OVLIVNĚNÍ MORTALITY U PACIENTŮ S PLICNÍ HYPERTENZÍ

PAVEL JANSA

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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]

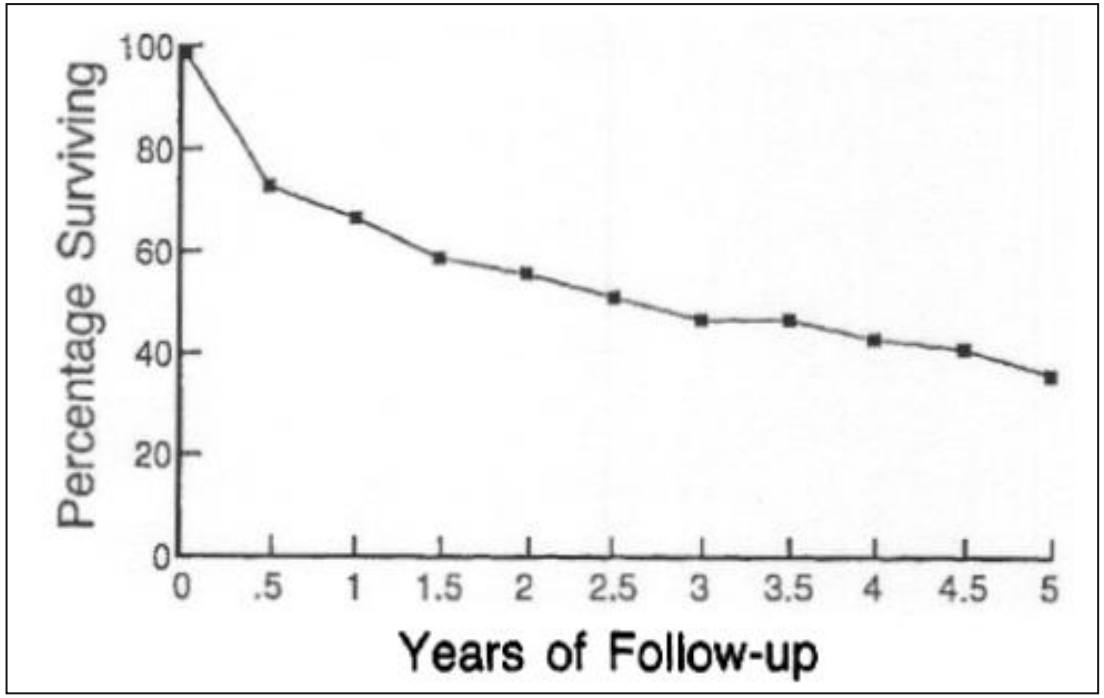
GROUP 5 PH with unclear and/or multifactorial mechanisms — 5 %
- 5.1 Haematological disorders[d]
- 5.2 Systemic disorders[e]
- 5.3 Metabolic disorders[f]
- 5.4 Chronic renal failure with or without haemodialysis
- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

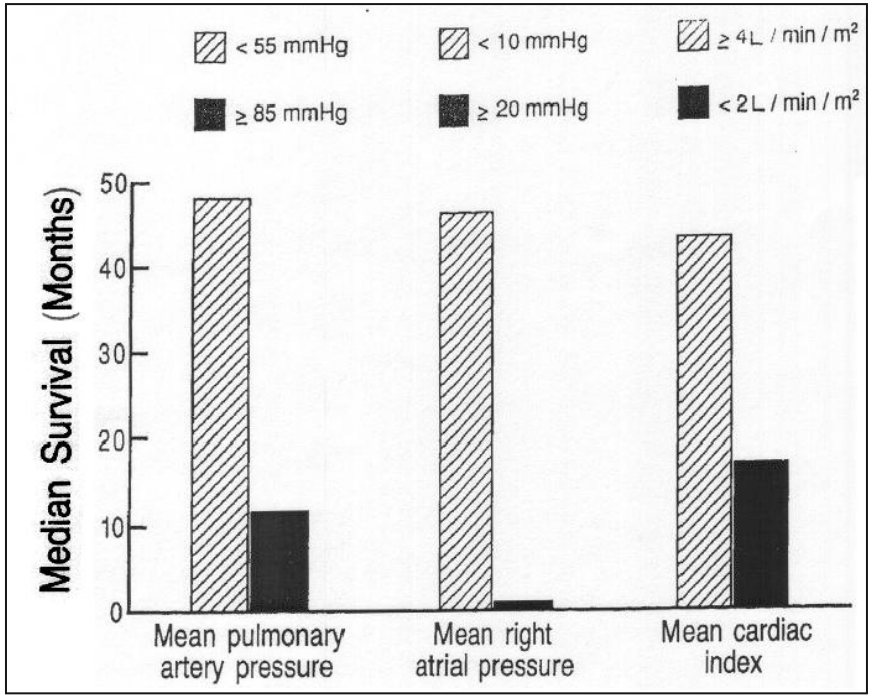
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
EHJ 2022, *ERJ* 2022

Survival in Patients with Primary Pulmonary Hypertension

Results from a National Prospective Registry

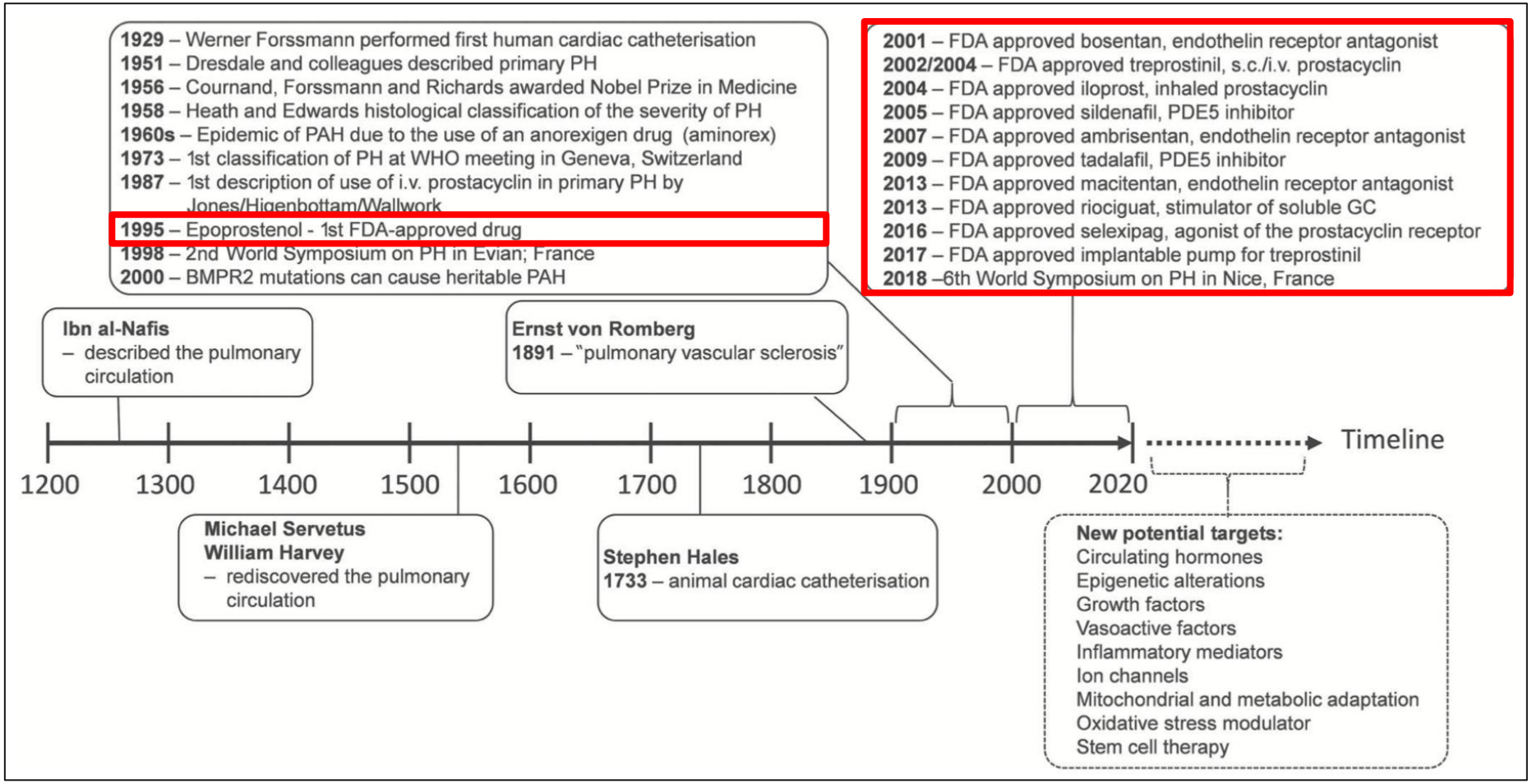
32 US clinical centers
187 patients diagnosed between 1981 and 1985, FU through 1988 (106 died)
Mean age 36±15 years, male:female 1:1.7
Median survival 2.8 yrs, 1-, 3- and 5-yr survival rates 68, 48 and 34%





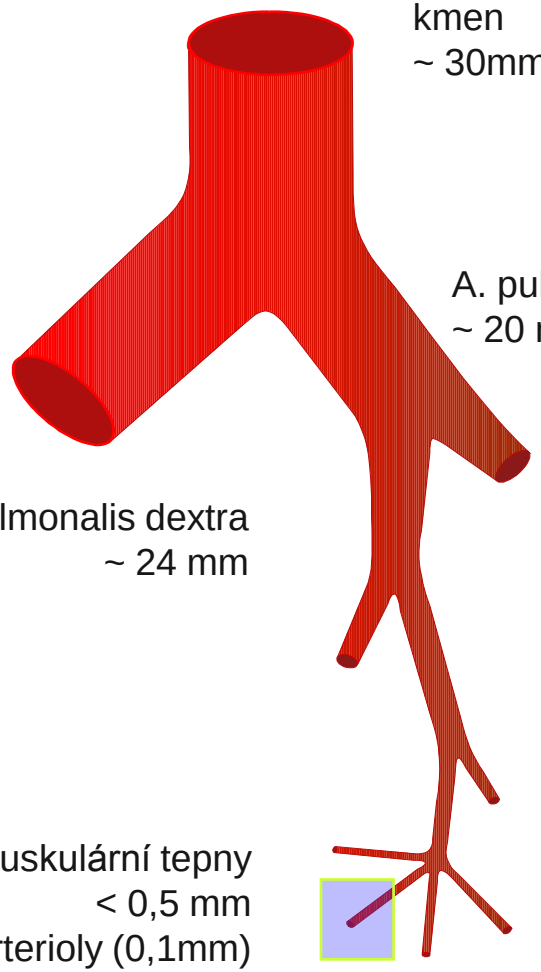
D'Alonzo et al. *Ann Internal Med* 1991;115:343

EVOLUCE LÉČBY PLICNÍ ARTERIÁLNÍ HYPERTENZE

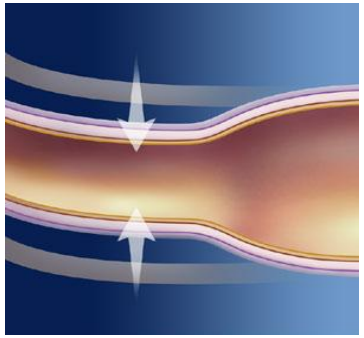


Sommer N et al. Current and future treatments of pulmonary arterial hypertension. *Br J Pharmacol.* 2020;1–25

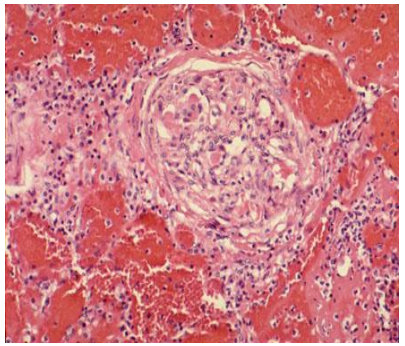
PODSTATA PLICNÍ ARTERIÁLNÍ HYPERTENZE (PAH)

