NE KAŽDÁ PLICNÍ HYPERTENZE JE PAH

PAVEL JANSA

pavel.jansa@vfn.cz

KOMPLEXNÍ KARDIO VASKULÁRNÍ CENTRUM VFN Praha

European Reference Network for rare or low prevalence complex diseases

Network Respiratory Diseases (ERN-LUNG)

Member General University Hospital in Prague — Czechia

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

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

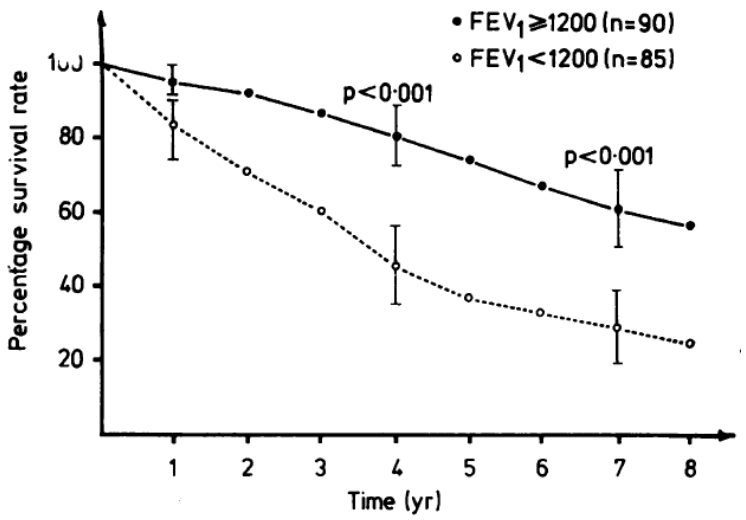
Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
Combined post- and pre-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise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

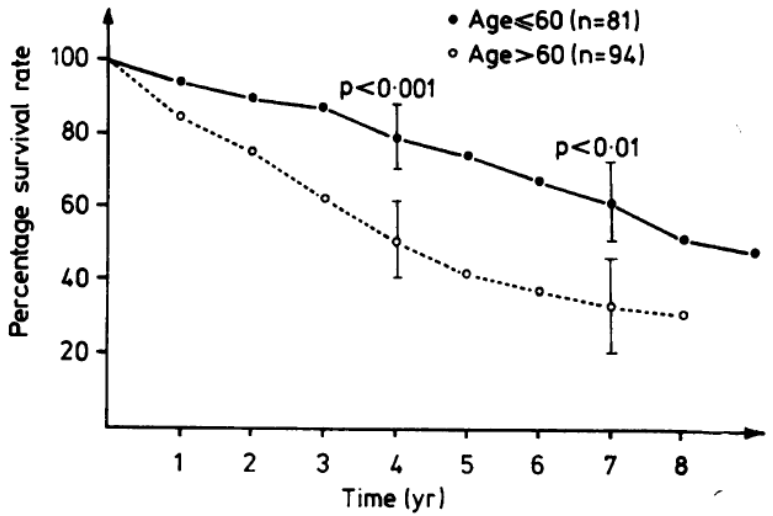
Cor et Vasa 58 (2016)
EHJ 2022, *ERJ* 2022

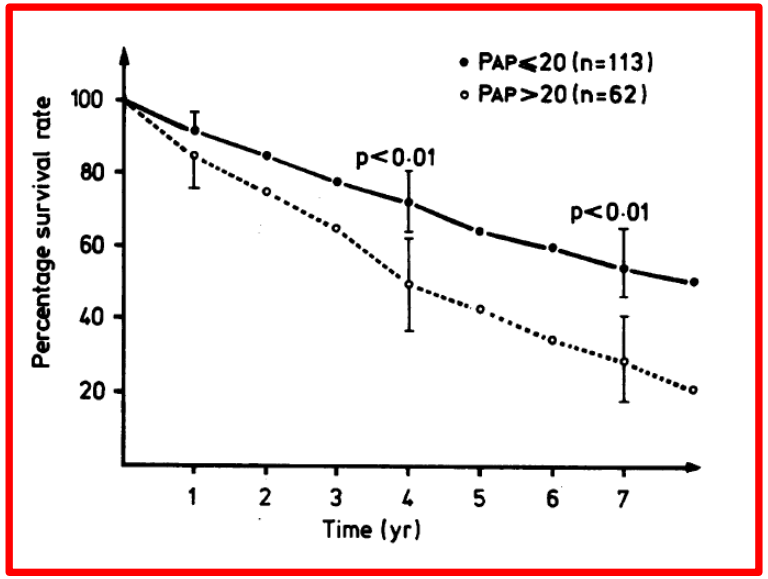
Prognostic value of pulmonary artery pressure in chronic obstructive pulmonary disease

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

N=175, RHC 1968-1972, FEV1 40.2 ±11.1 %, PAMP 19.8 ± 7.6 mmHg, CI 3.24 ± 0.93 L/min/m^2
Rekatetrizace: N=64 (po 5.5 letech)



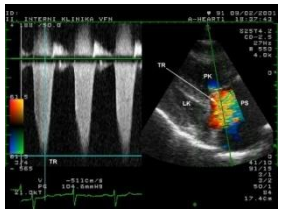




Thorax 1981 ;36:752-758

Suspekce

Plicní hypertenze podle ECHO možná nebo pravděpodobná

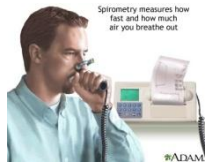


Postižení myokardu nebo chlopní levého srdce

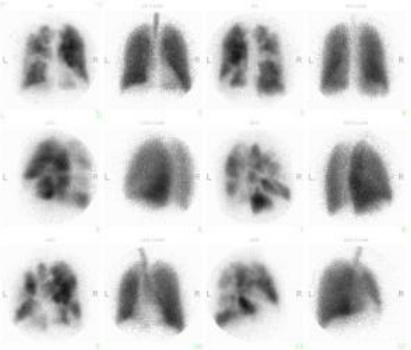
Bez postižení myokardu nebo chlopní levého srdce

Detekce

Skupina 2



Plicní funkce



Výrazná redukce

Normální nebo nevýznamná redukce

V/Q plicní scintigrafie

Skupina 3

Nesegmentární defekty

Segmentární defekty

hemodynamika

CT angiografie, angiografie, hemodynamika

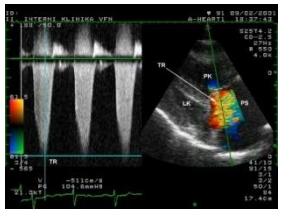
Konfirmace

Skupina 1

Skupina 4

Suspekce

Plicní hypertenze podle ECHO možná nebo pravděpodobná

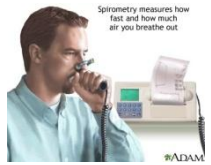


Postižení myokardu nebo chlopní levého srdce

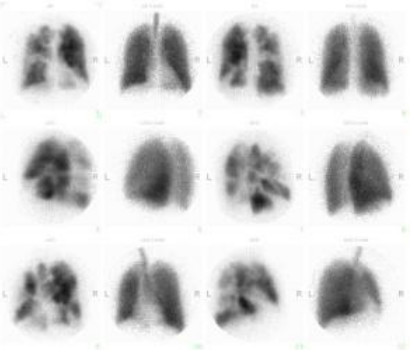
Bez postižení myokardu nebo chlopní levého srdce

Detekce

Skupina 2 → **PH centrum (těžká PH)**



Plicní funkce



Výrazná redukce

Normální nebo nevýznamná redukce → V/Q plicní scintigrafie

Skupina 3

Nesegmentární defekty

Segmentární defekty

Konfirmace

PH centrum (těžká PH)

