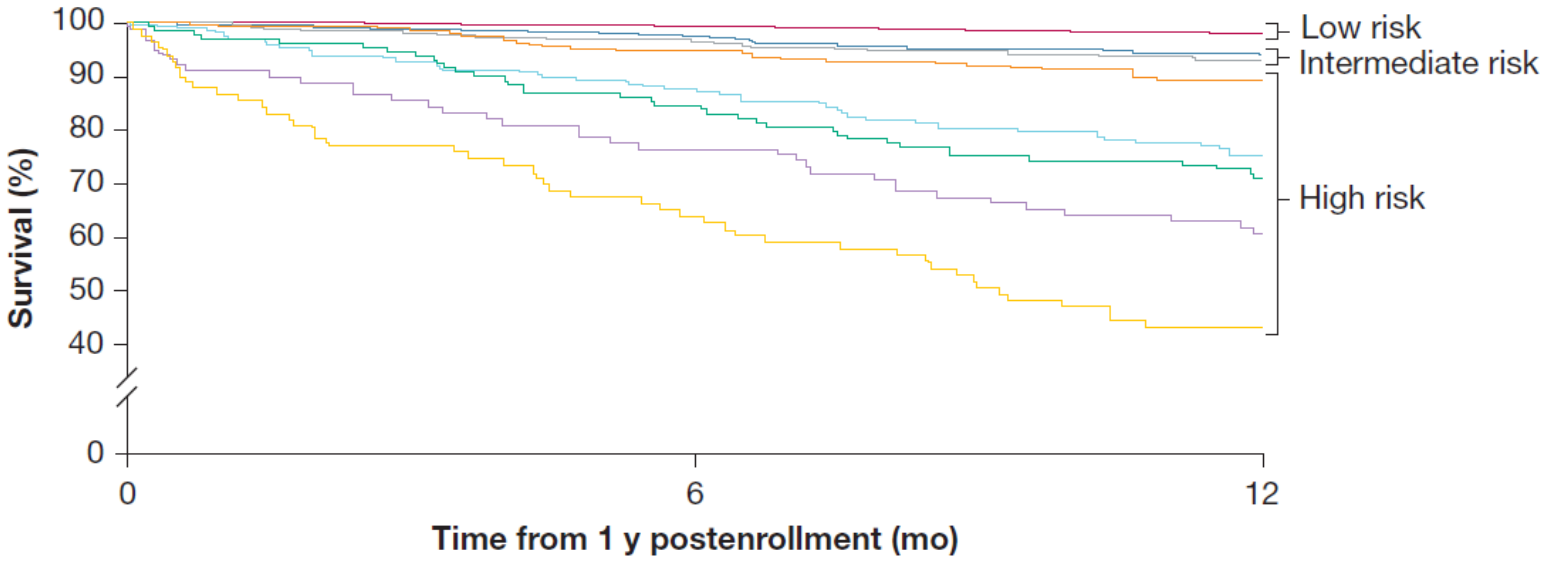
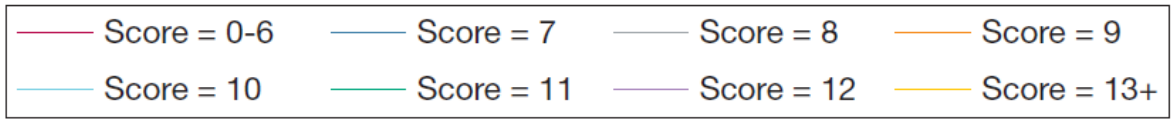


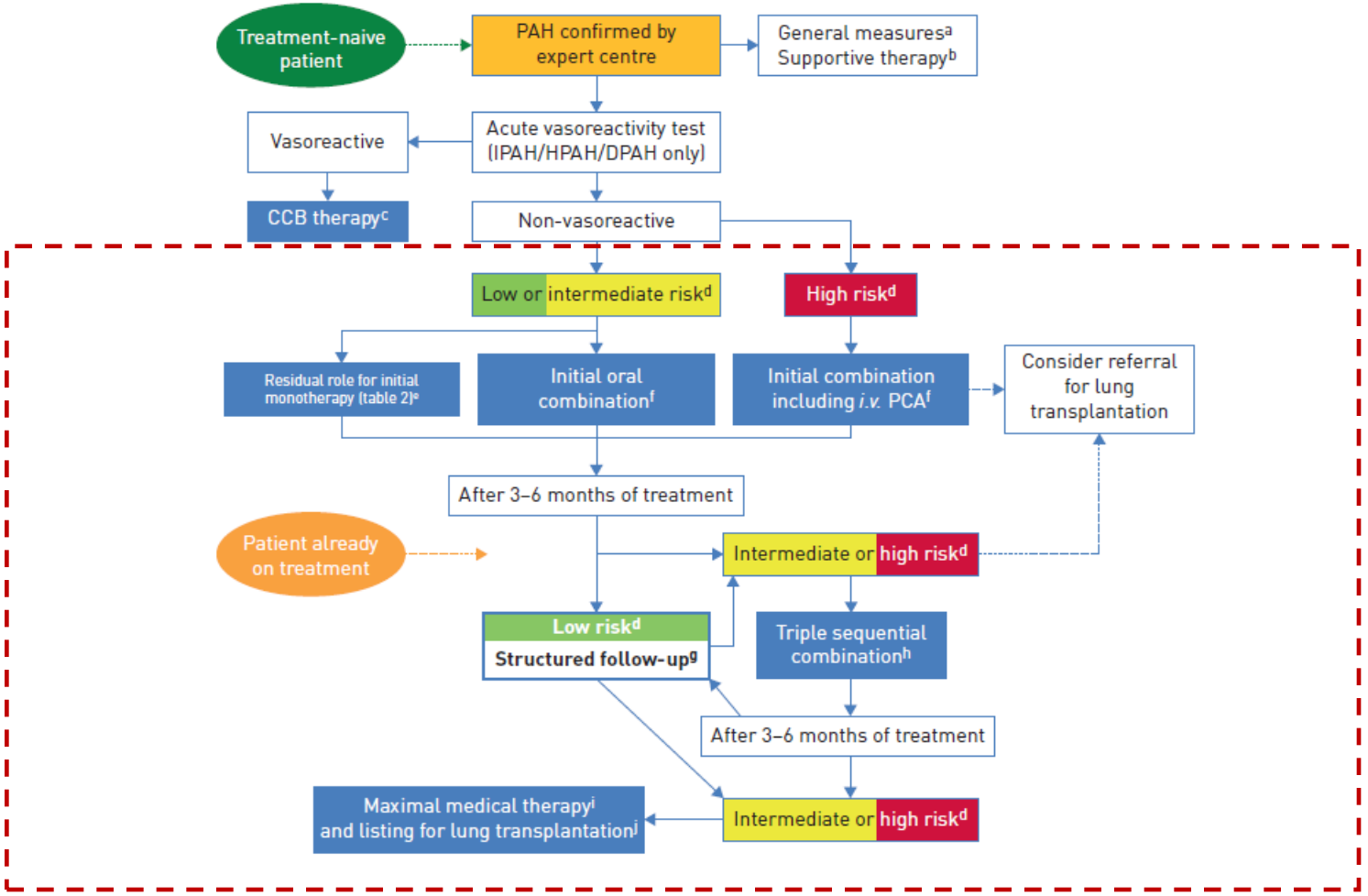
No. at risk			
Score = 0-6	1,073	1,056	1,032
Score = 7	386	368	344
Score = 8	306	292	275
Score = 9	266	248	230
Score = 10	195	166	142
Score = 11	130	109	87
Score = 12	90	68	54
Score = 13+	83	53	34



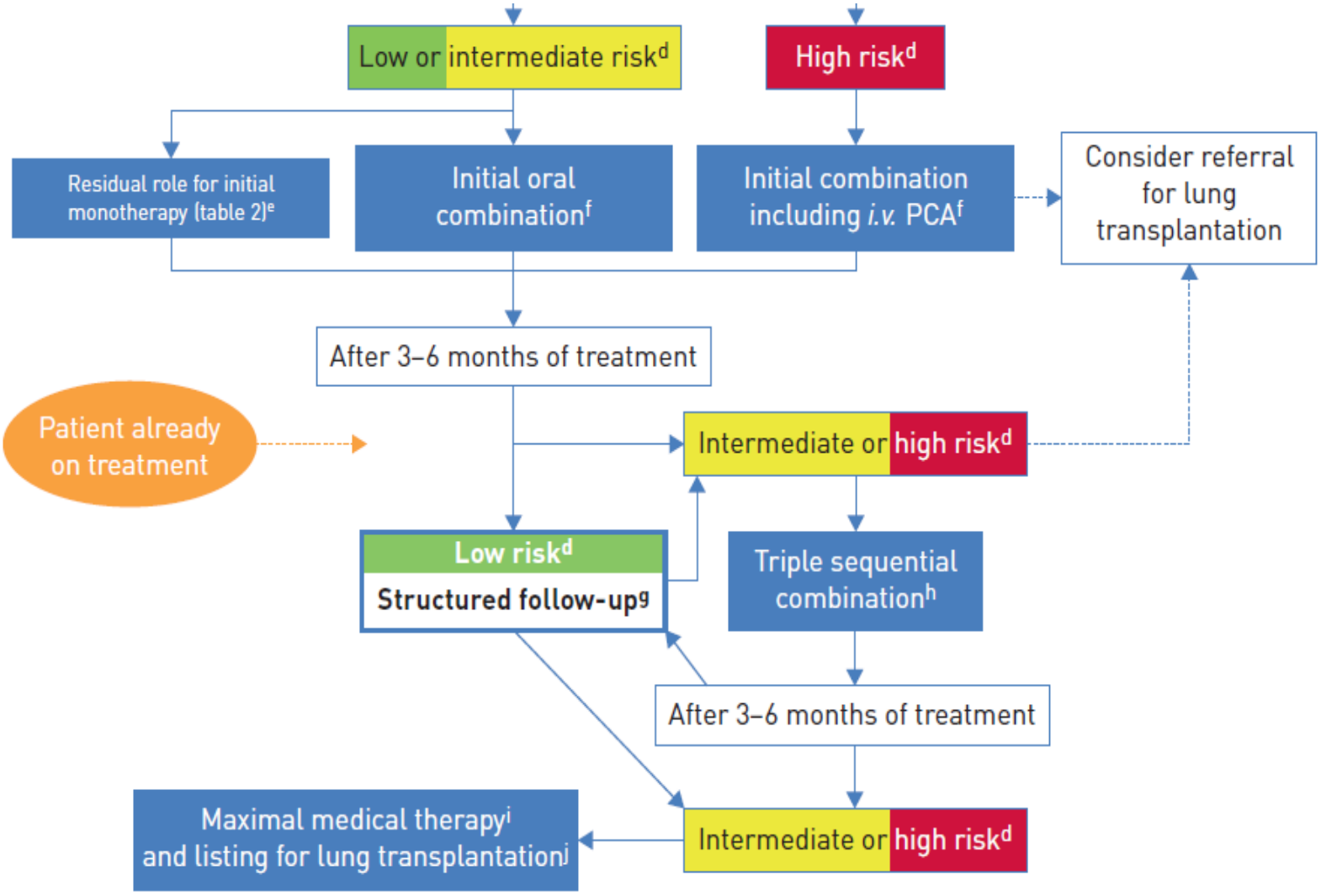
Treatment algorithm

Risk stratification and medical therapy of pulmonary arterial hypertension

Nazzareno Galiè[1], Richard N. Channick[2], Robert P. Frantz[3], Ekkehard Grünig[4], Zhi Cheng Jing[5], Olga Moiseeva[6], Ioana R. Preston[7], Tomas Pulido[8], Zeenat Safdar[9], Yuichi Tamura[10] and Vallerie V. McLaughlin[11]

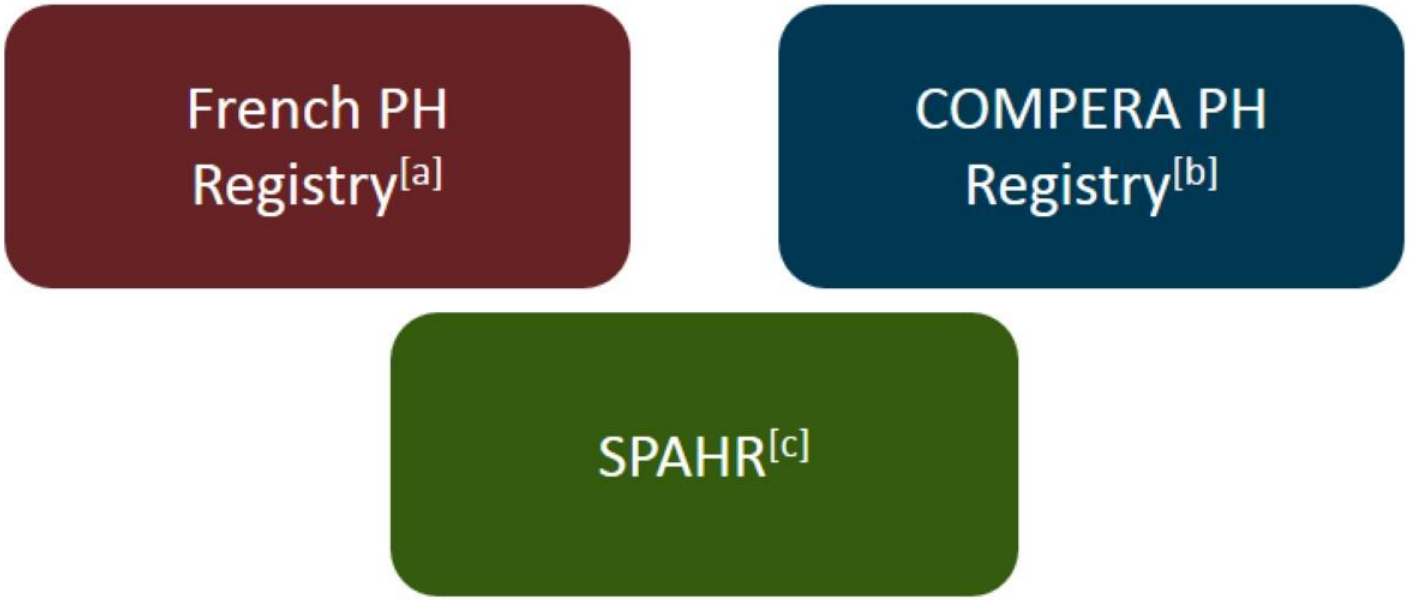


ERJ Express. Published on December 13, 2018 as doi: 10.1183/13993003.01889-2018


Low or intermediate risk[d]
High risk[d]
Residual role for initial monotherapy (table 2)[e]
Initial oral combination[f]
Initial combination including *i.v.* PCA[f]
Consider referral for lung transplantation
After 3–6 months of treatment
Patient already on treatment
Intermediate or high risk[d]
Low risk[d]
Structured follow-up[g]
Triple sequential combination[h]
After 3–6 months of treatment
Maximal medical therapy[i] and listing for lung transplantation[j]
Intermediate or high risk[d]

Registry Studies

- Evaluated prognostic impact of low-risk status in PAH



a. Boucly A, et al. *Eur Respir J.* 2017;50:1700889.
b. Hoeper MM, et al. *Eur Respir J.* 2017;50:1700740.
c. Kylhammar D, et al. *Eur Heart J.* 2017 [Epub ahead of print].