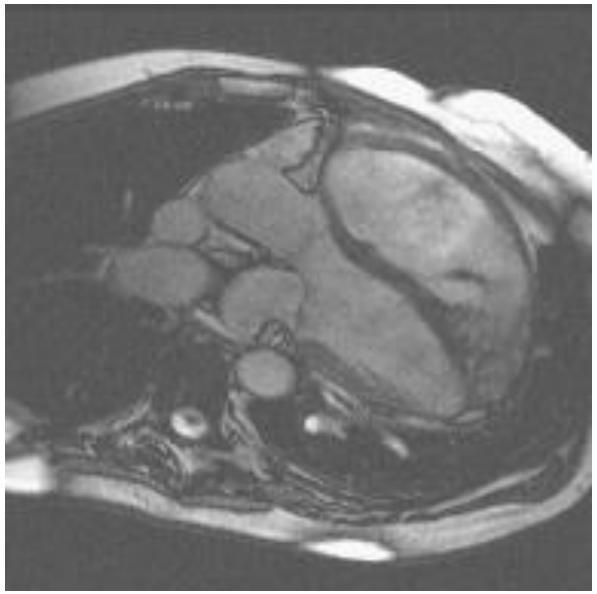


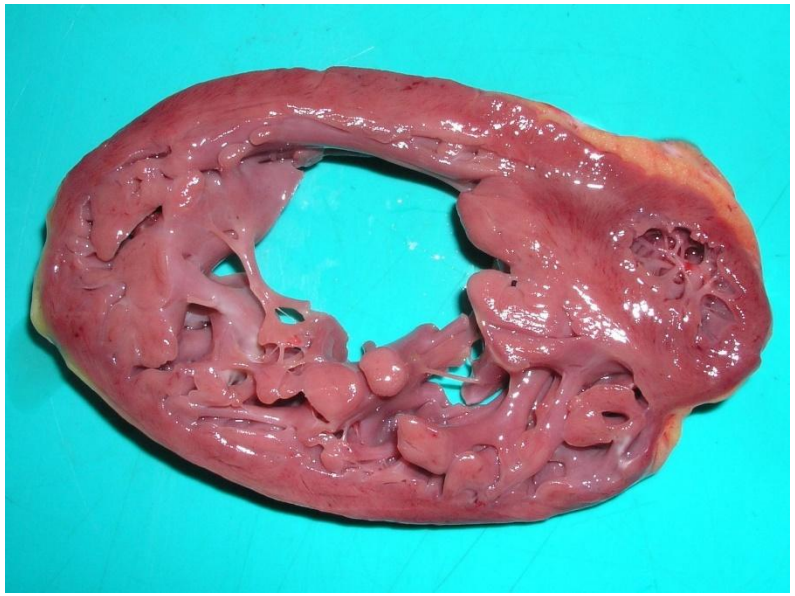
Vazokonstrikce



Remodelace



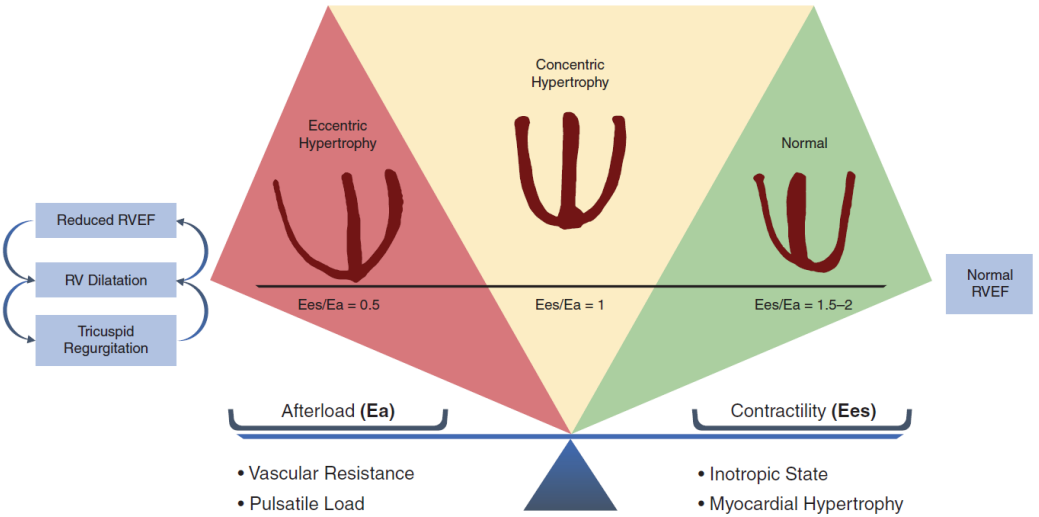


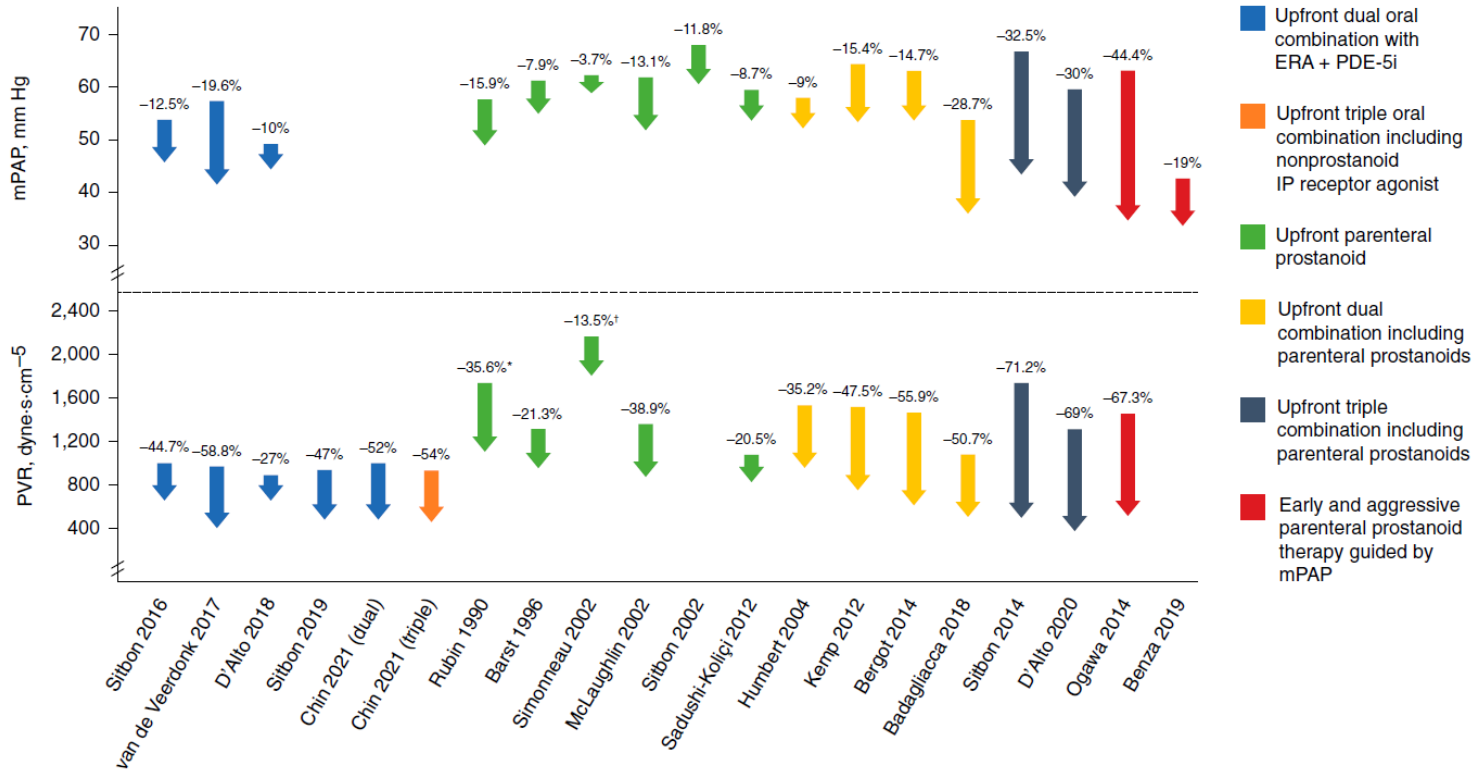


Zdroj: VFN, archiv autora

VÝVOJ TAKTIKY TERAPIE PAH

Aggressive Afterload Lowering to Improve the Right Ventricle
A New Target for Medical Therapy in Pulmonary Arterial Hypertension?





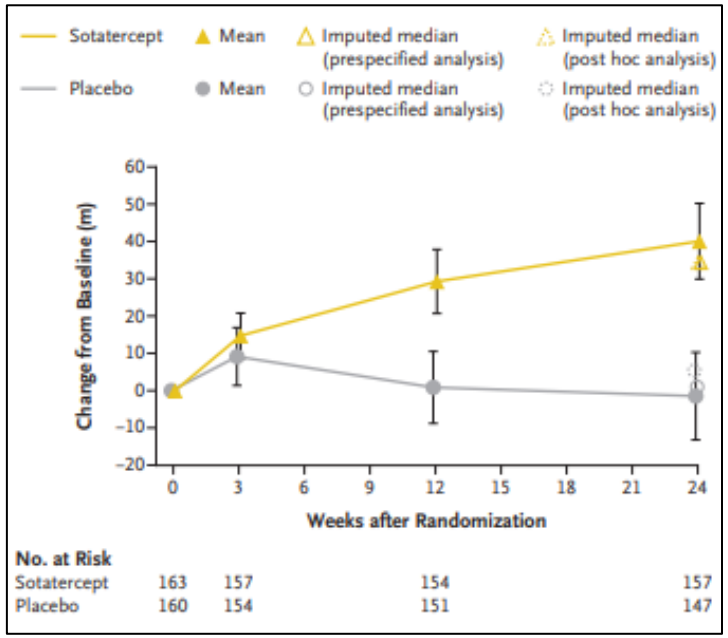
Vizza CD et al. *Am J Respir Crit Care Med* Vol 205, Iss 7, pp 751–760, Apr 1, 2022

Phase 3 Trial of Sotatercept for Treatment of Pulmonary Arterial Hypertension

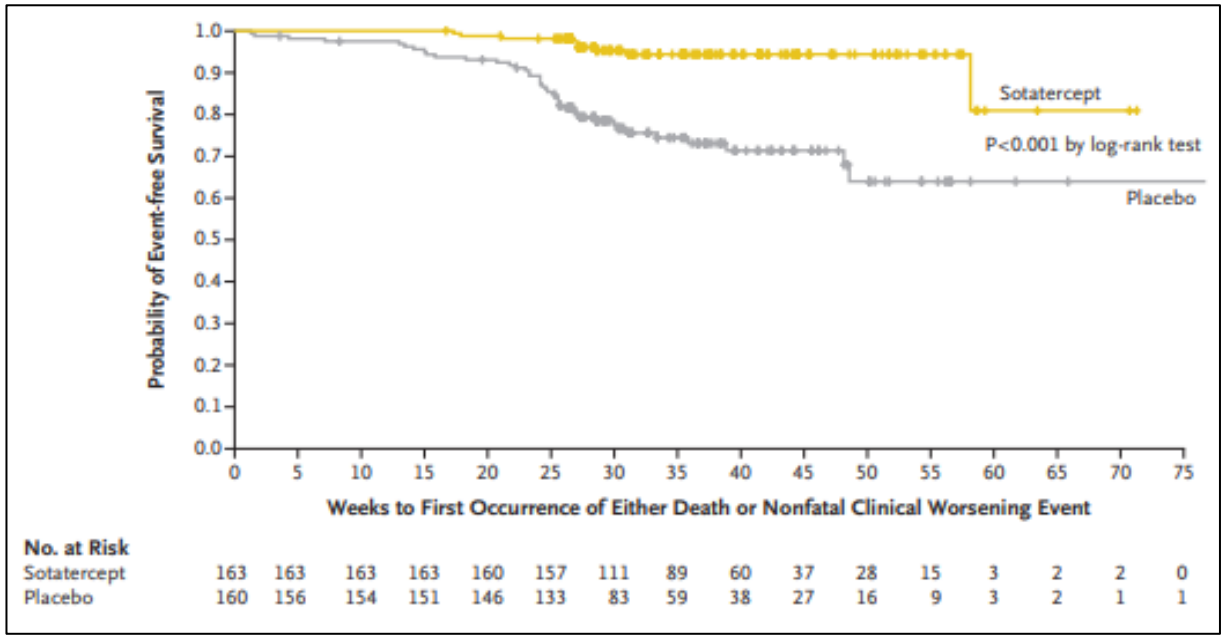
Marius M. Hoeper, M.D., David B. Badesch, M.D., H. Ardeschir Ghofrani, M.D., J. Simon R. Gibbs, M.D., Mardi Gomberg-Maitland, M.D., Vallerie V. McLaughlin, M.D., Ioana R. Preston, M.D., Rogerio Souza, M.D., Ph.D., Aaron B. Waxman, M.D., Ph.D., Ekkehard Grünig, M.D., Grzegorz Kopeć, M.D., Ph.D., Gisela Meyer, M.D., et al., for the STELLAR Trial Investigators*

PAH, 163 Sotatercept vs 160 placebo, 24 weeks, NYHA II 49% + NYHA III 51 %
Monotherapy 4 %, double 35 %, triple 61 %

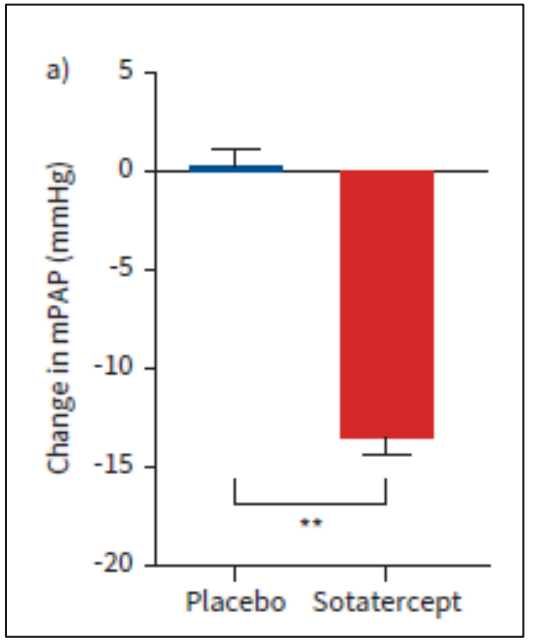
6MWT



TTCW



Hemodynamika

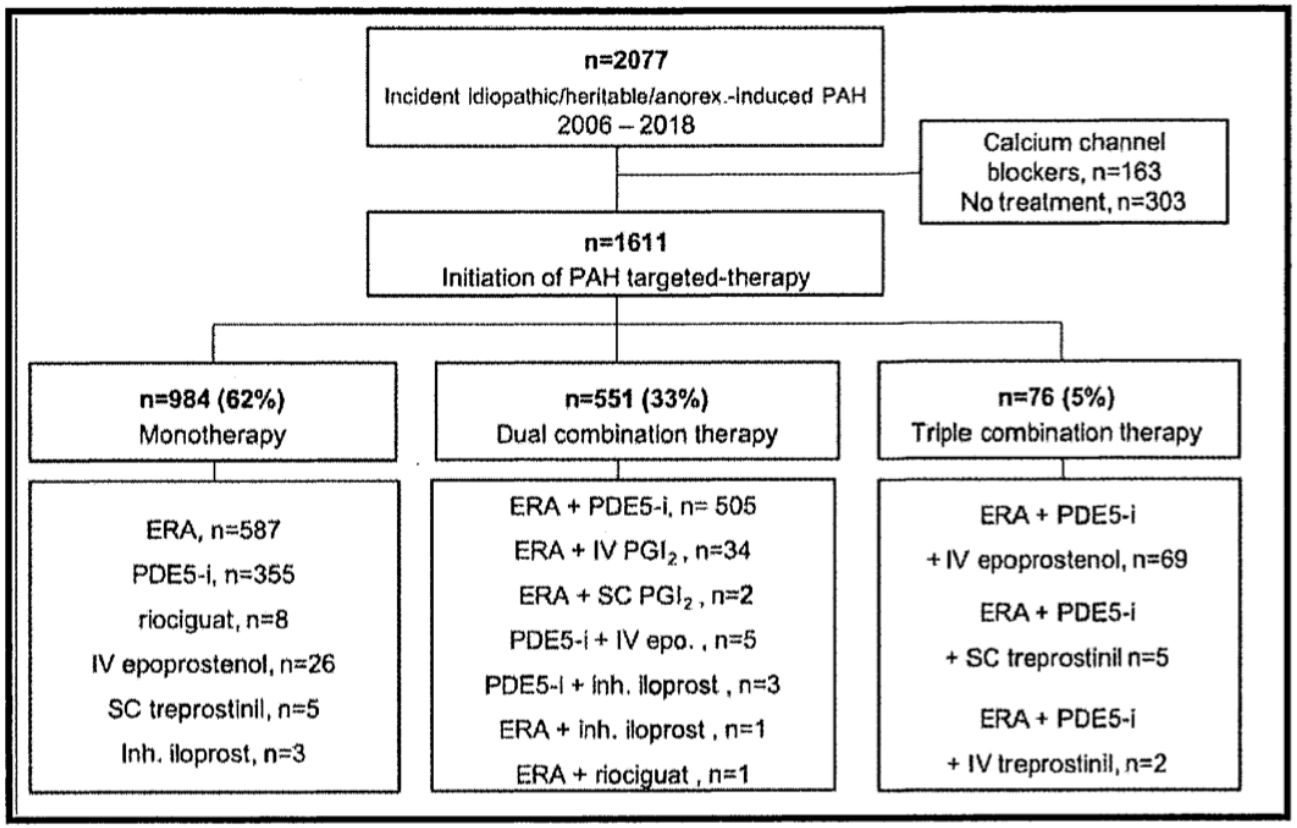


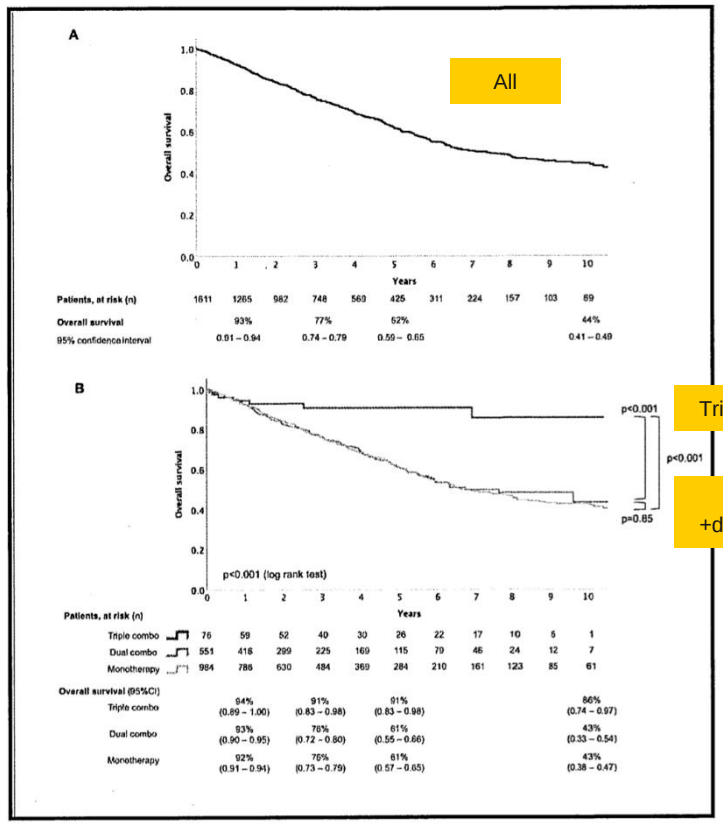
Hoeper M et al. *N Engl J Med* 2023, 388:1478-1490, Souza R et al. *Eur Respir J* 2023; 62: 2301107

Association between Initial Treatment Strategy and Long-Term Survival in Pulmonary Arterial Hypertension

