

# KDY A JAK LÉČIT PLICNÍ HYPERTENZI ASOCIOVANOU S ONEMOCNĚNÍM PLIC

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European  
Reference  
Network

for rare or low prevalence  
complex diseases

• **Network**  
Respiratory Diseases  
(ERN-LUNG)

• **Member**  
General University  
Hospital in Prague –  
Czechia



# 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).

Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG).

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**Councils:** Council on Cardiovascular Genomics.

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**Patient Forum**

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2015

Definition	Characteristics <sup>a</sup>
PH	PAPm $\geq$ 25 mmHg
Pre-capillary PH	PAPm $\geq$ 25 mmHg PAWP $\leq$ 15 mmHg
Post-capillary PH	PAPm $\geq$ 25 mmHg PAWP >15 mmHg
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR $\leq$ 3 WU <sup>c</sup>
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG $\geq$ 7 mmHg and/or PVR >3 WU <sup>c</sup>

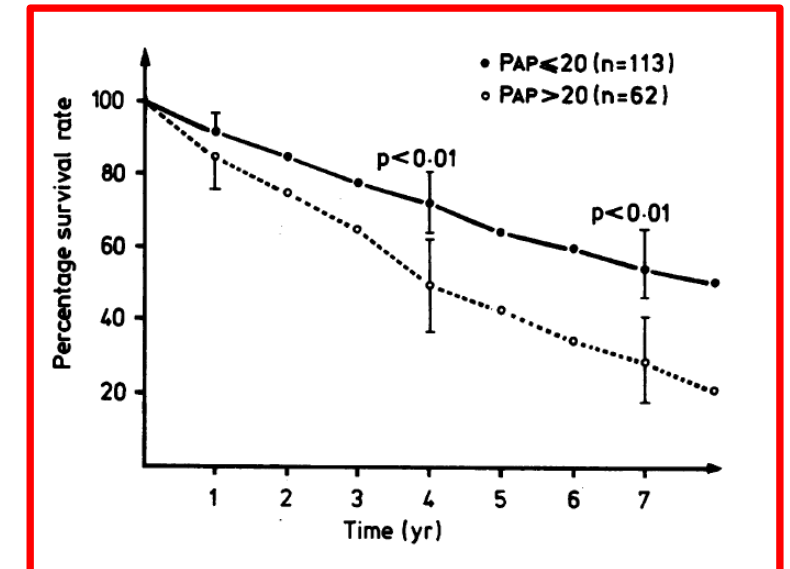
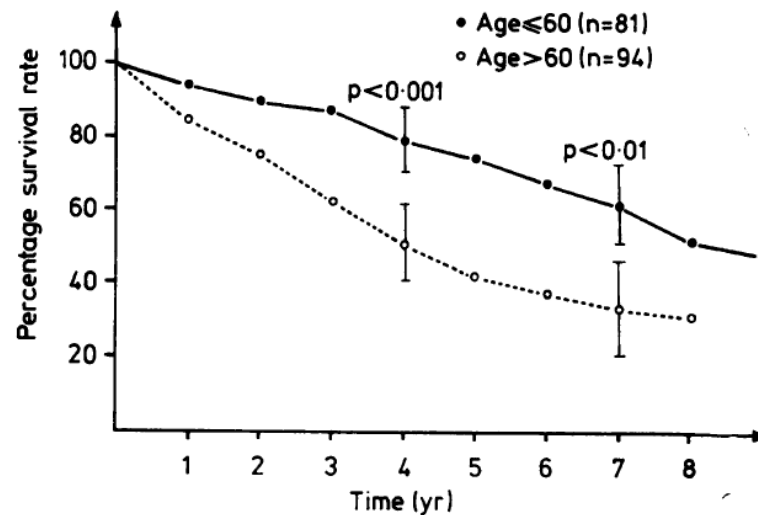
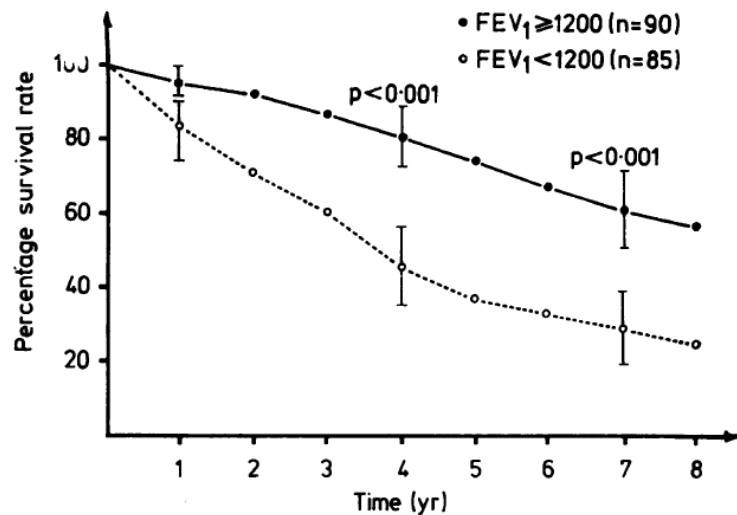
2022

Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP $\leq$ 15 mmHg PVR >2 WU
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR $\leq$ 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

# Prognostic value of pulmonary artery pressure in chronic obstructive pulmonary disease

E WEITZENBLUM, C HIRTH, A DUCOLONE, R MIRHOM,  
J RASAHOLINJANAHARY, M EHRHART

N=175, RHC 1968-1972, FEV<sub>1</sub> 40.2 ± 11.1 %, PAMP 19.8 ± 7.6 mmHg, CI 3.24 ± 0.93 L/min/m<sup>2</sup>  
Rekatetrizace: N=64 (po 5.5 letech)



# 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<sup>a</sup>
- 1.3 Associated with drugs and toxins<sup>a</sup>
- 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<sup>b</sup>
- 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<sup>c</sup>

## GROUP 5 PH with unclear and/or multifactorial mechanisms

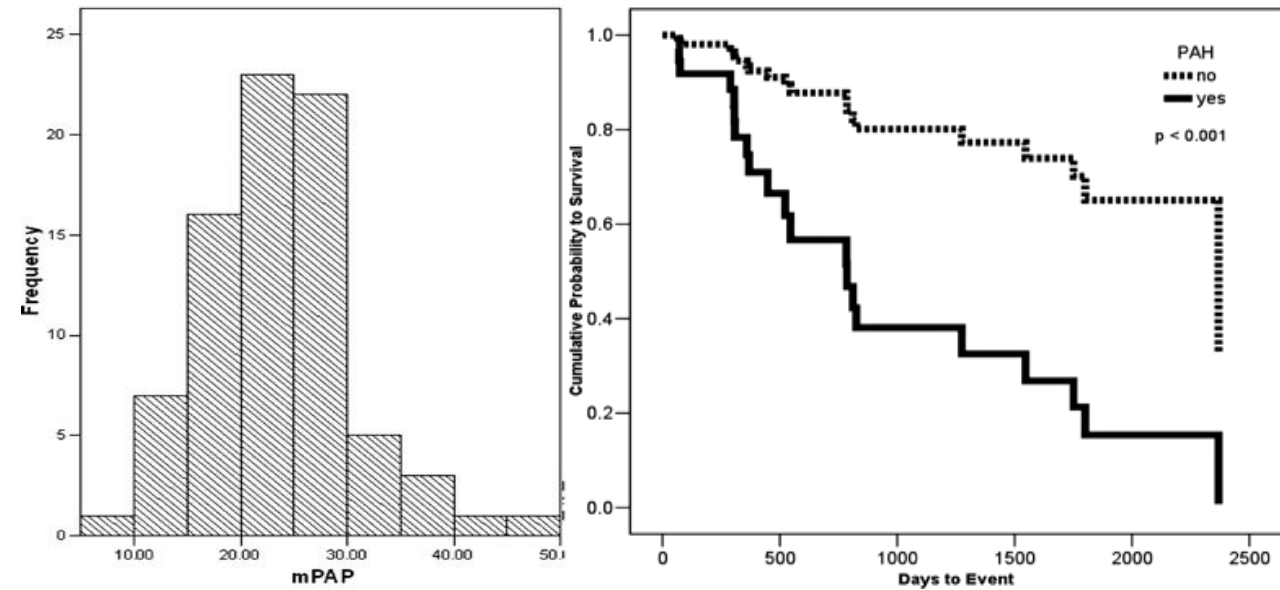
5 %

- 5.1 Haematological disorders<sup>d</sup>
- 5.2 Systemic disorders<sup>e</sup>
- 5.3 Metabolic disorders<sup>f</sup>
- 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*

