PLICNÍ (ARTERIÁLNÍ) HYPERTENZE

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European Heart Journal (2014) **35**, 3033–3080
doi:10.1093/eurheartj/ehu283

ESC GUIDELINES

2014 ESC Guidelines on the diagnosis and management of acute pulmonary embolism

The Task Force for the Diagnosis and Management of Acute Pulmonary Embolism of the European Society of Cardiology (ESC)

Endorsed by the European Respiratory Society (ERS)

Authors/Task Force Members: Stavros V. Konstantinides* (Chairperson) (Germany/Greece), Adam Torbicki* (Co-chairperson) (Poland), Giancarlo Agnelli (Italy), Nicolas Danchin (France), David Fitzmaurice (UK), Nazzareno Galiè (Italy), J. Simon R. Gibbs (UK), Menno V. Huisman (The Netherlands), Marc Humbert† (France), Nils Kucher (Switzerland), Irene Lang (Austria), Mareike Lankeit (Germany), John Lekakis (Greece), Christoph Maack (Germany), Eckhard Mayer (Germany), Nicolas Meneveau (France), Arnaud Perrier (Switzerland), Piotr Pruszczyk (Poland), Lars H. Rasmussen (Denmark), Thomas H. Schindler (USA), Pavel Svitil (Czech Republic), Anton Vonk Noordegraaf (The Netherlands), Jose Luis Zamorano (Spain), Maurizio Zompatori (Italy)

European Heart Journal Advance Access published September 15, 2015

European Heart Journal
doi:10.1093/eurheartj/ehv317

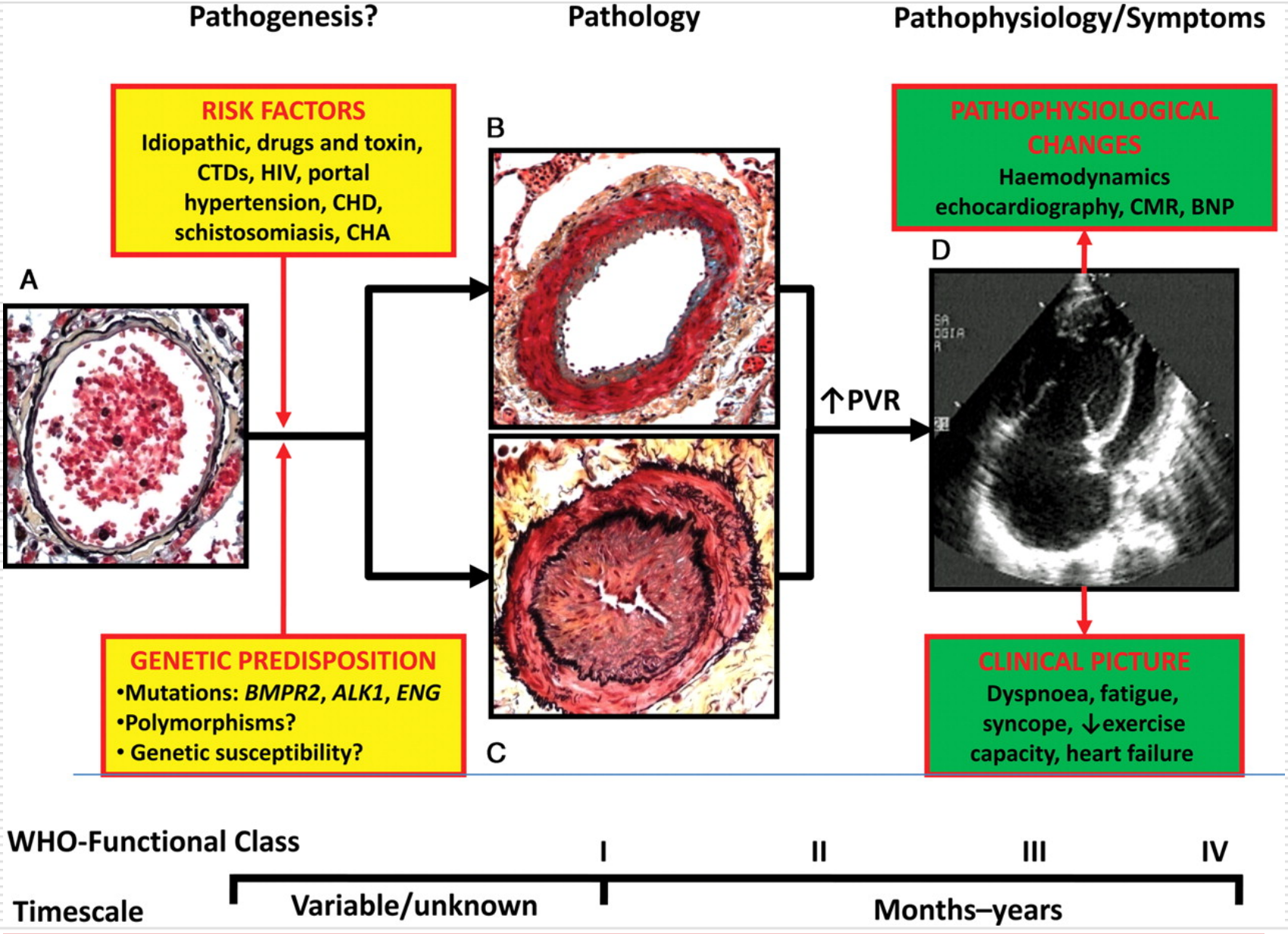
ESC/ERS GUIDELINES

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)

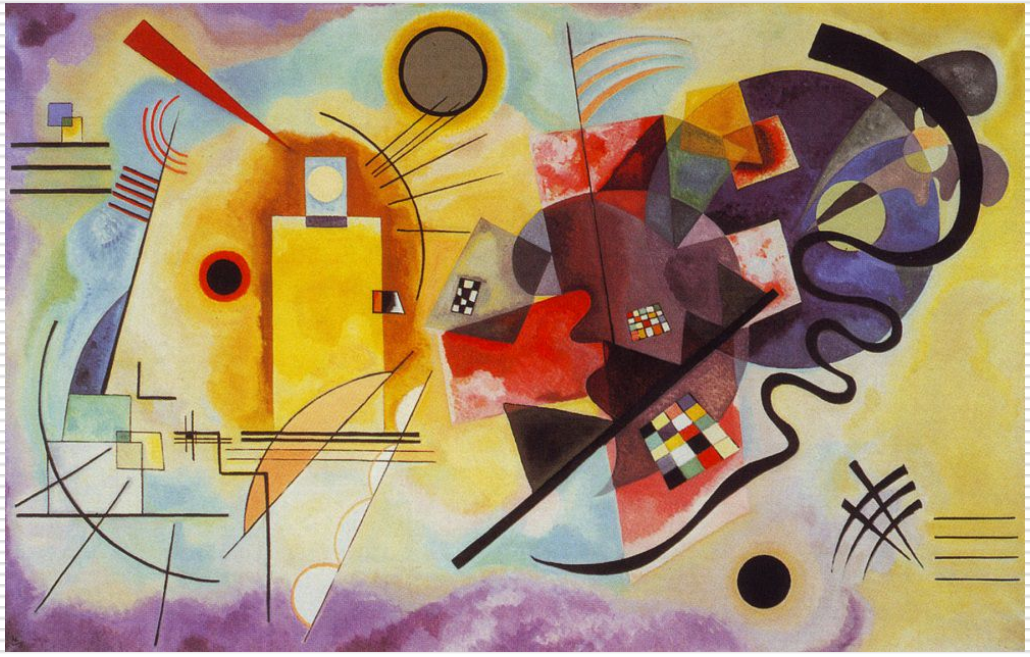
Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

