PŘETRVÁVAJÍCÍ DUŠNOST PO AKUTNÍ PLICNÍ EMBOLII

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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

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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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European Heart Journal (2022) **00**, 1–114
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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).

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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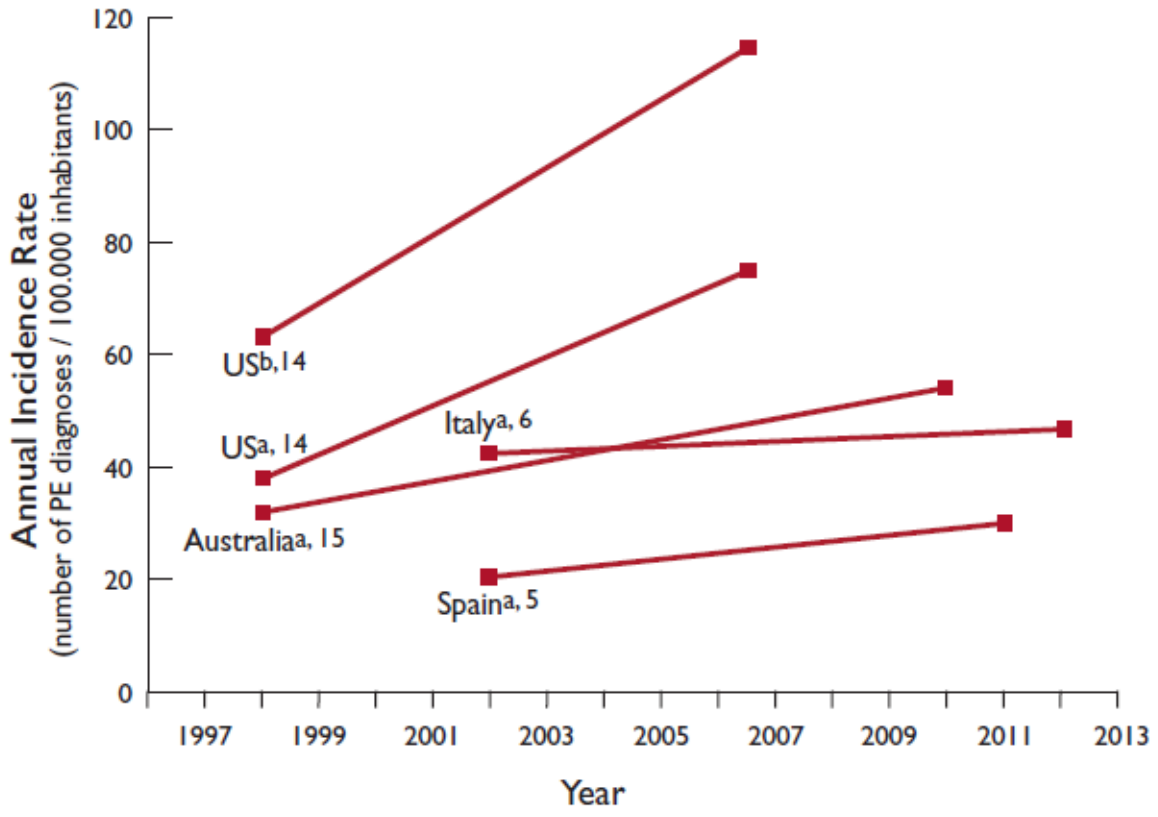
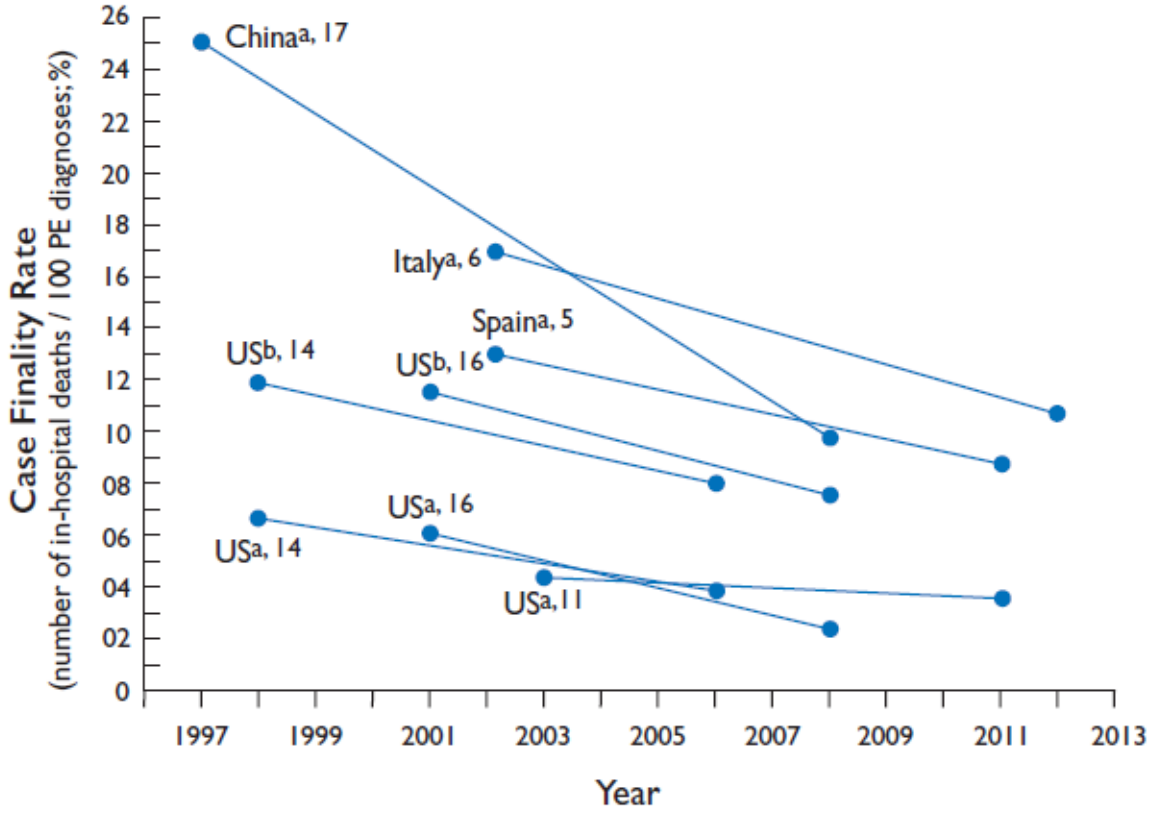
Councils: Council on Cardiovascular Genomics.

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

Patient Forum

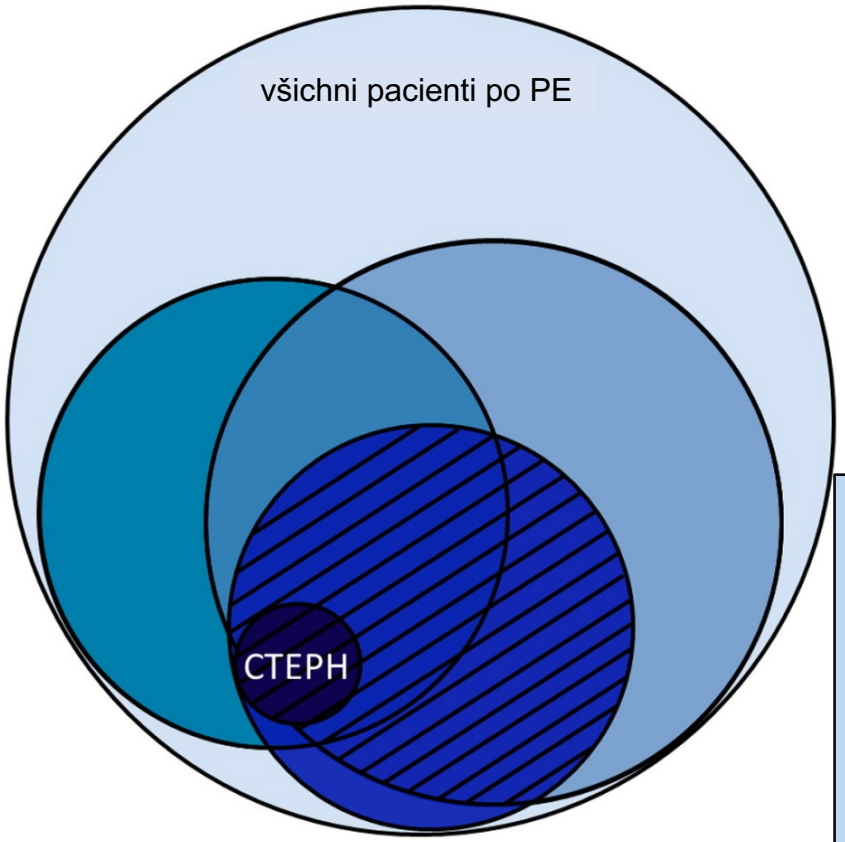
2019 ESC Guidelines for the diagnosis and management of acute pulmonary embolism developed in collaboration with the European Respiratory Society (ERS)

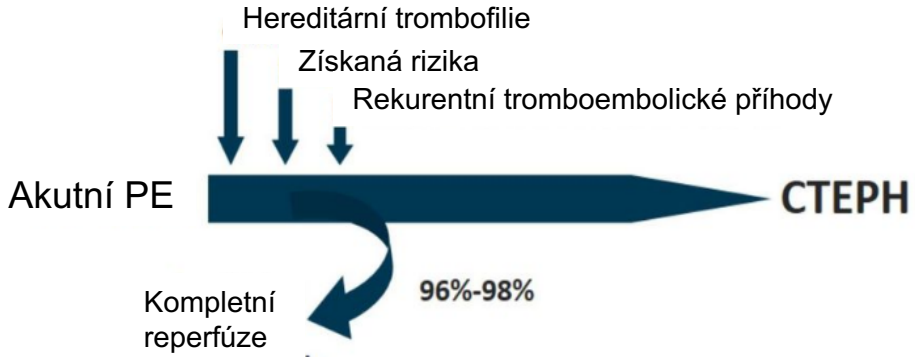




CHRONICKÉ KOMPLIKACE PO AKUTNÍ PLICNÍ EMBOLII

- 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

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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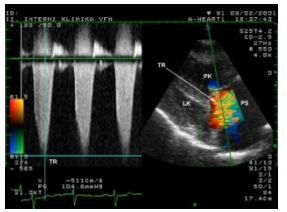
EHJ 2022, *ERJ* 2022

Suspekce

Plicní hypertenze podle ECHO možná nebo pravděpodobná



Postižení myokardu nebo chlopní levého srdce

Skupina 2

Bez postižení myokardu nebo chlopní levého srdce

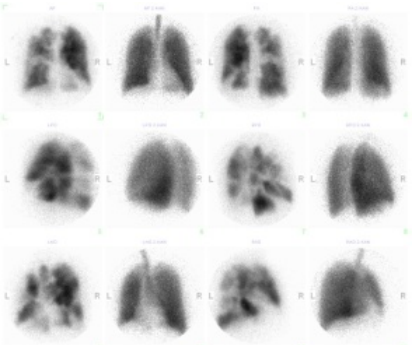
Detekce

Plicní funkce

Výrazná redukce

Skupina 3

Normální nebo nevýznamná redukce



V/Q plicní scintigrafie

Nesegmentární defekty

hemodynamika

Skupina 1

Segmentární defekty

CT angiografie, angiografie, hemodynamika

Skupina 4

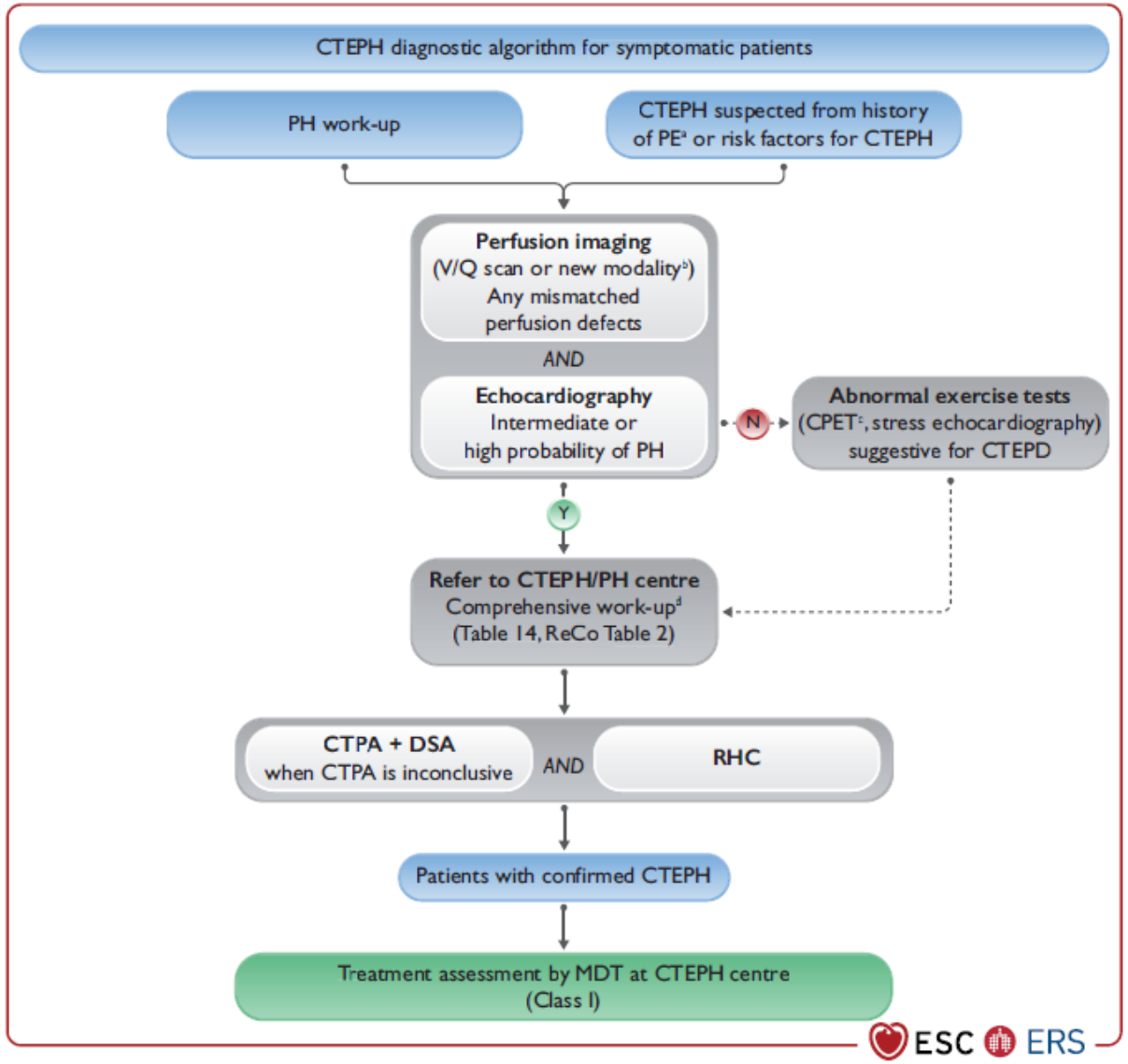
Konfirmace

DIAGNOSTICKÝ ALGORITMUS CTEPH (2022)

Suspekce

Detekce

Konfirmace



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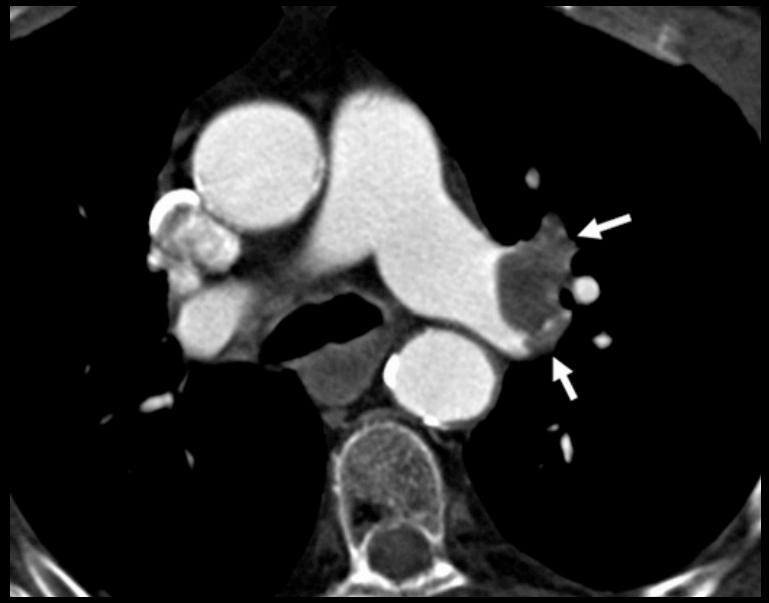
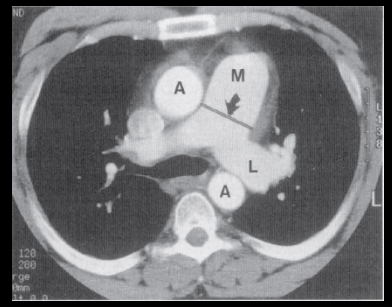
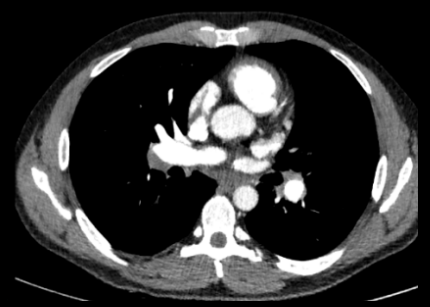
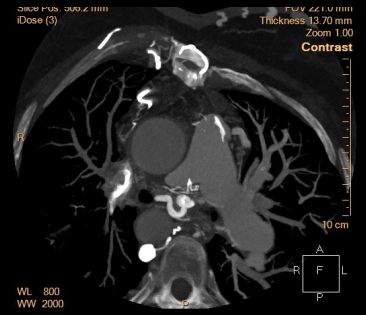
PLICNÍ EMBOLIE