French Registry 2006-2018

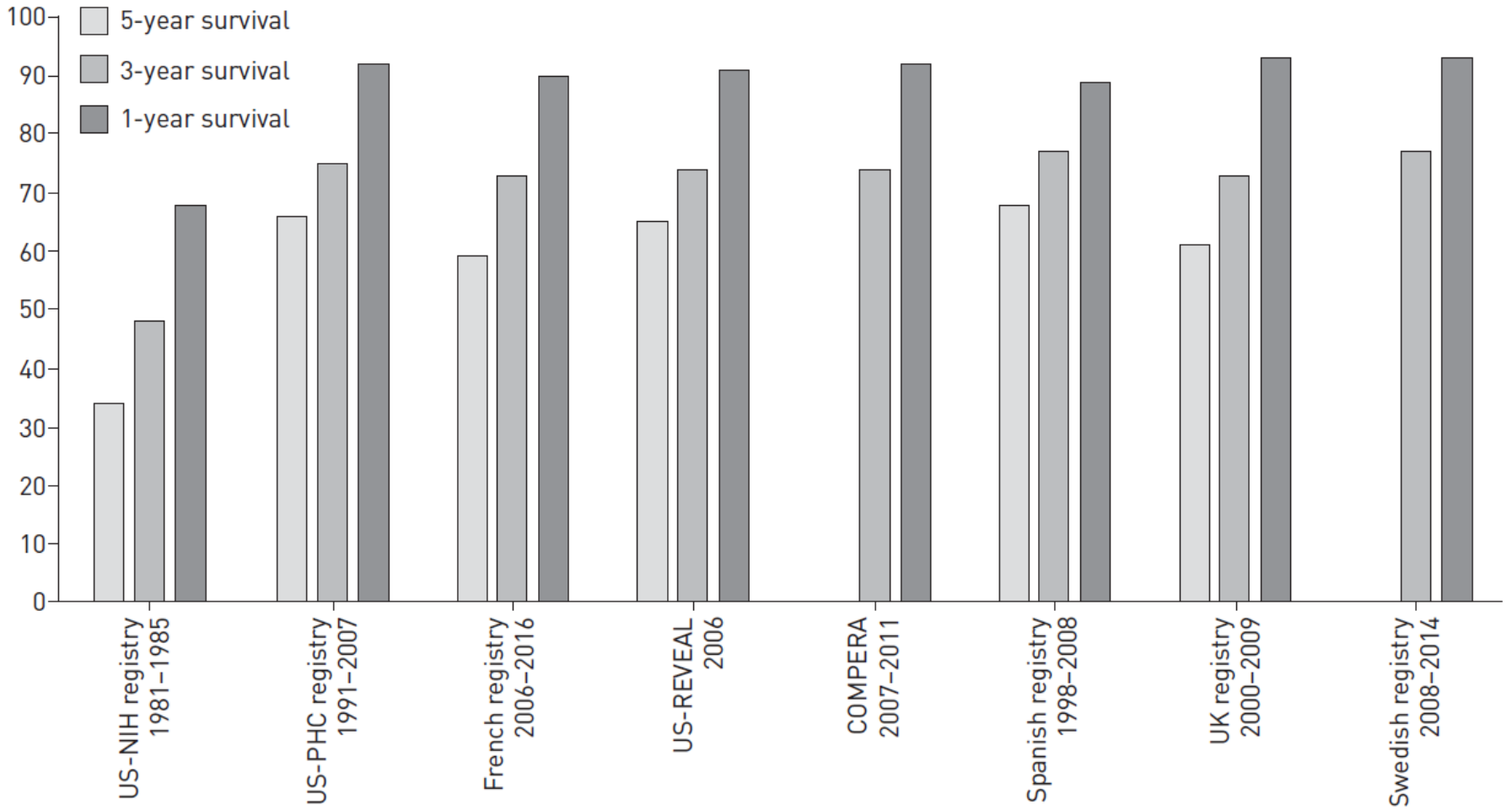
N=1611, 984 monotherapy, 551 dual combination therapy, 76 triple cpmbination therapy





Boucly A et al, *Am J Respir Crit Care Med* 2021 Oct 1;204(7):842-854

REGISTRY U PLICNÍ ARTERIÁLNÍ HYPERTENZE



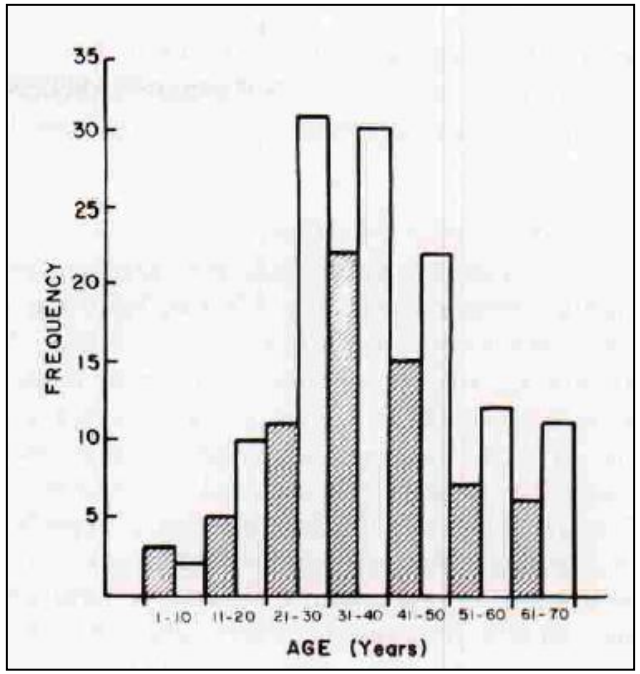
Swinnen K et al. *Eur Respir Rev* 2019; 28: 190050

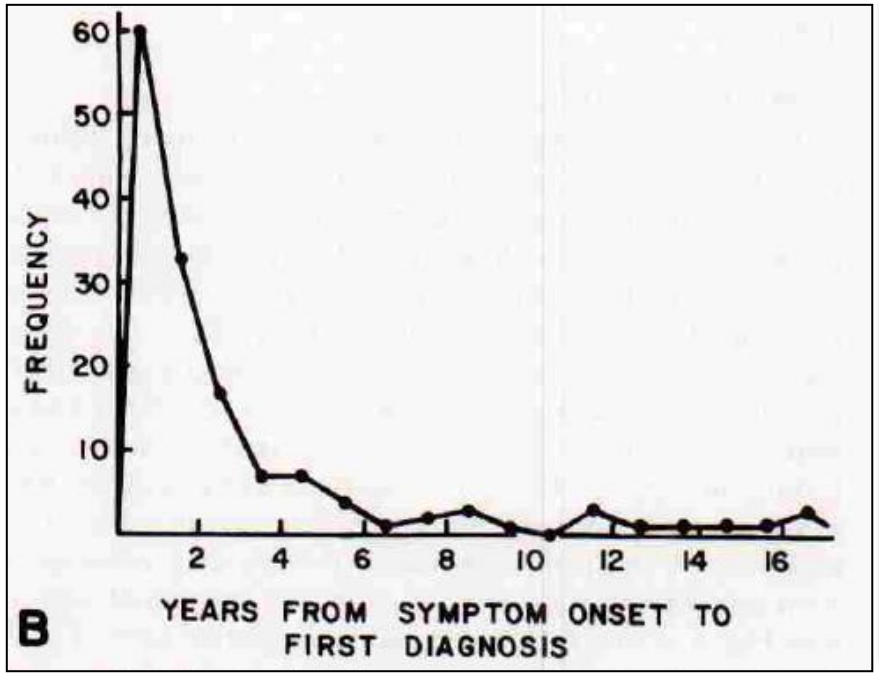
Primary Pulmonary Hypertension

A National Prospective Study

STUART RICH, M.D.; DAVID R. DANTZKER, M.D.; STEPHEN M. AYRES, M.D.; EDWARD H. BERGOFSKY, M.D.; BRUCE H. BRUNDAGE, M.D.; KATHERINE M. DETRE, M.D., Dr.P.H.; ALFRED P. FISHMAN, M.D.; ROBERTA M. GOLDRING, M.D.; BERTRON M. GROVES, M.D.; SPENCER K. KOERNER, M.D.; PAUL C. LEVY, Sc.D.; LYNNE M. REID, M.D.; CAROL E. VREIM, Ph.D.; and GEORGE W. WILLIAMS, Ph.D.; Bethesda, Maryland

32 center v USA, 187 pacientů diagnostikovaných v letech 1981 až 1985, FU do 1988 (106 úmrtí)
Průměrný věk 36±15 roku, M:F 1:1.7
Medián přežití 2.8 roku, pravděpodobnost přežití 1, 3, 5 let: 68 %, 48 %, 34%





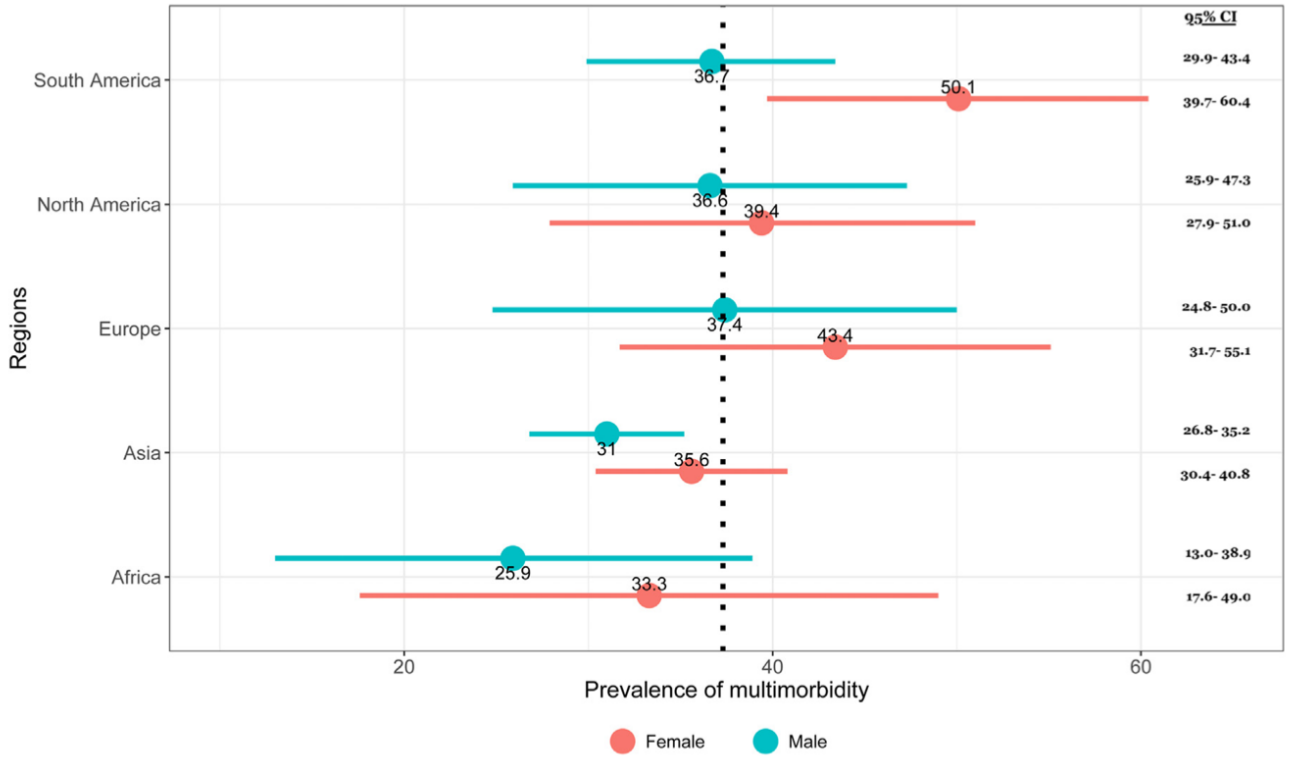
MEDICAL AND FAMILY HISTORY

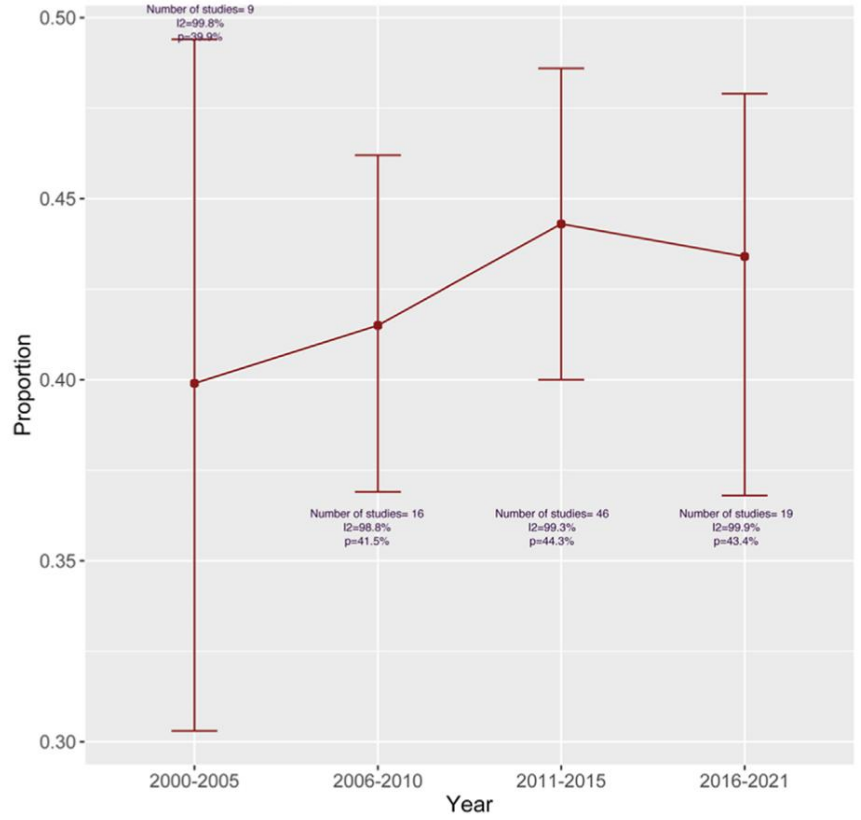
Forty-five percent of the patients were previous or current cigarette smokers, and only 5% had histories of appetite suppressant drug use. Fifty-four percent of the female patients had taken oral contraceptives at some time. There were 2.3 live births per female patient in the registry. None of these frequencies appear to differ dramatically from those found in the general population. There were 12 cases (6%) of familial pulmonary hypertension (disease affecting a first-order blood relative), 7 in men and 5 in women. Patients who had positive family histories were usually diagnosed sooner after the onset of symptoms than were the other registry patients (0.68 compared with 2.56 years; $p = 0.0002$). There were no differences, however, in their ages or hemodynamic findings.

Annals of Internal Medicine 1987, 107, 216-223

Global and regional prevalence of multimorbidity in the adult population in community settings: a systematic review and meta-analysis

126 studií, 15.4 milionů osob z 54 zemí (32.1% mužů, průměrný věk 56.94 ± 10.84 roku)
Definice multimorbidity: více než jedna komorbidita





Chawdhury SR et al. *EClinicalMedicine* 2023, Feb 16;57:101860

Comorbid Conditions and Outcomes in Patients With Pulmonary Arterial Hypertension

A REVEAL Registry Analysis

2959 pacientů s PAH, 78.9 % žen, průměrný věk 52.7 roku v době zařazení
45.9% idiopatická PAH, 51.1%+7% NYHA III+IV
Vznik databáze: 2006 (incidentní a prevalentní pacienti s PAH)

Characteristic	All Patients (N = 2,959)	Hypertension (n = 1,021)	Obesity[a] (n = 956)	Type 2 Diabetes (n = 324)	COPD (n = 498)	Sleep Apnea (n = 599)	Depression (n = 408)	Thyroid Disease[b] (n = 667)	None of the Analyzed Comorbidities[c] (n = 786)
Age at enrollment, mean ± SD, y	52.7 ± 14.7	58.9 ± 12.9	53.3 ± 13.3	58.6 ± 12.3	59.7 ± 12.8	56.3 ± 12.4	53.0 ± 13.4	56.4 ± 13.9	46.3 ± 14.9
Female sex	2,334 (78.9)	802 (78.6)	784 (82.0)	241 (74.4)	364 (73.1)	439 (73.3)	348 (85.3)	606 (90.9)	601 (76.5)
White	2,138 (72.3)	734 (71.9)	710 (74.3)	218 (67.3)	378 (75.9)	469 (78.3)	326 (79.9)	532 (79.8)	528 (67.2)
Etiology									
Idiopathic	1,358 (45.9)	515 (50.4)	505 (52.8)	184 (56.8)	249 (50.0)	353 (58.9)	187 (45.8)	285 (42.7)	330 (42.0)
APAH									
CTD	787 (26.6)	302 (29.6)	201 (21.0)	46 (14.2)	141 (28.3)	104 (17.4)	111 (27.2)	237 (35.5)	187 (23.8)
CHD	285 (9.6)	46 (4.5)	57 (6.0)	19 (5.9)	53 (10.6)	37 (6.2)	29 (7.1)	49 (7.3)	126 (16.0)
PoPH	175 (5.9)	43 (4.2)	54 (5.6)	37 (11.4)	23 (4.6)	31 (5.2)	23 (5.6)	28 (4.2)	45 (5.7)

Obesity was defined as BMI ≥ 30 kg/m^2
Thyroid disease was defined as patients with hyperthyroidism or hypothyroidism and/or patients having taken synthetic thyroid replacement for hypothyroidism.