ÚVOD

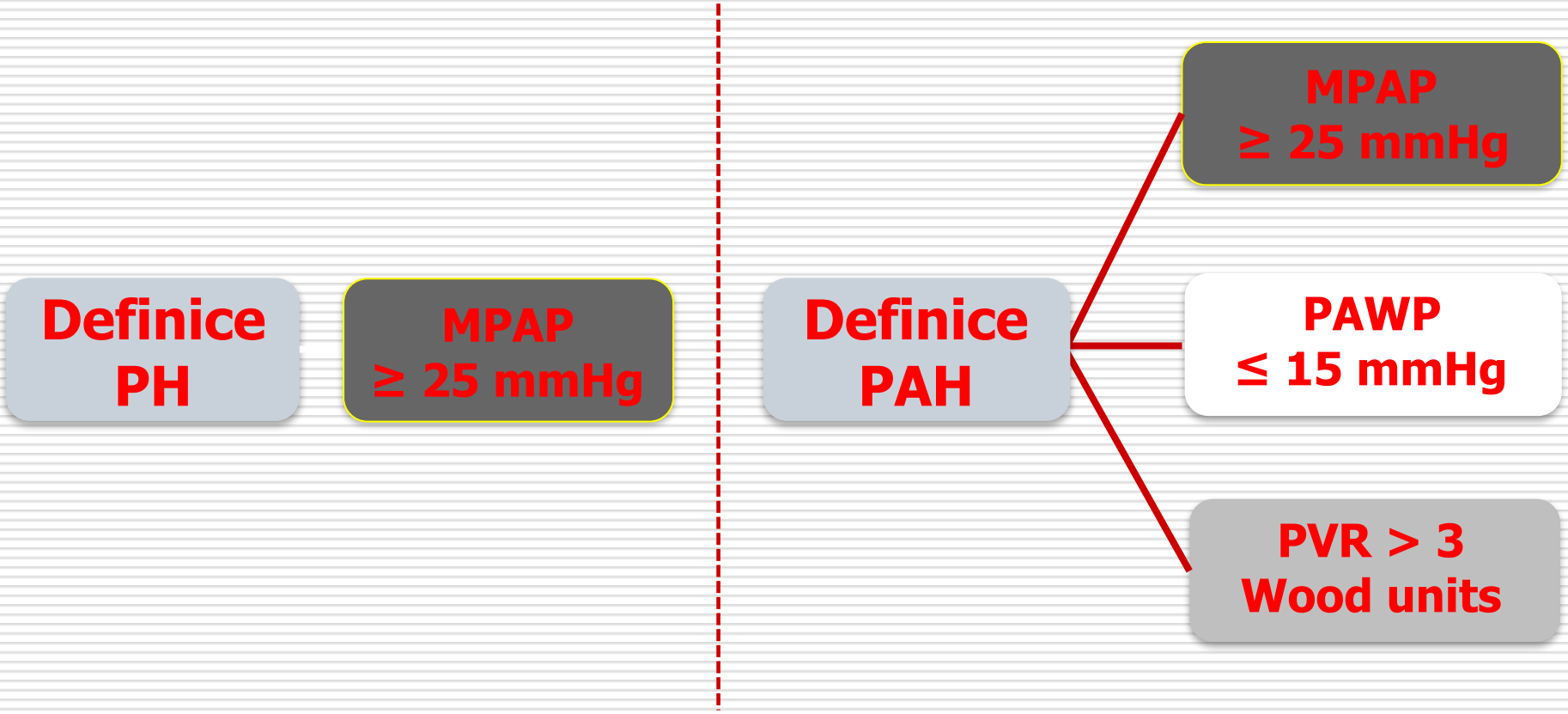


Galiè N et al. Eur Heart J 2010;31:2080-2086

DEFINICE, KLASIFIKACE A PROGNÓZA



Hemodynamická definice plicní hypertenze



PAP: pulmonary arterial pressure; PAWP: pulmonary artery wedge pressure; PVR: pulmonary vascular resistance

Hoeper MM, et al. J Am Coll Cardiol 2013; 62:D42-50.

Plicní hypertenze - klasifikace a epidemiologie

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Updated Clinical Classification of Pulmonary Hypertension

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Definition	Characteristics[a]	Clinical group(s)[b]
PH	PAPm ≥25 mmHg	All
Pre-capillary PH	PAPm ≥25 mmHg PAWP ≤15 mmHg	1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
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]	

PAH **prevalence** and **incidence** are in the range of 15–60 subjects per million population and 5–10 cases per million/year

Prevalence of PAH in at risk populations

- CHD: 4–15%
- Systemic sclerosis: 8–10%
- Portal hypertension: 0.5–10%
- HIV: 0.5%
- Sickle cell disease: 2%
- BMPR2 mutation carriers: 20%

1. Pulmonary arterial hypertension
   - 1.1 Idiopathic PAH
   - 1.2 Heritable PAH
   - 1.2.1 BMPR2
   - 1.2.2 ALK-1, ENG, SMAD9, CAV1, KCNK3
   - 1.2.3 Unknown
   - 1.3 Drug and toxin induced
   - 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 diseases
   - 1.4.5 Schistosomiasis

1′ Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis
1″. Persistent pulmonary hypertension of the newborn (PPHN)

2. Pulmonary hypertension due to left heart disease
   - 2.1 Left ventricular systolic dysfunction
   - 2.2 Left ventricular diastolic dysfunction
   - 2.3 Valvular disease
   - 2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

3. Pulmonary hypertension due to lung diseases and/or hypoxia
   - 3.1 Chronic obstructive pulmonary disease
   - 3.2 Interstitial lung disease
   - 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
   - 3.4 Sleep-disordered breathing
   - 3.5 Alveolar hypoventilation disorders
   - 3.6 Chronic exposure to high altitude
   - 3.7 Developmental lung diseases

4. Chronic thromboembolic pulmonary hypertension (CTEPH)

5. Pulmonary hypertension with unclear multifactorial mechanisms
   - 5.1 Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy
   - 5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
   - 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
   - 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH

Plicní arteriální hypertenze – prognóza onemocnění

CHEST Original Research
PULMONARY VASCULAR DISEASE

An Evaluation of Long-term Survival From Time of Diagnosis in Pulmonary Arterial Hypertension From the REVEAL Registry

Raymond L. Benza, MD; Dave P. Miller, MS; Robyn J. Barst, MD, FCCP; David B. Badesch, MD, FCCP; Adaani E. Frost, MD, FCCP; and Michael D. McGoon, MD, FCCP

CHEST 2012; 142(2):448–456

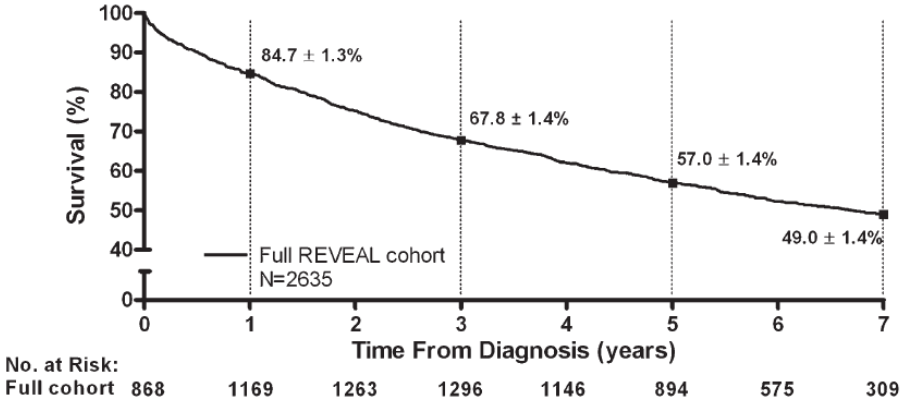


FIGURE 2. Seven-year survival from time of diagnostic right-sided heart catheterization for full REVEAL Registry cohort, using left truncation methods. ■ = estimated survival estimate ± SE at each particular time point. See Figure 1 legend for expansion of abbreviation.

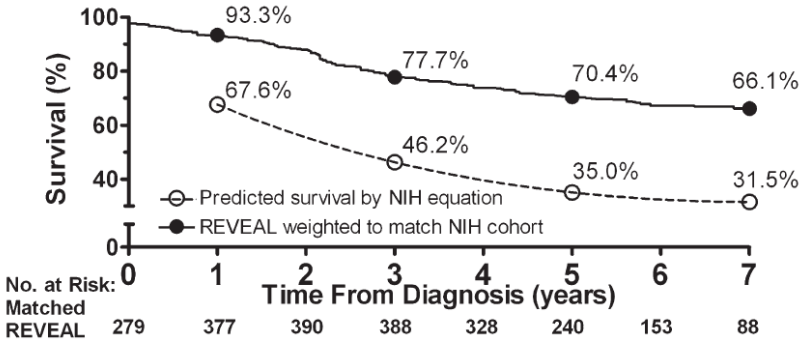


FIGURE 5. Seven-year survival from time of diagnostic RHC of REVEAL Registry cohort weighted to match age, sex, and mean pulmonary artery pressure distribution of NIH cohort. This cohort consisted of patients who met the NIH criteria (ie, had IPAH or FPAH and a pulmonary capillary wedge pressure of ≤ 12 mm Hg), and initiated an endothelin receptor antagonist, phosphodiesterase-5 inhibitor, or prostacyclin analogue within 6 months of diagnostic RHC. See Figure 1 and 4 legends for expansion of abbreviations.

NIH: Incidence 2/1 mil./1 rok, 187 pacientů (průměrný věk 36 let, Ž/M 2:1) sledovaných přes 7 let, mPAP 60 mmHg, CI 2.3 l/min, PVR 26 WU (Rich, Ann Intern Med, 1987)

Characteristic	REVEAL Registry Patients "Traditional Definition" Diagnosed After November 2001 (N = 2,635)	NIH Cohort (N = 187)	Unweighted Comparison Cohort[a] (n = 755)	Weighted Comparison Cohort[b] (n = 755)
Female sex, %	77	63	77	62
Age, y	50 ± 17	36 ± 15	47 ± 18	34 ± 16
mPAP, mm Hg	50 ± 14	60 ± 18	53 ± 13	60 ± 15
mRAP, mm Hg	9.4 ± 6.0	9.7 ± 6.0	9.8 ± 6.0	9.9 ± 5.0
Cardiac index, L/min/m²	2.3 ± 0.9	2.3 ± 0.9	2.2 ± 0.9	2.3 ± 1.1

Patient: _____ Date: _____

WHO Group I Subgroup	APAH-CTD +1	APAH-PoPH +2	FPAH +2	
Demographics & Comorbidities	Renal insufficiency +1	Male age >60 yrs +2		
NYHA/WHO Functional Class	I -2	III +1	IV +2	
Vital Signs	SBP <110 mm Hg +1	HR >92 BPM +1		
6-Minute Walk Test	≥440 m -1	<165 m +1		
BNP	<50 pg/mL -2	>180 pg/mL +1		
Echocardiogram	Pericardial effusion +1			
Pulmonary Function Test	% pred. DLco ≥80 -1	% pred. DLco ≤32 +1		
Right-heart Catheterization	mRAP >20 mm Hg within 1 yr +1	PVR >32 Wood units +2		

APAH=associated PAH; BNP=brain natriuretic peptide; BPM=beats per minute; CTD=connective tissue disease; DLco=carbon monoxide diffusing capacity; FPAH=familial PAH; HR=heart rate; mRAP=mean right atrial pressure; NYHA=New York Heart Association; PAH=pulmonary arterial hypertension; PoPH=portopulmonary hypertension; PVR=pulmonary vascular resistance; SBP=systolic blood pressure; WHO=World Health Organization.