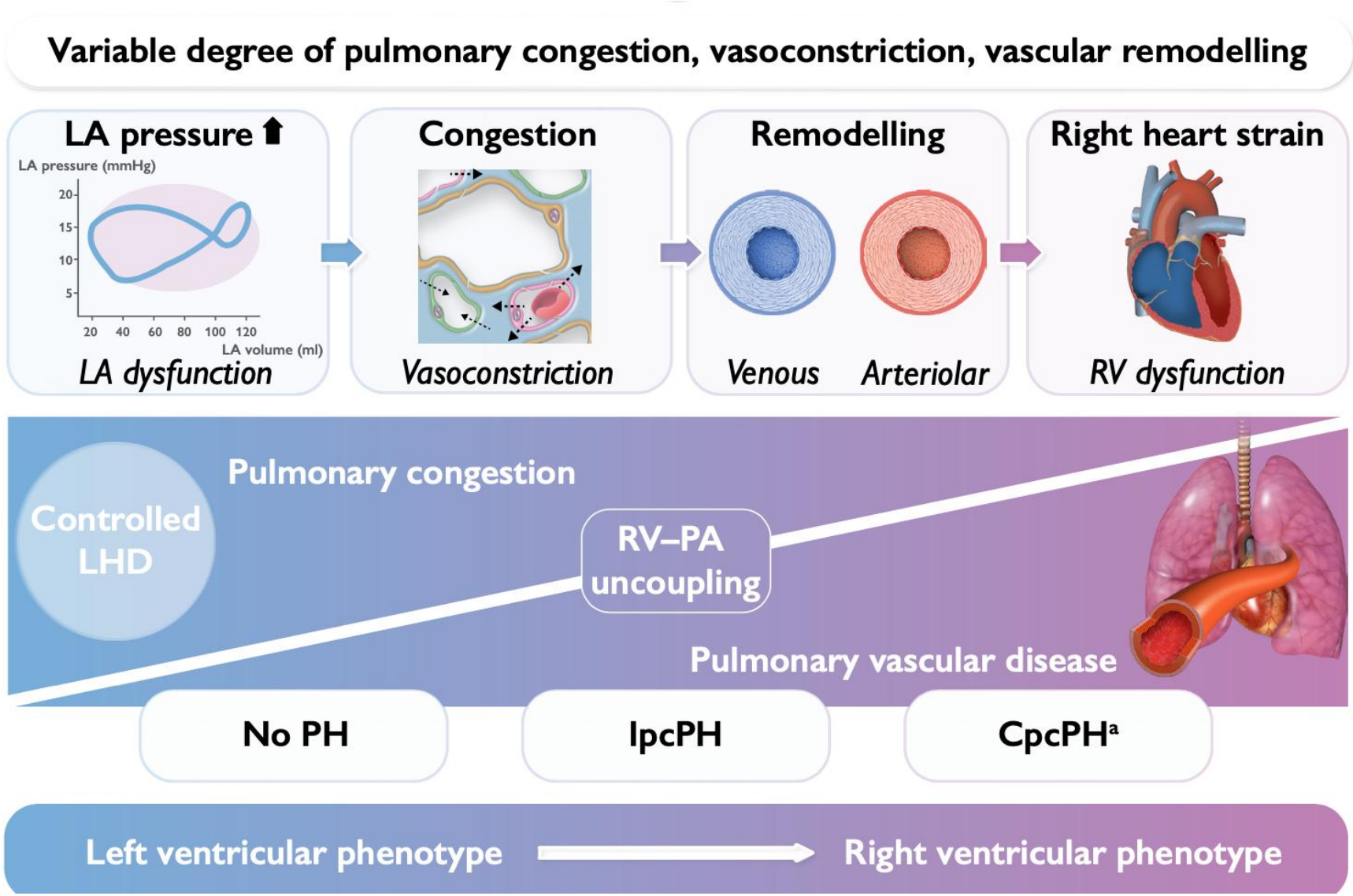
hemodynamika

CT angiografie, angiografie, hemodynamika

PH centrum ← Skupina 1

Skupina 4 → **PH (CTEPH) centrum**

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

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

HEMODYNAMICKÁ DEFINICE PH 2022

ESC
ERS

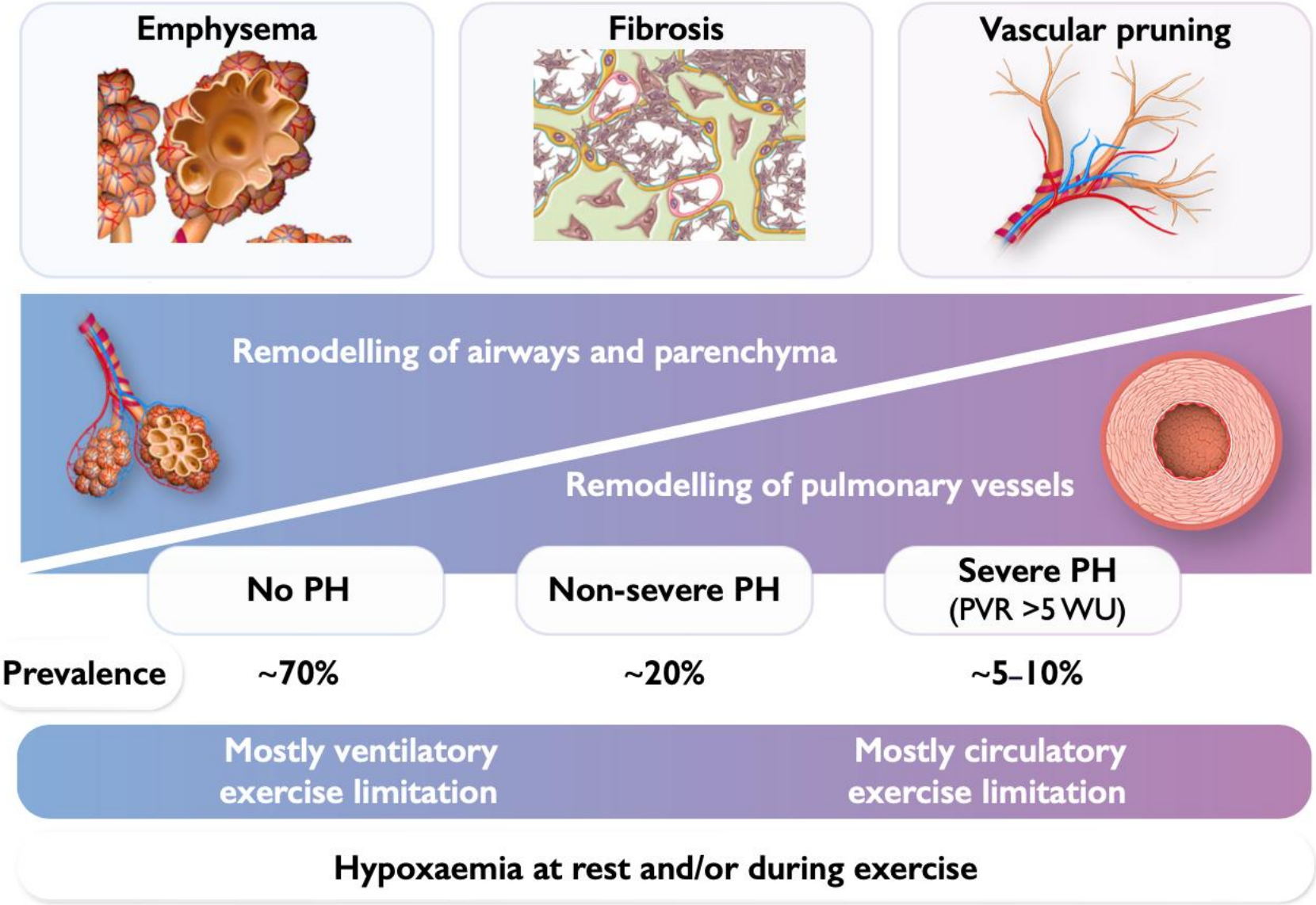
Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
Combined post- and pre-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise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

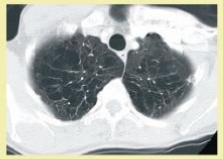


Těžká PH
PVR > 5 WU

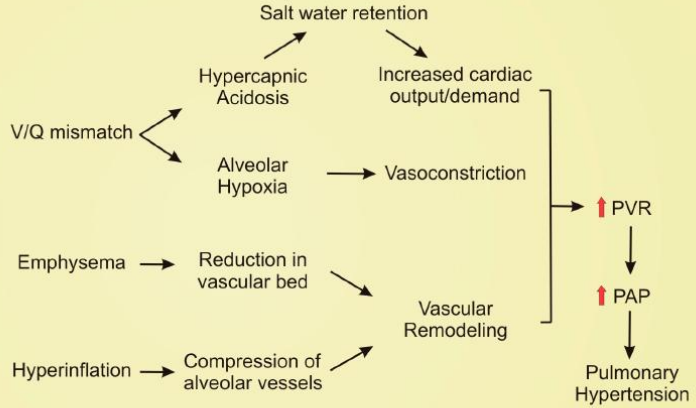
- Bez vztahu k plicním objemům
- Hypoxémie
- Nízká DLco

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

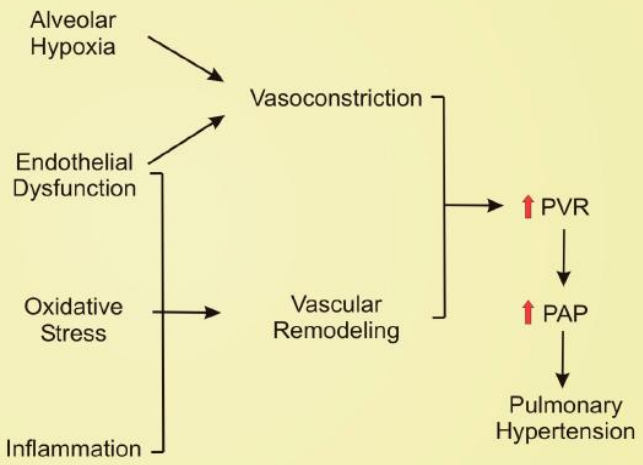
PATOFYZIOLOLGIE PH U PLICNÍCH ONEMOCNĚNÍ



Chronic Obstructive Pulmonary Disease

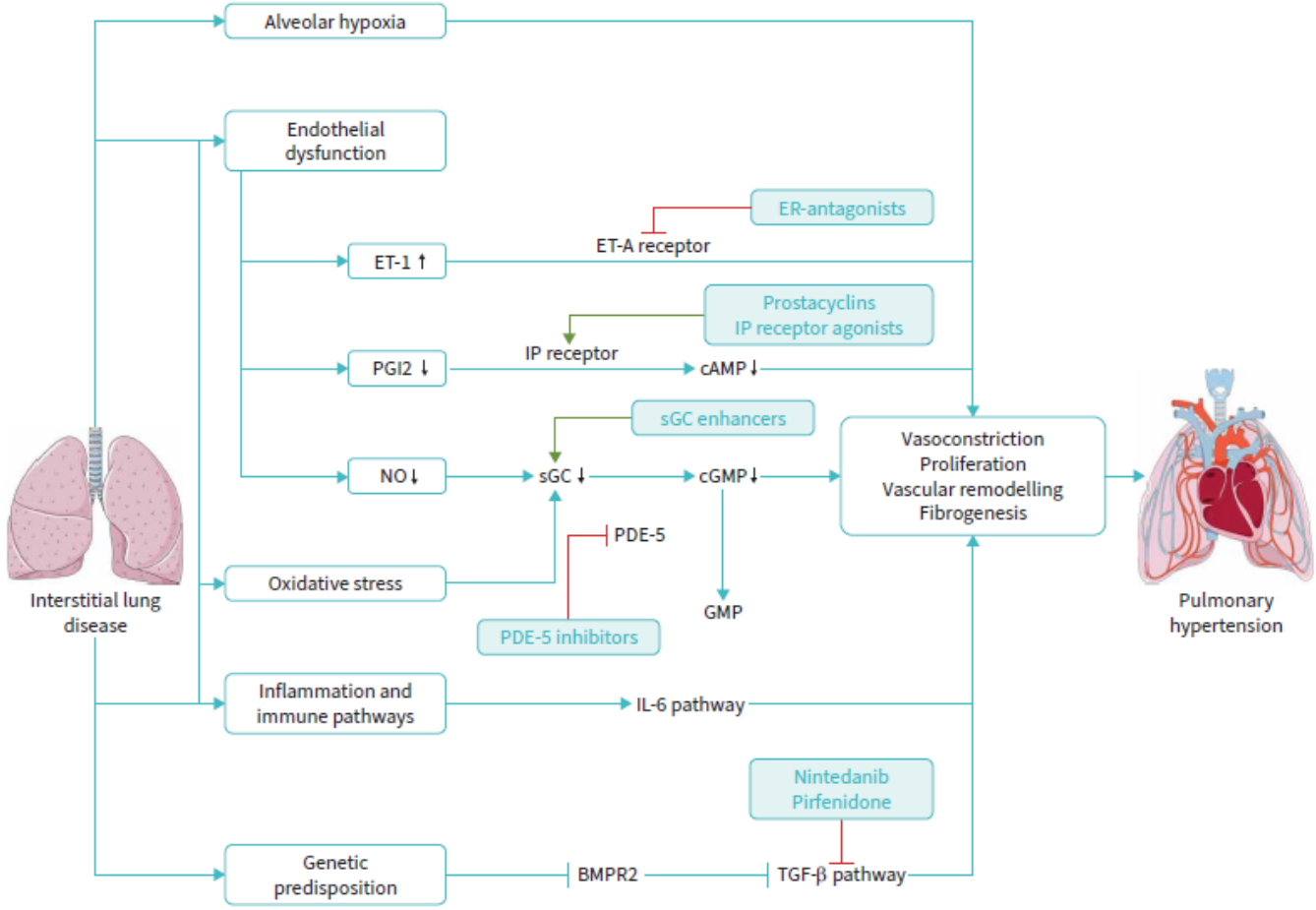


Interstitial Lung Disease



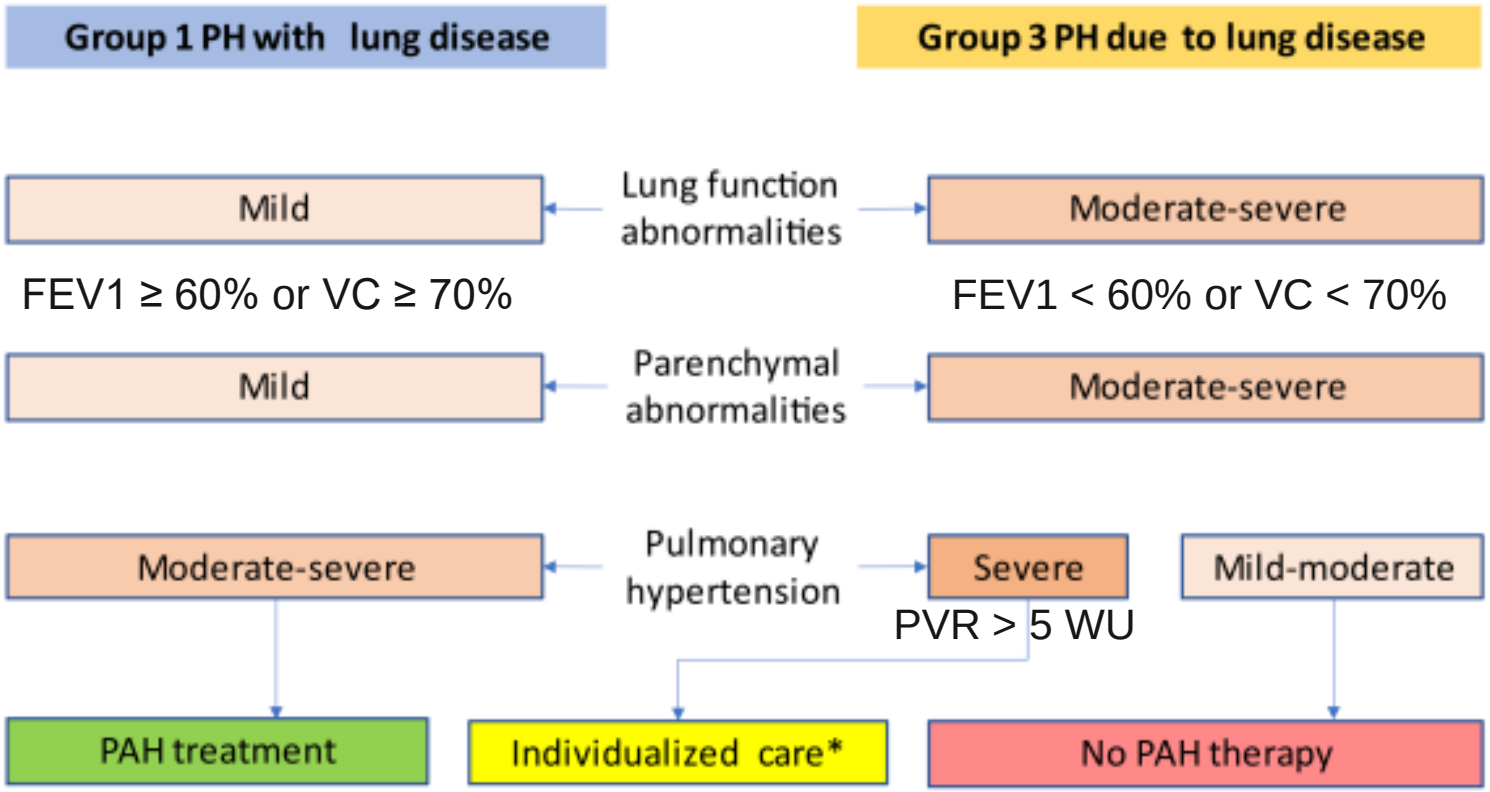
PVR≤5WU

PVR>5WU



Gonzales J et al. *Pharmaceuticals 2023*, 16, 418, Dhont S et al. *ERJ Open Res* 2022; 8: 00272-2022