Poms AD et al. *CHEST* 2013; 144(1):169–176

Comorbid Conditions and Outcomes in Patients With Pulmonary Arterial Hypertension

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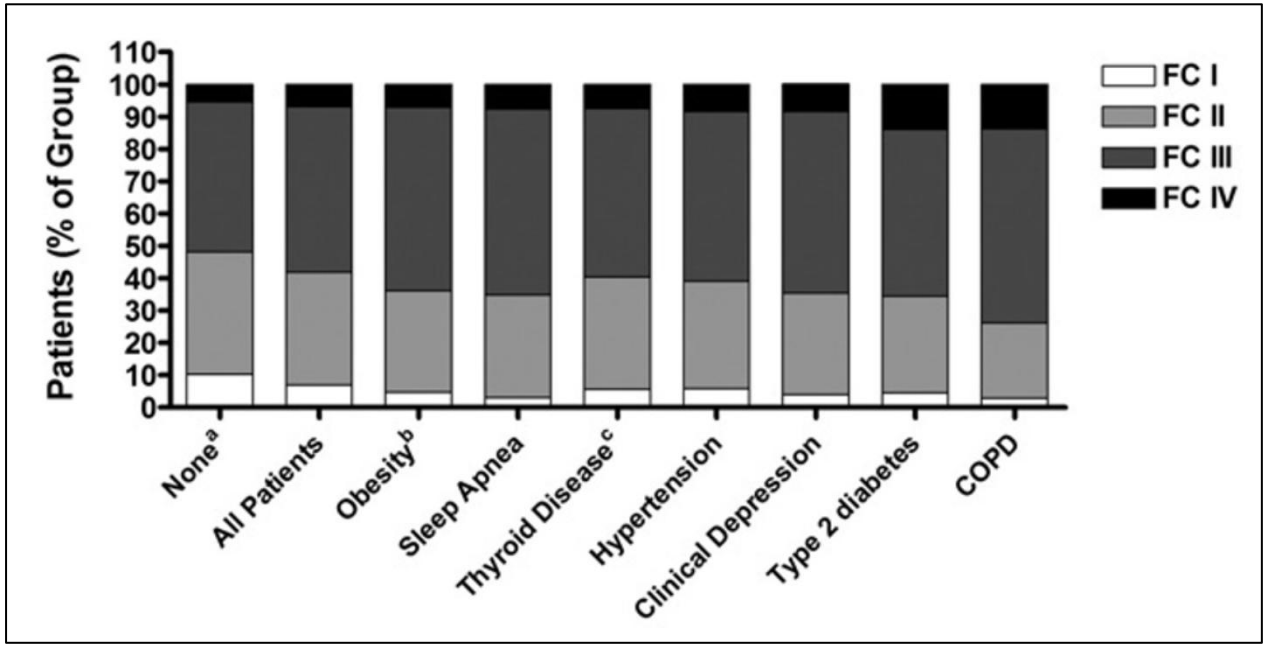
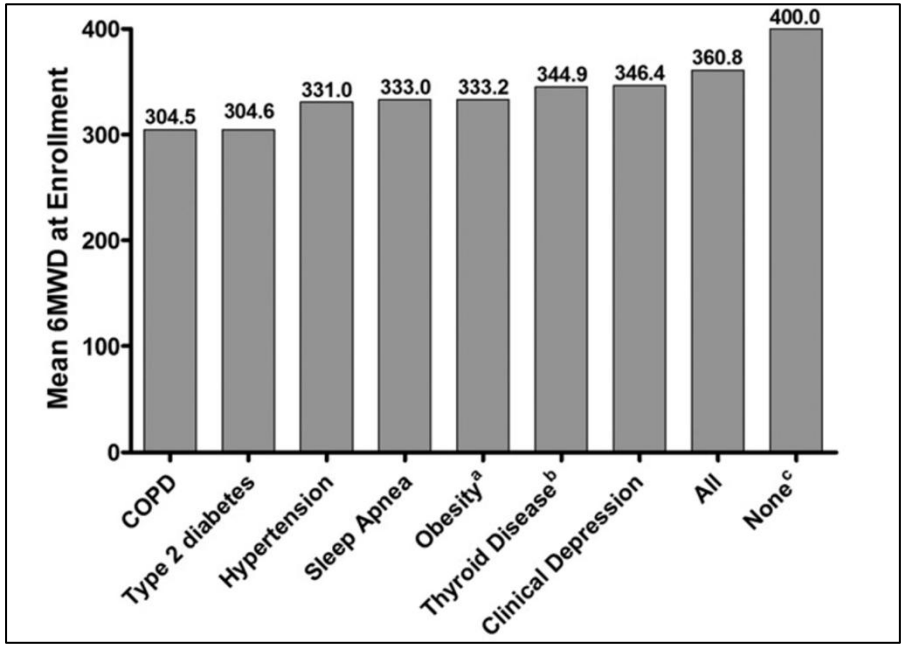


NYHA při zařazení



6MWD při zařazení

Poms AD et al. *CHEST* 2013; 144(1):169–176

Comorbid Conditions and Outcomes in Patients With Pulmonary Arterial Hypertension

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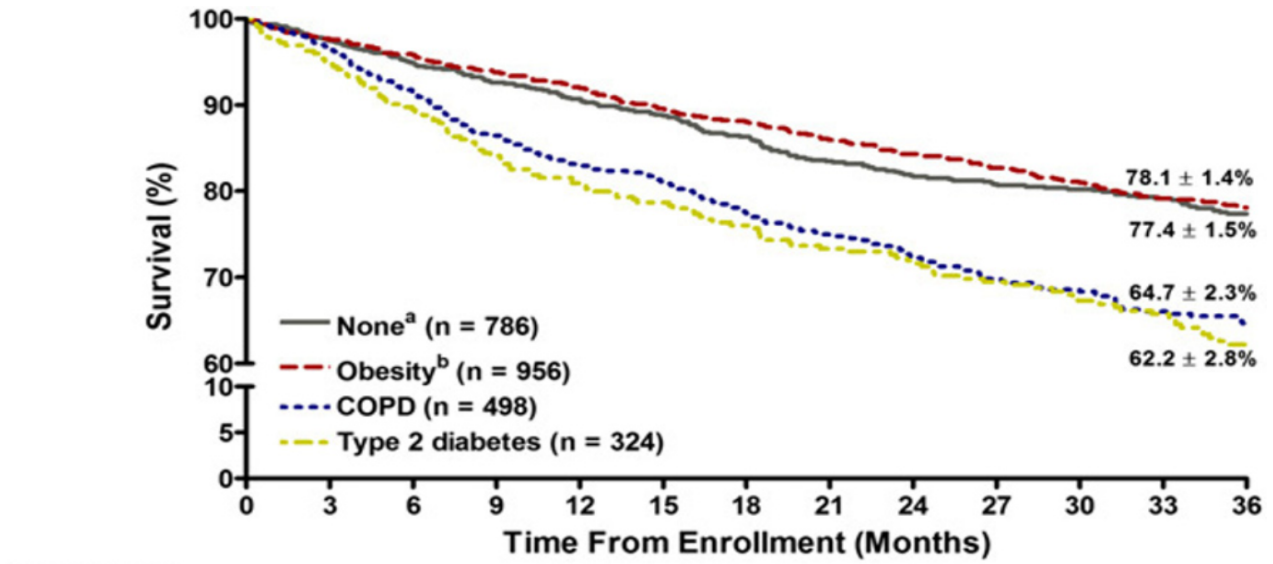


Number at risk:													
None	786	764	744	723	705	681	644	609	593	562	525	512	483
Obesity	956	929	910	882	858	823	790	736	713	677	626	610	589
COPD	498	480	452	423	406	392	358	330	314	289	267	256	239
Type 2 diabetes	324	305	287	268	254	243	230	216	210	195	179	173	158

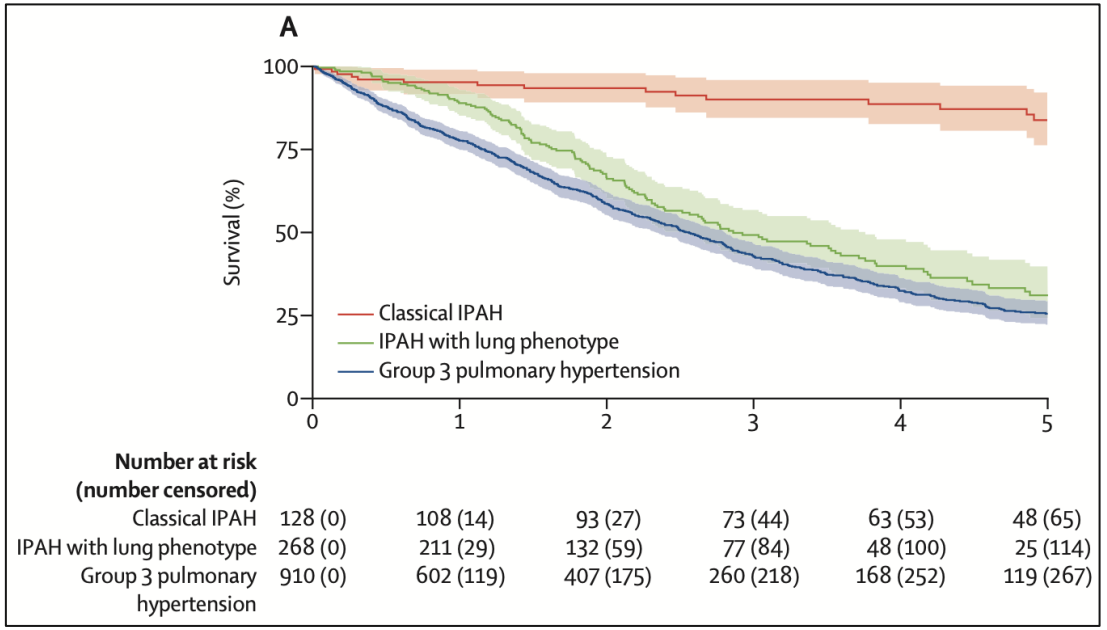
přežití

Poms AD et al. *CHEST* 2013; 144(1):169–176

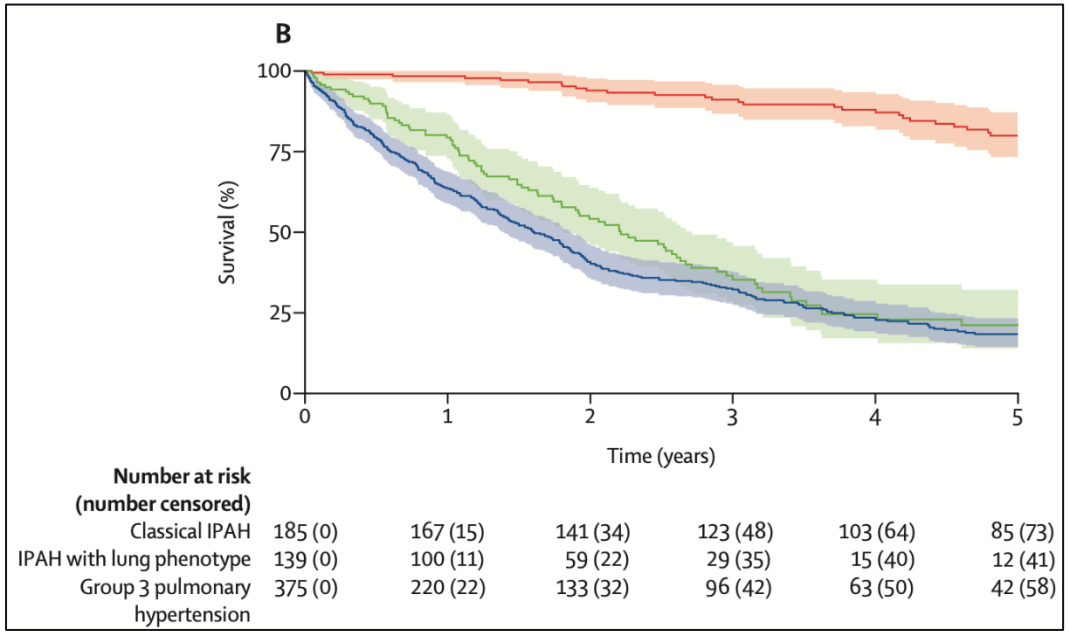
Phenotyping of idiopathic pulmonary arterial hypertension: a registry analysis

COMPERA (n=1306) and **ASPIRE** (699) registries, patient characteristics, response to therapy, survival

- classical IPAH (DLCO ≥ 45%, absence of cardiopulmonary comorbidities)
- IPAH + lung phenotype (DLCO < 45%, smoking history)
- PH due to lung disease (group 3 pulmonary hypertension)



COMPERA registry



ASPIRE registry

Hoeper MM et al. *Lancet Respir Med* 2022 Oct;10(10):937-948

Kazuistika ♀ J.Š., *1948

Anamnéza

- CHOPN
- FVC 1,45l (83%). VC 1,41l (77%), FEV1 0,71l (51%), FEV1/VCmax 48, TLCO 35%, KCO 44%
- Permanentní fibrilace síní
- ICHS, PCI v 2007
- Arteriální hypertenze

Leden 2023, Všeobecná fakultní nemocnice v Praze

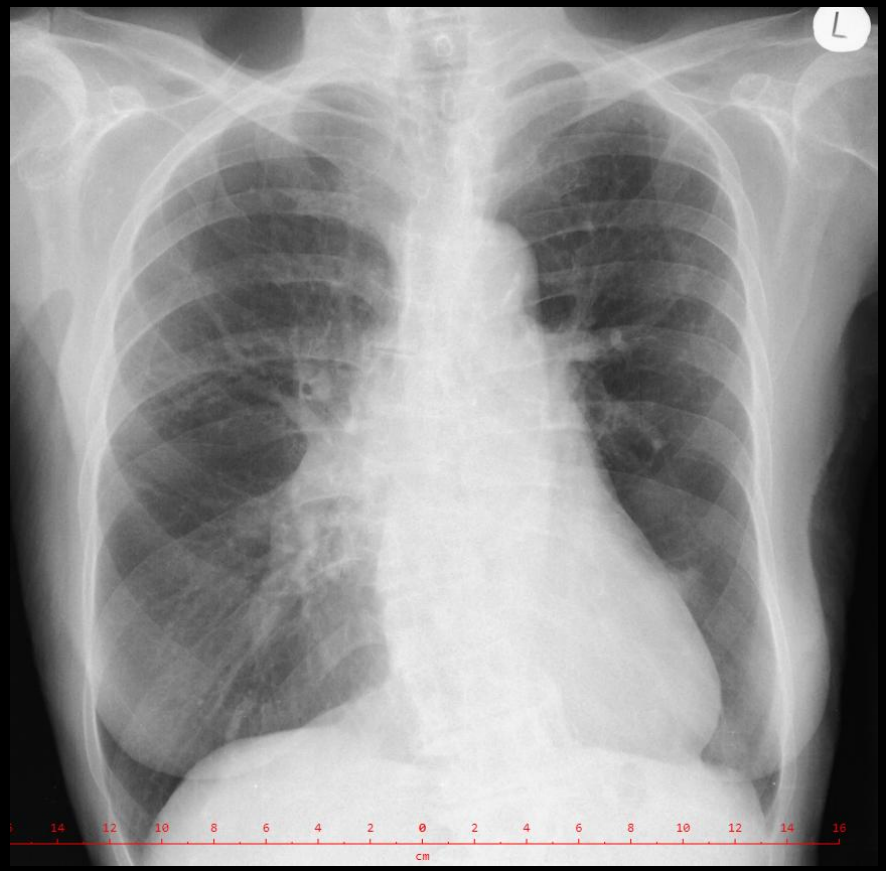
NYHA III, váha 42 kg, váha154 cm, TK 190/110