SUM OF ABOVE

(Starting Score) + 6

= RISK SCORE

Risk scores range from 0 (lowest risk) to 22 (highest risk)

	LOW RISK	AVERAGE RISK	MODERATE HIGH RISK	HIGH RISK	VERY HIGH RISK
RISK SCORE	1–7	8	9	10–11	≥12
PREDICTED 1-YEAR SURVIVAL	95%–100%	90%–<95%	85%–<90%	70%–<85%	<70%

United Therapeutics EUROPE LIMITED

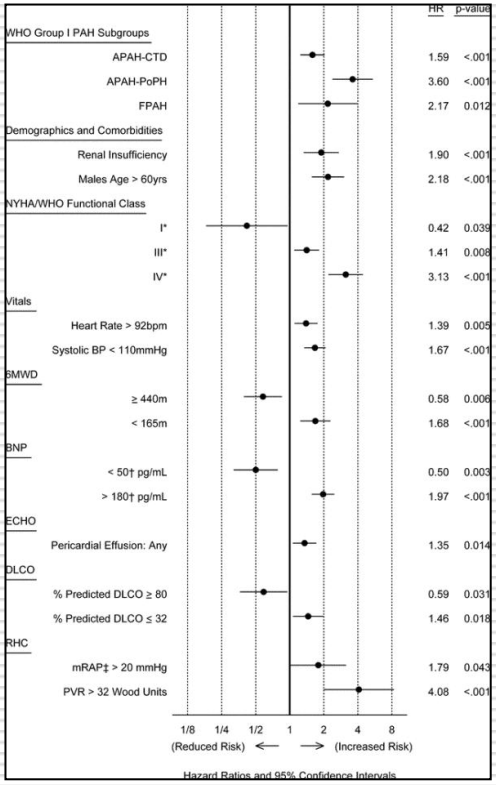
EU/REM/JUL12/248(1). Date of preparation: July 2013.

15 negativních faktorů

4 protektivní faktory

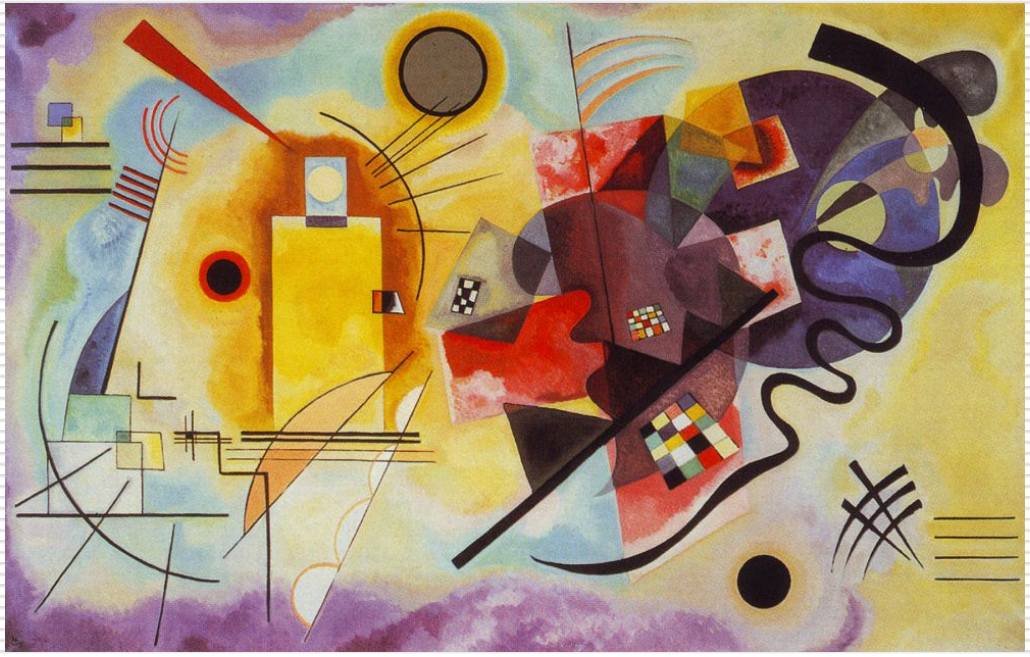
Kalk. rizikové skóre 0-22

Průměrné REVEAL skóre 7.4

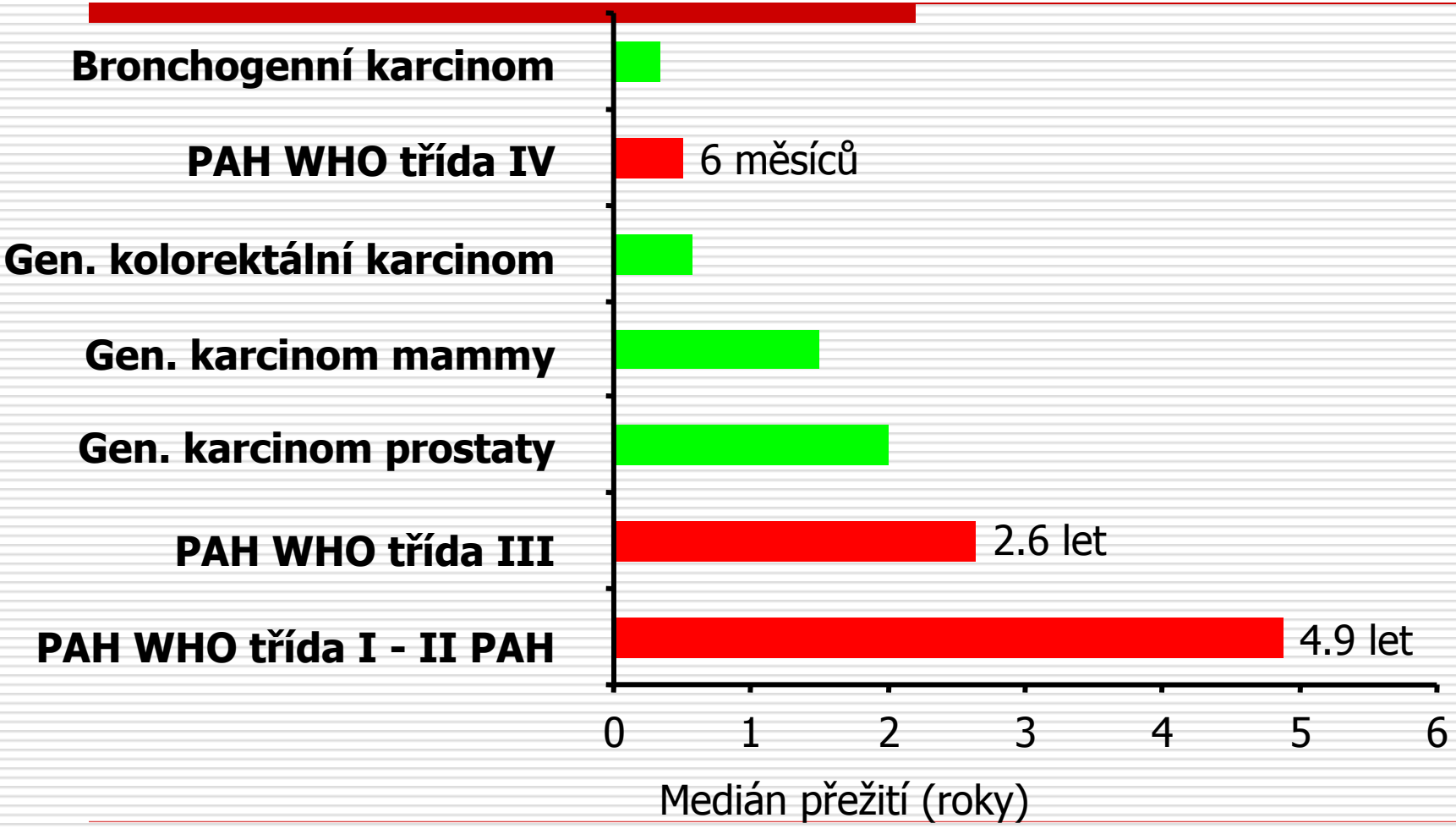


Benza RL; Miller DP; Gomberg-Maitland M; Frantz RP; et al. Predicting survival in pulmonary arterial hypertension: insights from the Registry to Evaluate Early and Long-Term Pulmonary Arterial Hypertension Disease Management (REVEAL). Circulation. 122(2):164-72, 2010 Jul 13.