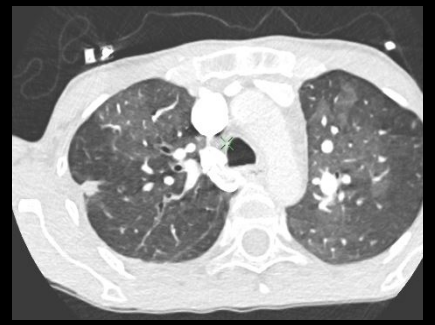
CTEPH

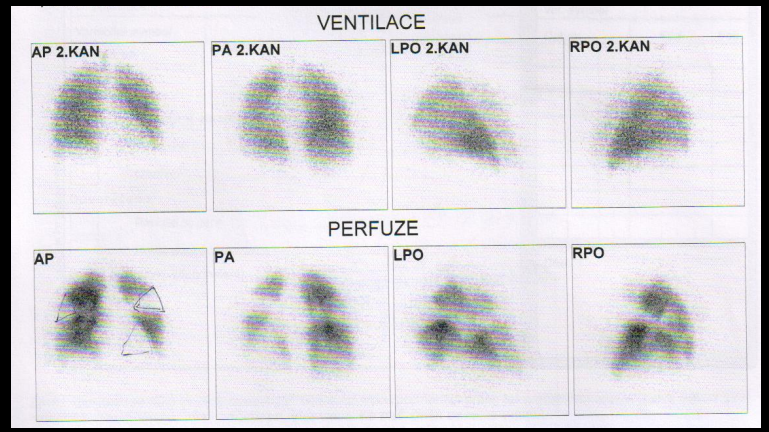


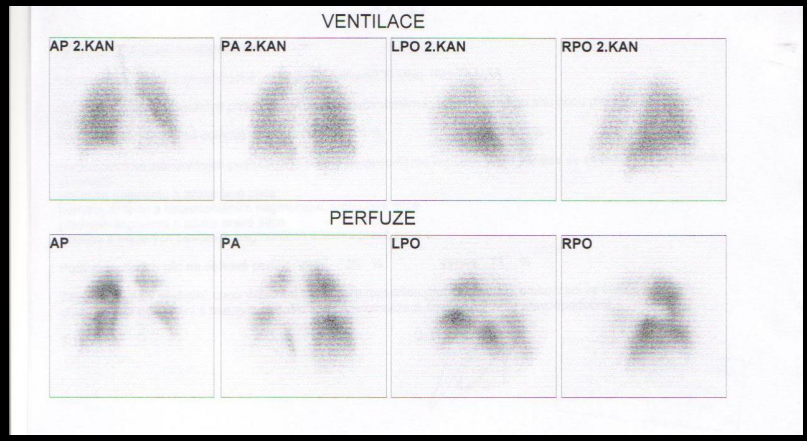






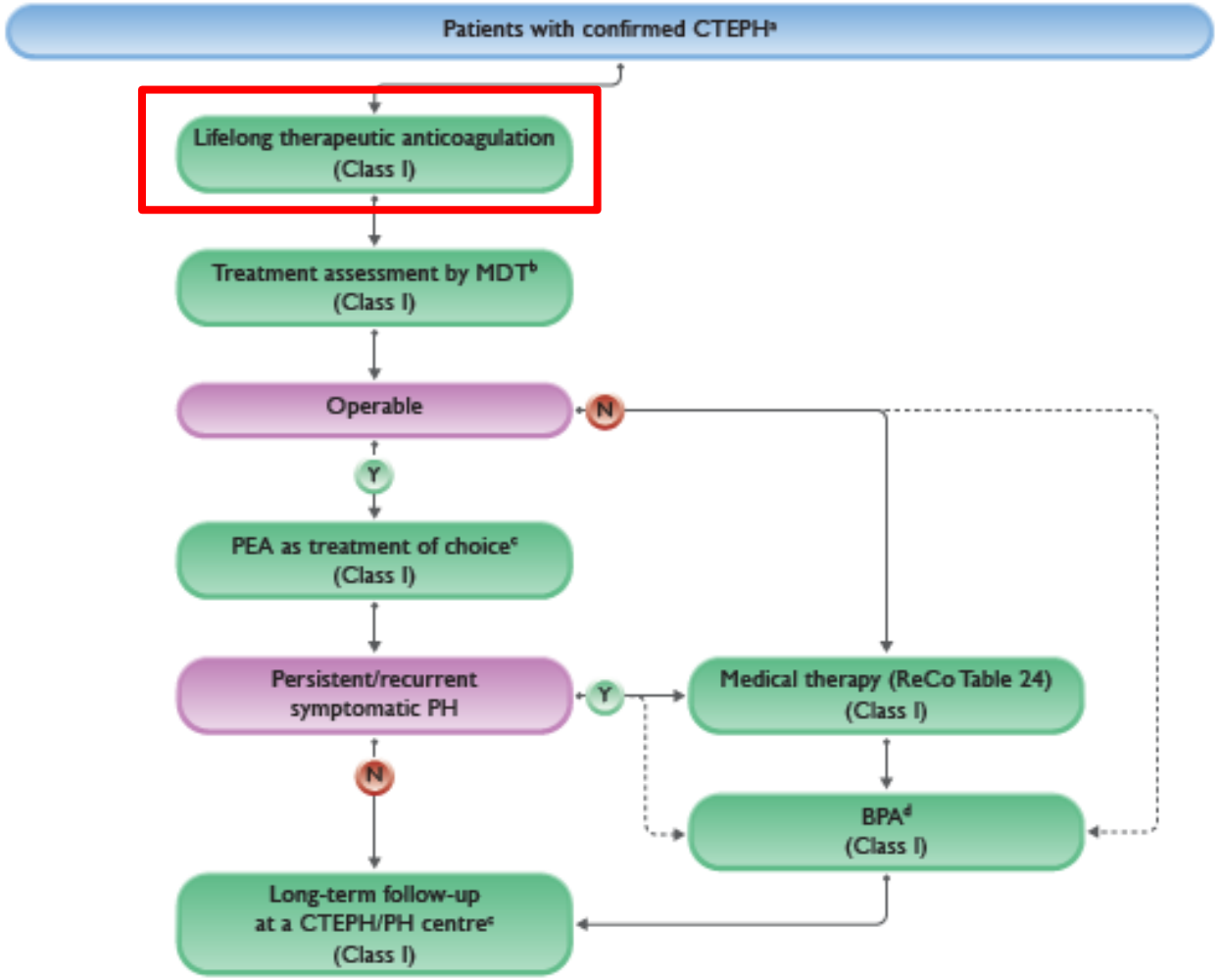


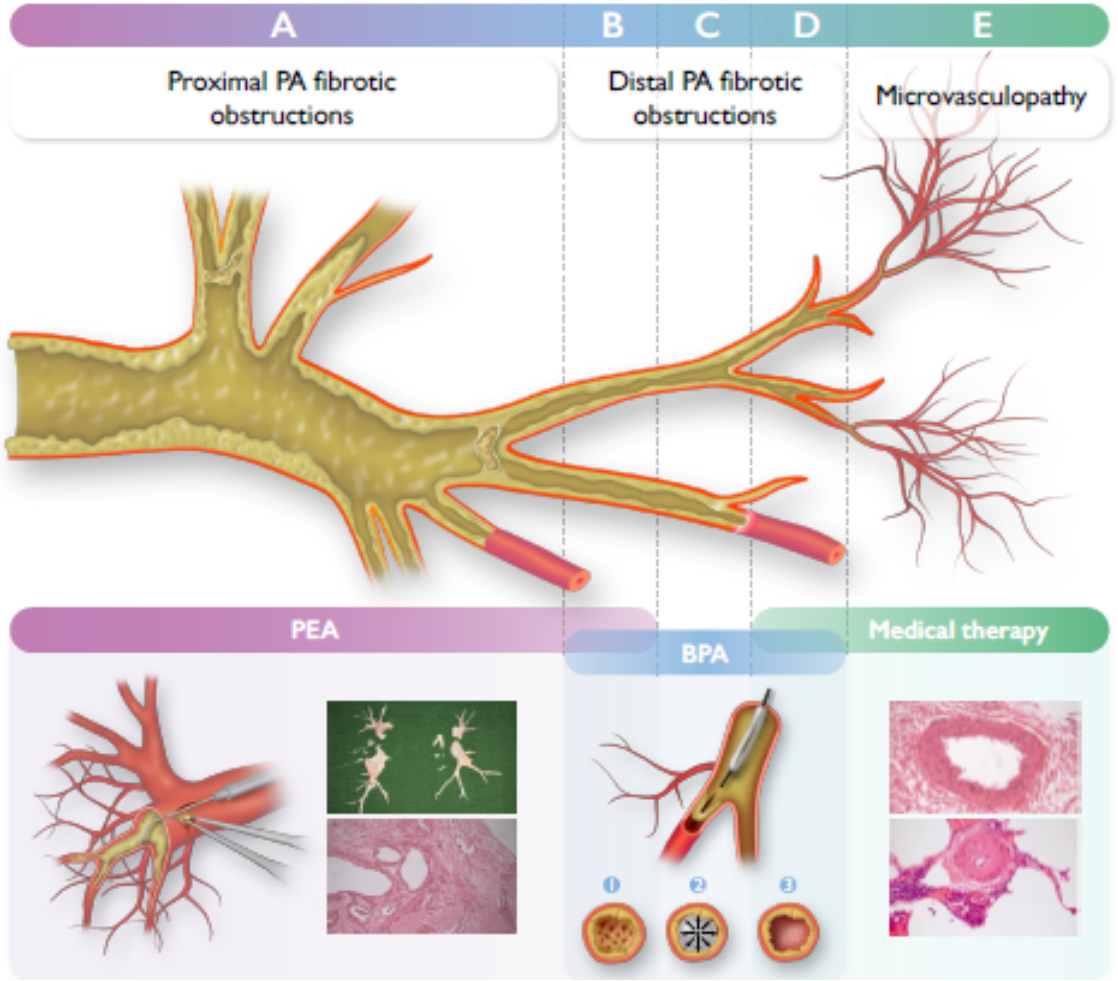




Zdroj: VFN

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





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Doporučené postupy Evropské kardiologické společnosti (ESC)/Evropské respirační společnosti (ERS) pro diagnostiku a léčbu plicní hypertenze, verze 2022.

Tabulka 24 doporučení – Doporučení pro chronickou tromboembolickou plicní hypertenzi a chronické tromboembolické plicní onemocnění bez plicní hypertenze

Doporučení	Třída[a]	Úroveň[b]
CTEPH		
U všech pacientů s CTEPH je doporučeno doživotní podávání terapeutických dávek antikoagulační léčby.	I	C
U všech pacientů s CTEPH je doporučeno testování antifosfolipidového syndromu.	I	C
U pacientů s CTEPH a antifosfolipidovým syndromem je doporučena antikoagulace pomocí VKA.	I	C
Je doporučeno, aby všichni pacienti s CTEPH byli vyšetřeni týmem pro CTEPH za účelem posouzení multimodální léčby.	I	C
PEA je doporučena jako léčba volby u pacientů s CTEPH a fibrózní obstrukcí v plicních tepnách přístupných chirurgickému zákroku.	I	B
BPA je doporučena u pacientů, kteří jsou technicky inoperabilní nebo mají reziduální PH po PEA a distální obstrukce vhodné pro BPA.	I	B
Riociguat je doporučen u symptomatických pacientů s inoperabilní CTEPH nebo přetrvávající/recidivující PH.	I	B
Dlouhodobé sledování je doporučeno po PEA a BPA, stejně jako u pacientů s CTEPH na medikamentózní léčbě.	I	C

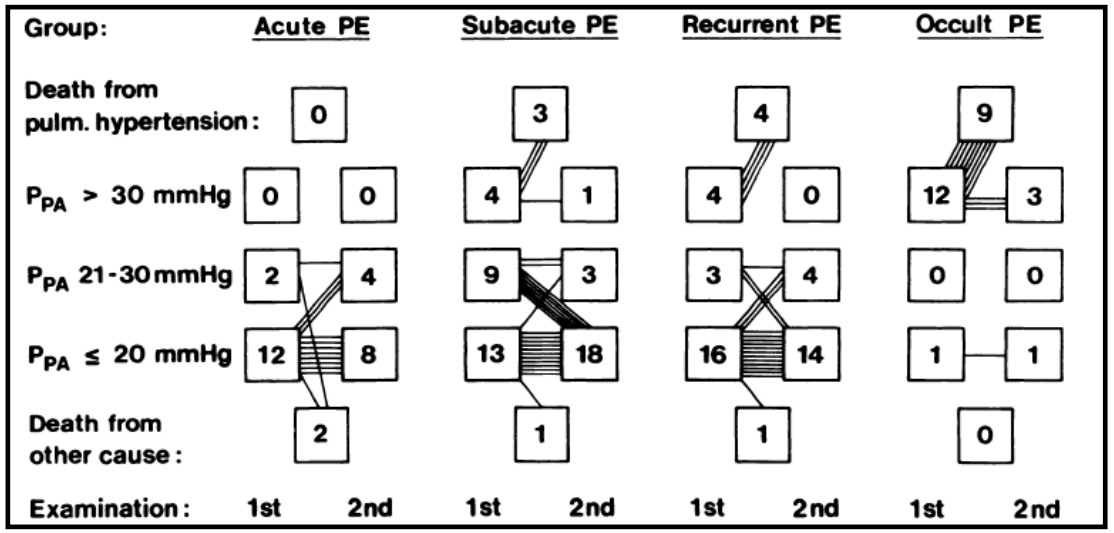
Humbert M et al. *Eur Heart* J 2022 Oct 11;43(38):3618-3731, Jansa P et al. *Cor Vasa* 2023;65:236–286

CTEPH – PROGNÓZA NELÉČENÉHO ONEMOCNĚNÍ

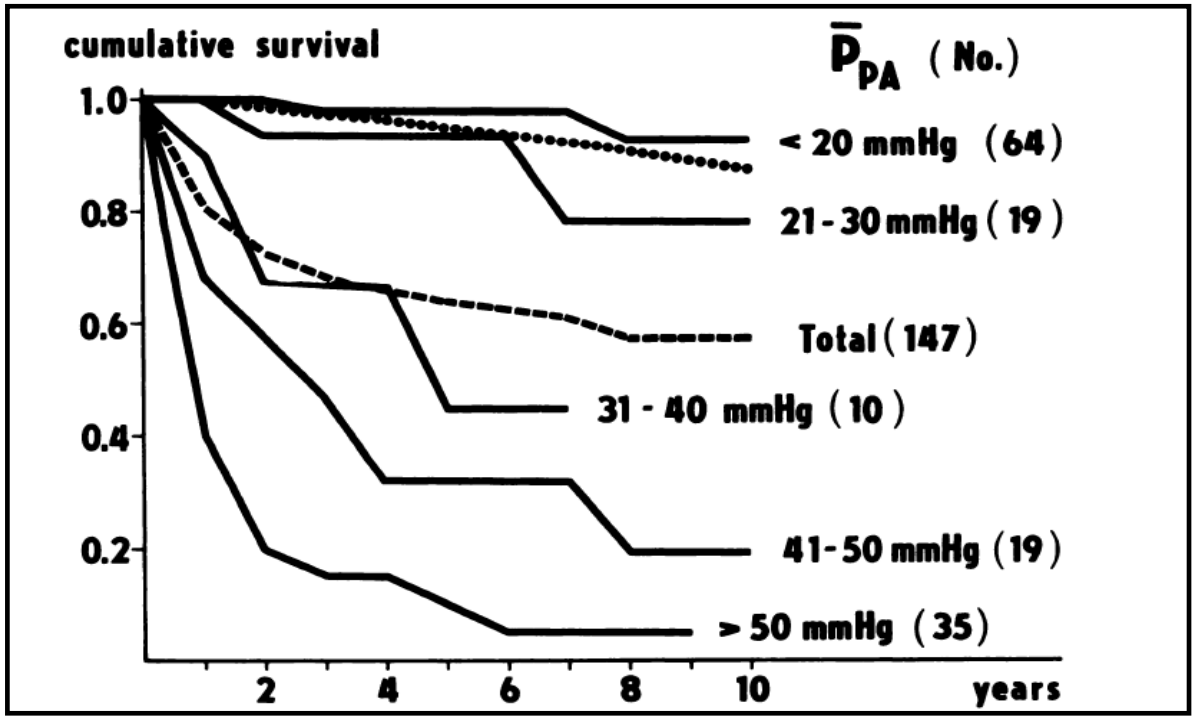
Longterm Follow-up of Patients with Pulmonary Thromboembolism*