COMPERA Registry
Baseline Characteristics

Patients: 1588	Mean age: 64 years
Female: 64%	I/D/H-PAH: 67%

29% of patients had died within 5 years after PAH diagnosis

Hoeper MM, et al. *Eur Respir J*. 2017;50:1700740.

Mortality in pulmonary arterial hypertension: prediction by the 2015 European pulmonary hypertension guidelines risk stratification model

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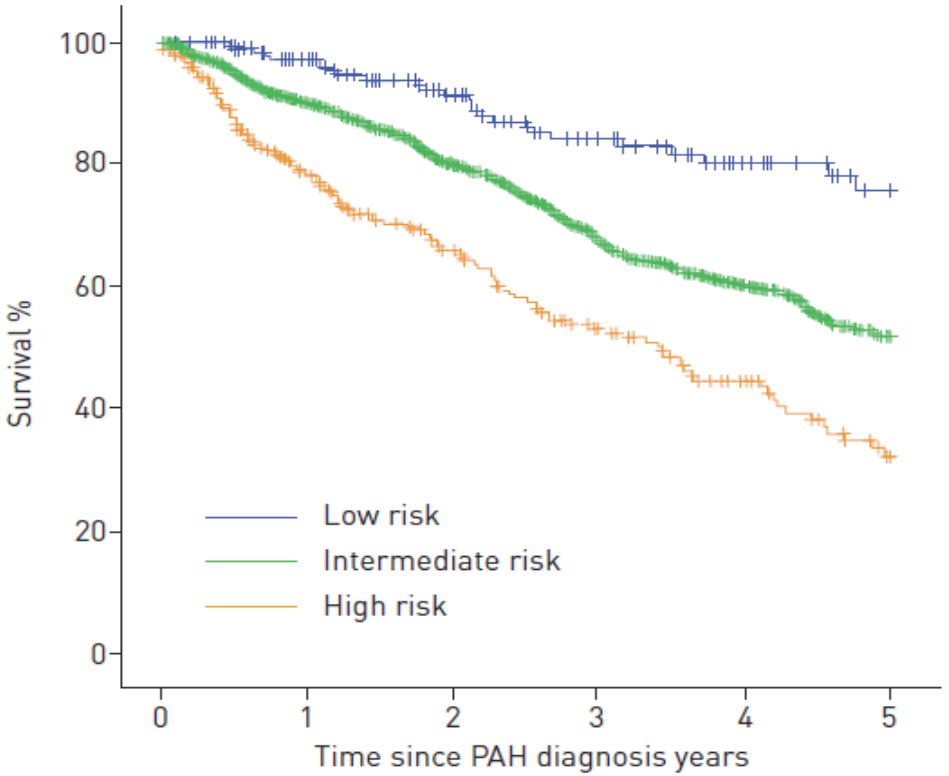

Years after enrolment	Survival %			Cases left n		
	Low risk	Intermediate risk	High risk	Low risk	Intermediate risk	High risk
0	100	100	100	196	1116	276
1	97.2	90.1	78.8	156	764	170
2	91.5	80.3	66.0	111	540	117
3	84.2	68.1	53.2	75	376	77
4	80.2	60.1	44.7	47	252	47
5	75.9	51.9	32.4	31	149	24

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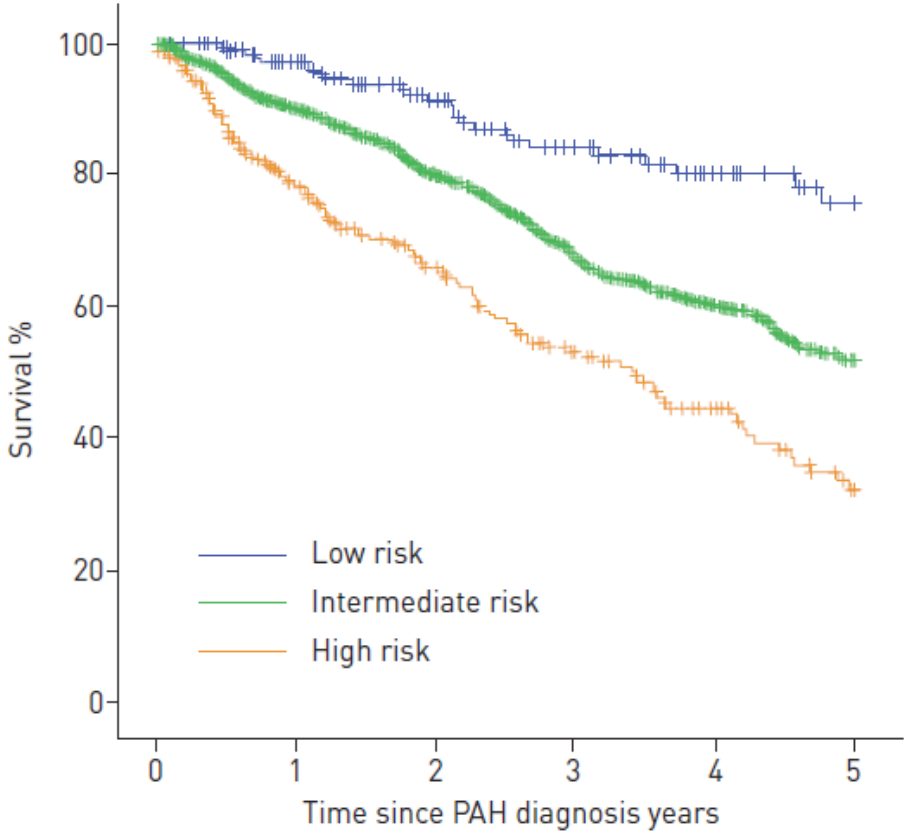

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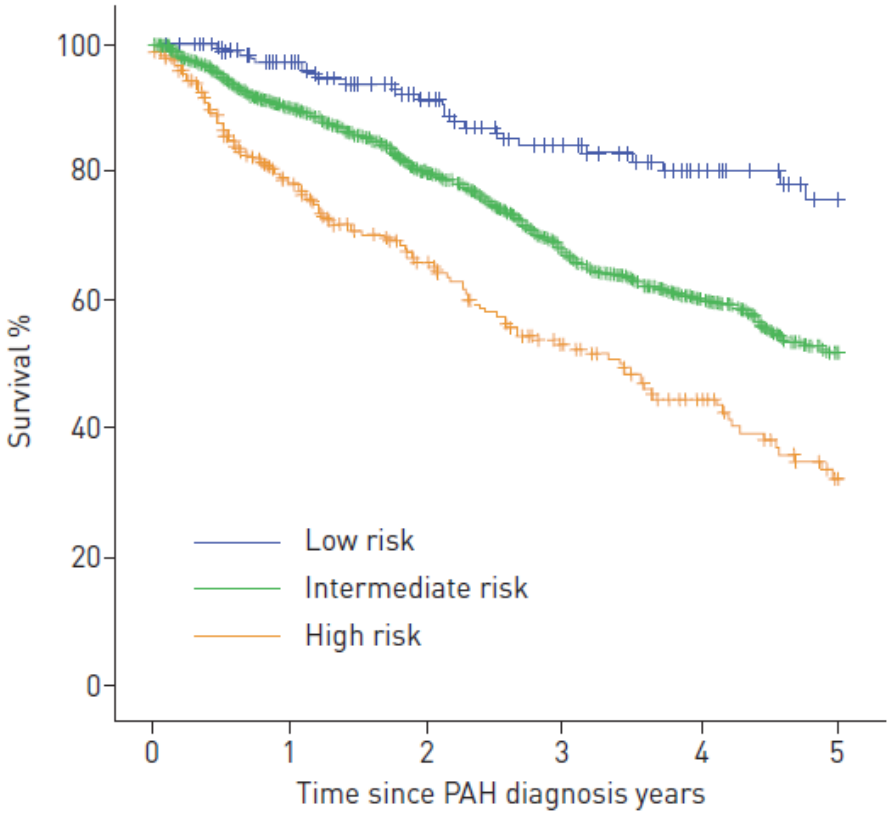

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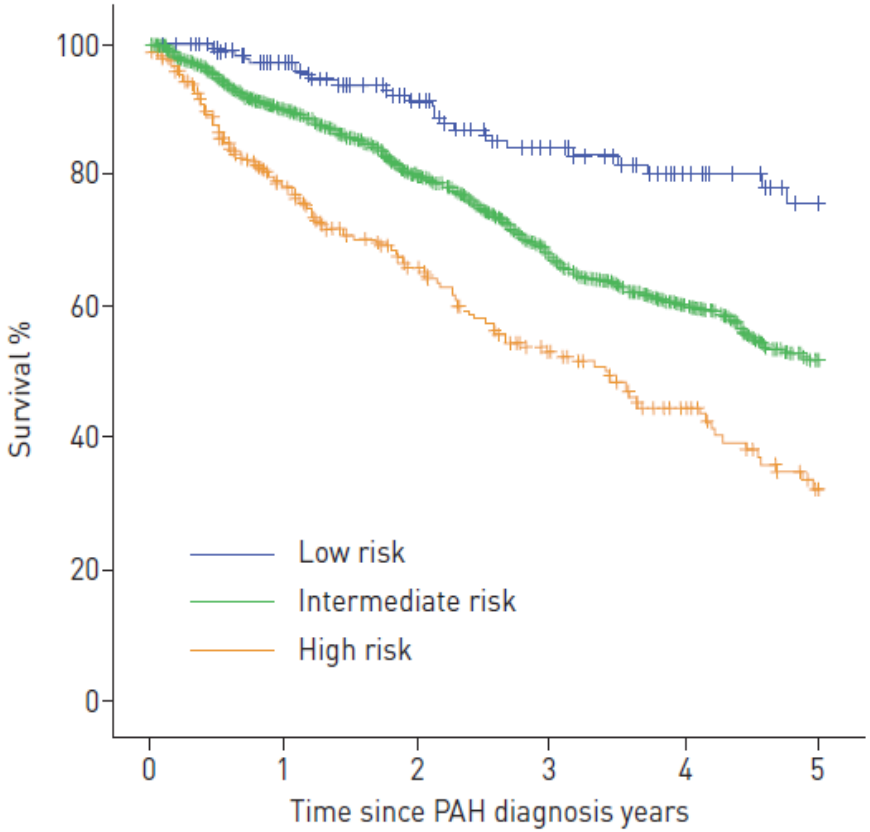

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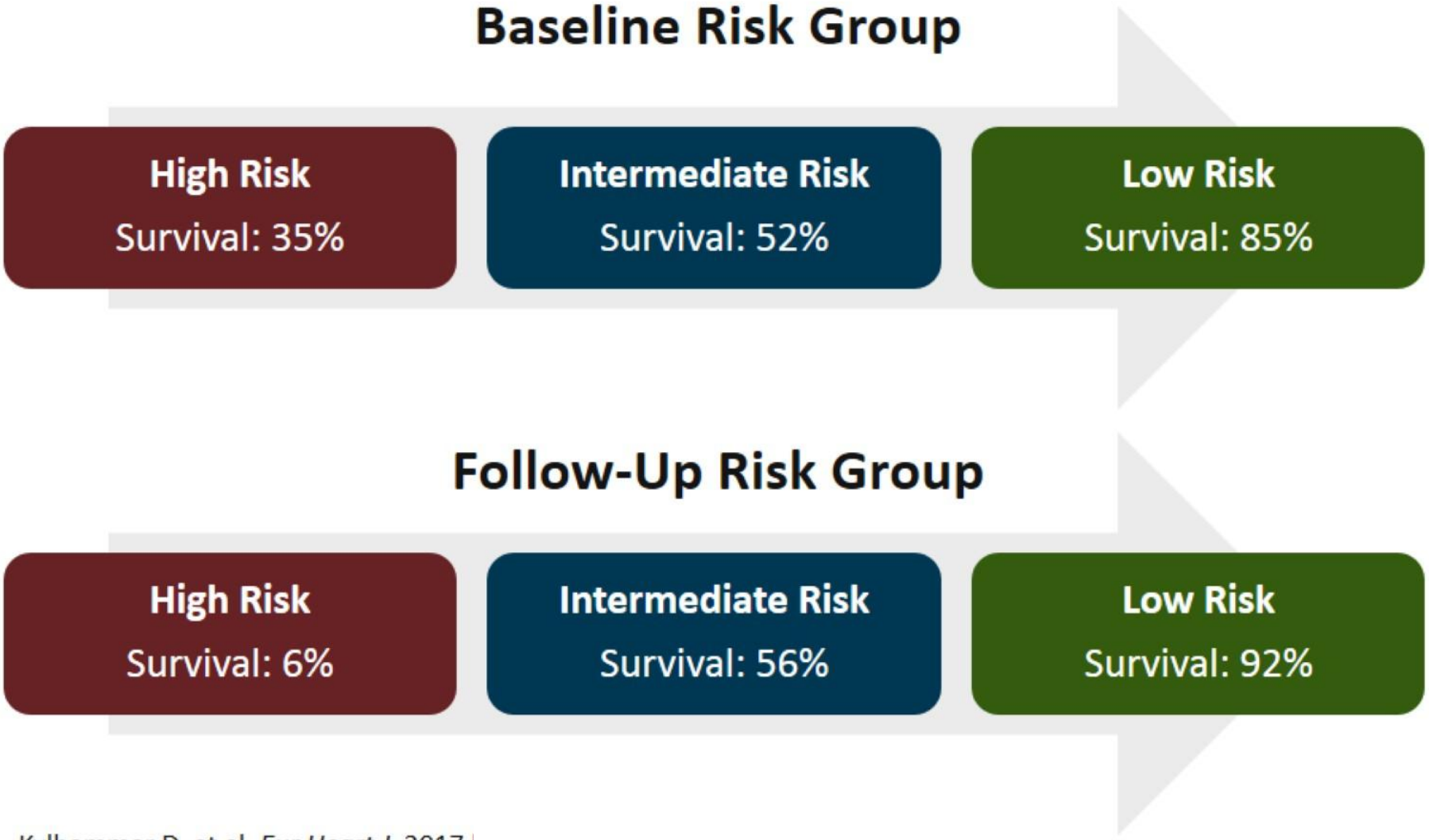
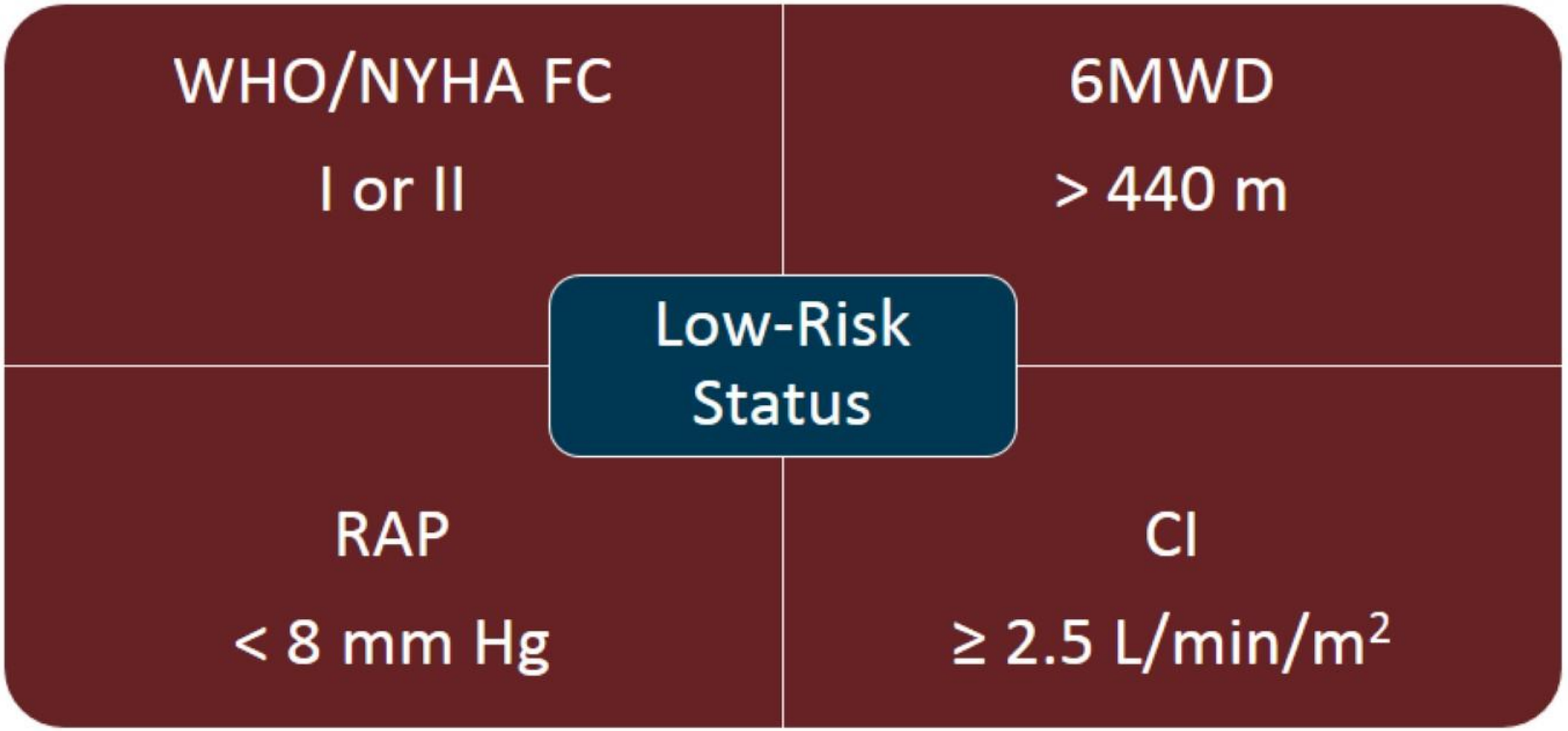
SPAHR
5-Year Survival