PRŮBĚH ONEMOCNĚNÍ

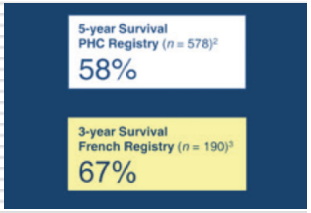


PAH – prognóza neléčeného onemocnění



D'Alonzo et al. Ann Internal Med 1991; Kato et al. Cancer 2001

Podle registru NIH bylo mediánové přežití pacientů s PAH 2,8 roku s 1, 3, 5-letým přežitím 68%, 48% a 34%.



CHEST
Original Research
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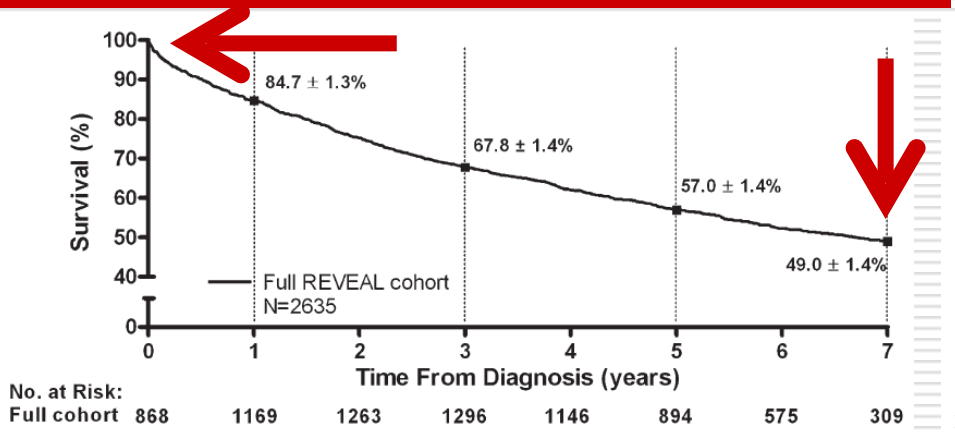

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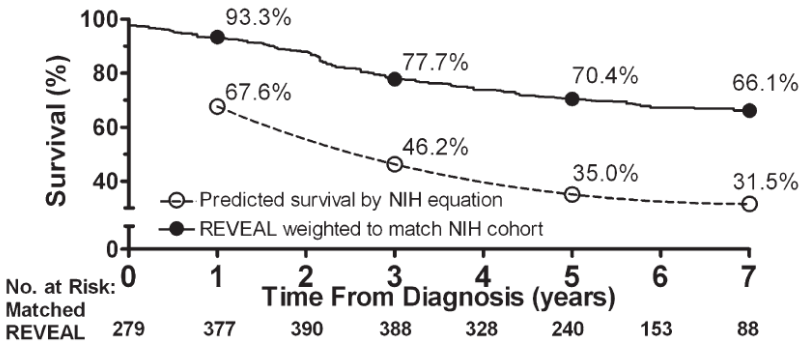

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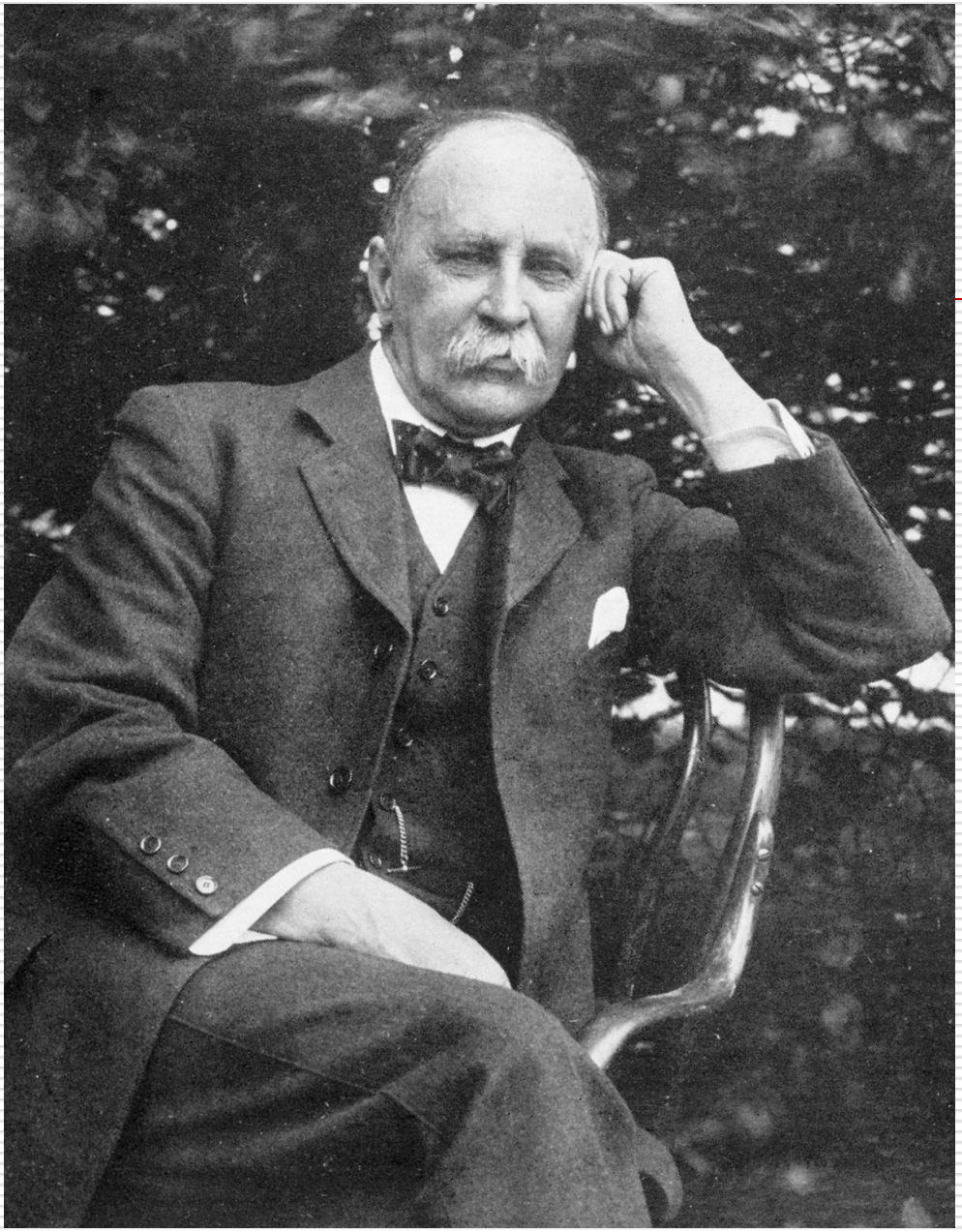
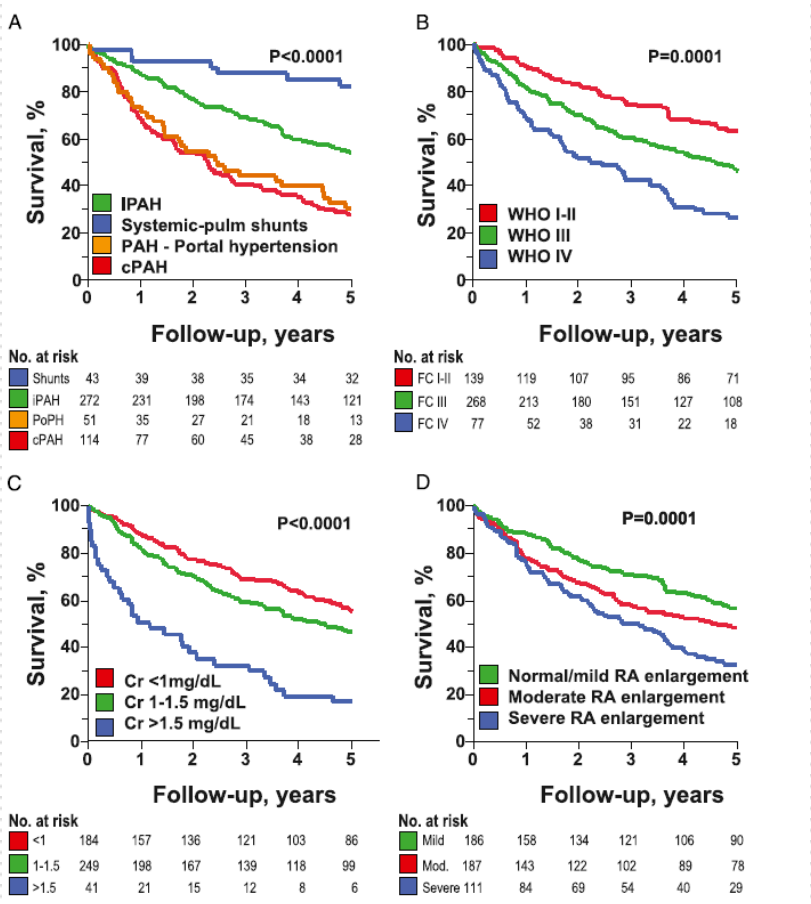
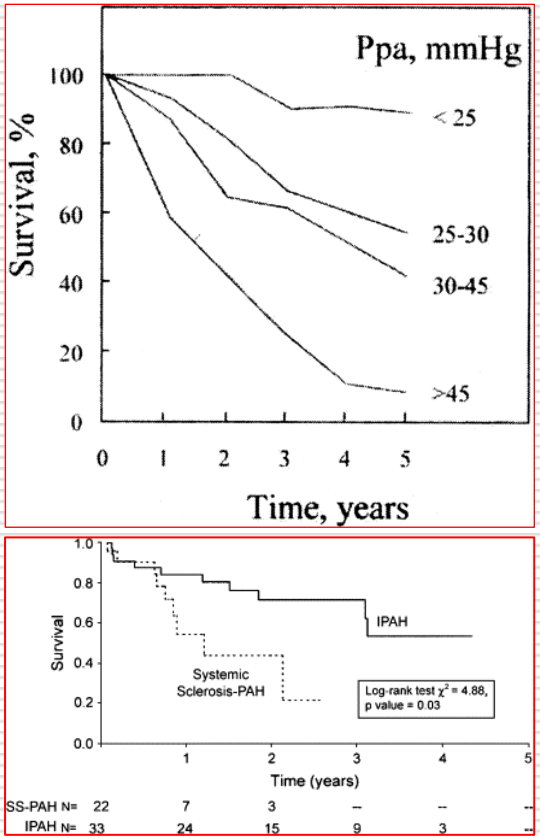
PROGNÓZA