Late Prognosis and Evolution of Hemodynamic and Respiratory Data

N=76, M/F = 50/26, věk 48.2 roku
hemodynamické vyšetření 1964-1979

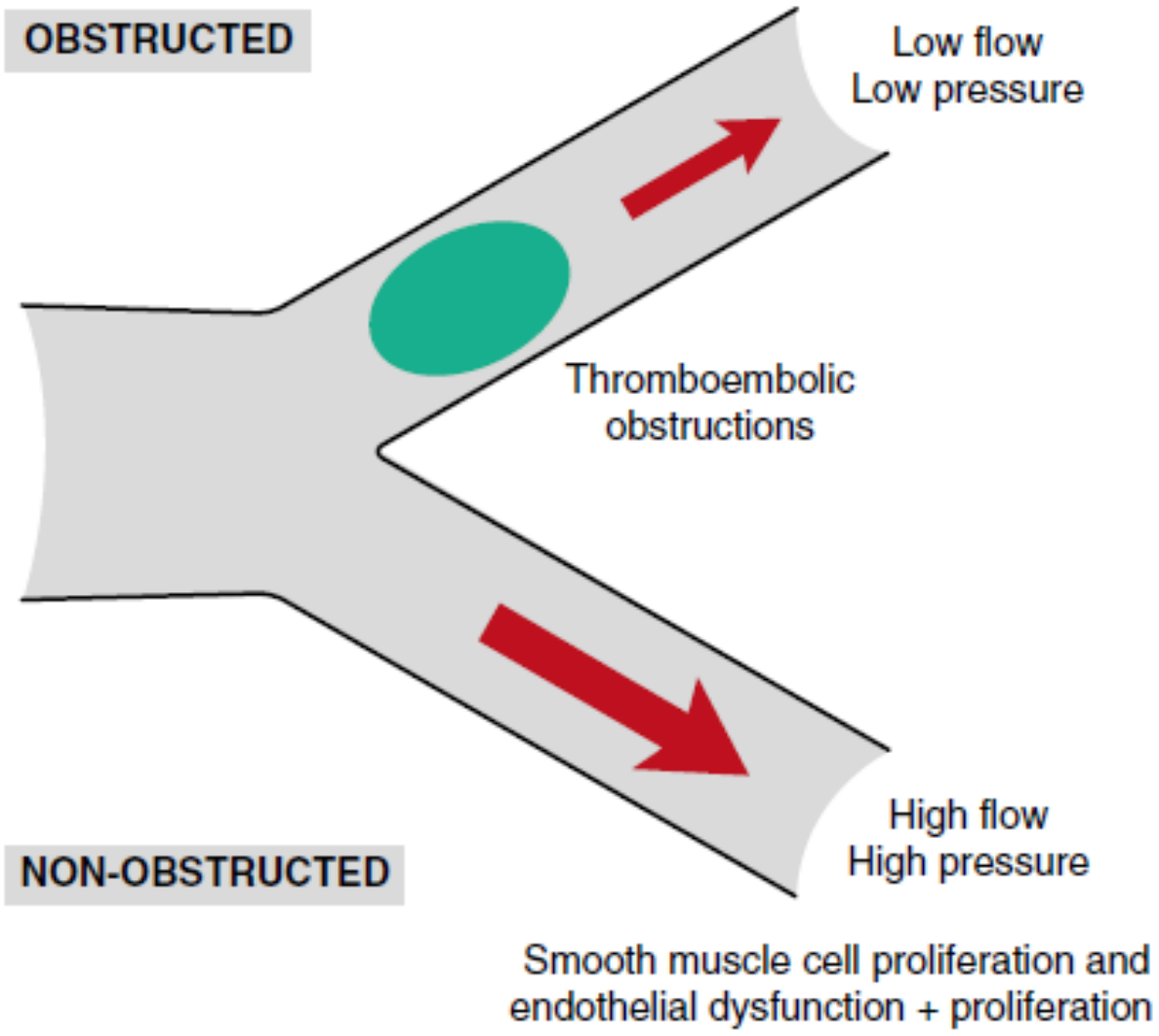


5leté přežití: PAMP > 50 mmHg 10%

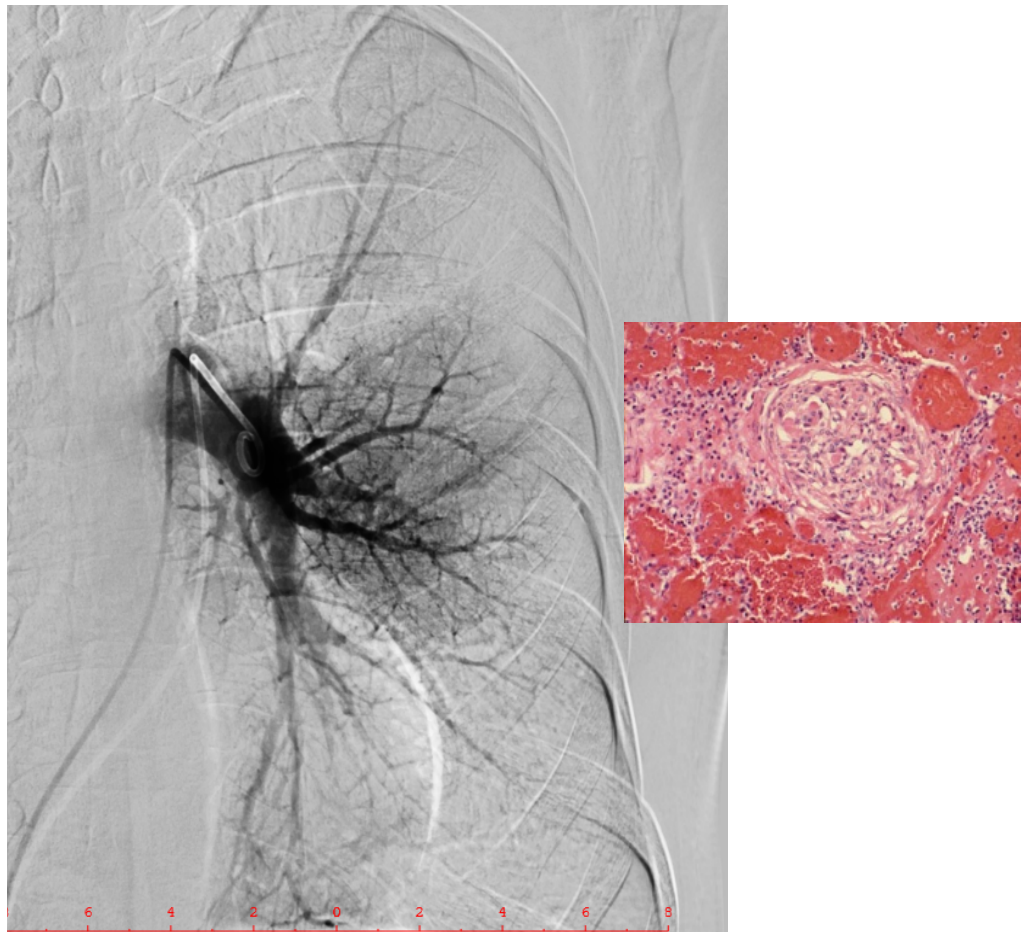


Riedel M, Stanek V, Widimsky J, Prerovsky I, *Chest* 1982, 81: 151-8

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

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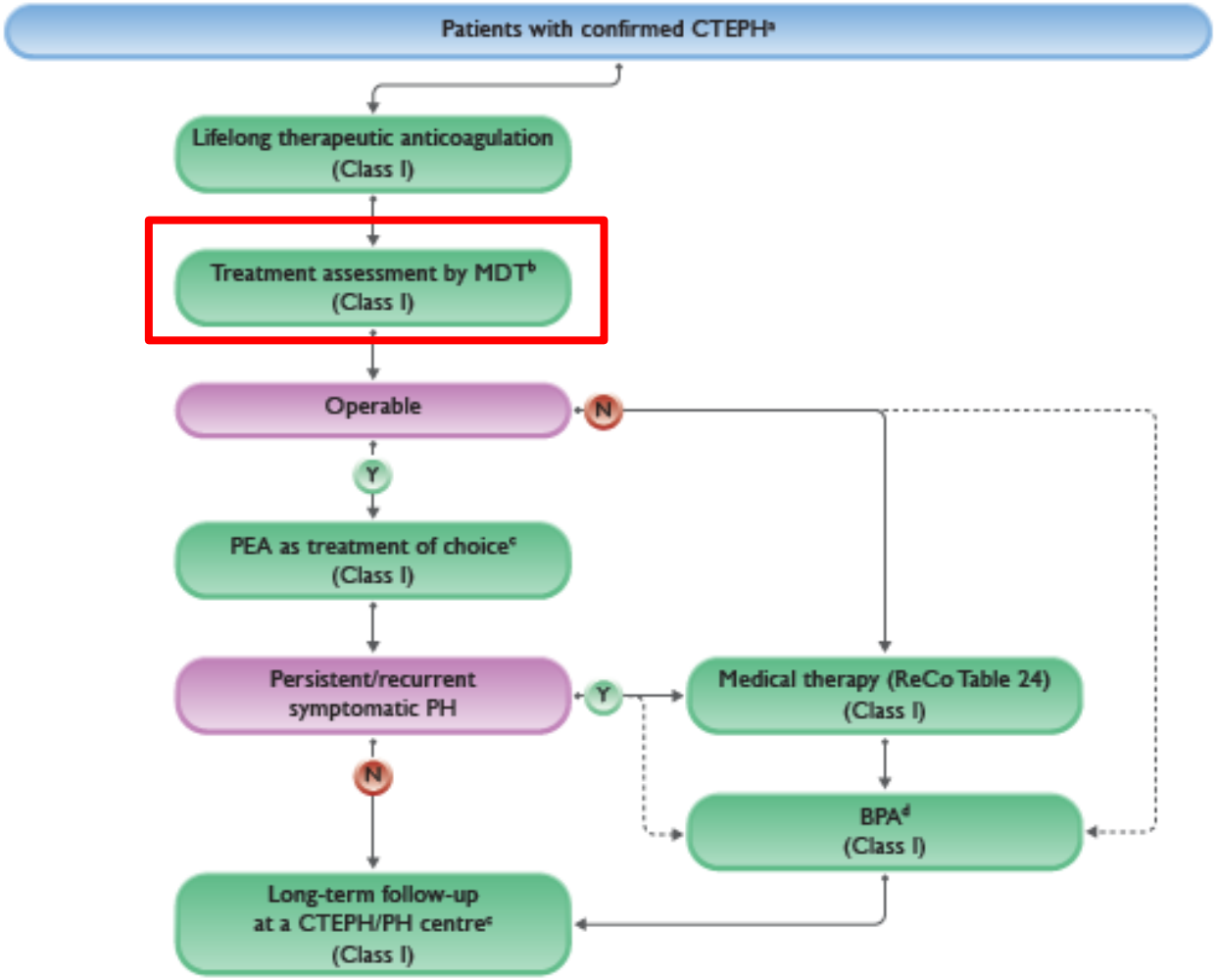