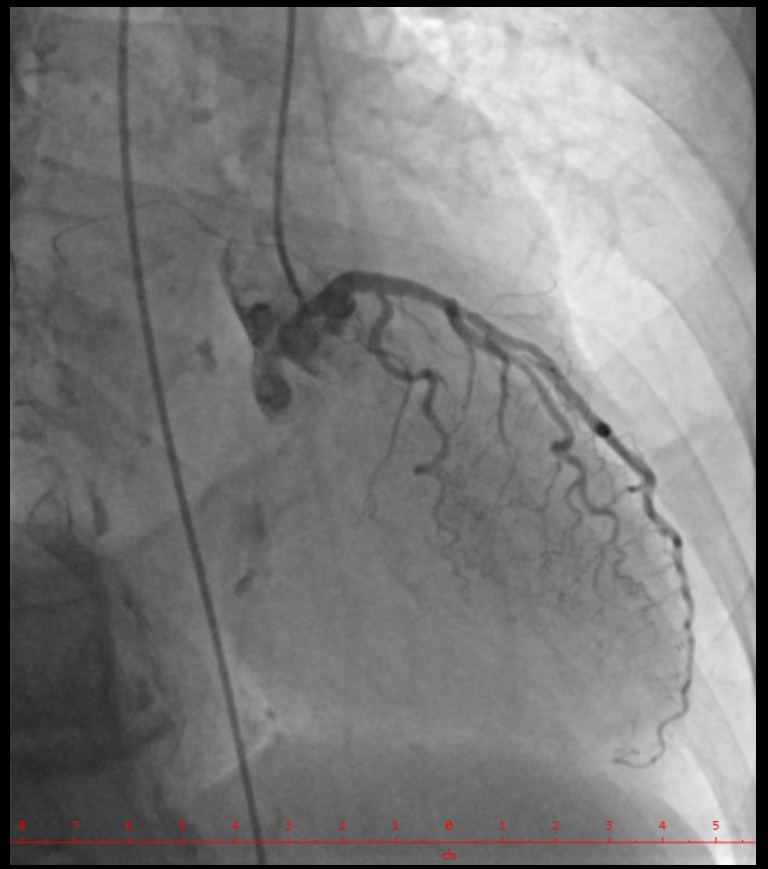
ECHO: EFLK 68 %, LAVi 54 mL/m^2, PASP 78 mmHg
V/Q scintigrafie: souhlasné defekty
6MWT 225 m

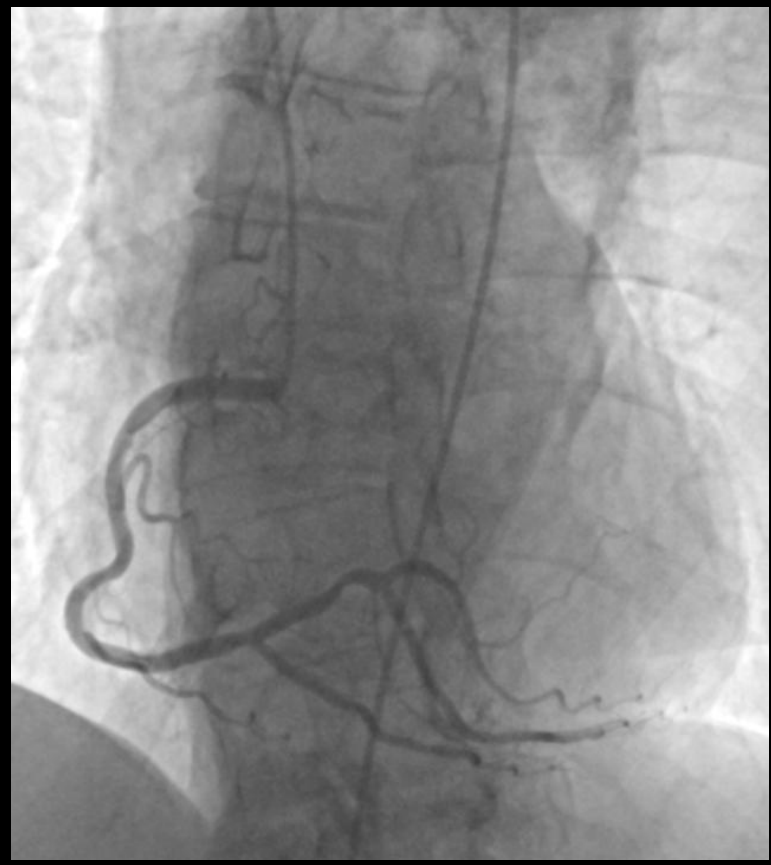
Pro BNP 4377 pg/mL

Hemodynamika:
RA 10 PA 83/42/55, PCW 18, CO 2.70 L/min, CI 1.97 L/min/m^2, PVR 13.7 WU, HR 81/min

Kazuistika ♀ J.Š., *1948







Kazuistika ♀ J.Š., *1948

Klinický závěr:

PAH s kardiopulmonálními komorbiditami

x

PH skupiny 2+3 s těžkou prekapilární komponentou

Léčba PH: Sildenafil 20 mg 3x1 tbl

	NYHA	6MWD (m)	proBNP (pg/mL)	PAMP (mmHg)	PCWP (mmHg)	CO (L/min)	CI (L/min/m^2)	PVR (WU)	SaO_2 (%)	SvO_2 (%)	BP (mmHg)
PH diagnóza (1/2023)	III	225	4377	55	18	2.7	1.97	13.7	95	63	145/94

Kazuistika ♀ J.Š., *1948

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PH Reevaluace (6/2023)	III	229	2348	39	15	3.97	2.39	6.0	97	66	138/89

Kazuistika ♀ J.Š., *1948

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KLINICKÁ KLASIFIKACE PLICNÍ HYPERTENZE (2022)

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- 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

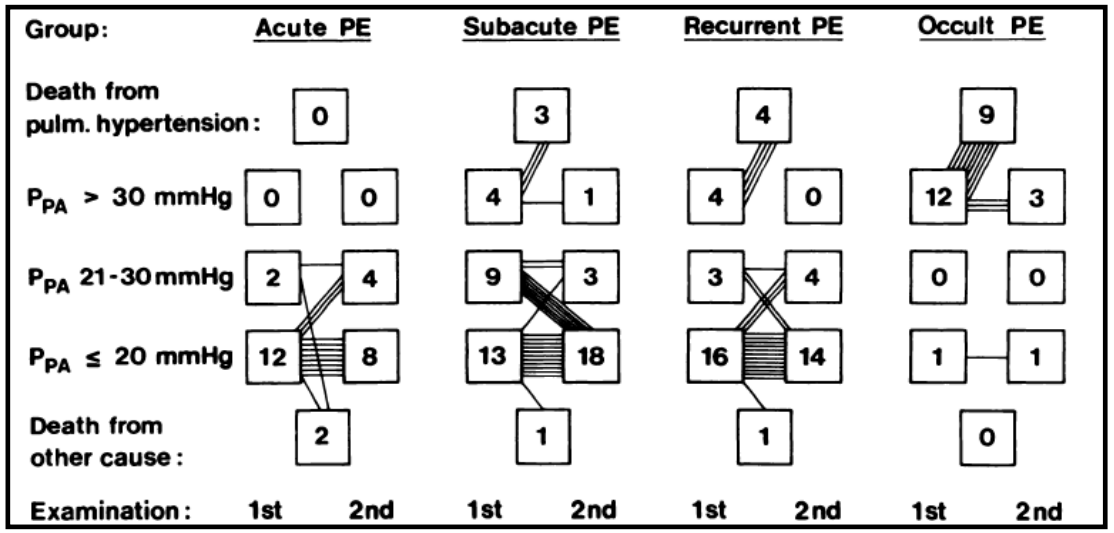
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension
EHJ 2022, *ERJ* 2022

CTEPH – PROGNÓZA NELÉČENÉHO ONEMOCNĚNÍ

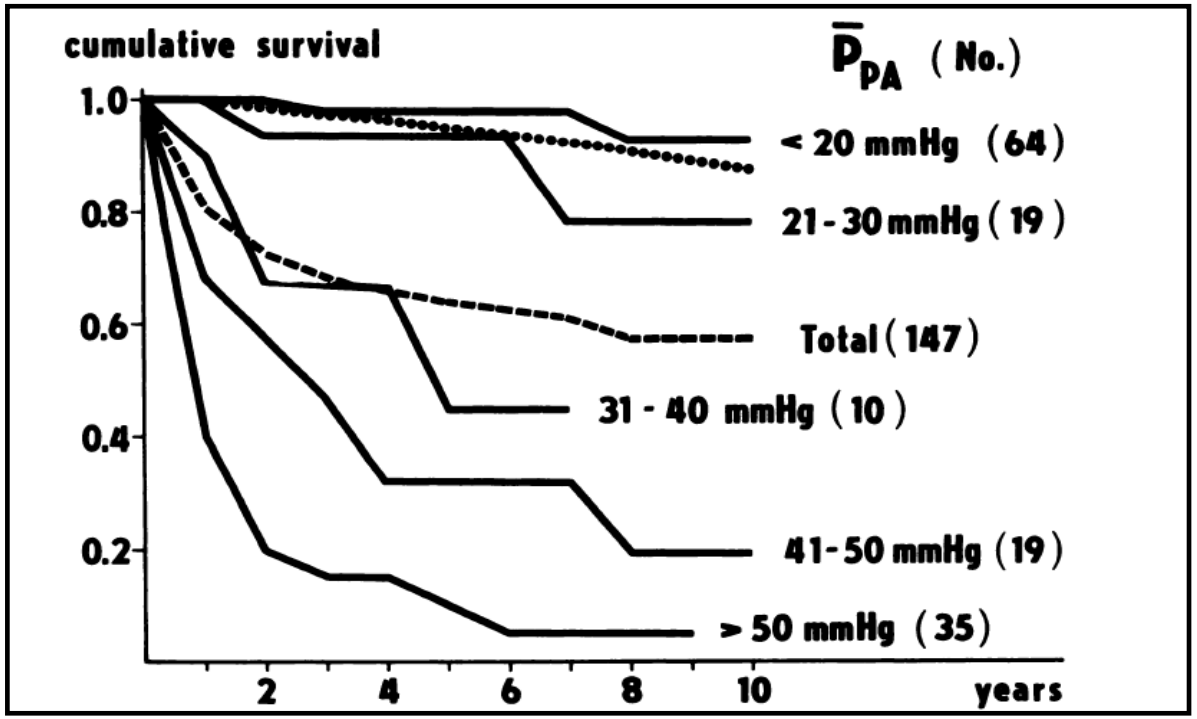
Longterm Follow-up of Patients with Pulmonary Thromboembolism*