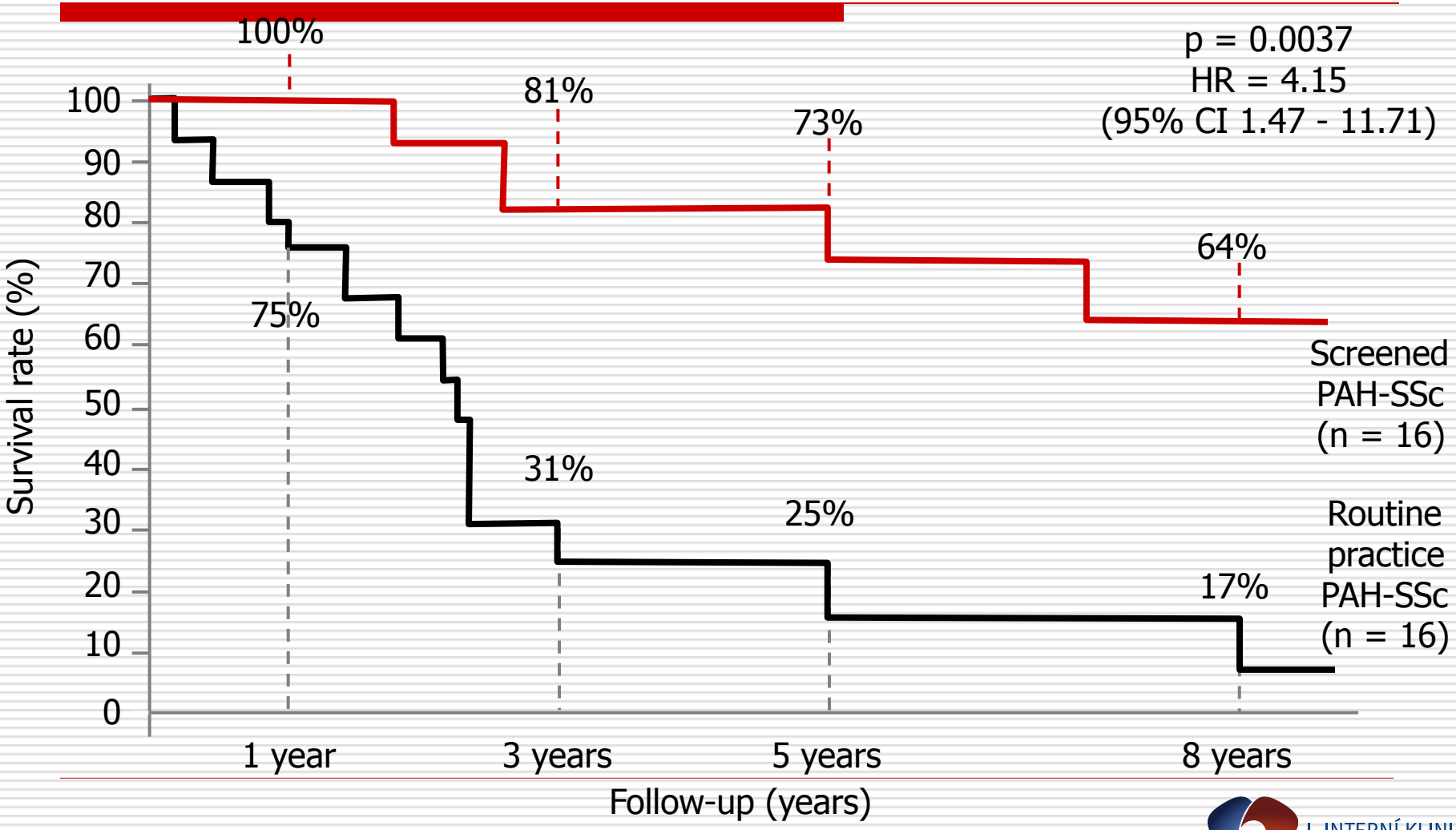
“Man is as old as his arteries”

Plicní hypertenze - prognóza





Screening PH u pacientů se SSc



CI: Confidence interval; HR: Hazard ratio; SSc: Systemic Sclerosis

Humbert M, et al. Arthritis Rheum 2011; 63:3522-30.

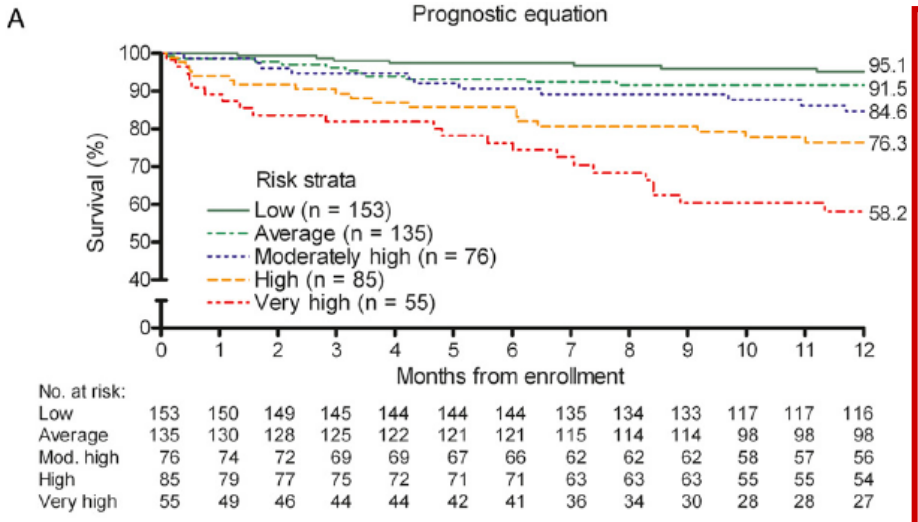
I. INTERNÍ KLINIKA KARDIOLOGIE
FAKULTNÍ NEMOCNICE OLOMOUC

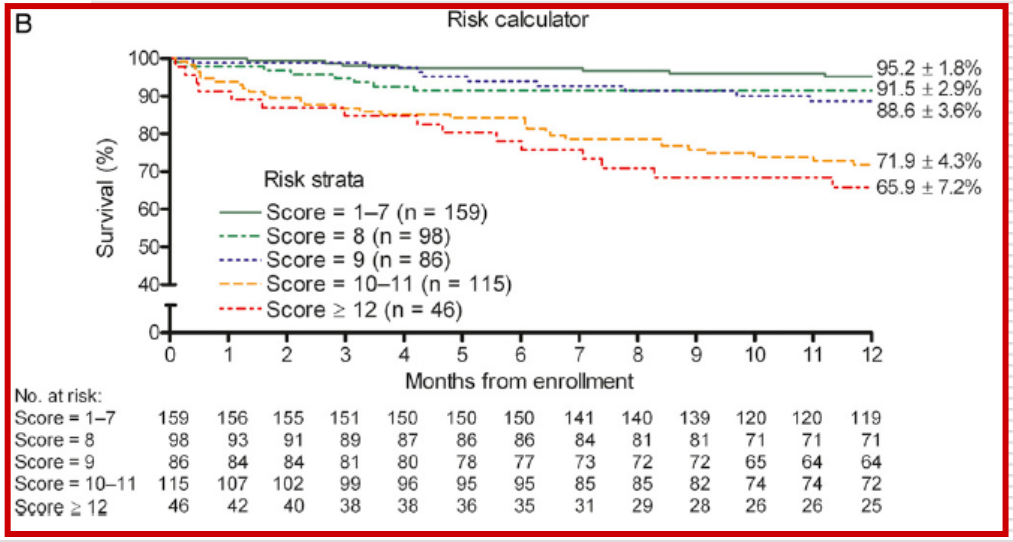
CHEST Original Research

PULMONARY VASCULAR DISEASE

The REVEAL Registry Risk Score Calculator in Patients Newly Diagnosed With Pulmonary Arterial Hypertension

Raymond L. Benza, MD; Mardi Gomberg-Maitland, MD, FCCP; Dave P. Miller, MS; Adaani Frost, MD, FCCP; Robert P. Frantz, MD; Aimee J. Foreman, MA; David B. Badesch, MD, FCCP; and Michael D. McGoon, MD, FCCP





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WHO Group I Subgroup	APAH-CTD +1	APAH-PoPH +2	FPAH +2	
Demographics & Comorbidities	Renal insufficiency +1	Male age >60 yrs +2		
NYHA/WHO Functional Class	I -2	III +1	IV +2	
Vital Signs	SBP <110 mm Hg +1	HR >92 BPM +1		
6-Minute Walk Test	≥440 m -1	<165 m +1		
BNP	<50 pg/mL -2	>180 pg/mL +1		
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United Therapeutics EUROPE LIMITED

EU/REM/JUL12/248(1). Date of preparation: July 2013.