# Prevalence and Outcomes of Pulmonary Arterial Hypertension in Advanced Idiopathic Pulmonary Fibrosis\*

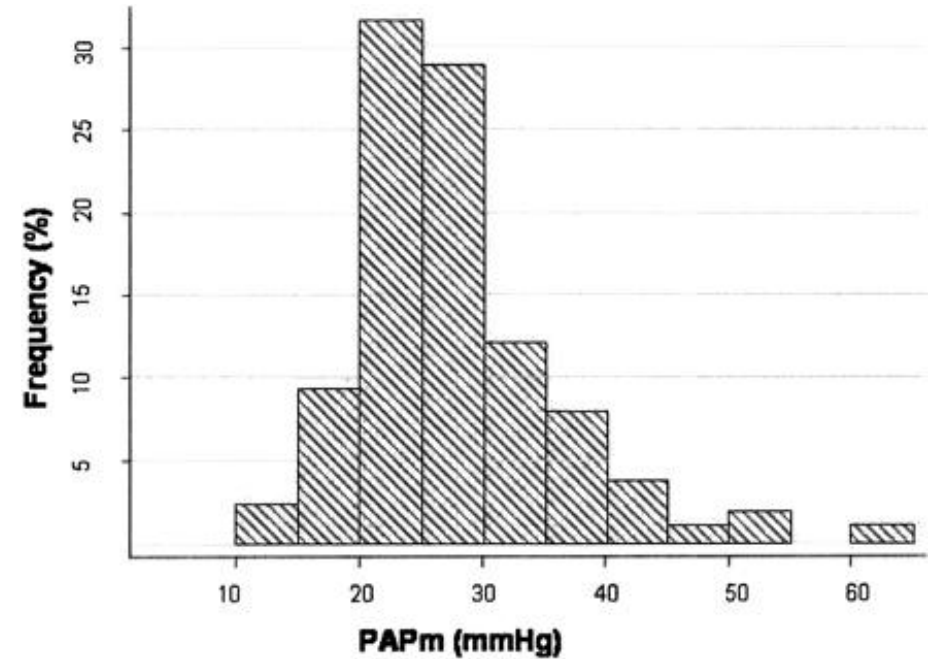
Christopher J. Lettieri, MD; Steven D. Nathan, MD, FCCP; Scott D. Barnett, PhD; Shahzad Ahmad, MD; and Andrew F. Shorr, MD, MPH, FCCP



n=79  
plicní hypertenze 31.6 % (PAMP 29.5 mmHg)

# Pulmonary Hemodynamics in Advanced COPD Candidates for Lung Volume Reduction Surgery or Lung Transplantation\*

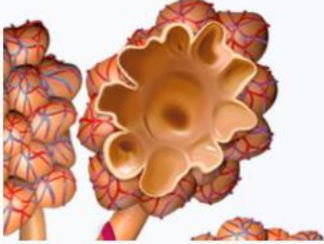
Gabriel Thabut, MD; Gaëlle Dauriat, MD; Jean Baptiste Stern, MD; Damien Logeart, MD; Antoine Lévy, MD; Rolana Marrash-Chahla, MD; and Hervé Mal, MD



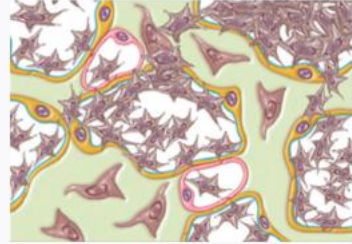
n=247  
PAMP (mmHg): 50 % > 25, 12 % > 35, 5 % > 40 mmHg

# PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

Emphysema



Fibrosis



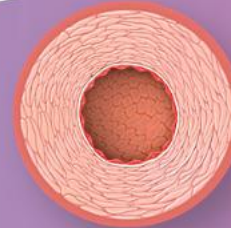
Vascular pruning



Remodelling of airways and parenchyma



Remodelling of pulmonary vessels



No PH

Non-severe PH

Severe PH  
(PVR >5 WU)

Prevalence

~70%

~20%

~5-10%

Mostly ventilatory  
exercise limitation

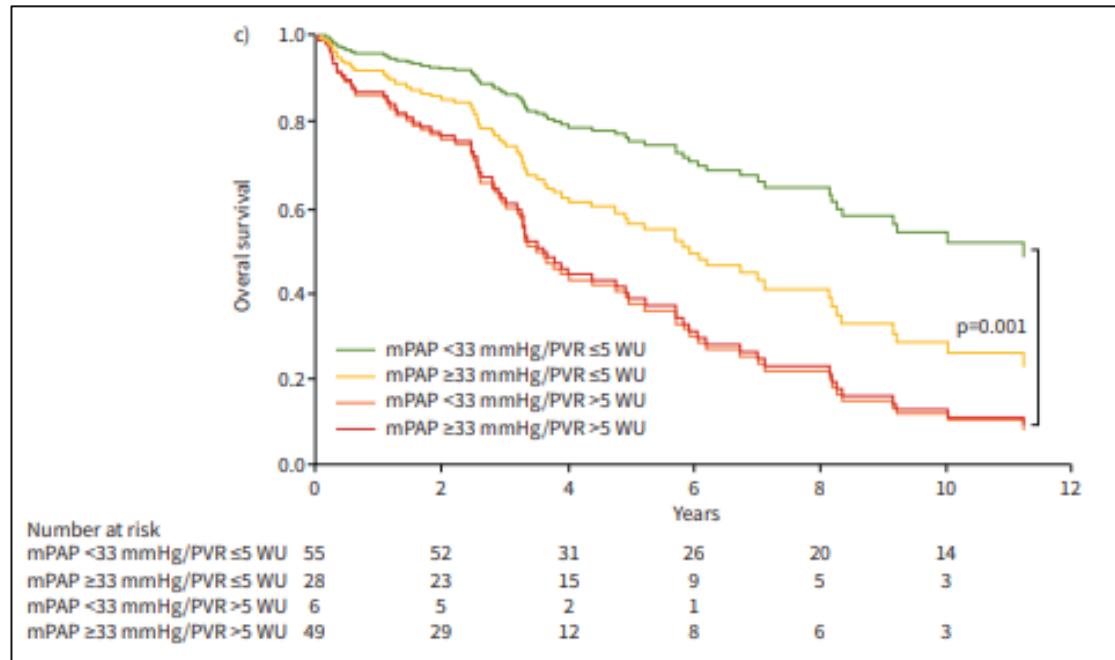
Mostly circulatory  
exercise limitation

Hypoxaemia at rest and/or during exercise

**Těžká PH**  
PVR > 5 WU

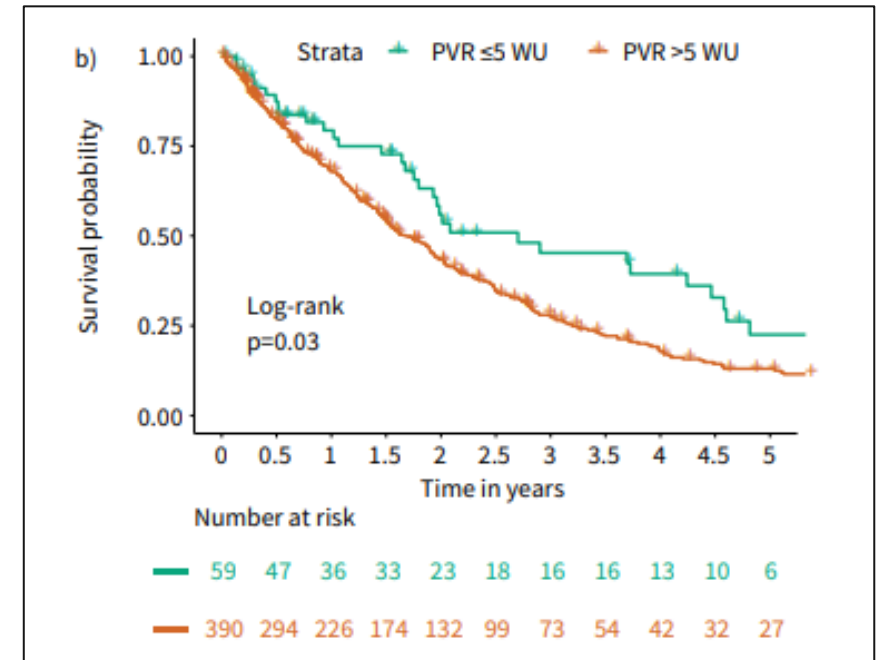
- Bez vztahu k plicním objemům
- Hypoxémie
- Nízká DLco