Kylhammar D, et al. *Eur Heart J.* 2017

French PH Registry
Low-Risk Criteria

- Determined the association between number of low-risk criteria achieved within 1 year of diagnosis and long-term prognosis



Boucly A, et al. *Eur Respir J.* 2017

French PH Registry
Baseline Characteristics

Patients: 1017	Mean age: 57 years
Female: 59%	Idiopathic PAH: 75%

23% of patients had died
after a median follow-up of 34 months

Boucly A, et al. *Eur Respir J.* 2017

French PH Registry
Number of Low-Risk Criteria Achieved and Survival

- Each low-risk criterion independently predicted transplant-free survival at first reevaluation

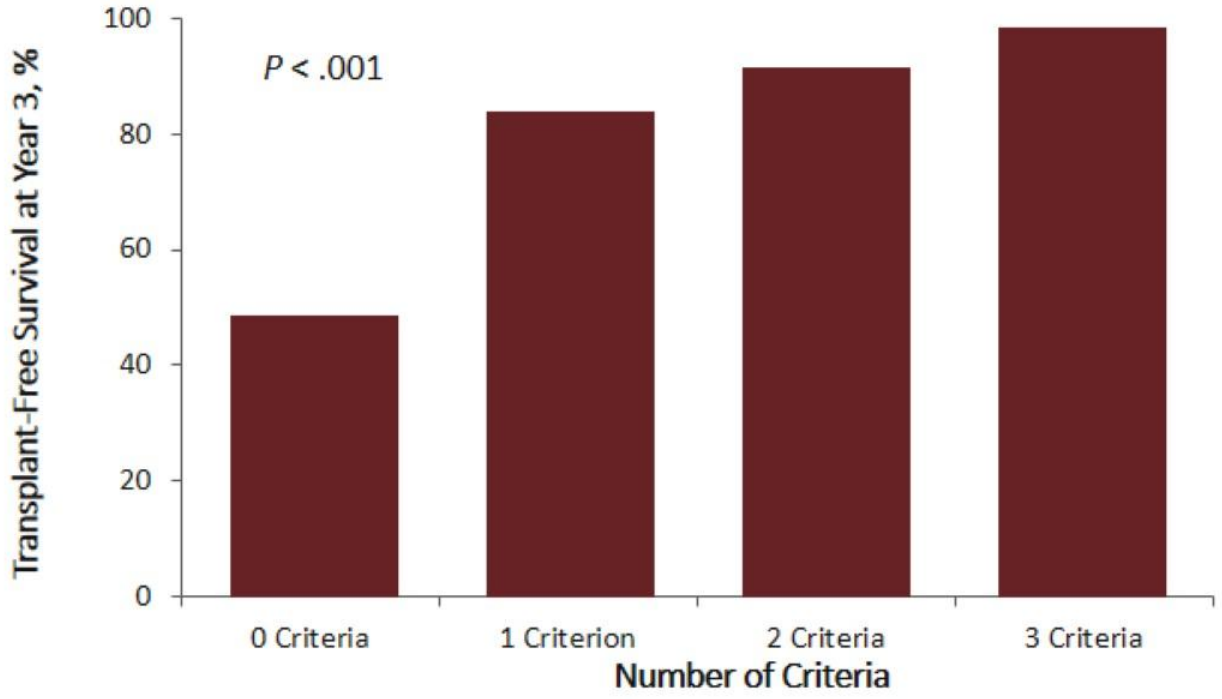
Number of Low-Risk Criteria Achieved at First Re-Evaluation	Transplant-Free Survival at 3 Years, %
4	97
3	93
2	81
1	68
0	40

Boucly A, et al. *Eur Respir J.* 2017

French PH Registry

Number of Noninvasive Low-Risk Criteria Achieved and Survival

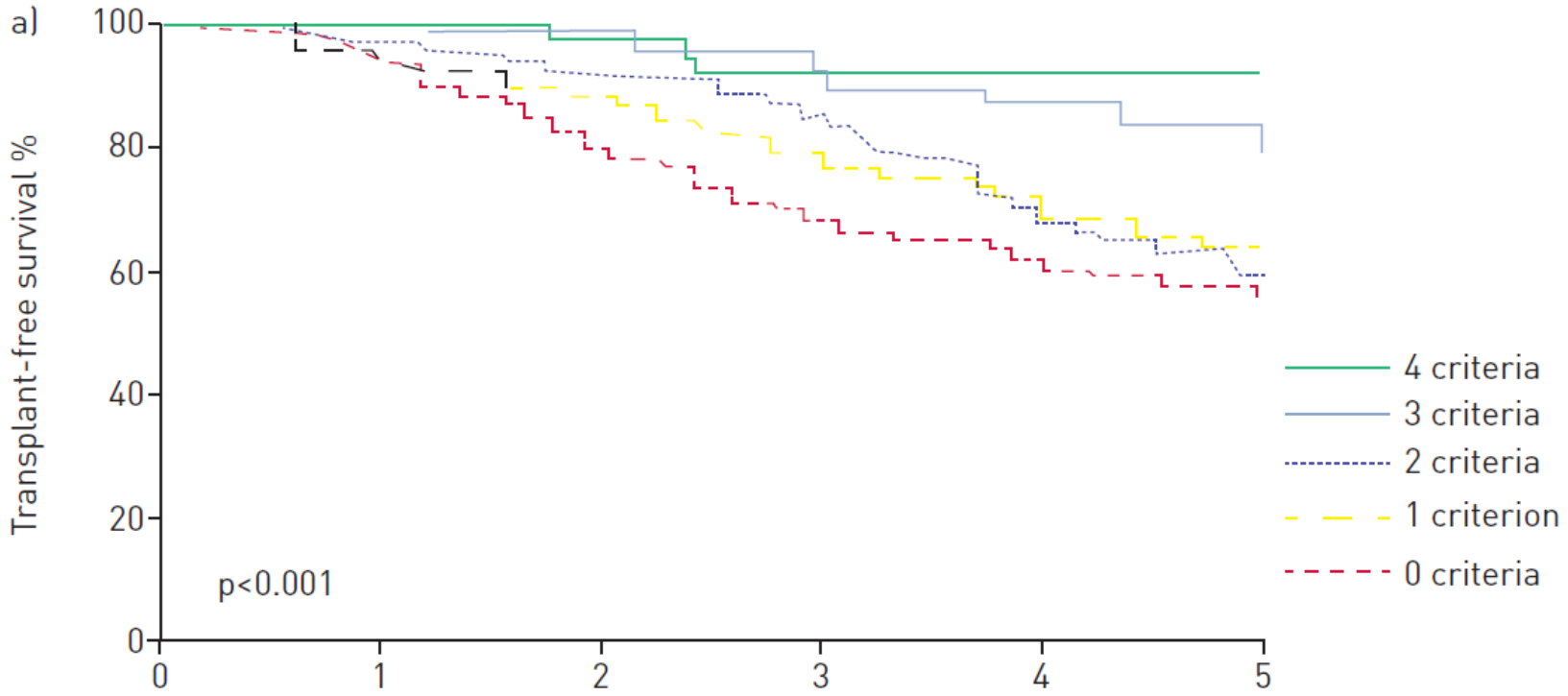
- Noninvasive low-risk criteria: WHO/NYHA FC I to II, 6MWD > 440 m, BNP < 50 ng/L or NT-proBNP < 300 ng/L



Boucly A, et al. *Eur Respir J.* 2017;50:1700889.

Risk assessment, prognosis and guideline implementation in pulmonary arterial hypertension

Athénaïs Boucly[1,2,3], Jason Weatherald[2,3,4], Laurent Savale[1,2,3], Xavier Jaïs[1,2,3], Vincent Cottin[5], Grégoire Prevot[6], François Picard[7], Pascal de Groote[8], Mitja Jevnikar[1,2,3], Emmanuel Bergot[9], Ari Chaouat[10,11], Céline Chabanne[12], Arnaud Bourdin[13], Florence Parent[1,2,3], David Montani[1,2,3], Gérald Simonneau[1,2,3], Marc Humbert[1,2,3] and Olivier Sitbon[1,2,3]



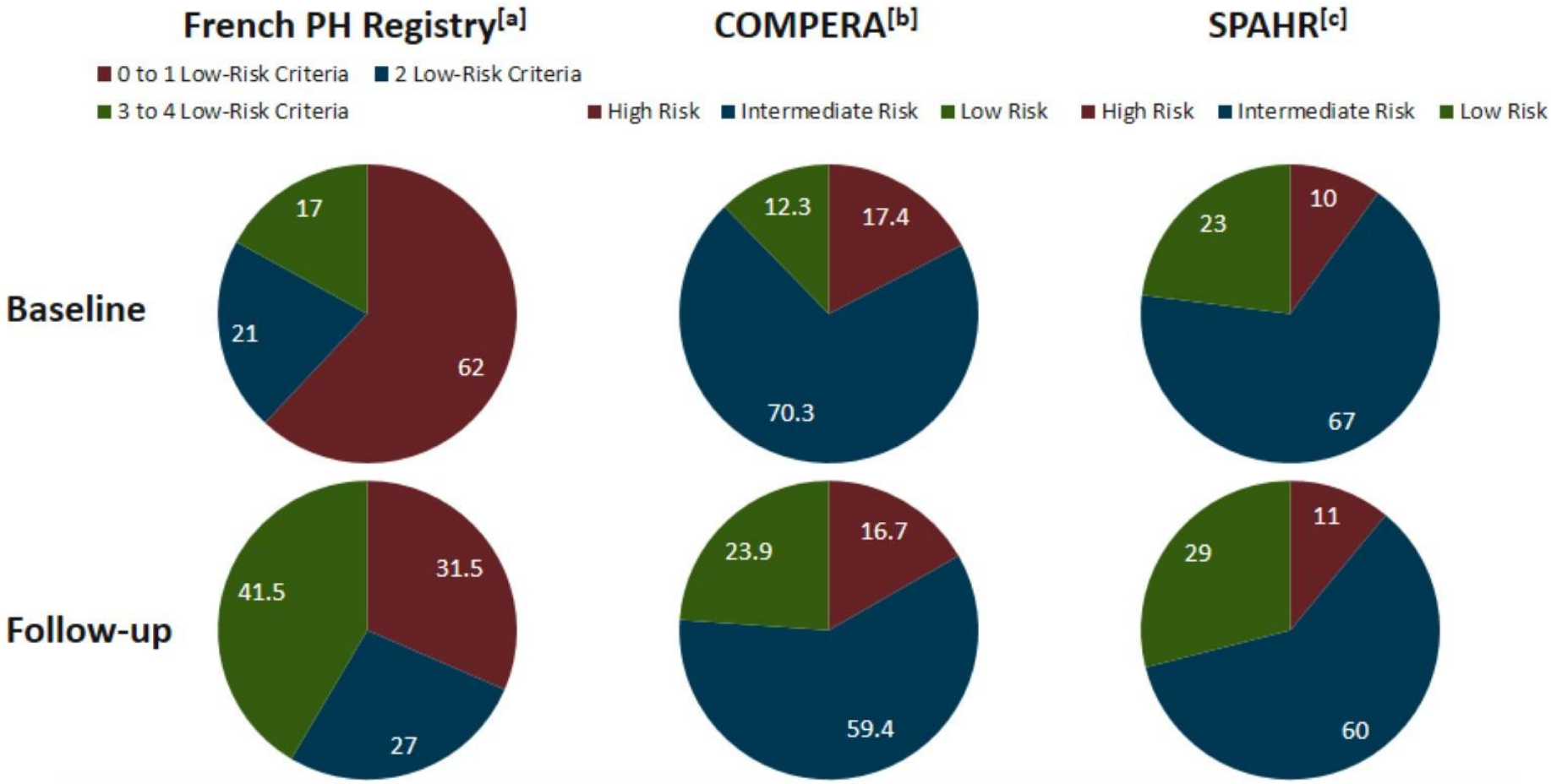
Transplant-free survival according to the number of low-risk criteria (World Health Organization/New York Heart Association functional class I–II I; 6-min walking distance >440 m; right atrial pressure <8 mmHg; cardiac index ⩾2.5 L·min^{-1}·m^{-2}) present at a) time of pulmonary arterial hypertension diagnosis; b) first re-evaluation within the first year after diagnosis.

Pharmacologic Agents for PAH
Multiple Pathways

Endothelin Pathway	Nitric Oxide Pathway	Prostacyclin Pathway
• Selective ERAs (ambrisentan) • Dual ERAs (bosentan, macitentan)	• PDE5 inhibitors (sildenafil, tadalafil) • Soluble guanylate cyclase stimulators (riociguat)	• Prostacyclin analogues (epoprostenol, treprostinil, iloprost) • Nonprostanoid receptor agonists (selexipag)

Humbert M, et al. *Circulation*. 2014;130:2189-2208.