Late Prognosis and Evolution of Hemodynamic and Respiratory Data

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

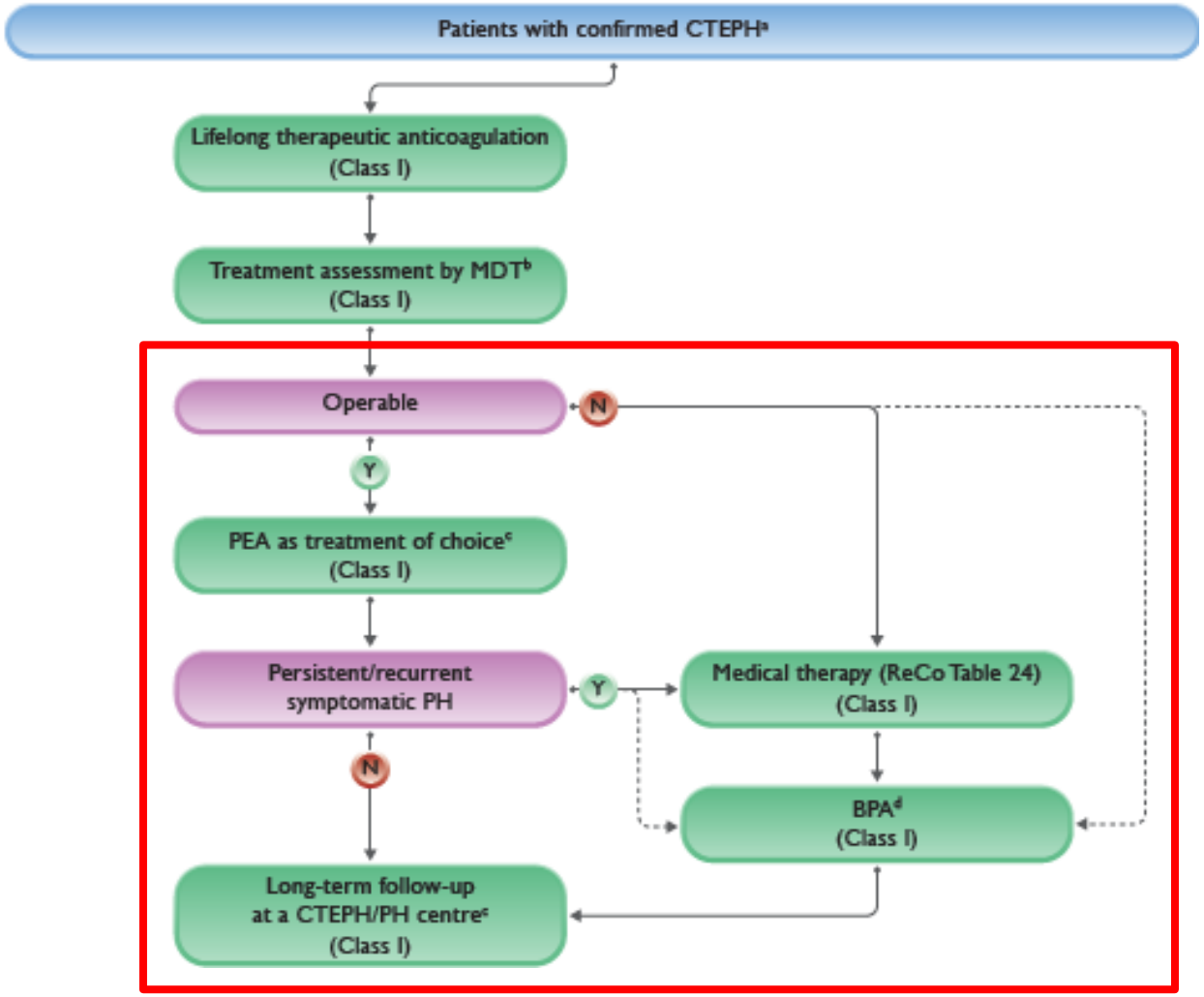


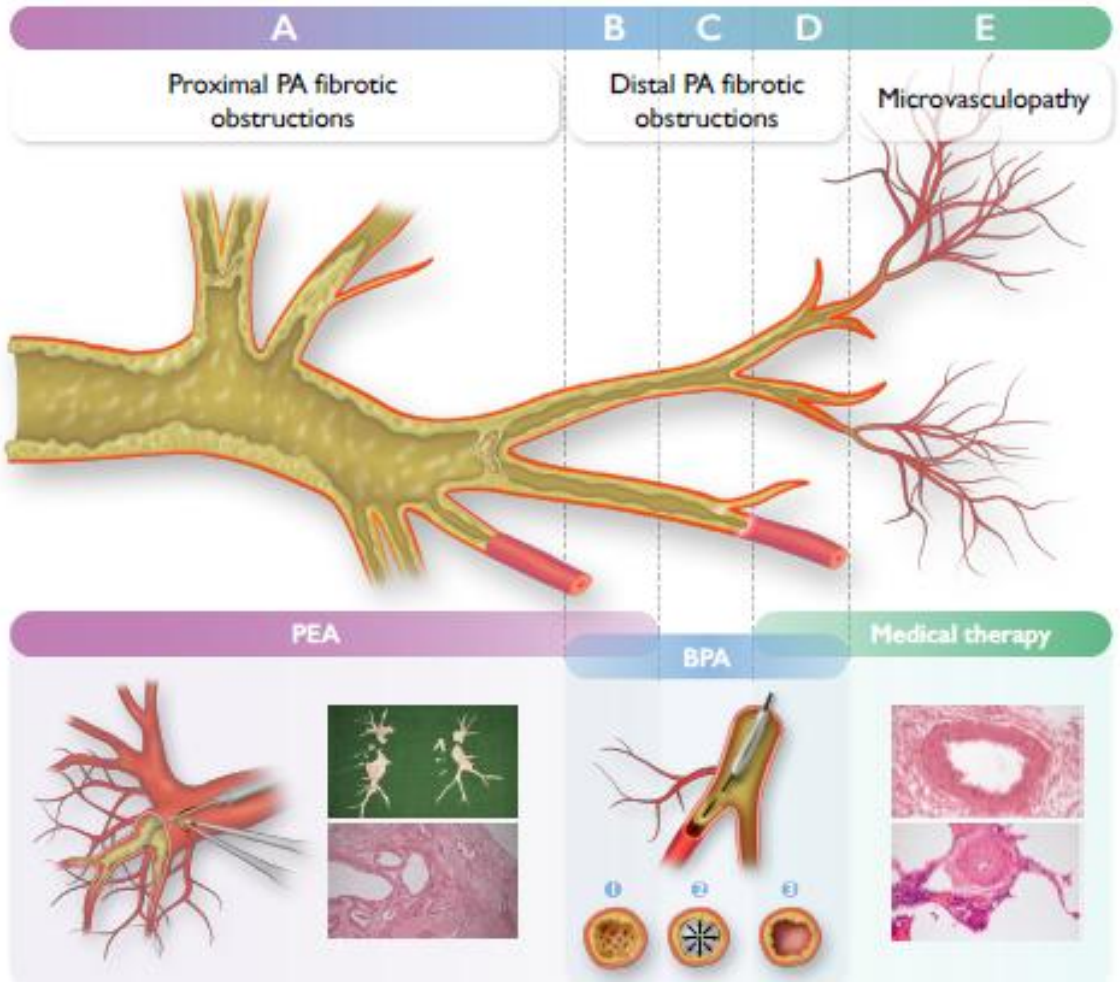
5leté přežití: PAMP > 50 mmHg 10%



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

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

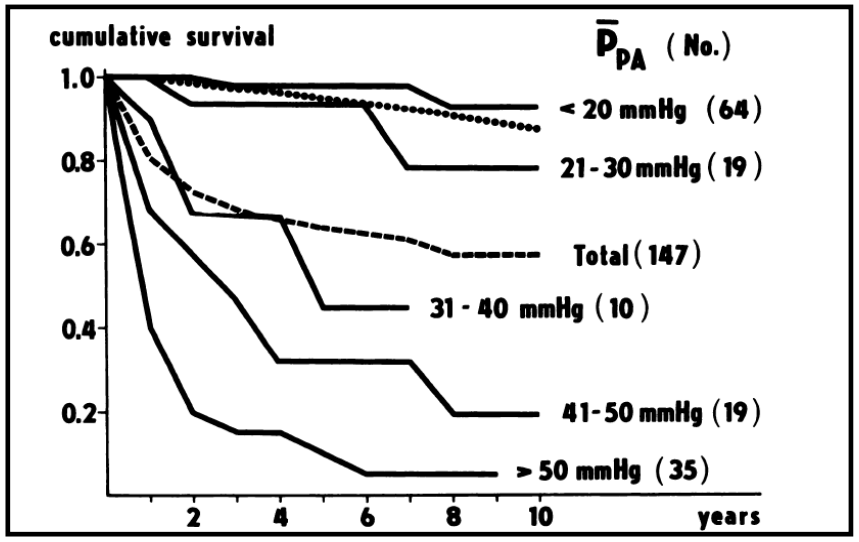




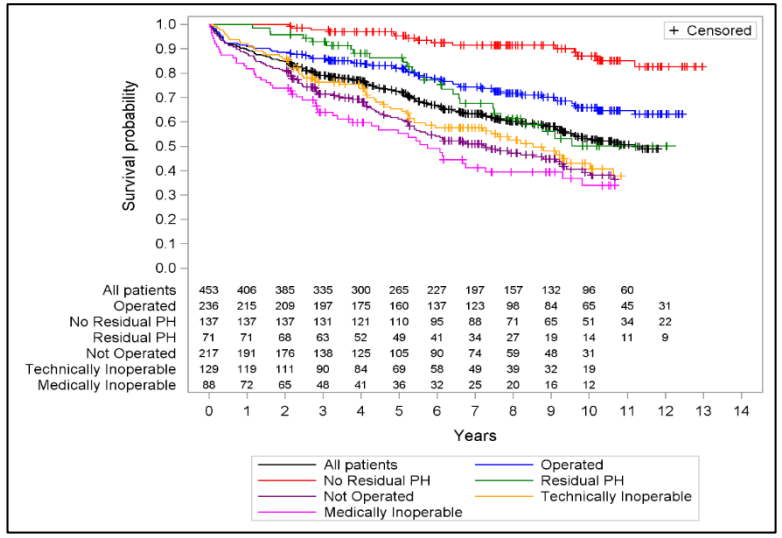
Humbert M et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *EHJ* 2022, *ERJ* 2022

„MULTIMODÁLNÍ“ LÉČBA CTEPH V ČR

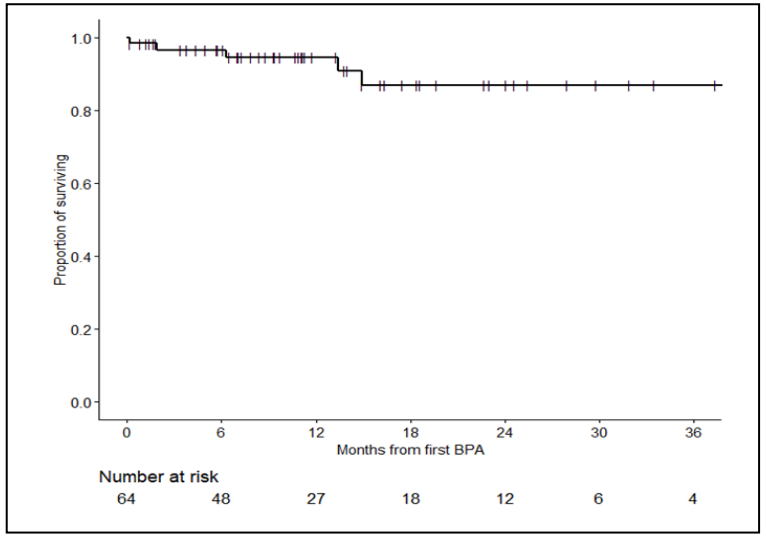
Pravděpodobnost přežití (%)
bez léčby CTEPH



Pravděpodobnost přežití (%)
2004-2016 (PEA)



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



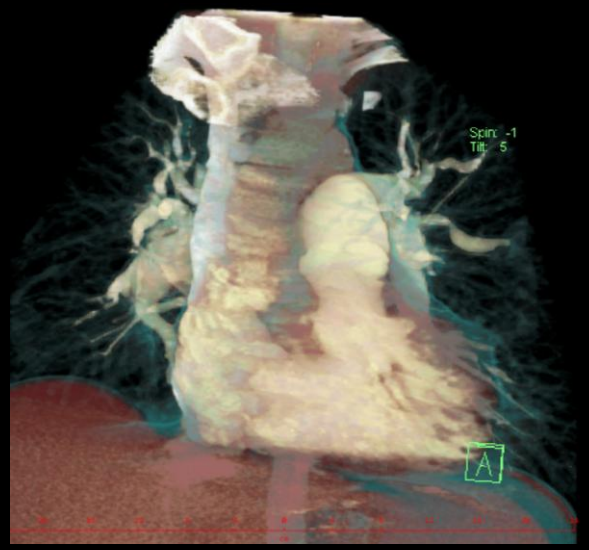
Riedel M, Stanek V, Widimsky J, Prerovsky I, *Chest* 1982, 81: 151-8
Jansa P et al. *Eur Respir J* 2020, 56: 1543; doi: 10.1183/13993003
Jansa P, Heller S et al. *J. Clin. Med*. 2020, 9, 3608; doi:10.3390/jcm9113608

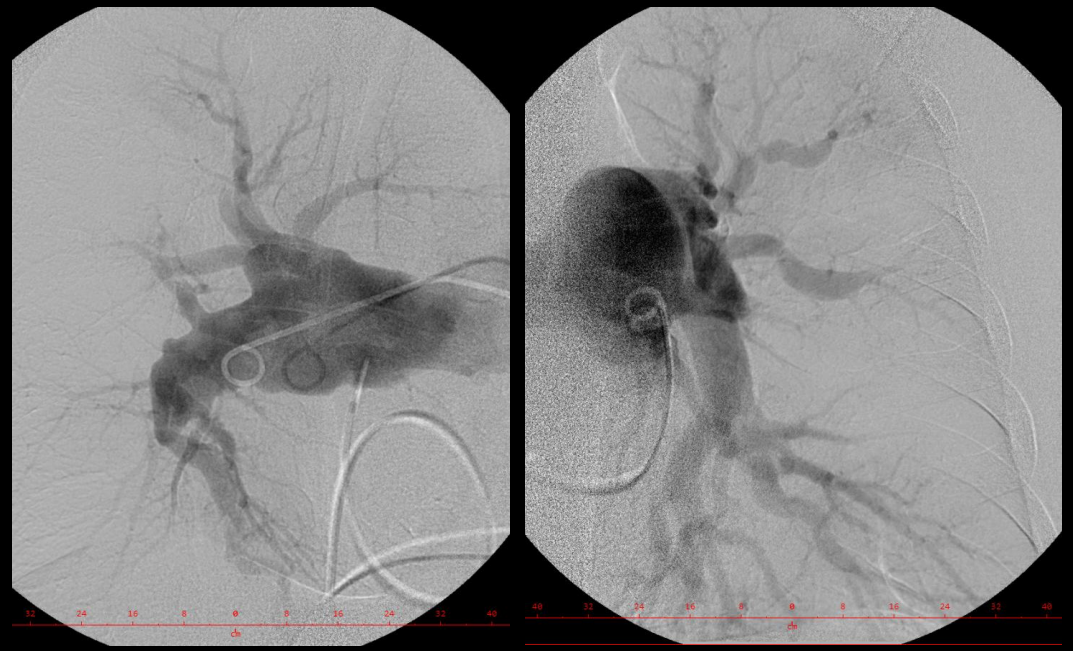
Kazuistika: ♂ ZP, * 6.června 1960

Březen 2009, VFN v Praze

CTEPH, NYHA II-III, 6MWD 460 m, B6
Plicní funkce: TLC 6,63 (89,3%), FEV1 3,72 (95,4%), FVC 4,71 (97,3%), TLCO 10,97 (97%)

Hemodynamika:
RAP 20, PAMP 87, PCWP 14, CO 5.34, CI 2.27, PVR 13,67WU

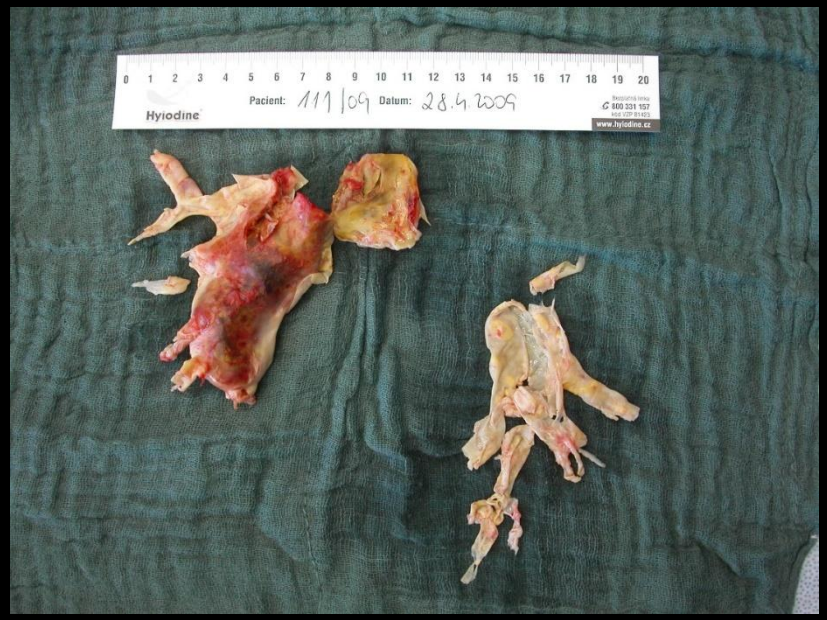




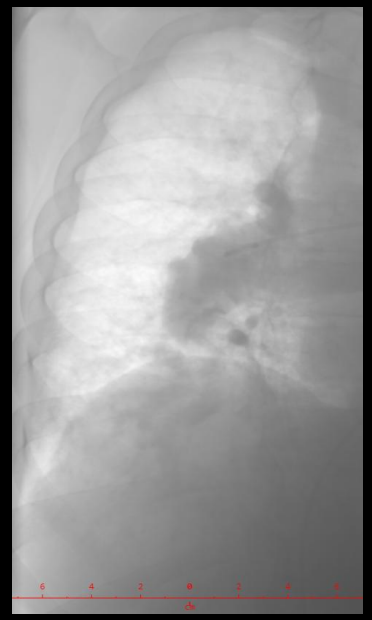
Obrazová dokumentace: VFN Praha

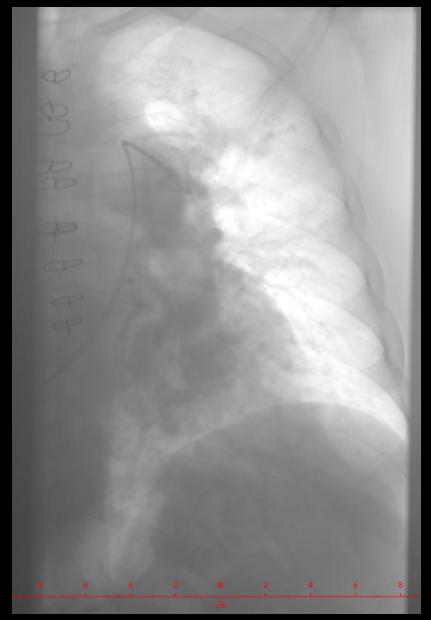
Kazuistika: ♂ ZP, * 6.června 1960

PEA 28.4.2009



Angiografie (4/2015)





	NYHA	6MWD (m)	PAMP (mmHg)	PCWP (mmHg)	CO (L/min)	CI (L/min/m^2)	PVR (WU)
Před PEA (3/2009)	II-III	460	87	14	5.34	2.27	13.7
Po PEA (4/2015)	II	495	65	8	6.0	2.6	9.5

Obrazová dokumentace: VFN Praha, archiv Prof. Lindnera

Kazuistika: ♂ ZP, * 6.června 1960

PEA 28.4.2009**, riociguat od 5/2015**

6xBPA (10/2019…10/2021):

Říjen 2019	A6, A8, A9, A10 vpravo
Listopad 2019	A5, A8, A9, A10 vlevo
Leden 2020	A10, A9, A8, A3 vpravo
Březen 2020	A6, A4, A5, A2, A1 vlevo
Červen 2020	A4, left A2a, A4, A6, A10 vpravo
Říjen 2021	A1, A2, A6a,b, A10 vlevo

	NYHA	6MWD (m)	PAMP (mmHg)	PCWP (mmHg)	CO (L/min)	CI (L/min/m^2)	PVR (WU)
Před PEA (3/2009)	II-III	460	87	14	5.34	2.27	13.7
Po PEA (4/2015)	II	495	65	8	6.0	2.6	9.5
Po rio (4/2019)	II	487	60	9	7.7	2.8	7.7
Po 6 BPA (5/2022)	II	493	53	19	6.4	2.7	5.3

Kazuistika: ♂ ZP, * 6-června-1960

PEA 28.4.2009**, riociguat od 5/2015**

6xBPA (10/2019…10/2021):

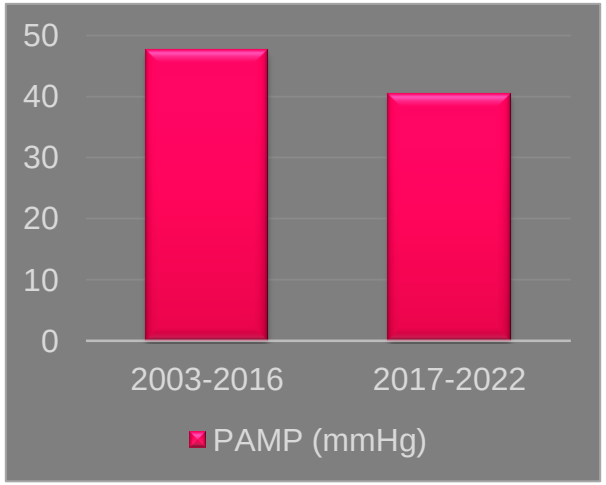
Říjen 2019	A6, A8, A9, A10 vpravo
Listopad 2019	A5, A8, A9, A10 vlevo
Leden 2020	A10, A9, A8, A3 vpravo
Březen 2020	A6, A4, A5, A2, A1 vlevo
Červen 2020	A4, left A2a, A4, A6, A10 vpravo
Říjen 2021	A1, A2, A6a,b, A10 vlevo