## 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



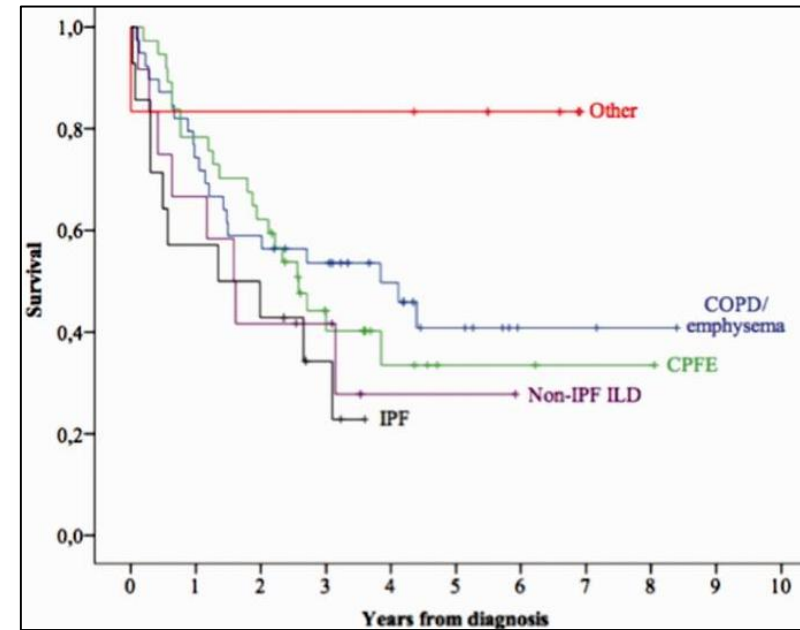
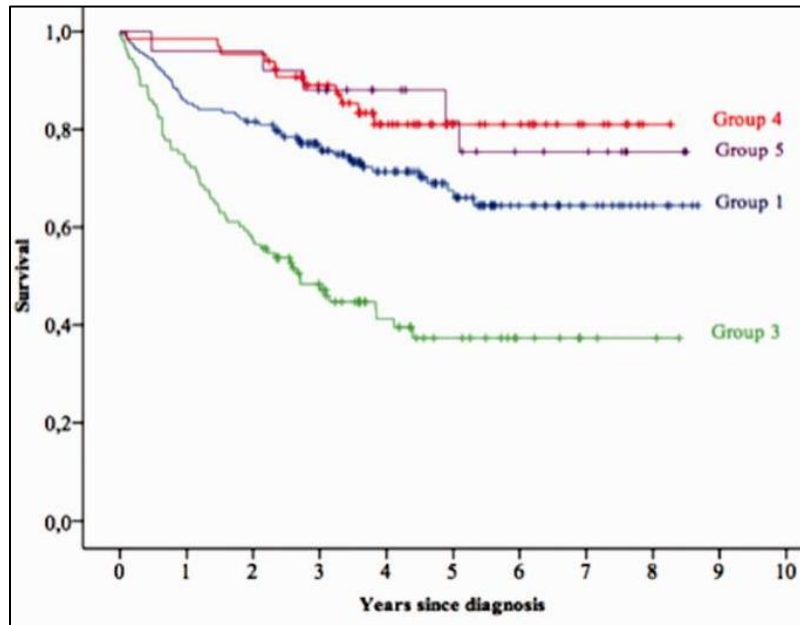
N=449, věk 73, 65 % mužů  
 PAMP 39 (33, 46) mmHg, PVR 7.6 (6.0, 10.6) WU,  
 TLC 72 (60, 86) %, FEV1 69 (55, 81) %



# Pulmonary hypertension in chronic lung diseases: comparison to other pulmonary hypertension groups

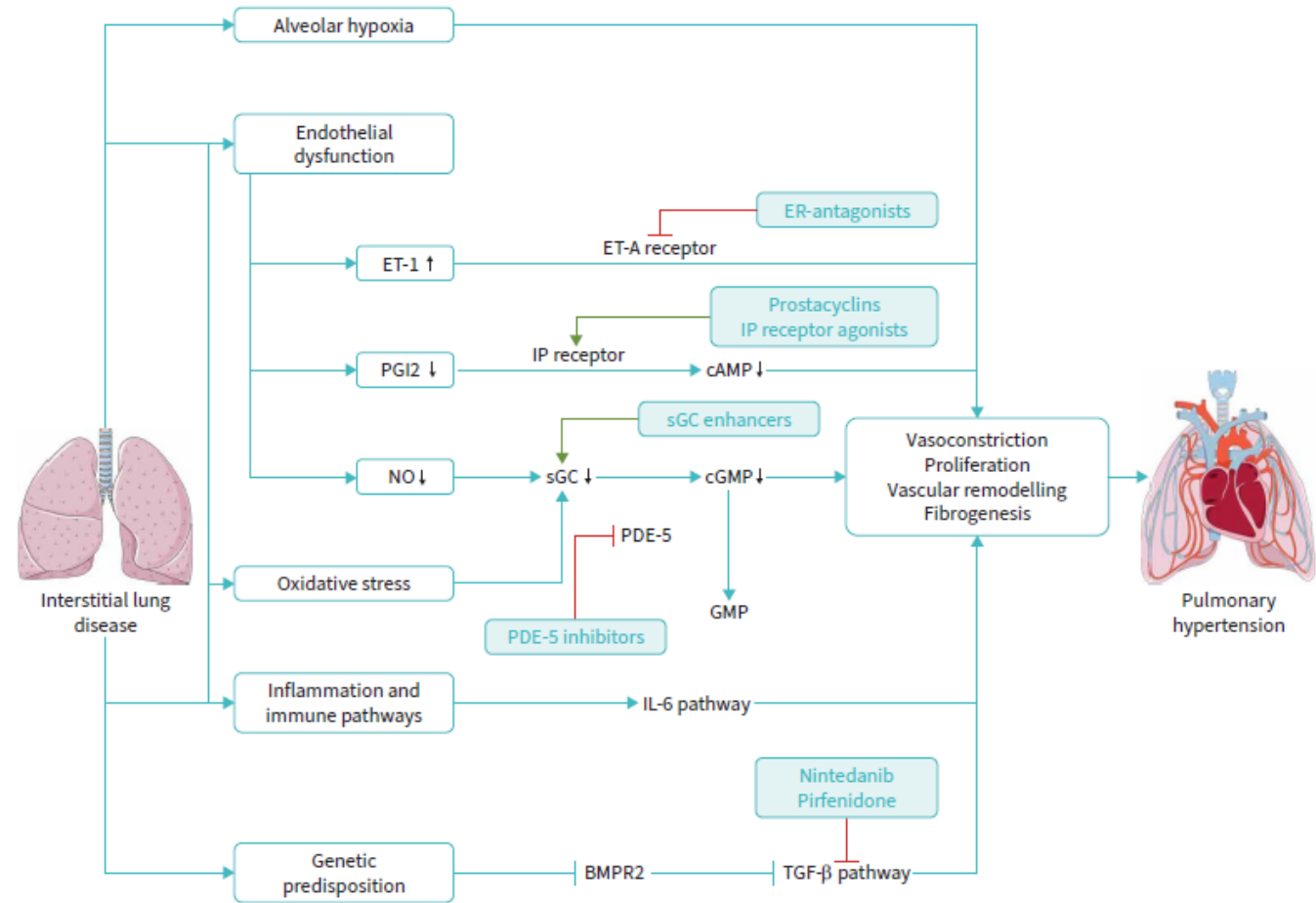
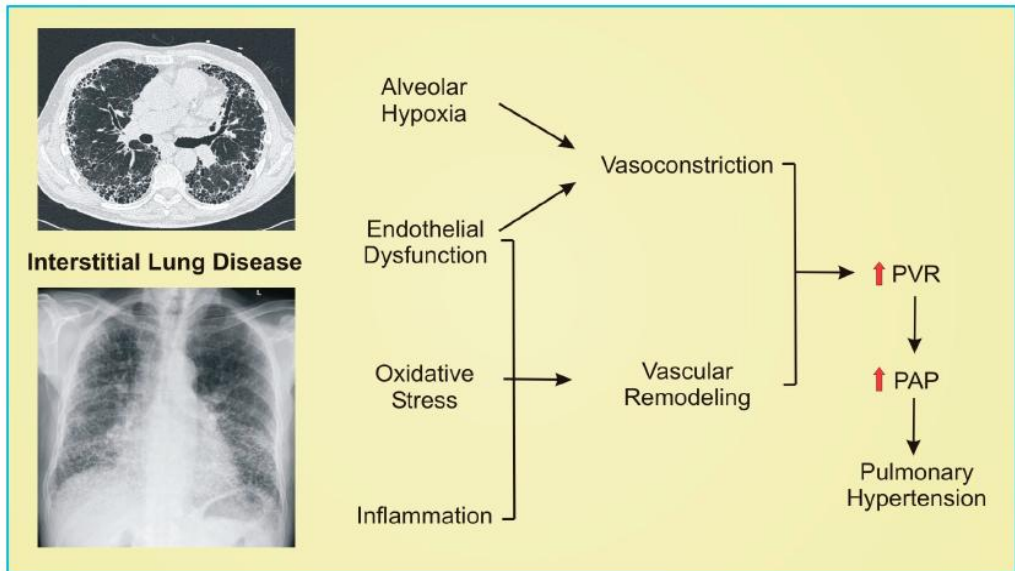
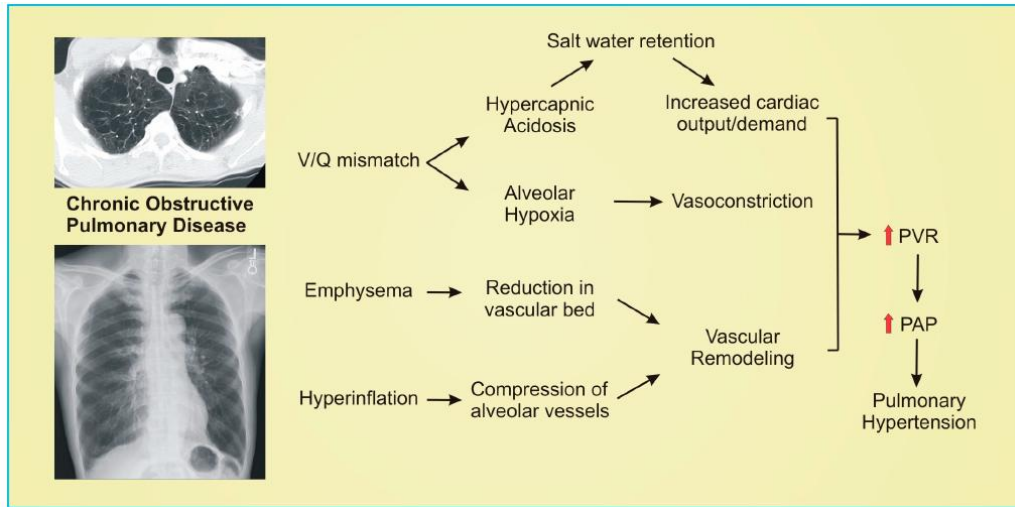
N=363, 2006-2014

