ČASTĚJŠÍ TYPY PLICNÍ HYPERTENZE (SKUPINA 2, 3, 4)

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European Reference Network for rare or low prevalence complex diseases

Network Respiratory Diseases (ERN-LUNG)

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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)

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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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ESC/ERS GUIDELINES

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

Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS).

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† Representing the Association for European Paediatric and Congenital Cardiology (AEPC)

ESC Clinical Practice Guidelines (CPG) Committee: listed in the Appendix.

ESC subspecialty communities having participated in the development of this document:

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

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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

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

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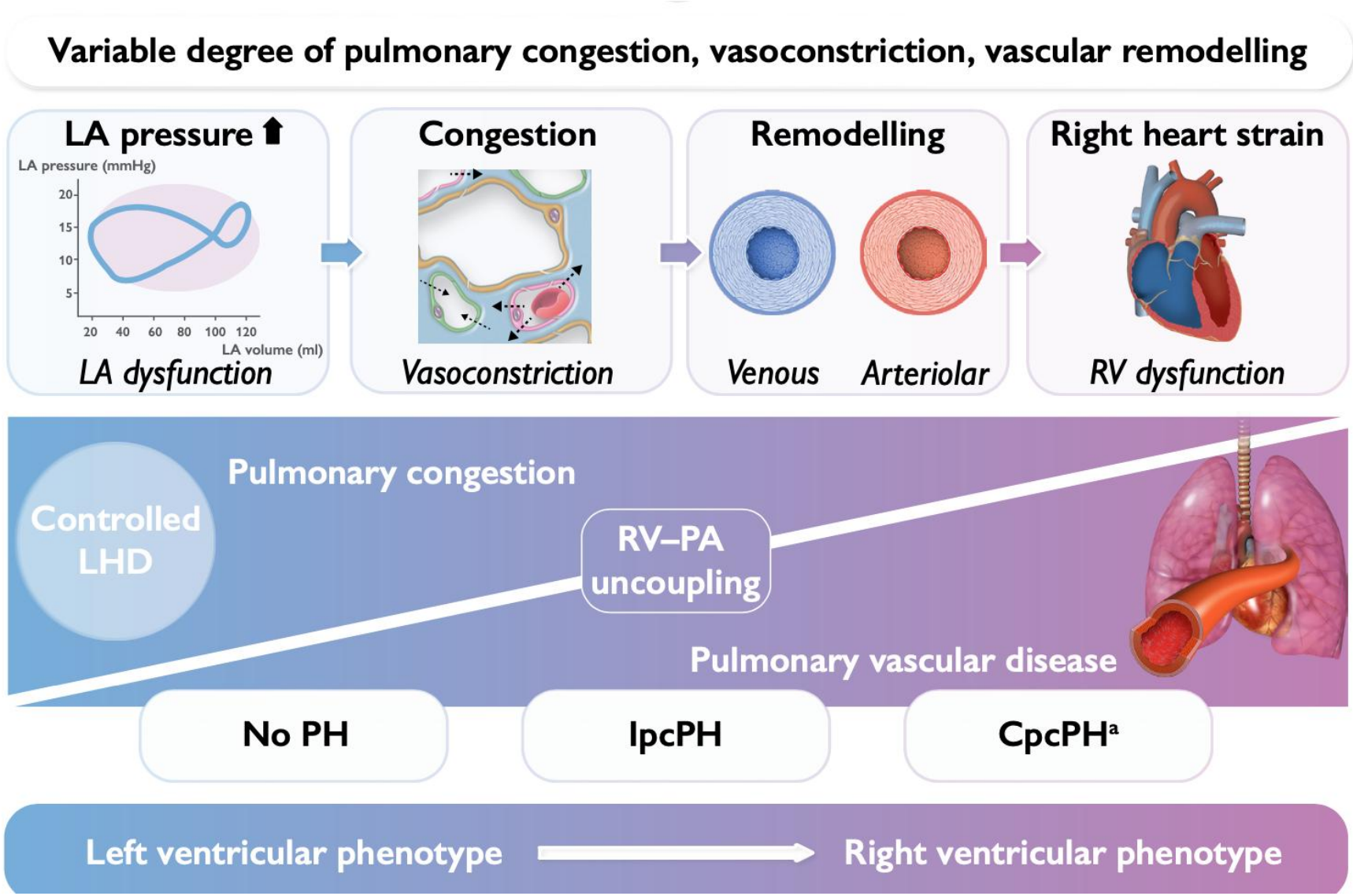
PLICNÍ HYPERTENZE ASOCIOVANÁ S ONEMOCNĚNÍM LEVÉHO SRDCE



Srdeční selhání se zachovalou EF (36-83 % pacientů)

Srdeční selhání s redukovanou EF (40-72 % pacientů)

PVR > 2 WU
(20-30 % pacientů)

Těžká prekapilární komponenta

PVR > 5 WU

- Středně těžká až těžká PH
- Dilatace a/nebo dysfunkce PK

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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

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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 ONEMOCNĚNÍM LEVÉHO SRDCE

- **Terapie základního onemocnění levé komory srdeční, diuretika**
- **Specifická terapie PAH (?)**

  - ERA všeobecně nedoporučovány (zhoršení symptomatologie v důsledku tekutinové retence)

	GRADE			
Recommendations	**Quality of evidence**	**Strength of recommendation**	**Class[a]**	**Level[b]**
No recommendation can be given for or against the use of PDE5is in patients with HFpEF and combined post- and pre-capillary PH	Low	None	–	–
The use of PDE5is in patients with HFpEF and isolated post-capillary PH is not recommended	Low	Conditional	**III**	**C**
In patients with LHD and CpcPH with a severe pre-capillary component (e.g. PVR >5 WU), an individualized approach to treatment is recommended			**I**	**C**

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Pulmonary hypertension due to left heart disease

First author or study [ref.]	Study drug	Dose	Subjects n	Duration	Population	Primary outcome	Result
Guazzi [74]	Sildenafil	50 mg 3 times a day	44	12 months	HFpEF	PVR, RV performance, CPET	Improvement
LEPHT [75]	Riociguat	0.5, 1 or 2 mg 3 times a day	201	16 weeks	HFrEF	mPAP *versus* placebo	No change
Hoendermis [73]	Sildenafil	60 mg 3 times a day	52	12 weeks	HFpEF	mPAP *versus* placebo	No change
SIOVAC [77]	Sildenafil	40 mg 3 times a day	231	24 weeks	VHD	Composite clinical score#	Worsening in active group
MELODY-1 [76]	Macitentan	10 mg once daily	48	12 weeks	HF (EF >30%); 75% HFpEF	Safety and tolerability	+10% fluid retention in active group

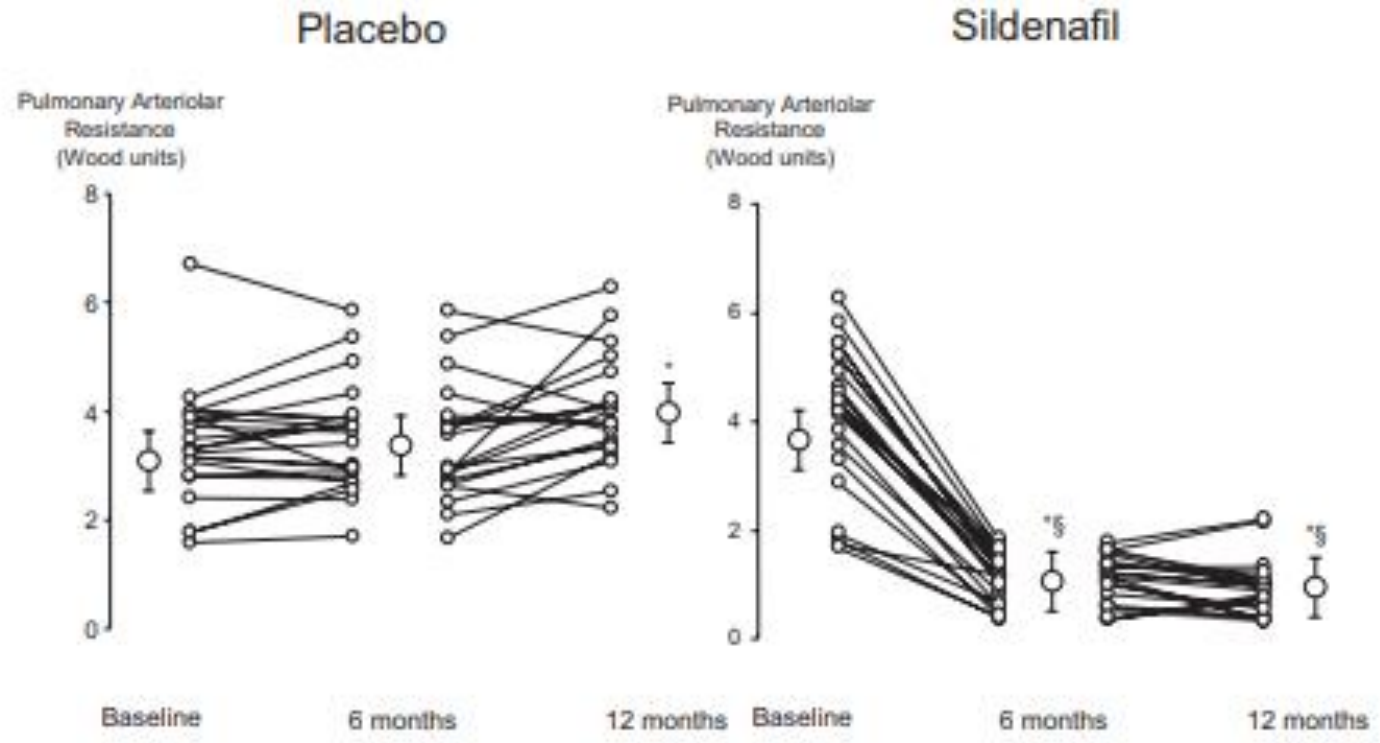
Vachiéry JL et al. *Eur Respir J* 2019 Jan; 53(1): 1801897

Pulmonary Hypertension in Heart Failure With Preserved Ejection Fraction

A Target of Phosphodiesterase-5 Inhibition in a 1-Year Study

Marco Guazzi, MD, PhD; Marco Vicenzi, MD;
Ross Arena, PhD; Maurizio D. Guazzi, MD, PhD, FESC

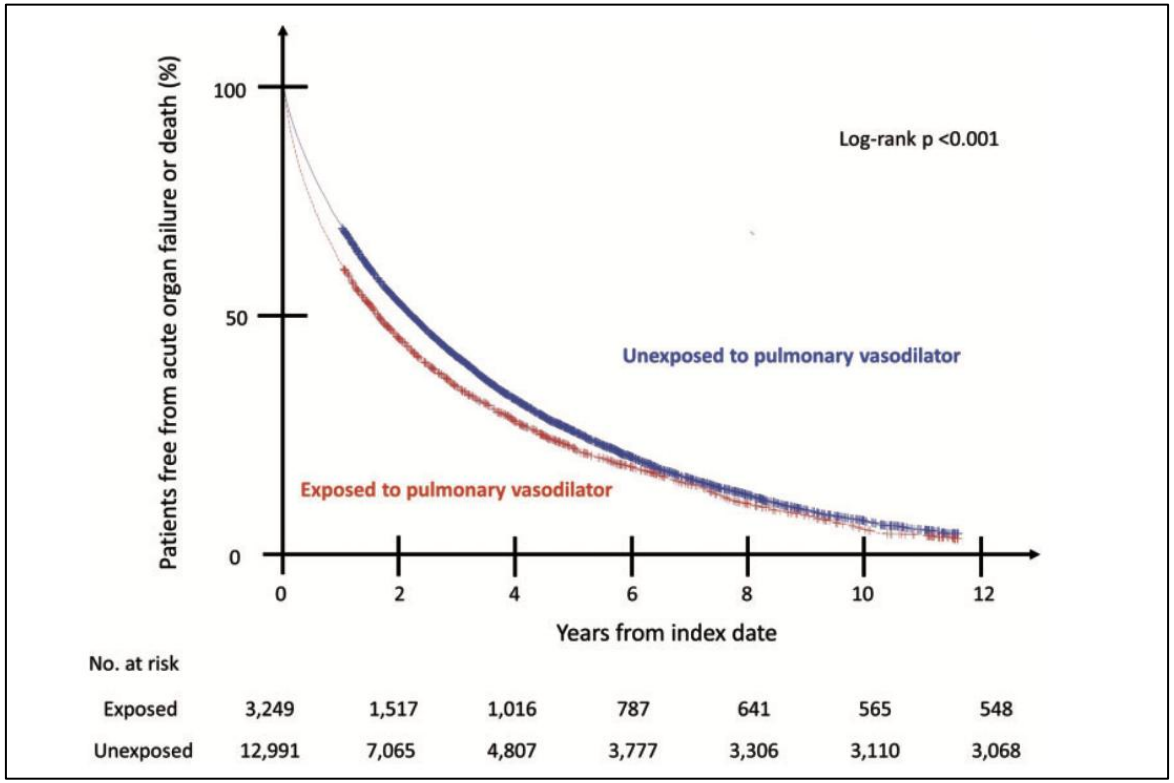
N=44 (randomizace 1:1), EF ≥ 50 %, PASP > 40 mmHg, Sildenafil 3x50 mg
Měsíc 6: sildenafil PAMP pokles o 42±13 %, obdobně měsíc 12



Circulation. 2011;124:164-174

Outcomes of pulmonary vasodilator use in Veterans with pulmonary hypertension associated with left heart disease and lung disease

2006-2016, n=132552, PH skupiny 2/3, 2.5 % (n=3249) specifická vazodilatační léčba PH
Úmrtí nebo selhání orgánů