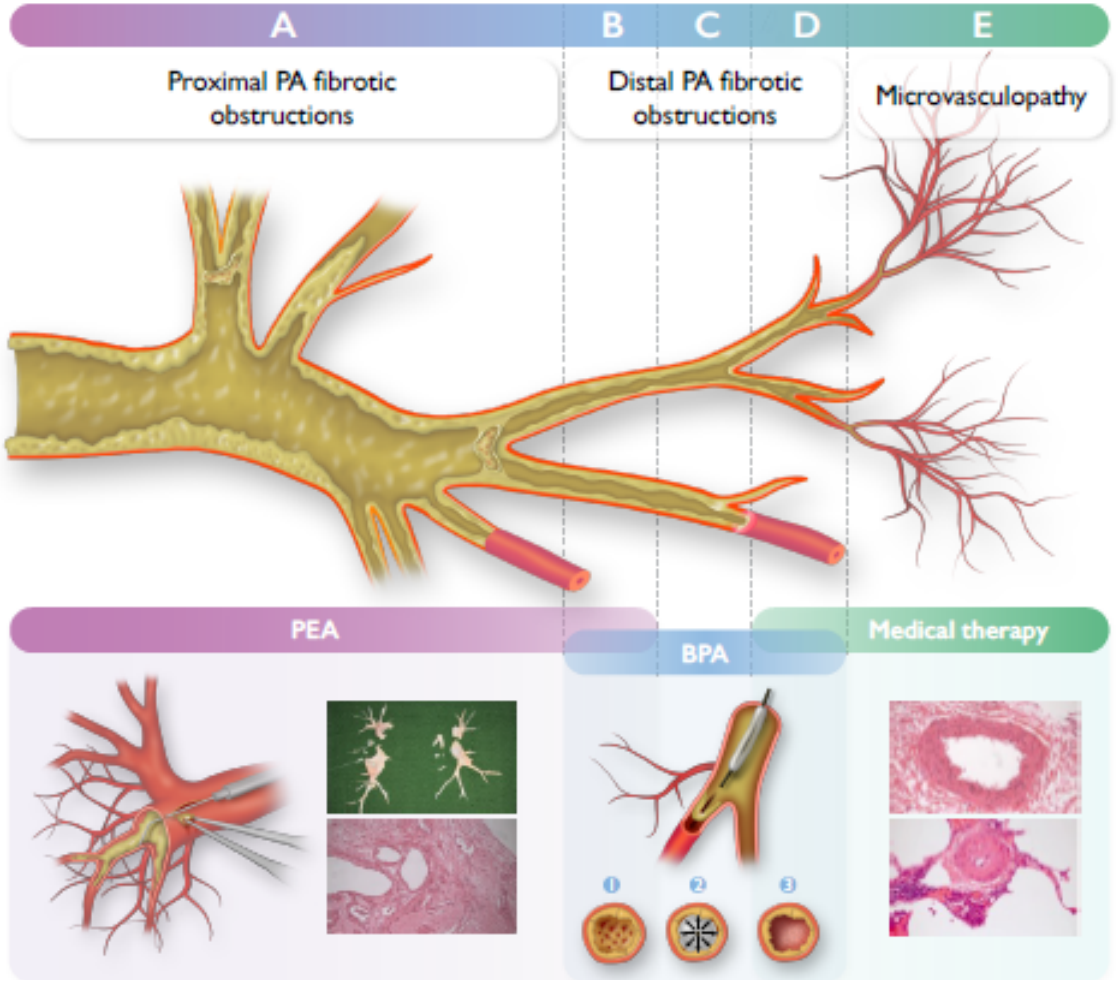

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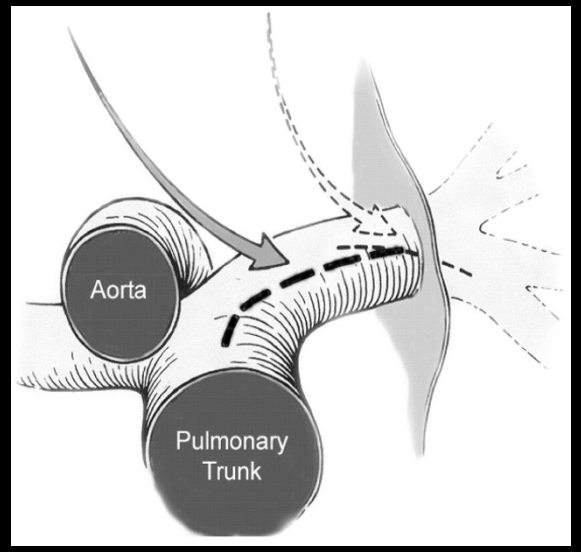
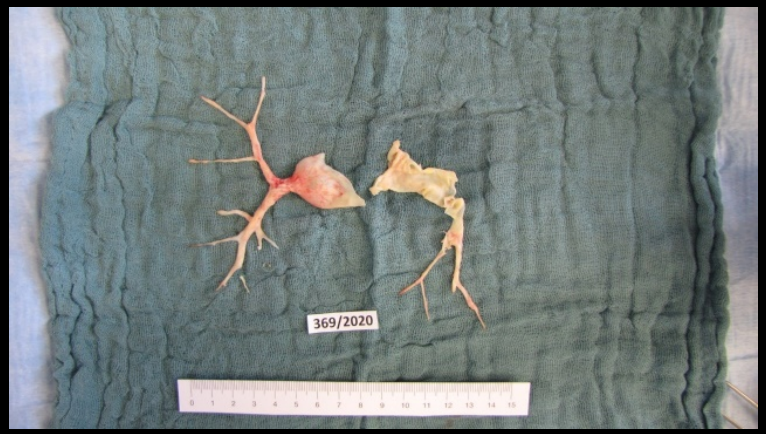
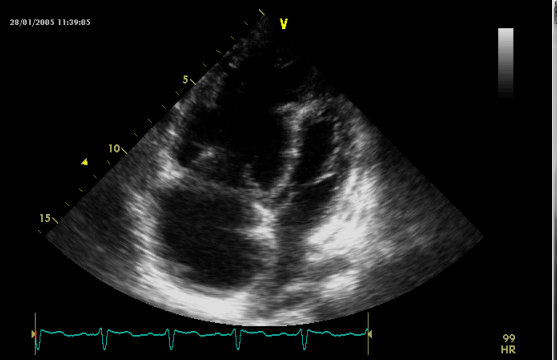
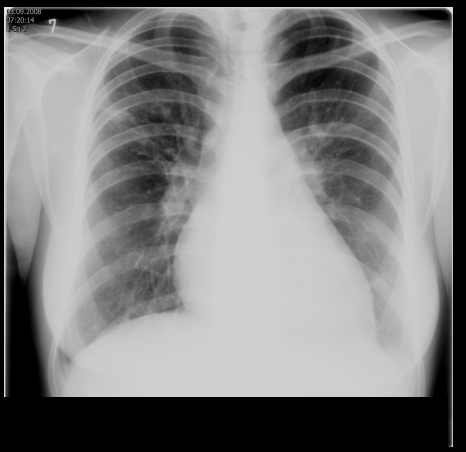
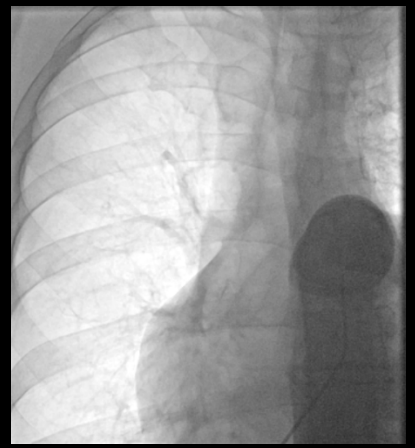
PLICNÍ ENDARTEREKTOMIE (PEA)

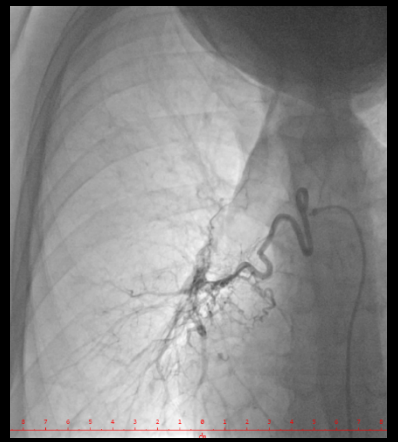


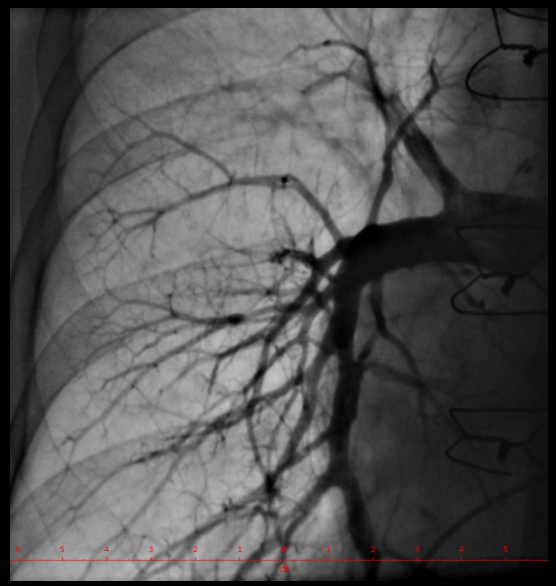


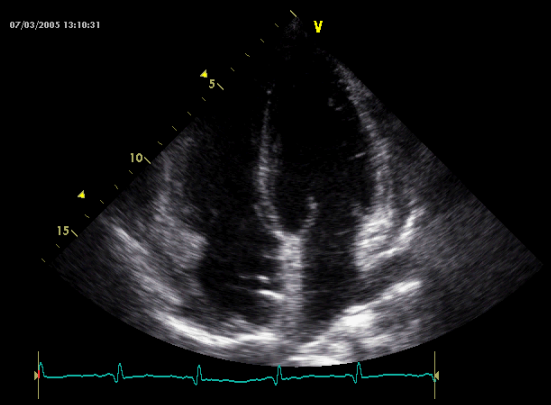


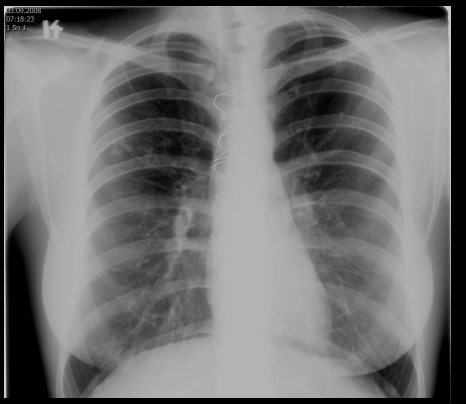








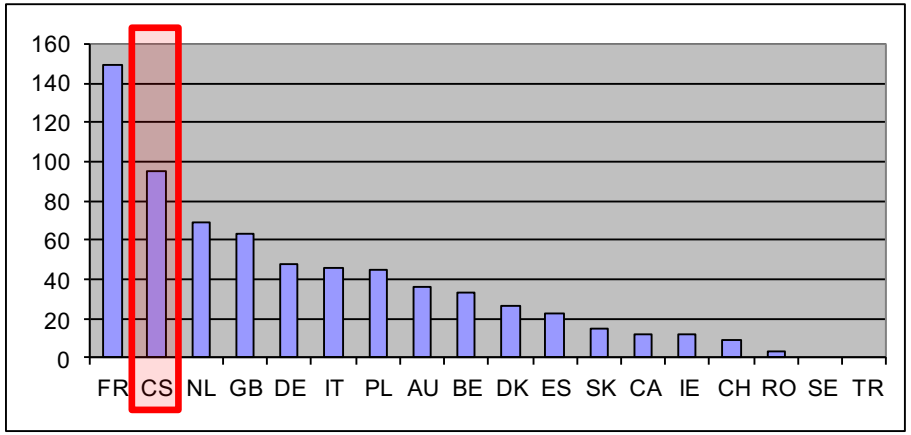


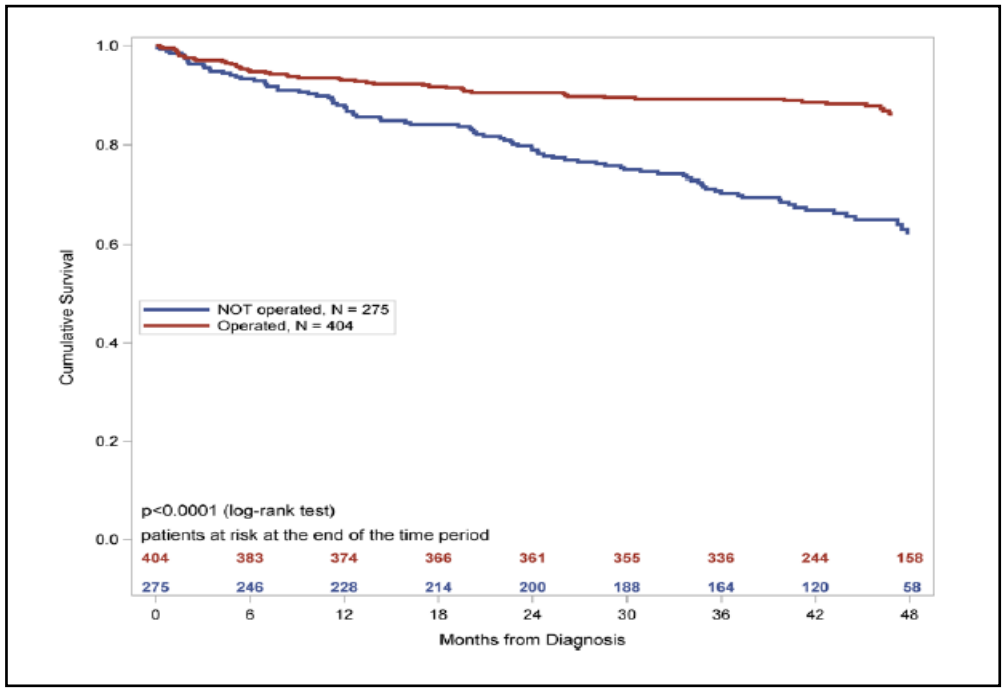


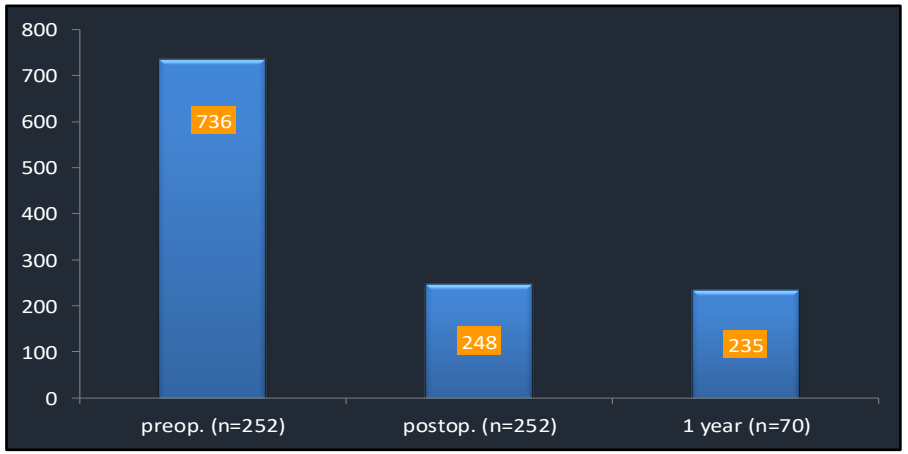
Zdroj: Všeobecná fakultní nemocnice v Praze

INTERNATIONAL CTEPH REGISTRY

N=679 (nově diagnostikovaní 2007-2009, Evropa+Kanada)
NYHA FC III+IV 81.4 %, věk 63 let (51; 72)
Operováno 56.8 %
PAMP 47 (38; 55) mmHg, CI 2.2 (1.8; 2.7) $L.min^{-1}.m^{-2}$
PVR 709 (480; 988) $dyn.s.cm^{-5}$







Pepke Zaba J et al. *Circulation* 2011 Nov 1;124(18):1973-81; Delcroix M et al. *J Thorac Cardiovasc Surg* 2011;141:702-1

EFEKT CHIRURGICKÉ LÉČBY CTEPH NA PŘEŽITÍ

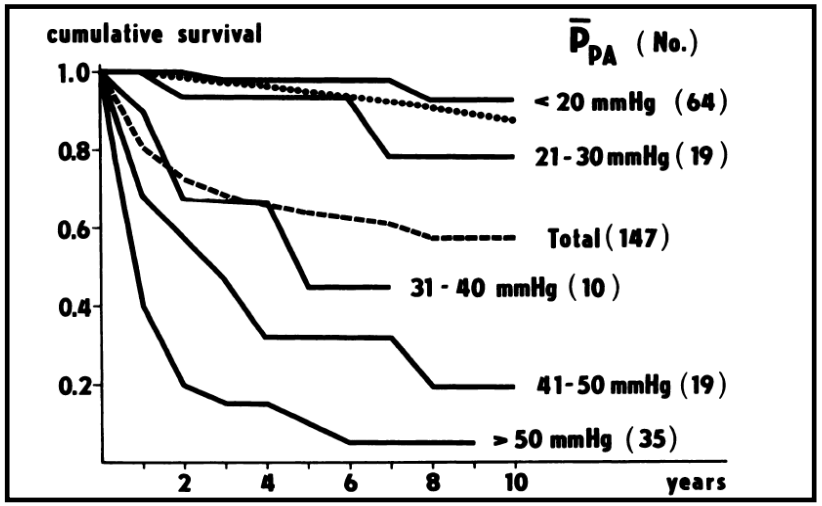
N=623 (nově diagnostikovaní 2003-2019)
N=456 (nově diagnostikovaní 2003-2016), NYHA III+IV 91.8 %, věk 63.2 (49; 77), operováno 52.1 %
PAMP 50.0 (34; 81) mmHg, CI 2.1 (1; 4) $L.min^{-1}.m^{-2}$, PVR 682.4 (244; 2197) $dyn.s.cm^{-5}$

