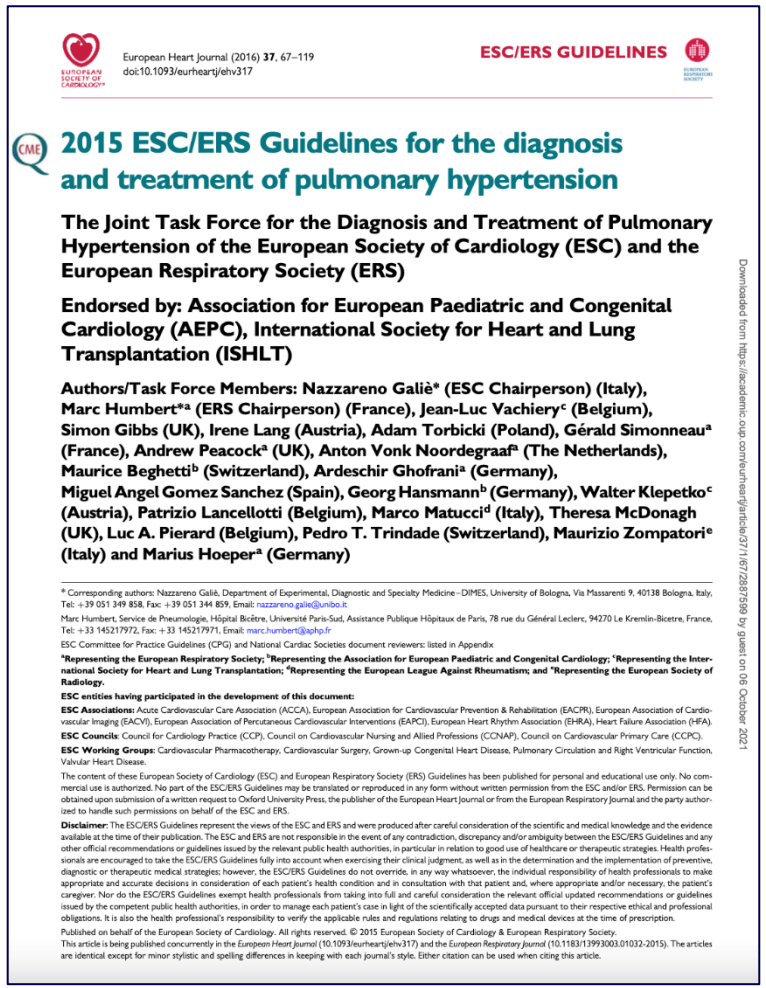
DOPORUČENÉ POSTUPY ESC/ERS 2022 PRO DIAGNOSTIKU A LÉČBU PLICNÍ HYPERTENZE

PAVEL JANSA (VFN Praha)

MARTIN HUTYRA (FN Olomouc)

RELEVANTNÍ DOKUMENTY



European Heart Journal (2016) **37**, 67–119
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

Authors/Task Force Members: Nazzareno Galiè* (ESC Chairperson) (Italy), Marc Humbert*[a] (ERS Chairperson) (France), Jean-Luc Vachiery[c] (Belgium), Simon Gibbs (UK), Irene Lang (Austria), Adam Torbicki (Poland), Gérald Simonneau[a] (France), Andrew Peacock[a] (UK), Anton Vonk Noordegraaf[a] (The Netherlands), Maurice Beghetti[b] (Switzerland), Ardeschir Ghofrani[a] (Germany), Miguel Angel Gomez Sanchez (Spain), Georg Hansmann[b] (Germany), Walter Klepetko[c] (Austria), Patrizio Lancellotti (Belgium), Marco Matucci[d] (Italy), Theresa McDonagh (UK), Luc A. Pierard (Belgium), Pedro T. Trindade (Switzerland), Maurizio Zompatori[e] (Italy) and Marius Hoeper[a] (Germany)

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Marc Humbert, Service de Pneumologie, Hôpital Bicêtre, Université Paris-Sud, Assistance Publique Hôpitaux de Paris, 78 rue du Général Leclerc, 94270 Le Kremlin-Bicetre, France, Tel: +33 145217972, Fax: +33 145217971, Email: marc.humbert@aphp.fr

ESC Committee for Practice Guidelines (CPG) and National Cardiac Societies document reviewers: listed in Appendix

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ESC entities having participated in the development of this document:

ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

ESC Councils: Council for Cardiology Practice (CCP), Council on Cardiovascular Nursing and Allied Professions (CCNAP), Council on Cardiovascular Primary Care (CCPC).

ESC Working Groups: Cardiovascular Pharmacotherapy, Cardiovascular Surgery, Grown-up Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Valvular Heart Disease.

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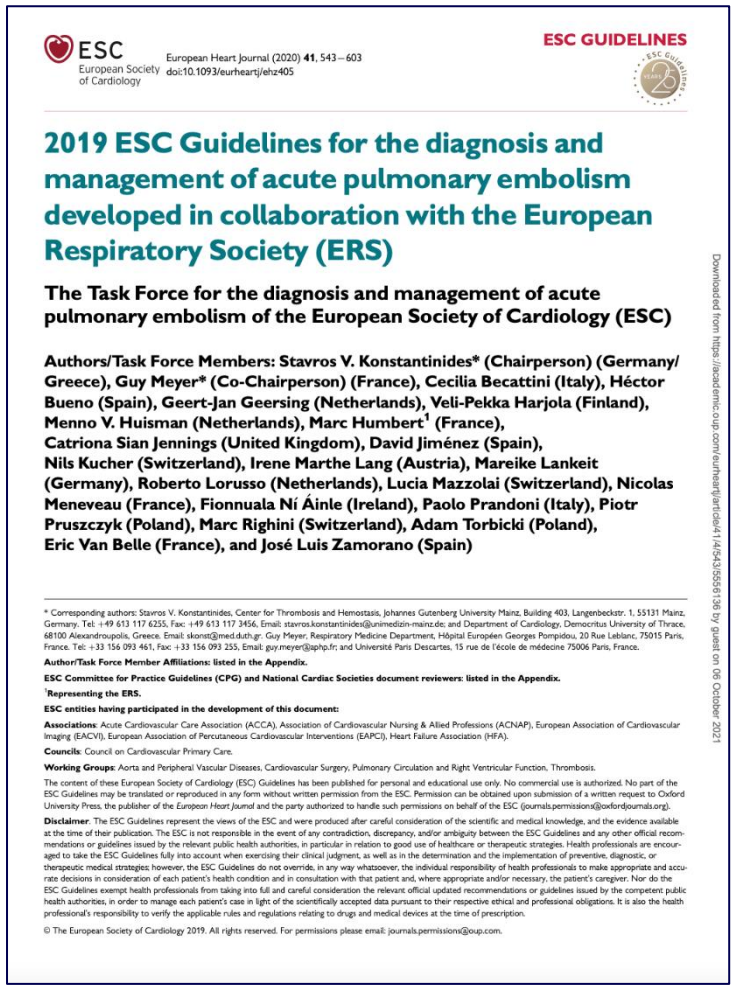
This article is being published concurrently in the *European Heart Journal* (10.1093/eurheartj/ehv317) and the *European Respiratory Journal* (10.1183/13993003.01032-2015). The articles are identical except for minor stylistic and spelling differences in keeping with each journal's style. Either citation can be used when citing this article.



ESC European Society of Cardiology
European Heart Journal (2020) **41**, 543–603
doi:10.1093/eurheartj/ehz405

ESC GUIDELINES

2019 ESC Guidelines for the diagnosis and management of acute pulmonary embolism developed in collaboration with the European Respiratory Society (ERS)

The Task Force for the diagnosis and management of acute pulmonary embolism of the European Society of Cardiology (ESC)

Authors/Task Force Members: Stavros V. Konstantinides* (Chairperson) (Germany/Greece), Guy Meyer* (Co-Chairperson) (France), Cecilia Becattini (Italy), Héctor Bueno (Spain), Geert-Jan Geersing (Netherlands), Veli-Pekka Harjola (Finland), Menno V. Huisman (Netherlands), Marc Humbert[†] (France), Catriona Sian Jennings (United Kingdom), David Jiménez (Spain), Nils Kucher (Switzerland), Irene Marthe Lang (Austria), Mareike Lankeit (Germany), Roberto Lorusso (Netherlands), Lucia Mazzolai (Switzerland), Nicolas Meneveau (France), Fionnuala Ní Áinle (Ireland), Paolo Prandoni (Italy), Piotr Pruszczyk (Poland), Marc Righini (Switzerland), Adam Torbicki (Poland), Eric Van Belle (France), and José Luis Zamorano (Spain)

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[†]**Representing the ERS.**

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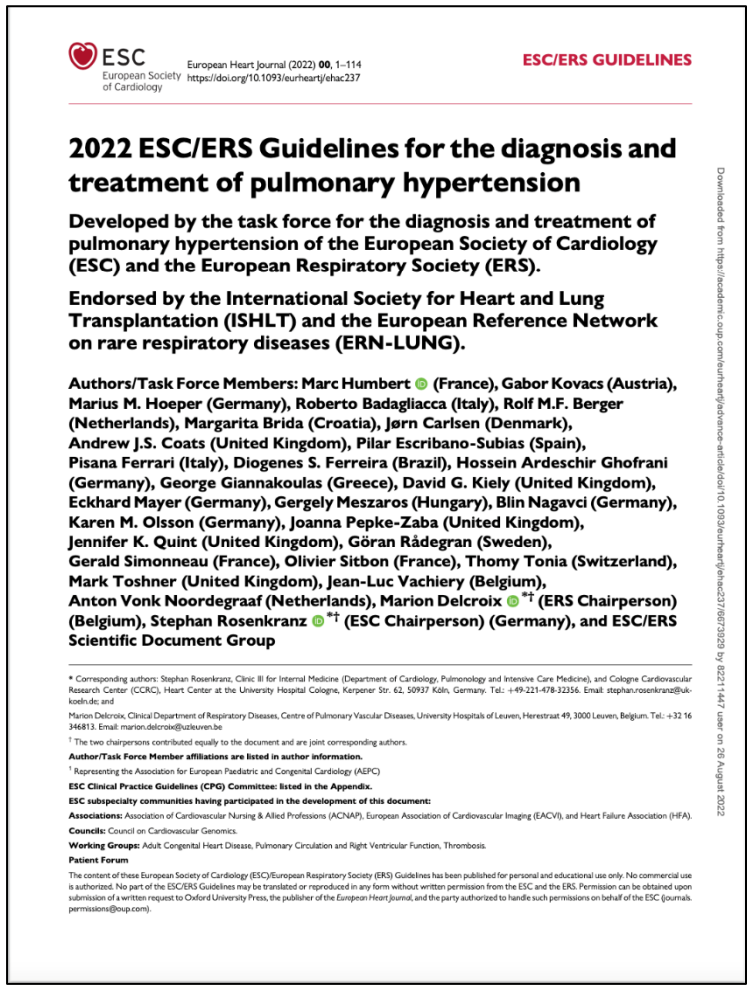

ESC European Society of Cardiology
European Heart Journal (2022) **00**, 1–114
https://doi.org/10.1093/eurheartj/ehac237

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Authors/Task Force Members: Marc Humbert (France), Gabor Kovacs (Austria), Marius M. Hoeper (Germany), Roberto Badagliacca (Italy), Rolf M.F. Berger (Netherlands), Margarita Brida (Croatia), Jørn Carlsen (Denmark), Andrew J.S. Coats (United Kingdom), Pilar Escribano-Subias (Spain), Pisana Ferrari (Italy), Diogenes S. Ferreira (Brazil), Hossein Ardeschir Ghofrani (Germany), George Giannakoulas (Greece), David G. Kiely (United Kingdom), Eckhard Mayer (Germany), Gergely Meszaros (Hungary), Blin Nagavci (Germany), Karen M. Olsson (Germany), Joanna Pepke-Zaba (United Kingdom), Jennifer K. Quint (United Kingdom), Göran Rådegran (Sweden), Gerald Simonneau (France), Olivier Sitbon (France), Thomy Tonia (Switzerland), Mark Toshner (United Kingdom), Jean-Luc Vachiery (Belgium), Anton Vonk Noordegraaf (Netherlands), Marion Delcroix *[†] (ERS Chairperson) (Belgium), Stephan Rosenkranz *[†] (ESC Chairperson) (Germany), and ESC/ERS Scientific Document Group

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[‡] Representing the Association for European Paediatric and Congenital Cardiology (AEPC)

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Associations: Association of Cardiovascular Nursing & Allied Professions (ACNAP), European Association of Cardiovascular Imaging (EACVI), and Heart Failure Association (HFA).

Councils: Council on Cardiovascular Genomics.

Working Groups: Adult Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Thrombosis.

Patient Forum

www.escardio.org www.kardio-cz.cz

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- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
EHJ 2022, *ERJ* 2022

2015

Definition	Characteristics[a]
PH	PAPm ≥25 mmHg
Pre-capillary PH	PAPm ≥25 mmHg PAWP ≤15 mmHg
Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU[c]
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU[c]

2022

Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
Combined post- and pre-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise >3 mmHg/L/min

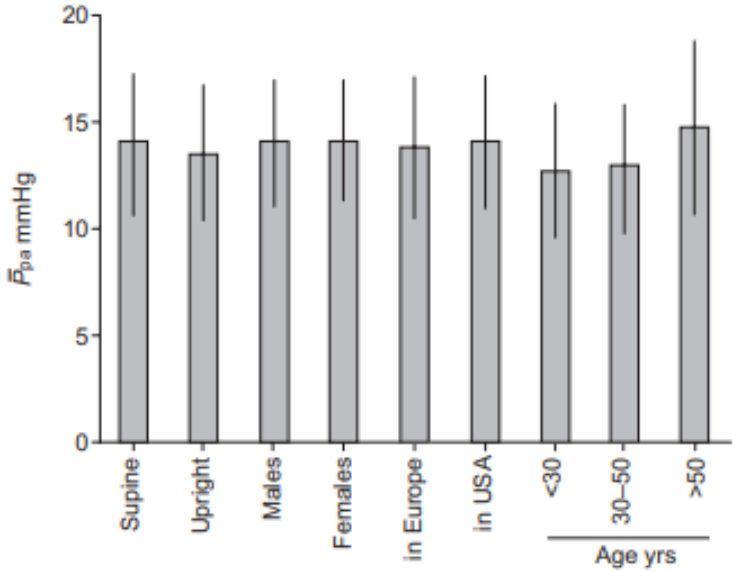
Cor et Vasa 58 (2016)
EHJ 2022, *ERJ* 2022

Pulmonary arterial pressure during rest and exercise in healthy subjects: a systematic review

G. Kovacs*, A. Berghold#, S. Scheidl* and H. Olschewski*

n=1.187 , 47 studií, 13 zemí, hemodynamické vyšetření v klidu

$\bar{P}_{pa}$ mmHg	14.0±3.3
Systolic P_{pa} mmHg	20.8±4.4
Diastolic P_{pa} mmHg	8.8±3.0
P_{paw} mmHg	8.0±2.9
Heart rate min^{-1}	76±14
Cardiac output $L \cdot min^{-1}$	7.3±2.3
Cardiac index $L \cdot min^{-1} \cdot m^{-2}$	4.1±1.3
PVR $dyn \cdot s \cdot cm^{-5}$	74±30



Eur Respir J 2009; 34: 888–894

Chronic Cor Pulmonale

Report of an Expert Committee*

Contents

Foreword

IN THE early months of 1960, the Director-General of the World Health Organization appointed an Expert Committee to inquire into and write a report on the subject of cor pulmonale.

In order to facilitate the preliminary study, as well as the actual deliberations of the Committee, two consultants were appointed, to prepare a survey of the subject: Professor H. Denolin, Chargé de cours á l'Université de Bruxelles; and Dr. C. M. Fletcher, Senior Lecturer in Medicine, Postgraduate Medical School, London. This, a sixty-page report, was put together by Drs. Denolin and Fletcher during July, 1960, and made available to Committee members shortly thereafter.

The membership of the Expert Committee was as follows: Dr. J. Dankmeijer, Professor of Anatomy, Embryology and Physical Anthropology, University of Leiden, the Netherlands; Dr. F. Herles, Professor of Medicine, II Internal Clinic, Charles University, Prague, Czechoslovakia; Dr. M. Ibrahim, formerly Professor of Cardiology, Faculty of Medicine, Cairo University, Cairo, Province of Egypt, United Arab Republic; Dr. D. D. Reid, Professor of Epidemiology, Department of Medical Statistics and Epidemiology, London School of Hygiene and Tropical Medicine, London, England; Dr. D. W. Richards, Lambert Professor of Medicine, College of Physicians and Surgeons, Columbia University, New

*Reprinted by permission from the World Health Organization Technical Report Series No. 213. Reprints of the original report may be obtained for $0.30 from World Health Organization, Palais Des Nations, Geneva.