Need for Early Diagnosis and Intensification of Disease Management

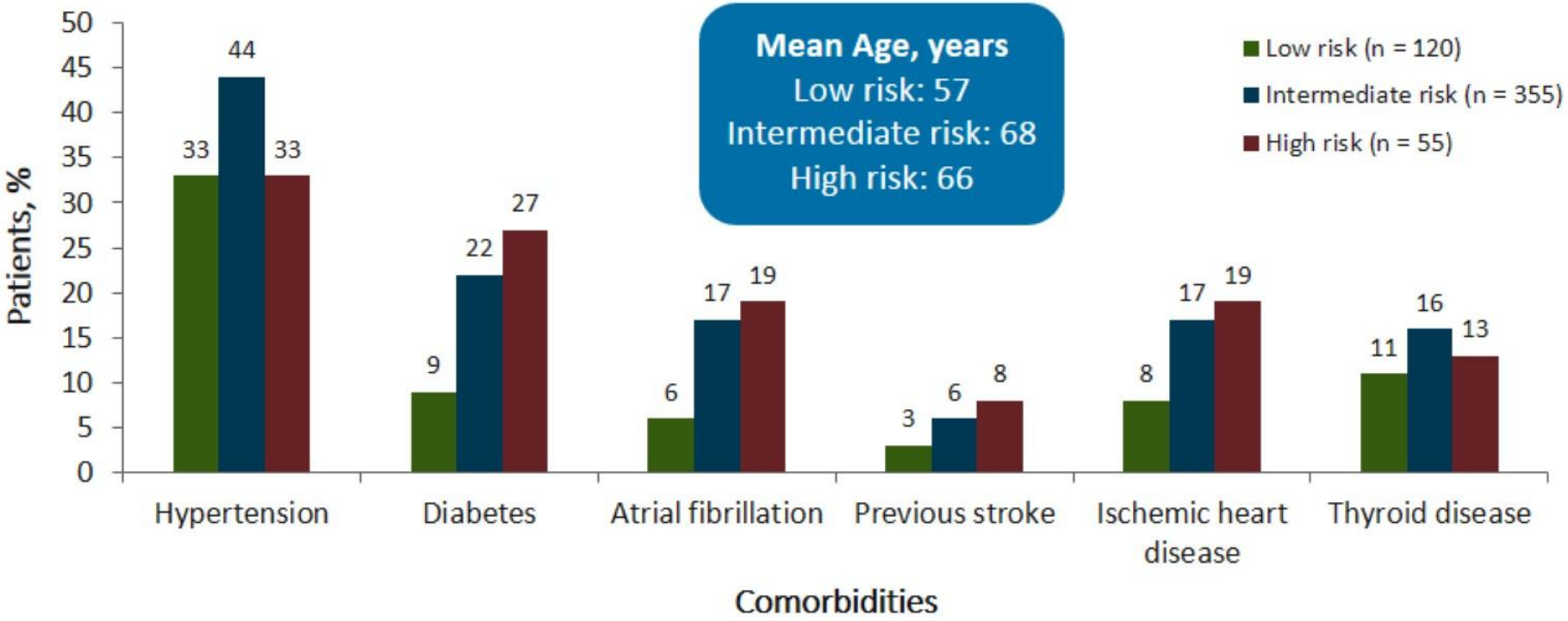


Most patients are diagnosed late in the disease process and do not reach low-risk criteria at first follow-up

a. Boucly A, et al. *Eur Respir J.* 2017;50:1700889; b. Hoeper MM, et al. *Eur Respir J.* 2017;50:1700740; c. Kylhammar D, et al. *Eur Heart J.* 2017

PAH Population Is Heterogeneous

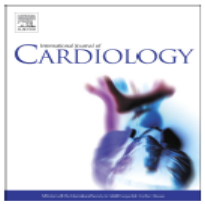
- Baseline patient characteristics from SPAHR



Aggressive initial therapy may not be possible in elderly patients with comorbidities as in younger patients without comorbidities

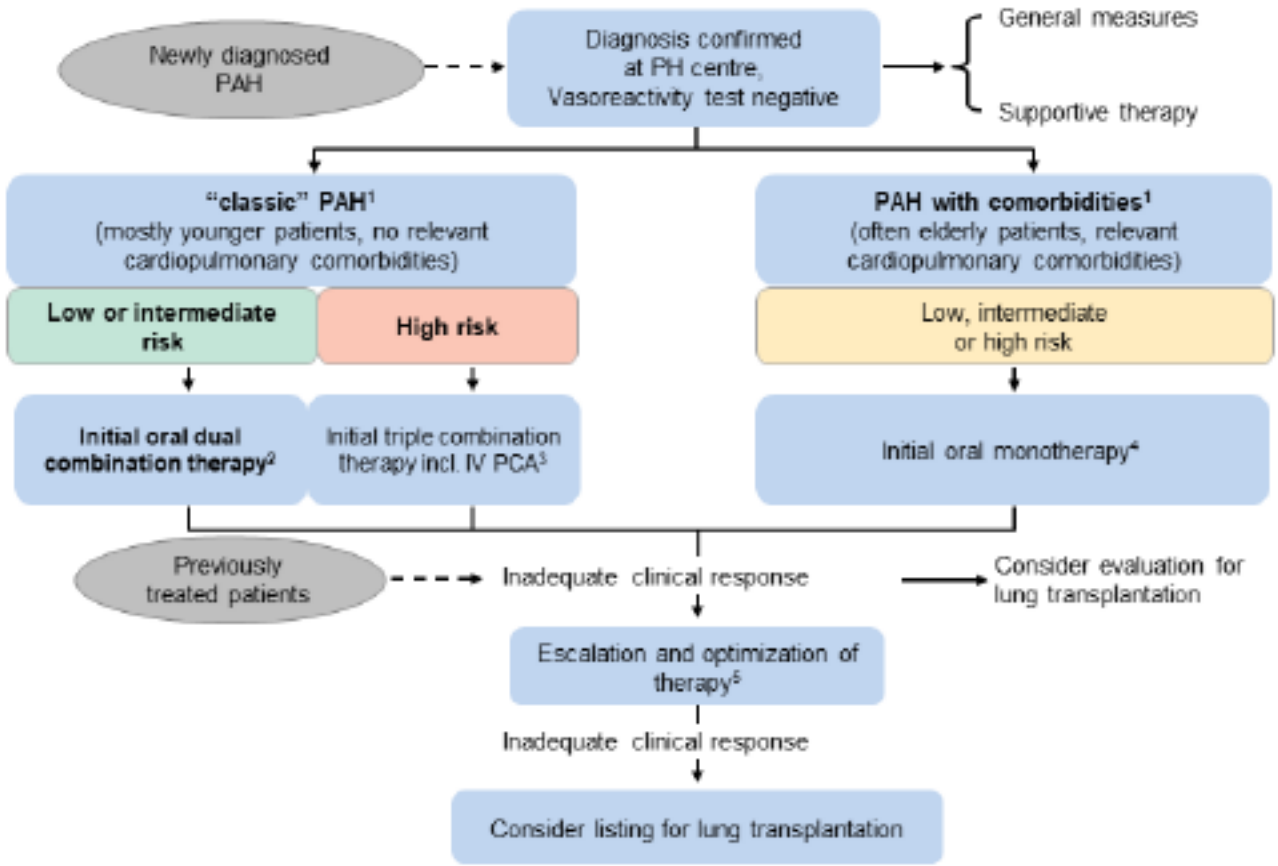
Kylhammar D, et al. *Eur Heart J.* 2017

Accepted Manuscript



Targeted therapy of pulmonary arterial hypertension: Updated recommendations from the Cologne Consensus Conference 2018

Marius M. Hoeper, Christian Apitz, Ekkehard Grünig, Michael Halank, Ralf Ewert, Harald Kaemmerer, Hans-Joachim Kabitz, Christian Kähler, Hans Klose, Hanno Leuchte, Silvia Ulrich, Karen M. Olsson, Oliver Distler, Stephan Rosenkranz, H. Ardeschir Ghofrani



♀ 38yo, SLE-PAH, NYHA II, HR 80bpm, BP 105/74

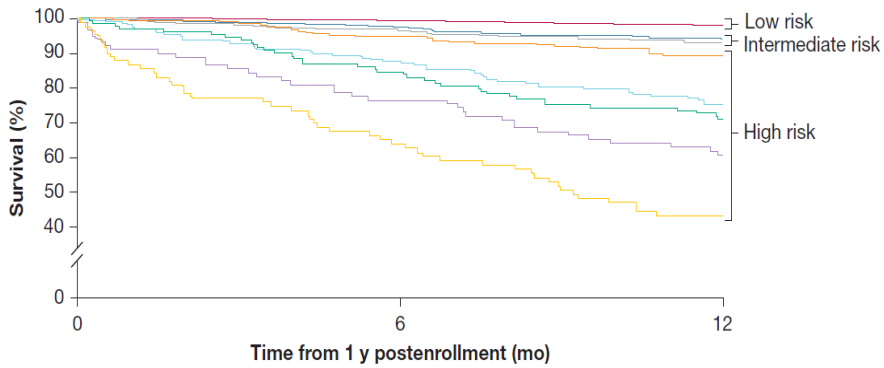
♀ 38yo, SLE-PAH, NYHA II, HR 80bpm, BP 105/74

- SLE-PAH since 2006 on cellcept and steroids
- Triple oral PAH therapy
- Referred for LUTX evaluation with the clinical suspicion of PVOD

REVEAL 2.0 — Updated PAH Risk Score

WHO Group I Subgroup	CTD-PAH	PoPH	Heritable	1
	+1	+3	+2	
Demographics	Males age >60 y			
	+2			
Comorbidities	eGFR <60 mL/min/1.73 m² or renal inefficiency (if eGFR is unavailable)			
	+1			
NYHA/WHO Functional Class	I	III	IV	
	−1	+1	+2	
Vital Signs	SBP <110 mm Hg	HR >96 BPM		1
	+1	+1		
All-cause Hospitalizations ≤6 mo	All-cause hospitalizations within 6 mo			1
	+1			
6-Minute Walk Test	≥440 m	320 to <440 m	<165 m	
	−2	−1	+1	
BNP	<50 pg/mL or NT-proBNP <300 pg/mL	200 to <800 pg/mL	≥800 pg/mL or NT-proBNP ≥1,100 pg/mL	
	−2	+1	+2	
Echocardiogram	Pericardial effusion			
	+1			
Pulmonary Function Test	% predicted D_{LCO} <40%			
	+1			
Right Heart Catheterization	mRAP ≥20 mm Hg within 1 y	PVR <5 Wood units		
	+1	−1		
			SUM OF ABOVE	3
			+	6
			= RISK SCORE	9

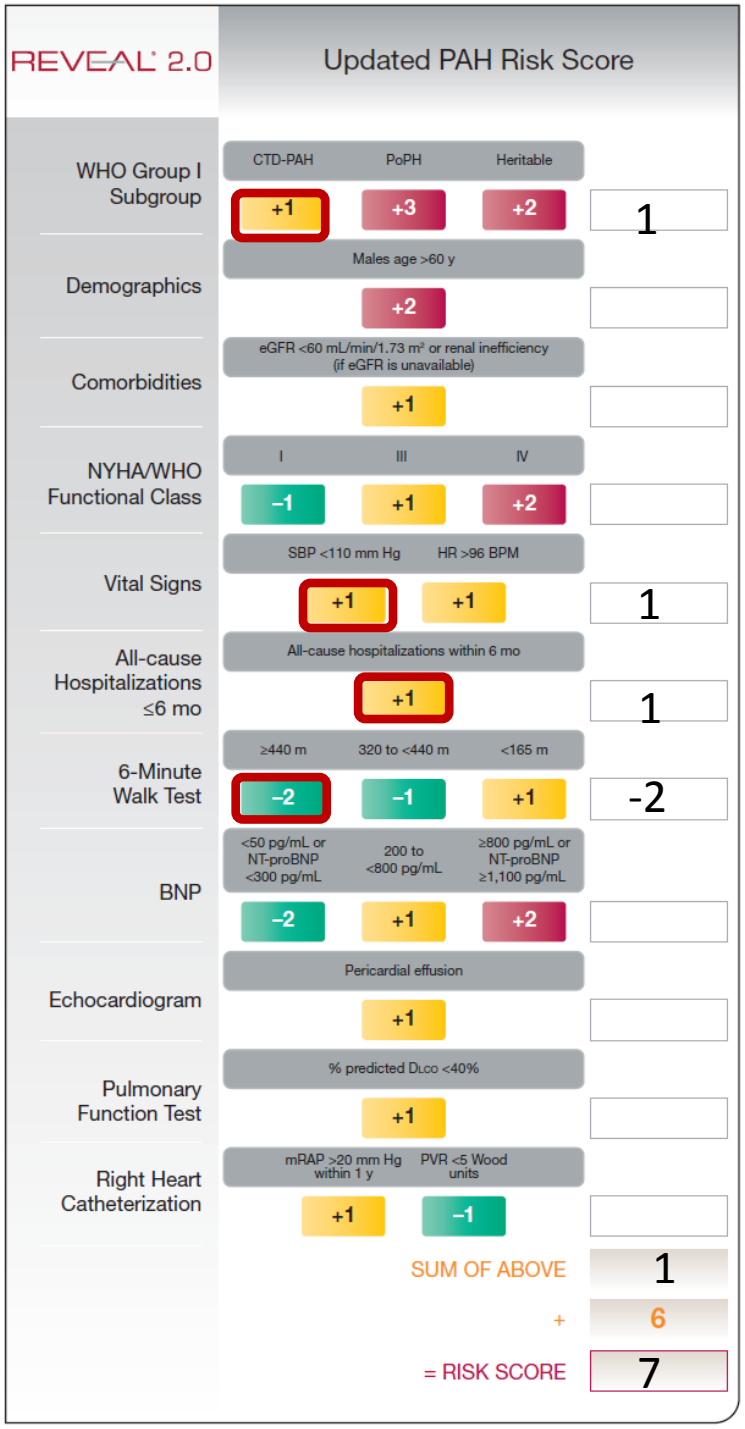
A



No. at risk

Score = 0-6	1,073	1,056	1,032
Score = 7	386	368	344
Score = 8	306	292	275
Score = 9	266	248	230
Score = 10	195	166	142
Score = 11	130	109	87
Score = 12	90	68	54
Score = 13+	83	53	34

Score = 0-6 — Score = 7 — Score = 8 — Score = 9
Score = 10 — Score = 11 — Score = 12 — Score = 13+