Gillmeyer KR et al. *Pulmonary Circulation* 2021; 11(2) 1–12

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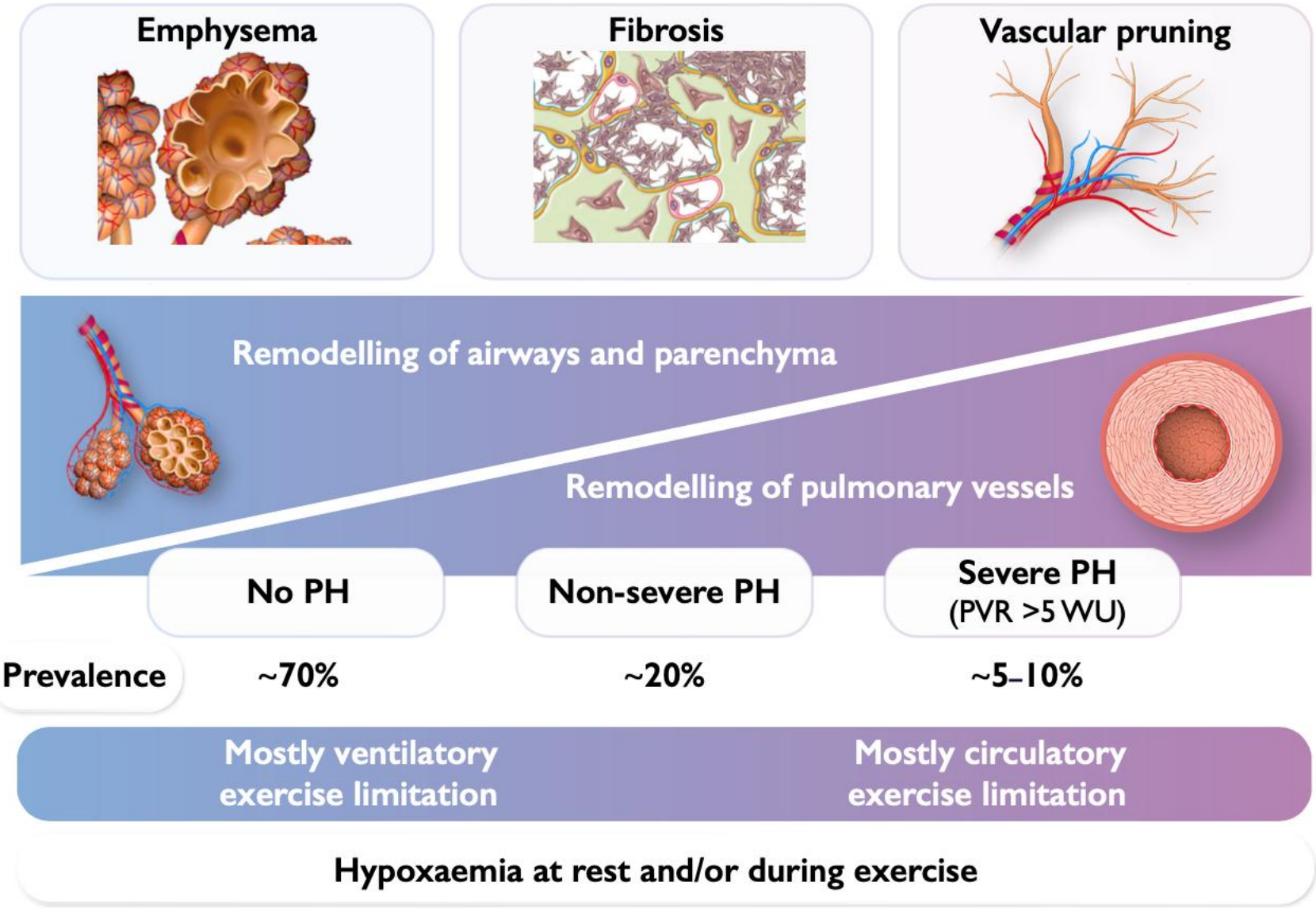
PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

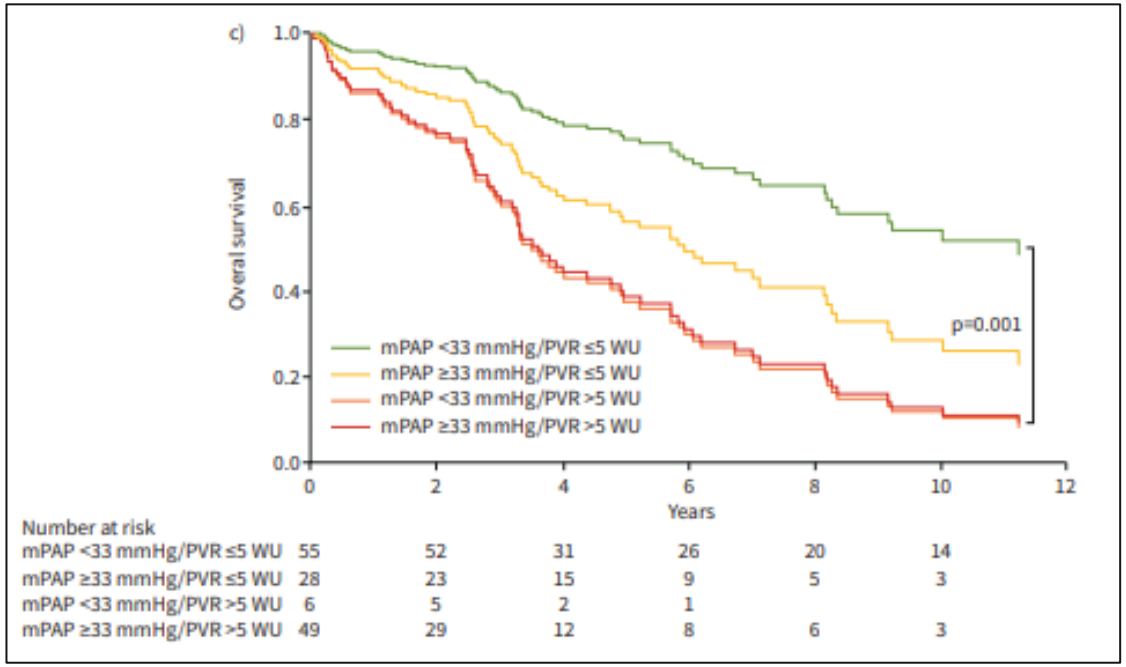


Těžká PH
PVR > 5 WU

- Bez vztahu k plicním objemům
- Hypoxémie
- Nízká DLco

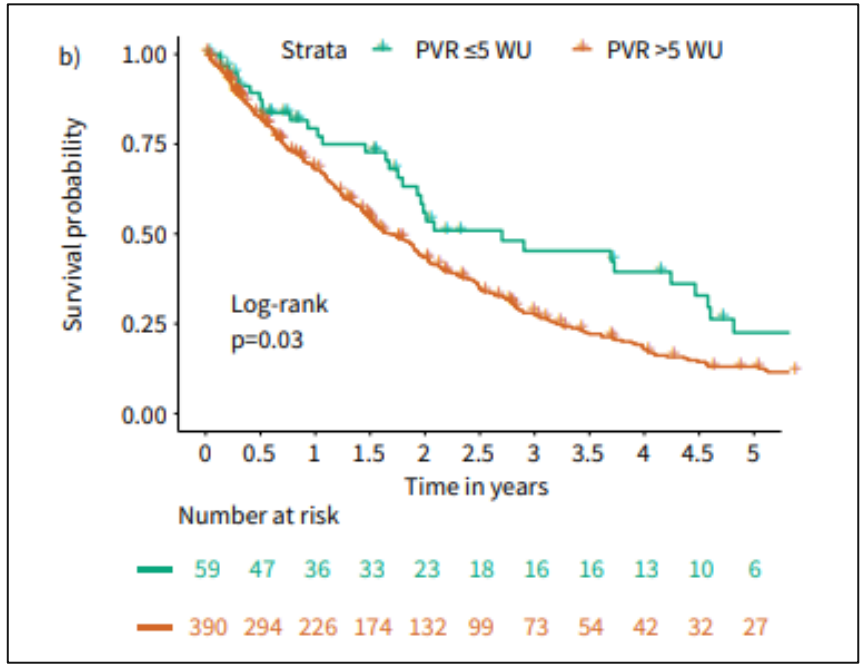
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

Elevated pulmonary vascular resistance predicts mortality in COPD patients



N=139, věk 68, 55 % mužů
PAMP 35 (27–43) mmHg, PVR 4.3 (2.9–7.3) WU, FEV1 56±20%

Pulmonary vascular resistance predicts mortality in patients with pulmonary hypertension associated with interstitial lung disease: results from the COMPERA registry



N=449, věk 73, 65 % mužů
PAMP 39 (33, 46) mmHg, PVR 7.6 (6.0, 10.6) WU,
TLC 72 (60, 86) %, FEV1 69 (55, 81) %

Zeder K et al. *Eur Respir J* 2021; 58: 2100944,
Olsson KM et al. . *Eur Respir J* 2021; 58: 2101483

PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

Léčba: Léčba základního plicního onemocnění
DDOT pokud indikována
CPAP pokud indikován
Plicní RHB programy
U indikovaných pacientů zvážení LuTx
U kombinované etiologie PH adekvátní léčba
Specifická vazodilatační léčba?

Inhaled treprostinil may be considered in patients with PH associated with ILD[734]	IIb	B
The use of ambrisentan is not recommended in patients with PH associated with IPF[740]	III	B
The use of riociguat is not recommended in patients with PH associated with IIP[181]	III	B
The use of PAH medication is not recommended in patients with lung disease and non-severe PH[e]	III	C

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

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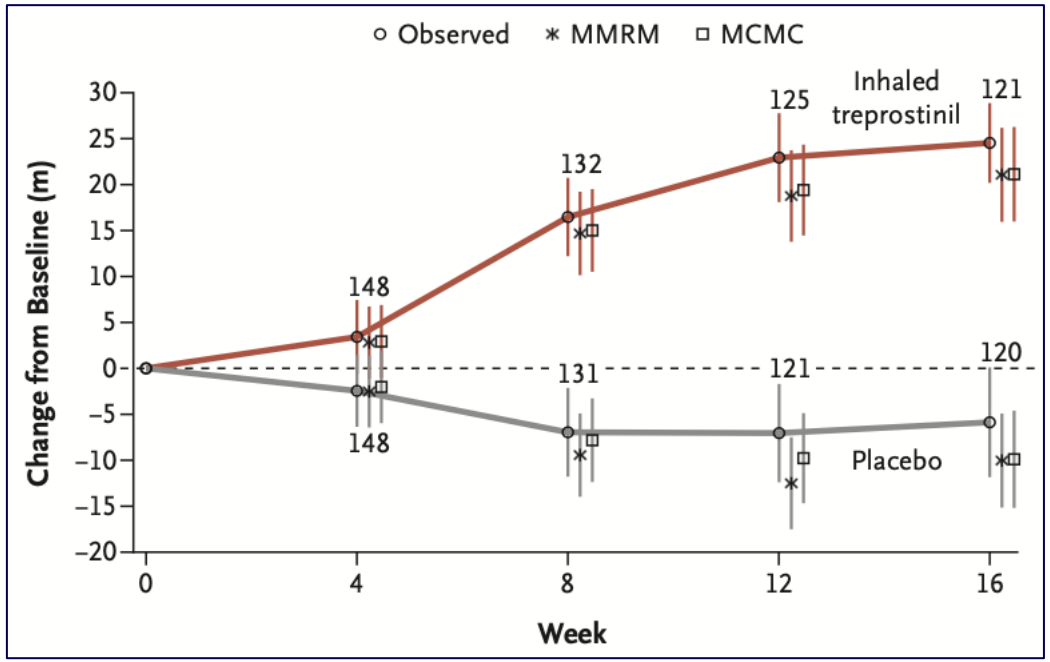
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

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	

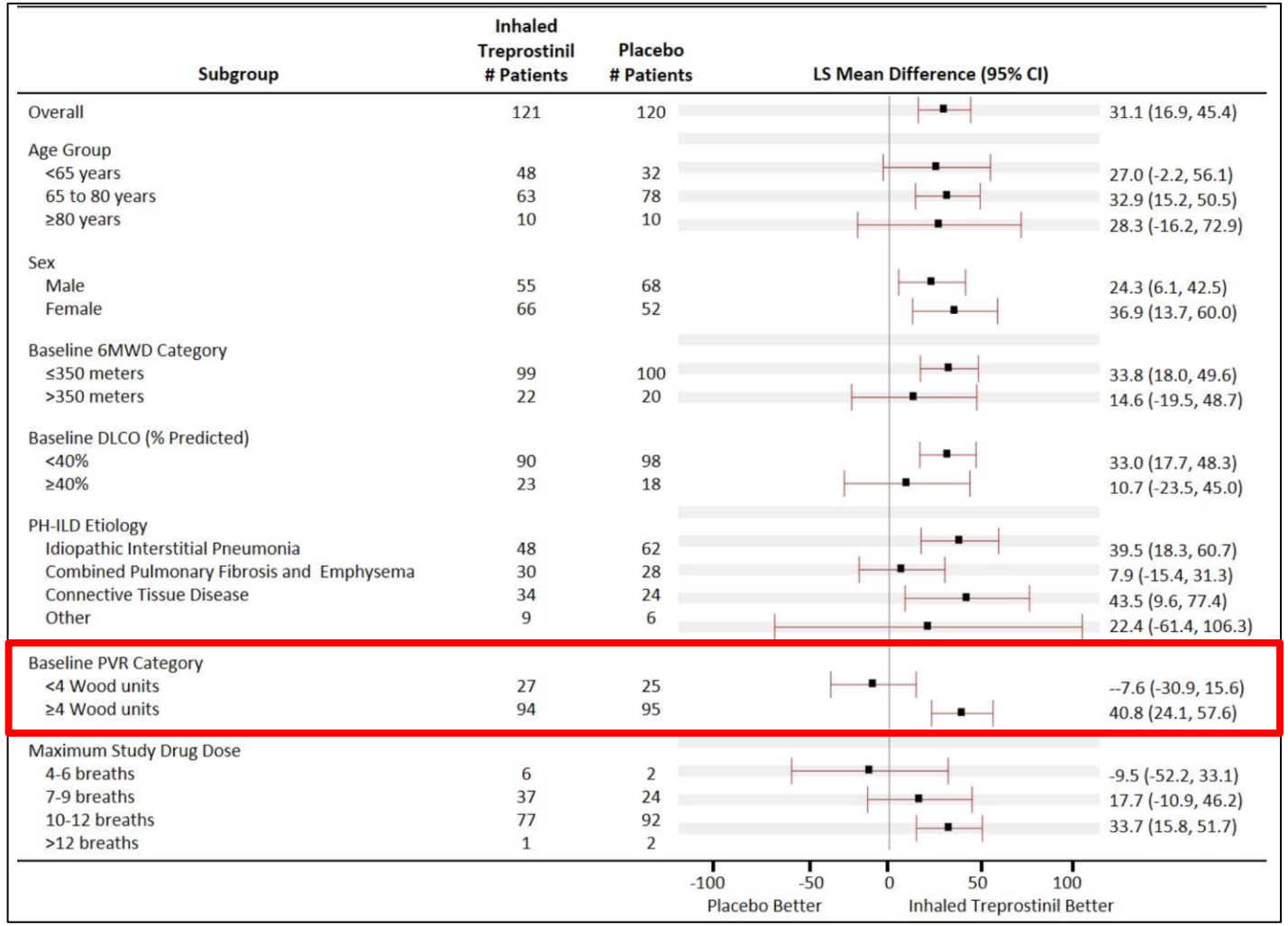
Waxman A et al. *N Engl J Med* 2021;384:325-34.