The Challenge to Decide between Pulmonary Hypertension Due to Chronic Lung Disease and PAH with Chronic Lung Disease



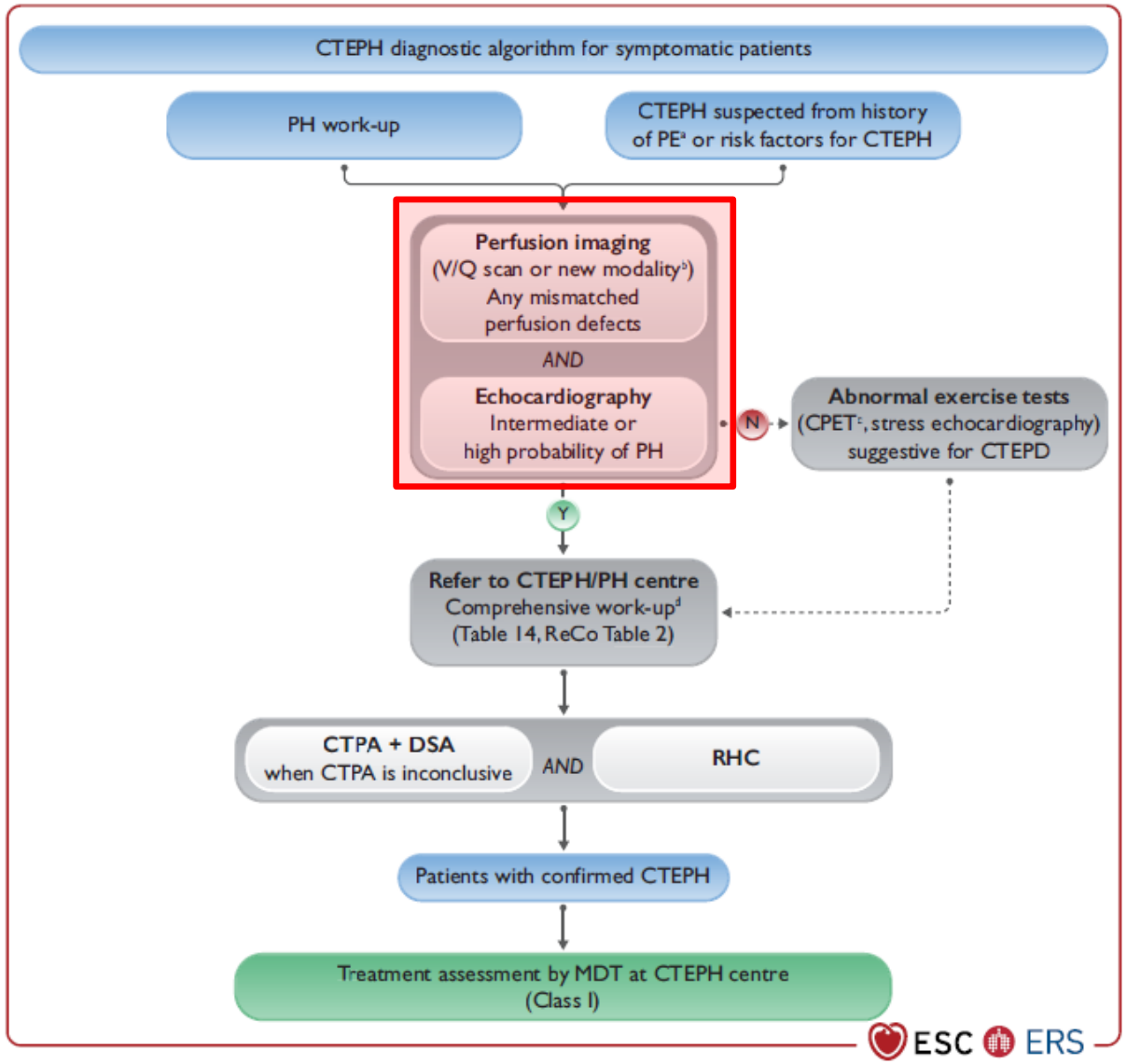
Olschewski H. *Diagnostics* 2021, 11, 311

DIAGNOSTICKÝ ALGORITMUS CTEPH (2022)

Suspekce

Detekce

Konfirmace



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

Ventilation–Perfusion Scintigraphy Is More Sensitive than Multidetector CTPA in Detecting Chronic Thromboembolic Pulmonary Disease as a Treatable Cause of Pulmonary Hypertension

Nina Tunariu[1], Simon J.R. Gibbs[2,3], Zarni Win[4], Wendy Gin-Sing[2], Alison Graham[1], Philip Gishen[1], and Adil AL-Nahhas[3,4]

227 patients referred to the PH service between 2000 and 2005. Group A - CTEPH, Group B - non CTEPH

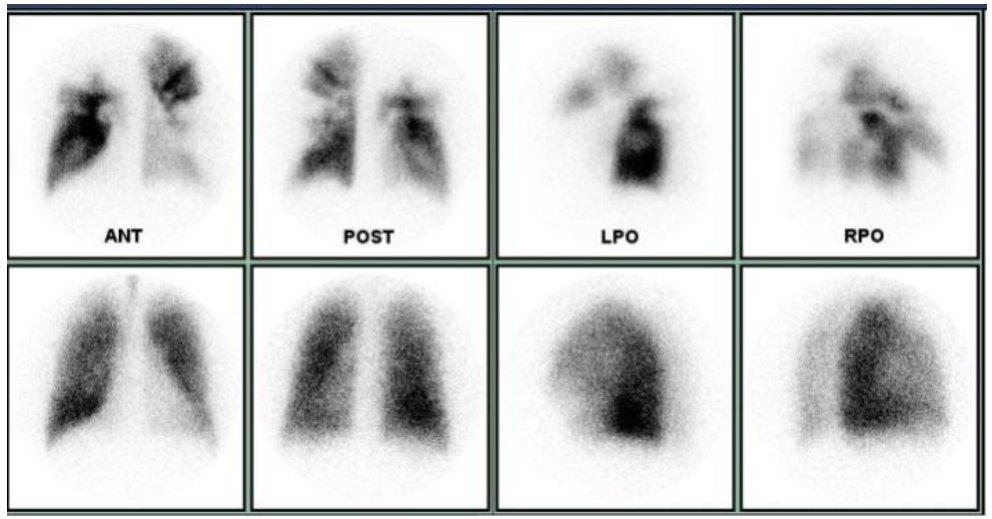


TABLE 1
Summary of V/Q Scans and CTPA Results

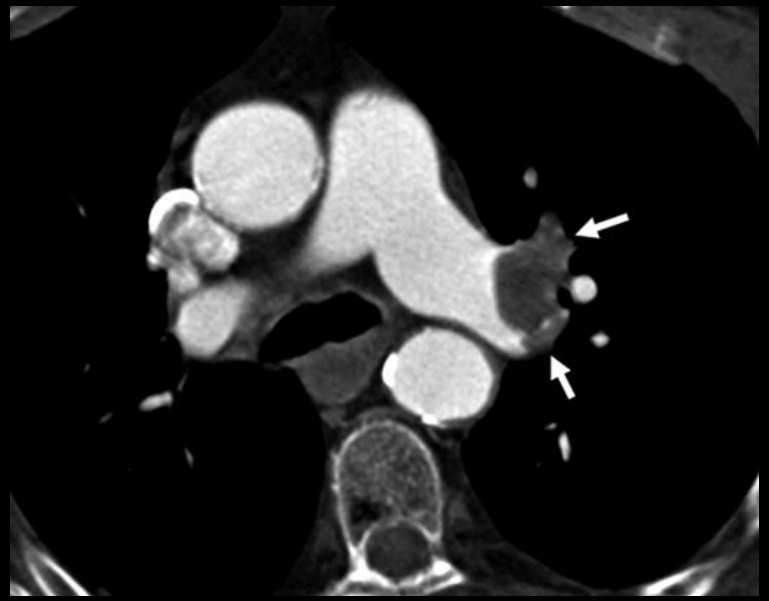
	V/Q			CTPA	
Group	Low probability	Intermediate probability	High probability	Negative	Positive
A (*n* = 78)	2	1	75	38	40
B (*n* = 149)	134	7	8	148	1

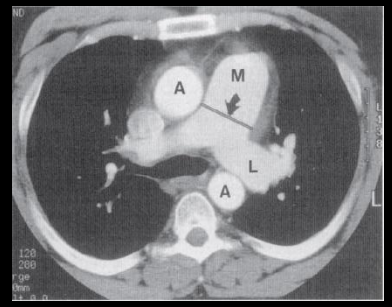
VQ scintigraphy: sensitivity 96-97%, specificity 90-95%
CT angiography: sensitivity 51%, specificity 99%