Skupina 1 164 (45.2%), skupina 3 109 (30%), skupina 4 65 (17.9%), skupina 5 25 (6.9%)



	Overall	Group 1	Group 3	Group 4	Group 5	P <sup>§</sup>
Pericardial effusion (n (%))	57 (15.7)	26 (15.8)	18 (16.5)	8 (12.3)	5 (20)	0.82
mPAP <sup>†</sup> (mmHg)	40 (25–83)	38 (25–83)	41 (26–72)	42 (26–68)	35 (26–63)	0.01
PCWP <sup>†</sup> (mmHg)	9 (1–15)	9 (1–15)	10 (2–15)	8 (1–15)	11 (3–15)	0.13
PVR <sup>†</sup> (WU)	9.1 (3.3–45.5)	8.5 (3.3–28.5)	9.7 (3.9–23.1)	10.3 (4.5–45.5)	7.4 (3.9–17.9)	0.001
CI <sup>†</sup> (L/min/m <sup>2</sup> )	2.4 (0.8–5.6)	2.6 (1.1–5.2)	2.4 (1.1–4)	2.2 (0.8–3.7)	3 (1.7–5.6)	0.001
RAP <sup>†</sup> (mmHg)	6 (0–27)	6 (0–27)	6 (1–21)	5 (0–21)	6 (0–16)	0.54
SvO <sub>2</sub> <sup>†</sup> (%)	64 (25–94)	66 (33–93)	61 (40–93)	61 (25–94)	65 (34–89)	<0.001

# PATOFYZIOLOGIE PH U PLICNÍCH ONEMOCNĚNÍ



PVR ≤ 5WU

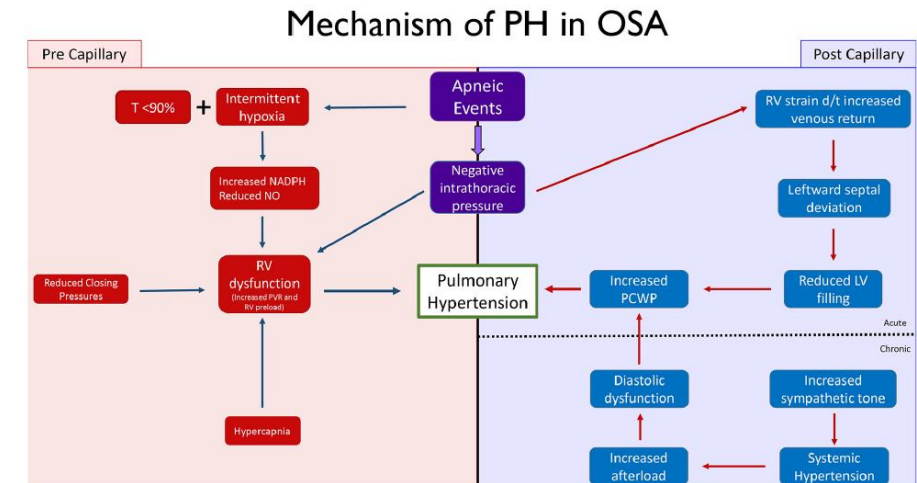
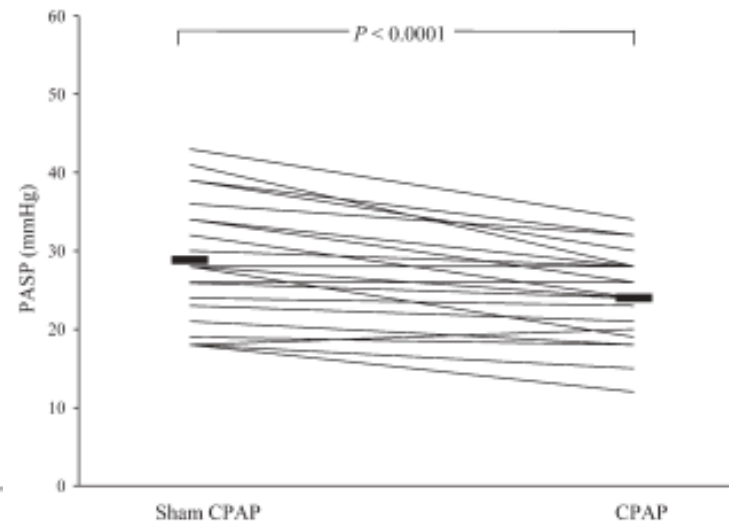
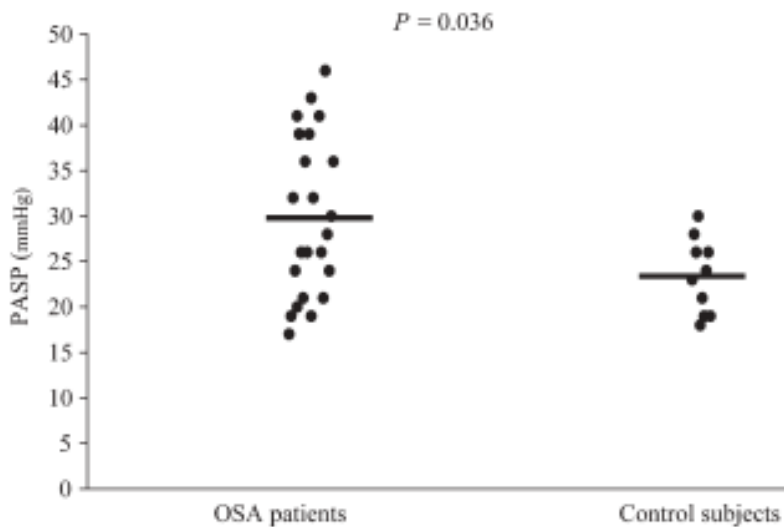
PVR > 5WU

**Léčba:** Léčba základního plicního onemocnění , DDOT pokud indikována , CPAP pokud indikován, plicní RHB programy, Zvážení LuTx  
 U kombinované etiologie PH adekvátní léčba, specifická vazodilatační léčba?

## Pulmonary hypertension in obstructive sleep apnoea: effects of continuous positive airway pressure

A randomized, controlled cross-over study

23 pacientů s OSA (AHI 44.1 + 29.3), 10 kontrol  
 Randomizace: 12 týdnů účinná vs předstíraná léčba přetlakem, neinvazivní odhad tlaků v plicnici  
 Efekt významnější u pacientů s diastolickou dysfunkcí



# PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

**Léčba:** Léčba základního plicního onemocnění , DDOT pokud indikována , CPAP pokud indikován, plicní RHB programy, Zvážení LuTx  
U kombinované etiologie PH adekvátní léčba, specifická vazodilatační léčba?

Trial Study	Therapy	Target	Outcome
Chronic Obstructive Pulmonary Disease-Associated Pulmonary Hypertension (COPD-PH)			
Blanco et al., 2010	Sildenafil	NO	Reduced mean PAP
Blanco et al., 2013	Sildenafil and pulmonary rehabilitation	NO	No improvement in cycle endurance time
Goudie et al., 2014	Tadalafil	NO	No improvement in 6MWD
SPHERIC-1 Vitulo et al., 2017	Sildenafil	NO	Reduced PVR
Stolz et al., 2008	Bosentan	ET-1	No improvement in 6MWD
Valerio et al., 2009	Bosentan	ET-1	Reduced mean PAP and PVR, Increased 6MWD, and reduced BODE index

# PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

**Léčba:** Léčba základního plicního onemocnění , DDOT pokud indikována , CPAP pokud indikován, plicní RHB programy, Zvážení LuTx  
U kombinované etiologie PH adekvátní léčba, specifická vazodilatační léčba?