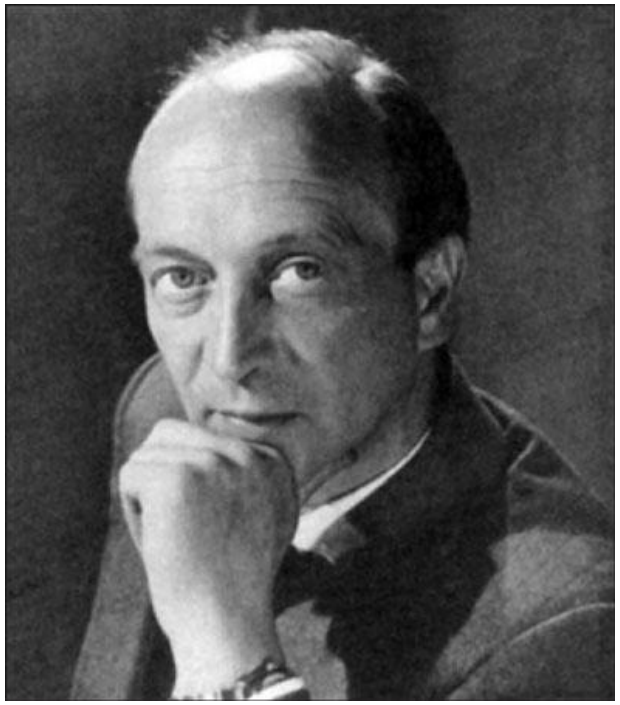
The following are regarded as the upper limits of normal values with the reference point* 10 cm. above the level of the back in the supine position:

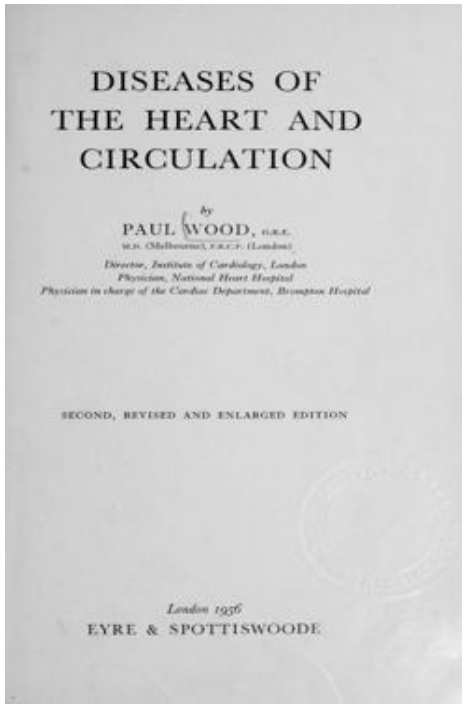
	Systolic mm. Hg	Diastolic mm. Hg	Mean mm. Hg
Right atrium	..	..	6
Right ventricle	25	6	..
Pulmonary artery	25	12	15
Pulmonary arteriolar wedge pressure	..	..	9

The total pulmonary resistance lies between 150 and 300 dynes sec. cm.$^{-5}$

Circulation, Volume XXVII, April 1963

DEFINICE PLICNÍ HYPERTENZE




DISEASES OF
THE HEART AND
CIRCULATION

by

PAUL WOOD, O.B.E.

M.D. (Melbourne), F.R.C.P. (London)

Director, Institute of Cardiology, London
Physician, National Heart Hospital
Physician in charge of the Cardiac Department, Brompton Hospital

SECOND, REVISED AND ENLARGED EDITION

London 1956
EYRE & SPOTTISWOODE

PULMONARY HYPERTENSION WITH SPECIAL REFERENCE TO THE VASOCONSTRICTIVE FACTOR*

BY

PAUL WOOD

From the Institute of Cardiology, National Heart Hospital, and the Brompton Hospital

Received July 2, 1958

The normal pulmonary blood pressure in a series of 60 normal controls studied at the Institute of Cardiology and at the Brompton hospital was 16/7 mm. Hg with reference to the sternal angle, the mean being 11 mm. and the range 8/2–28/14 mm. The mean cardiac output was 8 litres a minute, and the common range 5·5 to 10·5 litres a minute. The left atrial pressure averaged 2 to 3 mm. Hg.

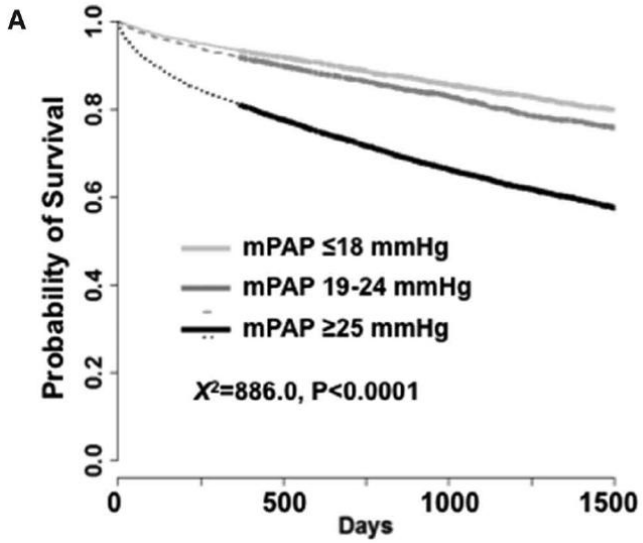
The pulmonary vascular resistance, which in simple units is the pulmonary artery pressure minus the left atrial pressure in mm. Hg divided by the pulmonary blood flow in litres a minute, was therefore about 1 unit or 80 dynes sec./cm.$^{-5}$.

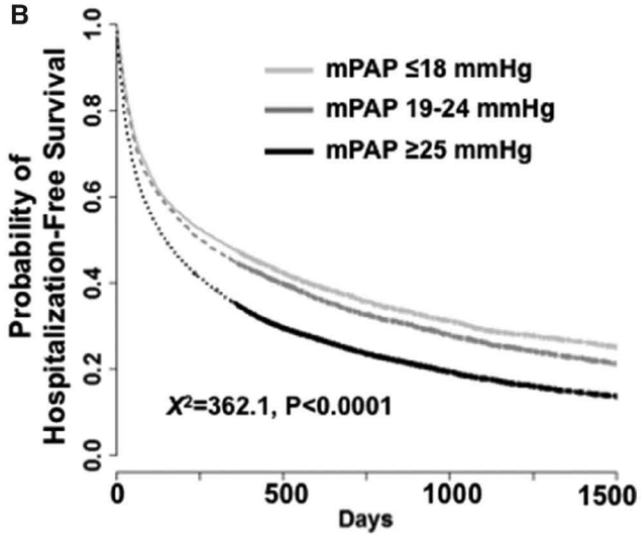
Definition of Pulmonary Hypertension. Pulmonary hypertension literally implies a pulmonary blood pressure above 30/15 mm. which is the upper limit of the normal range. In practice serious pulmonary hypertension usually means a pressure at or around systemic level, but rarely in excess of 150 mm.

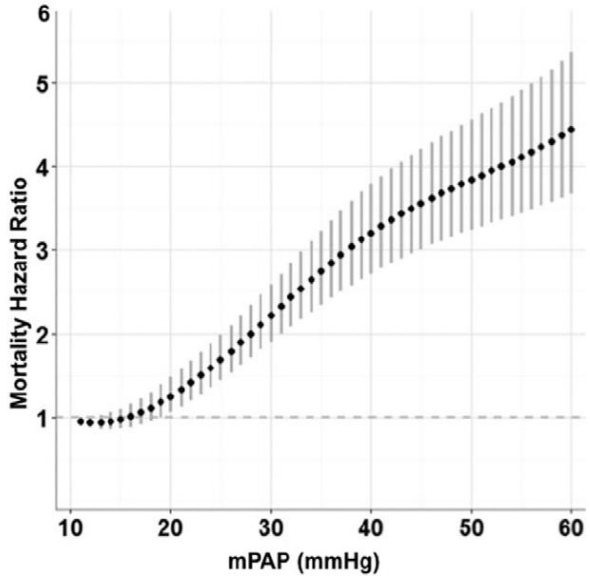
Paul Wood: *Br Heart J* 1958, 20, 557-570
Cor et Vasa 58 (2016)

Association of Borderline Pulmonary Hypertension With Mortality and Hospitalization in a Large Patient Cohort: Insights From the Veterans Affairs Clinical Assessment, Reporting, and Tracking Program

N=21.727, US veterans, right heart catheterization 2007–2012, median follow-up 908 days
Association between mPAP and outcomes of all-cause mortality and hospitalization
3 groups
(1) referent (≤18mm Hg; n=4 207)
(2) borderline PH (19–24 mm Hg; n=5 030)
(3) PH (≥25 mm Hg; n=12 490)

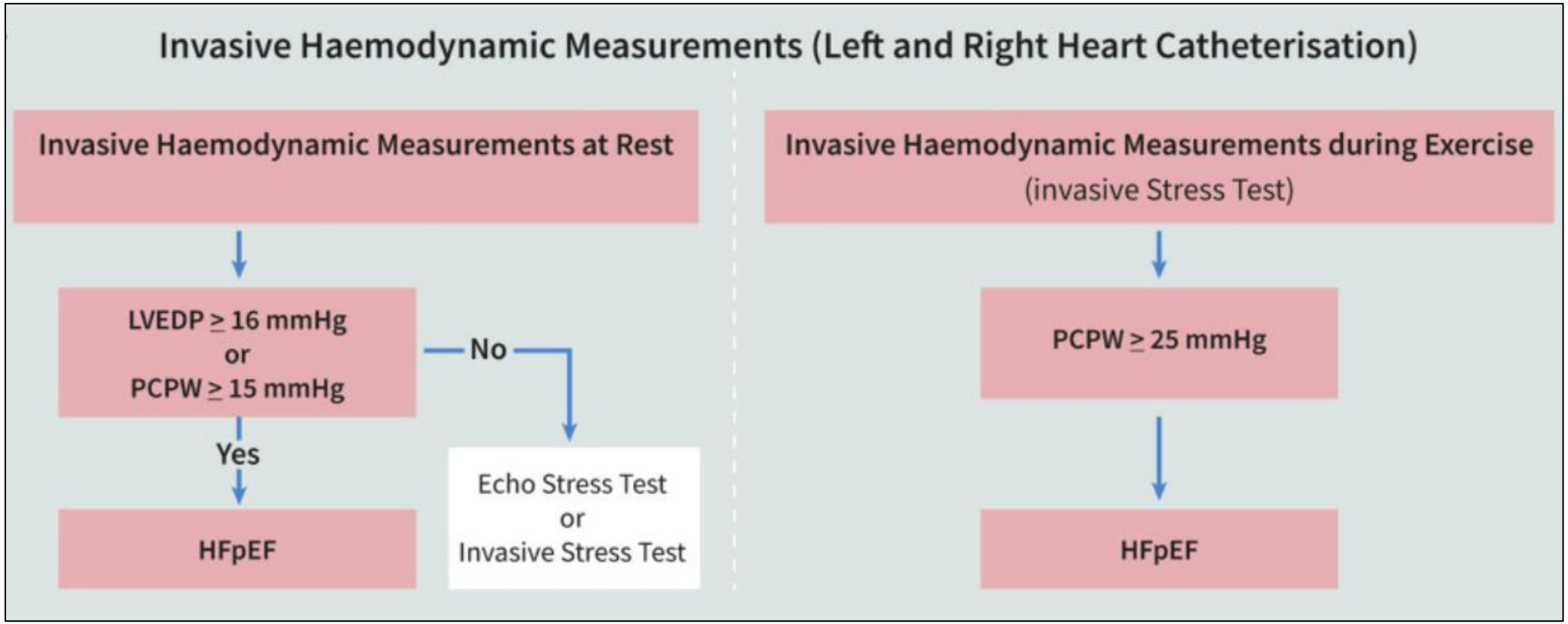






Maron BA et al. *Circulation*. 2016;133:1240-1248

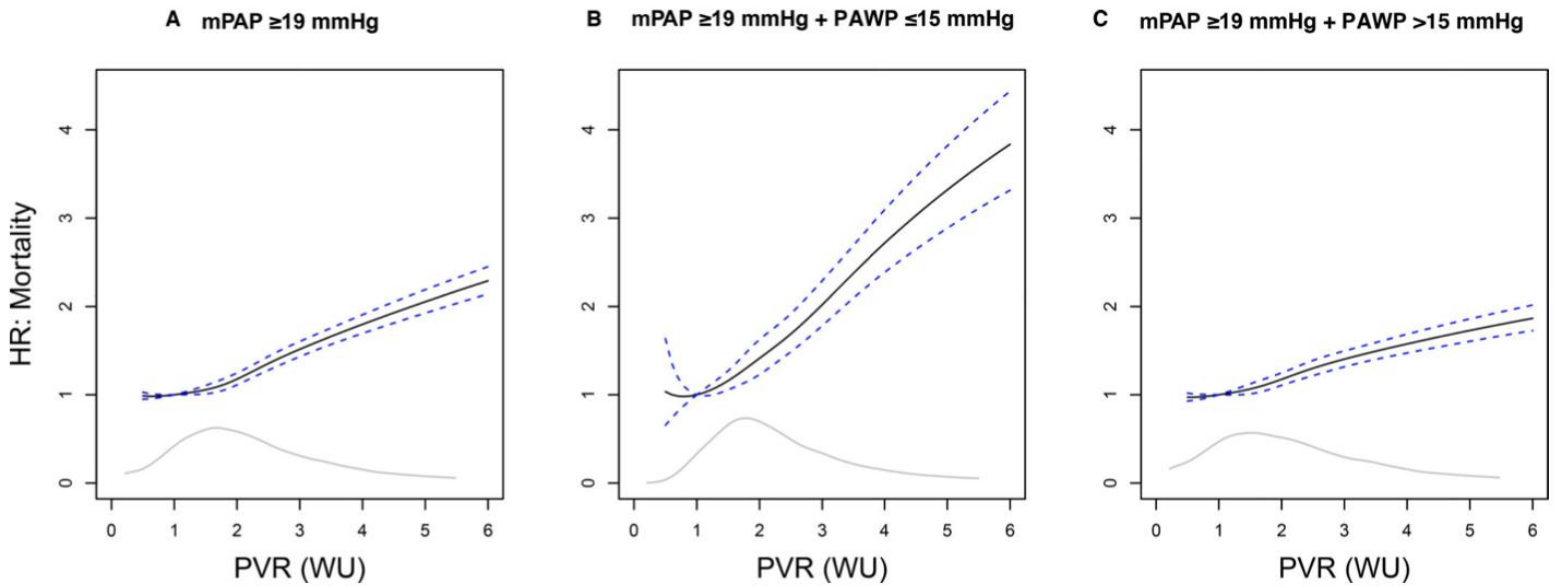
How to diagnose heart failure with preserved ejection fraction: the HFA–PEFF diagnostic algorithm: a consensus recommendation from the Heart Failure Association (HFA) of the European Society of Cardiology (ESC)

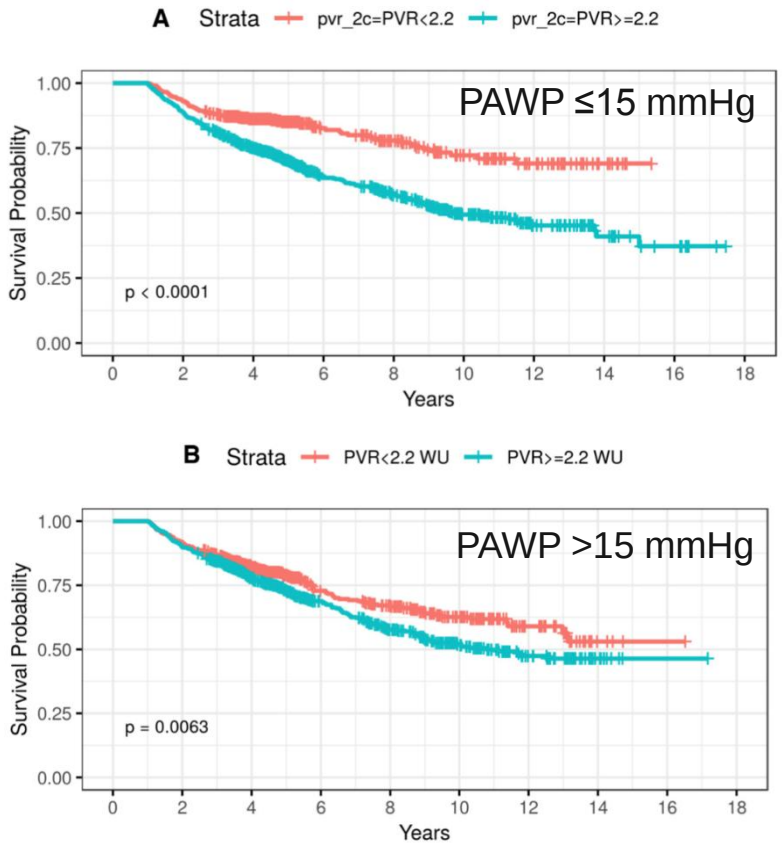


Pieske B et al. *European Heart Journal* (2019) 40, 3297–3317

The Association Between Pulmonary Vascular Resistance and Clinical Outcomes in Patients with Pulmonary Hypertension: A Retrospective Cohort Study