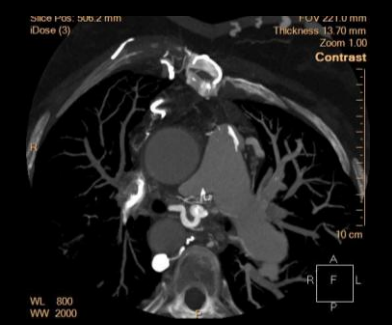
J Nucl Med 2007; 48:680–684

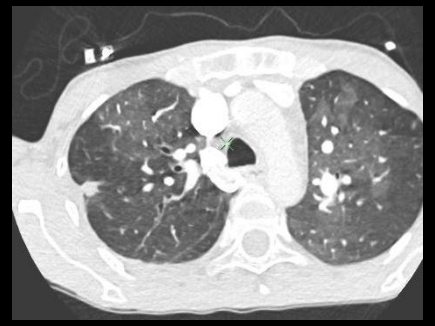
PLICNÍ EMBOLIE

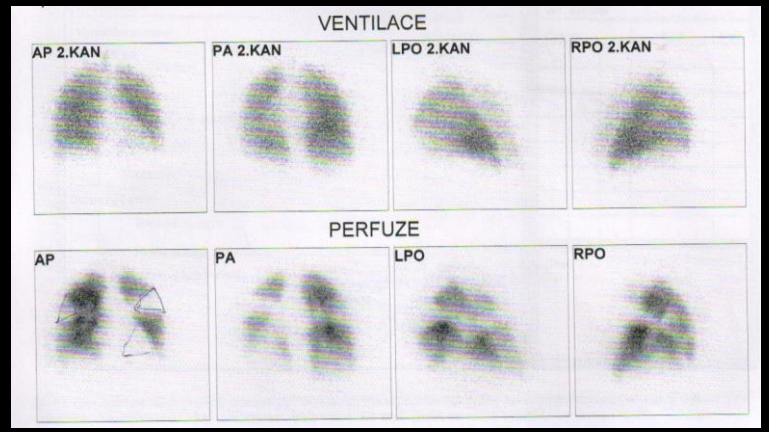
CTEPH

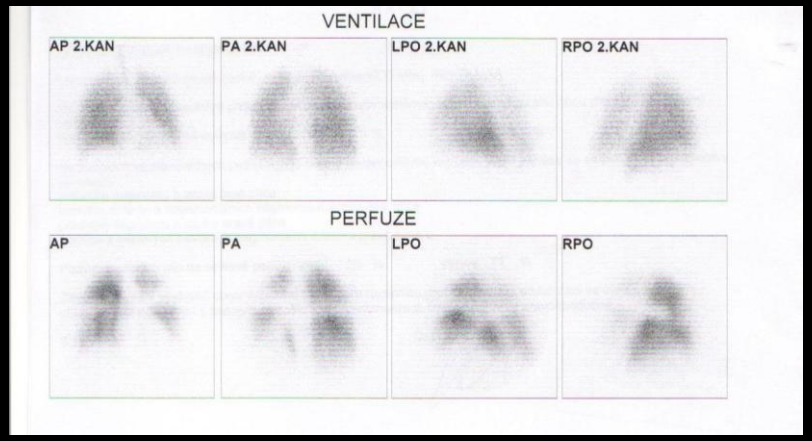












Zdroj: VFN

EUROPEAN RESPIRATORY *journal*
FLAGSHIP SCIENTIFIC JOURNAL OF ERS

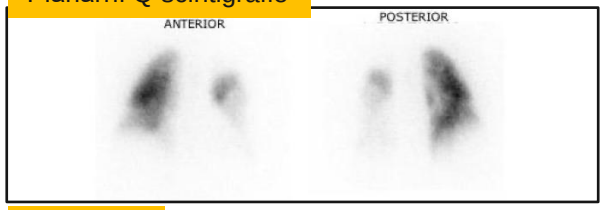
DETEKCE A DIAGNOSTIKA CTEPH

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

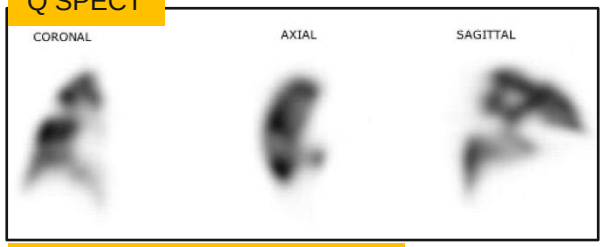
DECT a **MR perfúze** – nenahrazuje scintigrafii (limitovaná dostupnost, zkušenost, validace)

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

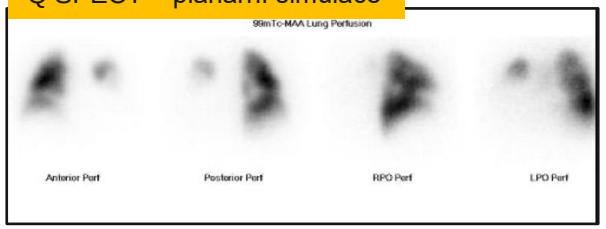
Planární Q scintigrafie



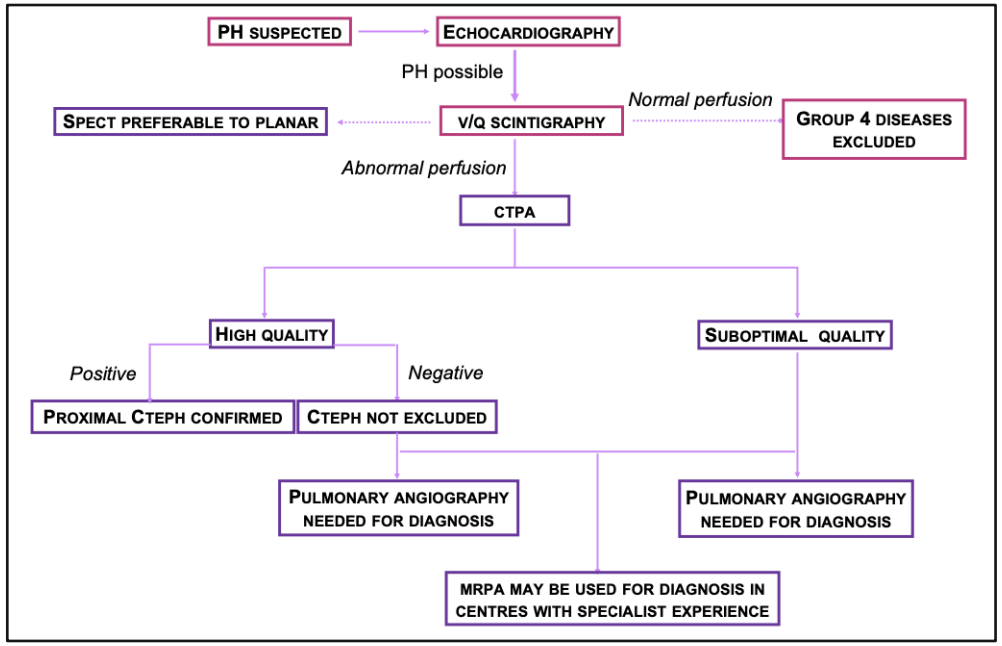
Q SPECT



Q SPECT – planární simulace



DECT

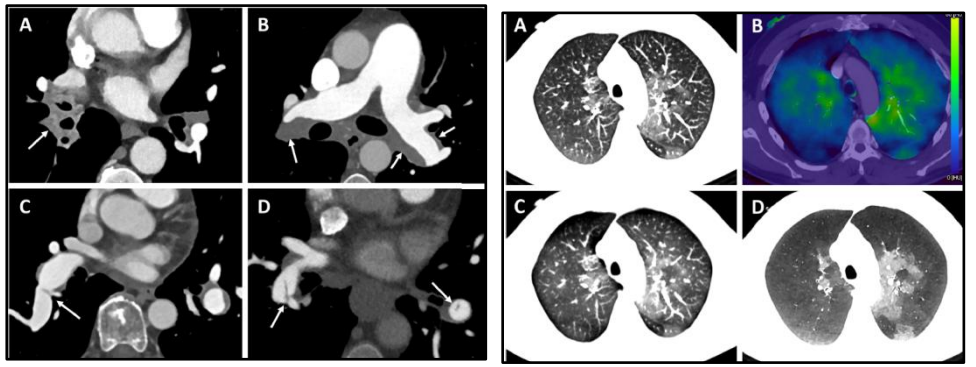


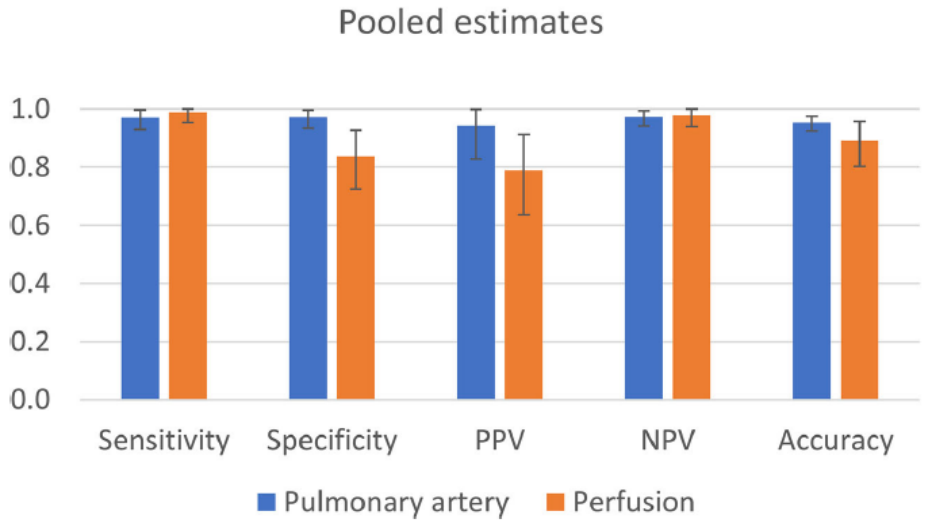
Eur Respir J 2021 Jun 17;57(6):2002828

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



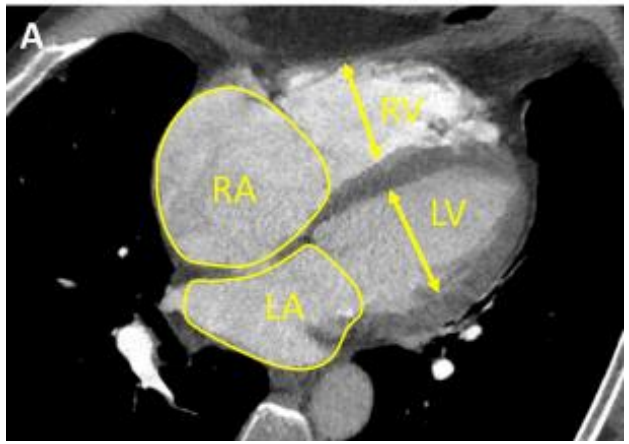


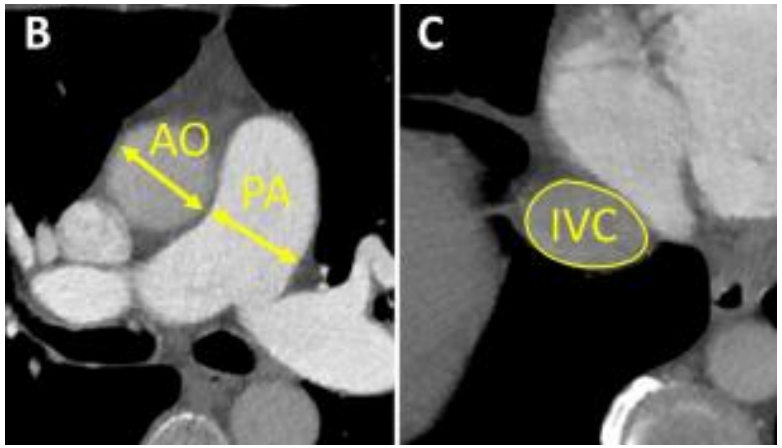
Lambert L et al. *Eur Radiol* 2022, Apr 28. doi: 10.1007/s00330-022-08804-5

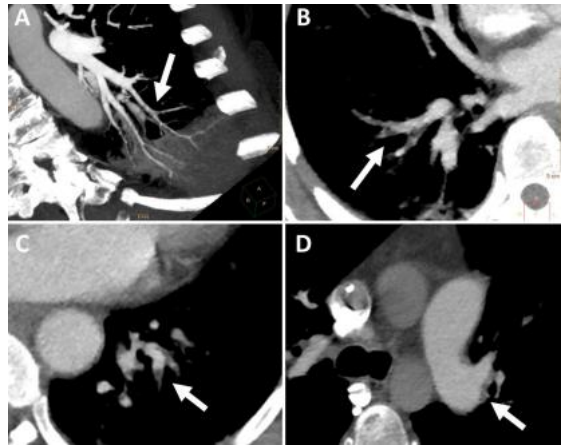
Even non-expert radiologists report chronic thromboembolic pulmonary hypertension (CTEPH) on CT pulmonary angiography with high sensitivity and almost perfect agreement

Jan Hrdlicka[1] · Martin Jurka[1] · Bianka Bircakova[1] · David Ambroz[2] · Pavel Jansa[2] · Andrea Burgetova[1] · Lukas Lambert[1]

51 patients with CTEPH, 49 patients without CTEPH
Three radiologists with different levels of experience in CT imaging (**R1**:15 years, **R2**:6 years, and **R3**:3 years)







	R1		R2		R3		Overall	
	(%)	95%CI	(%)	95%CI	(%)	95%CI	(%)	95%CI
Sensitivity	100	93–100	100	93–100	100	93–100	100	98–100
Specificity	100	93–100	96	86–100	96	86–100	97	93–99
PPV	100	-	96	87–99	96	87–99	97	94–99
NPV	100	-	100	-	100	-	100	-
Accuracy	100	97–100	98	93–100	98	93–100	99	97–100