Trial Study	Therapy	Target	Outcome
<b>Pulmonary Fibrosis-Associated Pulmonary Hypertension (PF-PH)</b>			
STEP-IPF Zisman et al., 2010	Sildenafil	NO	No improvement in 6MWD
Behr et al., 2021	Sildenafil and Pirfenidone	NO	No improvement in 6MWD, respiratory hospitalization, or mortality
RISE-IIP Nathan et al., 2019	Riociguat	NO	No improvement in 6MWD; increased adverse events and mortality
iNO-PF Nathan et al., 2020	Pulsed inhaled NO	NO	Increased moderate/vigorous physical activity
ARTEMIS-IPF Raghu et al., 2013	Ambrisentan	ET-1	No improvement in lung function, respiratory hospitalization, or death; Increased harm
BPHIT Corte et al., 2014	Bosentan	ET-1	No decrease to PVR index of 20% or more
INCREASE Waxman et al., 2021	Inhaled Treprostinil	Prostacyclin	Improvement in 6MWD

# PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

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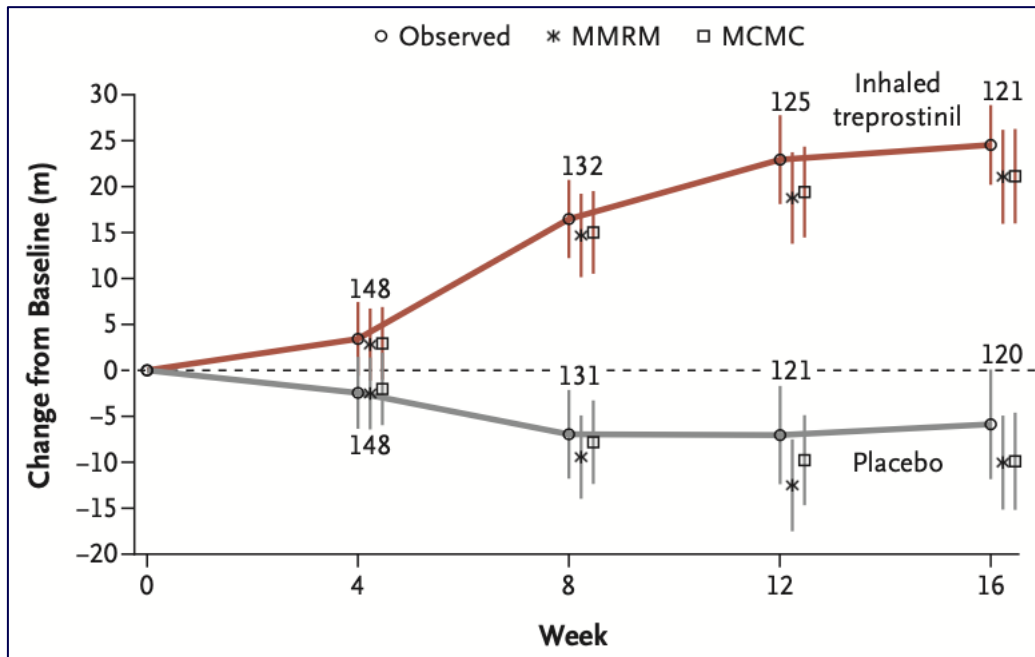
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, *EHJ* 2022, *ERJ* 2022

Gonzales J et al. Pharmacology and Emerging Therapies for Group 3 Pulmonary Hypertension Due to Chronic Lung Disease. *Pharmaceuticals* 2023, 16, 418

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 %, 68 (21 %) předčasně ukončilo studii



End Point	Inhaled Treprostinil (N=163)	Placebo (N=163)	Treatment Effect (95% CI)	P Value
<b>Primary end point</b>				
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
<b>Secondary end points§</b>				
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††

# 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

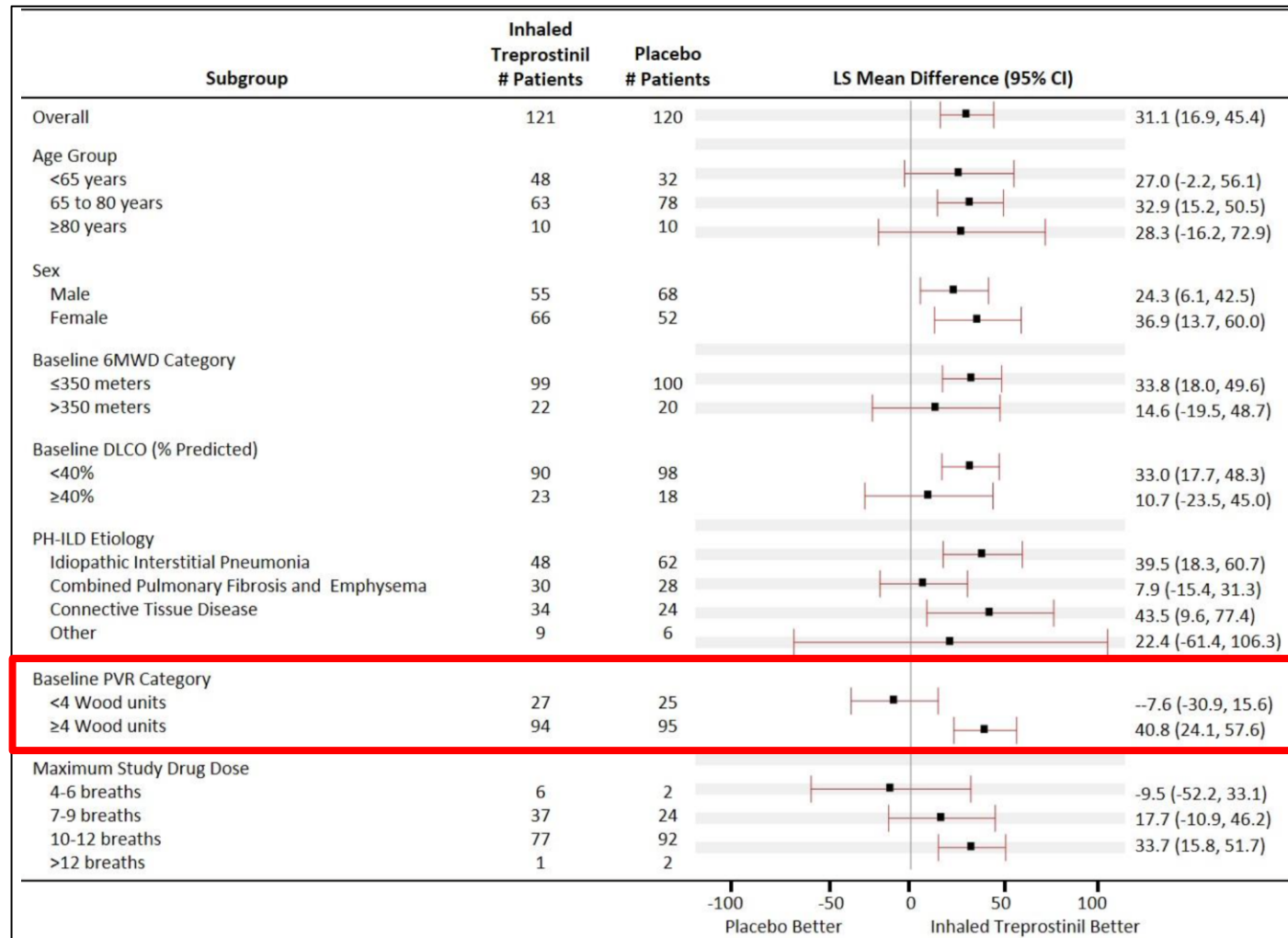
	Inhaled Treprostinil (N=163)	Placebo (N=163)	All Patients (N=326)
6-minute walk distance, meters; mean (range)	254.1 (100-538)	265.1 (30-505)	259.6 (30-538)
Median	256.0	260.0	259.0
Pulmonary vascular resistance, Woods units; mean (range)	6.369 (3.11-18.05)	6.013 (3.06-17.62)	6.191 (3.06-18.05)
Median	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 <sub>1</sub> % 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	



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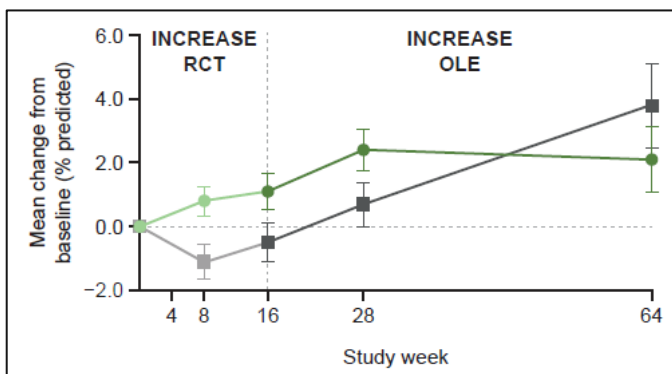
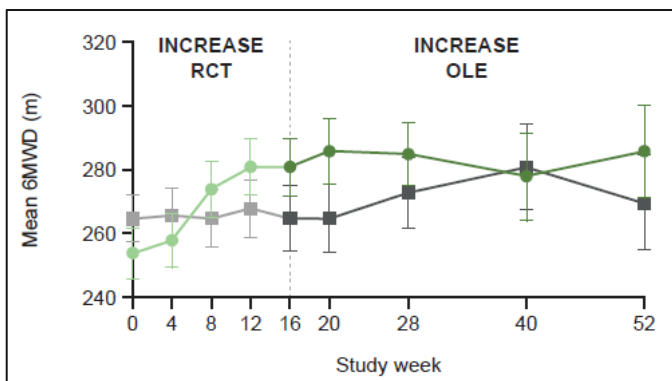
# Long-term inhaled treprostinil for PH-ILD: INCREASE open-label extension study