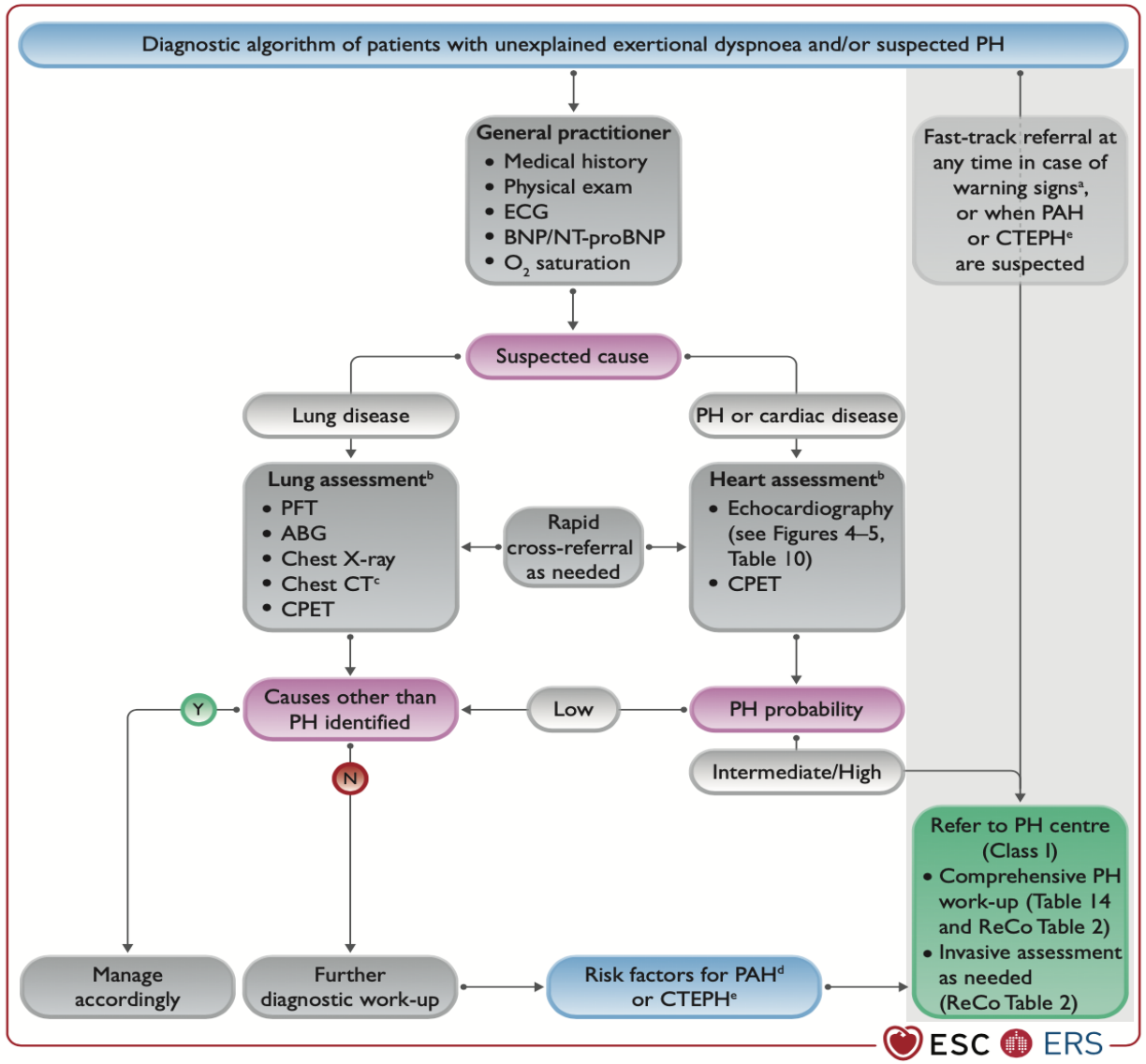
N=40.082; male 96.7 %, age 66.5 [61.1–73.5] yr
history of heart failure (N=23,201 [57.9%]) and chronic obstructive pulmonary disease (N=13,348 [33.3%])





Maron BA et al. *Lancet Respir Med.* 2020 September ; 8(9): 873–884

DIAGNOSTICKÝ ALGORITMUS PH (2022)



Suspekce

Detekce

Konfirmace

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

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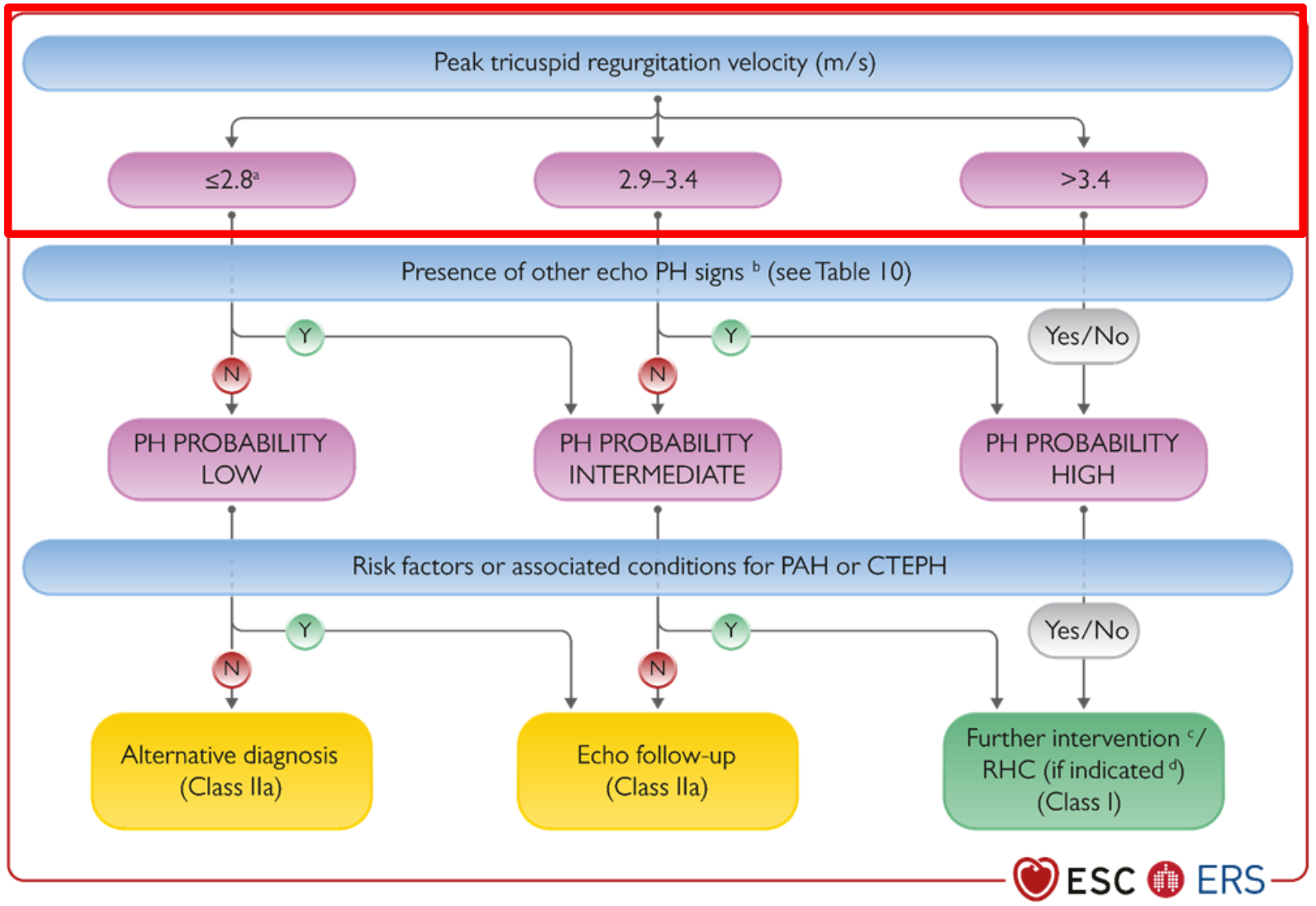

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022

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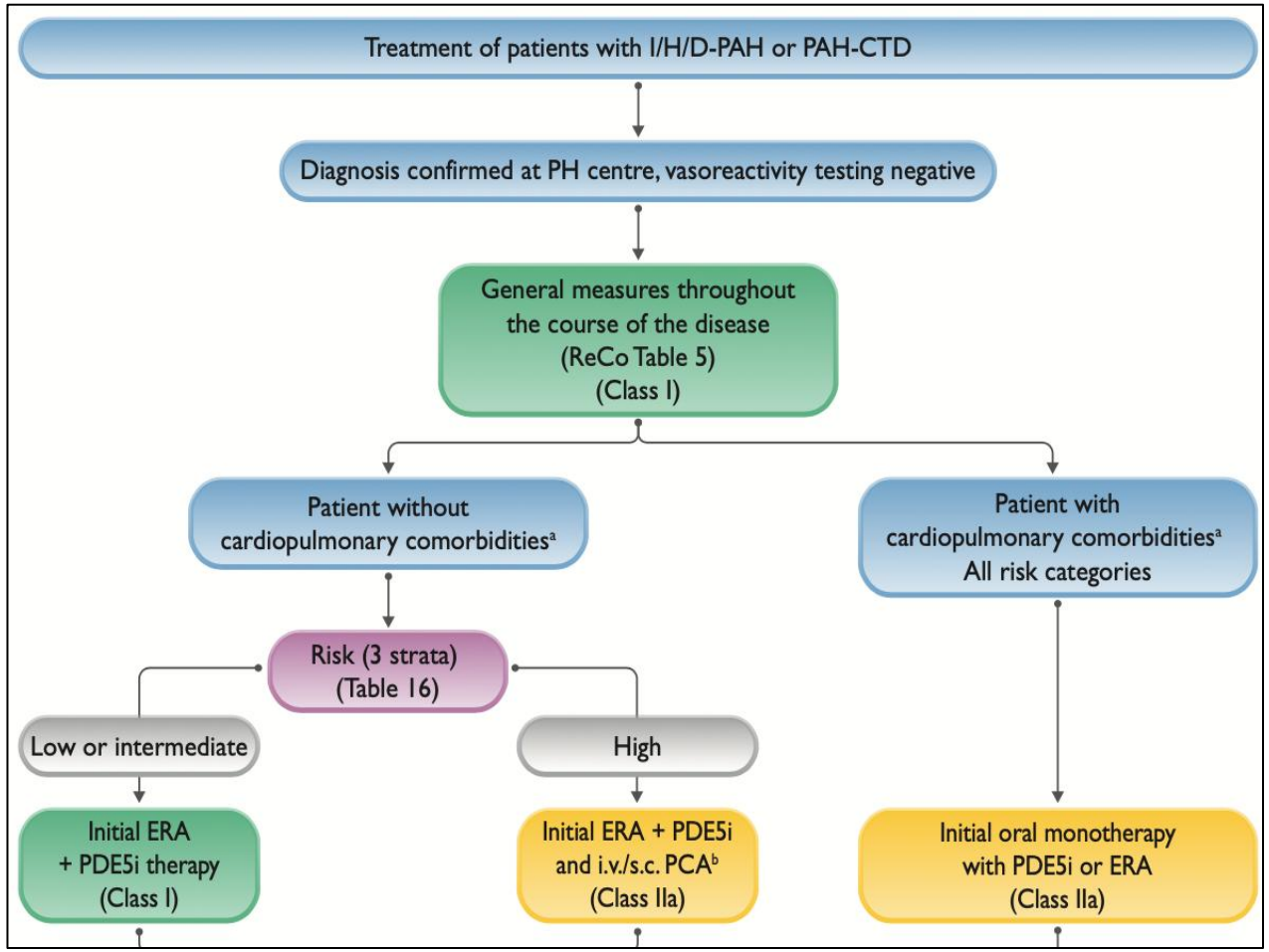

Determinants of prognosis (estimated 1-year mortality)	Low risk (<5%)	Intermediate risk (5–20%)	High risk (>20%)
Clinical observations and modifiable variables			
Signs of right HF	Absent	Absent	Present
Progression of symptoms and clinical manifestations	No	Slow	Rapid
Syncope	No	Occasional syncope[a]	Repeated syncope[b]
WHO-FC	I, II	III	IV
6MWD[c]	>440 m	165–440 m	<165 m
CPET	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[d]	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^2 TAPSE/sPAP >0.32 mm/mmHg No pericardial effusion	RA area 18–26 cm^2 TAPSE/sPAP 0.19–0.32 mm/mmHg Minimal pericardial effusion	RA area >26 cm^2 TAPSE/sPAP <0.19 mm/mmHg Moderate or large pericardial effusion
cMRI[e]	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 mmHg CI ≥2.5 L/min/m^2 SVI >38 mL/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 L/min/m^2 SVI 31–38 mL/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 L/min/m^2 SVI <31 mL/m^2 SvO_2 <60%

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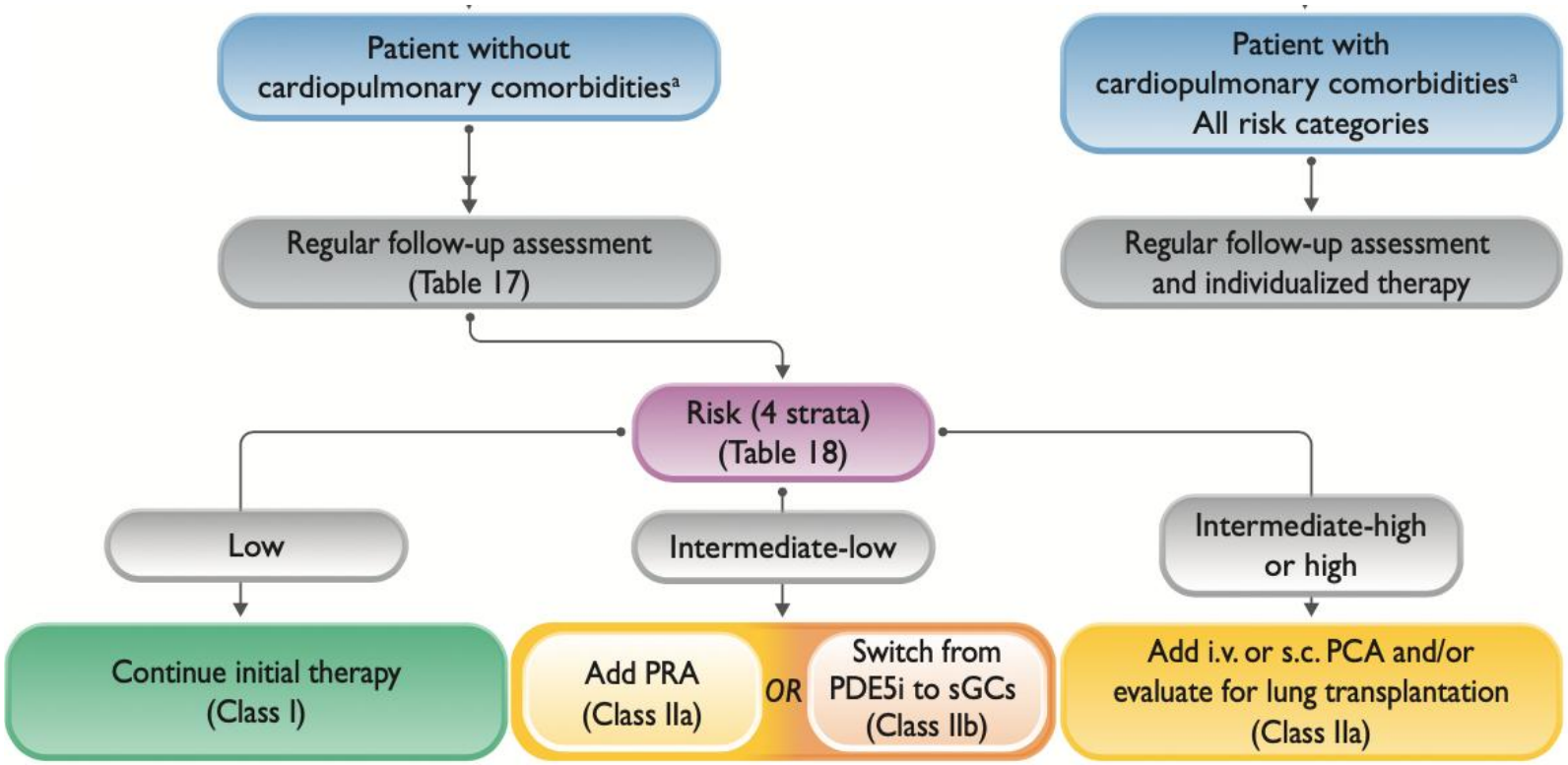
TERAPEUTICKÝ ALGORITMUS PH (2022)