Eur Radiol 2023, Aug 22. doi: 10.1007/s00330-023-10098-0

PLICNÍ HYPERTENZE V ČR

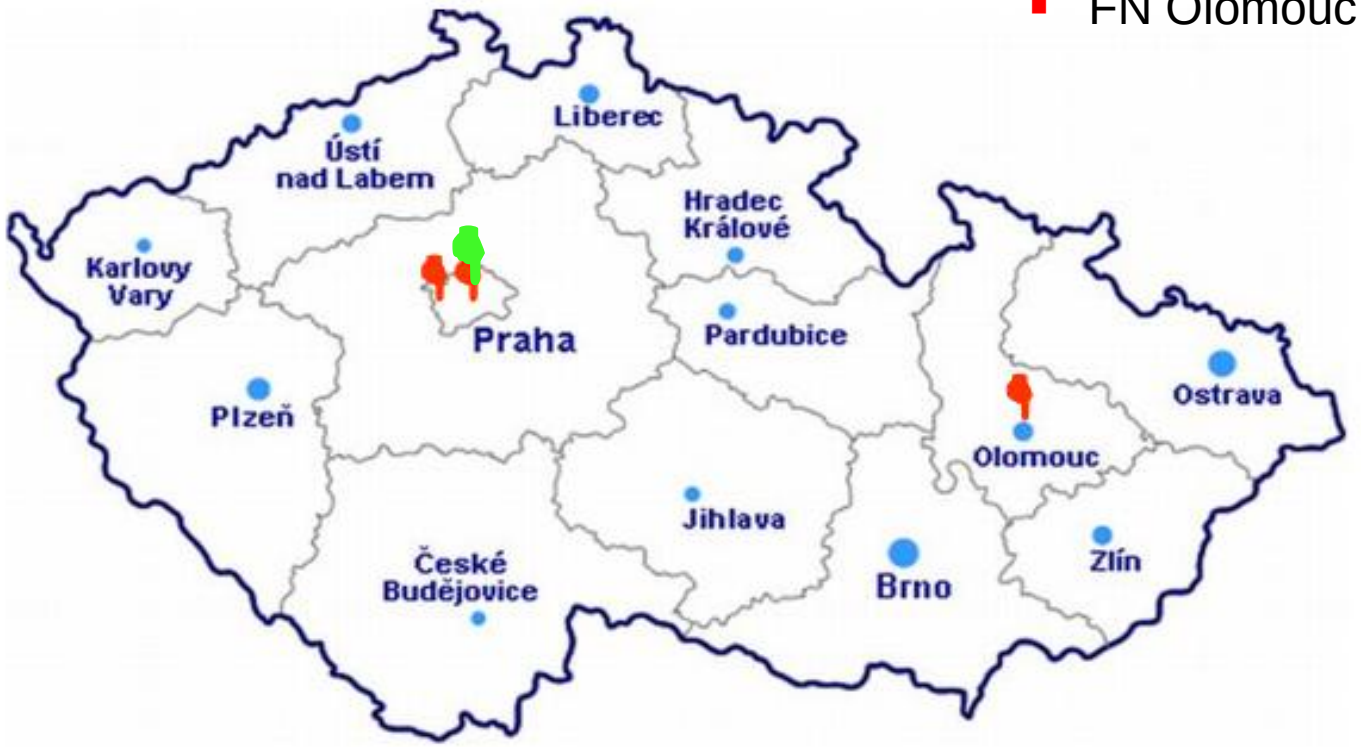
European Reference Network
for rare or low prevalence complex diseases

Network
Respiratory Diseases
(ERN-LUNG)

Member
General University Hospital in Prague — Czechia

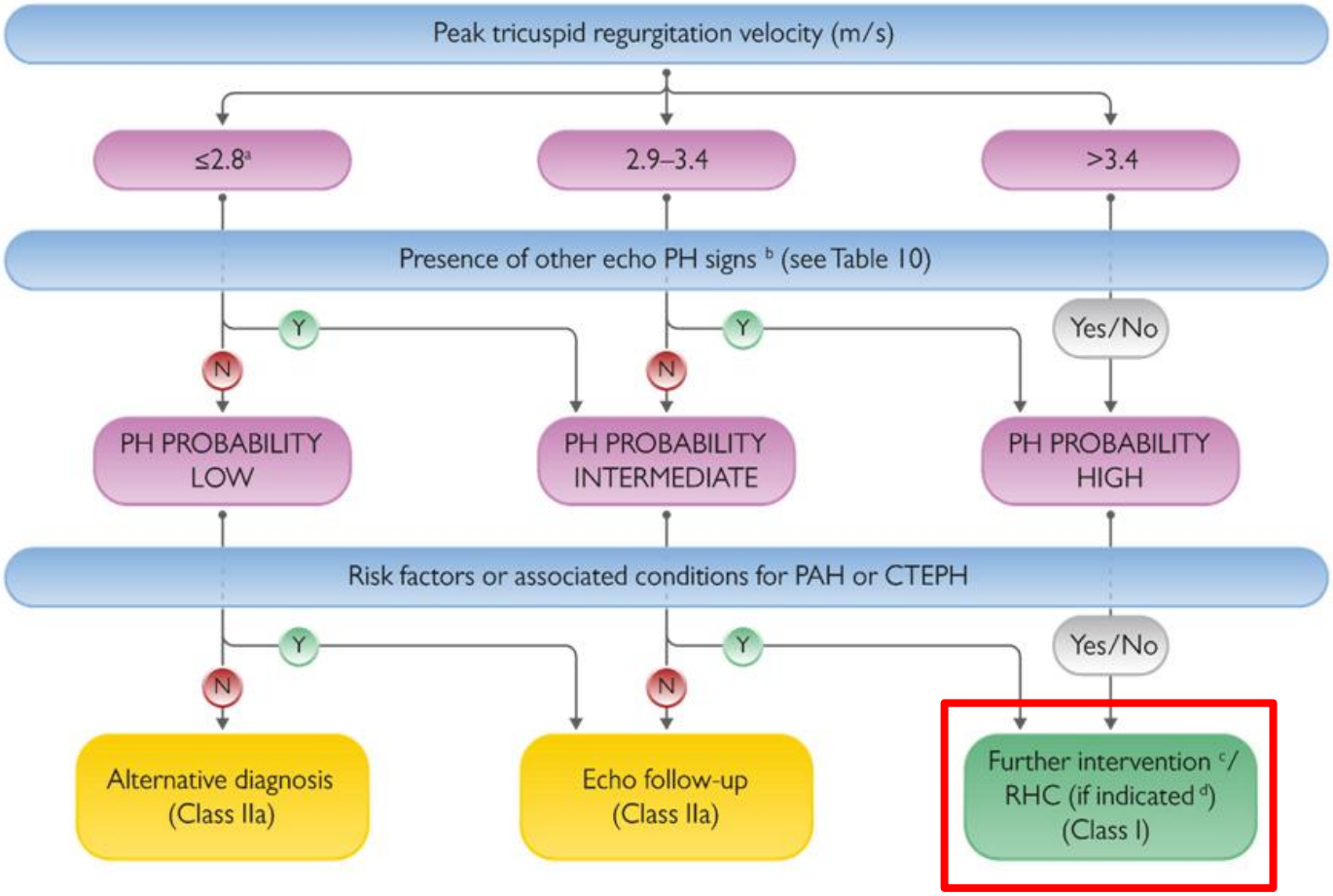
1 expertní centrum pro CTEPH
(PEA/BPA, CTEPH MDT team)
- VFN Praha

3 expertní centra pro PAH
- VFN Praha
- IKEM Praha
- FN Olomouc



Vyšetření k odeslání pacienta: ECHO, plicní funkce (spiro, objemy, DLco), V/Q scinti plic

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



Measured variables	Normal value
Right atrial pressure, mean (RAP)	2–6 mmHg
Pulmonary artery pressure, systolic (sPAP)	15–30 mmHg
Pulmonary artery pressure, diastolic (dPAP)	4–12 mmHg
Pulmonary artery pressure, mean (mPAP)	8–20 mmHg
Pulmonary arterial wedge pressure, mean (PAWP)	≤15 mmHg
Cardiac output (CO)	4–8 L/min
Mixed venous oxygen saturation (SvO_2)[a]	65–80%
Arterial oxygen saturation (SaO_2)	95–100%
Systemic blood pressure	120/80 mmHg
Calculated parameters	
Pulmonary vascular resistance (PVR)[b]	0.3–2.0 WU
Pulmonary vascular resistance index (PVRI)	3–3.5 WU·m^2
Total pulmonary resistance (TPR)[c]	<3 WU
Cardiac index (CI)	2.5–4.0 L/min·m^2
Stroke volume (SV)	60–100 mL
Stroke volume index (SVI)	33–47 mL/m^2
Pulmonary arterial compliance (PAC)[d]	>2.3 mL/mmHg

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

HEMODYNAMIC INDICES

Pressures

RA, RV, PA and PCWP

Cardiac output

Thermodilution

Fick

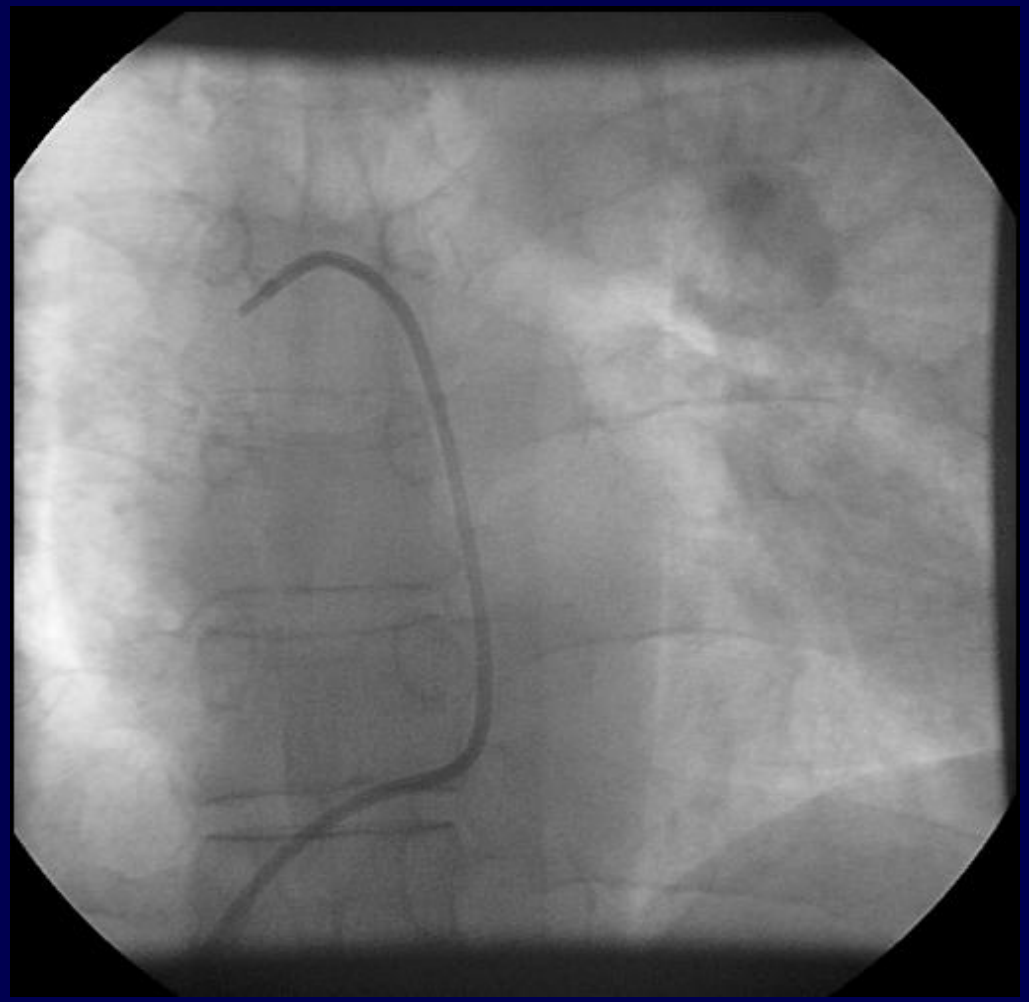
Saturations

MvO_2,SaO_2, (IVC, SVC, RA, RV)