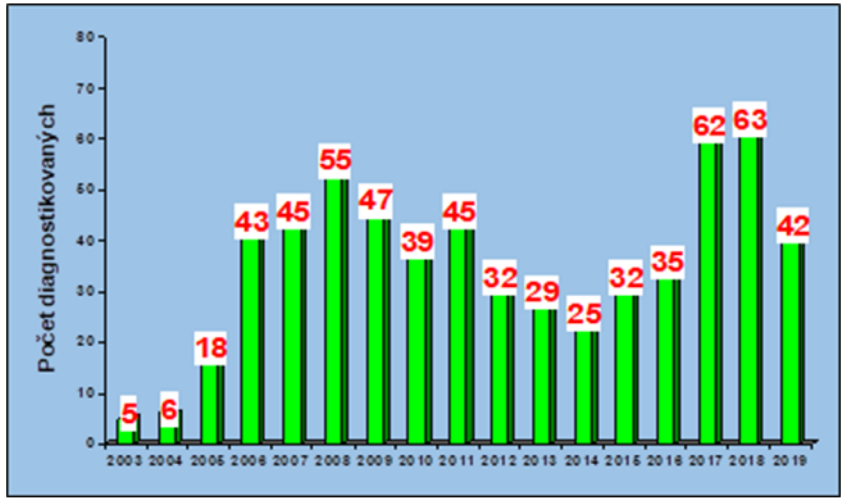
Do roku 2003

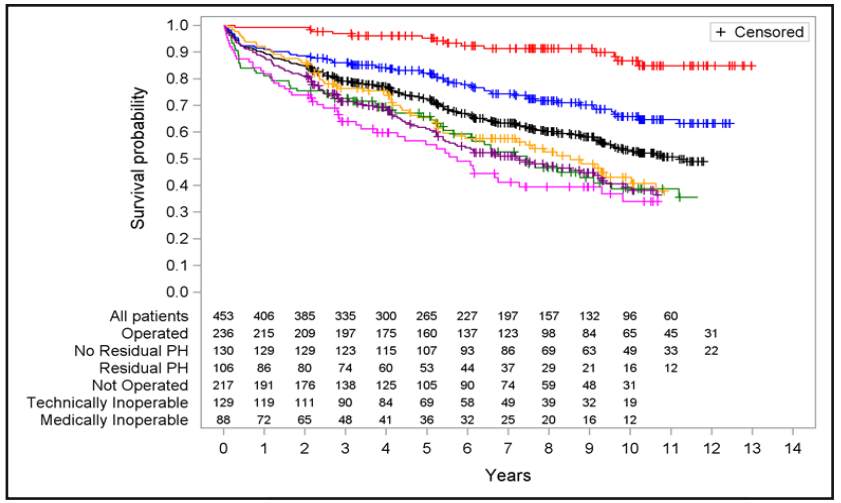
5leté přežití: PAMP > 50 mmHg 10%

2004-2016

5leté přežití: PEA 95 % vs ostatní 55-65 %

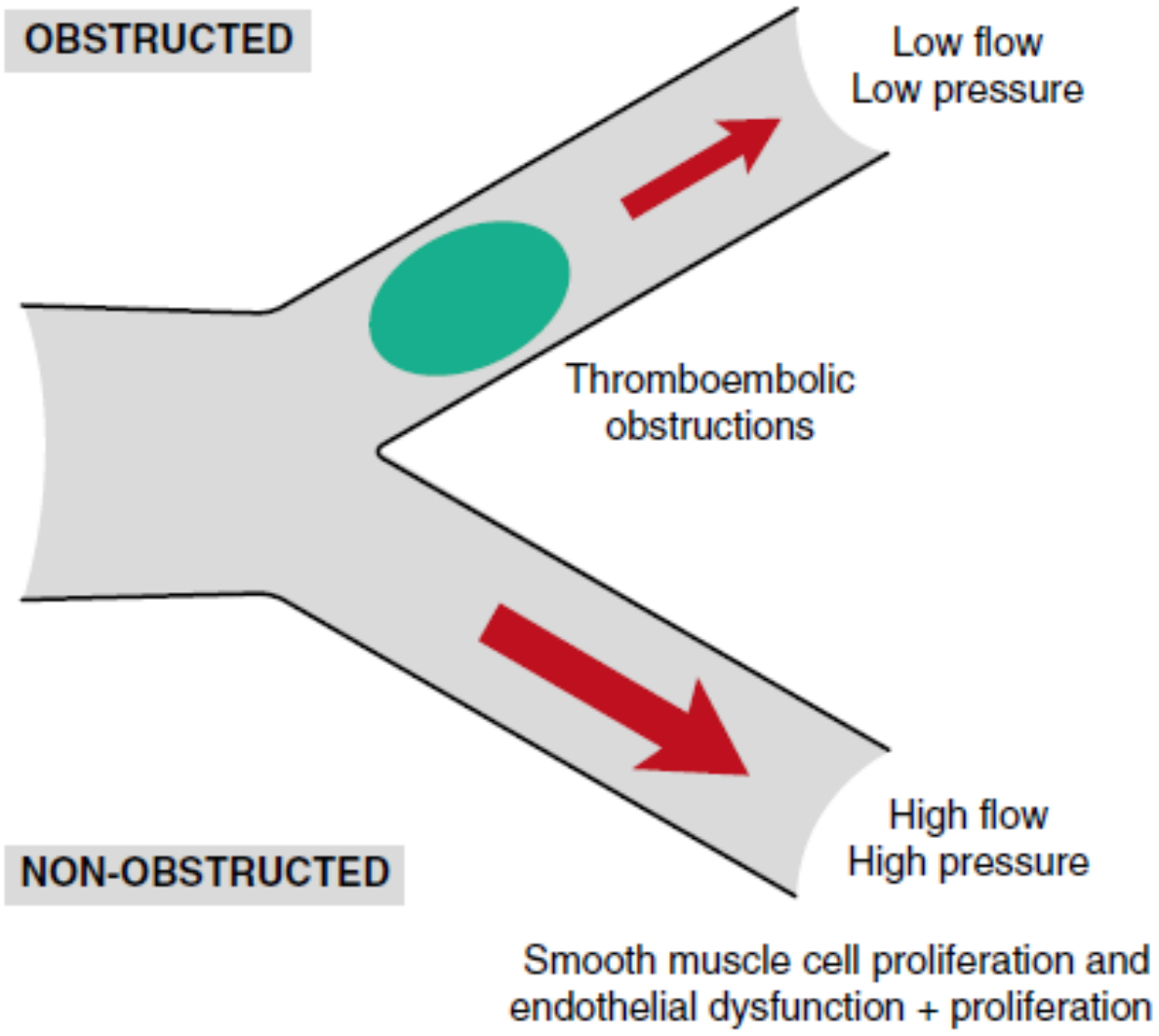




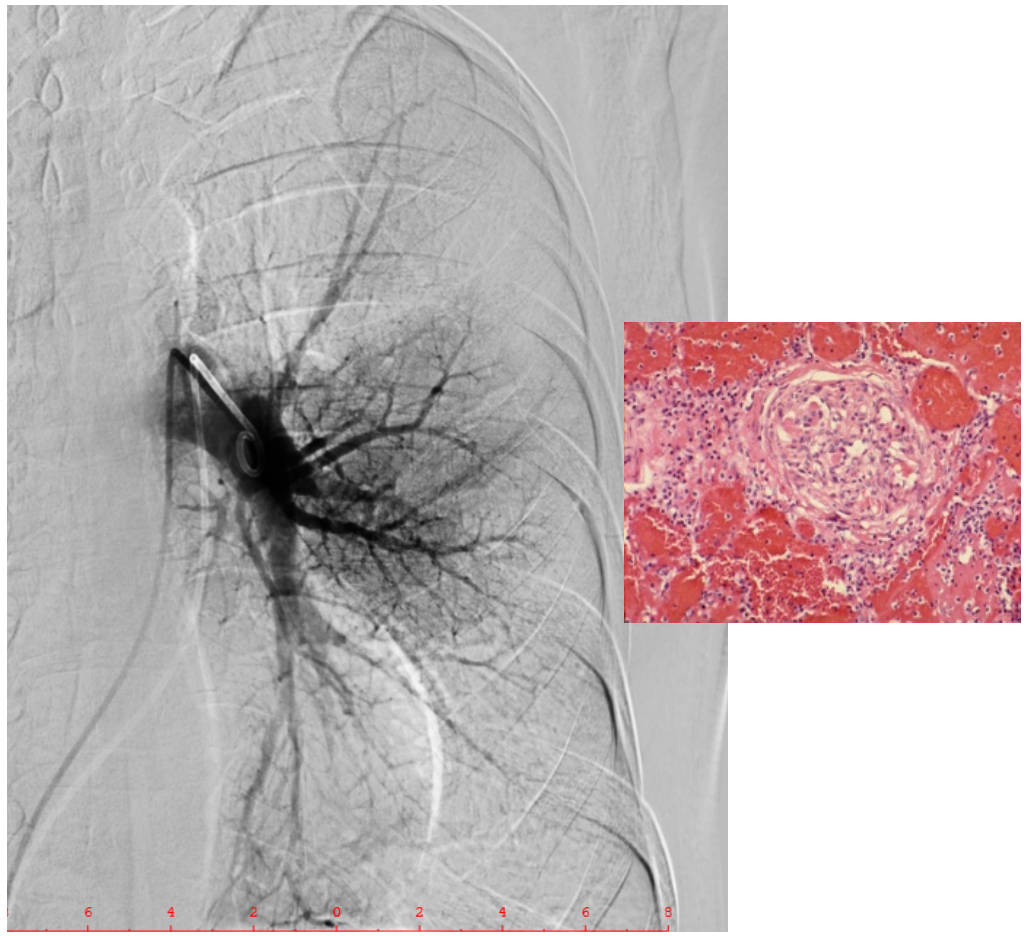


Riedel M, Stanek V, Widimsky J, Prerovsky I, *Chest* 1982, 81: 151-8
Jansa P et al. Epidemiology of chronic thromboembolic pulmonary hypertension (CTEPH) in the Czech Republic. *Pulmonary Circulation* 2022;12:e12038

PATOFYZIOLOGIE CTEPH

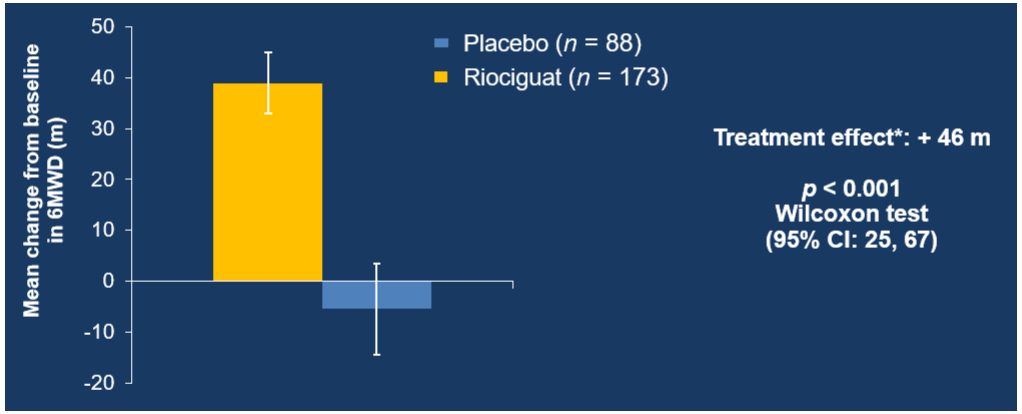


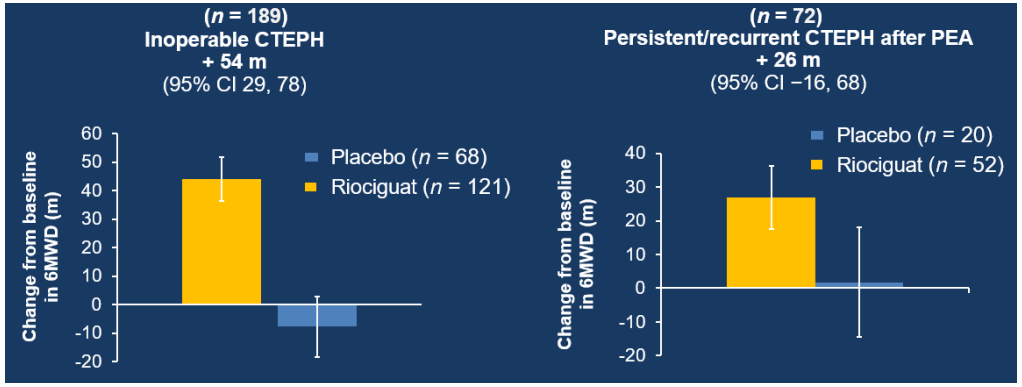
Smooth muscle cell proliferation and
endothelial dysfunction + proliferation



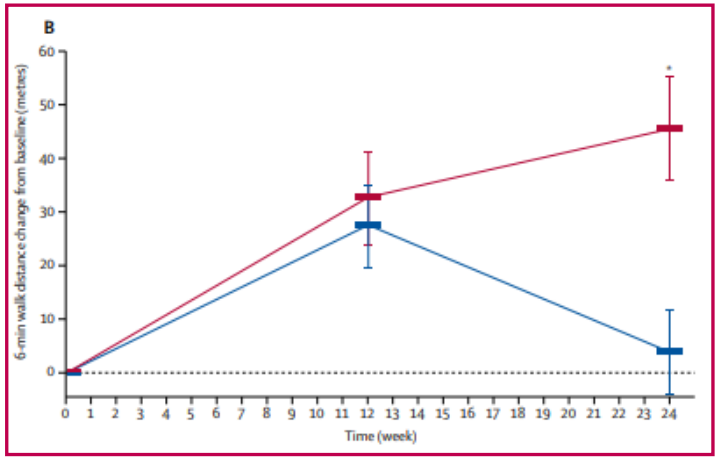
Lang et al. *Ann Am Thorac Soc* 2016 Vol 13, Supplement 3, pp S215–S221

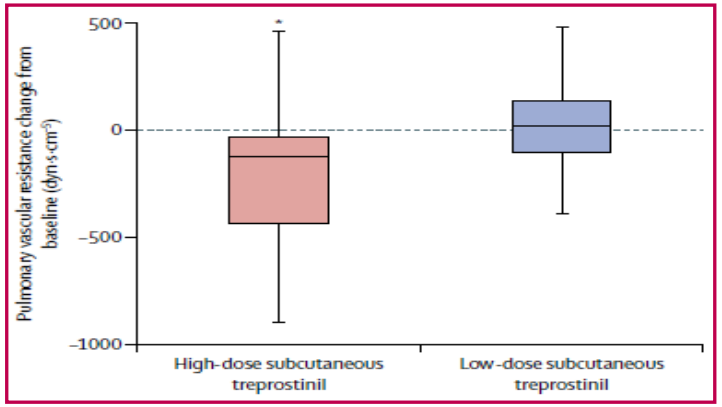
REGISTROVANÁ FARMAKOTERAPIE CTEPH





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

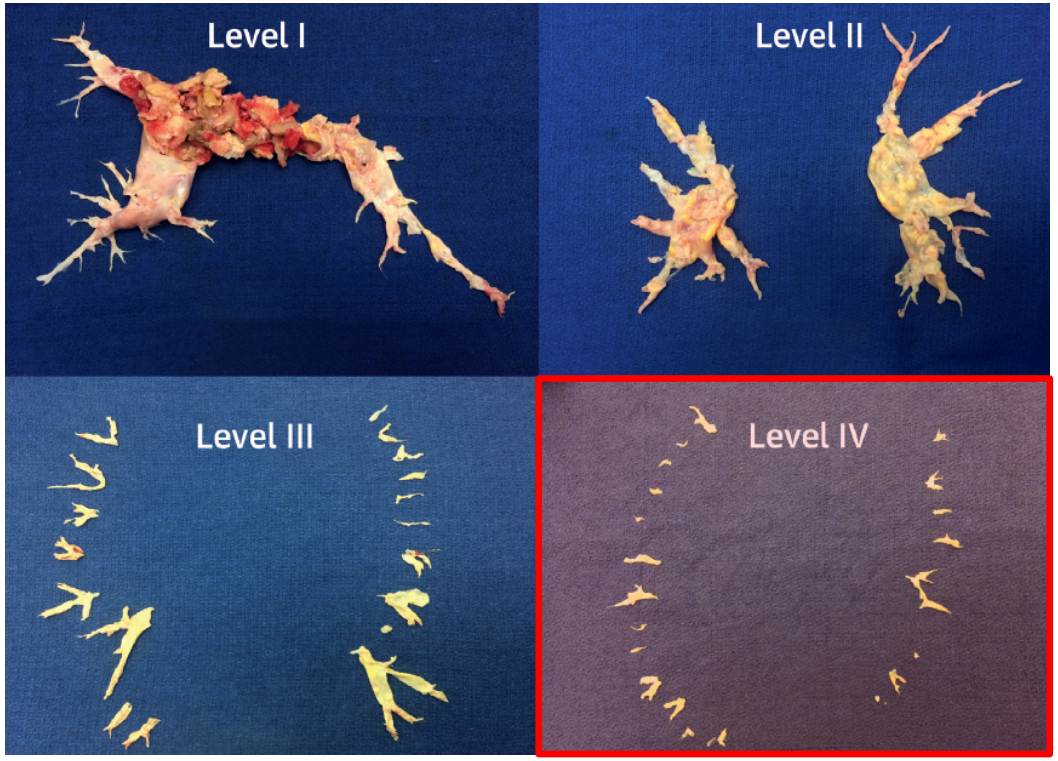


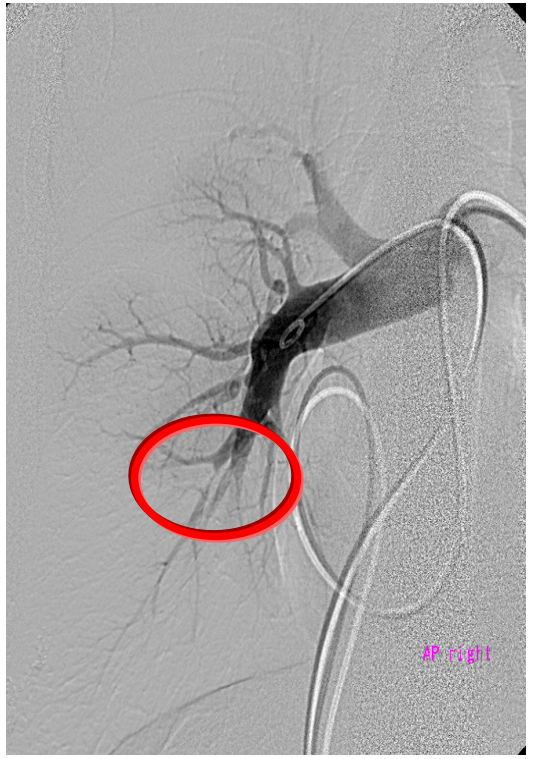


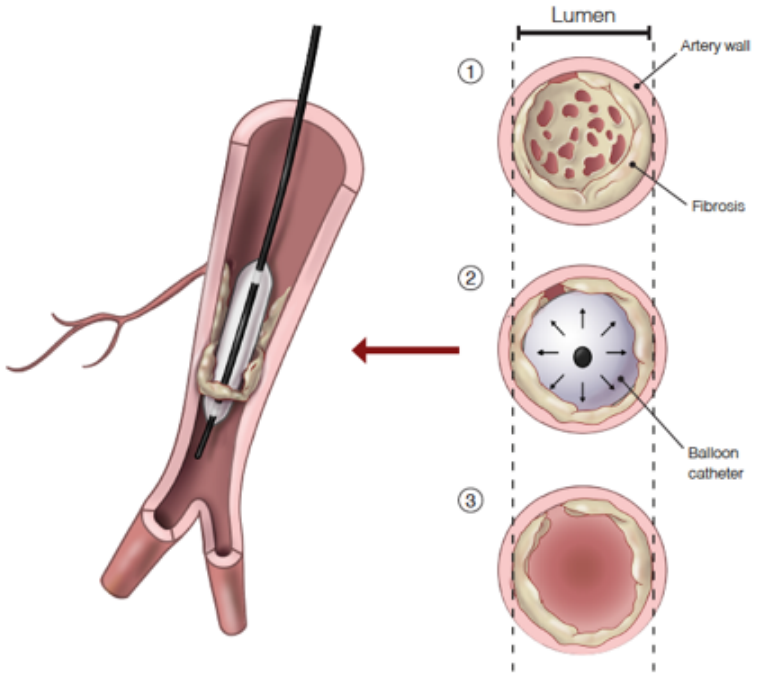
Studie CTREPH (treprostinil), n=105
Inoperabilní, perzistentní CTEPH, věk 64, 24 týdnů