REVEAL 2.0
Updated PAH Risk Score
WHO Group I Subgroup
CTD-PAH
PoPH
Heritable
+1
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Males age >60 y
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Comorbidities
eGFR <60 mL/min/1.73 m² or renal inefficiency (if eGFR is unavailable)
+1
NYHA/WHO Functional Class
I
III
IV
–1
+1
+2
Vital Signs
SBP <110 mm Hg
HR >96 BPM
+1
+1
1
All-cause Hospitalizations ≤6 mo
All-cause hospitalizations within 6 mo
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Pericardial effusion
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% predicted DLCO <40%
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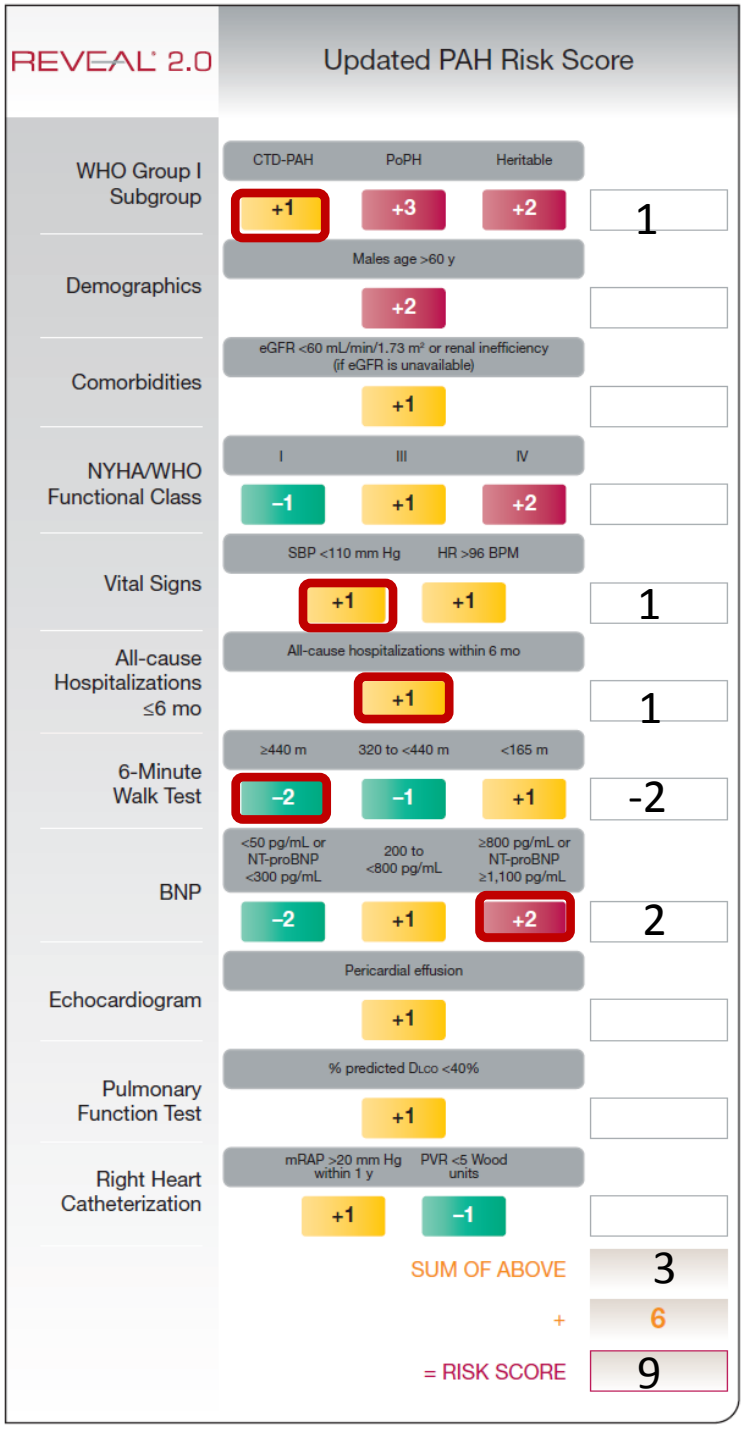
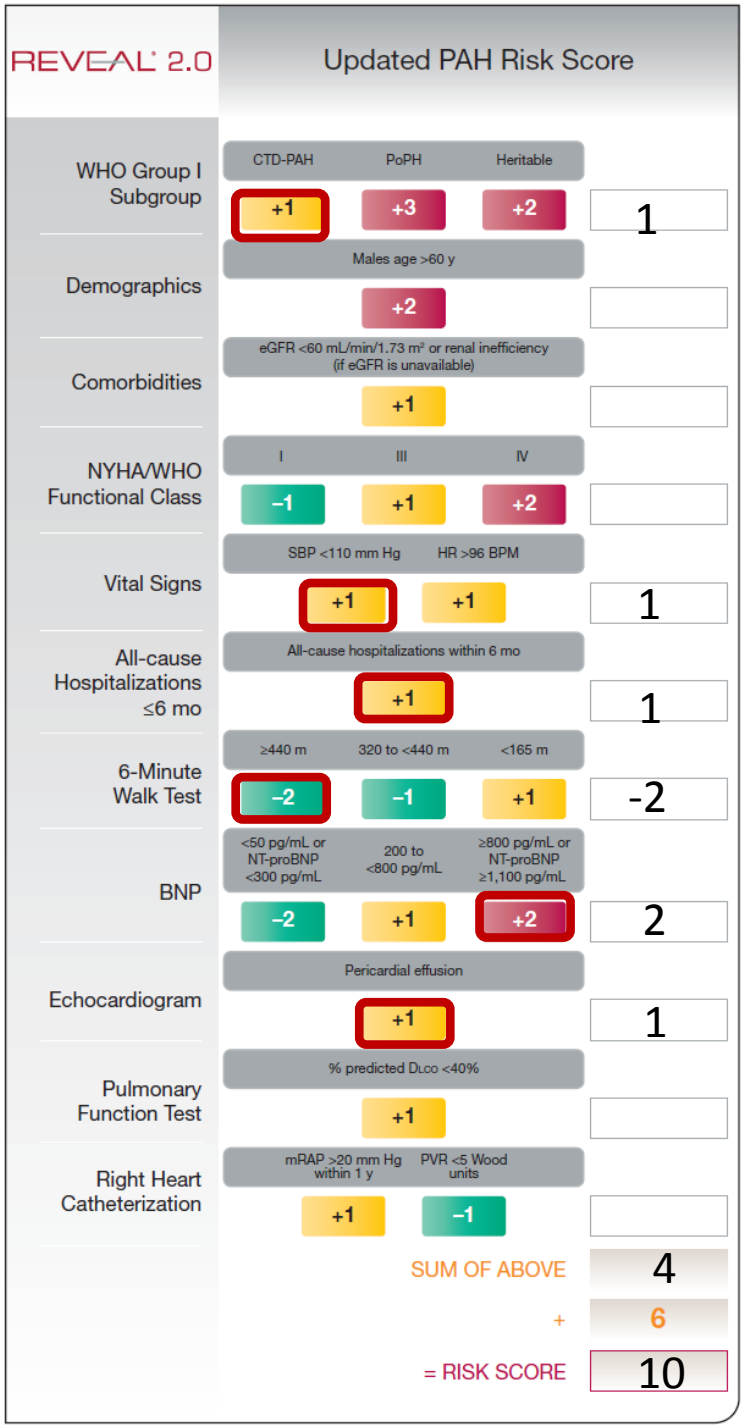
PVR <5 Wood units
+1
–1
SUM OF ABOVE
1
+
6
= RISK SCORE
7

6min WT: 483m



6min WT: 483m

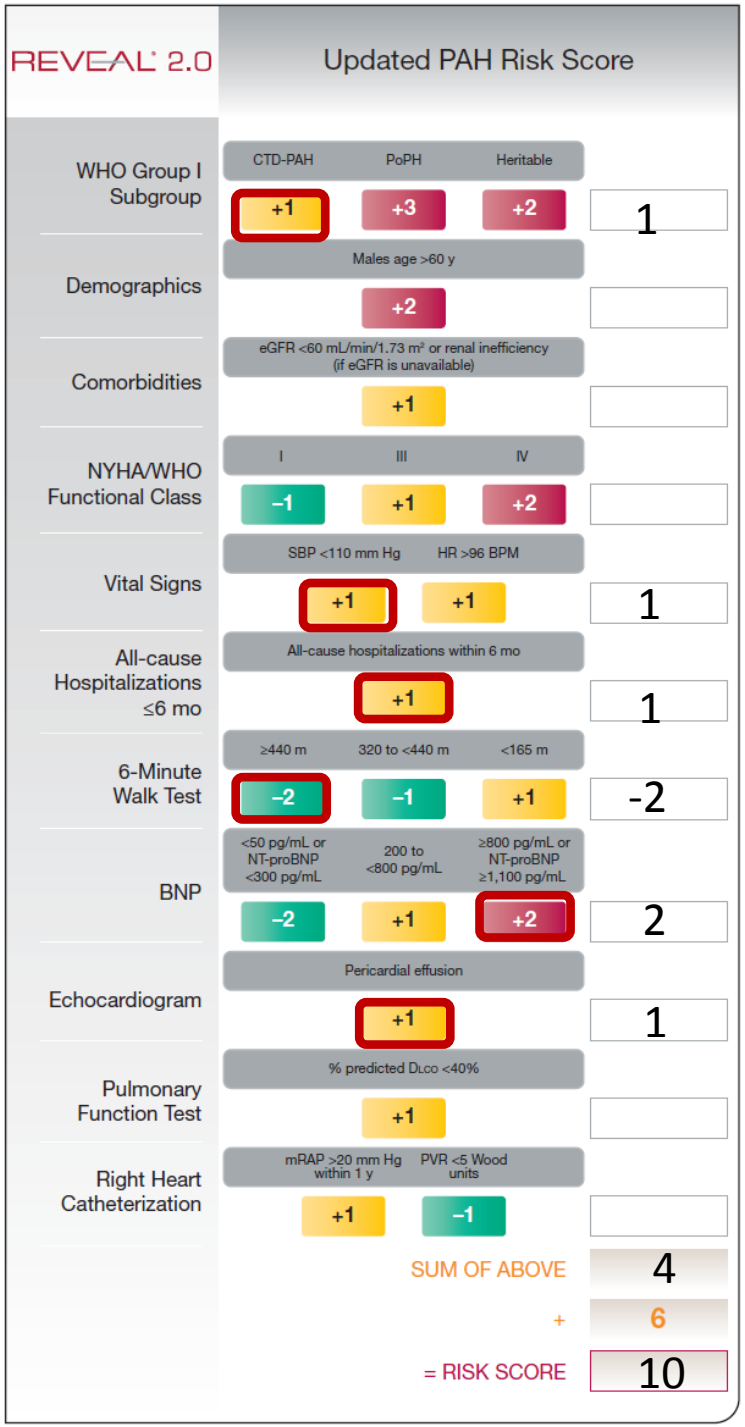
NT-proBNP: 1350



6min WT: 483m

NT-proBNP: 1350

US: TAPSE 17mm, TR 4.4m/sec, RV 60mm, pericardial effusion



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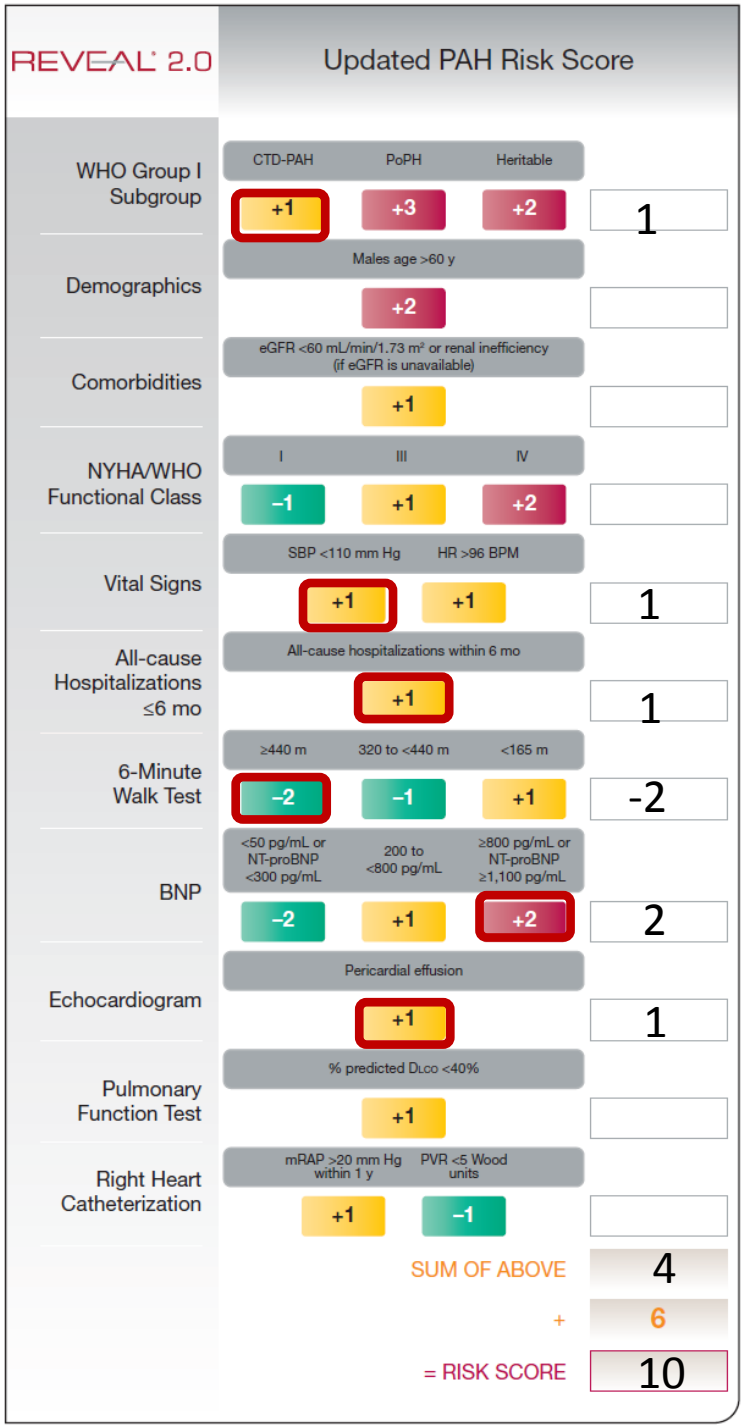
RHC: RAP 5mmHg, mPAP 44mmHg, CI 3L/m^2, PVR 8WU



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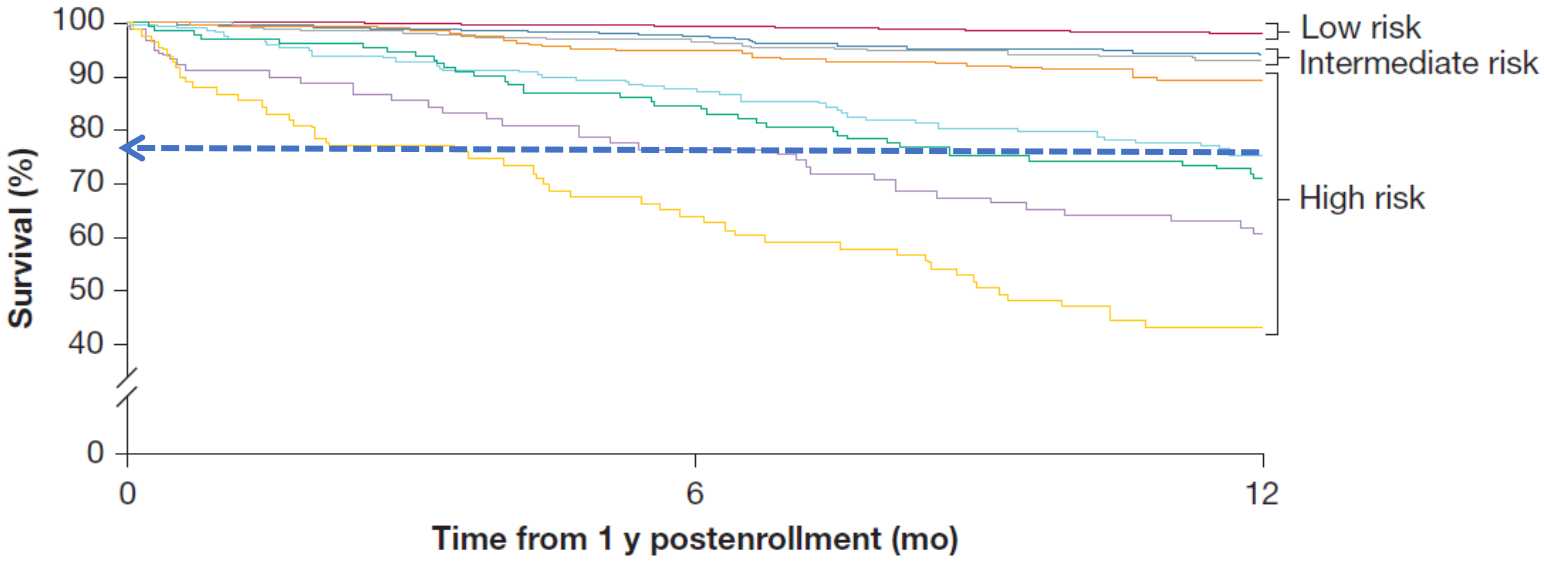
Lung perfusion scan: normal