Determinants of prognosis	Low risk	Intermediate–low risk	Intermediate–high risk	High risk
Points assigned	1	2	3	4
WHO-FC	I or II[a]	-	III	IV
6MWD, m	>440	320–440	165–319	<165
BNP or NT-proBNP,[a] ng/L	<50 <300	50–199 300–649	200–800 650–1100	>800 >1100

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

SKRÍNINK A ČASNÁ DETEKCE PAH

Recommendations	Class[a]	Level[b]
Systemic sclerosis		
In patients with SSc, an annual evaluation of the risk of having PAH is recommended[183,186]	I	B
Other		
Counselling regarding the risk of PAH and annual screening are recommended in individuals who test positive for PAH-causing mutations and in first-degree relatives of patients with HPAH[33]	I	B
In patients referred for liver transplantation, echocardiography is recommended as a screening test for PH	I	C
Further tests (echocardiography, BNP/NT-proBNP, PFTs, and/or CPET) should be considered in symptomatic patients with CTD, portal hypertension, or HIV to screen for PAH[172]	IIa	B

SKRÍNINK PLICNÍ ARTERIÁLNÍ HYPERTENZE

- Systémová sklerodermie (každý rok)
- Prvostupňoví příbuzní nemocných s PAH
- HIV v případě přítomnosti symptomů podezřelých z plicní hypertenze
- Jaterní onemocnění před transplantací jater
- Stavy po korekci zkratové vrozené srdeční vady

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KLINICKÁ KLASIFIKACE PLICNÍ HYPERTENZE (2022)

GROUP 1 Pulmonary arterial hypertension (PAH) — 1 %

- 1.1 Idiopathic
  - 1.1.1 Non-responders at vasoreactivity testing
  - 1.1.2 Acute responders at vasoreactivity testing
- 1.2 Heritable[a]
- 1.3 Associated with drugs and toxins[a]
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
- 1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
- 1.6 Persistent PH of the newborn

GROUP 2 PH associated with left heart disease — 70 %

- 2.1 Heart failure:
  - 2.1.1 with preserved ejection fraction
  - 2.1.2 with reduced or mildly reduced ejection fraction[b]
- 2.2 Valvular heart disease
- 2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH

GROUP 3 PH associated with lung diseases and/or hypoxia — 20 %

- 3.1 Obstructive lung disease or emphysema
- 3.2 Restrictive lung disease
- 3.3 Lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoventilation syndromes
- 3.5 Hypoxia without lung disease (e.g. high altitude)
- 3.6 Developmental lung disorders

GROUP 4 PH associated with pulmonary artery obstructions — 4 %

- 4.1 Chronic thrombo-embolic PH
- 4.2 Other pulmonary artery obstructions[c]

GROUP 5 PH with unclear and/or multifactorial mechanisms — 5 %

- 5.1 Haematological disorders[d]
- 5.2 Systemic disorders[e]
- 5.3 Metabolic disorders[f]
- 5.4 Chronic renal failure with or without haemodialysis
- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

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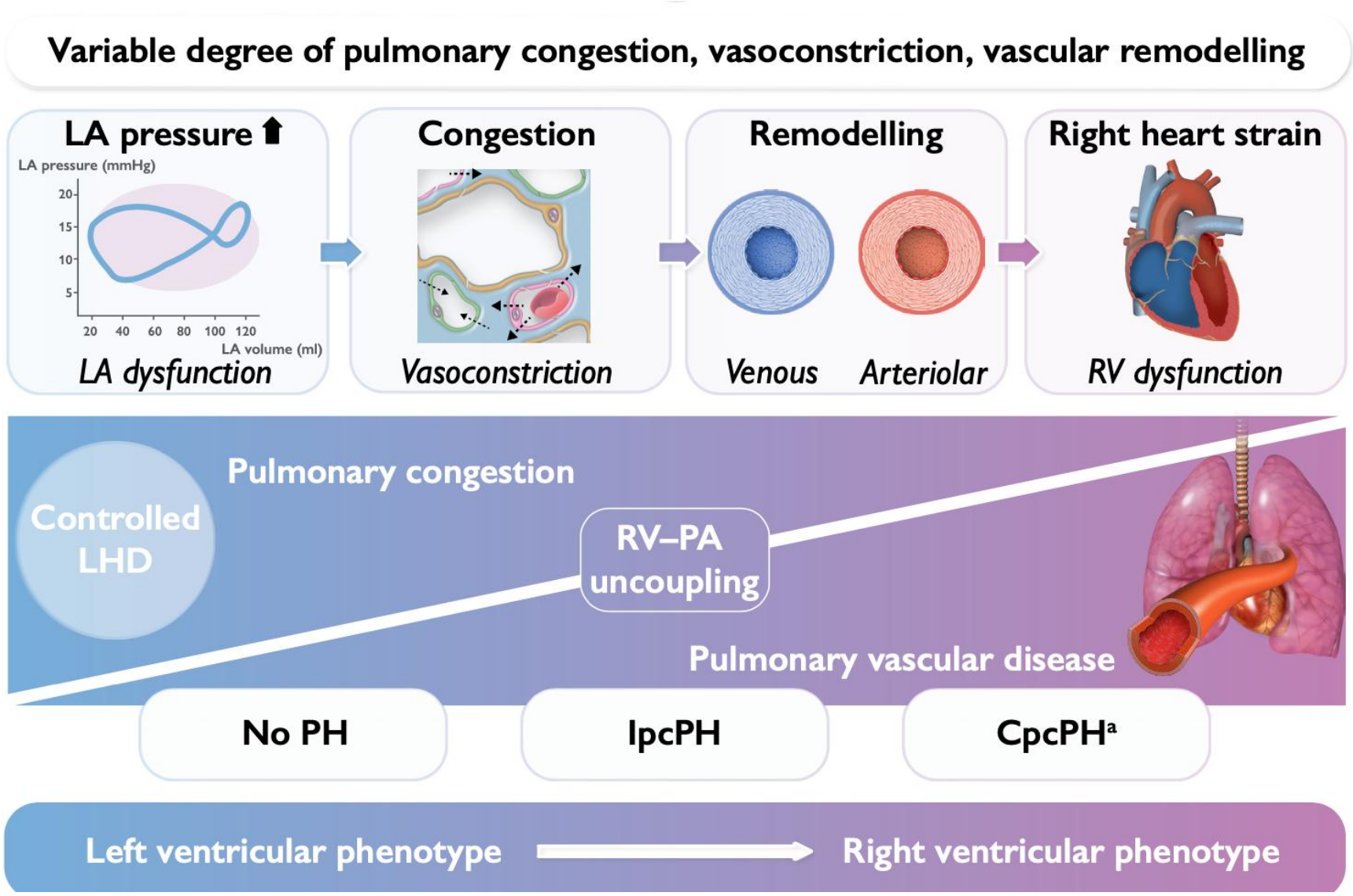
EHJ 2022, *ERJ* 2022

PLICNÍ HYPERTENZE ASOCIOVANÁ S ONEMOCNĚNÍM LEVÉHO SRDCE



PVR > 2 WU
(20-30 % pacientů)

Těžká prekapilární komponenta

PVR > 5 WU

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PLICNÍ HYPERTENZE ASOCIOVANÁ S ONEMOCNĚNÍM LEVÉHO SRDCE

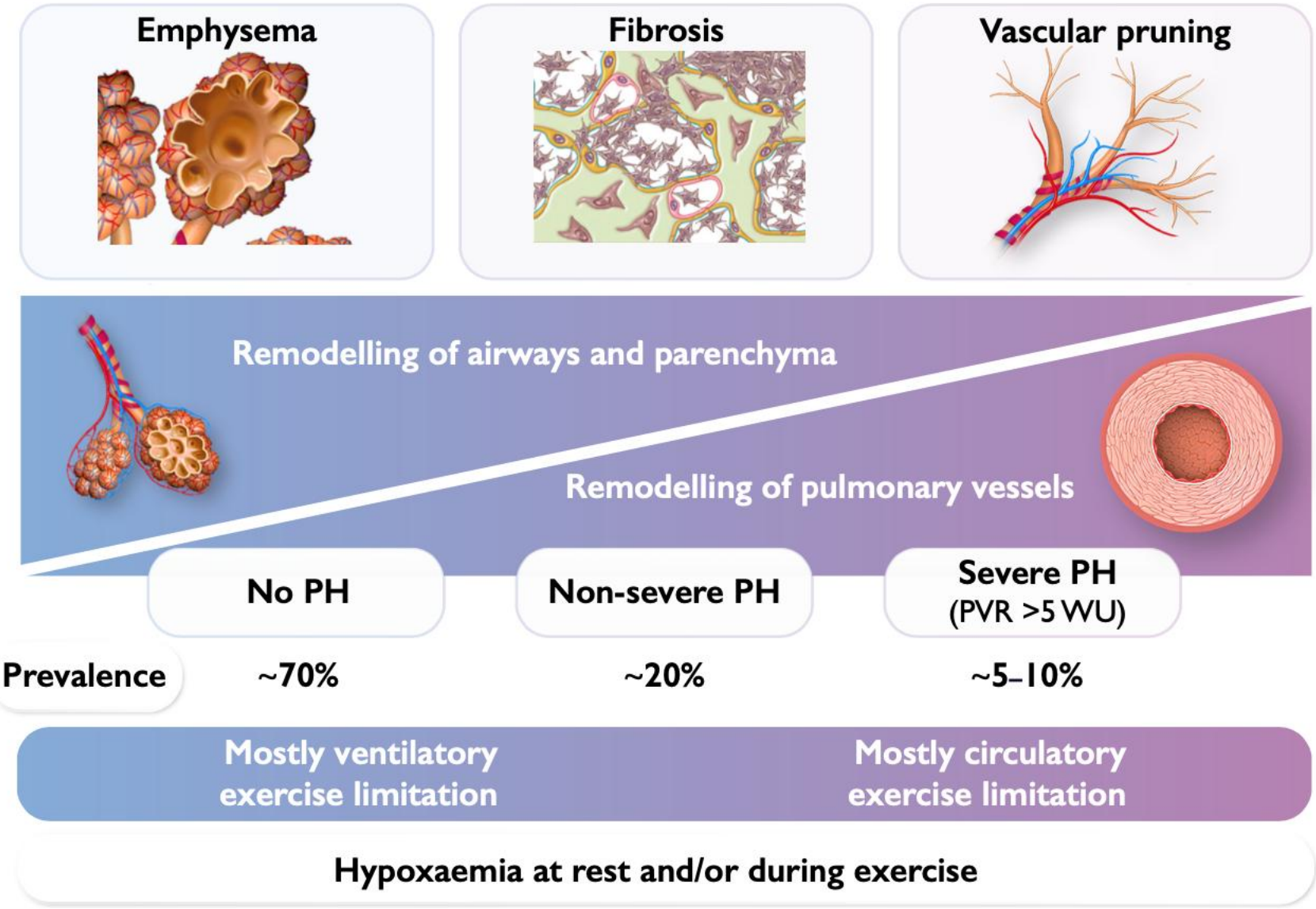
Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
Combined post- and pre-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise >3 mmHg/L/min

Latentní postkapilární komponenta:

PAWP ＞ 18 mmHg po volumové výzvě

2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
(*European Heart Journal*; 2022 – doi: 10.1093/eurheartj/ehac237 and *European Respiratory Journal*; 2022 – doi: 10.1183/13993003.00879-2022)

PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI



Těžká PH
PVR > 5 WU

- Bez vztahu k plicním objemům
- Hypoxémie
- Nízká DLco

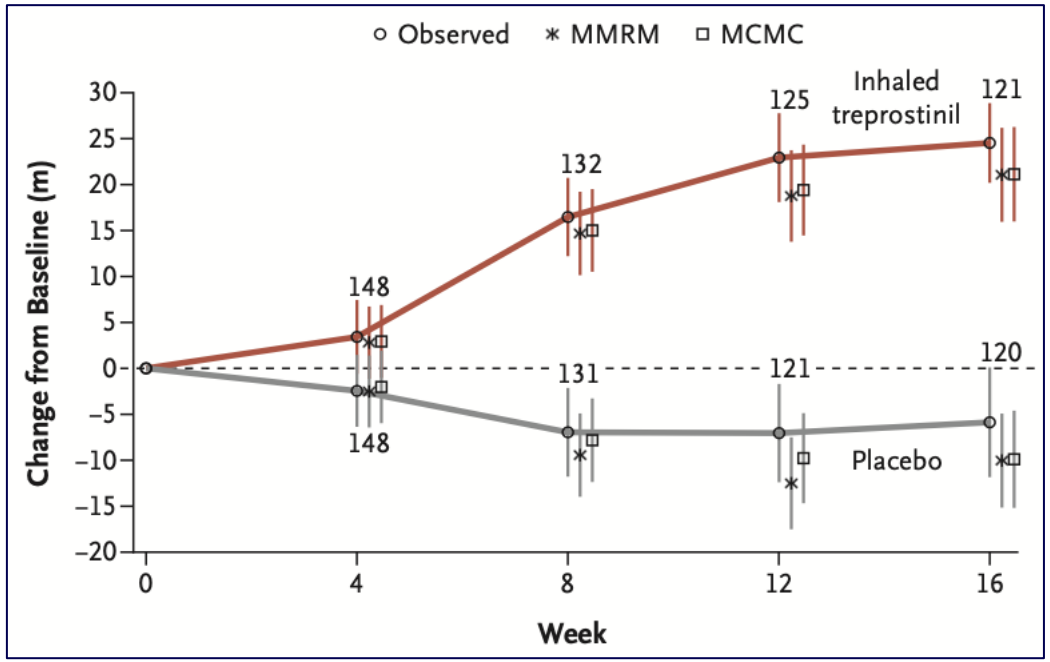
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Inhaled Treprostinil in Pulmonary Hypertension Due to Interstitial Lung Disease

N=326, PH u ILD, inhal. treprostinil 12 vdechů (celkem 72 µg) 4x denně vs placebo
Prekapilární PH, PVR více než 3 WU, FVC méně než 70 %



End Point	Inhaled Treprostinil (N=163)	Placebo (N=163)	Treatment Effect (95% CI)	P Value
Primary end point				
Change in peak 6-minute walk distance from baseline to wk 16 — m†	21.08±5.12	−10.04±5.12	31.12±7.25 (16.85 to 45.39)‡	<0.001
Secondary end points§				
Change in plasma concentration of NT-proBNP from baseline to wk 16¶				
Mean (±SD) change — pg/ml	−396.35±1904.90	1453.95±7296.20		
Median — pg/ml	−22.65	20.65		
Range — pg/ml	−11,433.0 to 5373.1	−5483.3 to 87,148.3		
Ratio to baseline	0.85±0.06	1.46±0.11	0.58±0.06 (0.47 to 0.72)‖	<0.001
Occurrence of clinical worsening — no. (%)			0.61 (0.4 to 0.92)**	0.04
Any event	37 (22.7)	54 (33.1)		
Hospitalization for cardiopulmonary indication	18 (11.0)	24 (14.7)		
Decrease in 6-minute walk distance of >15% from baseline	13 (8.0)	26 (16.0)		
Death from any cause	4 (2.5)	4 (2.5)		
Lung transplantation	2 (1.2)	0		
Least-squares mean change in peak 6-minute walk distance from baseline to wk 12 — m†	18.77±4.99	−12.52±5.01	31.29±7.07 (17.37 to 45.21)‡	<0.001
Least-squares mean change in trough 6-minute walk distance from baseline to wk 15 — m	9.3±5.5	−12.7±5.5	21.99±7.7 (6.85 to 37.14)‡	0.005††

Waxman A et al. *N Engl J Med* 2021;384:325-34.

Supplementary Appendix

This appendix has been provided by the authors to give readers additional information about their work.