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

BALÓNKOVÁ PLICNÍ ANGIOPLASTIKA V LÉČBĚ CTEPH





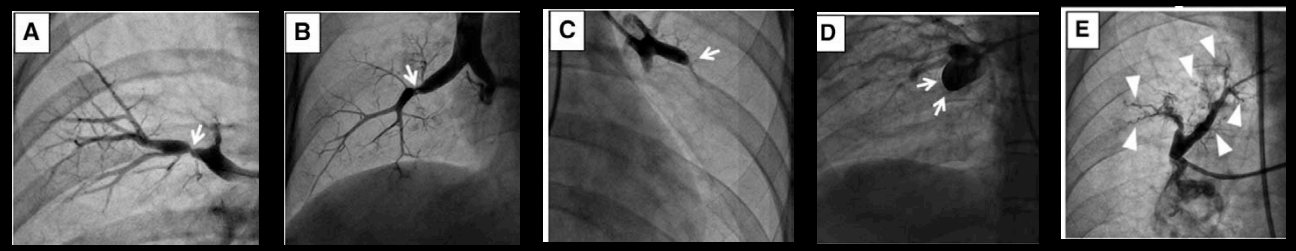


Mahmud et al. *JACC* 2018 Vol 71, 2468-8; Maren Hötten Master Thesis in Scientific Design, 2016; VFN

BALÓNKOVÁ PLICNÍ ANGIOPLASTIKA V LÉČBĚ CTEPH

Novel Angiographic Classification of Each Vascular Lesion in Chronic Thromboembolic Pulmonary Hypertension Based on Selective Angiogram and Results of Balloon Pulmonary Angioplasty

Takashi Kawakami, MD, PhD; Aiko Ogawa, MD, PhD; Katsumasa Miyaji, MD, PhD; Hiroki Mizoguchi, MD, PhD; Hiroto Shimokawahara, MD, PhD; Takanori Naito, MD; Takashi Oka, MD; Kei Yunoki, MD, PhD; Mitsuru Munemasa, MD, PhD; Hiromi Matsubara, MD, PhD

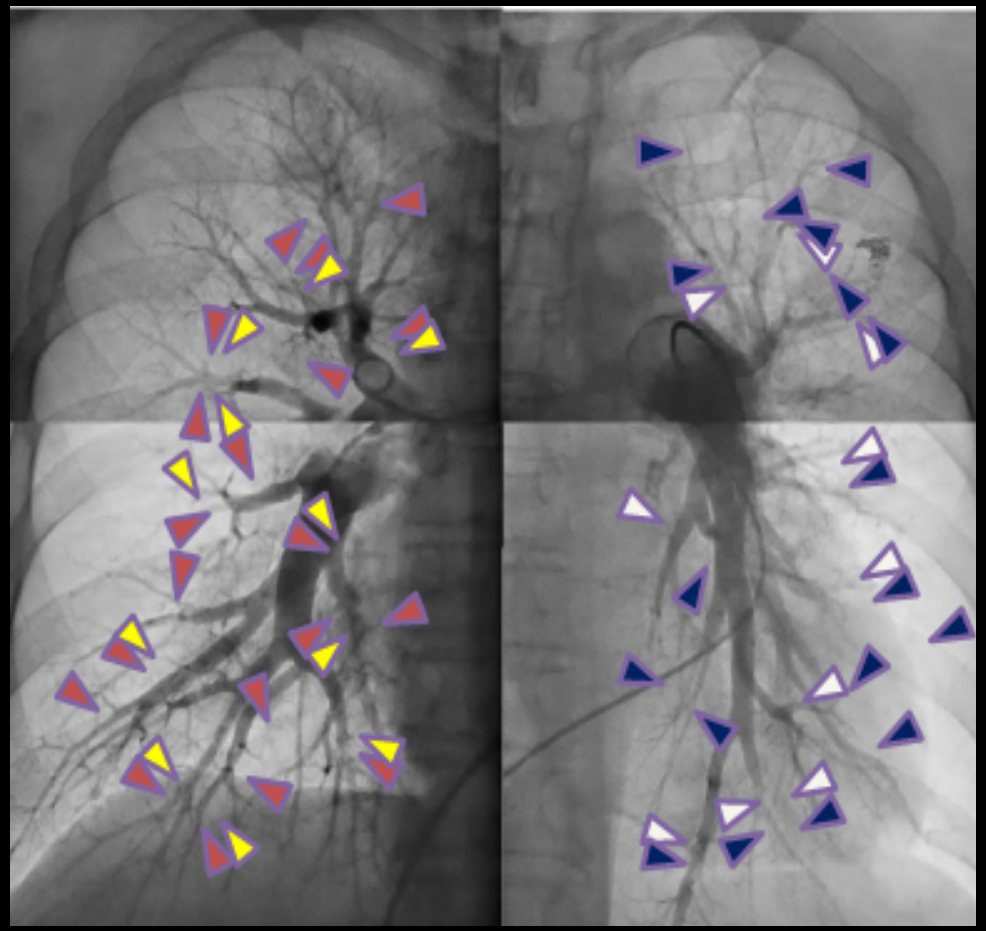


A Ring-like, B Web, C Subtotal lesion, D Total occlusion, E Tortuous lesion

Segmentární a subsegmentární větve

Subsegmentární a distální větve

↑ úspěšnost, ↓ komplikace

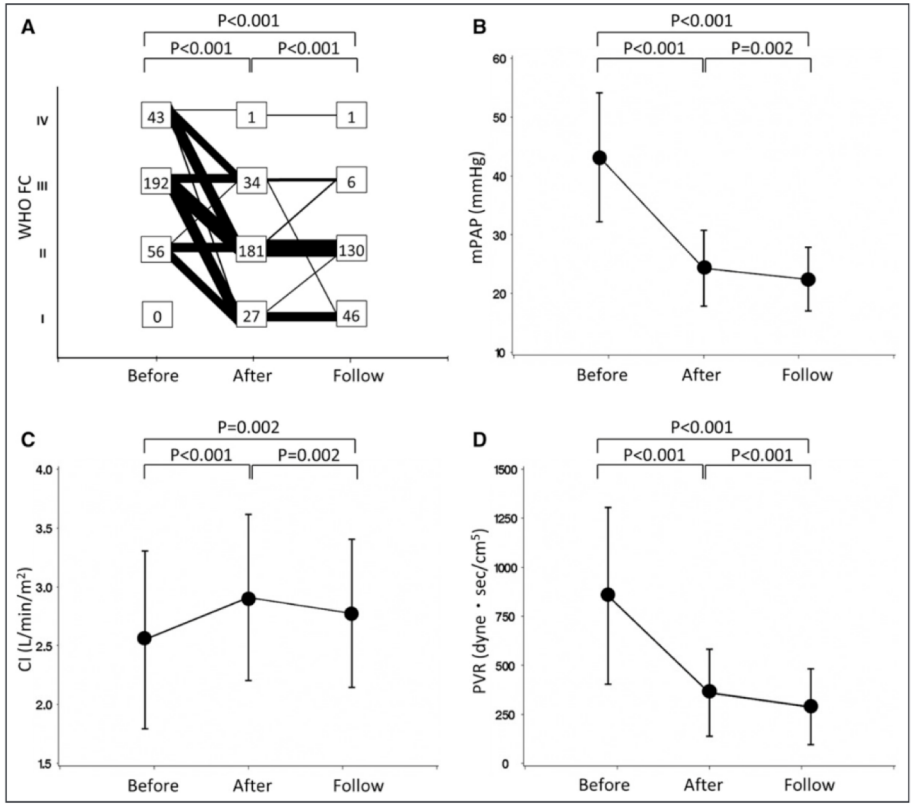


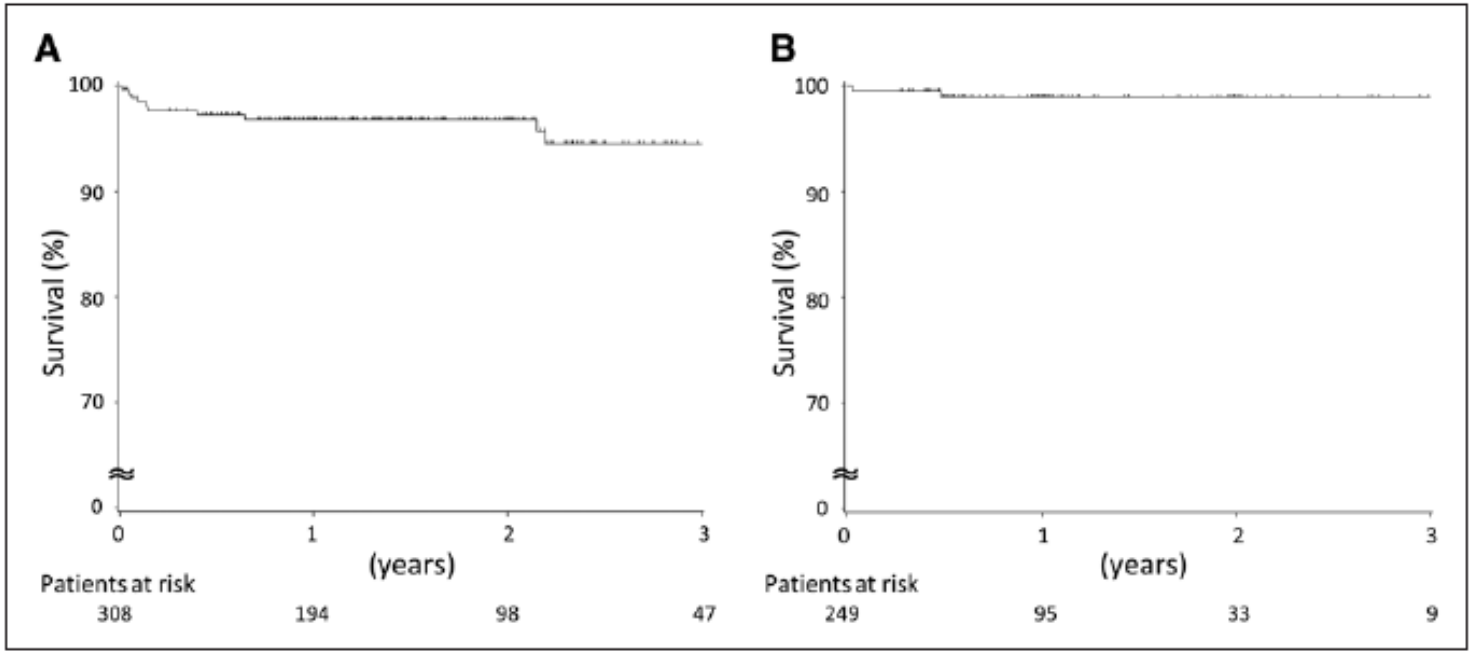
Circ Cardiovasc Interv. 2016;9:e003318
Okayama Medical Centre
VFN

Balloon Pulmonary Angioplasty for Chronic Thromboembolic Pulmonary Hypertension

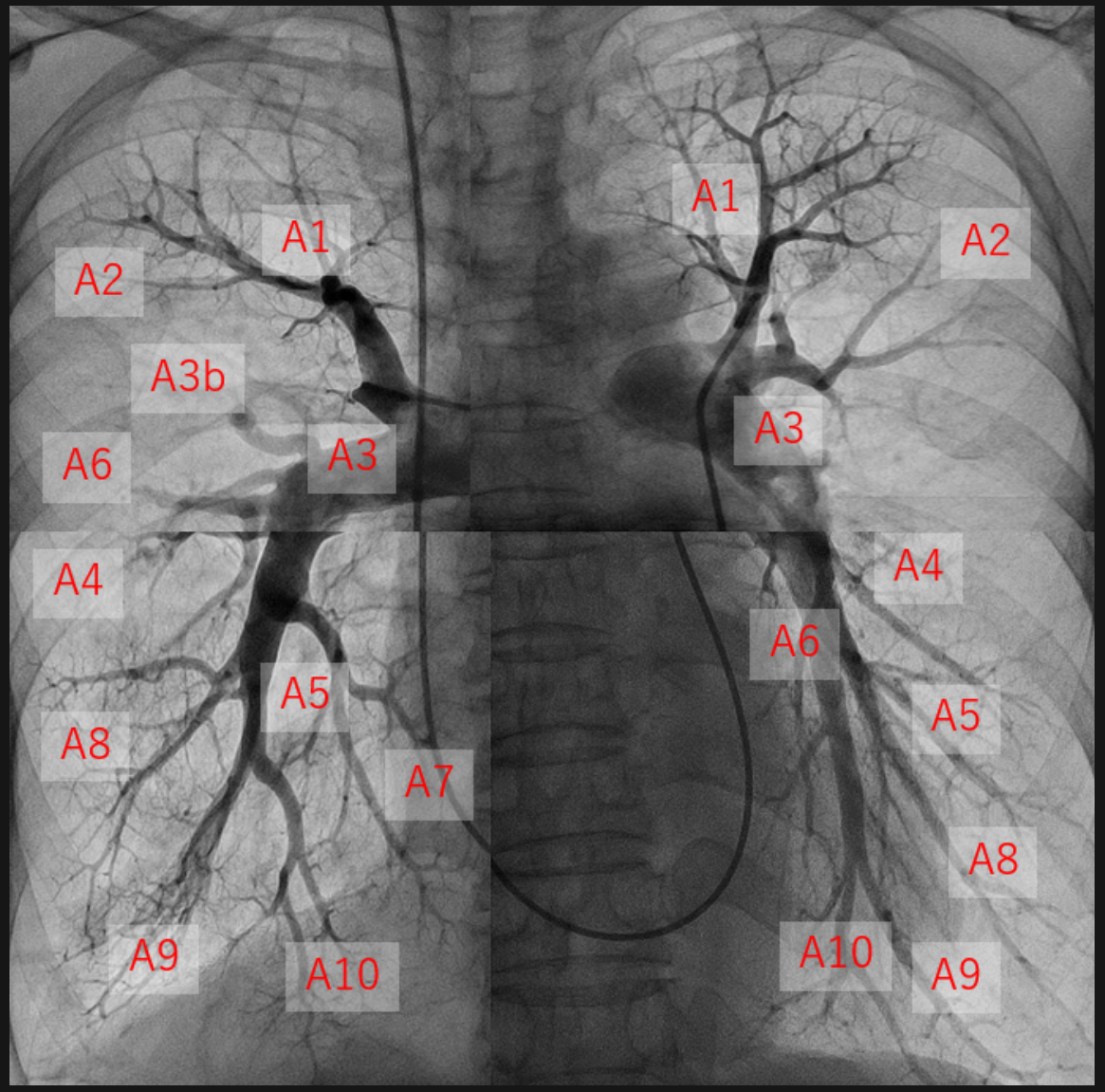
Results of a Multicenter Registry

N=308, M/F 62/246, věk 62 , 7 institucí, 1408 výkonů, inoperabilní 76 %, reziduální 4.5 %, operabilní 19.5 %
Farmakoterapie před BPA: 72 %
PAMP 43.2±11.0 → 24.3±6.4 mm Hg, komplikace: 36.3%, časná mortalita – 8 pacientů