A



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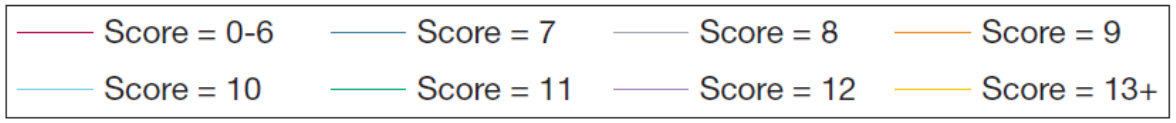
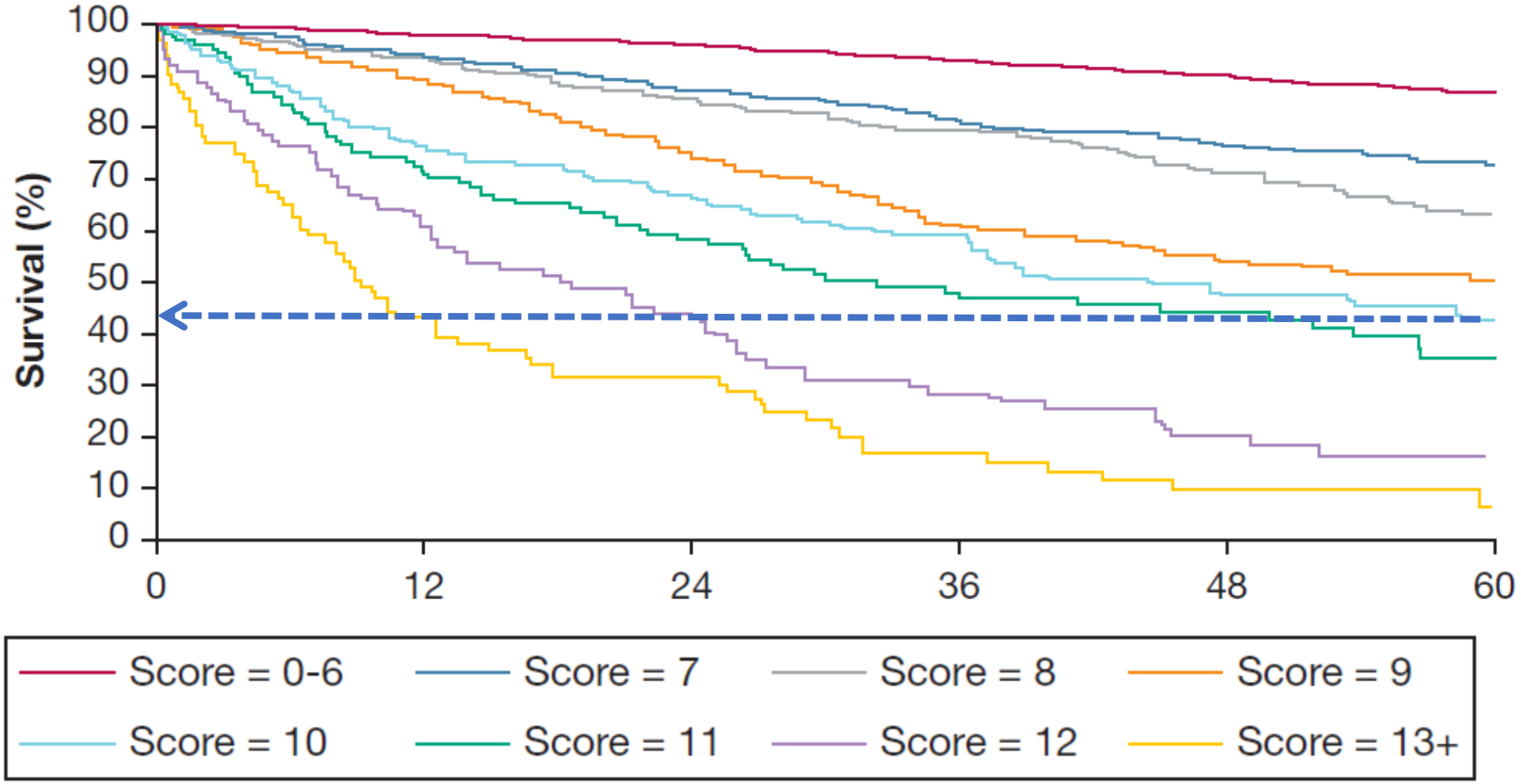


Predicting Survival in Patients With Pulmonary Arterial Hypertension

The REVEAL Risk Score Calculator 2.0 and Comparison With ESC/ERS-Based Risk Assessment Strategies

Raymond L. Benza, MD; Mardi Gomberg-Maitland, MD; C. Greg Elliott, MD; Harrison W. Farber, MD; Aimee J. Foreman, MA; Adaani E. Frost, MD; Michael D. McGoon, MD; David J. Pasta, MS; Mona Selej, MD; Charles D. Burger, MD; and Robert P. Frantz, MD



Risk Assessment in PAH

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	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 36–44.9	Peak VO_2 <11 ml/min/kg (<35 % pred.) VE/VCO_2 ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/ml	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65 %	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60 %

[a]Most of the proposed variables and cut-off values are based on expert opinion.
[b]Occasional syncope during brisk or heavy exercise, or occasional orthostatic syncope in an otherwise stable patient.
[c]Repeated episodes of syncope, even with little or regular physical activity.

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1 2 3

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COMPERA score: 13/8= 1,63

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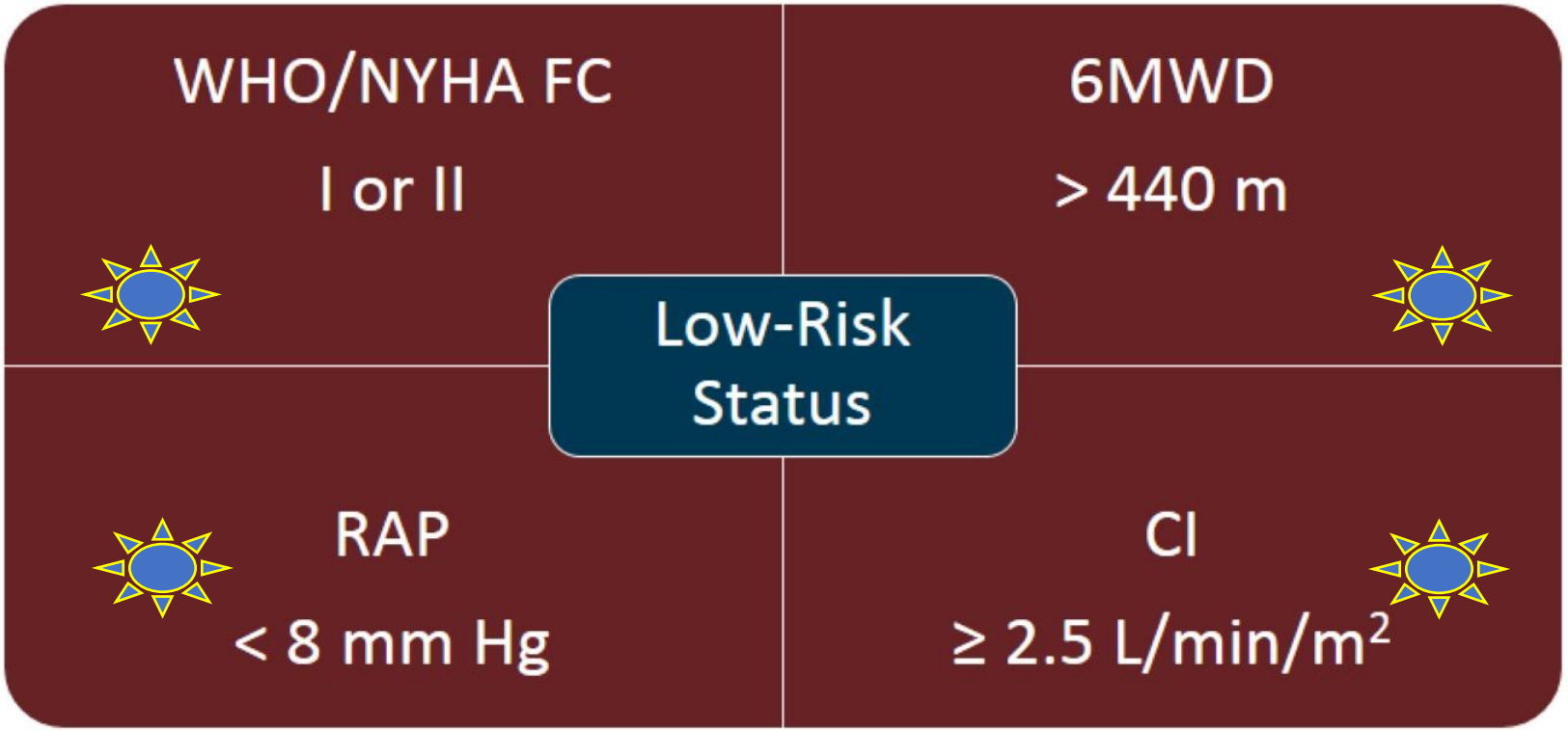

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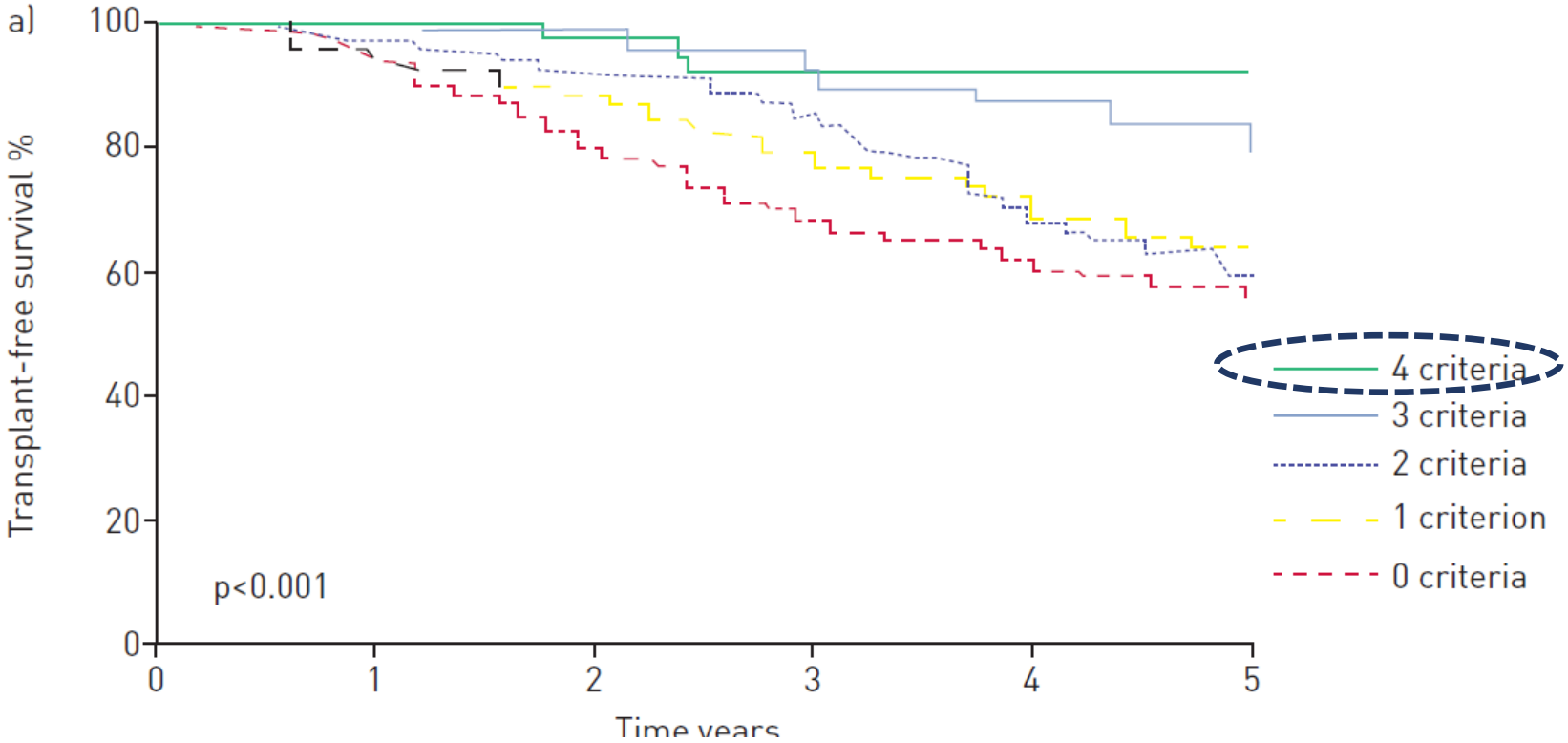

FIGURE 3 Transplant-free survival according to the number of low-risk criteria (World Health Organization/New York Heart Association functional class I–II I; 6-min walking distance >440 m; right atrial pressure <8 mmHg; cardiac index ⩾2.5 L·min^{-1}·m^{-2}) present at a) time of pulmonary arterial hypertension diagnosis; b) first re-evaluation within the first year after diagnosis.