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

Subgroup	Inhaled Treprostinil # Patients	Placebo # Patients	LS Mean Difference (95% CI)
Overall	121	120	31.1 (16.9, 45.4)
Age Group			
<65 years	48	32	27.0 (-2.2, 56.1)
65 to 80 years	63	78	32.9 (15.2, 50.5)
≥80 years	10	10	28.3 (-16.2, 72.9)
Sex			
Male	55	68	24.3 (6.1, 42.5)
Female	66	52	36.9 (13.7, 60.0)
Baseline 6MWD Category			
≤350 meters	99	100	33.8 (18.0, 49.6)
>350 meters	22	20	14.6 (-19.5, 48.7)
Baseline DLCO (% Predicted)			
<40%	90	98	33.0 (17.7, 48.3)
≥40%	23	18	10.7 (-23.5, 45.0)
PH-ILD Etiology			
Idiopathic Interstitial Pneumonia	48	62	39.5 (18.3, 60.7)
Combined Pulmonary Fibrosis and Emphysema	30	28	7.9 (-15.4, 31.3)
Connective Tissue Disease	34	24	43.5 (9.6, 77.4)
Other	9	6	22.4 (-61.4, 106.3)
Baseline PVR Category			
<4 Wood units	27	25	--7.6 (-30.9, 15.6)
≥4 Wood units	94	95	40.8 (24.1, 57.6)
Maximum Study Drug Dose			
4-6 breaths	6	2	-9.5 (-52.2, 33.1)
7-9 breaths	37	24	17.7 (-10.9, 46.2)
10-12 breaths	77	92	33.7 (15.8, 51.7)
>12 breaths	1	2	



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

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

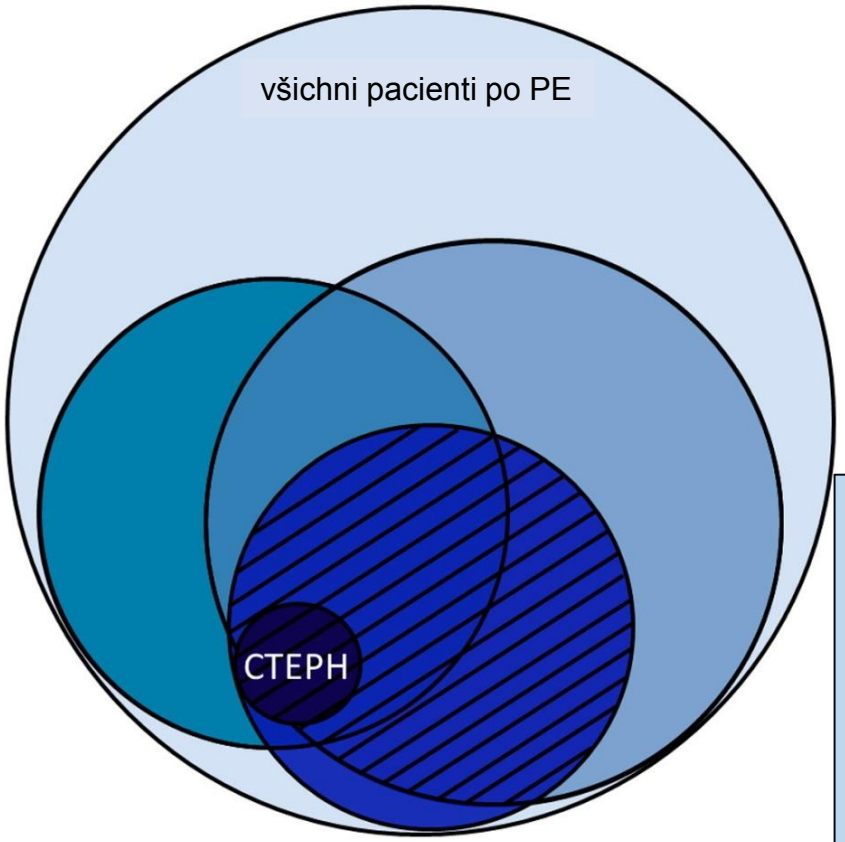
GROUP 4 PH associated with pulmonary artery obstructions — 4 %
- 4.1 Chronic thrombo-embolic PH
- 4.2 Other pulmonary artery obstructions[c]

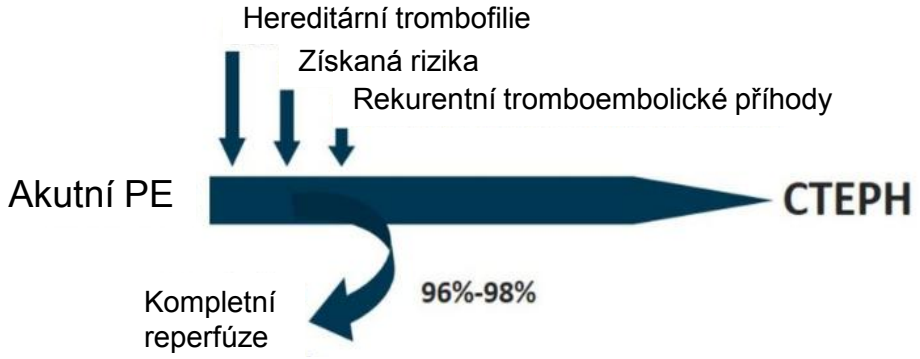
GROUP 5 PH with unclear and/or multifactorial mechanisms — 5 %
- 5.1 Haematological disorders[d]
- 5.2 Systemic disorders[e]
- 5.3 Metabolic disorders[f]
- 5.4 Chronic renal failure with or without haemodialysis
- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

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

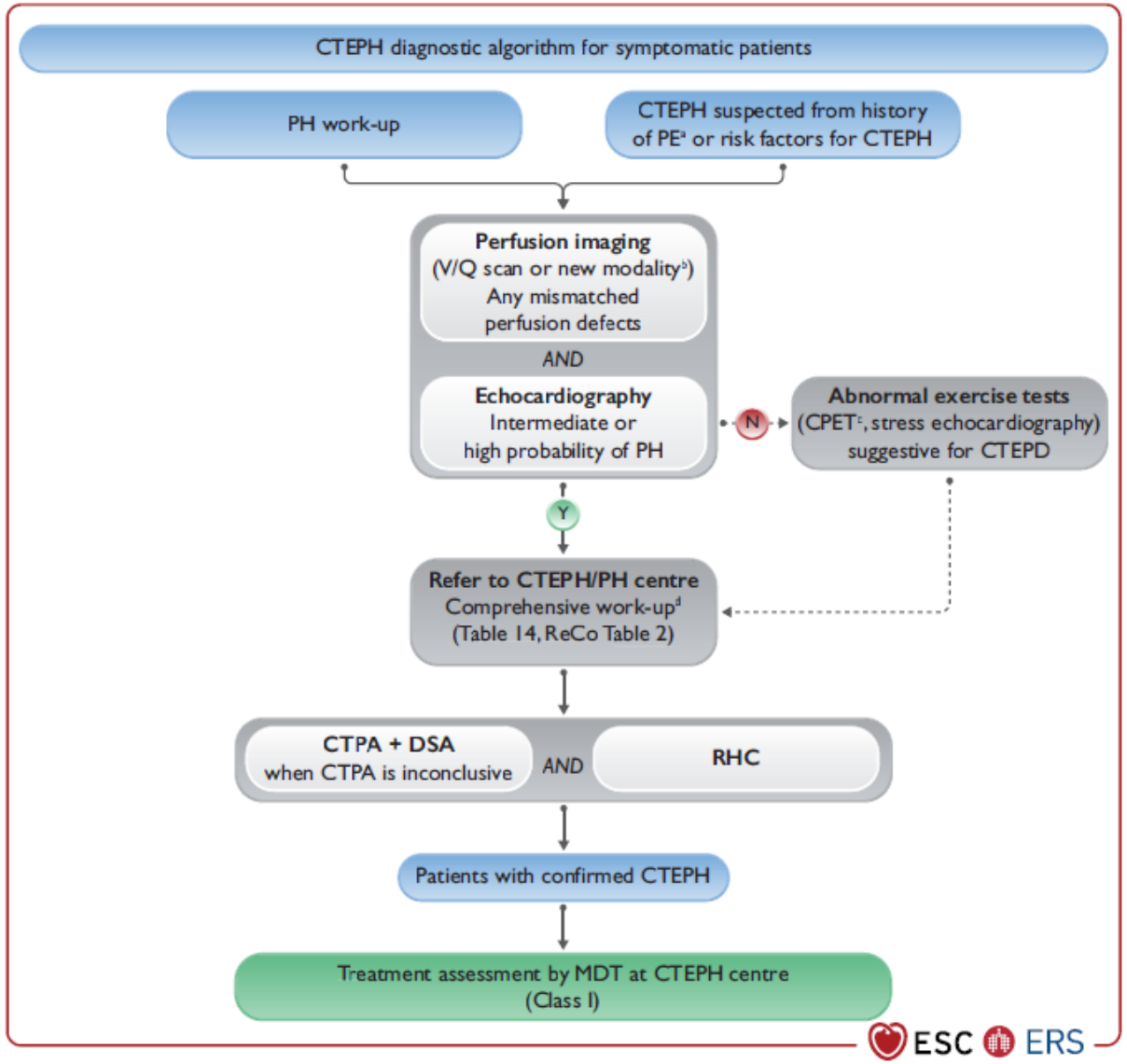
Konsekvence: pravostranné srdeční selhání a smrt

CTEPD bez plicní hypertenze

Trombot. okluze+remodelace+symptomy (bez PH)

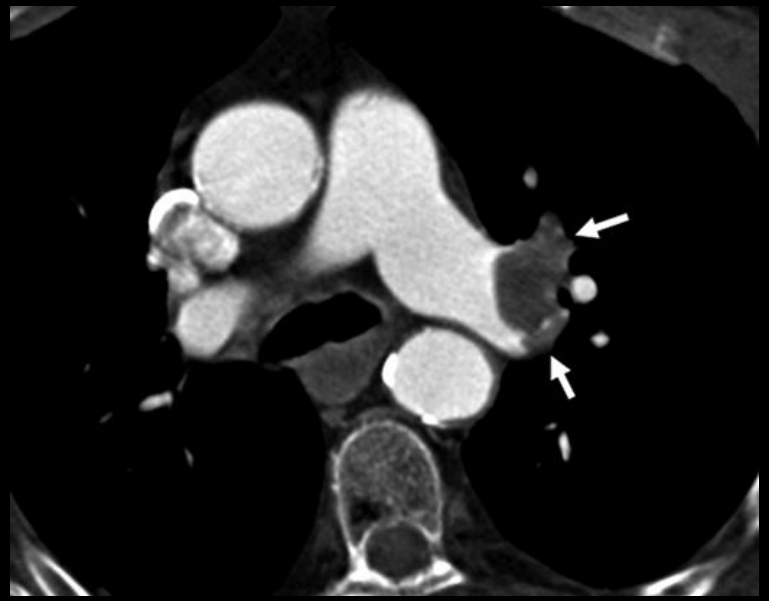
Klok FA et al. *Blood Rev* 2014 Nov;28(6):221-6, Delcroix M et al. *Eur Respir J* 2021 Jun 17;57(6):2002828

DIAGNOSTICKÝ ALGORITMUS CTEPH (2022)

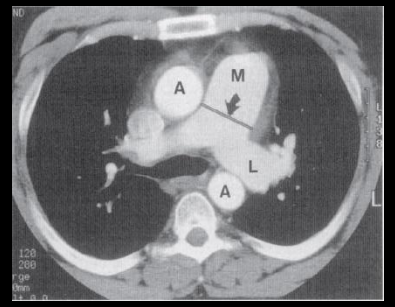


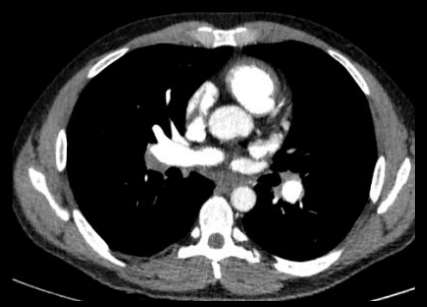
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