108 týdnů po skončení DB, N=242

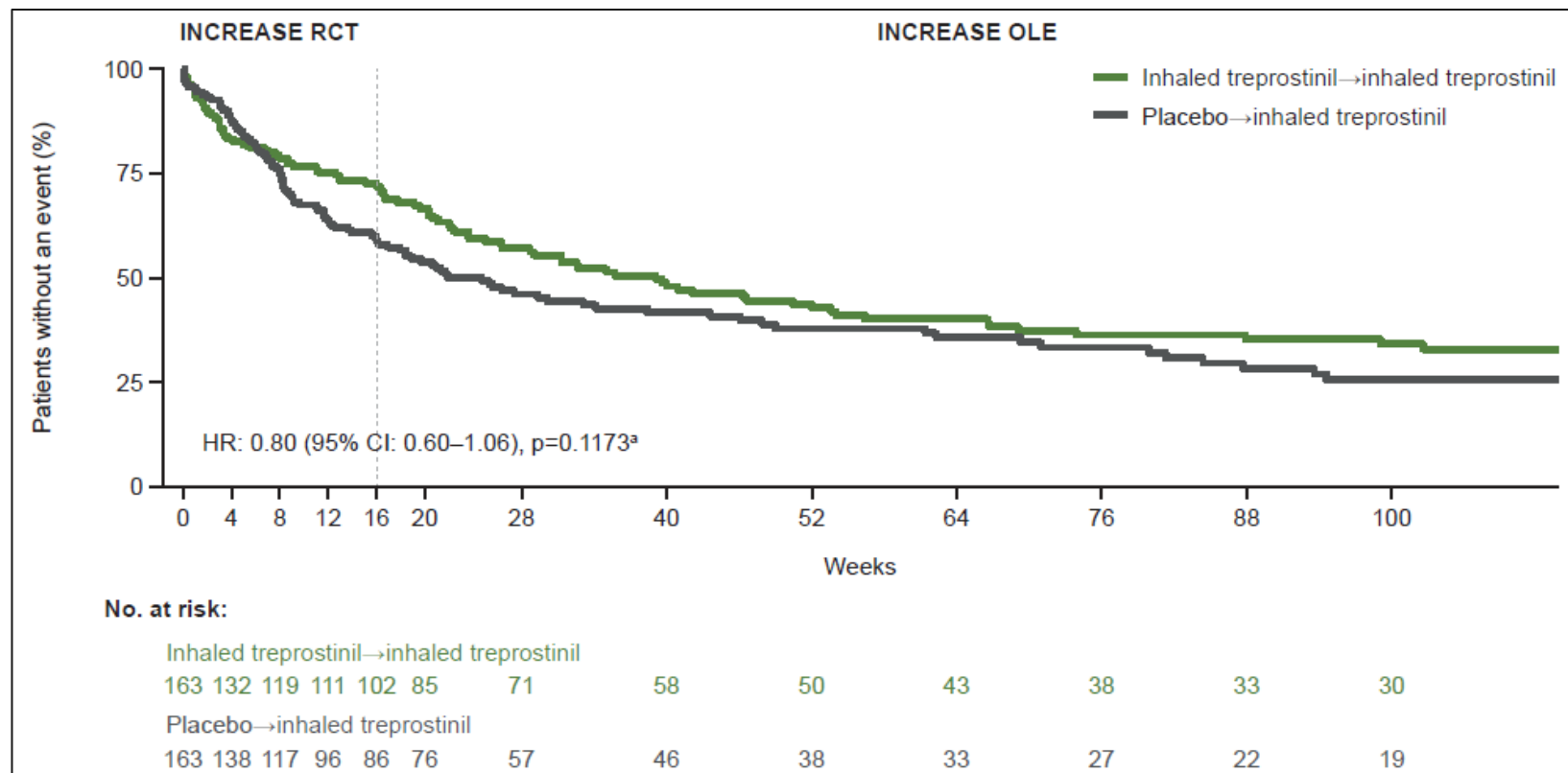
**Předčasné ukončení: 172 (71 %)**

úmrtí (n=56), rozhodnutí pacienta (n=41), nežádoucí účinky (n=29), ostatní (n=46)

6MWT (m)



FVC (%)



Čas do exacerbace (týdny)

# Pulmonary Hypertension in Patients with Chronic Fibrosing Idiopathic Interstitial Pneumonias

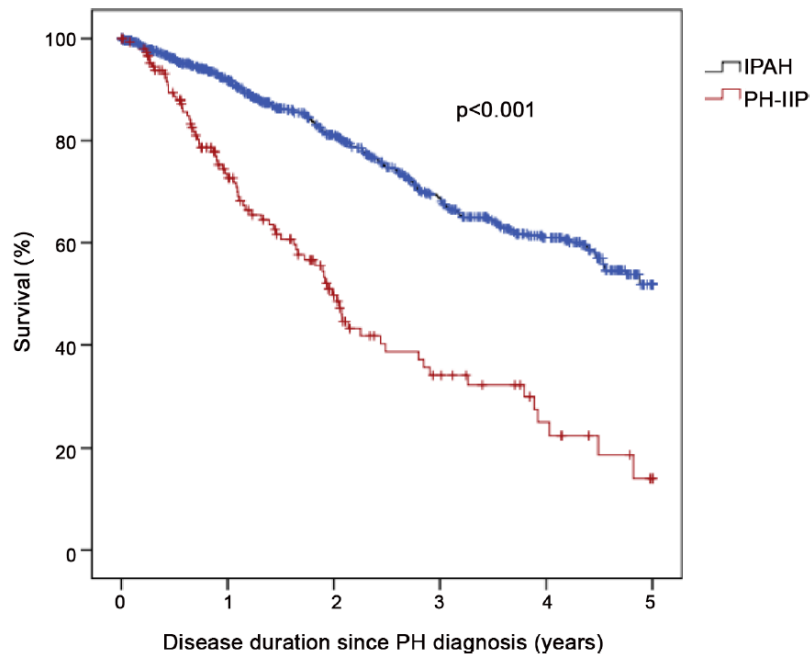
Registr COMPERA, IPAH (n = 798), PH-IIP (n = 151), PAPm  $\geq$  25 mmHg, PAWP  $\leq$  15 mmHg  
PH-IPP respondér na specifickou léčbu: 6MWD  $\geq$  20 m nebo zlepšení NYHA třídy

	IPAH (n = 798)	PH-IIP (n = 151)	p value
Female (n, %)	478 (59.9%)	56 (37.1%)	<0.001
Age, years (mean, SD)	64.5 $\pm$ 15.8	71.1 $\pm$ 10.7	<0.001
BMI, kg/m <sup>2</sup> (mean/SD)	28.0 $\pm$ 6.7	26.9 $\pm$ 5.0	0.033
6MWD, m (mean, SD)	303 $\pm$ 129	251 $\pm$ 116	<0.001
WHO Class I (n, %)	3 (0.4%)	0	*
WHO Class II (n, %)	98 (12.5%)	4 (2.7%)	*
WHO Class III (n, %)	568 (72.4%)	106 (71.6%)	*
WHO Class IV (n, %)	115 (14.7%)	38 (25.7%)	*
TLC (% pred)	95.3 $\pm$ 17.2	68.8 $\pm$ 17.0	<0.001
FVC (% pred)	82.1 $\pm$ 21.6	62.9 $\pm$ 20.0	<0.001
FEV <sub>1</sub> (% pred)	76.7 $\pm$ 21.4	67.7 $\pm$ 20.2	0.001
DLCO (% pred)	50.1 $\pm$ 20.5	28.5 $\pm$ 15.8	<0.001
paO <sub>2</sub> (mmHg) <sup>a</sup>	62.4 $\pm$ 13.4	56.3 $\pm$ 10.4	<0.001
paCO <sub>2</sub> (mmHg) <sup>a</sup>	34.5 $\pm$ 7.1	37.4 $\pm$ 5.5	0.001
RAP, mmHg	8.6 $\pm$ 5.0	5.9 $\pm$ 4.8	<0.001
mPAP, mmHg	45 $\pm$ 13	37 $\pm$ 9	<0.001
PAWP, mmHg	9.6 $\pm$ 3.4	8.0 $\pm$ 3.8	<0.001
PVR, dyn.s.cm <sup>-5</sup>	793 $\pm$ 433	649 $\pm$ 268	0.001
CI, L/min/m <sup>2</sup>	2.2 $\pm$ 0.7	2.1 $\pm$ 0.6	0.438
SvO <sub>2</sub> , %	63 $\pm$ 9	64 $\pm$ 8	0.063
Bilirubin (mg/dl)	0.7 (0.5–1.0)	0.6 (0.5–0.9)	0.011
Creatinine (mg/dl)	1.0 (0.8–1.3)	1.0 (0.8–1.3)	0.755
Uric acid (mg/dl)	7.2 (5.7–9.0)	7.1 (5.7–8.3)	0.224
NT-proBNP (ng/L)	1,627 (577–3,487)	1,029 (373–2901)	0.065
BNP (ng/L)	233 (93–469)	114 (59–256)	0.009