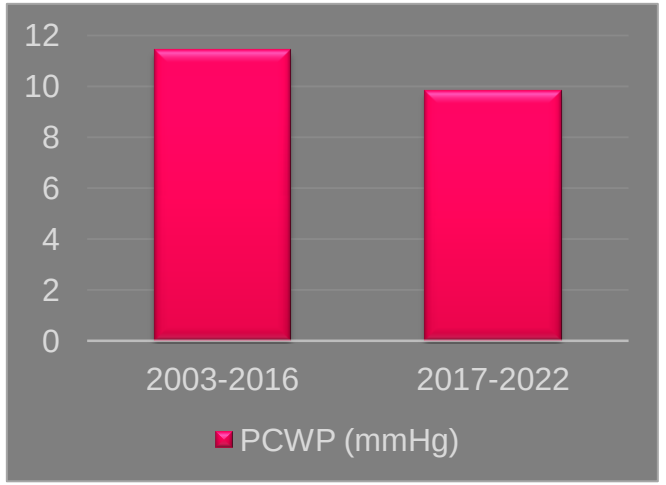
Fibrilace síní	diagnóza 1/2020, elektrická kardioverze v 3/2020, RFA v 3/2022 (recidiva)
OSA	diagnóza v 2021, léčen BiPAP

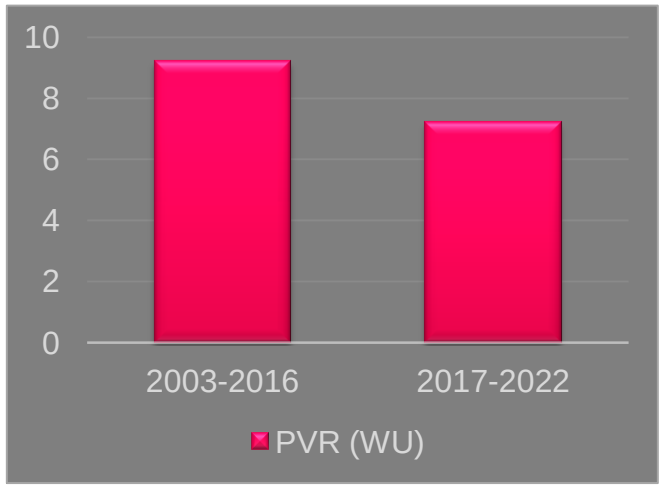
	NYHA	6MWD (m)	PAMP (mmHg)	PCWP (mmHg)	CO (L/min)	CI (L/min/m^2)	PVR (WU)
Před PEA (3/2009)	II-III	460	87	14	5.34	2.27	13.7
Před PEA (4/2015)	II	495	65	8	6.0	2.6	9.5
Po rio (4/2019)	II	487	60	9	7.7	2.8	7.7
Po 6 BPA (5/2022)	II	493	53	19	6.4	2.7	5.3

CTEPH IN THE CZECH REPUBLIC (2003-16 vs 2017-22)

	Year of diagnosis	
	2003-2016, N = 453	2017–2022, N = 321
All patients, n (%)	453 (100.0%)	321 (100.0%)
Operability, n (%)		
Operated	236 (52.1%)	114 (35.5%)
Not operated	217 (47.9%)	207 (64.5%)
Age at diagnosis [years]		
Mean (SD)	63.1 (12.55)	63.9 (13.14)
Median (Range)	65.2 (18.8 - 84.8)	66.9 (19.7 - 87.3)
Sex, n (%)		
Male	247 (54.5%)	169 (52.6%)
Female	206 (45.5%)	152 (47.4%)
BMI		
Mean (SD)	28.4 (5.42)	29.2 (5.79)
Median (Range)	27.8 (17.1 - 58.1)	28.8 (15.8 - 48.3)
Years from 1st PE to diagnosis		
Median (Range)	2.2 (0.0 - 43.2)	1.0 (0.0 - 39.0)
NYHA, n (%)		
I	2 (0.5%)	0 (0.0%)
II	34 (7.8%)	63 (19.9%)
III	385 (88.1%)	240 (75.9%)
IV	16 (3.7%)	13 (4.1%)
6MWD (m)		
Mean (SD)	339.7 (110.15)	387.3 (134.64)

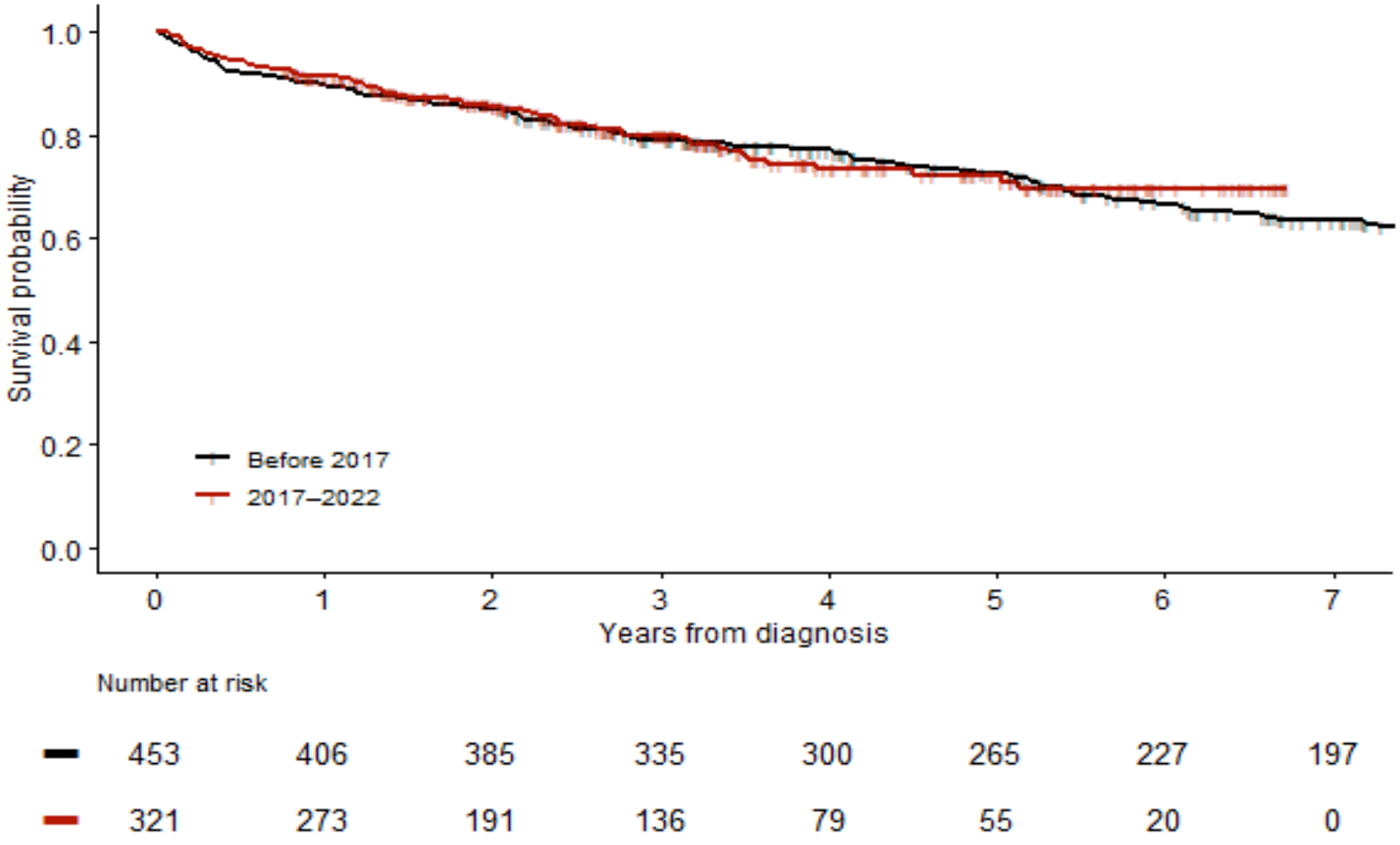






Manuscript in preparation

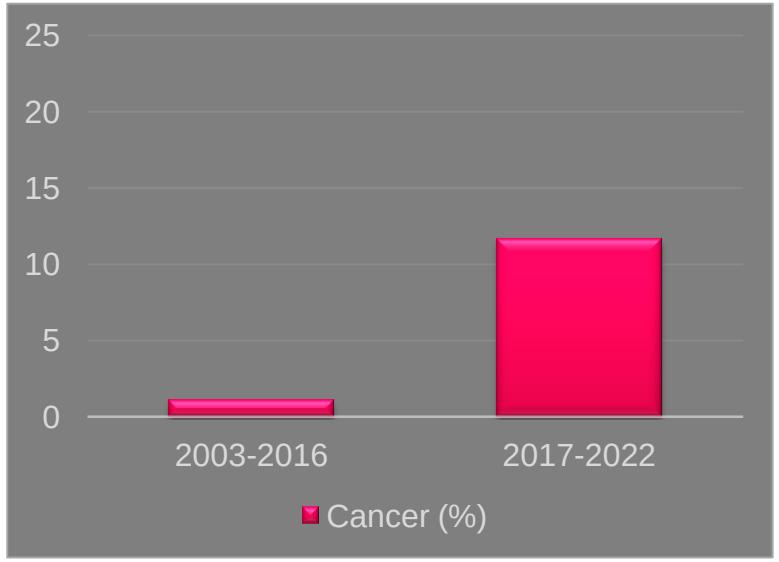
CTEPH IN THE CZECH REPUBLIC (2003-16 vs 2017-22)

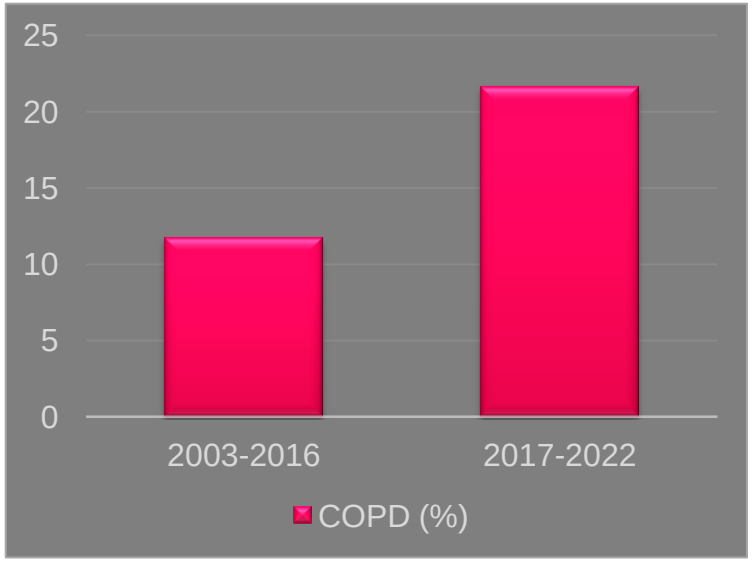


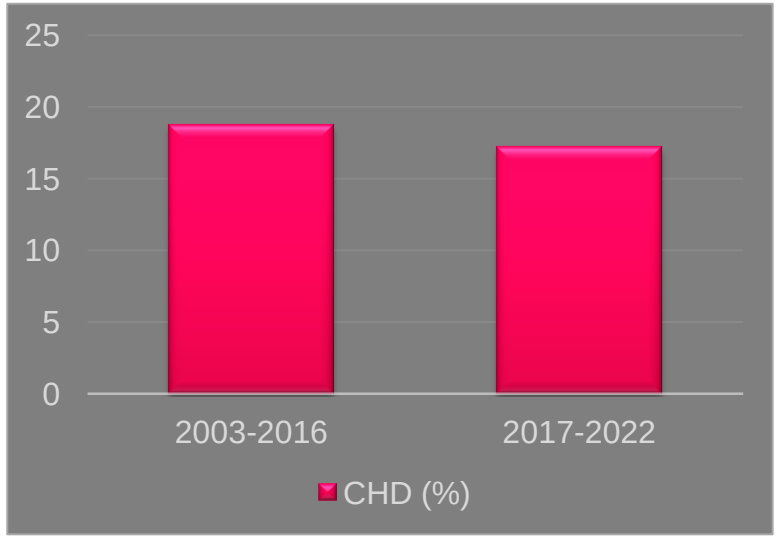
Survival from diagnosis (newly diagnosed CTEPH patients 2003-2016, n=453 vs 2017-2022, n=321)

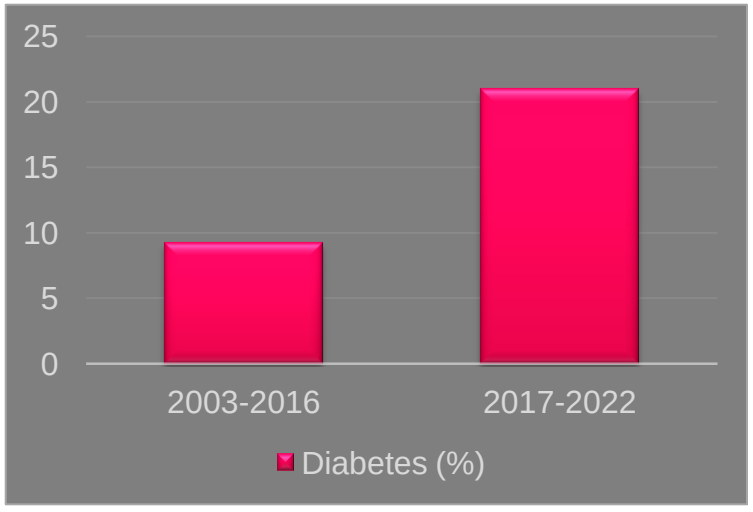
Manuscript in preparation

CTEPH IN THE CZECH REPUBLIC (2003-16 vs 2017-22)