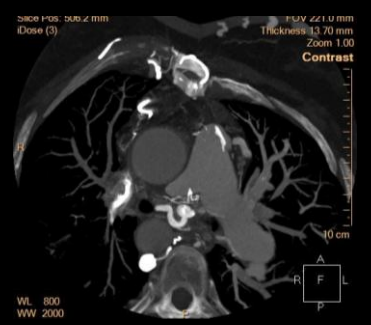
PLICNÍ EMBOLIE

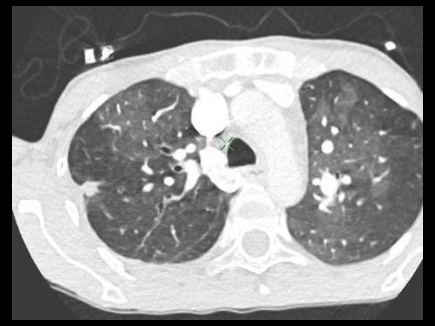


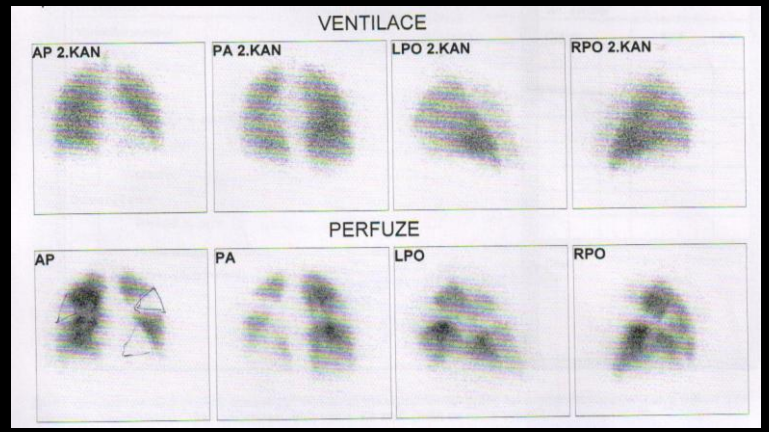
CTEPH

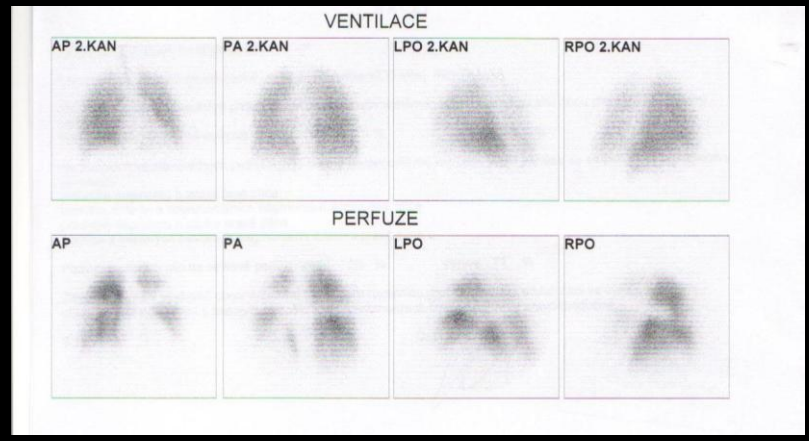












Zdroj: VFN

EUROPEAN RESPIRATORY *journal*
FLAGSHIP SCIENTIFIC JOURNAL OF ERS

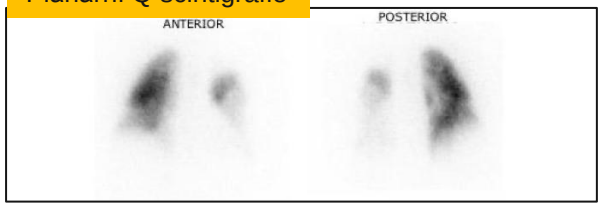
DIAGNOSTIKA

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

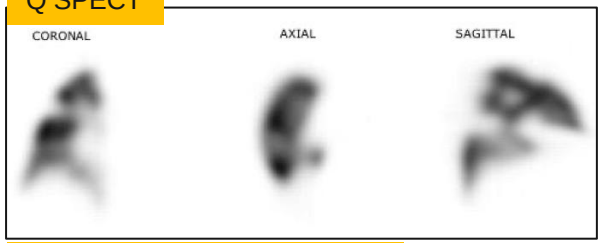
DECT a **MR perfúze** – nenahrazuje scintigrafii (limitovaná dostupnost, zkušenost, validace)

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

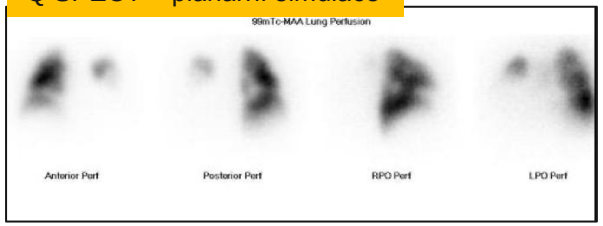
Planární Q scintigrafie

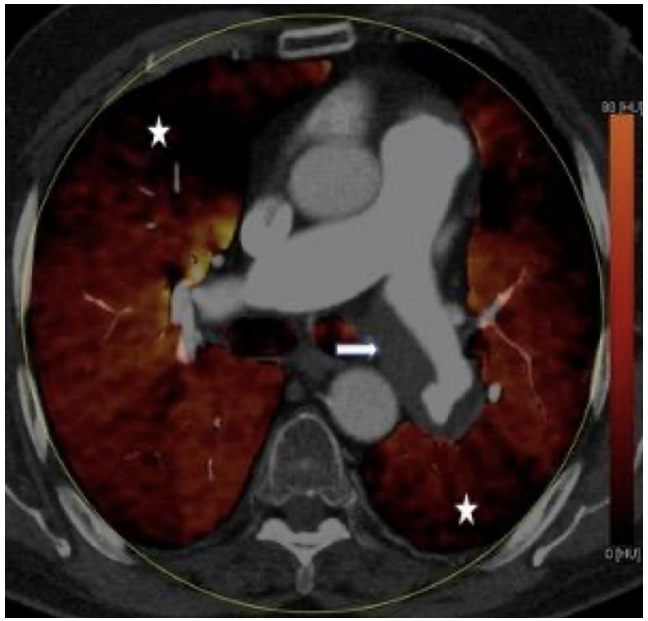


Q SPECT

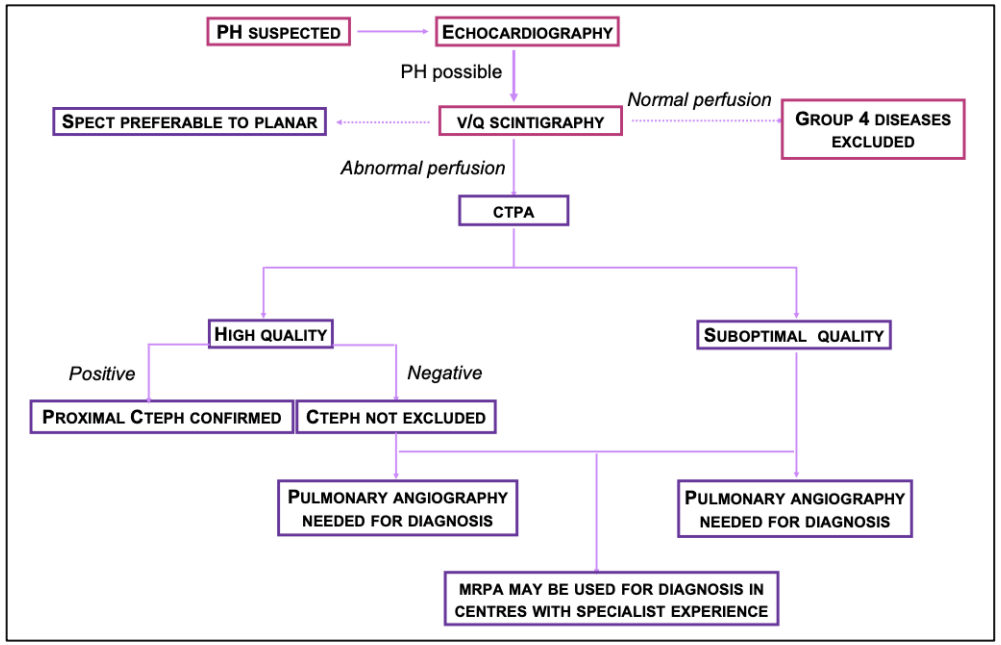


Q SPECT – planární simulace





DECT

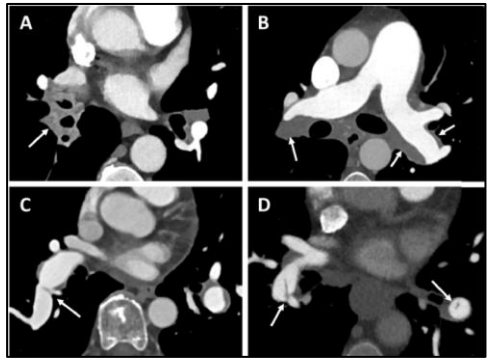


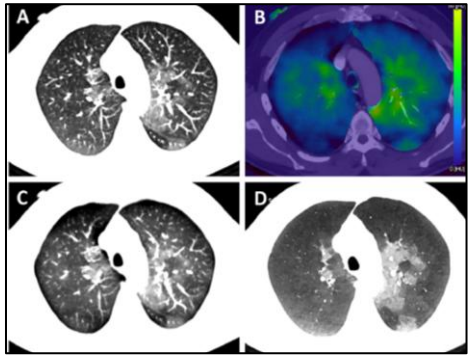
Eur Respir J 2021 Jun 17;57(6):2002828

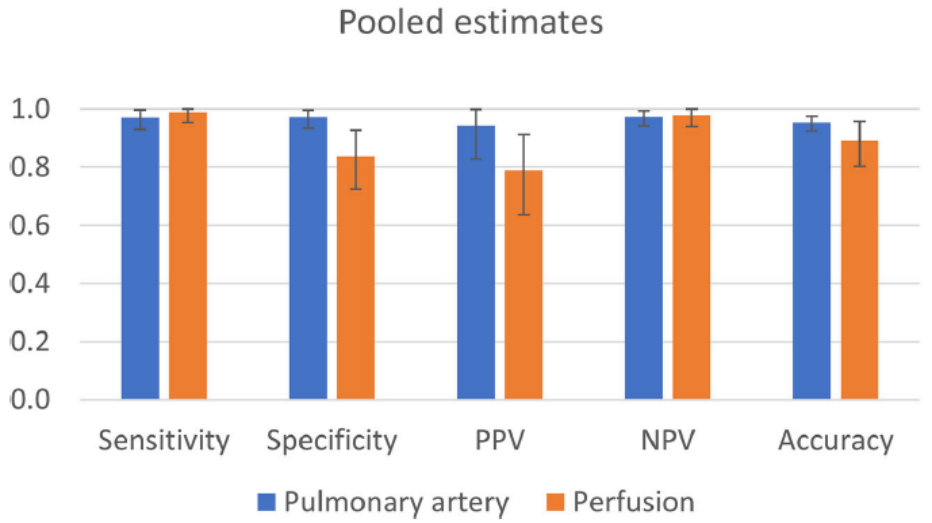
The diagnostic performance of CT pulmonary angiography in the detection of chronic thromboembolic pulmonary hypertension—systematic review and meta-analysis

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

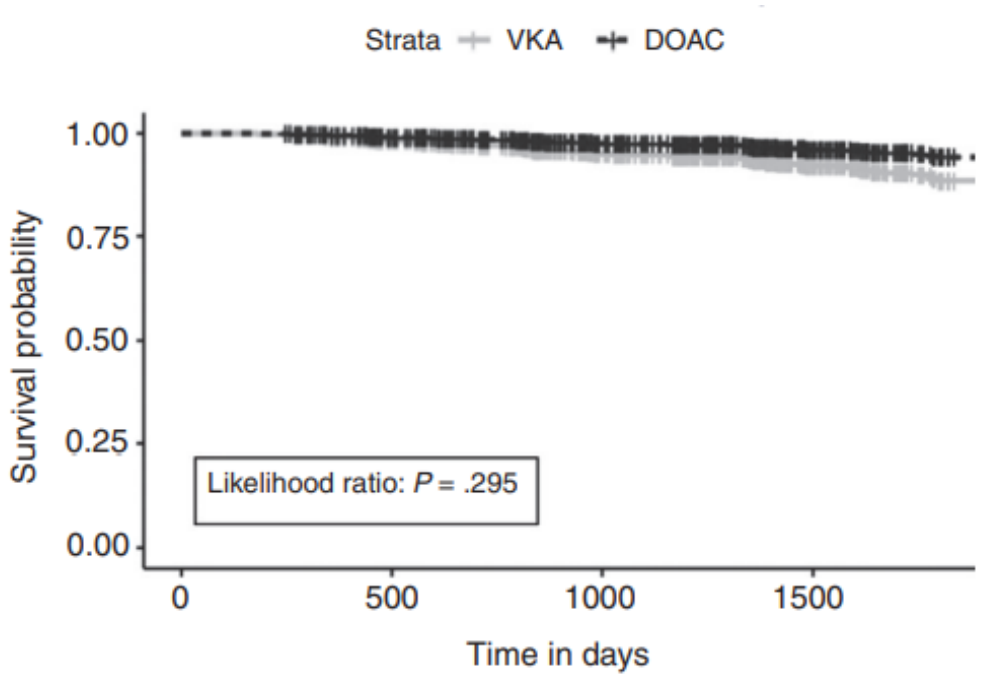
A multicenter study of anticoagulation in operable chronic thromboembolic pulmonary hypertension