Supplement to: Waxman A, Restrepo-Jaramillo R, Thenappan T, et al. Inhaled treprostinil in pulmonary hypertension due to interstitial lung disease. N Engl J Med 2021;384:325-34. DOI: 10.1056/NEJMoa2008470

	Inhaled Treprostinil (N=163)	Placebo (N=163)	All Patients (N=326)
6-minute walk distance, meters; mean (range)	254.1 (100-538)	265.1 (30-505)	259.6 (30-538)
Median	256.0	260.0	259.0
Pulmonary vascular resistance, Woods units; mean (range)	6.369 (3.11-18.05)	6.013 (3.06-17.62)	6.191 (3.06-18.05)
Median	5.570	5.060	5.275
NT-proBNP, pg/mL; mean (range)	1857.53 (10.2-21942.0)	1808.86 (23.0-16297.0)	1832.88 (10.2-21942.0)
Median*	550.50	420.80	503.85
Pulmonary arterial pressure, mmHg; mean (range)	37.2 (25-74)	36.0 (25-61)	36.6 (25-74)
Median	35.0	35.0	35.0
Pulmonary capillary wedge pressure, mmHg; mean (range)	10.1 (2-20)	9.6 (0-15)	9.8 (0-20)
Median	10.0	10.0	10.0
Pulmonary function tests			
FEV_1 % Predicted; mean (range)	63.9 (23, 120)	65.0 (22, 145)	
Median	63.0	63.0	
FVC % Predicted; mean (range)	62.5 (24, 130)	63.8 (20, 134)	
Median	60.0	61.0	
TLC % Predicted; mean (range)	62.9 (25, 126)	64.2 (30, 109)	
Median	62.0	62.5	
DLCO % Predicted; mean (range)	30.0 (5, 86)	28.1 (1, 86)	
Median	29.0	26.0	

Waxman A et al. *N Engl J Med* 2021;384:325-34.

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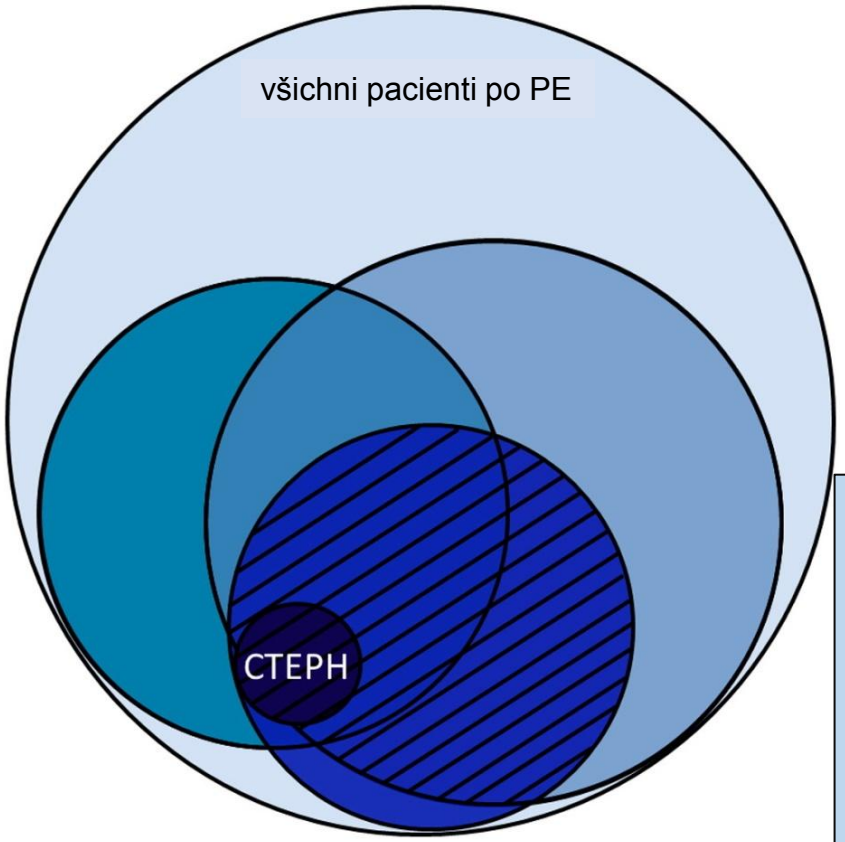
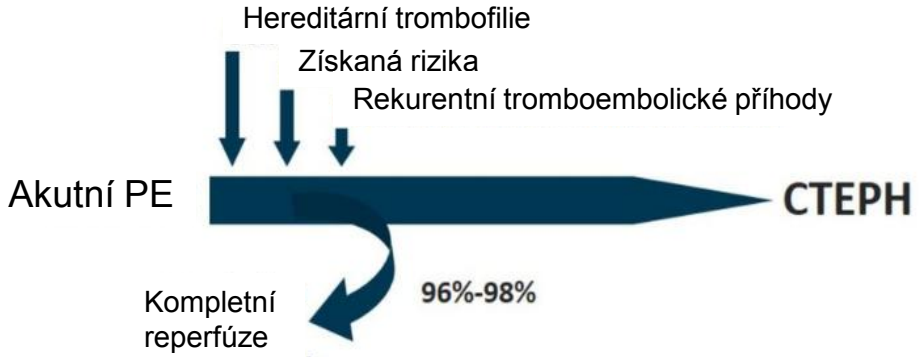
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
EHJ 2022, *ERJ* 2022

CHRONICKÉ KOMPLIKACE PO AKUTNÍ PLICNÍ EMBOLII

- všichni pacienti po PE
- symptomatičtí
- s perzistujícími tromby
- ↓ zátěžová kapacita
- CTEPH
- postembolický syndrom





CTEPD s plicní hypertenzí (=CTEPH)

Definice: prekapilární plicní hypertenze+symptomy

Příčina: trombotická okluze + remodelace

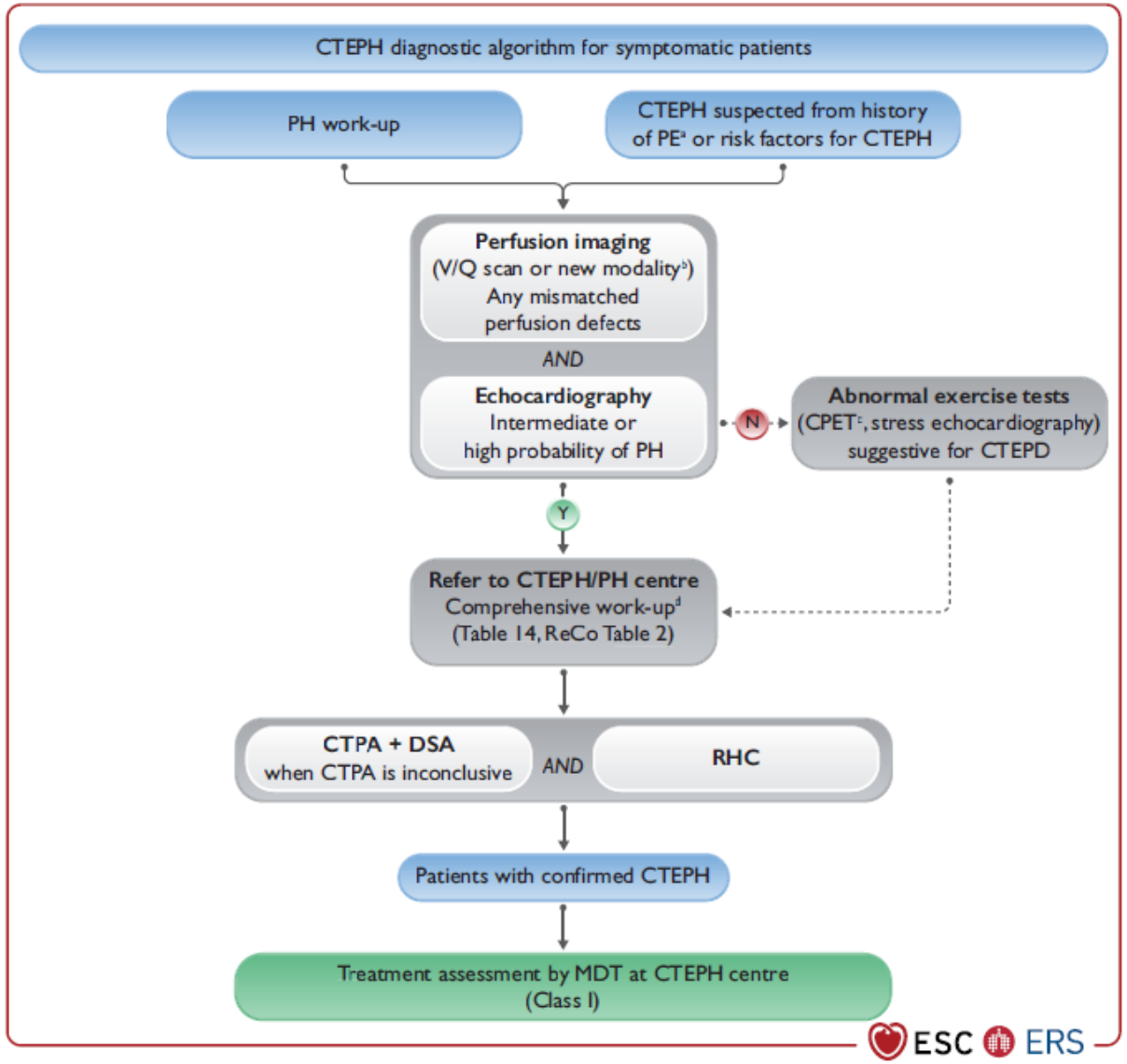
Konsekvence: pravostranné srdeční selhání a smrt

CTEPD bez plicní hypertenze

Trombot. okluze+remodelace+symptomy (bez PH)

Klok FA et al. *Blood Rev* 2014 Nov;28(6):221-6, Delcroix M et al. *Eur Respir J* 2021 Jun 17;57(6):2002828

DIAGNOSTICKÝ ALGORITMUS CTEPH (2022)



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EUROPEAN RESPIRATORY *journal*
FLAGSHIP SCIENTIFIC JOURNAL OF ERS

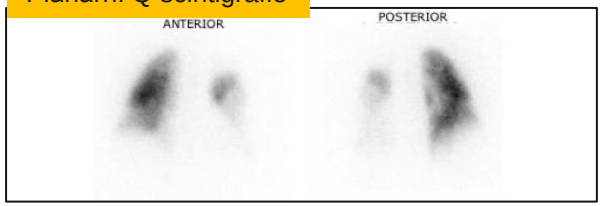
DIAGNOSTIKA

V/Q planární scinti je nadále klíčovou detekční zobrazovací metodou, **V/Q SPECT** je superiorní, plan.simulace

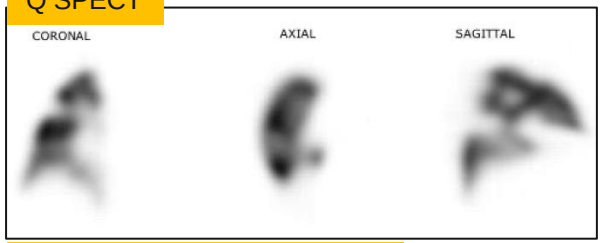
DECT a **MR perfúze** – nenahrazuje scintigrafii (limitovaná dostupnost, zkušenost, validace)

CTA může nahradit DSA u proximálních nálezů, pro zobrazení periferie není dostatečná

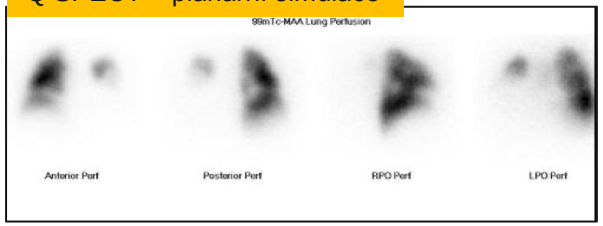
Planární Q scintigrafie

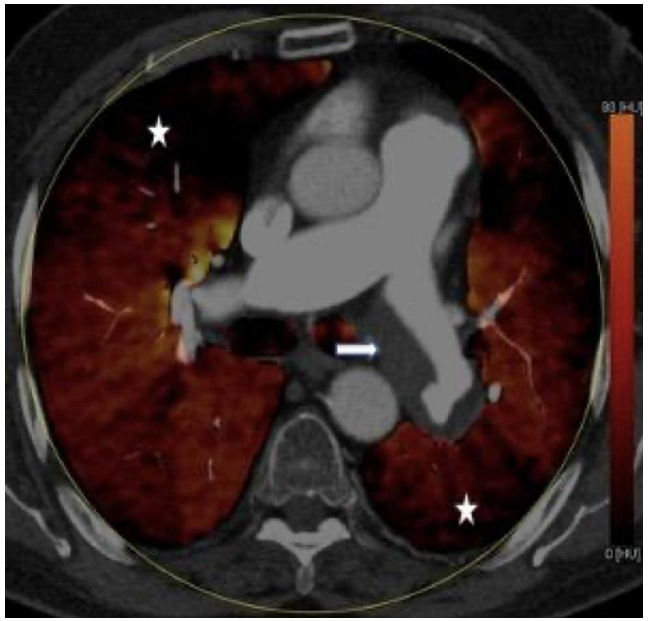


Q SPECT

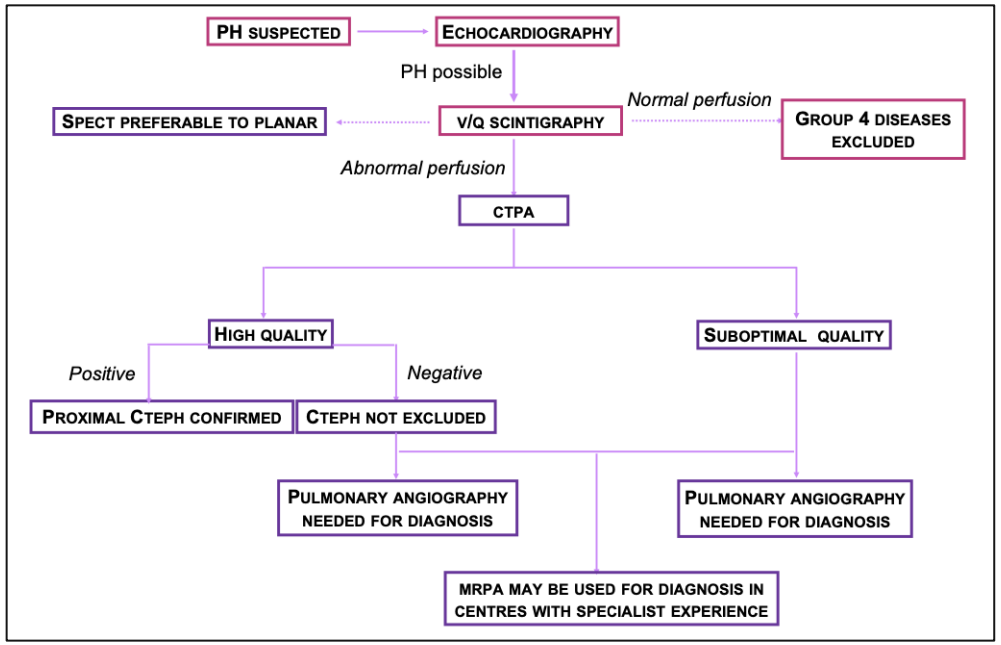


Q SPECT – planární simulace





DECT

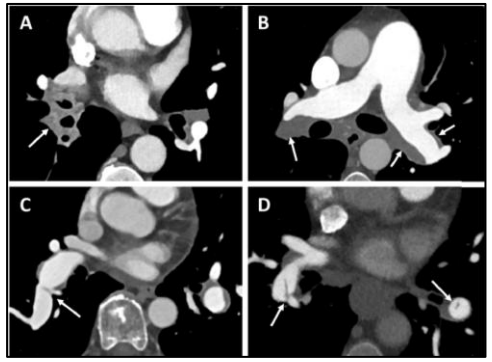


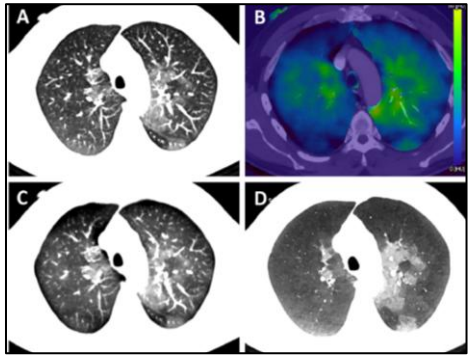
Eur Respir J 2021 Jun 17;57(6):2002828

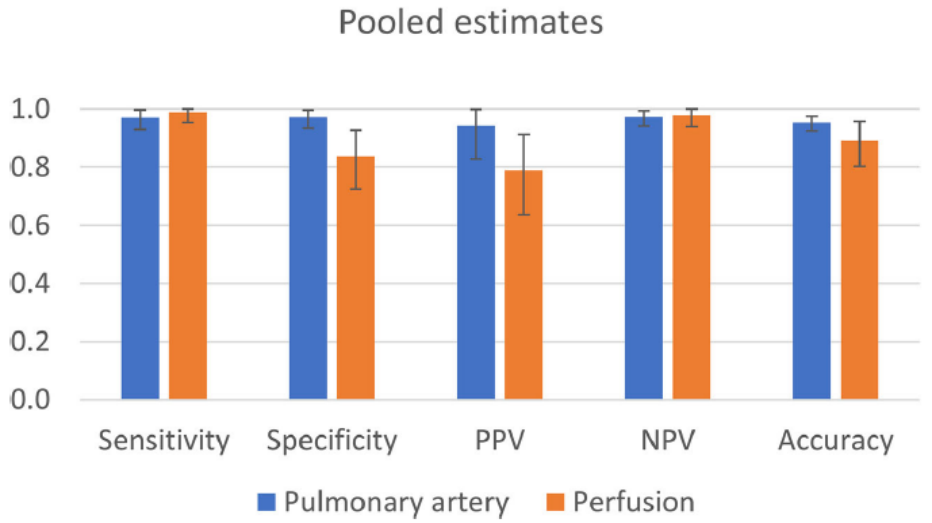
The diagnostic performance of CT pulmonary angiography in the detection of chronic thromboembolic pulmonary hypertension—systematic review and meta-analysis