Ogawa A et al. *Circ Cardiovasc Qual Outcomes*. 2017;10:e004029


A1
A2
A3b
A6
A4
A5
A8
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A10
A1
A2
A3
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A6
A5
A8
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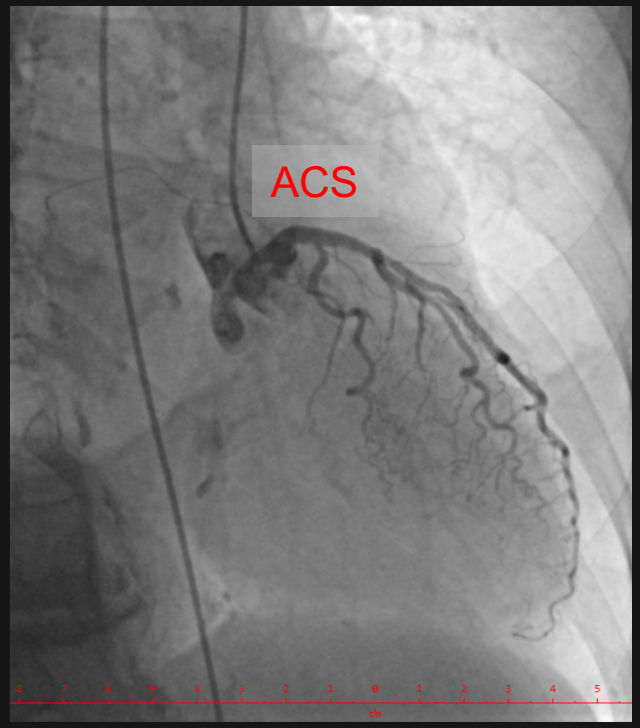
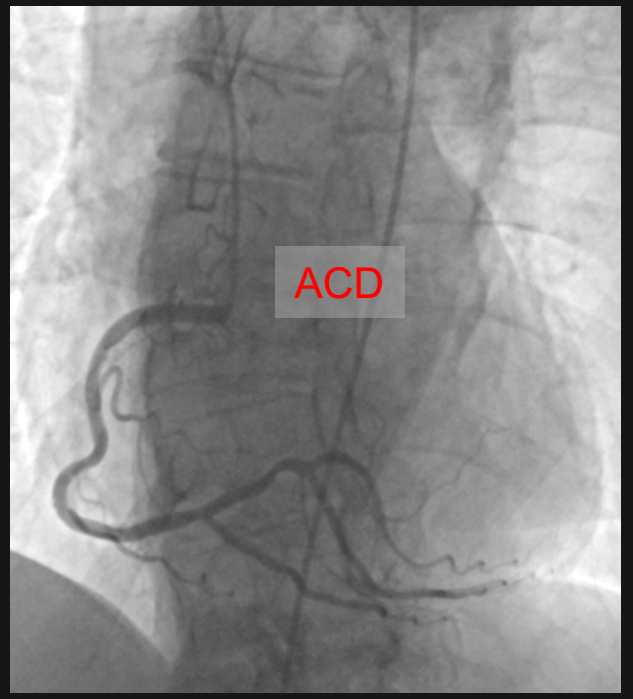
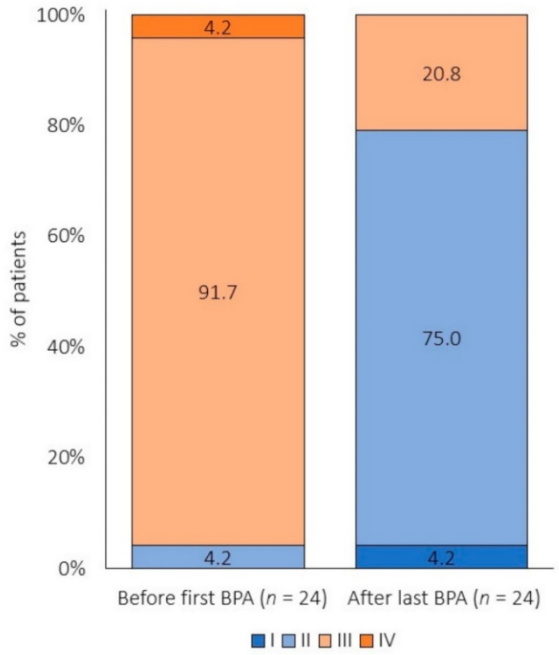
A9


ACS

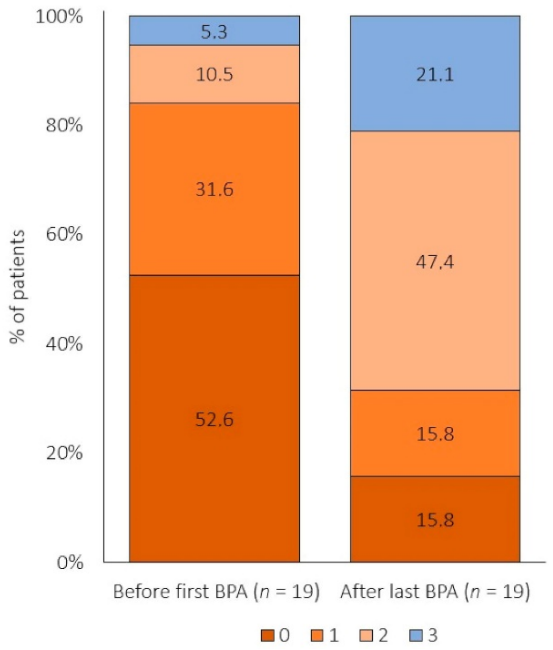

ACD

Balloon Pulmonary Angioplasty in Patients with Chronic Thromboembolic Pulmonary Hypertension: Impact on Clinical and Hemodynamic Parameters, Quality of Life and Risk Profile

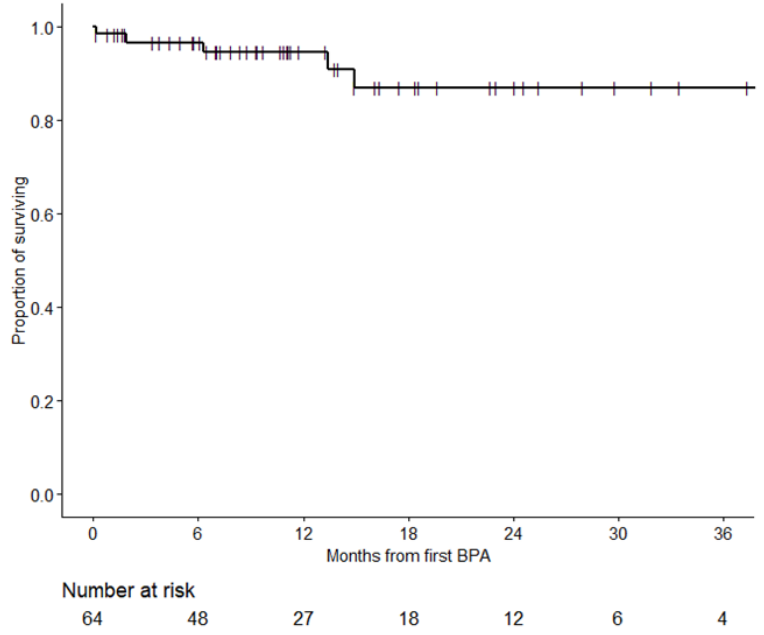
N=64, 47 inoperable (distal disease), 4 operable - refused surgery, 13 residual PH after PEA
160 BPAs, 749 lesions (9.9 % CTOs)
Improvement:
PAMP (−18%, $p < 0.001$), PVR (−32%, $p < 0.001$), SV (+17%, $p = 0.011$), QoL (+37% overall health status, $p < 0.001$)



NYHA functional class



Risk profile (NYHA, 6MWT, proBNP)



Survival

Jansa P, Heller S et al. *J. Clin. Med*. 2020, 9, 3608; doi:10.3390/jcm9113608

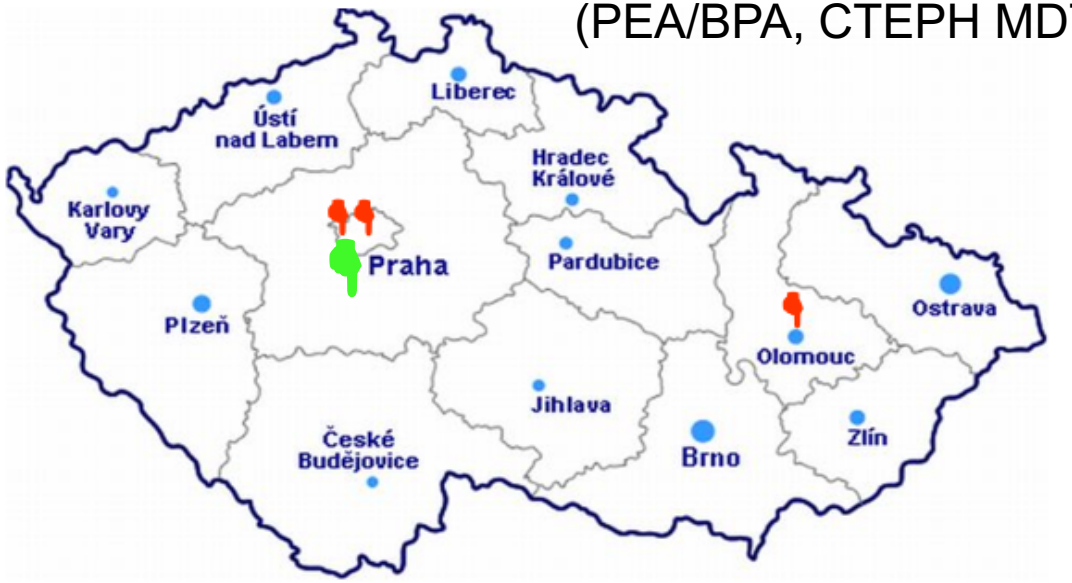
PLICNÍ HYPERTENZE V ČR

European Reference Network
for rare or low prevalence complex diseases

Network
Respiratory Diseases (ERN-LUNG)

Member
General University Hospital in Prague — Czechia

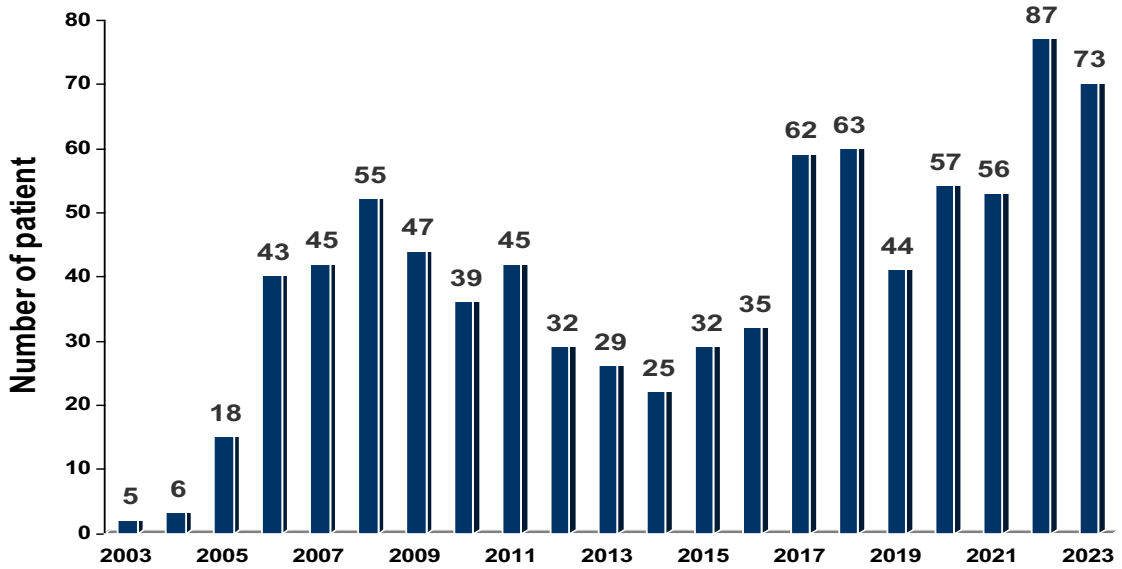
- 3 expertní centra pro PAH centres
- 1 expertní centrum pro CTEPH (PEA/BPA, CTEPH MDT team)



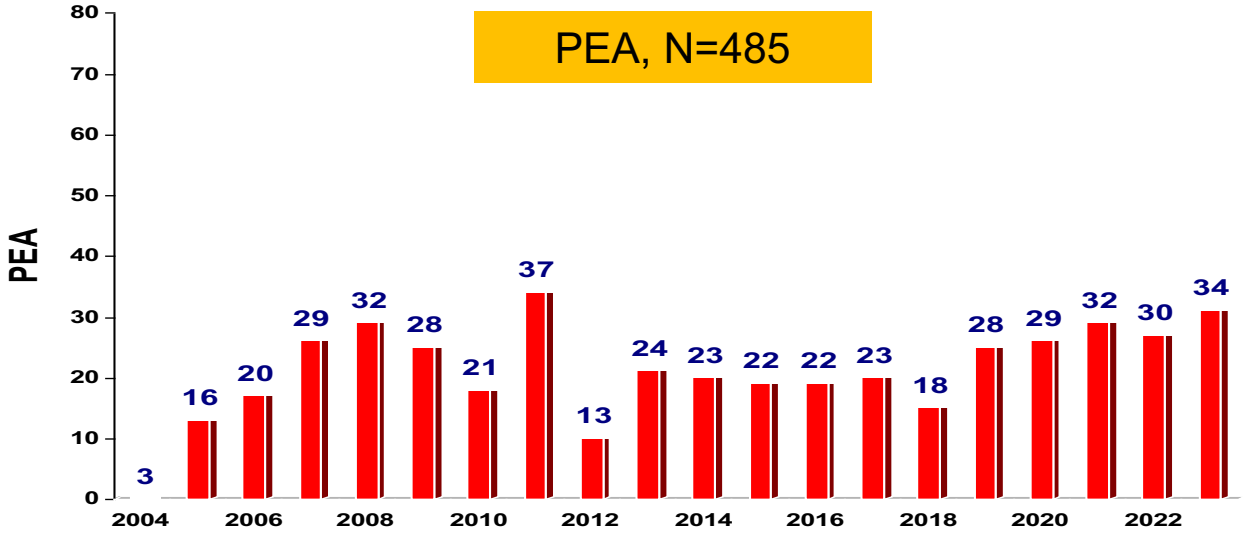
- II. Interní klinika VFN a 1.LF UK v Praze, 1. interní klinika FN Olomouc, Klinika kardiologie IKEM
- II. Interní klinika VFN a 1.LF UK v Praze

CTEPH IN THE CZECH REPUBLIC (2003-2023)

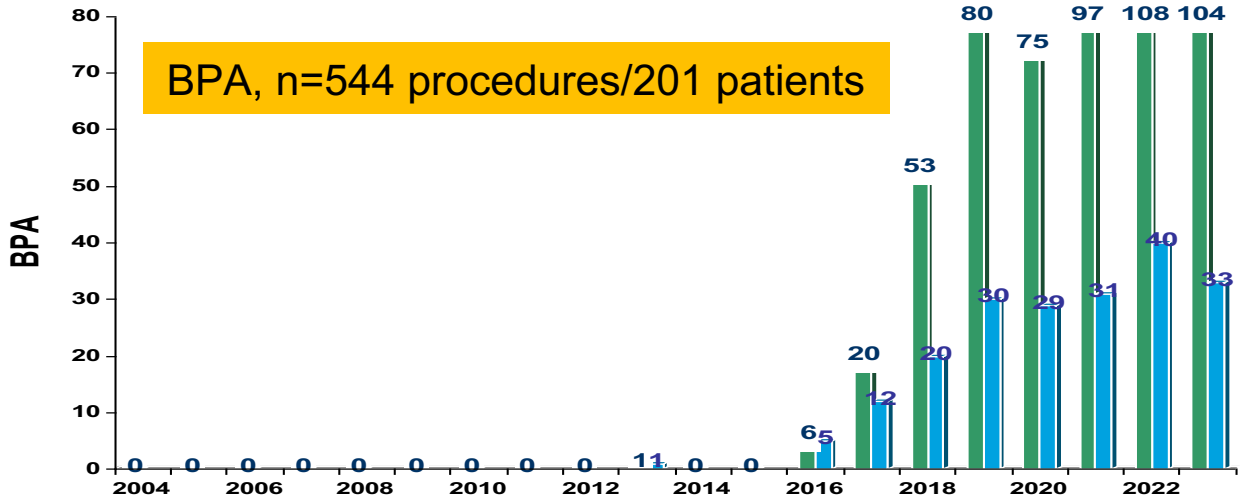
Newly diagnosed patients (n=892)