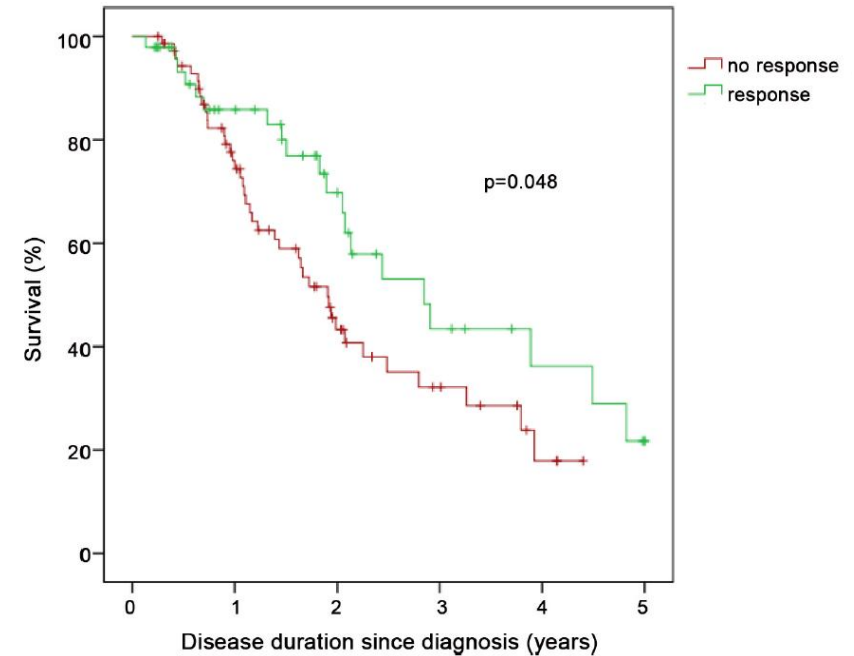
	IPAH (n = 798)	PH-IIP (n = 151)	p value
<b>Baseline</b>			
ERA monotherapy	172 (21.6%)	11 (7.3%)	<0.001
PDE-5 inhibitor monotherapy	461 (57.8%)	133 (88.1%)	<0.001
PCA monotherapy	13 (1.6%)	0	t.n.a.
Other monotherapy	29 (3.6%)	0	t.n.a.
ERA+PDE-5 inhibitor	85 (10.7%)	4 (2.6%)	0.001
Other double combination therapies	29 (3.6%)	3 (2.0%)	0.459
<b>1 year</b>			
ERA monotherapy	72 (13.6%)	4 (4.7%)	0.020
PDE-5 inhibitor monotherapy	228 (43.1%)	75 (87.2%)	<0.001
PCA monotherapy	0	1 (1.2%)	t.n.a.
Other monotherapy	8 (1.5%)	0	t.n.a.
ERA+PDE-5 inhibitor	129 (24.4%)	2 (2.3%)	<0.001
Other double combination therapies	42 (7.9%)	0	0.002
Triple combination therapy	33 (6.2%)	0	t.n.a.
No therapy	17 (3.2%)	4 (4.7%)	t.n.a.

# Pulmonary Hypertension in Patients with Chronic Fibrosing Idiopathic Interstitial Pneumonias

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Přežití IPAH vs PH-IPP



Přežití PH-IPP respondéři vs nonrespondéři

# PLICNÍ HYPERTENZE ASOCIOVANÁ S PLICNÍMI ONEMOCNĚNÍMI

## Léčba: Léčba základního plicního onemocnění

DDOT pokud indikována, CPAP pokud indikován, plicní RHB programy, zvažení LuTx

U kombinované etiologie PH adekvátní léčba, specifická vazodilatační léčba?



Inhaled treprostinil may be considered in patients with PH associated with ILD <sup>734</sup>	<b>IIb</b>	<b>B</b>
The use of ambrisentan is not recommended in patients with PH associated with IPF <sup>740</sup>	<b>III</b>	<b>B</b>
The use of riociguat is not recommended in patients with PH associated with IIP <sup>181</sup>	<b>III</b>	<b>B</b>
The use of PAH medication is not recommended in patients with lung disease and non-severe PH <sup>e</sup>	<b>III</b>	<b>C</b>
PDE5is may be considered in patients with severe PH associated with ILD (individual decision-making in PH centres)	<b>IIb</b>	<b>C</b>
The use of PDE5is in patients with ILD and non-severe PH is not recommended	<b>III</b>	<b>C</b>

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The use of PDE5is in patients with ILD and non-severe PH is not recommended		<b>III</b>	<b>C</b>

# 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<sup>a</sup>
- 1.3 Associated with drugs and toxins<sup>a</sup>
- 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<sup>b</sup>
- 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 %

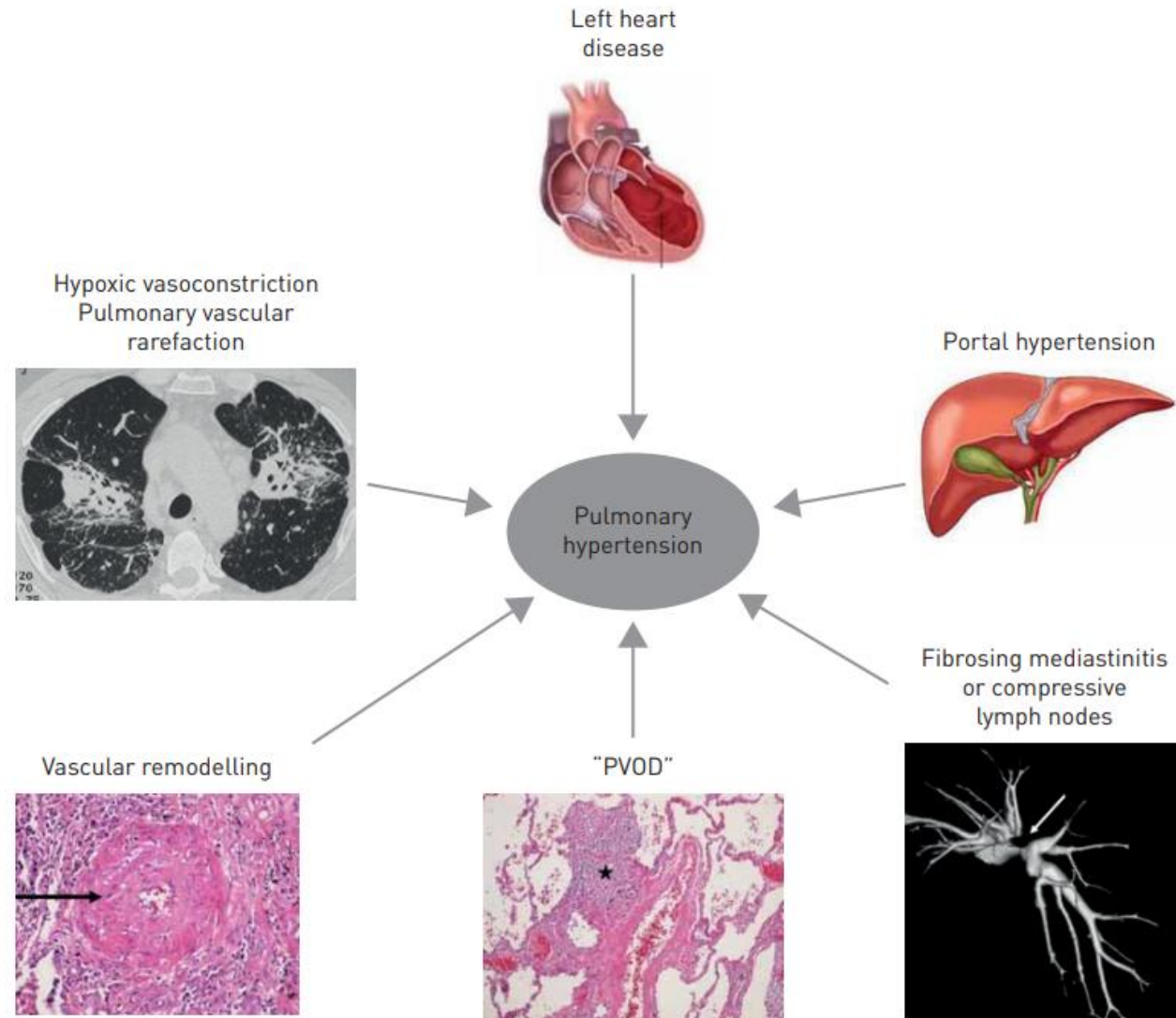
- 4.1 Chronic thrombo-embolic PH
- 4.2 Other pulmonary artery obstructions<sup>c</sup>