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



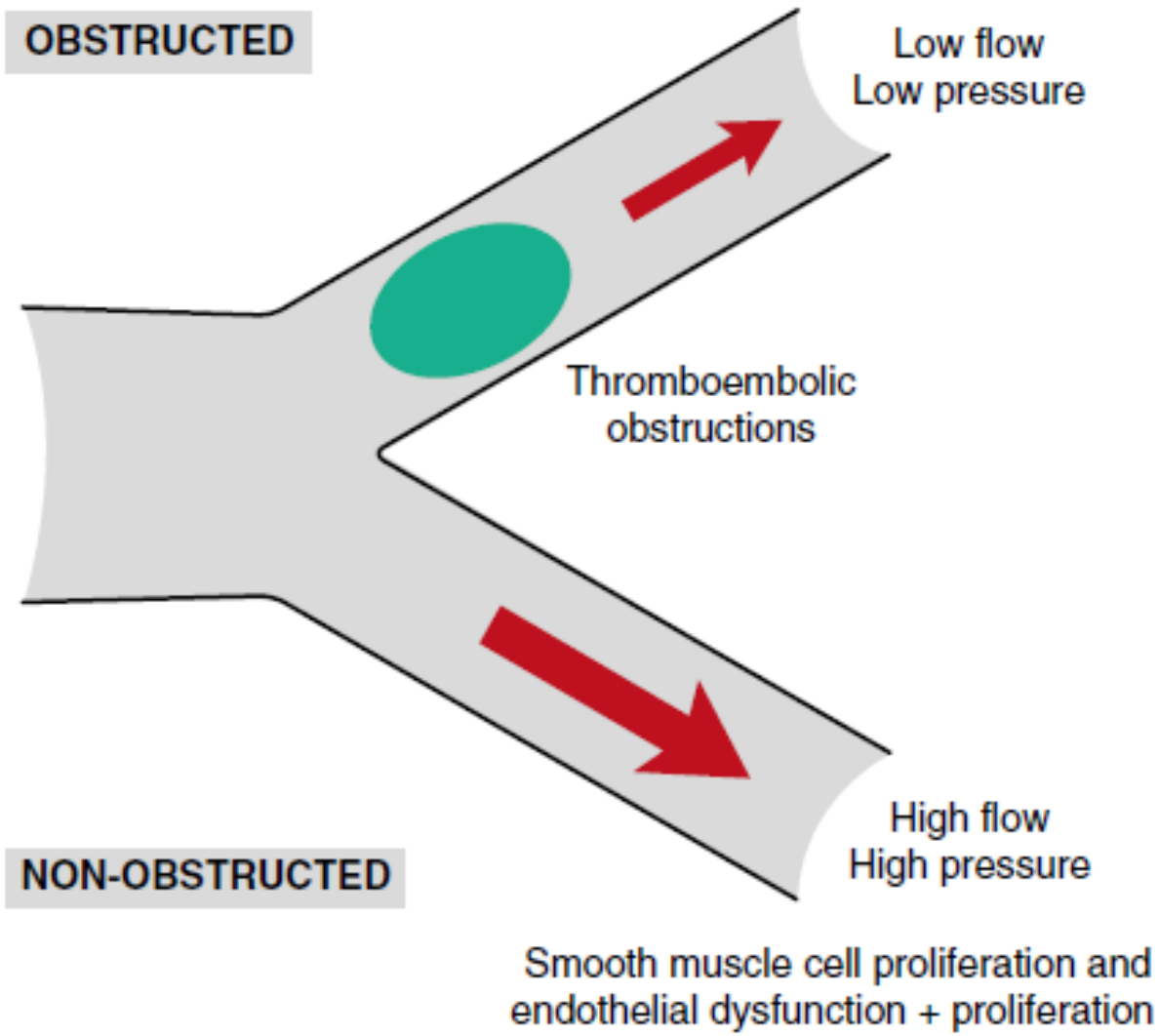
Bunclark K et al. *J Thromb Haemost.* 2020;18:114–122

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

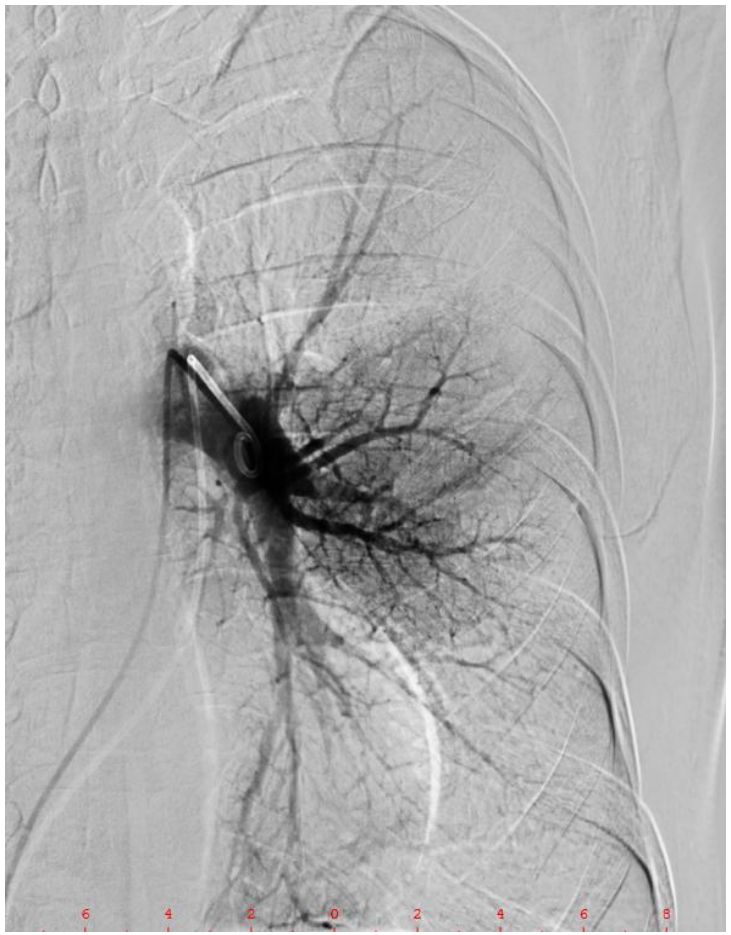
CTEPH and CTEPD without PH			
2015 Guidelines	**Class**	**2022 Guidelines**	**Class**
Lifelong anticoagulation is recommended in all patients with CTEPH	I	Lifelong, therapeutic doses of anticoagulation are recommended in all patients with CTEPH	I
		Antiphospholipid syndrome testing is recommended in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

PATOFYZIOLOGIE CTEPH

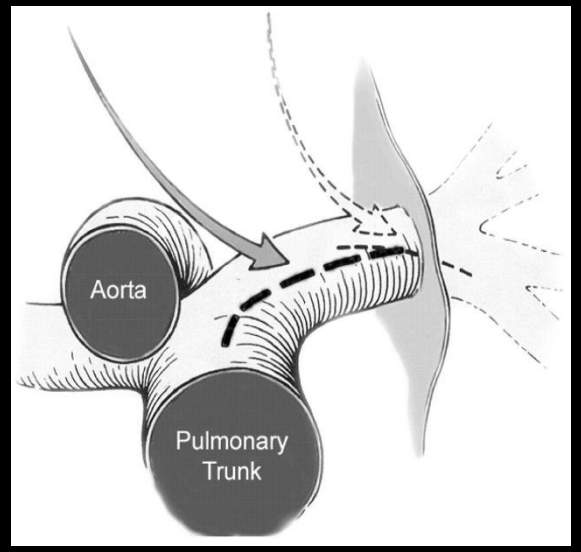


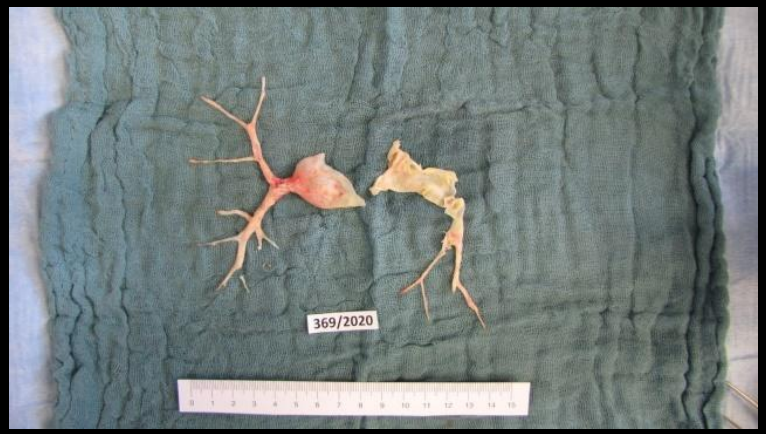
Smooth muscle cell proliferation and endothelial dysfunction + proliferation

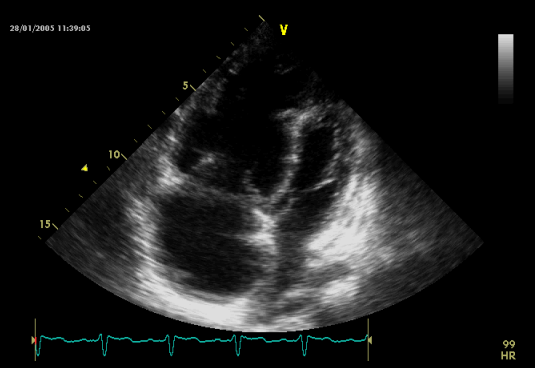


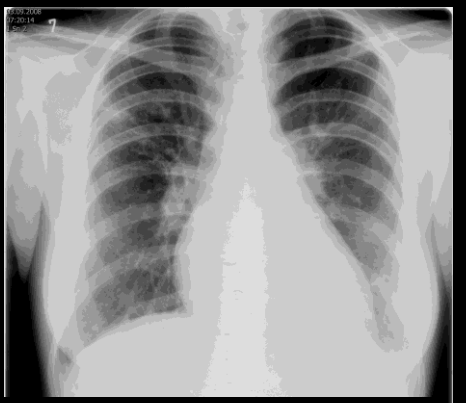
Lang et al. *Ann Am Thorac Soc* 2016 Vol 13, Supplement 3, pp S215–S221

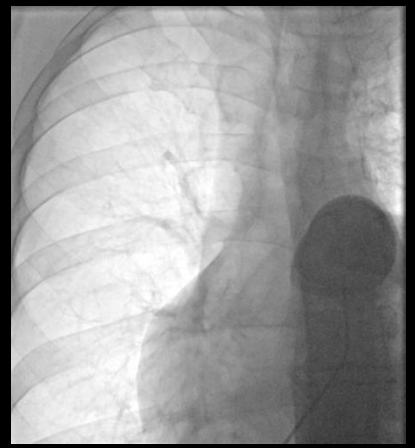
PLICNÍ ENDARTEREKTOMIE (PEA)