10 studií, 734 pacientů
CTA má vysokou senzitivitu a vysokou specificitu, pokud je prováděna expertním radiologem

Author	Year published	Design	Age (years) ± SD (range)	Male gender	Inclusion criteria	Sample size	Patients with CTEPH
Tunariu [8]	2007	R	42 (18–81)	37%	PH of any type	227	78
Bartalena [13]	2008	R	55 (22–87)	36%	PH of any type	107	37
Reichelt [14]	2009	R	59 (18–76)	48%	Suspected CTEPH	27	24
Nakazawa [15]	2011	P	58 (29–80)	67%	Suspected or known CTEPH	51	51
He [16]	2012	P	43 ± 15	43%	Suspected CTEPH	114	51
Dournes [17]	2014	R	67 ± 13	35%	PH of any type	40	14
Masy [18]	2018	R	59 ± 16	25%	PH of any type	80	36
Wang [11]	2020	P	42 ± 15	34%	Suspected CTEPH	150	51
Fathala [19]	2021	R	41 ± 10	37%	CTEPH (scintigraphy, PEA)	54	54
Schüssler [20]	2021	P	63 ± 15	31%	Suspected CTEPH	71	13







Lambert L et al. *Eur Radiol* 2022, Apr 28. doi: 10.1007/s00330-022-08804-5

2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

CTEPH and CTEPD without PH			
2015 Guidelines	**Class**	**2022 Guidelines**	**Class**
Lifelong anticoagulation is recommended in all patients with CTEPH	I	Lifelong, therapeutic doses of anticoagulation are recommended in all patients with CTEPH	I
		Antiphospholipid syndrome testing is recommended in patients with CTEPH	I
		In patients with CTEPH and antiphospholipid syndrome, anticoagulation with VKAs is recommended	I

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022

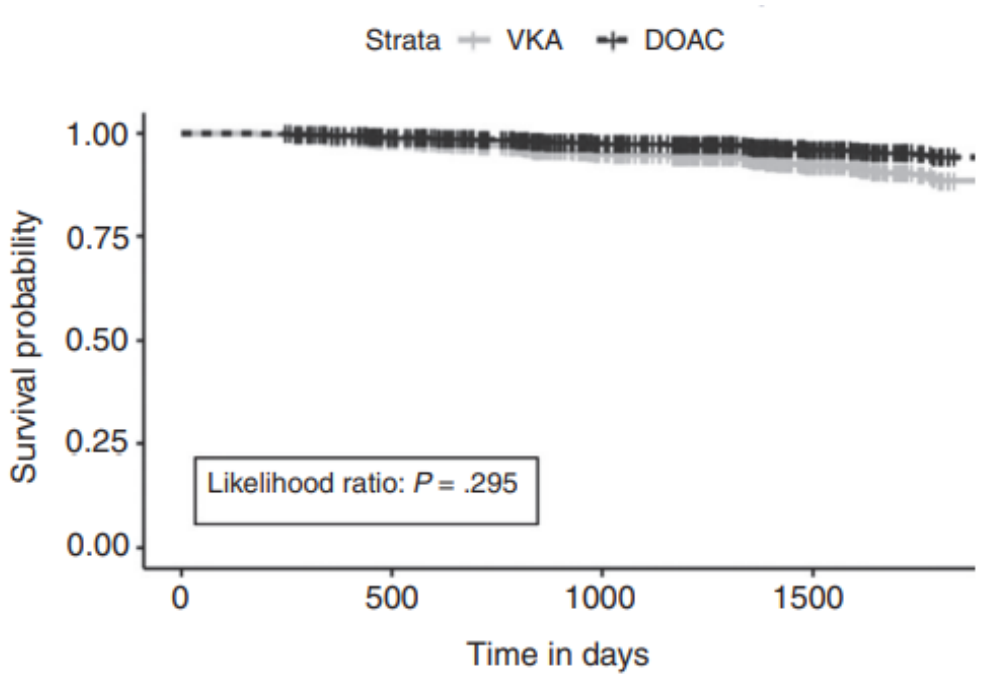
A multicenter study of anticoagulation in operable chronic thromboembolic pulmonary hypertension

Retrospective analysis, PEA 2007-2018, **794 VKA**, **206 DOACs**, mean observation period 612 days
Significant improvements in hemodynamics and functional status in both groups following PEA
Major bleeding events equivalent (P = 1)
VTE recurrence higher (P = .008) with DOACs (4.62%/person-year) than VKAs (0.76%/person-year), survival did not differ

Recurrent VTE & bleeding events

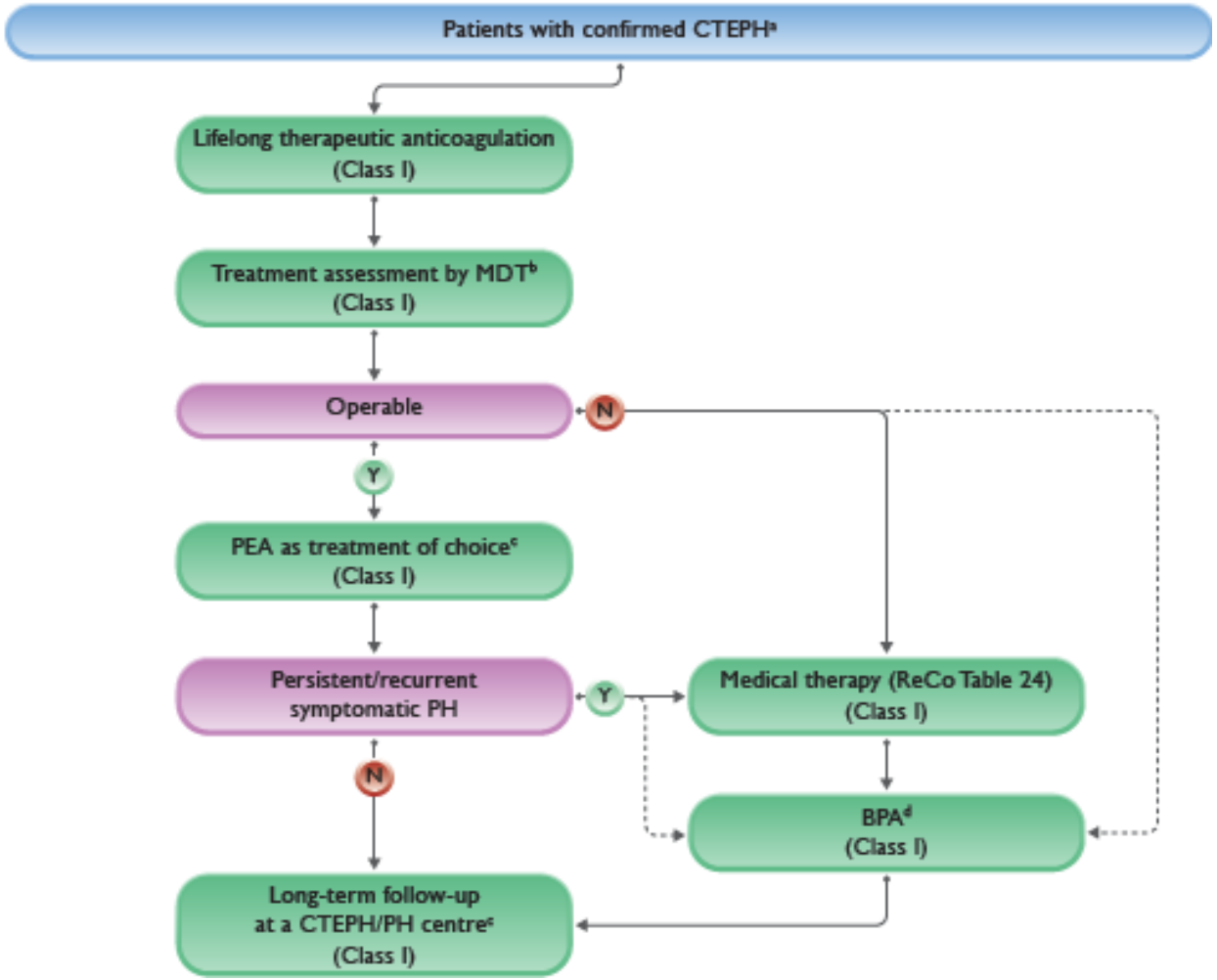
	VKA		DOAC	
	n	Events	*n*	Events
Recurrent VTE	-	-	-	-
Pulmonary embolism	11	11	10	10
Deep vein thrombosis	1	1	0	0
Major bleeding	-	-	-	-
Fatal events	3	3	0	0
Central nervous system	3	3	0	0
Retroperitoneal	1	1	0	0
Hemopericardium	0	0	0	0
Intraocular	0	0	0	0
Hemoptysis	0	0	0	0
Gastrointestinal	2	2	1	1
Hematuria	1	1	0	0
Clinically relevant non-major bleeding	-	-	-	-
Gastrointestinal	0	0	1	1
Large diffuse hematomas	2	2	0	0
Epistaxis	1	1	0	0

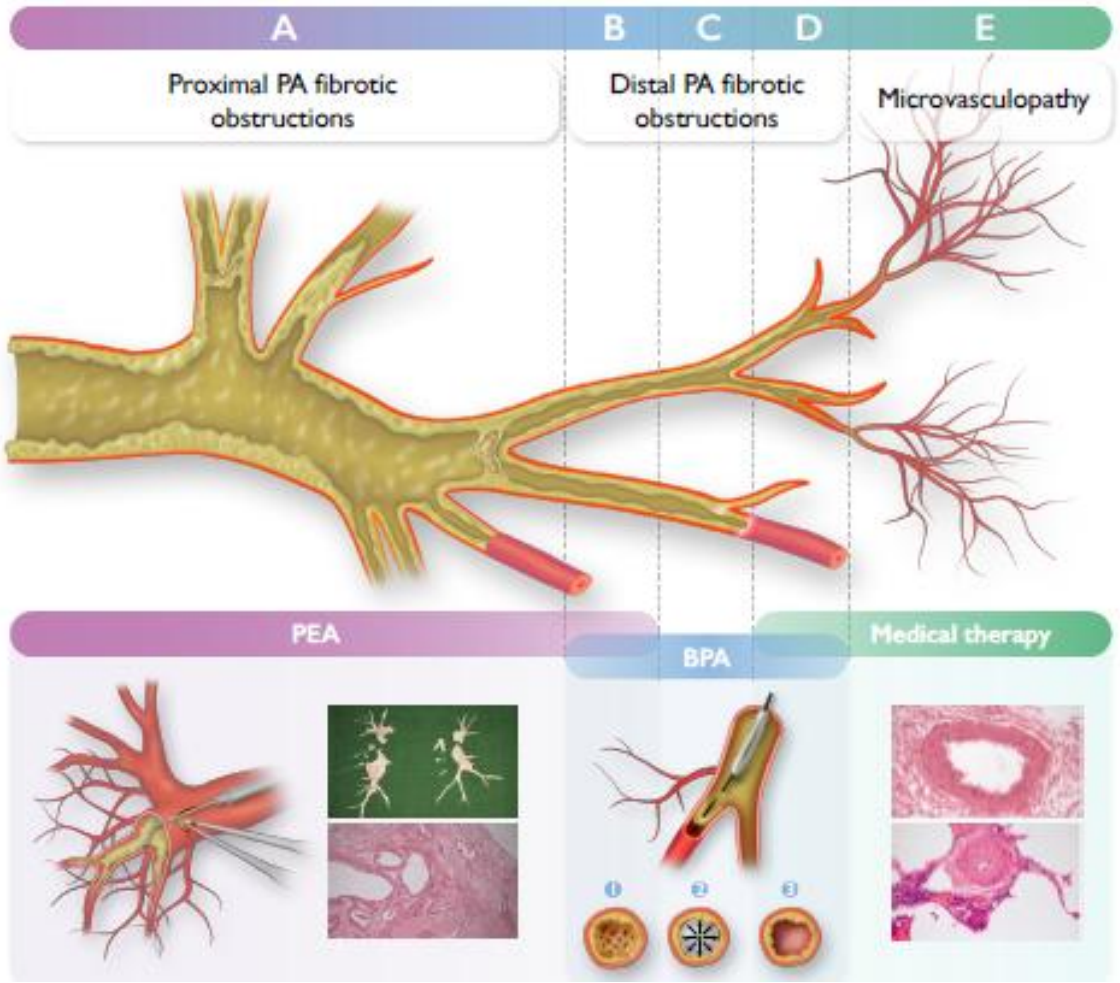
Survival



Bunclark K et al. *J Thromb Haemost.* 2020;18:114–122

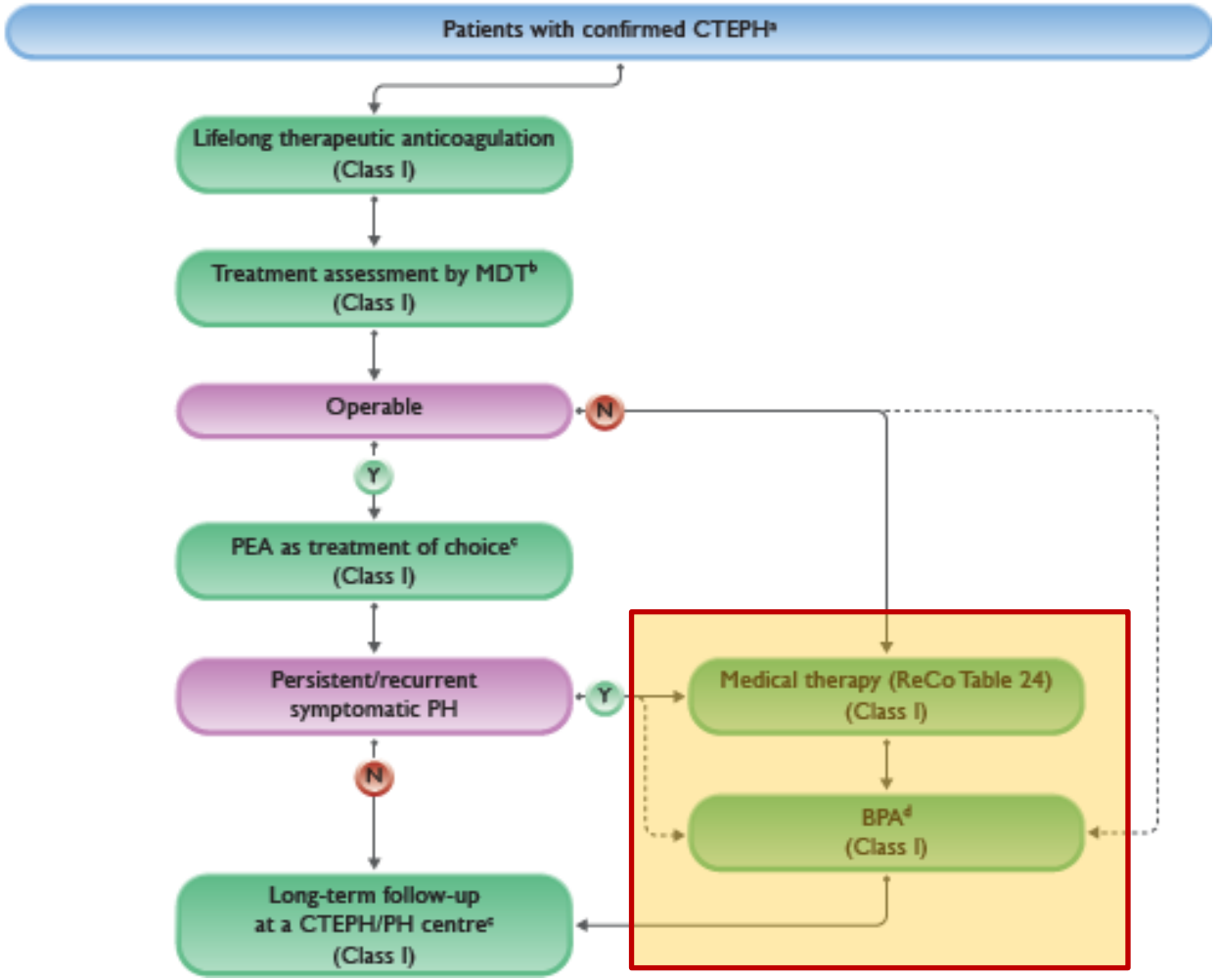
2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