15 negativních prediktorů

4 protektivní faktory

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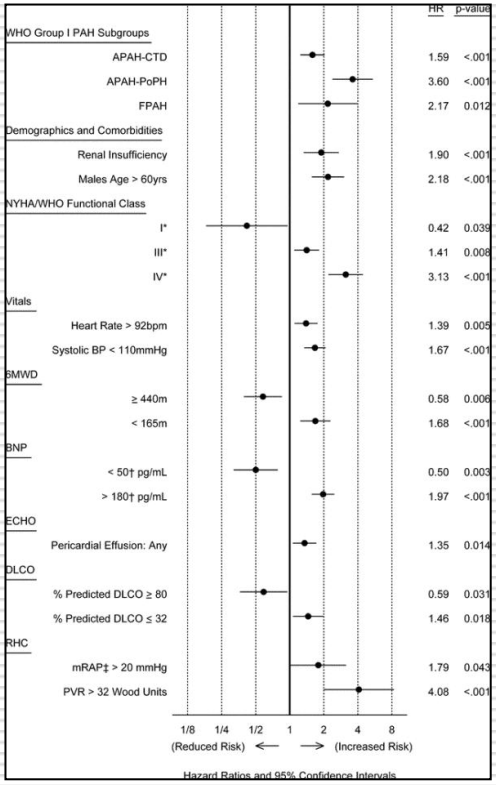

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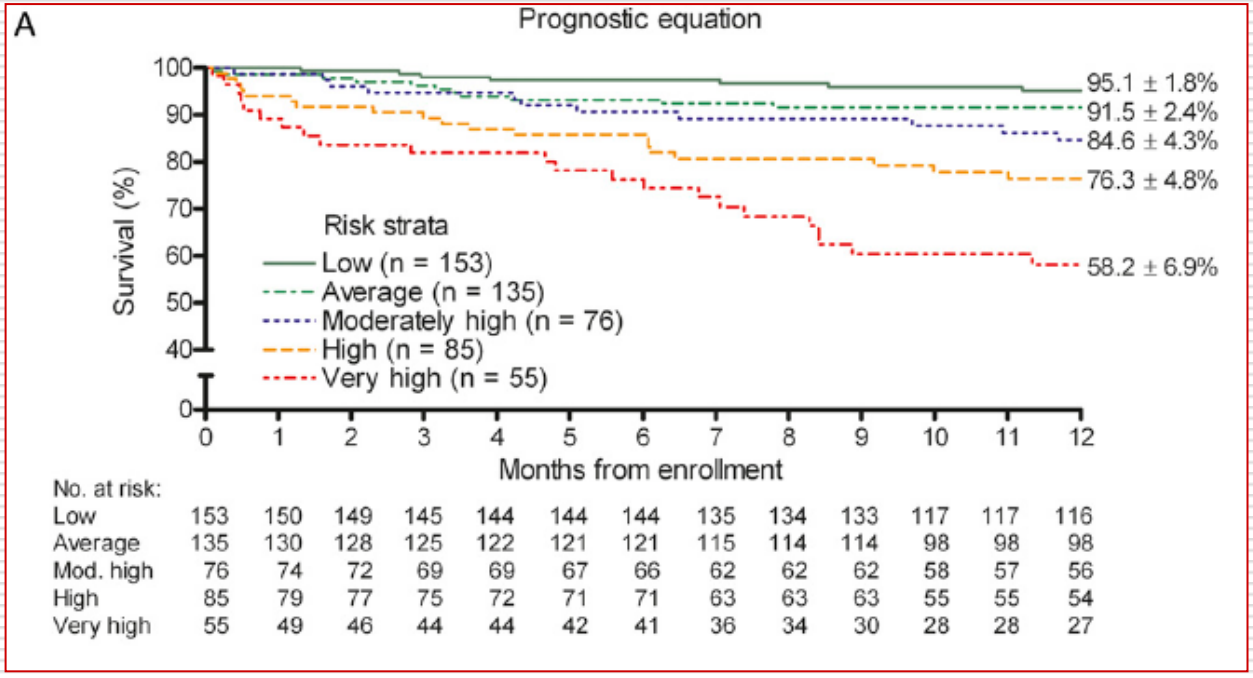
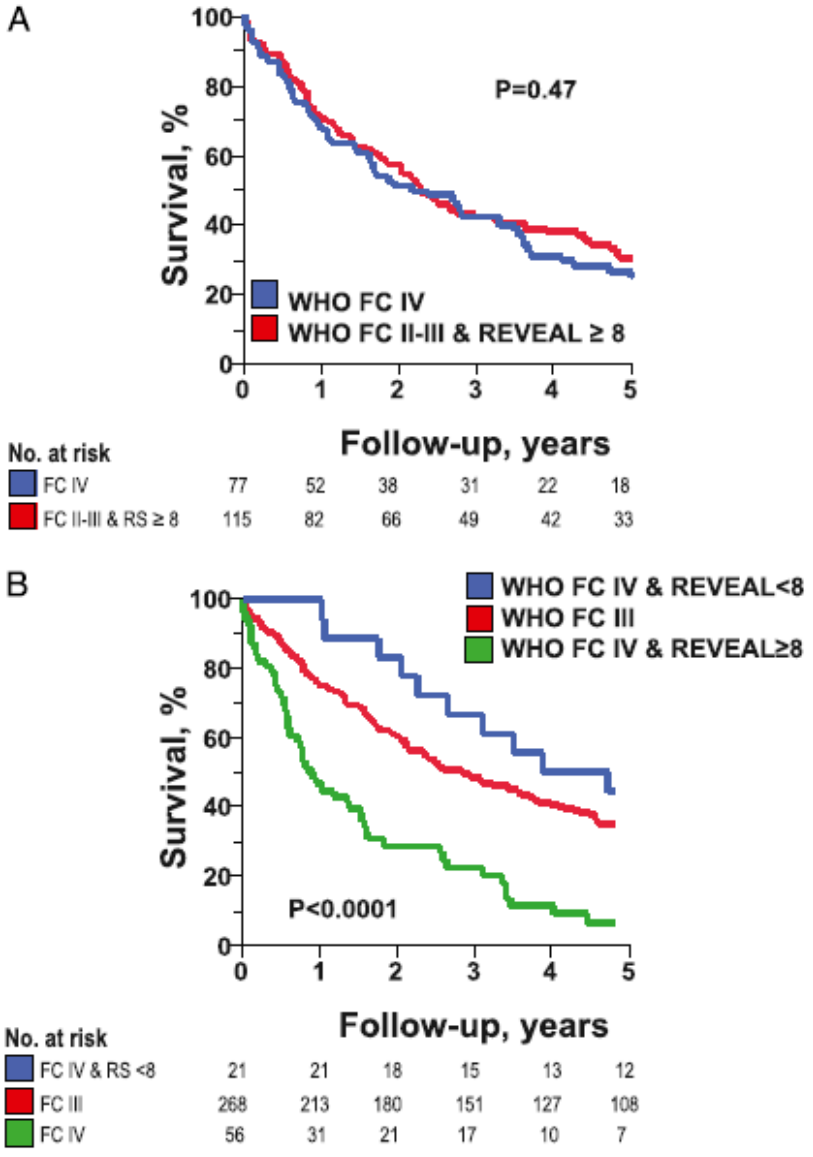
1-leté přežívání v rámci skupin rizikové stratifikace pacientů v registru REVEAL



Riziko přežívání	1-leté
Nízké	> 95%
Střední	90-95%
Vyšší	85-90%
Vysoké	70-85%
Velmi vysoké	<70%

Benza RL; Miller DP; Gomberg-Maitland M; Frantz RP; et al. Predicting survival in pulmonary arterial hypertension: insights from the Registry to Evaluate Early and Long-Term Pulmonary Arterial Hypertension Disease Management (REVEAL). Circulation. 122(2):164-72, 2010 Jul 13.