PEA, N=485



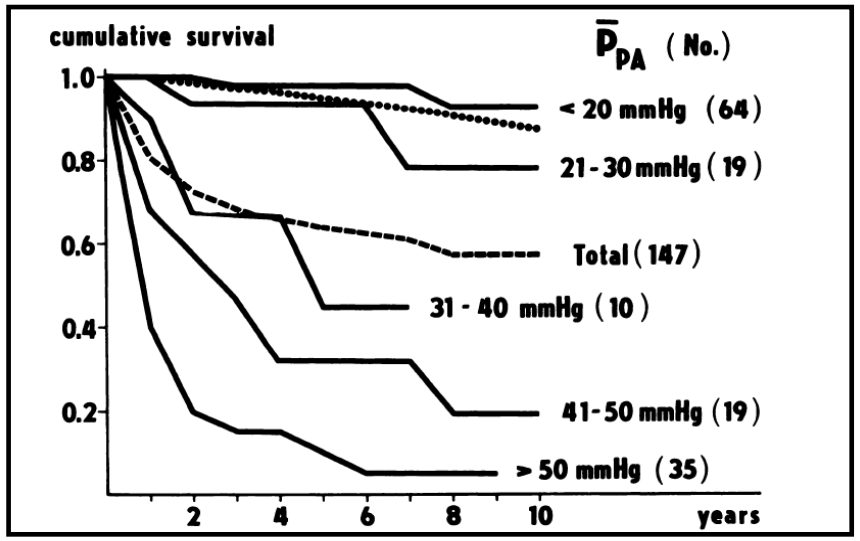
BPA, n=544 procedures/201 patients



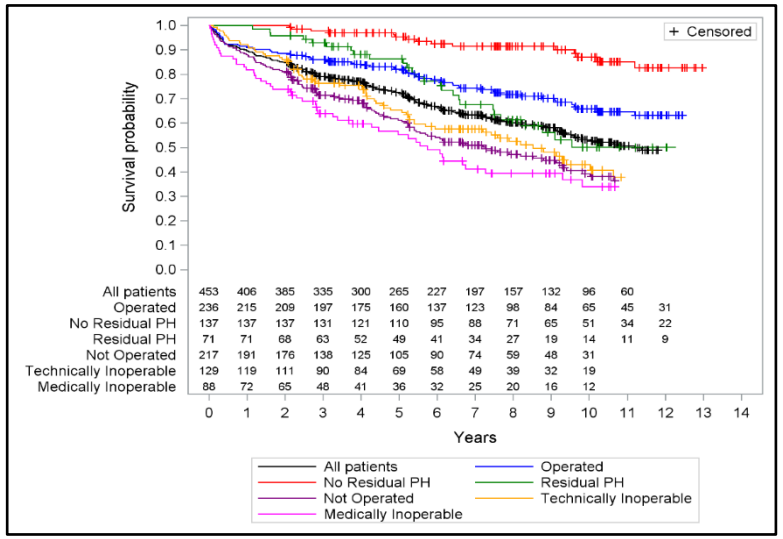
Jansa P et al. Epidemiology of chronic thromboembolic pulmonary hypertension (CTEPH) in the Czech Republic. *Pulmonary Circulation* 2022;12:e12038

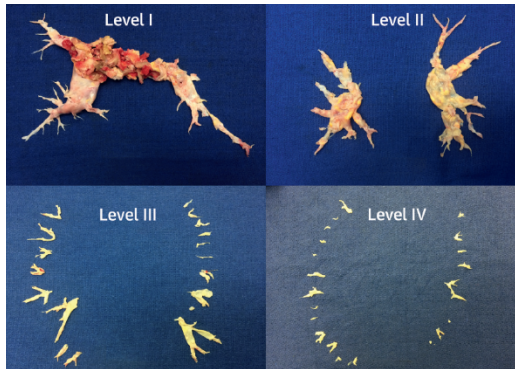
„MULTIMODÁLNÍ“ LÉČBA CTEPH

Pravděpodobnost přežití (%) **bez léčby CTEPH**

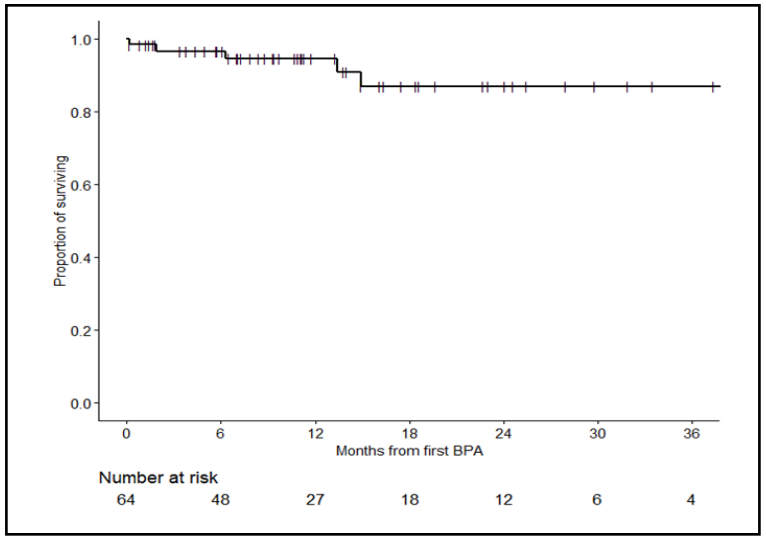


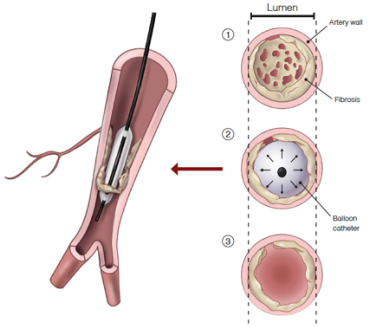
Pravděpodobnost přežití (%) **2004-2016 (PEA)**





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

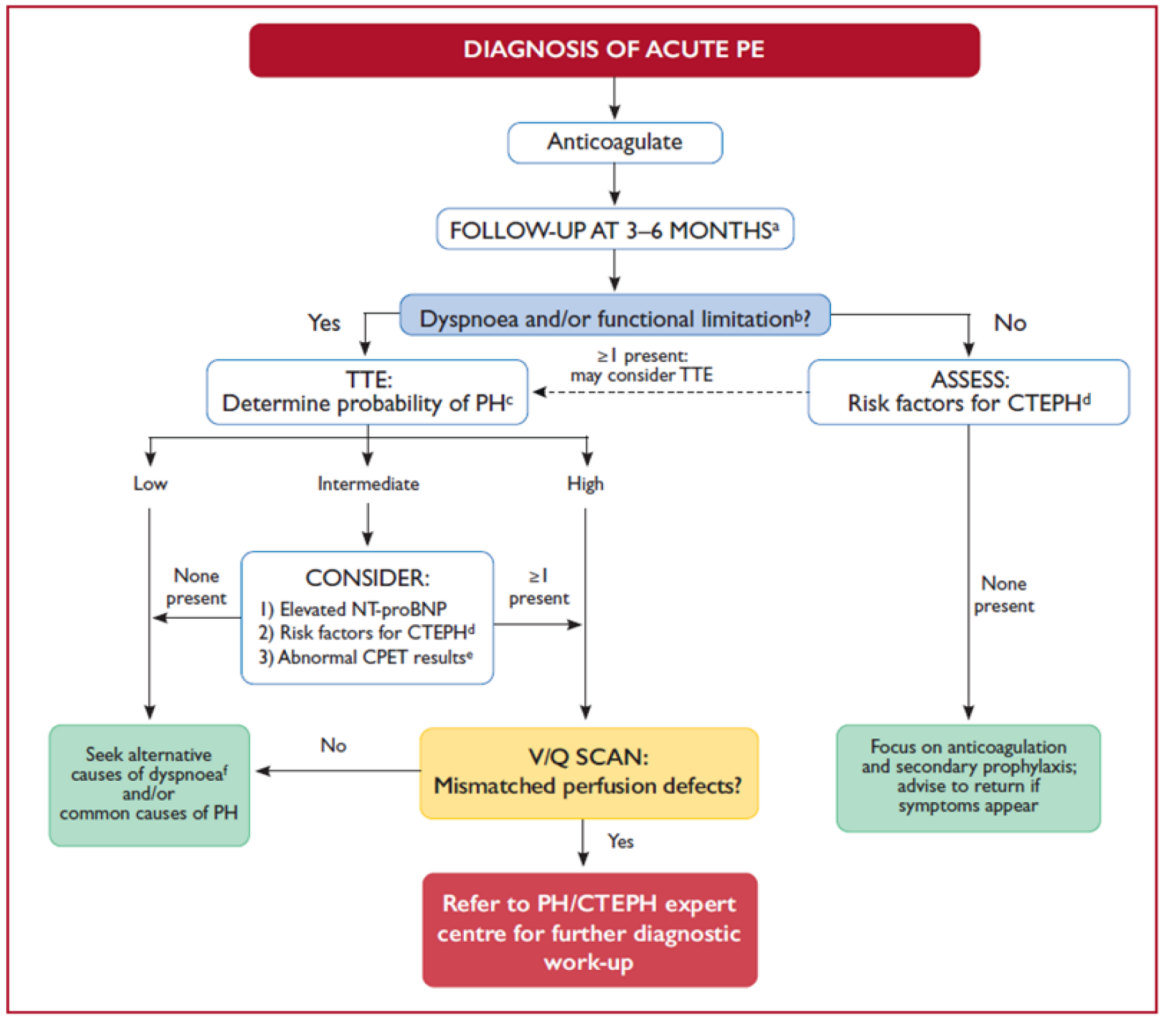
Epidemiology of chronic thromboembolic pulmonary hypertension (CTEPH) in the Czech Republic

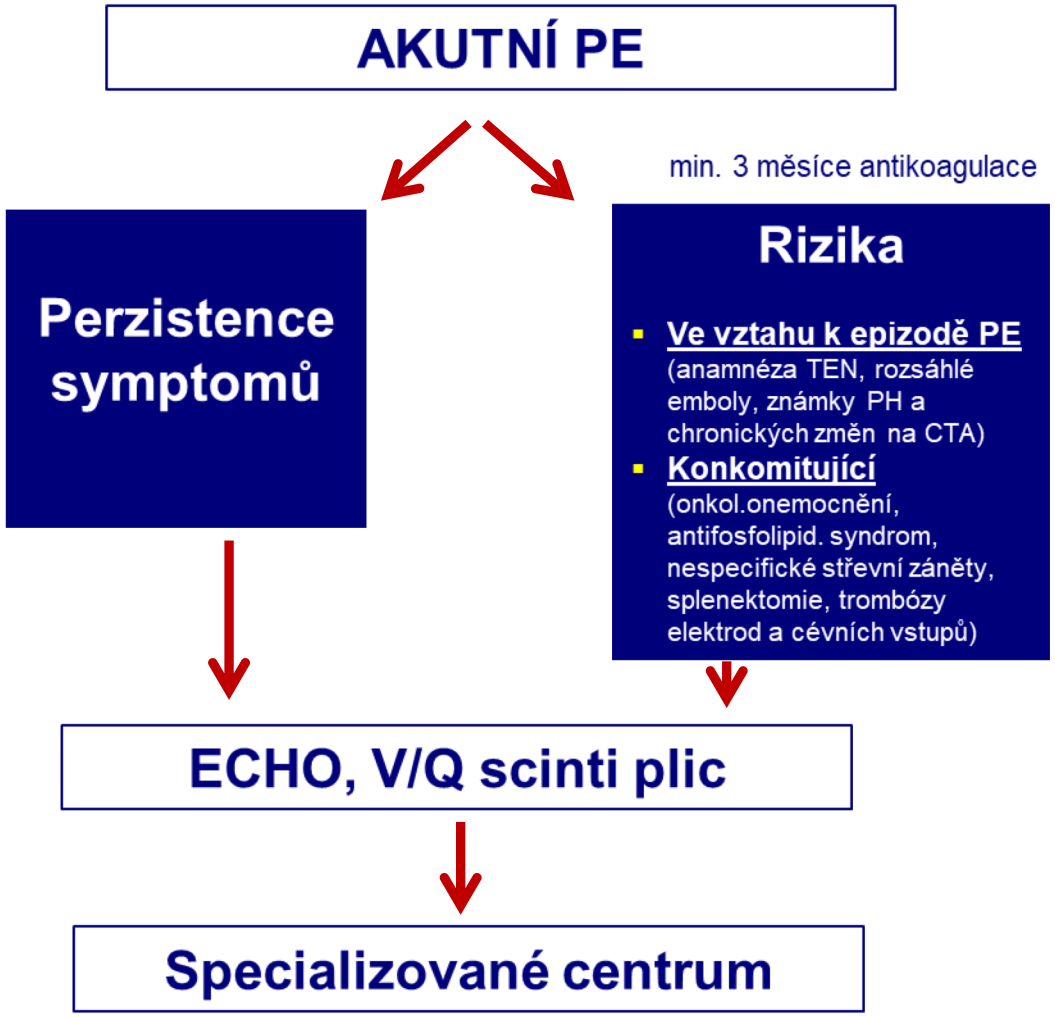
N=456 (newly diagnosed 2003-2016), NYHA III+IV 91.8 %, age 63.2 (49; 77), operated 52.1 %
PAMP 50.0 (34; 81) mmHg, CI 2.1 (1; 4) $L.min^{-1}.m^{-2}$, PVR 682.4 (244; 2197) $dyn.s.cm^{-5}$

Parameter	All, $N = 453$	Operated, $N = 236$	Not-operated		
			Overall not-operated, $N = 217$	Technically inoperable, $N = 129$	Medically inoperable, $N = 88$
Age, years, median (range)	65.2 (19; 85)	62.2 (21; 81)	69.9 (19; 85)	70.6 (19; 83)	67.9 (26; 85)
Sex, *n* (%), female	206 (45.5)	89 (37.7)	117 (53.9)	71 (55.0)	46 (52.3)
DVT history, *n* (%)	186 (41.9)	99 (42.5)	87 (41.2)	45 (35.4)	42 (50.0)
PE history, *n* (%)	354 (78.1)	190 (80.5)	164 (75.6)	94 (72.9)	70 (79.5)
Time from first PE to diagnosis, years, median (range)	2.2 (0; 43)	2.4 (0; 43)	2.0 (0; 42)	2.1 (0; 31)	1.9 (0; 42)
NYHA FC, *n*	437	234	203	115	88
FC I/II, *n* (%)	36 (8.3)	22 (9.4)	14 (6.9)	4 (3.5)	10 (11.3)
FC III/IV, *n* (%)	401 (91.8)	212 (90.6)	189 (93.1)	111 (96.5)	78 (88.6)
Median (range) 6MWD, *m* [*n*]	335.5 (40; 645) [372]	344.0 (106; 645) [206]	321.0 (40; 627) [166]	322.5 (40; 627) [94]	319.5 (50; 605) [72]
RHC, median (range)					
mPAP, mmHg [*n*]	48.0 (11; 91) [439]	51.0 (11; 87) [228]	43.0 (17; 91) [211]	44.0 (21; 91) [123]	43.0 (17; 73) [88]
PVR, dyn × s/cm^5 [*n*]	684.0 (83; 2197) [436]	756.8 (83; 2197) [228]	594.0 (112; 1723) [208]	607.2 (112; 1723) [121]	584.8 (133; 1525) [87]
CI, L/min/m^2, [*n*]	2.2 (1; 5) [429]	2.1 (1; 4) [223]	2.3 (1; 5) [206]	2.2 (1; 5) [120]	2.3 (1; 4) [86]
Median (range) BNP, pg/ml [*n*]	204.0 (10; 4828) [196]	254.5 (14; 4828) [94]	159.0 (10; 1531) [102]	197.0 (10; 1531) [55]	151.0 (11; 1526) [47]
Anticoagulation, *n*	446	233	213	127	86
NOAC, *n* (%)	13 (2.9)	4 (1.7)	9 (4.2)	5 (3.9)	4 (4.7)
Other anticoagulants, *n* (%)	433 (97.1)	229 (98.3)	204 (95.8)	122 (96.1)	82 (95.3)

Jansa P et al. *Pulmonary Circulation* 2022;12:e12038

ČASNÁ DIAGNÓZA CTEPH





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

SHRNUTÍ

- Výskyt CTEPH po PE: 2-4 %, až 30 % pacientů nemá anamnézu TEN
- CTEPD bez PH: symptomy + průkaz postembolických reziduí
- CTEPH (CTEPD s PH) : symptomy + průkaz postembolických reziduí + plicní hypertenze
- V/Q SPECT nahrazuje V/Q planární scintigrafii
- Role CTA v detekci a diagnostice
- Časná detekce v prvních 3-6 měsících po akutní PE
- Při léčbě NOAC vyloučit APS (10 % u CTEPH)
- Multimodální léčba CTEPH umožňuje kompletní vyléčení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