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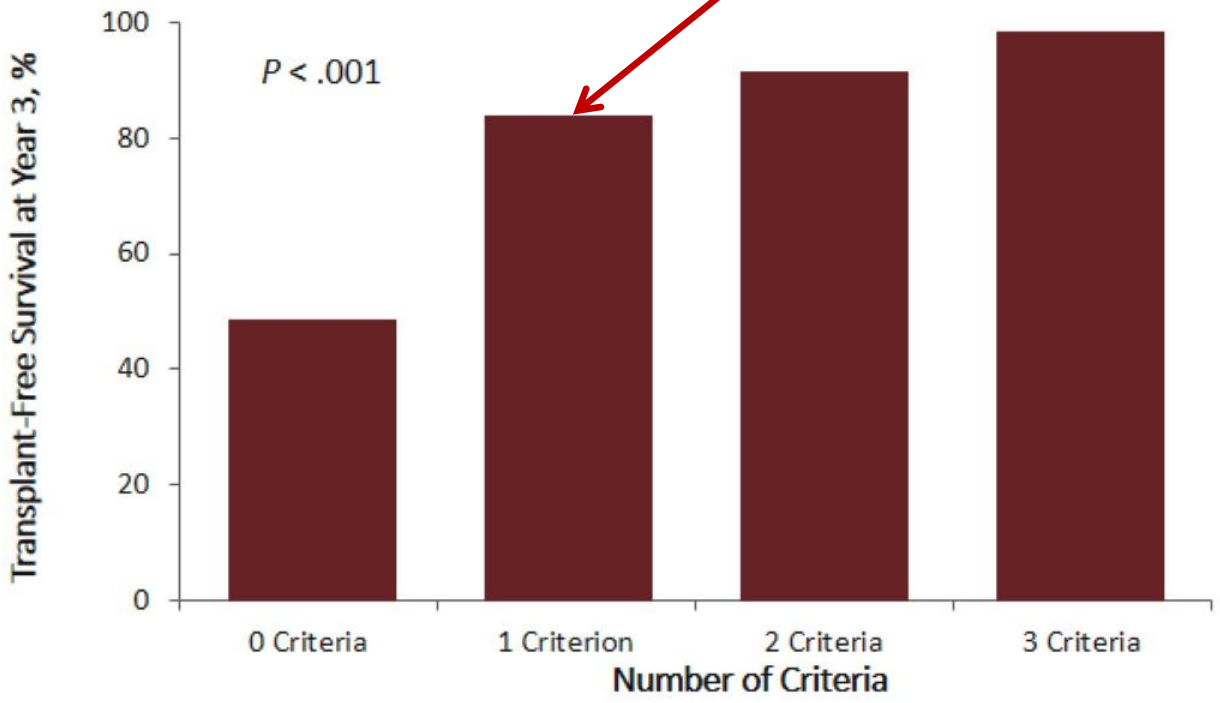

Boucly A, et al. *Eur Respir J.* 2017;50:1700889.

Upfront triple combination therapy in pulmonary arterial hypertension: a pilot study

Olivier Sitbon[1,2,3], Xavier Jaïs[1,2,3], Laurent Savale[1,2,3], Vincent Cottin[4], Emmanuel Bergot[5], Elise Artaud Macari[1,2,3], Hélène Bouvaist[6], Claire Dauphin[7], François Picard[8], Sophie Bulifon[1,2,3], David Montani[1,2,3], Marc Humbert[1,2,3] and Gérald Simonneau[1,2,3]

Subjects	19
Age years (range)	39.4±14.2 (18.1–63.1)
Females	17 (89)
Idiopathic/heritable/anorexigen-associated PAH	9/10/0
***BMPR2* mutation carrier n/n tested (%)**	10/13 (77)
NYHA functional class III/IV	8 (42)/11 (58)
6-min walk distance m	215±174
Haemodynamics	
Right atrial pressure mmHg	12.2±5.2
Mean pulmonary arterial pressure mmHg	67.7±15.8
Pulmonary capillary wedge pressure mmHg	8.3±3.4
Cardiac output $L \cdot min^{-1}$	2.83±0.77
Cardiac index $L \cdot min^{-1} \cdot m^{-2}$	1.64±0.34
Pulmonary vascular resistance $dyn \cdot s \cdot cm^{-5}$	1807±722
Mean blood pressure mmHg	91.7±12.2
Heart rate beats per min	92.3±10.7
Mixed venous oxygen saturation %	50.1±9.0

: 0 *Low-Risk Criteria*

French PH Registry
Low-Risk Criteria

- Determined the association between number of low-risk criteria achieved within 1 year of diagnosis and long-term prognosis

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Risk assessment, prognosis and guideline implementation in pulmonary arterial hypertension

Athénaïs Boucly[1,2,3], Jason Weatherald[2,3,4], Laurent Savale[1,2,3], Xavier Jaïs[1,2,3], Vincent Cottin[5], Grégoire Prevot[6], François Picard[7], Pascal de Groote[8], Mitja Jevnikar[1,2,3], Emmanuel Bergot[9], Ari Chaouat[10,11], Céline Chabanne[12], Arnaud Bourdin[13], Florence Parent[1,2,3], David Montani[1,2,3], Gérald Simonneau[1,2,3], Marc Humbert[1,2,3] and Olivier Sitbon[1,2,3]

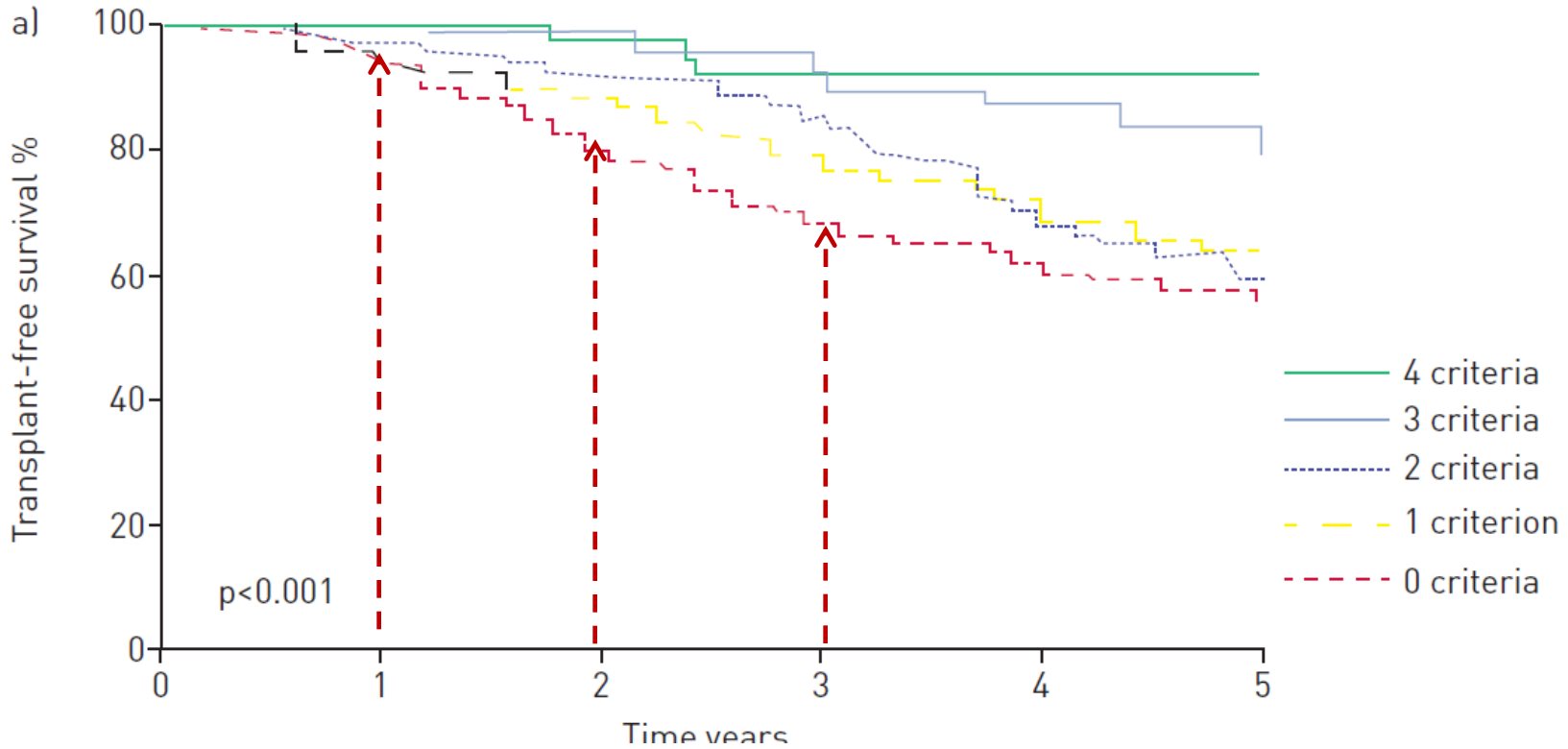


FIGURE 3 Transplant-free survival according to the number of low-risk criteria (World Health Organization/New York Heart Association functional class I–II I; 6-min walking distance >440 m; right atrial pressure <8 mmHg; cardiac index ⩾2.5 $L \cdot min^{-1} \cdot m^{-2}$) present at a) time of pulmonary arterial hypertension diagnosis; b) first re-evaluation within the first year after diagnosis.

Upfront triple combination therapy in pulmonary arterial hypertension: a pilot study

Olivier Sitbon[1,2,3], Xavier Jaïs[1,2,3], Laurent Savale[1,2,3], Vincent Cottin[4], Emmanuel Bergot[5], Elise Artaud Macari[1,2,3], Hélène Bouvaist[6], Claire Dauphin[7], François Picard[8], Sophie Bulifon[1,2,3], David Montani[1,2,3], Marc Humbert[1,2,3] and Gérald Simonneau[1,2,3]

	Baseline	Month 4 visit	Final follow-up visit#
NYHA FC I/II/III/IV n	0/0/8/10	1/16/1/0**	4/14/0/0**
6MWD m	227±171	463±94**	514±105**,¶
Haemodynamics			
RAP mmHg	11.9±5.2	4.9±4.9**	5.2±3.5**
mPAP mmHg	65.8±13.7	45.7±14.0**	44.4±13.4**
PCWP mmHg	8.4±3.5	6.7±3.2	7.9±2.8
Cardiac index $L \cdot min^{-1} \cdot m^{-2}$	1.66±0.35	3.49±0.69**	3.64±0.65**
PVR $dyn \cdot s \cdot cm^{-5}$	1718±627	564±260**	492±209**
Mean BP mmHg	92.1±12.5	80.1±11.7**	84.9±19.4
HR beats per min	92.3±10.7	83.9±9.8**	79.9±13.4**
Sv_{O_2} %	51.0±8.5	69.7±5.2**	72.2±4.0**
Dose of epoprostenol achieved $ng \cdot kg^{-1} \cdot min^{-1}$	0	15.9±1.9	19.6±6.0

- All patients initiated with upfront triple combination therapy were still alive after a mean follow up of 41.2±13.4 months.
- Overall survival estimates were 100%, 100% and 100% at 1, 2 and 3 years, and respective transplant-free survival estimates were 94%, 94% and 94%.
- Expected survival calculated from the French equation was 75% (95% CI 68–82%), 60% (95% CI 50–70%) and 49% (95% CI 38–60%) at 1, 2 and 3 years, respectively.

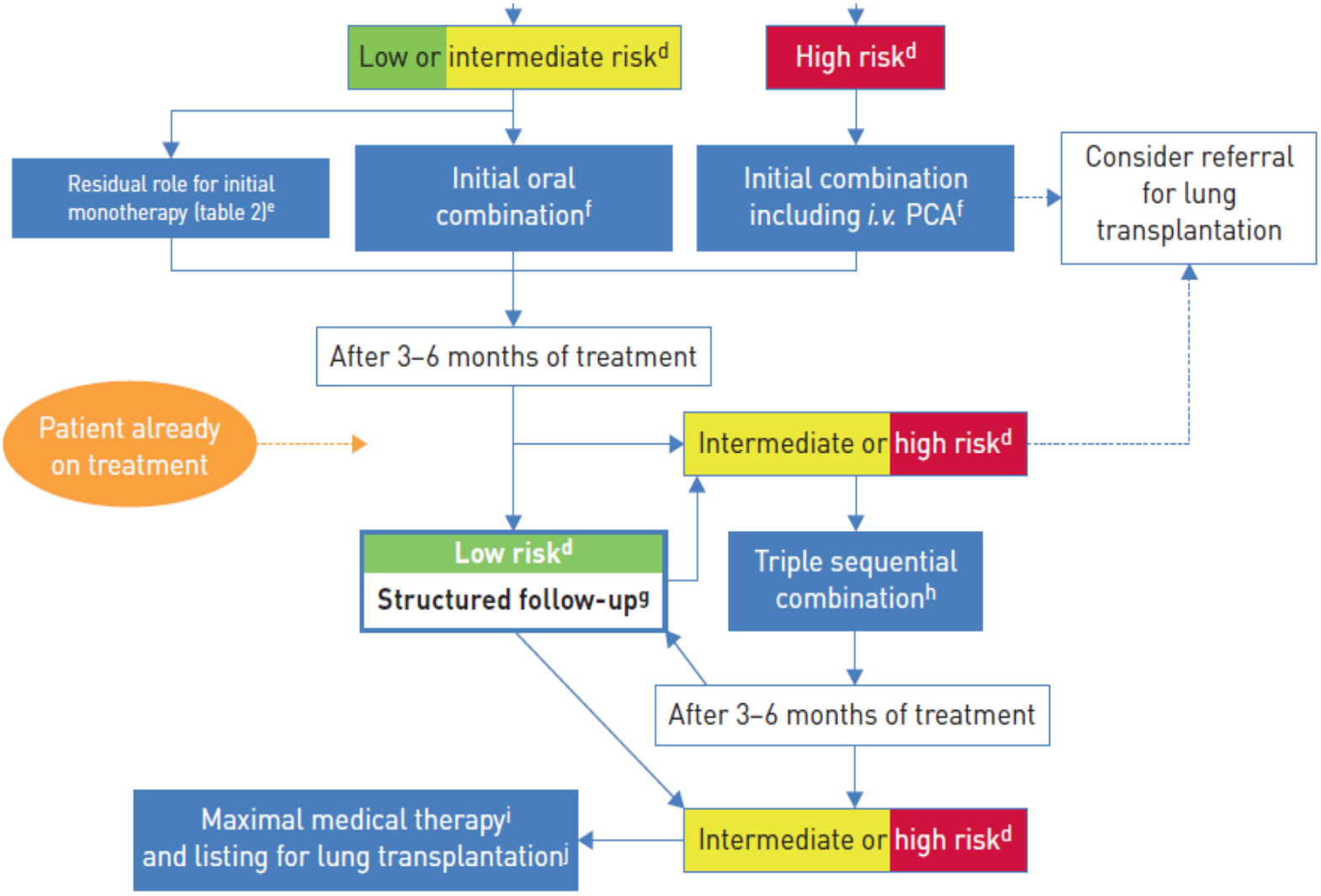
Eur Respir J 2014; 43: 1691–1697

Long-term patient survival with idiopathic/heritable pulmonary arterial hypertension treated at a single center in Japan

Aiko Ogawa [a], Kentaro Ejiri [b], Hiromi Matsubara [a,b,*]

- Extremely high doses of intravenous epoprostenol, (>100 ng/ kg/min) decreased significantly mean PAP from 63± 15 mmHg to 35± 10 mmHg by approximately 4 years.
- 1-, 2-, 3-, 5-, and 10-year survival rates were 98%, 96%, 96%, 96%, and 78% respectively.

Ogawa A, Ejiri K, Matsubara H. Long-term patient survival with idiopathic/ heritable pulmonary arterial hypertension treated at a single center in Japan. Life Sci 2014;118:414–9.