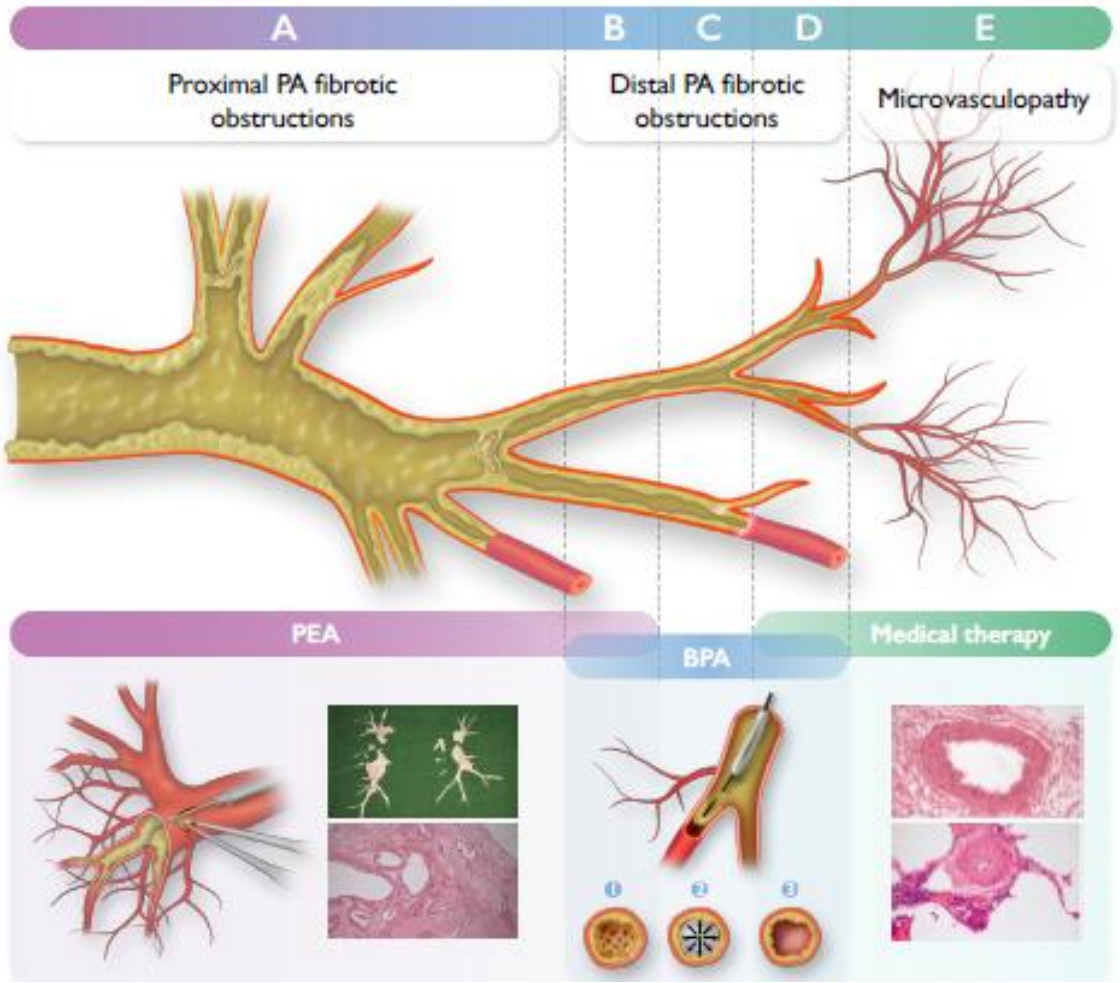




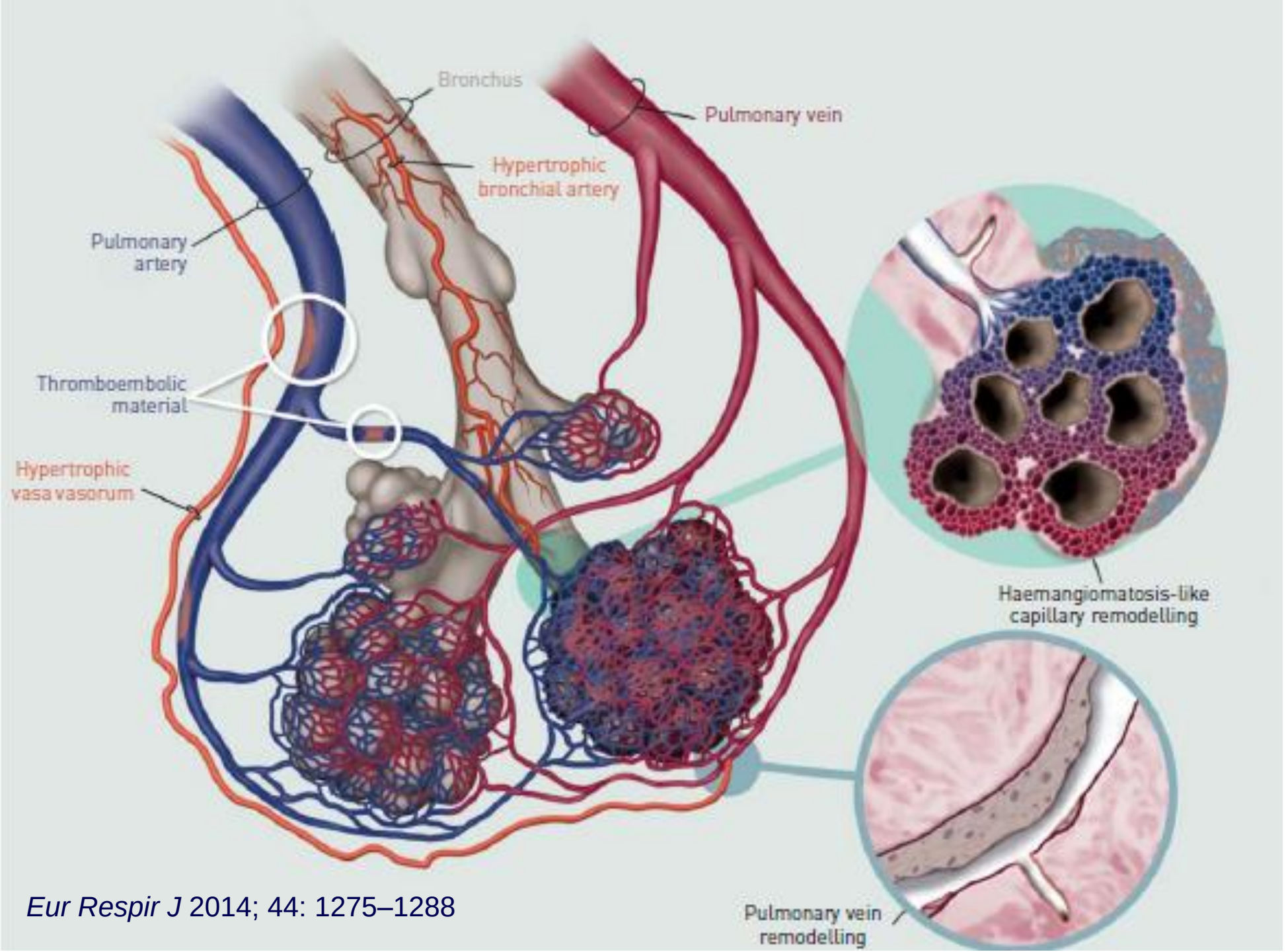
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022

2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension





Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022



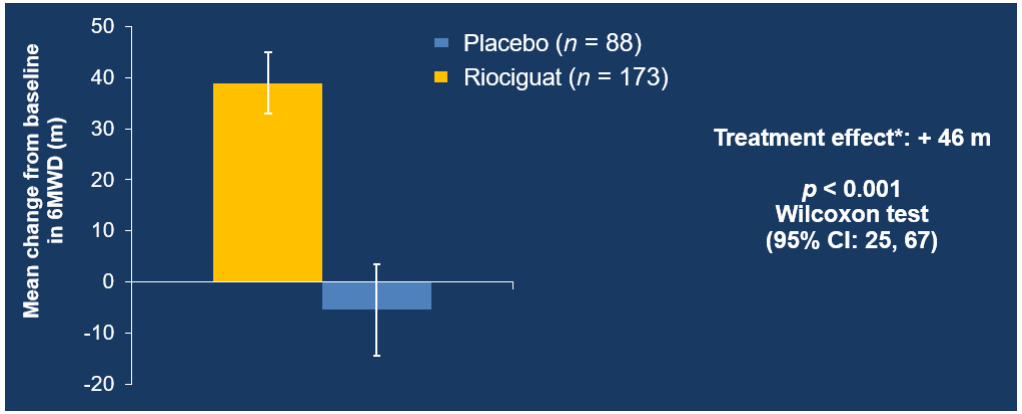
Eur Respir J 2014; 44: 1275–1288

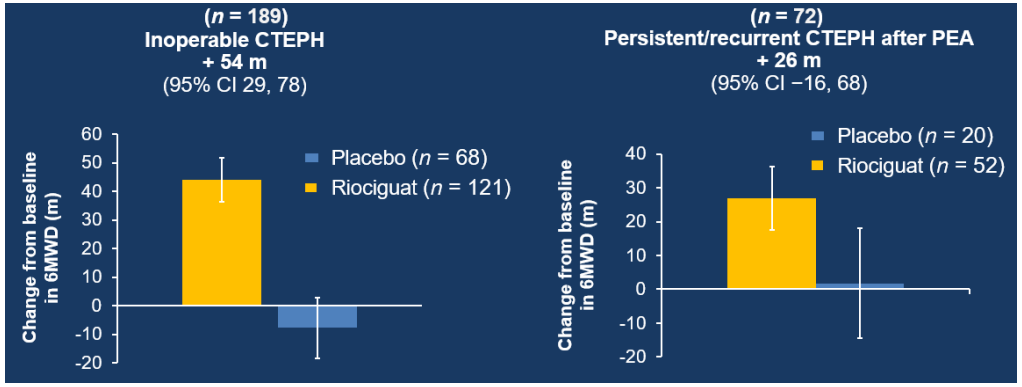
2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

CTEPH and CTEPD without PH (continued)			
2015 Guidelines	**Class**	**2022 Guidelines**	**Class**
Interventional BPA may be considered in patients who are technically non-operable or carry an unfavourable risk:benefit ratio for PEA	IIb	BPA is recommended in patients who are technically inoperable or have residual PH after PEA and distal obstructions amenable to BPA	I
Riociguat is recommended in symptomatic patients who have been classified as having persistent/recurrent CTEPH after surgical treatment or inoperable CTEPH by a CTEPH team including at least one experienced PEA surgeon	I	Riociguat is recommended for symptomatic patients with inoperable CTEPH or persistent/recurrent PH after PEA	I
		Treprostinil s.c. may be considered in patients in WHO-FC III–IV who have inoperable CTEPH or persistent/recurrent PH after PEA	IIb
Off-label use of drugs approved for PAH may be considered in symptomatic patients who have been classified as having inoperable CTEPH by a CTEPH team including at least one experienced PEA surgeon	IIb	Off-label use of drugs approved for PAH may be considered in symptomatic patients who have inoperable CTEPH	IIb

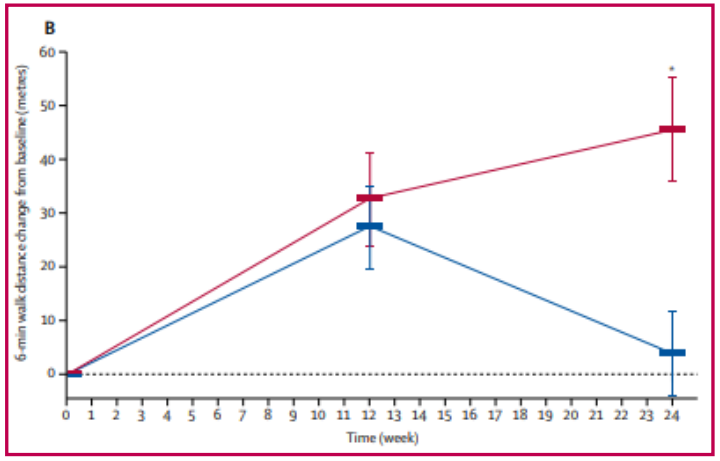
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022

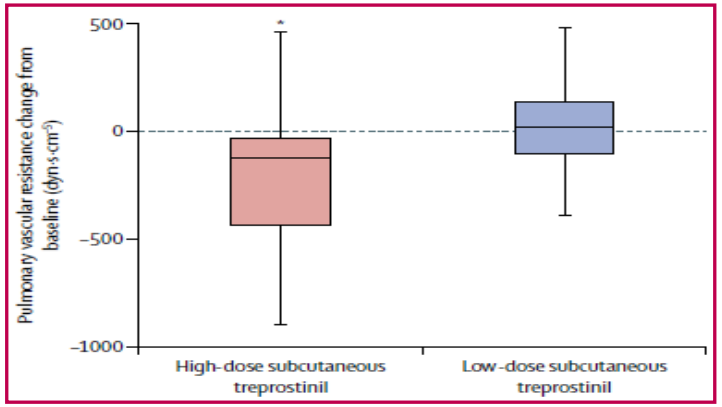
FARMAKOTERAPIE CTEPH





Studie CHEST (riociguat), n=261
Inoperabilní, perzistentní CTEPH, věk 59, 16 týdnů



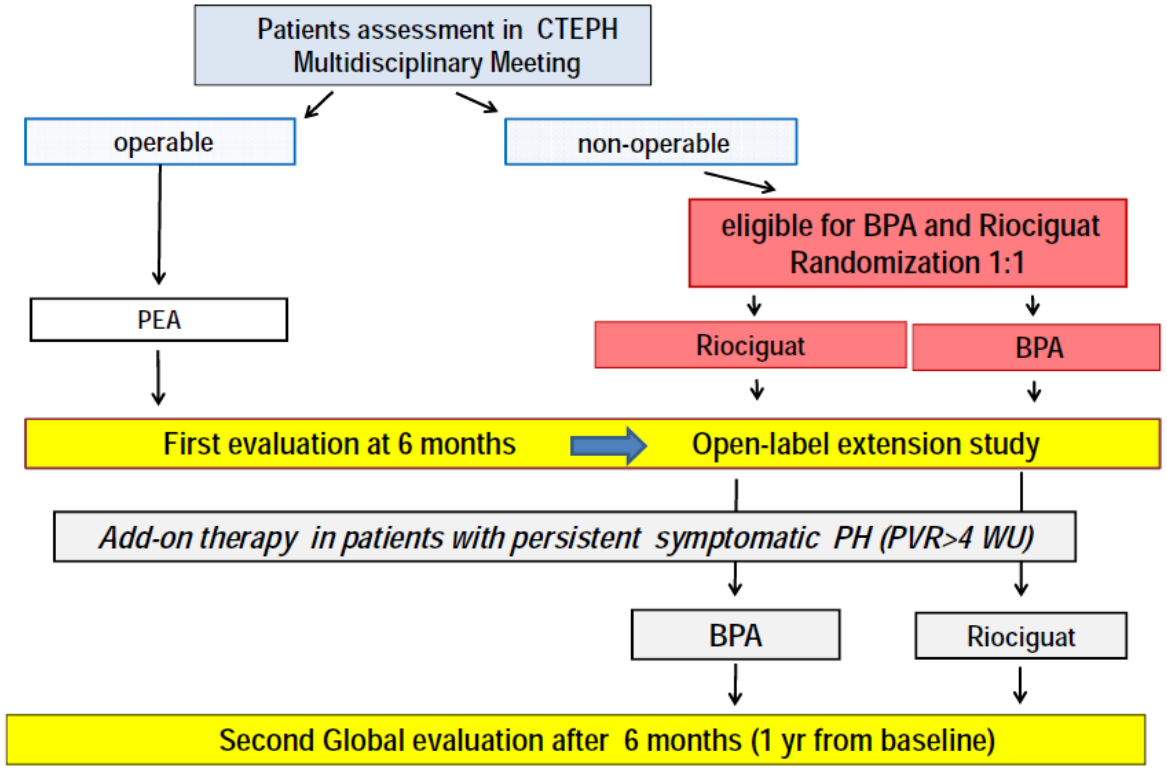


Studie CTREPH (treprostinil), n=105
Inoperabilní, perzistentní CTEPH, věk 64, 24 týdnů