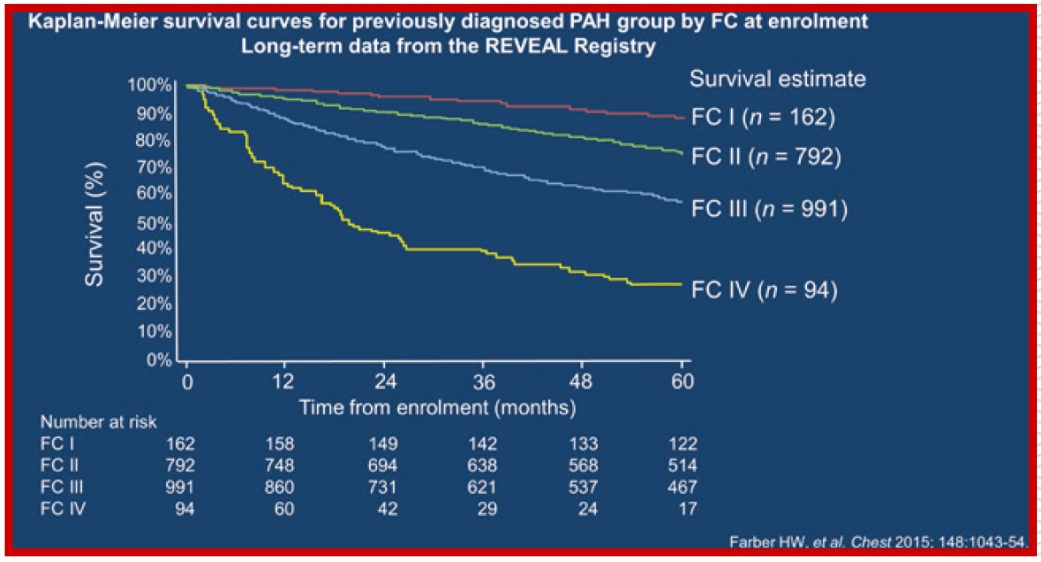


CHEST Original Research

PULMONARY VASCULAR DISEASE

Integration of Clinical and Hemodynamic Parameters in the Prediction of Long-term Survival in Patients With Pulmonary Arterial Hypertension

Garvan C. Kane, MD, PhD, FCCP; Hilal Maradit-Kremers, MD; Josh P. Slusser, BS; Chris G. Scott, MS; Robert P. Frantz, MD; and Michael D. McGoon, MD, FCCP



European Heart Journal Advance Access published September 15, 2015
European Heart Journal
doi:10.1093/eurheartj/ehv317
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)

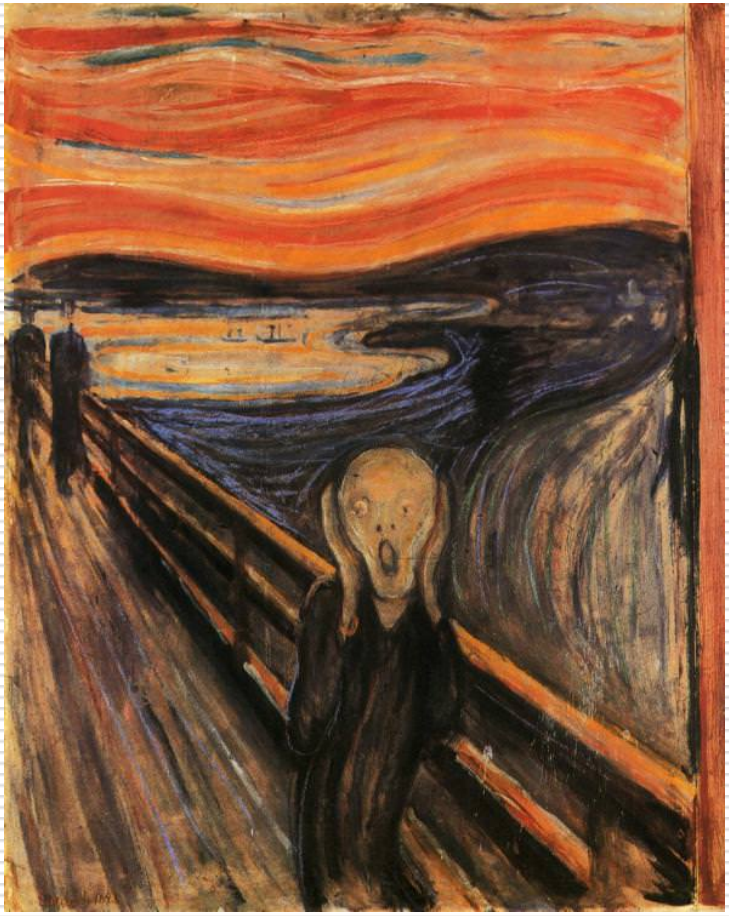
Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

- V klinických studiích a registrech bylo identifikováno množství prognostických faktorů.
- Faktory predikující přežití u pacientů s PAH jsou důležité pro jejich klinický management.
- Jsou založeny na hodnocení demografických, funkčních, laboratorních a hemodynamických parametrů.

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 ml/min/kg (35–65% pred.) VE/VCO_2 slope 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 slope ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60%

Galiè N, et al. Eur Heart J 2016; 37:67-119.

DIAGNÓZA



Screening plicní hypertenze

European Heart Journal Advance Access published September 15, 2015
European Heart Journal
doi:10.1093/eurheartj/ehv317
ESC/ERS GUIDELINES

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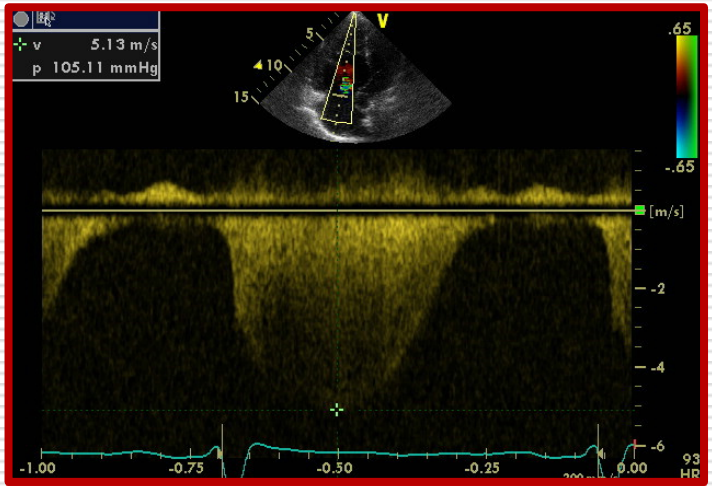
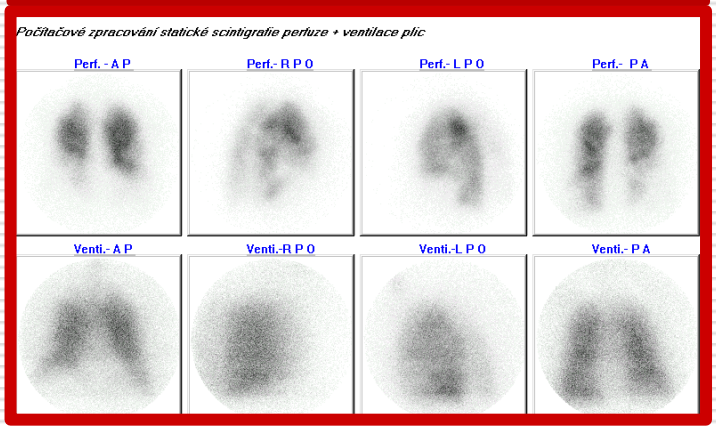
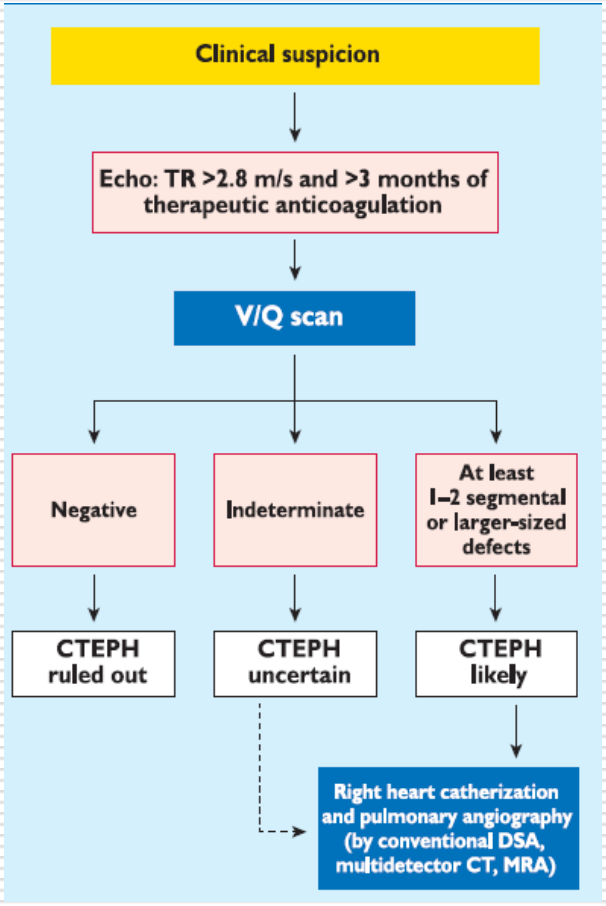
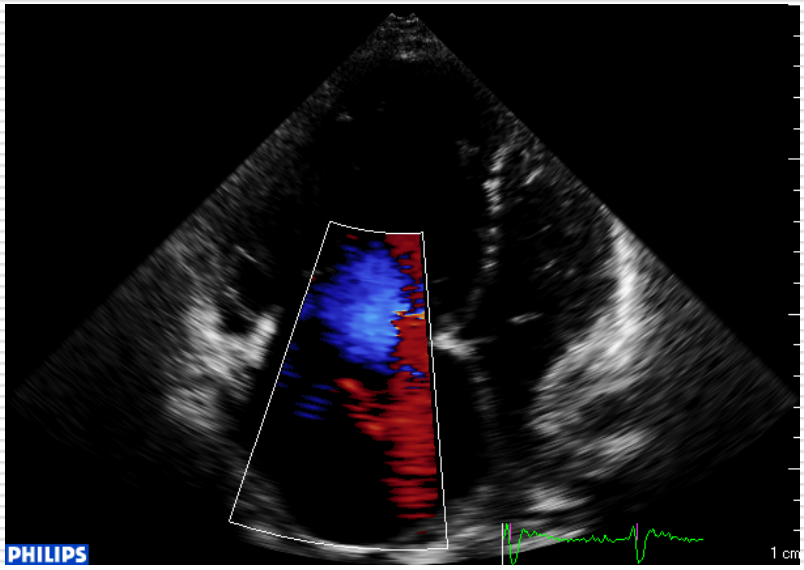