Low or intermediate risk[d]
High risk[d]
Residual role for initial monotherapy (table 2)[e]
Initial oral combination[f]
Initial combination including *i.v.* PCA[f]
Consider referral for lung transplantation
After 3–6 months of treatment
Patient already on treatment
Intermediate or high risk[d]
Low risk[d]
Structured follow-up[g]
Triple sequential combination[h]
After 3–6 months of treatment
Maximal medical therapy[i] and listing for lung transplantation[j]
Intermediate or high risk[d]

Adult Lung Transplants

Diagnoses (Transplants: January 1995 – June 2018)

Diagnosis	SLT (N=19,958)	BLT (N=43,572)	TOTAL (N=63,530)
COPD	7,750 (38.8%)	11,402 (26.2%)	19,152 (30.1%)
IIP	7,536 (37.8%)	9,047 (20.8%)	16,583 (26.1%)
CF	227 (1.1%)	9,447 (21.7%)	9,674 (15.2%)
ILD-not IIP	1,123 (5.6%)	2,486 (5.7%)	3,609 (5.7%)
A1ATD	814 (4.1%)	2,155 (4.9%)	2,969 (4.7%)
Retransplant	1,003 (5.0%)	1,553 (3.6%)	2,556 (4.0%)
IPAH	95 (0.5%)	1,768 (4.1%)	1,863 (2.9%)
Non CF-bronchiectasis	77 (0.4%)	1,637 (3.8%)	1,714 (2.7%)
Sarcoidosis	343 (1.7%)	1,197 (2.7%)	1,540 (2.4%)
PH-not IPAH	140 (0.7%)	838 (1.9%)	978 (1.5%)
LAM/tuberous sclerosis	161 (0.8%)	420 (1.0%)	581 (0.9%)
CTD	169 (0.8%)	395 (0.9%)	564 (0.9%)
OB	79 (0.4%)	460 (1.1%)	539 (0.8%)
Cancer	8 (0.0%)	30 (0.1%)	38 (0.1%)
Other	433 (2.2%)	737 (1.7%)	1,170 (1.8%)

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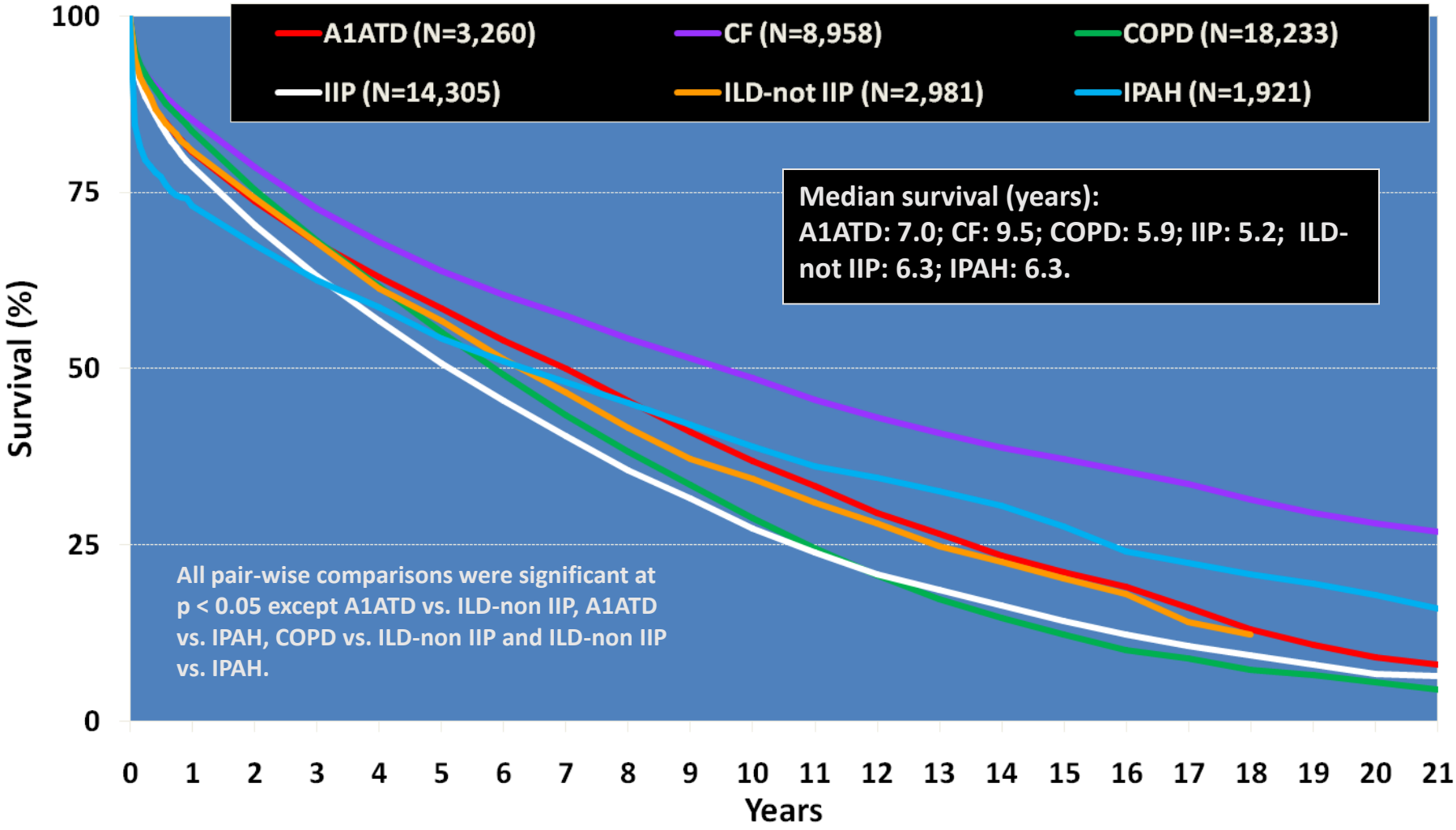
Case Mix (last 6 years)

- 2019: CF 3, PAH 2
- 2018: CF 3
- 2017: CF 9
- 2016: CF 5, PAH 1, A_1-AT 1
- 2015: CF 2, PAH 1, PF 3, ReTx 1
- 2014 :CF4, ReTx 1

- CF 27pts - PF 4pts - ReTx 2pts - PAH 2pts - A_1AT 1pt

Adult Lung Transplants
Kaplan-Meier Survival by Major Diagnosis
(Transplants: January 1990 – June 2016)



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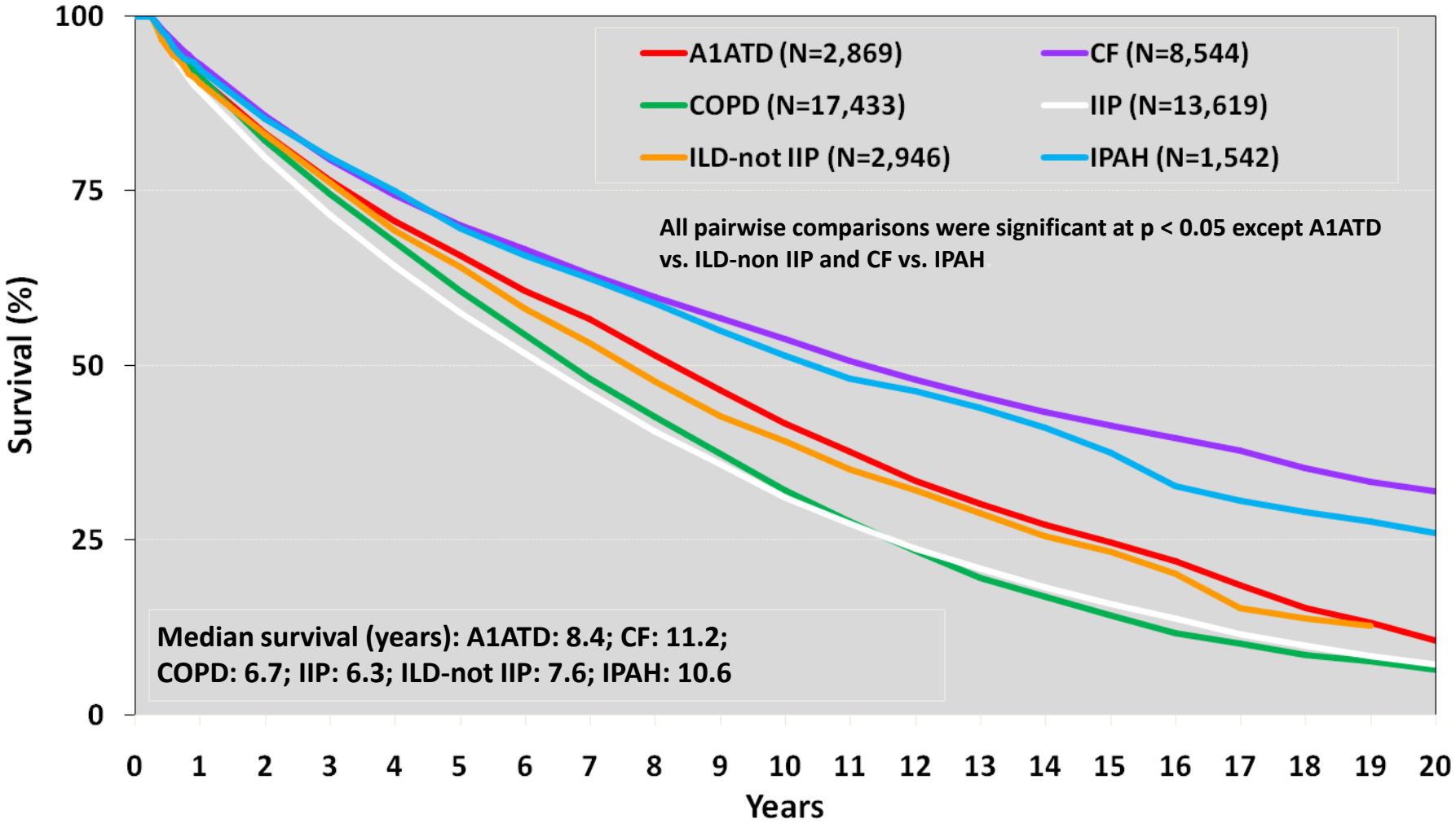
JHLT. 2018 Oct; 37(10): 1155-1206

Adult Lung Transplants

Kaplan-Meier Survival by Major Diagnosis Conditional on

Survival to 3 Months **(Transplants: January 1992 – June 2017)**



Πνευμονικά μοσχεύματα (1/1/17-13/12/2019)

- **35** Πνευμονικά Μοσχεύματα

- ΑΤΤΙΚΟ: 8
- Παπανικολάου: 4
- Παπαγεωργίου: 3
- Καβάλα, Κέρκυρα: 2
- 16 νοσοκομεία από 1
- 8/16 εκτός ΑΘΗ-ΘΕΣ

Δότες Αθήνας (5.000.000 πληθυσμός) 1/1/19-13/12/2019

- **18** Δότες

- ΑΤΤΙΚΟ: 8

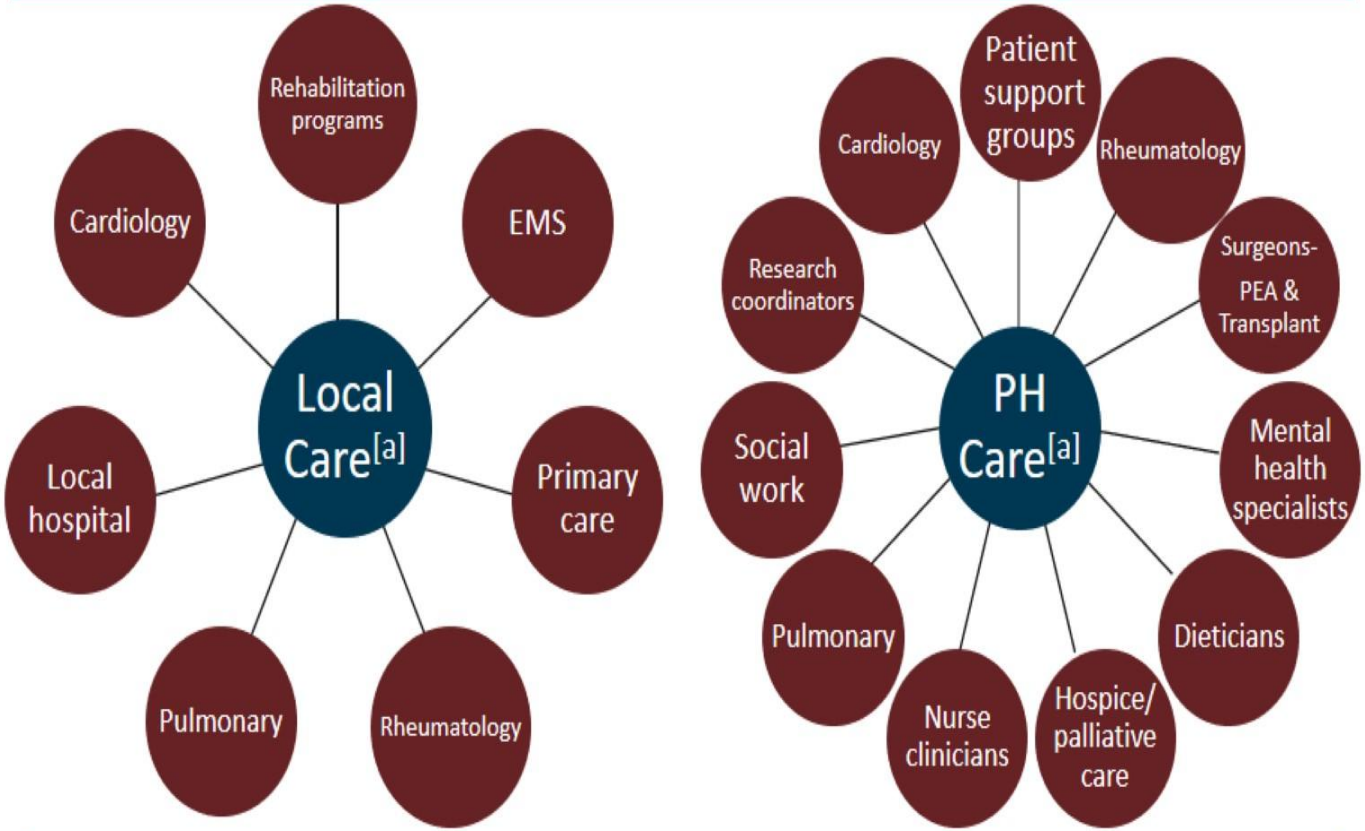
- ΩΚΚ, Ντυνάν: 2

- Υγεία, Κεντρική Κλινική, Ιατρικό Αμαρουσίου, Ευαγγελισμός, Ιπποκράτειο, Ερυθρός: 1

In conclusion

- Multi-parameter risk assessment is essential to determine prognosis and define the optimum treatment strategy for all patients with PAH
- Recent studies have provided strong evidence to support multi-parameter risk assessment in PAH pts at baseline and at follow-up
- The ultimate goal of treatment is to achieve a low risk profile

Coordination of Local and PH Center of Care
Collaborative Approach



Patients should be provided with access to accredited specialist centers which provide a multidisciplinary approach where there is a culture focused on narrative medicine, QoL, shared decision making and timely access to palliative care, and where there is participation in education[b]
- Patient Task Force, 6th World PH Symposium

a. McLaughlin VV, et al. *J Am Coll Cardiol*. 2015;65:1976-1997; b. McGoon, et al. *Eur Respir J*. 2019;53. pii:1801919.

Ευχαριστώ για την προσοχή σας