Ghofrani HA et al. *NEJM* 2013, 369: 319-329
Sadushi-Kolici R et al. *Lancet Respir* Med 2019, 7 (3): 239-248

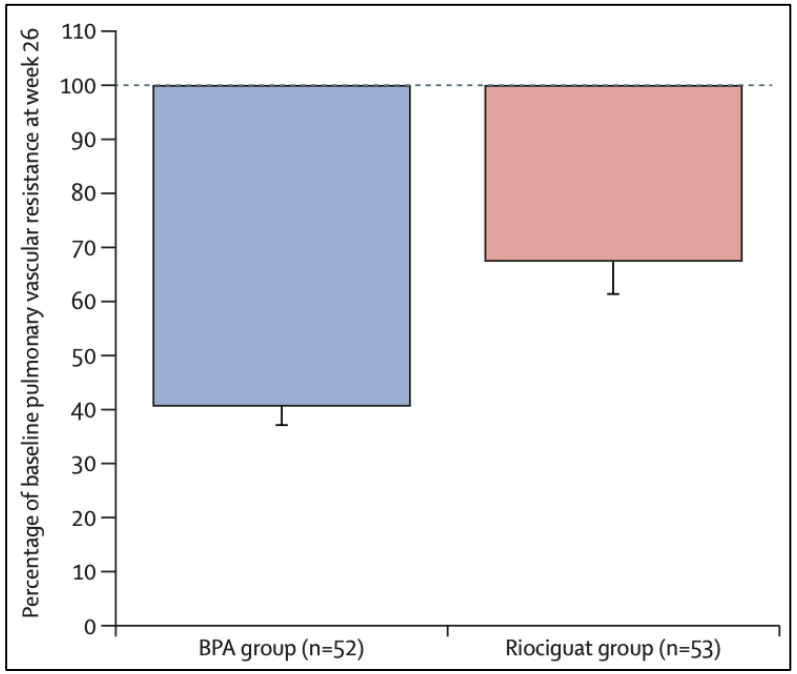
Balloon pulmonary angioplasty versus riociguat for the treatment of inoperable chronic thromboembolic pulmonary hypertension (RACE): a multicentre, phase 3, open-label, randomised controlled trial and ancillary follow-up study

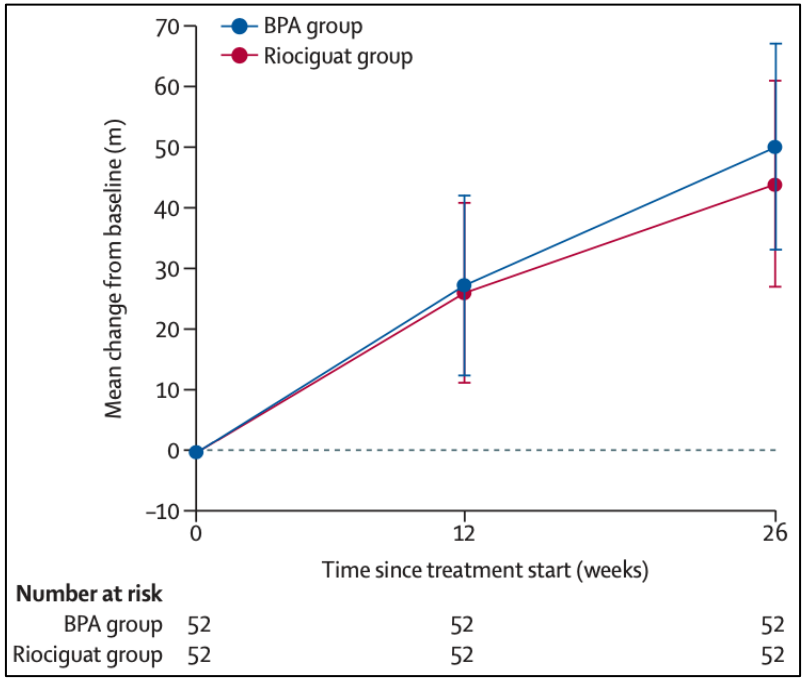
Nově diagnostikovaní pacienti s inoperabilní CTEPH, PVR > 320 dyn·s/cm^5.
2016-2019, N=105 (53 první linie riociguat, 52 první linie BPA)
Týden 52: Srovnatelný pokles PVR, BPA komplikace 14% u předléčených riociguatem vs 42% u první linie BPA



Jais X et al. *Lancet Respir Med* 2022. PublishedonlineAugust1,2022 https://doi.org/10.1016/S2213-2600(22)00214-4

Balloon pulmonary angioplasty versus riociguat for the treatment of inoperable chronic thromboembolic pulmonary hypertension (RACE): a multicentre, phase 3, open-label, randomised controlled trial and ancillary follow-up study





	BPA group (n=52)	Riociguat group (n=53)
Patients with ≥1 adverse event	33 (63%)	38 (72%)
Patients with ≥1 serious adverse event	26 (50%)	14 (26%)
Patients with ≥1 treatment-related serious adverse event	22 (42%)	5 (9%)
Most frequent adverse events (≥3 patients in either group)		
Gastro-oesophageal reflux	0	10 (19%)
Dizziness	1 (2%)	9 (17%)
Haemoptysis	8 (15%)	0
Headache	0	8 (15%)
Vomiting	0	8 (15%)
Cough	0	7 (13%)
Lung injury	8 (15%)	0

Změna PVR, týden 26

Změna 6MWD, týden 26

Komplikace, týden 26

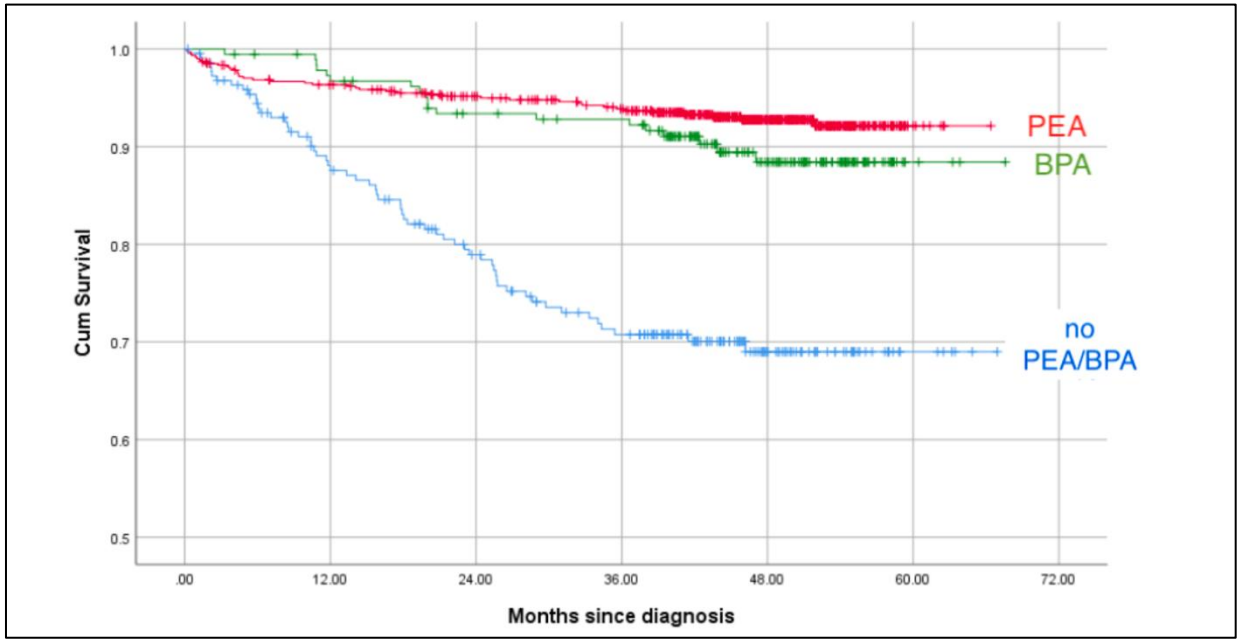
Týden 52:
Srovnatelný pokles PVR, BPA komplikace 14% u předléčených riociguatem vs 42% u první linie BPA

Jais X et al. *Lancet Respir Med* 2022. PublishedonlineAugust1,2022 https://doi.org/10.1016/S2213-2600(22)00214-4

ERS INTERNATIONAL CONGRESS 2021
virtual

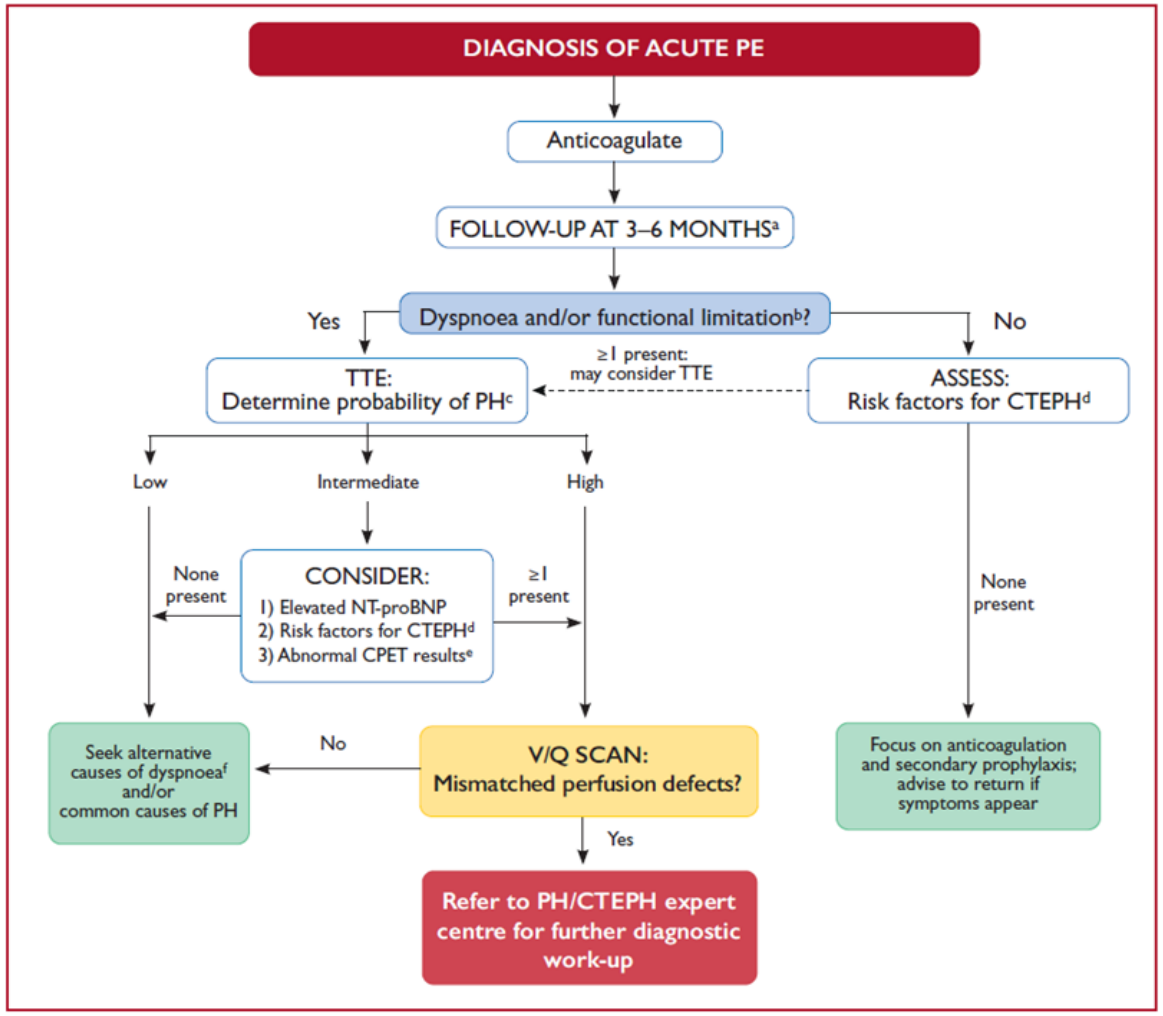
Current strategies for managing CTEPH

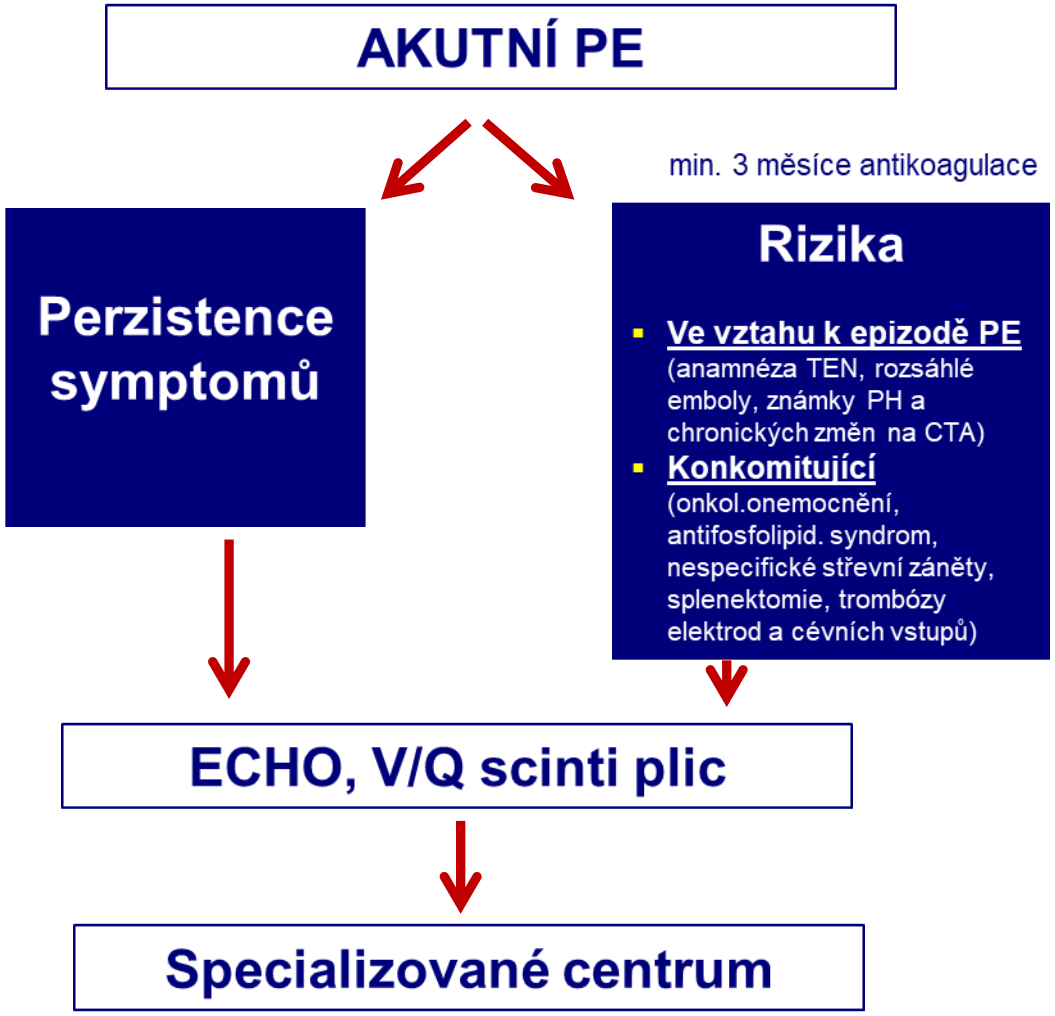
N=1009, newly diagnosed CTEPH patients, inclusion Feb 2015-Sep 2016, FU until Sep 2019
PEA 605 (60 %), 38 % PH drugs
BPA 185 (18 %), 78 % PH drugs
no PEA/BPA 219 (22 %) 76 % PH drugs



Delcroix M et al., ERS International Congress 2021

ČASNÁ DIAGNÓZA CTEPH





Konstantinides SV et al. *Eur Heart J* 2020 Jan 21;41(4):543-603

- Nová hemodynamická definice PH, kritéria zátěžové PH
- Kosmetické změny v diagnostické klasifikaci
- Modifikovaný diagnostický algoritmus PAH (suspekce, detekce, konfirmace)
- Modifikovaný terapeutický algoritmus PAH
  (diagnóza – 3 rizikové skupiny±komorbidity, FU 4 rizikové skupiny)
- Kritéria pro těžkou PH (resp. prekapilární komponentu) u PH skupiny 2, 3
  (PVR > 5 WU)
- Modifikovaná nomenklatura CTEPD/CTEPH
- Modifikovaný terapeutický algoritmus CTEPD/CTEPH
  (bez preference antikoagulace, multimodální léčba)

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022