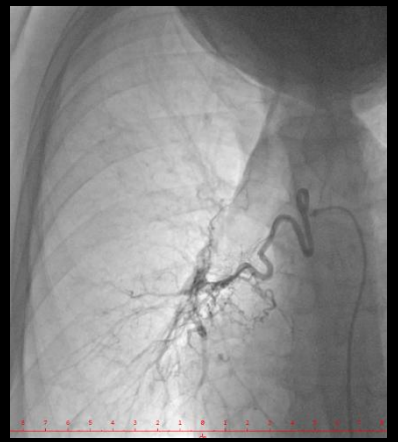


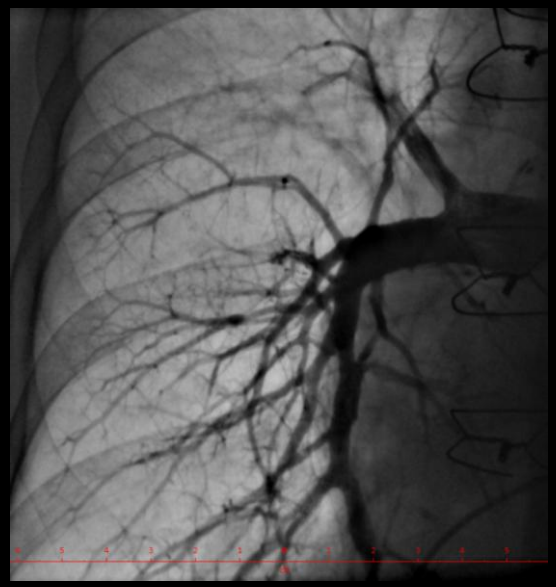


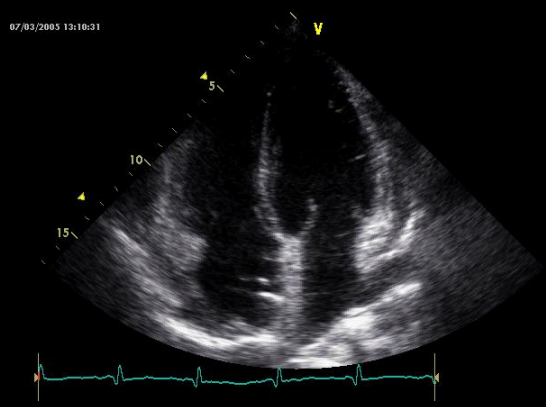


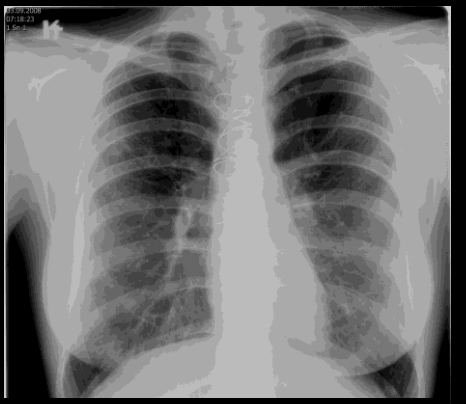






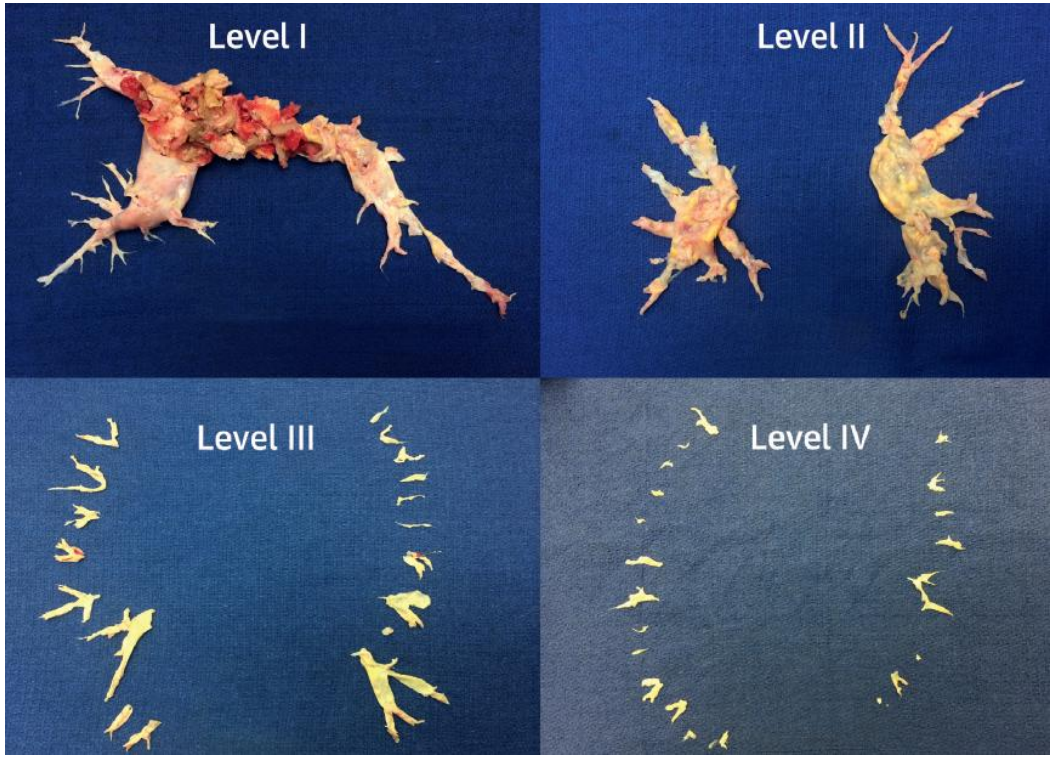


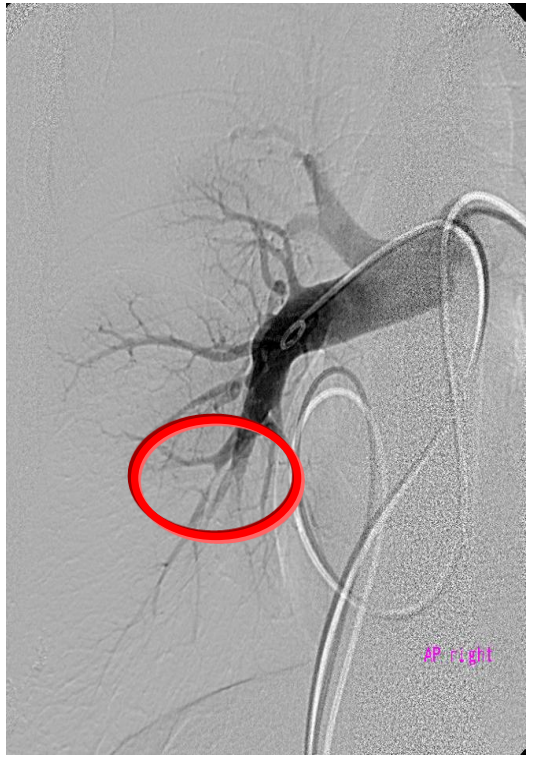


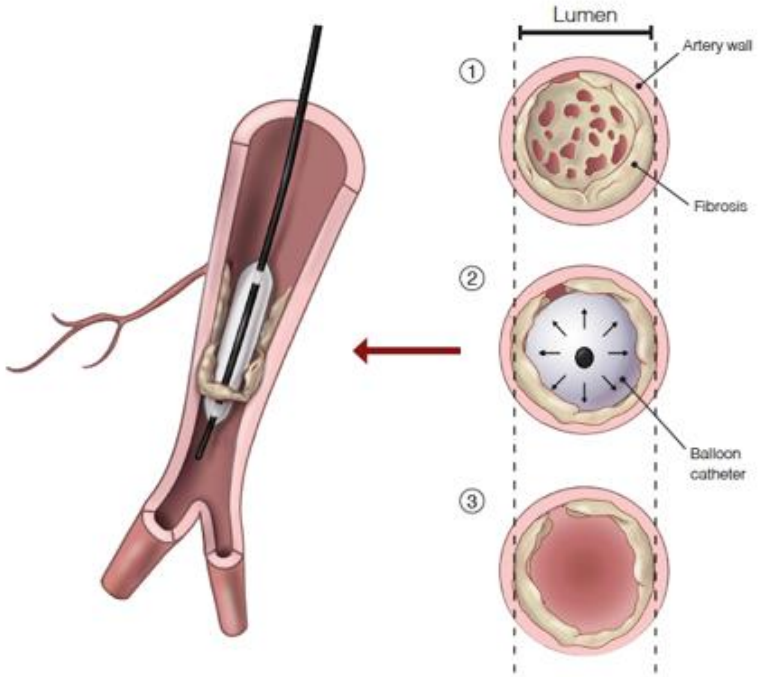


Zdroj: Všeobecná fakultní nemocnice v Praze

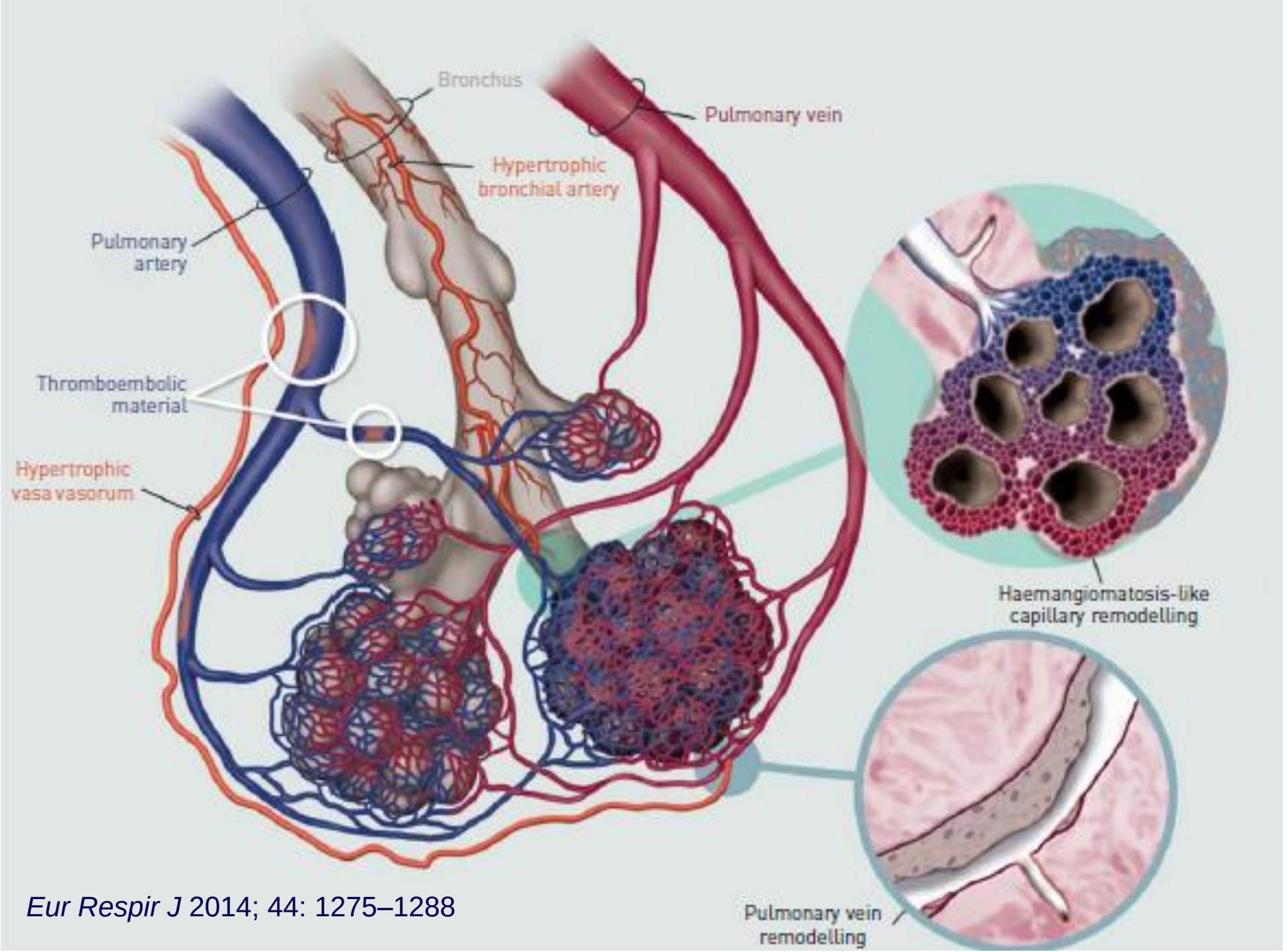
BALÓNKOVÁ PLICNÍ ANGIOPLASTIKA V LÉČBĚ CTEPH





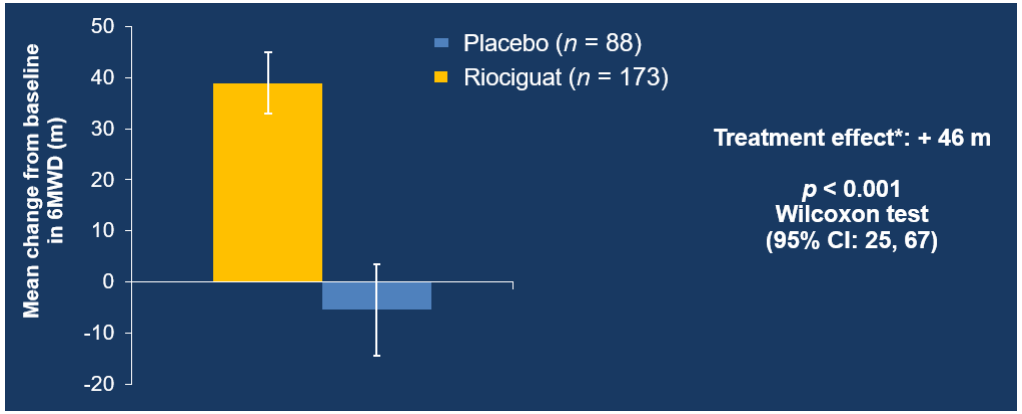


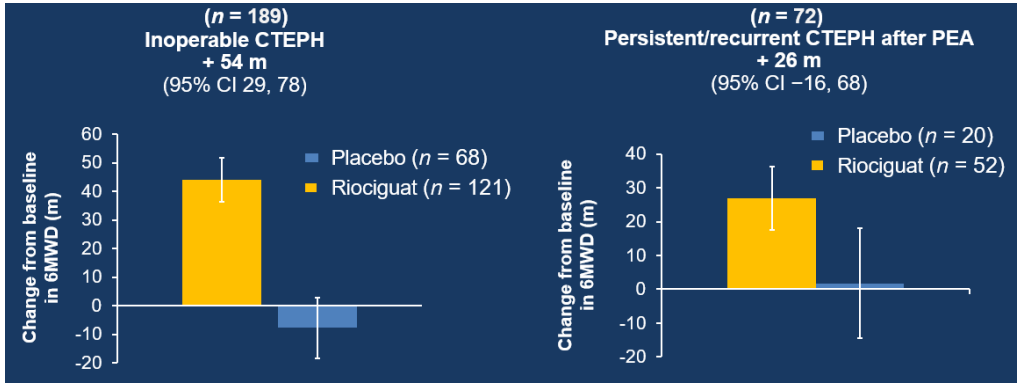
Mahmud et al. *JACC* 2018 Vol 71, 2468-8; Maren Hötten Master Thesis in Scientific Design, 2016; VFN