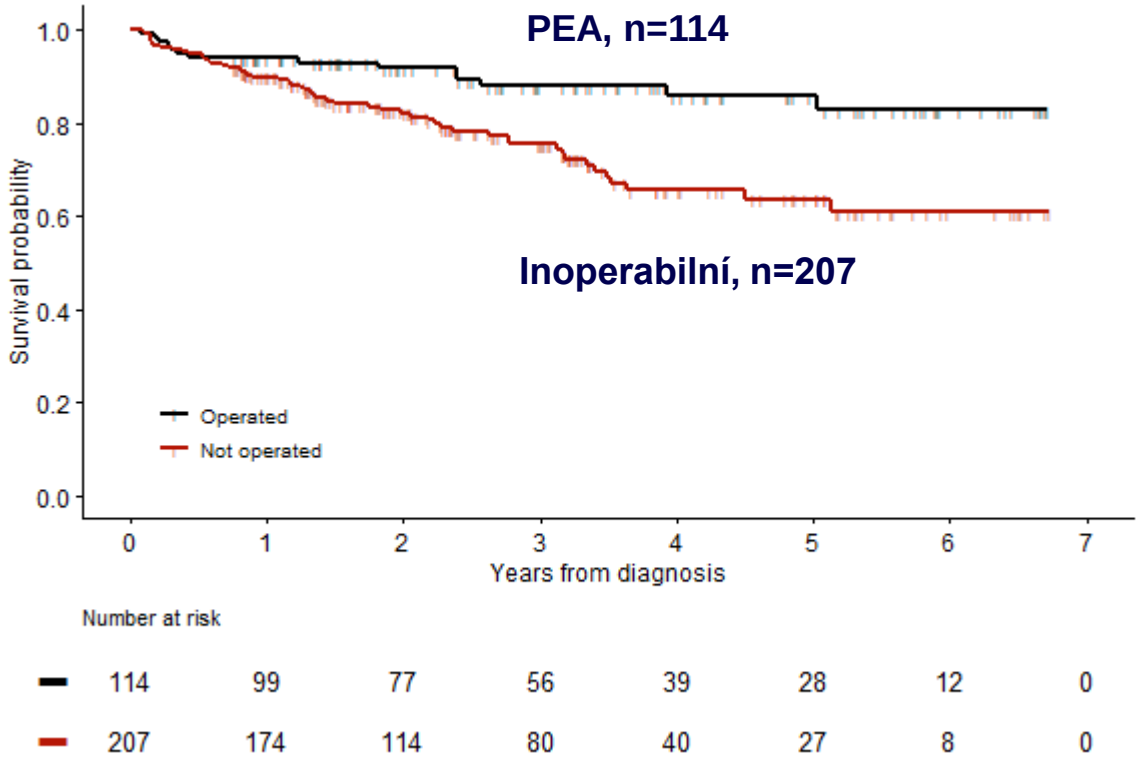






Manuscript in preparation

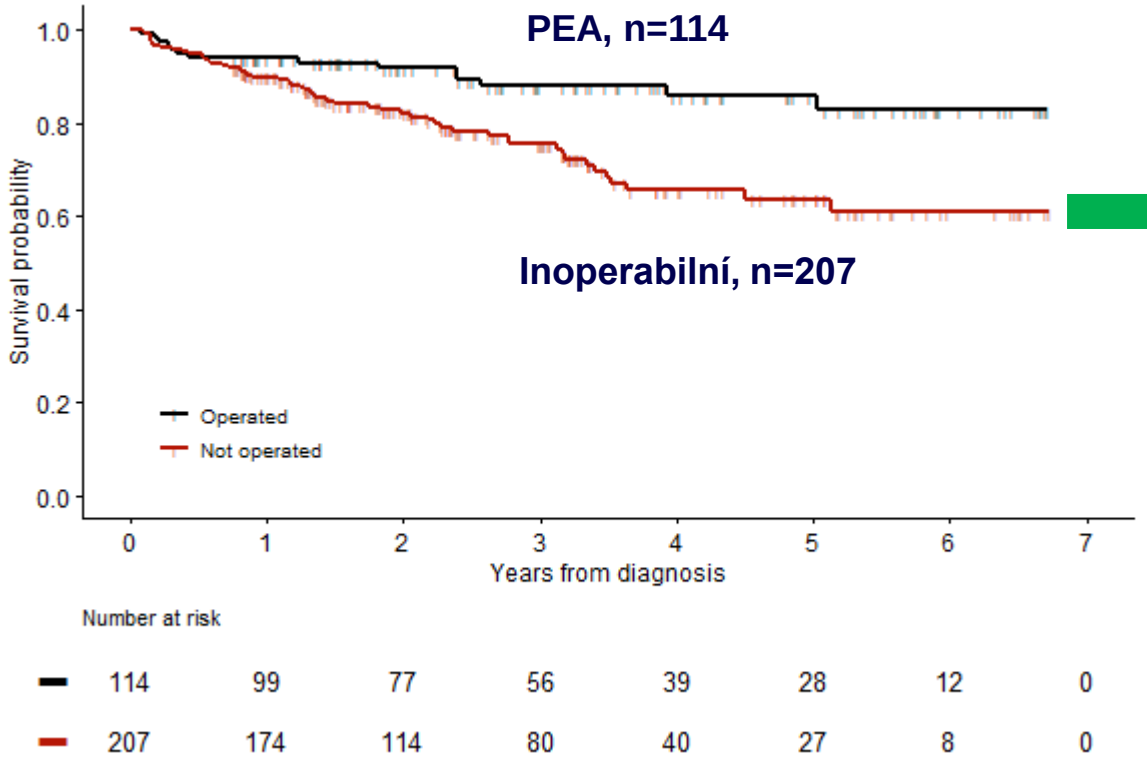
CTEPH IN THE CZECH REPUBLIC (2017-22)

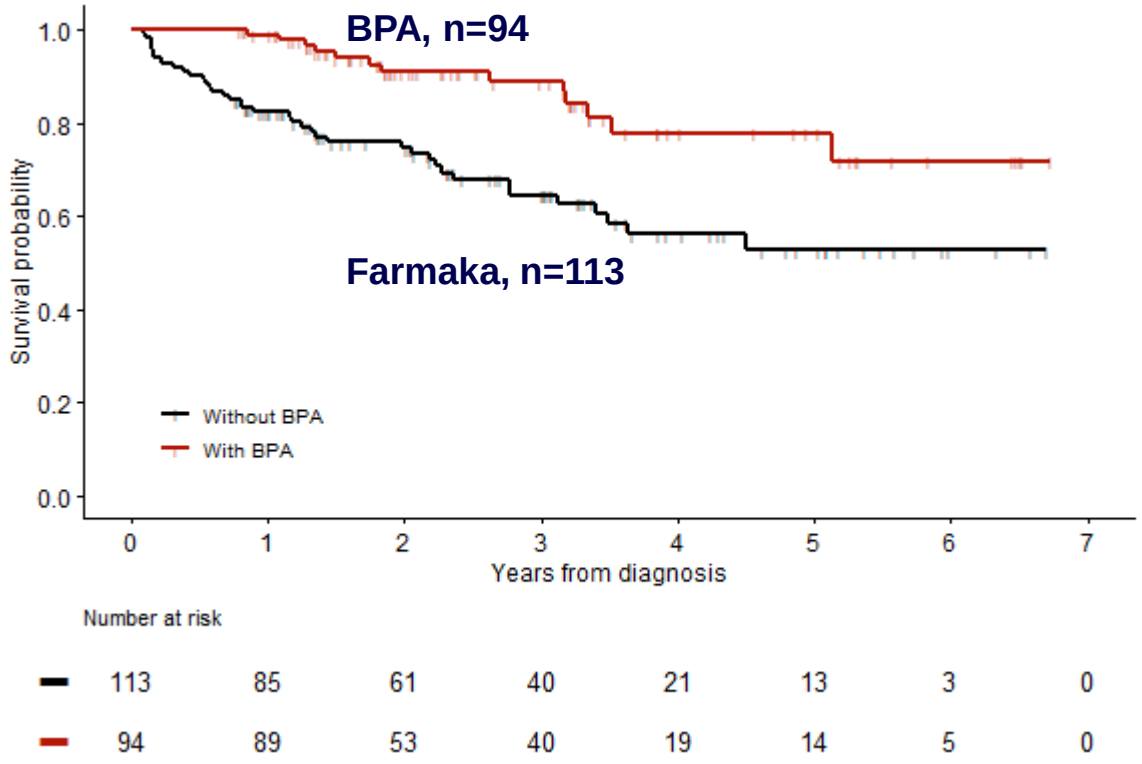


Operated (n=114) vs not operated (n=207)

Manuscript in preparation

CTEPH IN THE CZECH REPUBLIC (2017-22)



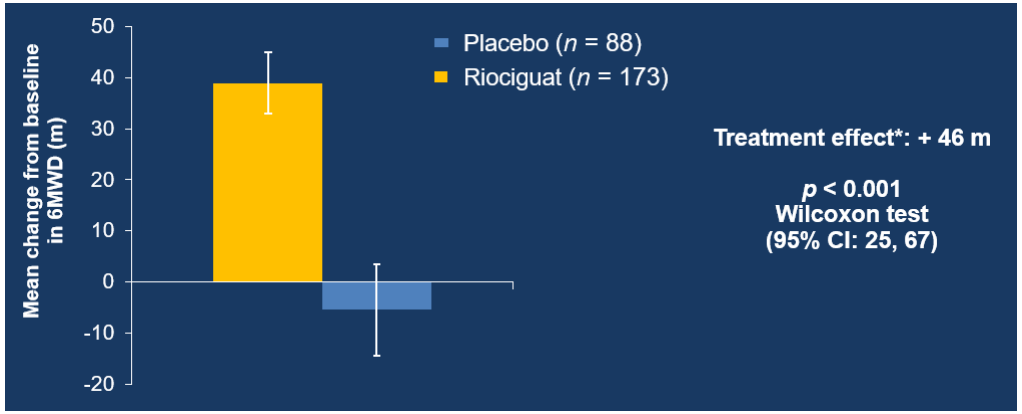


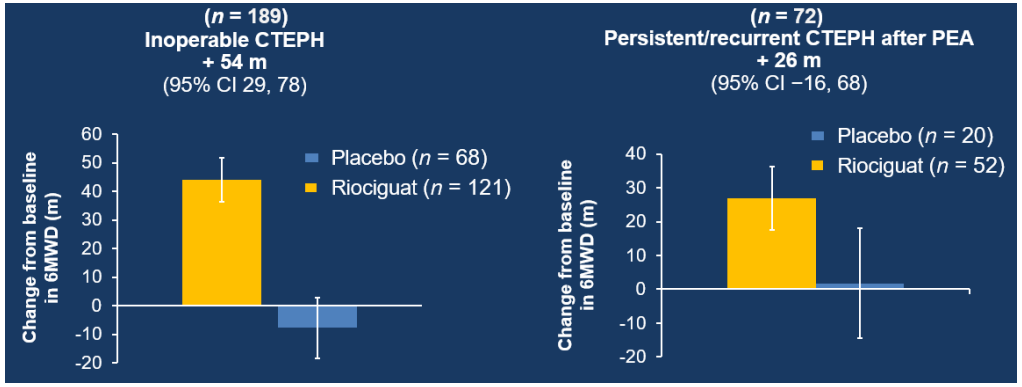
Operated (n=114) vs not operated (n=207)

Not operated + BPA (n=94) vs not operated-no BPA (n=113)

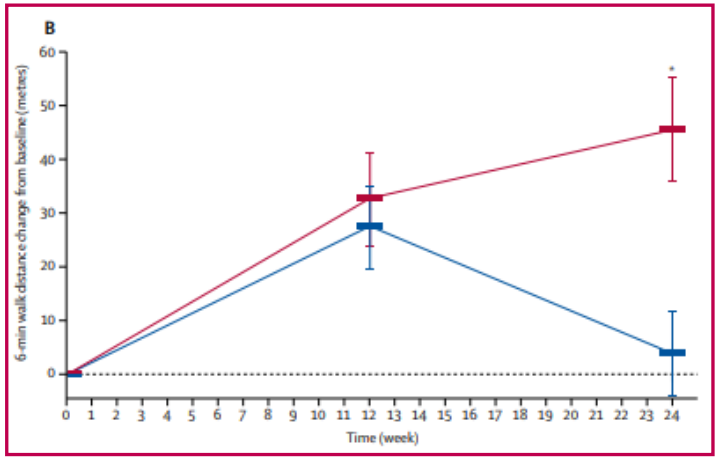
Manuscript in preparation

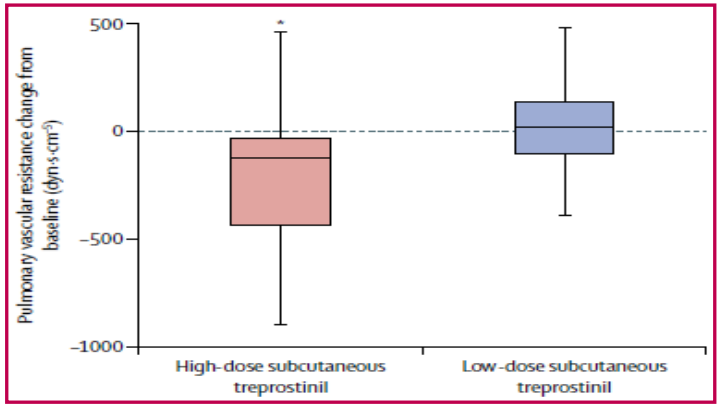
REGISTROVANÁ FARMAKOTERAPIE CTEPH





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





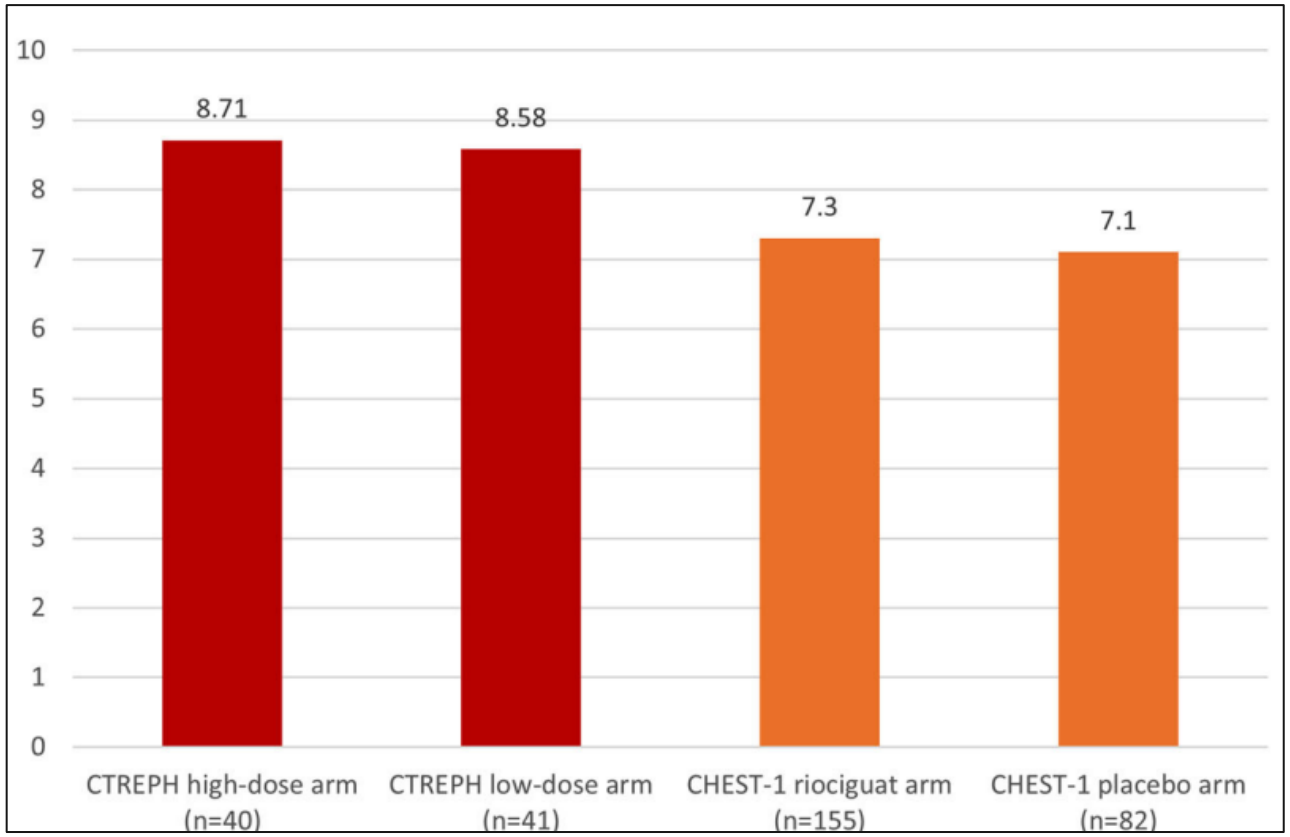
Studie CTREPH (treprostinil), n=105
Inoperabilní, perzistentní CTEPH, věk 64, 24 týdnů

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

The risk profile change in patients with severe chronic thromboembolic pulmonary hypertension treated with subcutaneous treprostinil

Pavel Jansa[1] | Grzegorz Kopeć[2] | Adam Torbicki[3] | Roela Sadushi-Kolici[4] | Ioana-Alexandra Campean[4] | Michael Halank[5] | Iveta Simkova[6] | Regina Steringer-Mascherbauer[7] | Barbara Salobir[8] | Walter Klepetko[9] | Jaroslav Lindner[10] | Irene M. Lang[4]

N=81 (40 high dose, 41 low dose), male sex 57 %, NYHA FC III+IV 95 %, PAPm 49.6 mmHg, PVR 893.5 dyn·s·cm^{-5}



Risk strata: low (score 1 - 7); average (8); moderately high (9); high (10 - 11); very high (≥12)

Jansa P, …Lang IM *Pulmonary Circulation.* 2023;13:e12274.

SHRNUTÍ

- Terapeutické možnosti PAH vyvinuté v posledních 30 letech (>10 přípravků, 3+1 signální cesty) zlepšují především symptomy
- Ovlivnění prognózy je limitováno absencí přímého vlivu léčby na pravou komoru, komorbiditami a pozdní diagnózou
- Pacienti s četnějšími a významnějšími komorbiditami jsou vylučováni z klinických studií
- Multimodální léčba CTEPH zásadně zlepšuje prognózu
- Prognosticky nejzávažnější jsou polymorbidní pacienti bez možnosti mechanické léčby ⇒ optimalizace farmakoterapie (prostanoidy)


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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World Pulmonary Hypertension Day