## GROUP 5 PH with unclear and/or multifactorial mechanisms

5 %

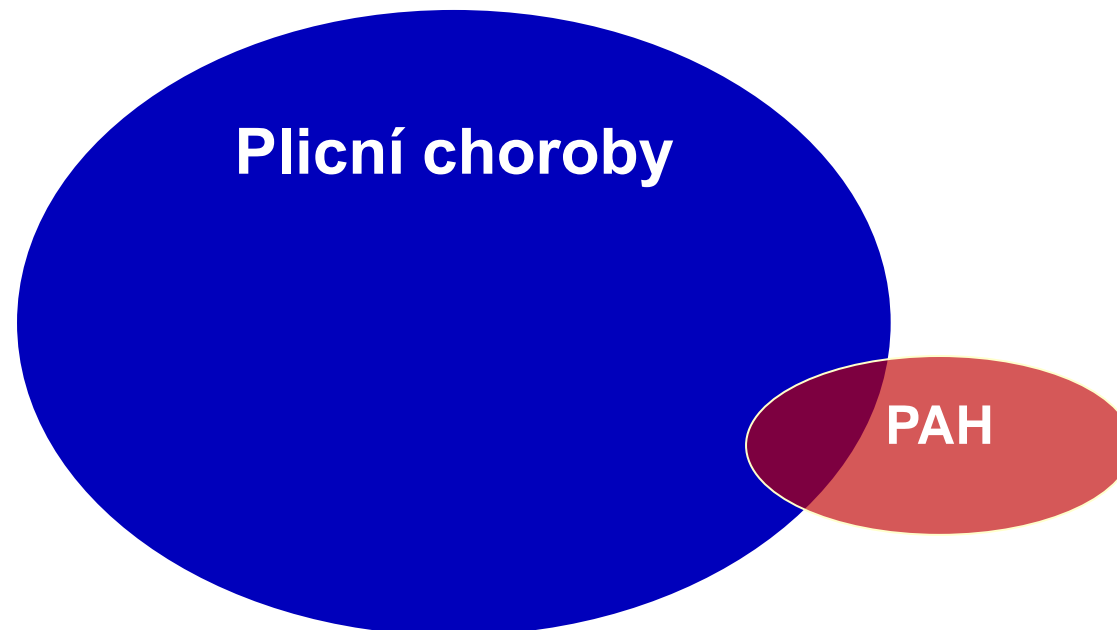
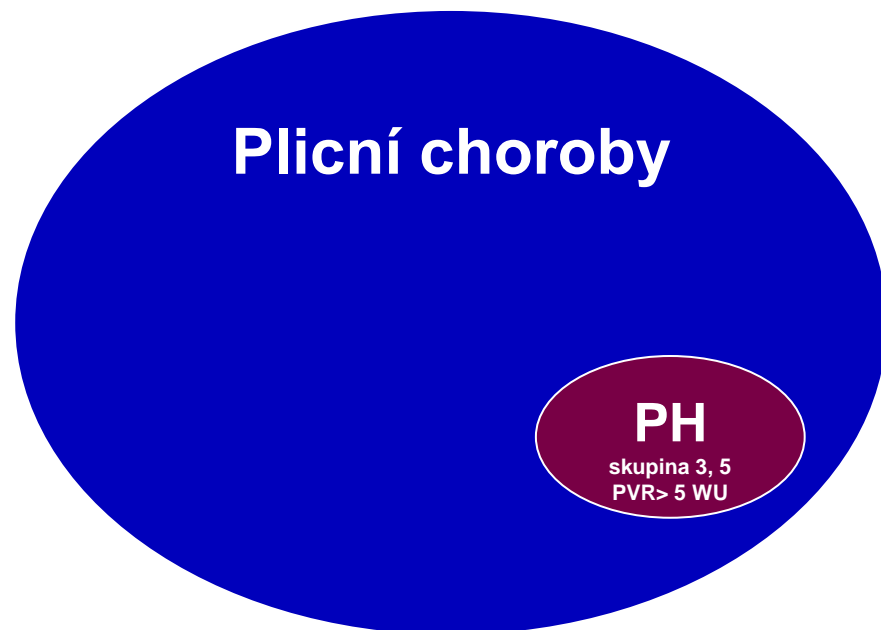
- 5.1 Haematological disorders<sup>d</sup>
- 5.2 Systemic disorders<sup>e</sup>
- 5.3 Metabolic disorders<sup>f</sup>
- 5.4 Chronic renal failure with or without haemodialysis
- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

# Which patients are SaPHe in sarcoidosis-associated pulmonary hypertension?





# PLICNÍM ONEMOCNĚNÍ A POSTIŽENÍ PLICNÍ CIRKULACE



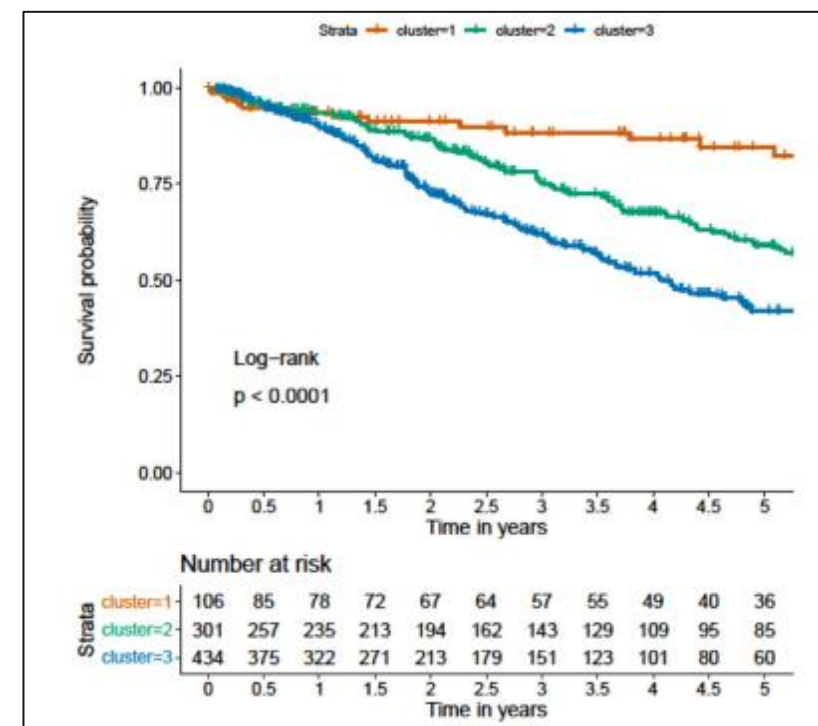
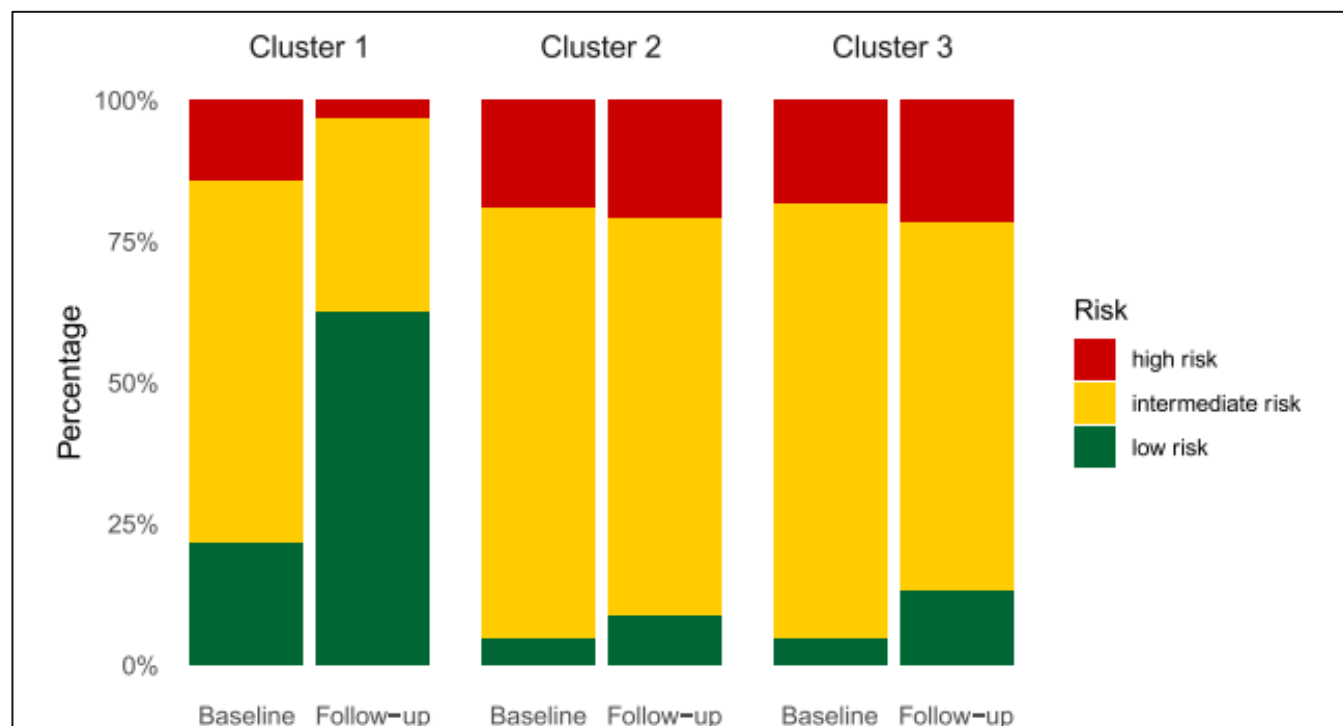
# Idiopathic pulmonary arterial hypertension phenotypes determined by cluster analysis from the COMPERA registry

N=841. Komorbidity: obezita, ICHS, arteriální hypertenze, DM

**Cluster 1** (n = 106; 12.6%): věk 45, 76% ženy, bez komorbidit, nekuřáci, DLCO  $\geq$ 45%

**Cluster 2** (n = 301; 35.8%): věk 75, 98% ženy, časté komorbidity, nekuřáci, DLCO  $\geq$ 45%

**Cluster 3** (n = 434; 51.6%): věk 72, 72% muži, časté komorbidity, kuřáci, DLCO <45%



# SHRNUTÍ

- Plicní hypertenze u plicních chorob je typicky prekapilární, lehká a obtížně léčitelná (PAMP > 20 mmHg, PVR > 2 WU)
- Diferenciální diagnostika je základem adekvátní léčby
  - symptomy, ECHO, plicní funkce, V/Q scinti
  - **kombinace etiologií (OSA, CTEPH, PAH) → adekvátní léčba**
- Těžká PH (PVR > 5 WU) skupiny 3 je indikací k diskusi s centrem pro PH k individuálnímu zvážení specifické léčby
  - **při úvaze o plicní cévní chorobě u IPP**  
→ **Ize zvážit PDE5i, treprostinil inh. (doporučení IIb)**