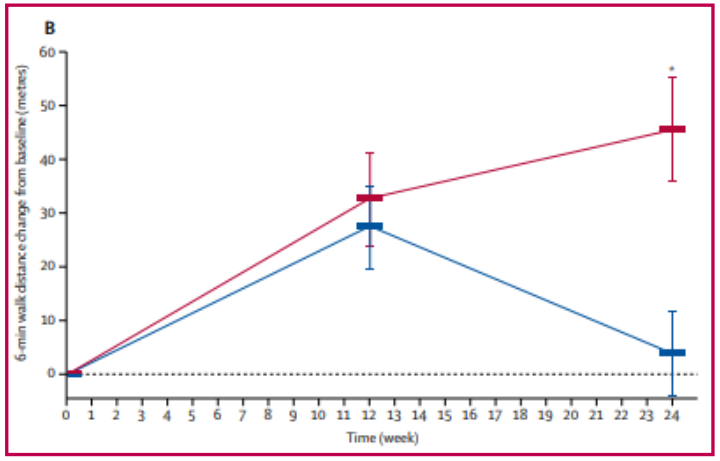
Eur Respir J 2014; 44: 1275–1288

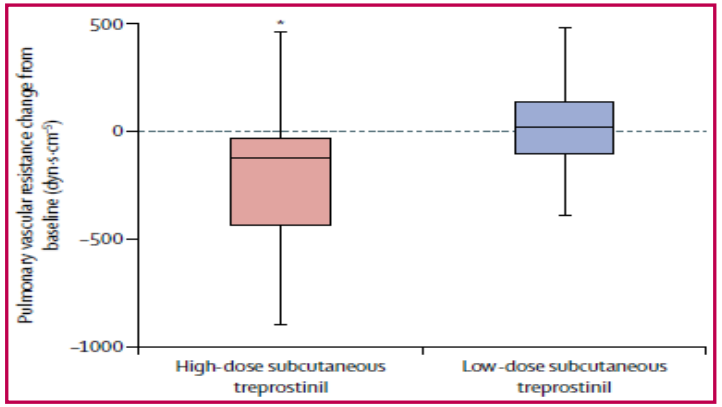
FARMAKOTERAPIE CTEPH





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

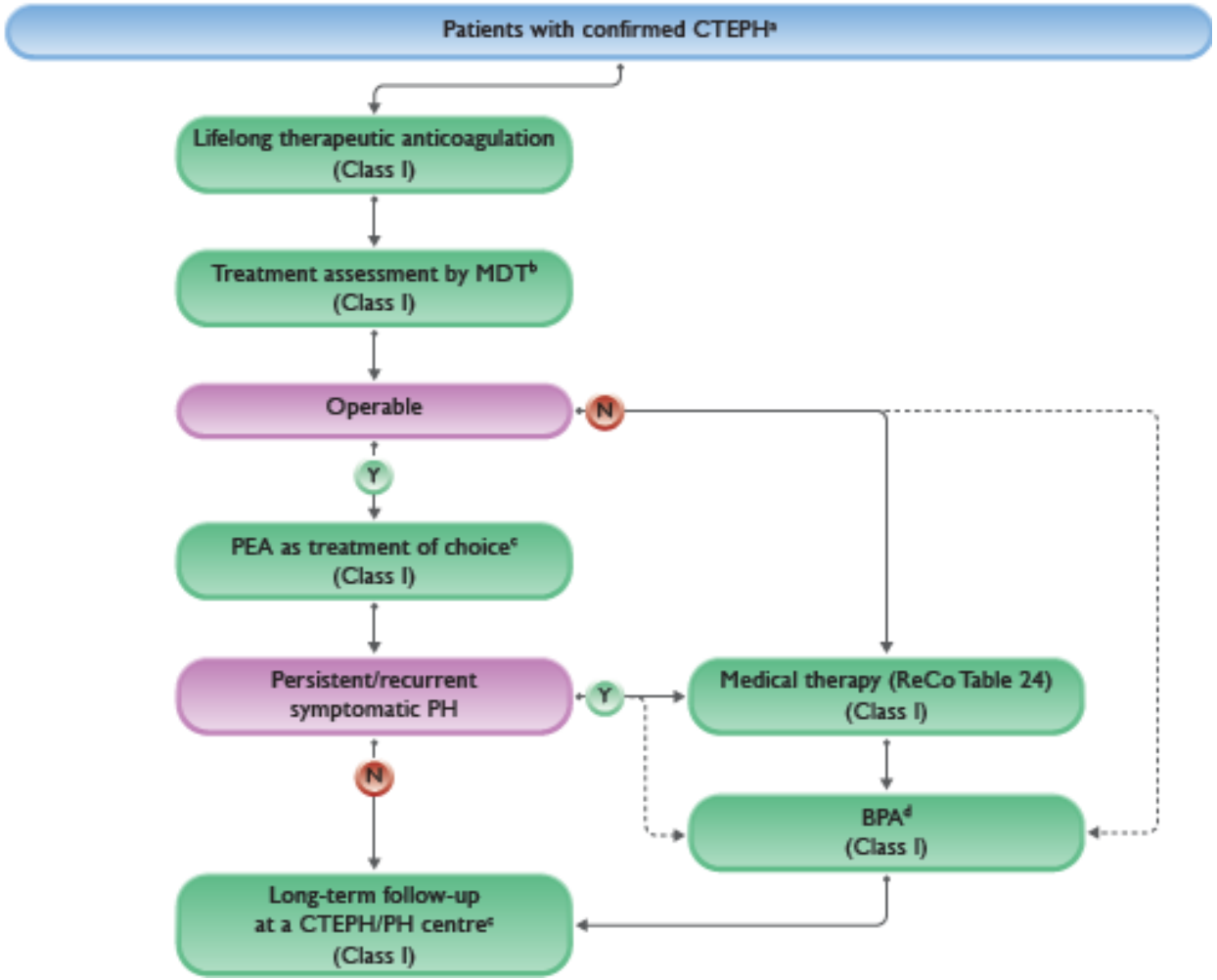


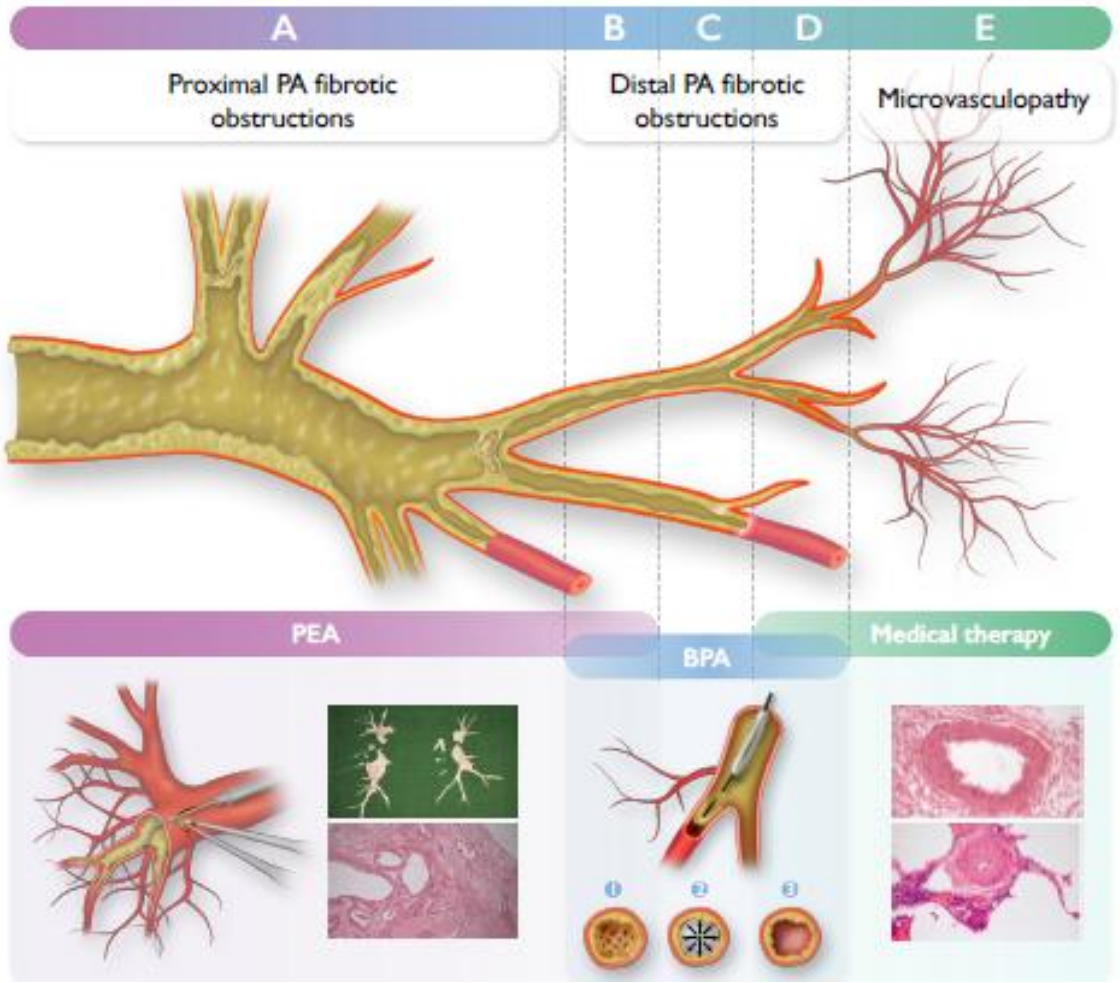


Studie CTREPH (treprostinil), n=105
Inoperabilní, perzistentní CTEPH, věk 64, 24 týdnů

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

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

Multidiscipinární CTEPH tým

- Chirurg se zkušeností s PEA
- Intervenční kardiolog se zkušeností s BPA
- Specialista na plicní hypertenzi
- Radiolog se zkušeností z vysokoobjemového CTEPH centra

Optimální počty výkonů

- PEA > 50 výkonů/rok
- BPA > 100 výkonů/rok nebo > 30 pacientů se zahájenou léčbou

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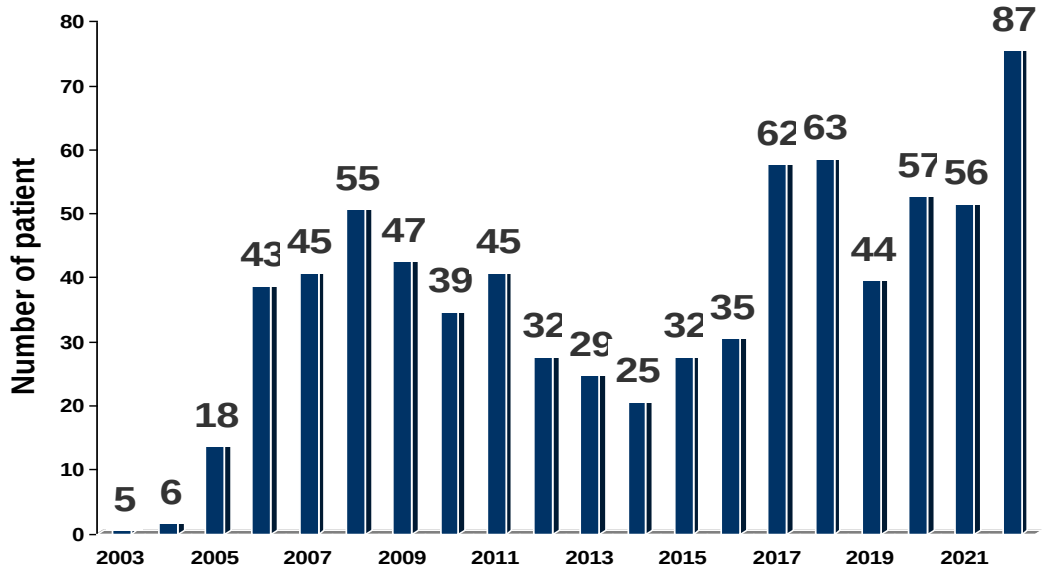
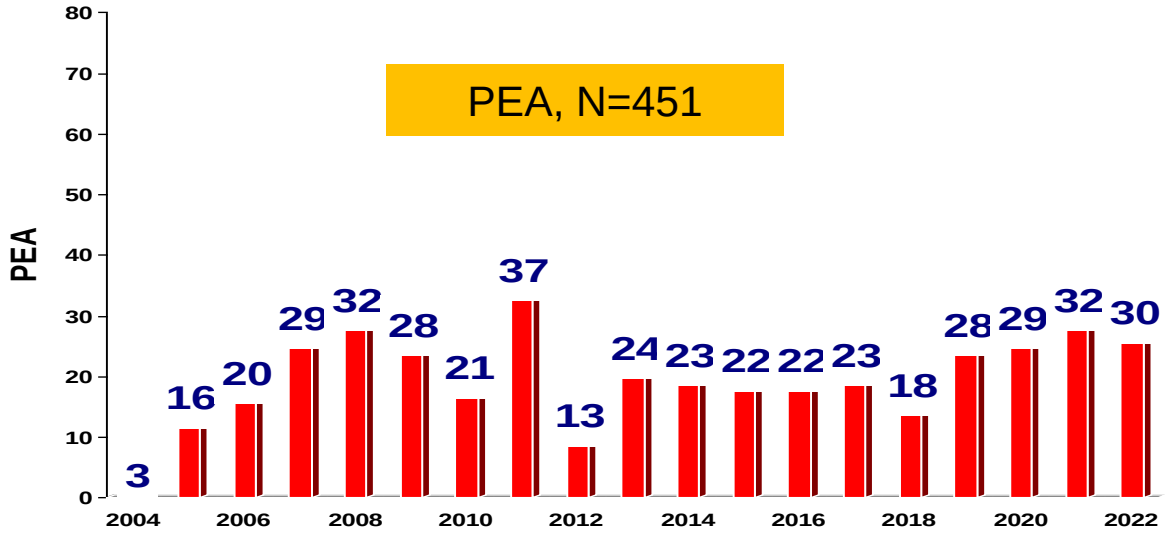
CTEPH V ČESKÉ REPUBLICE (2003-2022)

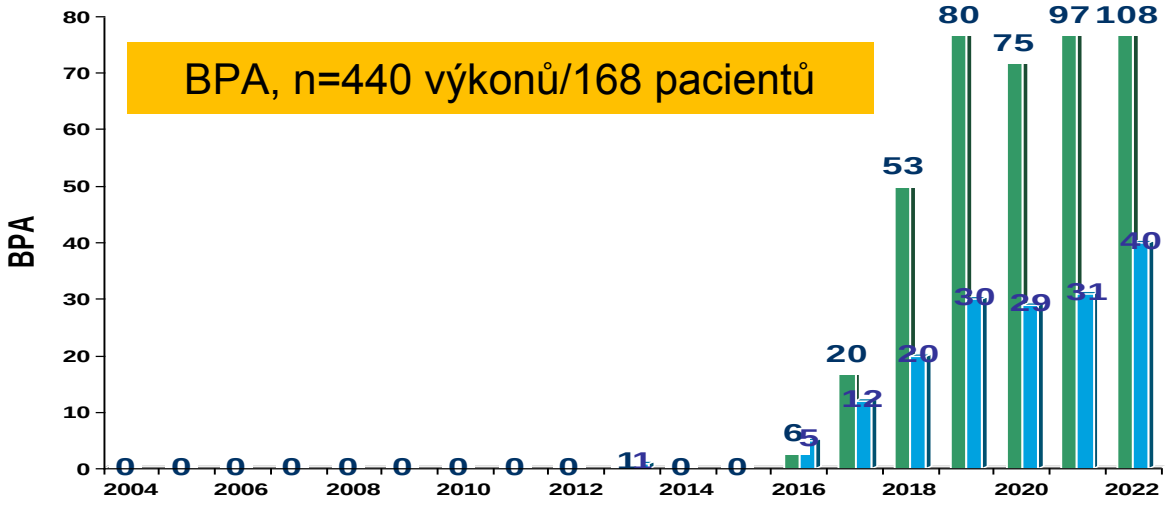
Nově diagnostikovaní pacienti (n=819)