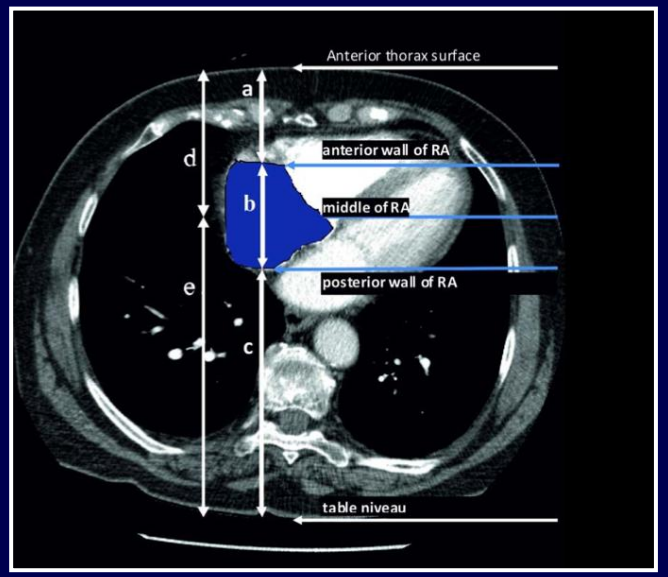
Vasodilatation testing

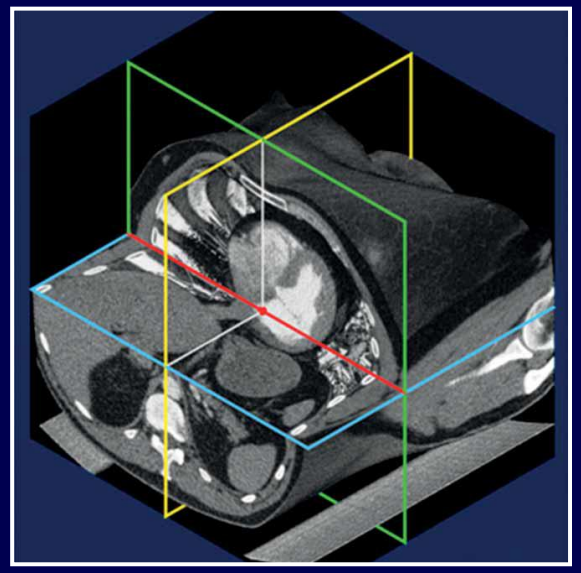
Fluid challenge

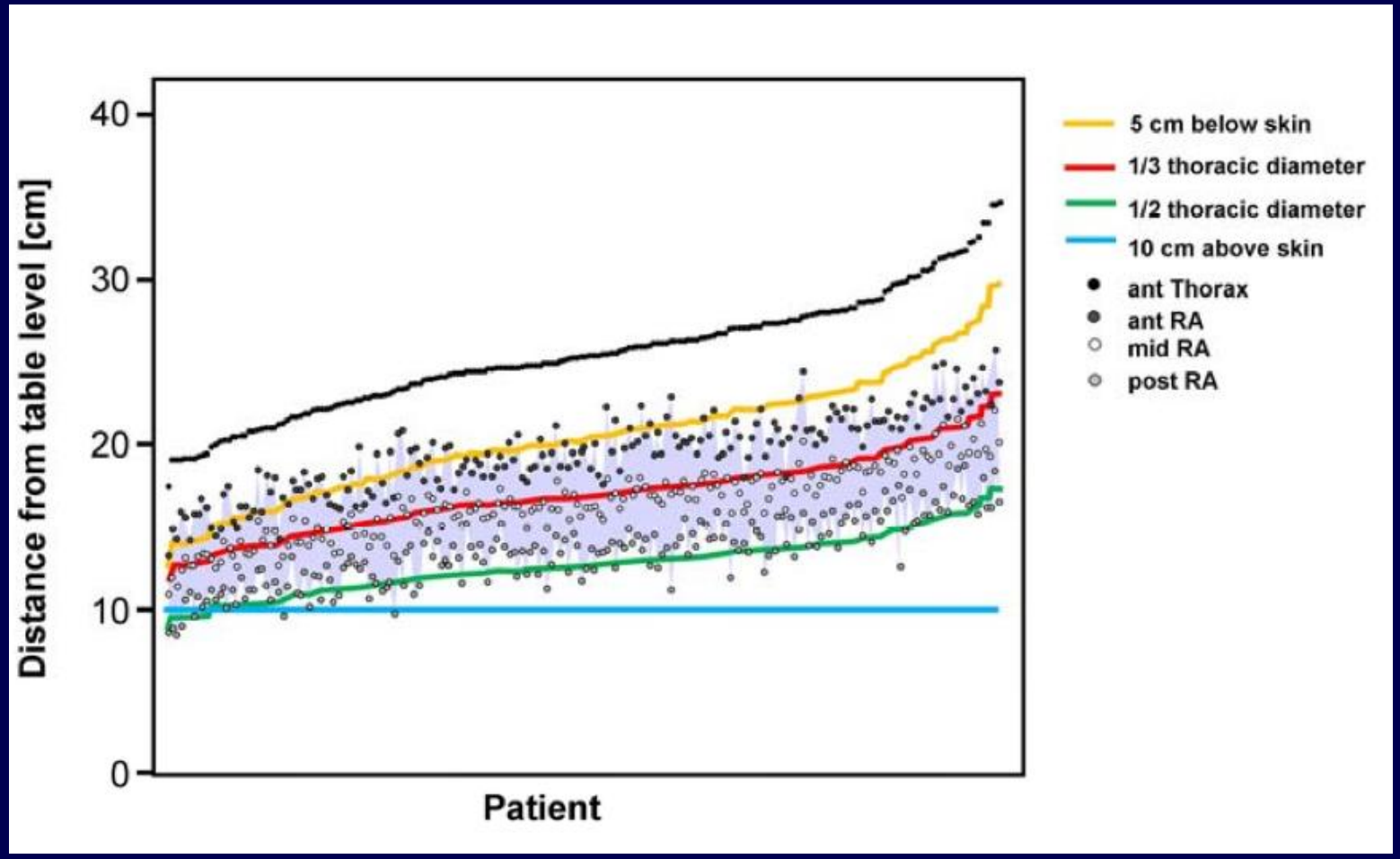
Hepatic vein catheterization



ABOUT ZERO REFERENCE LEVEL





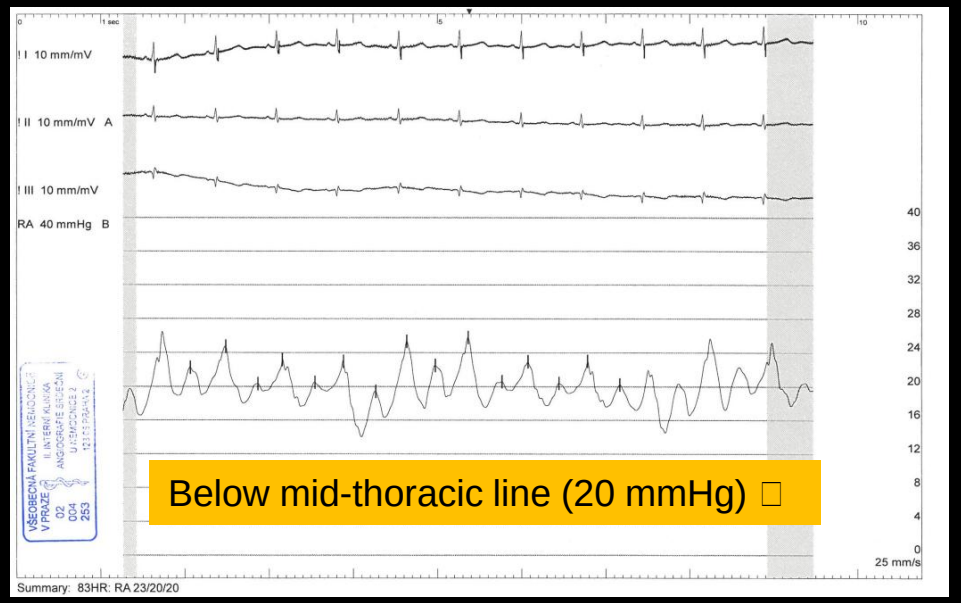


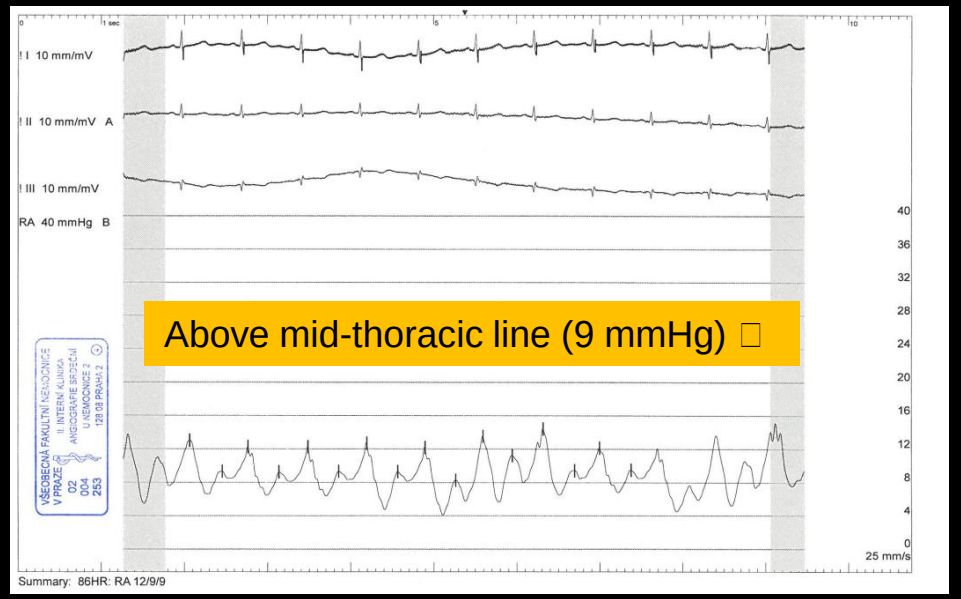
mid-thoracic line

Kovacs G et al., *Am J Respir Crit Care Med* 2014; 190: 252–257.