PEA, N=451



BPA, n=440 výkonů/168 pacientů

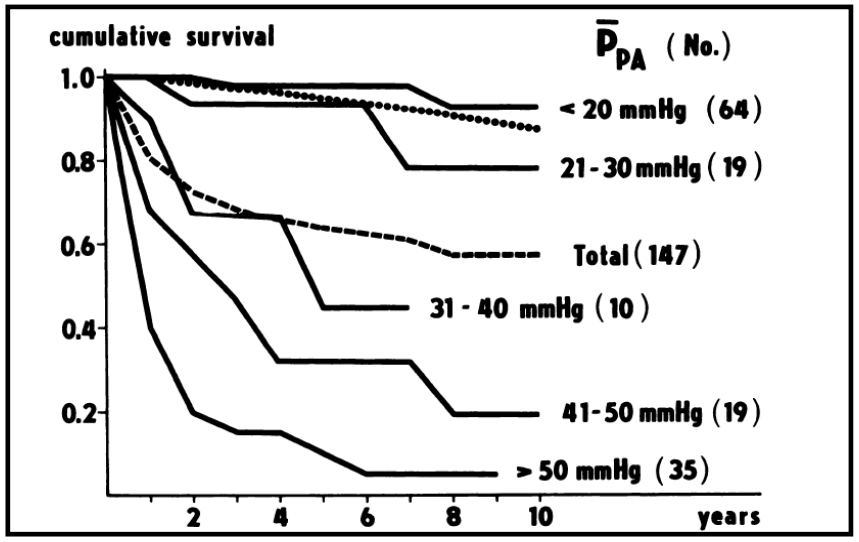


2017-2022:
nově diagnostikovaní 369.160 PEA, 162 BPA, 57 pouze farmakoterapie

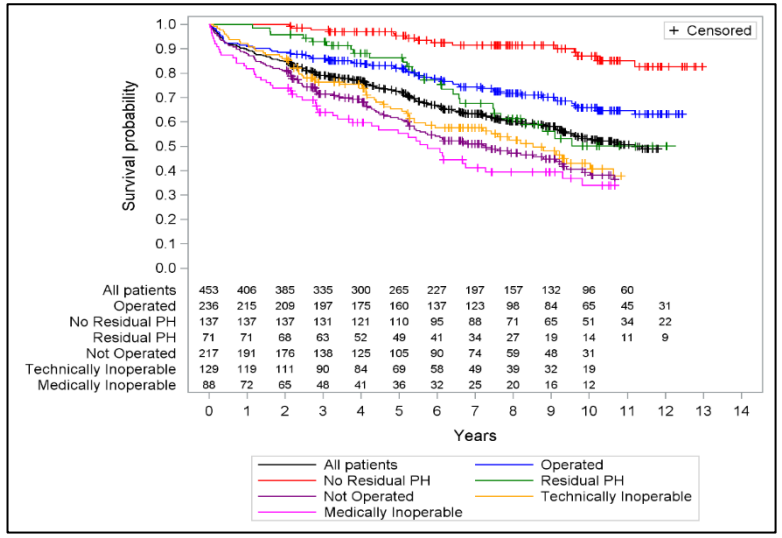
Jansa P et al. Epidemiology of chronic thromboembolic pulmonary hypertension (CTEPH) in the Czech Republic. *Pulmonary Circulation* 2022;12:e12038

„MULTIMODÁLNÍ“ LÉČBA CTEPH V ČR

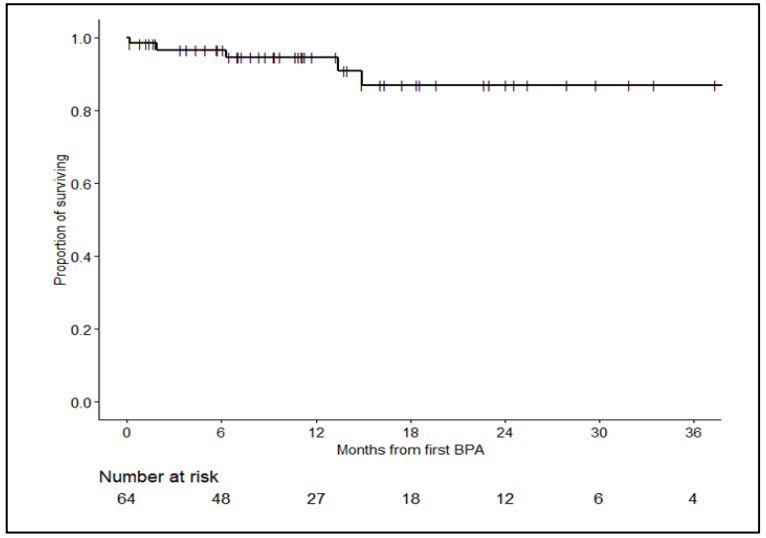
Pravděpodobnost přežití (%)
bez léčby CTEPH



Pravděpodobnost přežití (%)
2004-2016 (PEA)

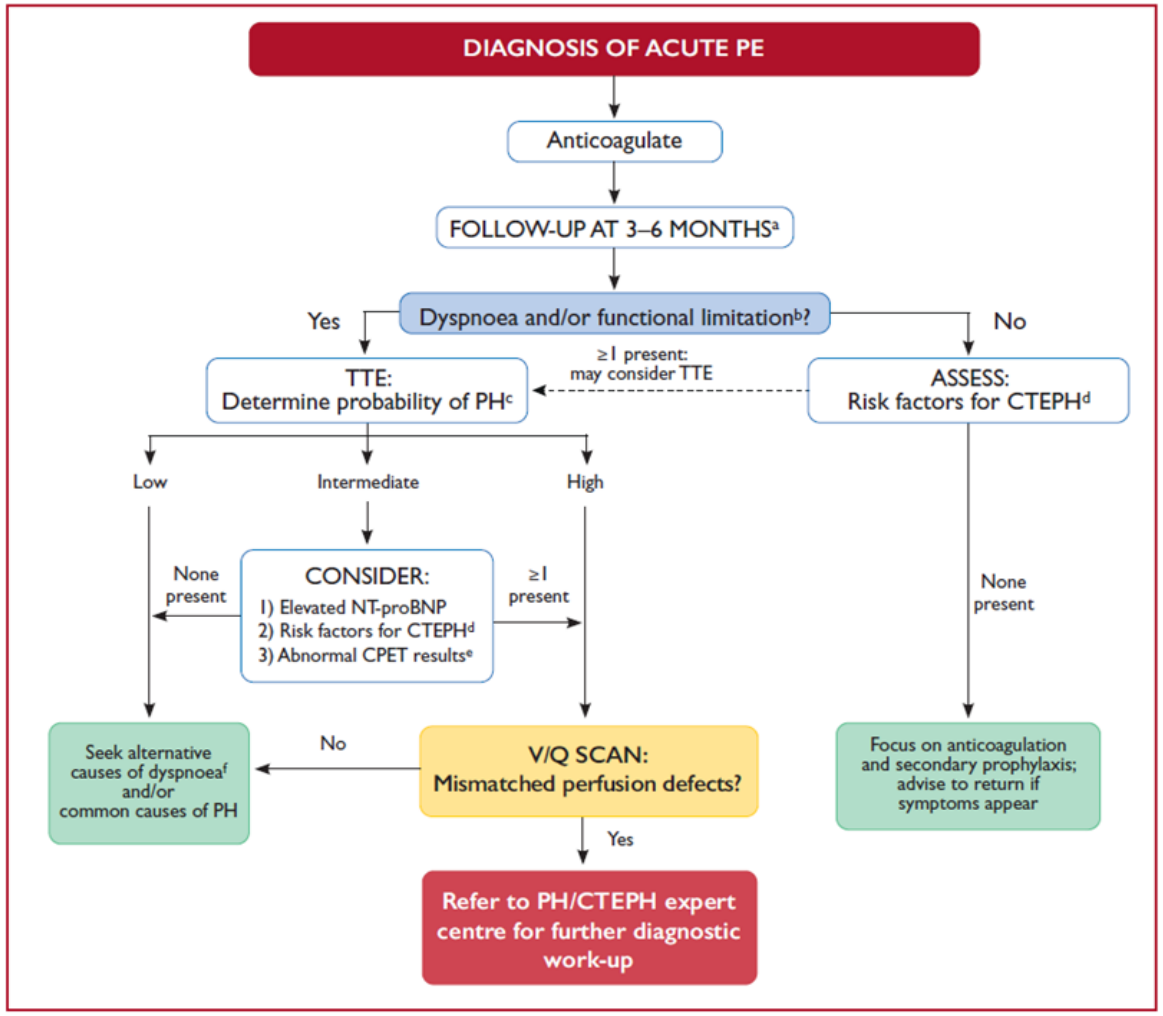


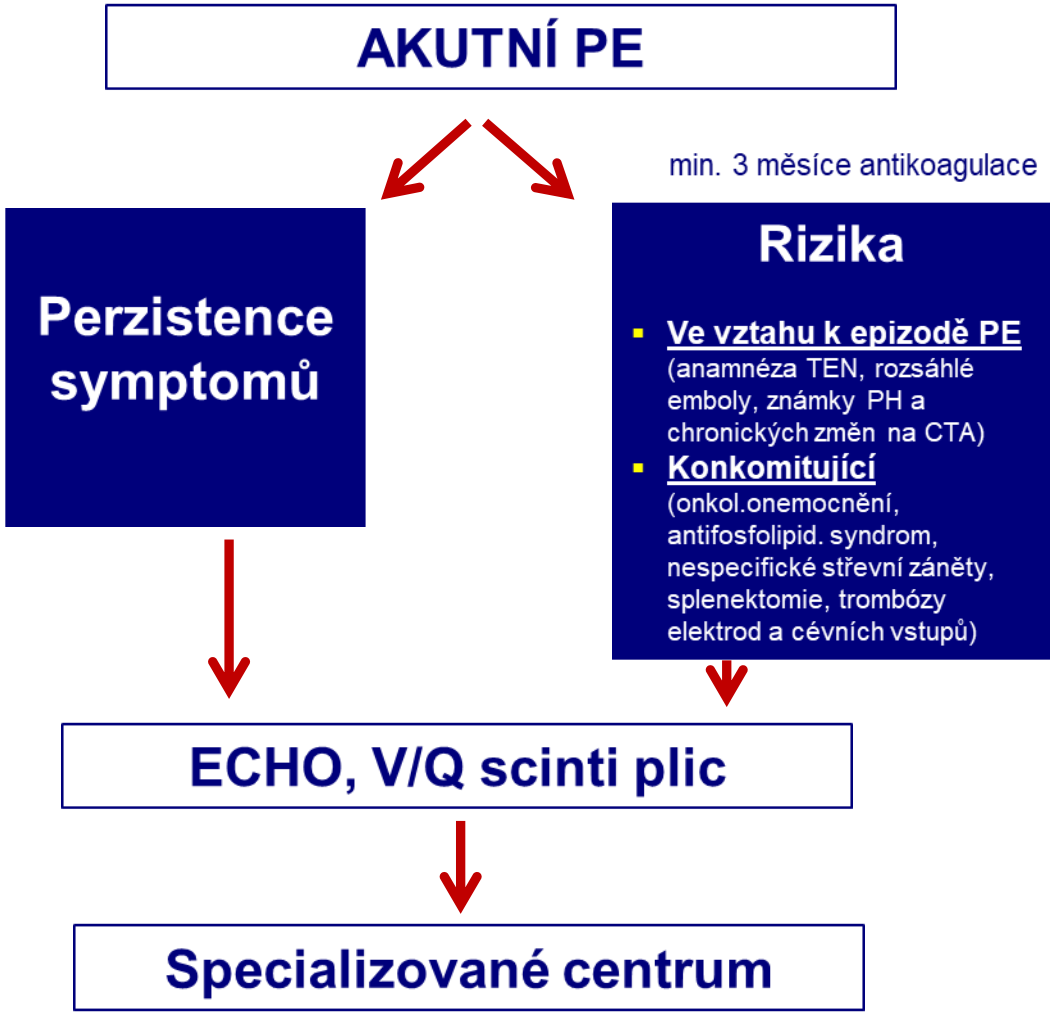
Pravděpodobnost přežití (%)
2016-2019 (BPA+farmakoterapie)



Riedel M, Stanek V, Widimsky J, Prerovsky I, *Chest* 1982, 81: 151-8
Jansa P et al. *Eur Respir J* 2020, 56: 1543; doi: 10.1183/13993003
Jansa P, Heller S et al. *J. Clin. Med*. 2020, 9, 3608; doi:10.3390/jcm9113608

ČASNÁ DIAGNÓZA CTEPH





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

- Volumová výzva k odhalení latentní postkapilární komponenty
- PH skupiny 2 s významnou prekapilární komponentou (PVR>5 WU) – specifická léčba?
- Těžká PH skupiny 3 (PVR>5 WU) – kombinovaná etioloogie? specifická léčba?
- Modifikovaná nomenklatura CTEPD/CTEPH
- Modifikovaný terapeutický algoritmus CTEPD/CTEPH (bez preference typu p.o. antikoagulace, multimodální léčba)
- Časná detekce CTEPD/CTEPH

Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022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Prof. MUDr. Michael Aschermann, DrSc.
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MUDr. Petr Kopecký
Kanylační centrum
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