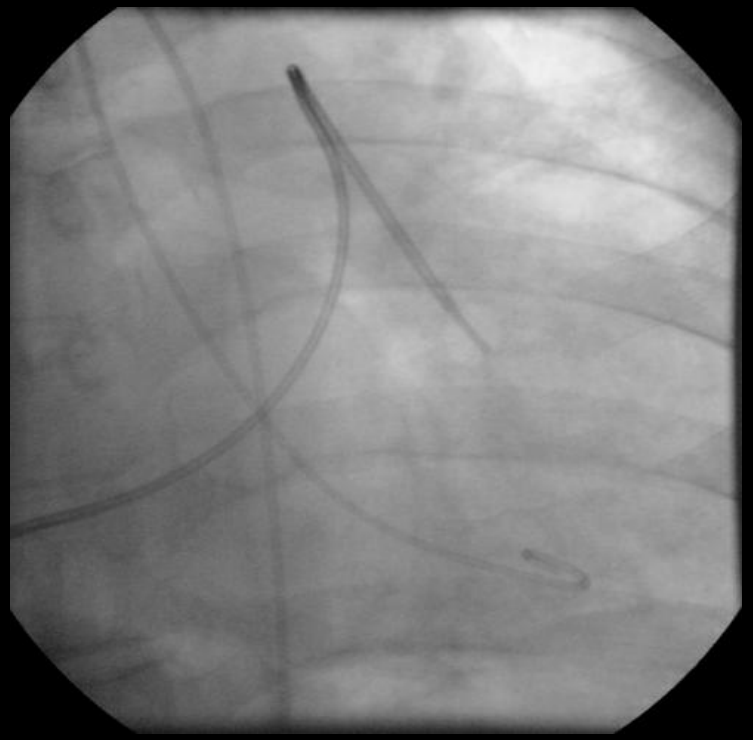
RAP (12 mmHg), mid-thoracic line

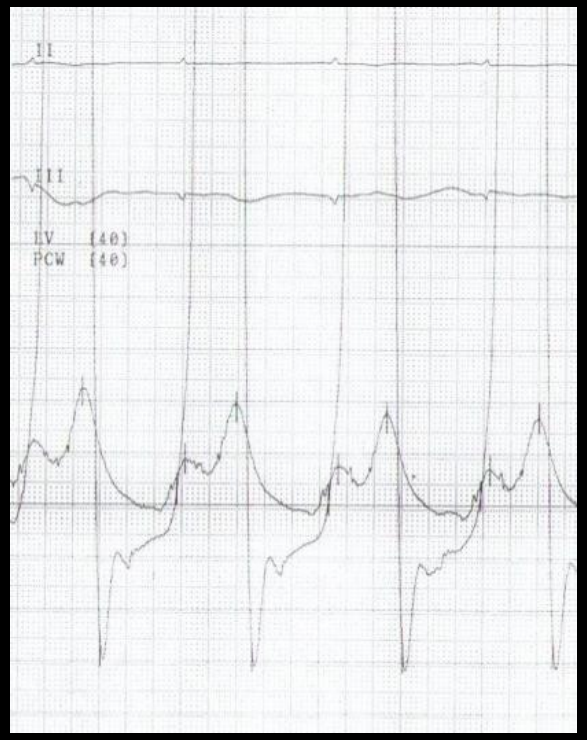
✔

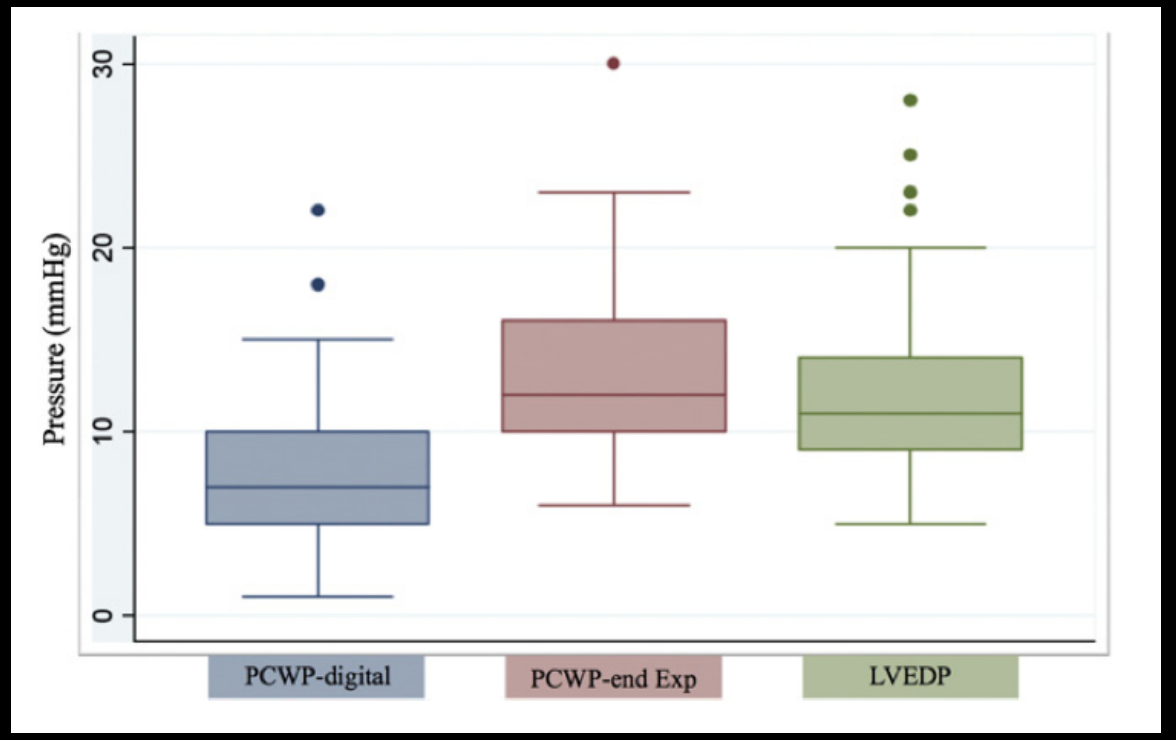




ABOUT PCWP

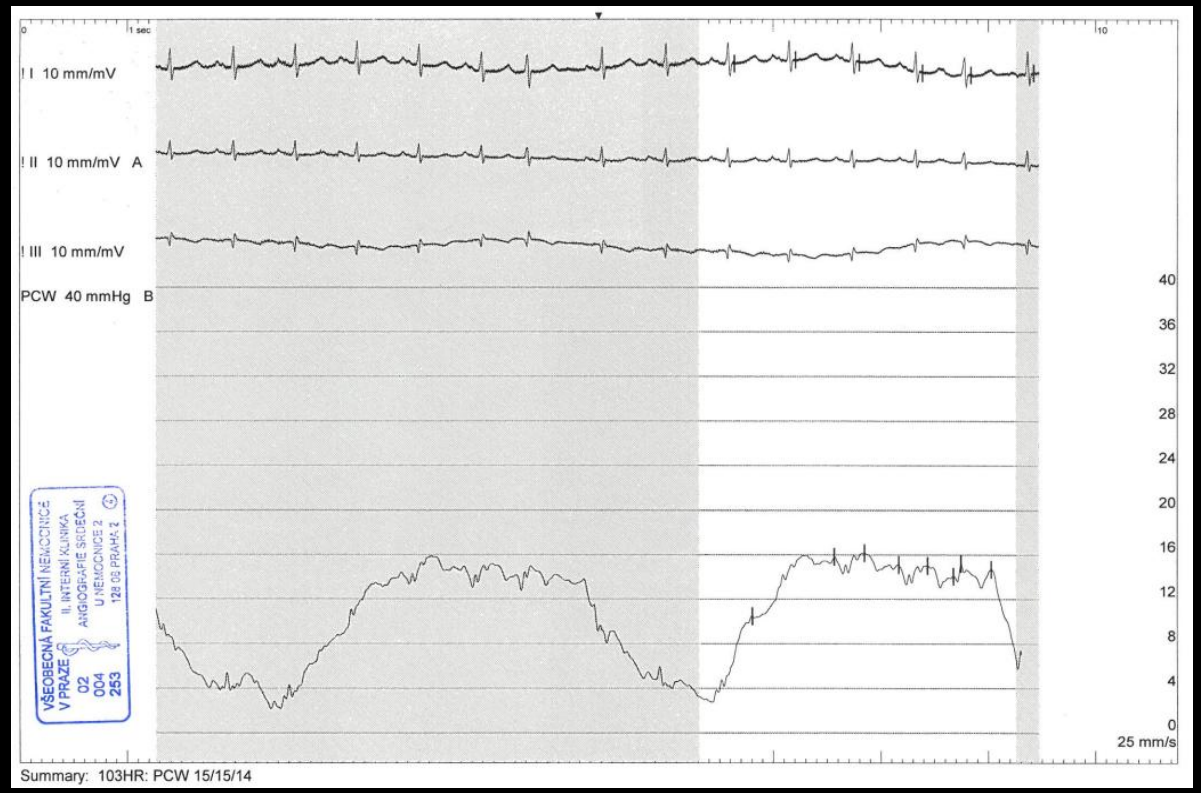




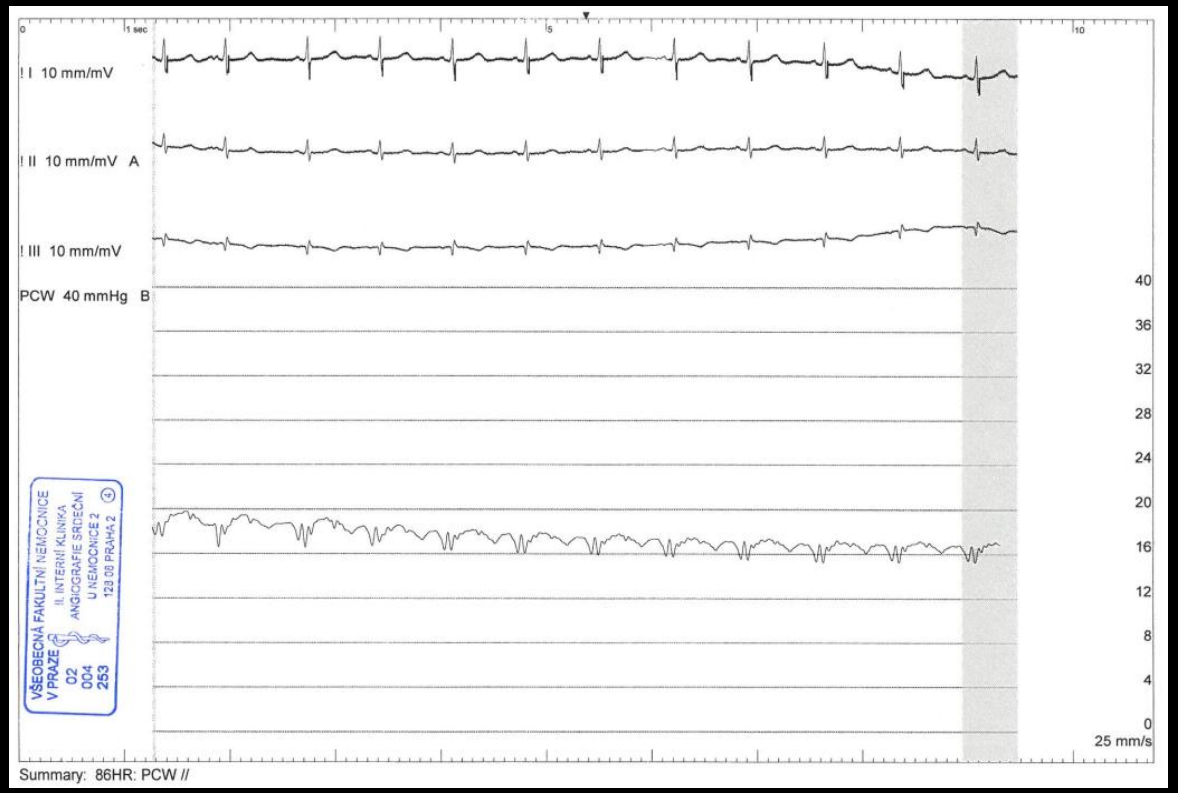


General University Hospital in Prague, Ryan JJ. Et al, *AHJ* 2012

PCWP (digital mean 14 mmHg)

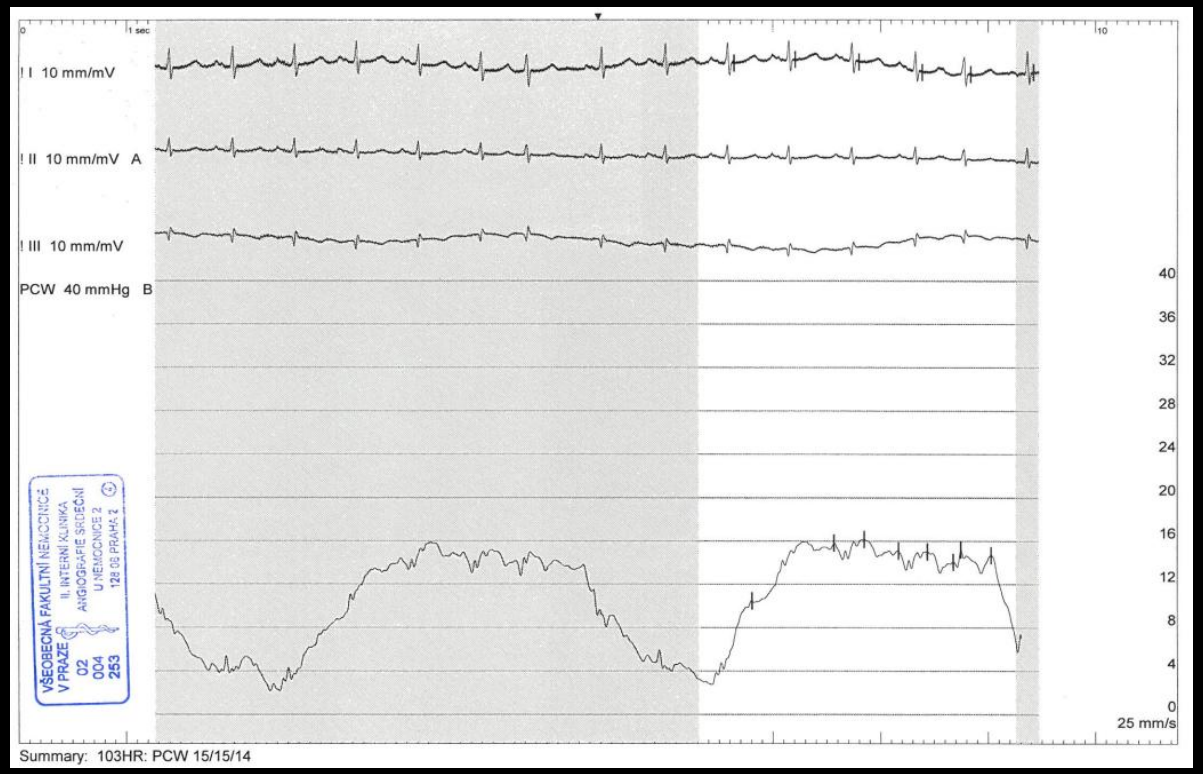


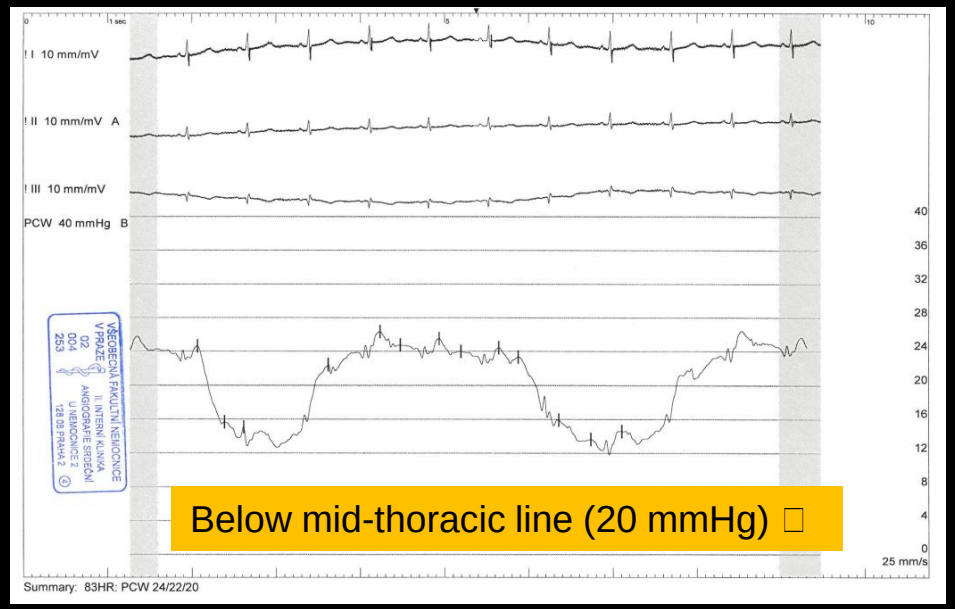
PCWP (end-exp mean 16 mmHg)



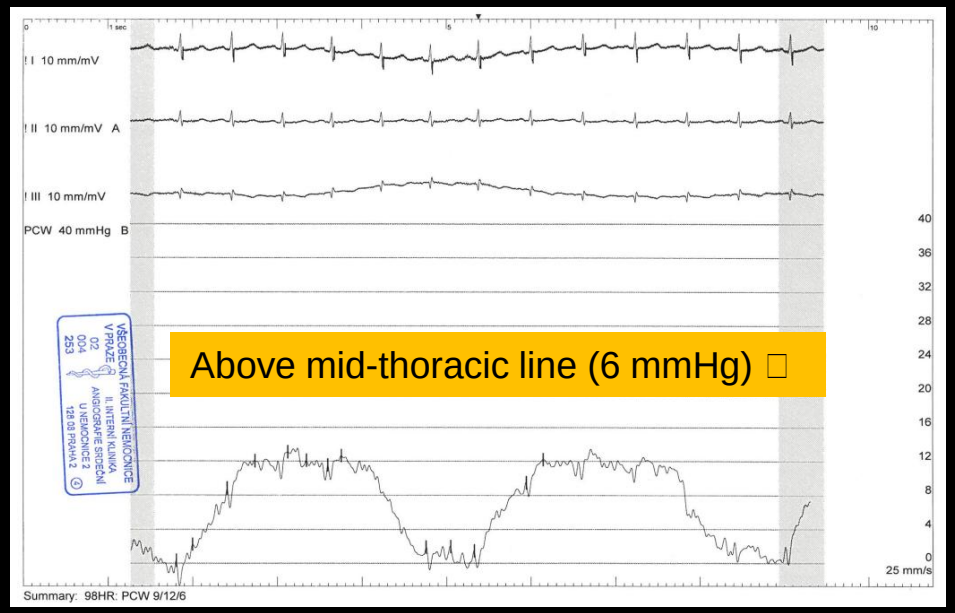
PCWP (14 mmHg), mid-thoracic line

✔





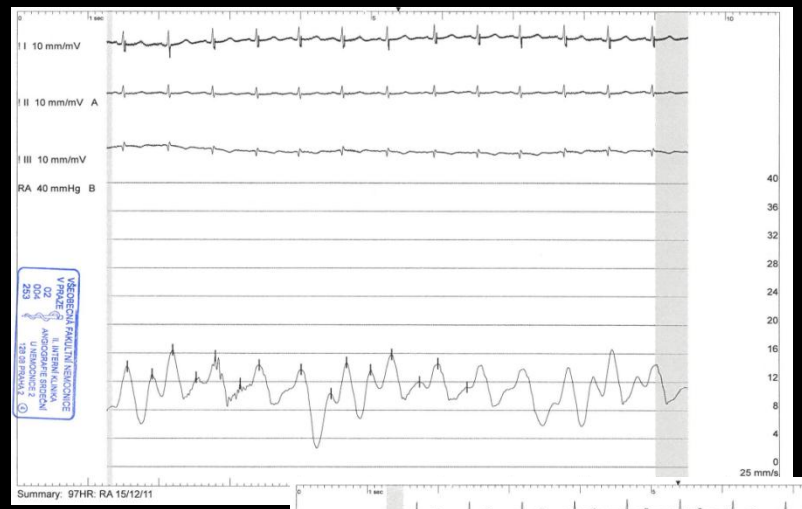
Below mid-thoracic line (20 mmHg) □

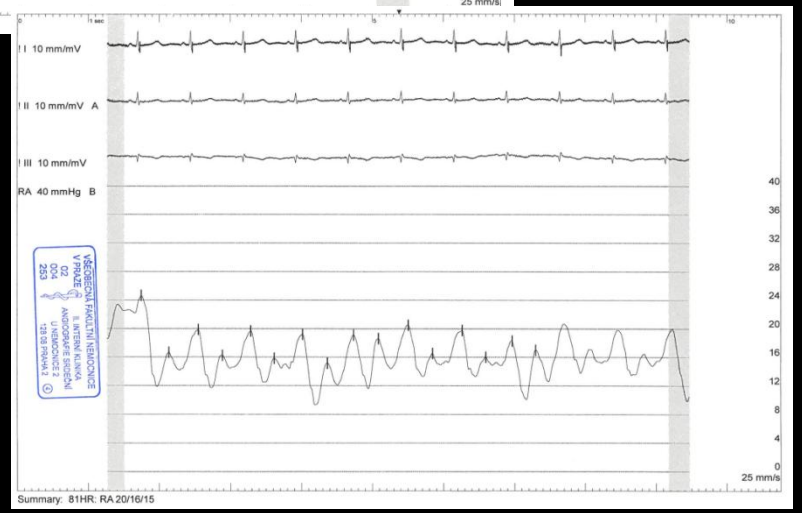


Above mid-thoracic line (6 mmHg) □

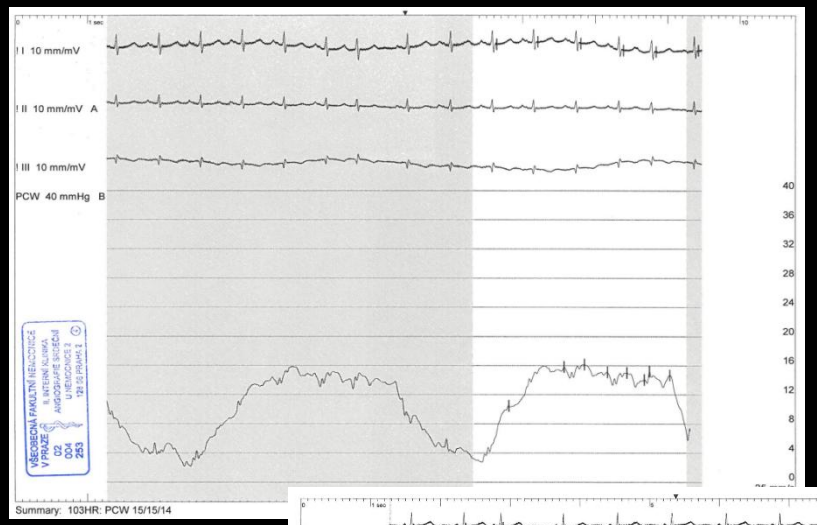
FLUID CHALLENGE

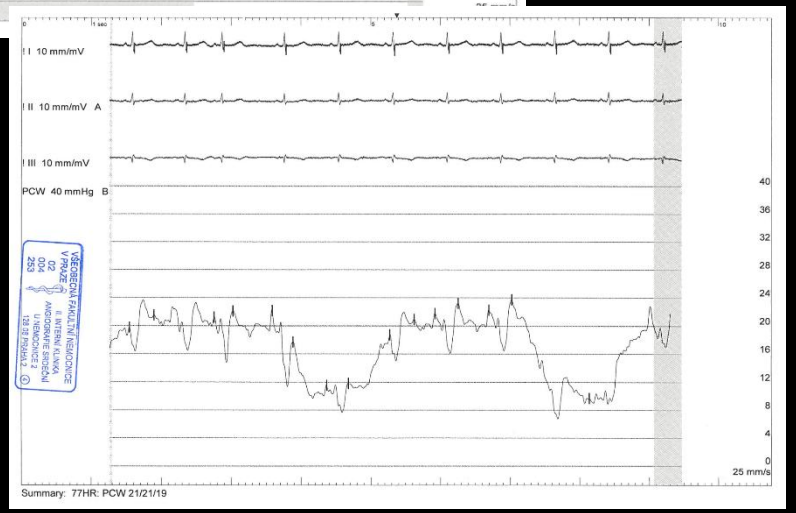
RAP before (11 mmHg)…after (15 mmHg)





PCWP before (14 mmHg)…after (19 mmHg)





SHRNUTÍ

- Nová hemodynamická definice PH (PAMP>20 mmHg, PVR > 2 WU)
- Pečlivá diferenciální diagnostika PH je zásadní pro adekvátní (specifickou) léčbu
- U skupiny PH 2 a 3 nutno odlišit pacienty s těžkou PH (PVR > 5 WU)
- U CTEPH v detekci V/Q scinti (SPECT), při hodnocení CT nutná zkušenost radiologa
- PSK je zlatým standardem v definitivní diagnostice PH (jednoduchá, spolehlivá a bezpečná)
- Pečlivá kalibrace, pečlivé hodnocení PCW, objemová výzva

European Reference Network

for rare or low prevalence complex diseases

Network
Respiratory Diseases (ERN-LUNG)

Member
General University Hospital in Prague — Czechia

pavel.jansa@vfn.cz

KURZY PLICNÍ HYPERTENZE

- REGISTRACE
- AKREDITACE
- PROGRAM 2022
- POŘADATEL KURZŮ

  Centrum pro plicní hypertenzi
  VFN Praha
- ORGANIZAČNÍ ZAJIŠTĚNÍ

V případě zájmu o tento odborný kurz
nás neváhejte kontaktovat prostřednictvím e-mailu v.strukova@gsymposion.cz,
budeme Vás informovat o nejbližším možném termínu.

6.-7. června 2025

www.pah-cz.cz