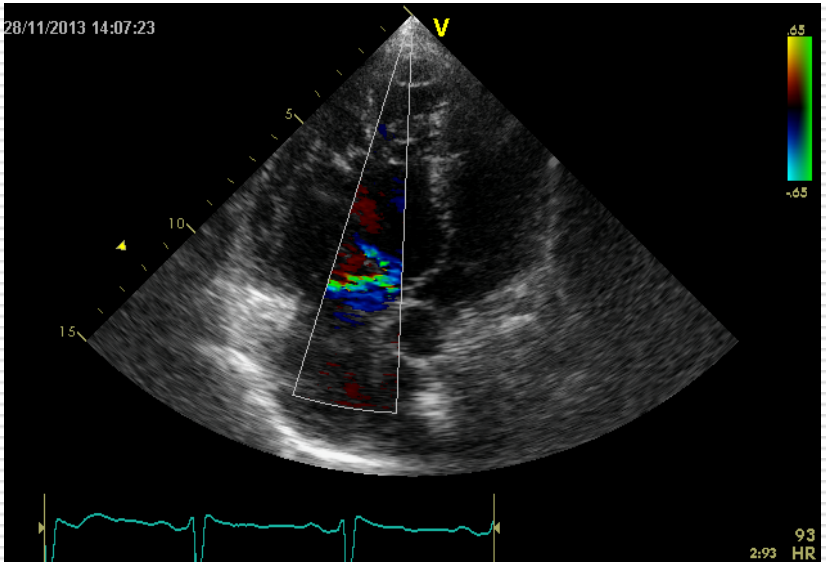


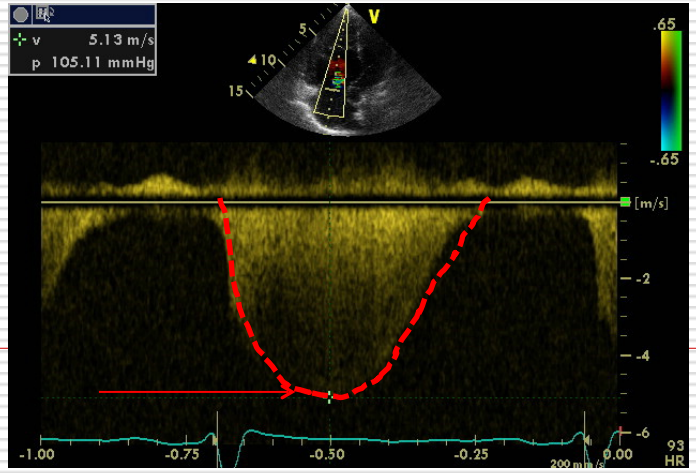


I. INTERNÍ KLINIKA
KARDIOLOGIE
FAKULTNÍ NEMOCNICE OLOMOUC

Echokardiografie - klíčové screeningové vyšetření







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KARDIOLOGIE
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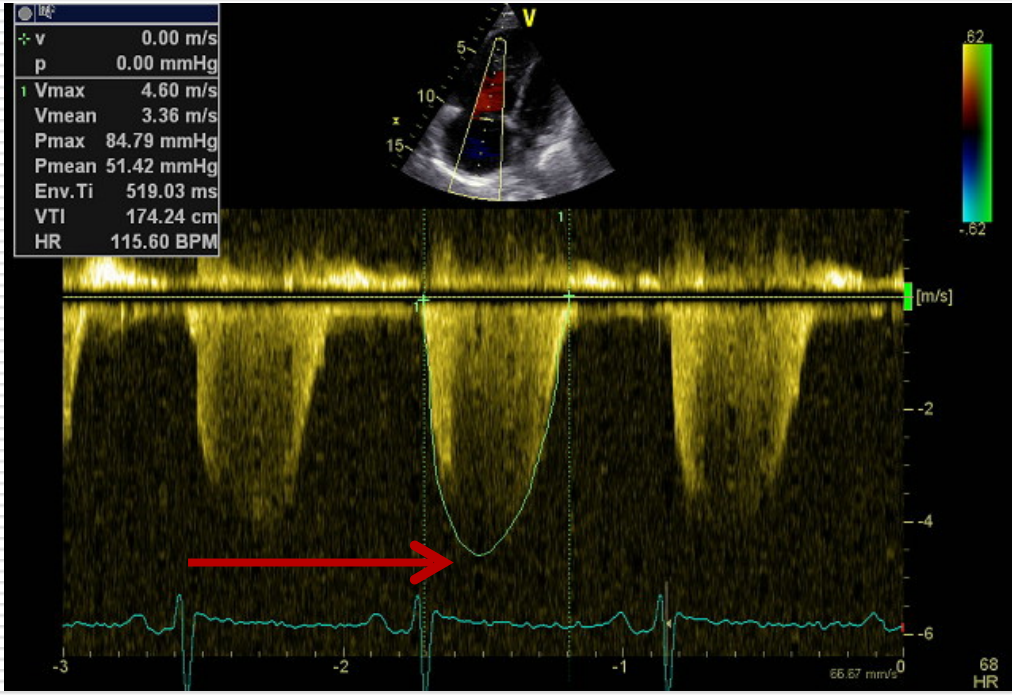
A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm.	



Morfologie a funkce pravé komory

European Heart Journal – Cardiovascular Imaging (2015) **16**, 233–271
doi:10.1093/ehjci/jev014

POSITION PAPER

Recommendations for Cardiac Chamber Quantification by Echocardiography in Adults: An Update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging

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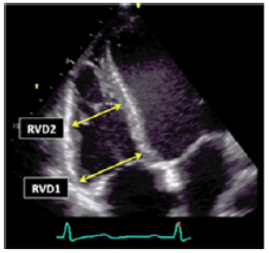
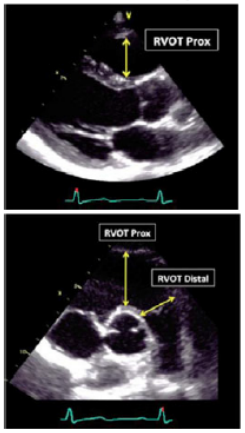
Rozměry DUTINY pravé komory

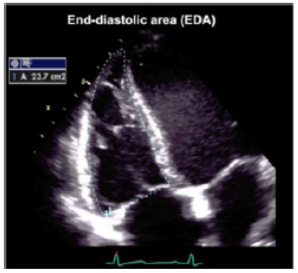
RV linear dimensions (inflow)*



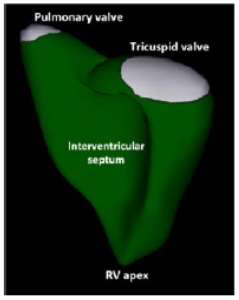
RV linear dimensions (outflow)*



RV areas (inflow)



3DE RV volumes



RV wall thickness

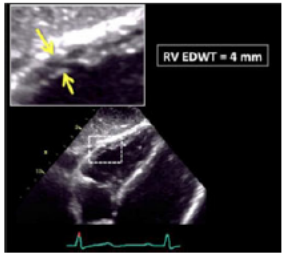
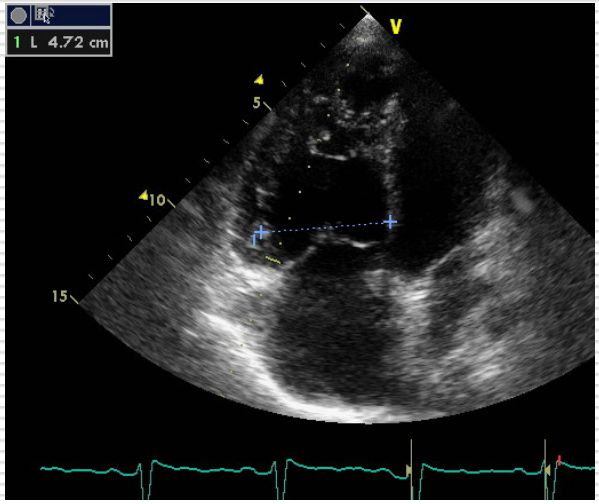


Table 8 Normal values for RV chamber size

Parameter	Mean ± SD	Normal range
RV basal diameter (mm)	33 ± 4	25–41
RV mid diameter (mm)	27 ± 4	19–35
RV longitudinal diameter (mm)	71 ± 6	59–83
RVOT PLAX diameter (mm)	25 ± 2.5	20–30
RVOT proximal diameter (mm)	28 ± 3.5	21–35
RVOT distal diameter (mm)	22 ± 2.5	17–27
RV wall thickness (mm)	3 ± 1	1–5
RVOT EDA (cm^2)		
Men	17 ± 3.5	10–24
Women	14 ± 3	8–20
RV EDA indexed to BSA (cm^2/m^2)		
Men	8.8 ± 1.9	5–12.6
Women	8.0 ± 1.75	4.5–11.5
RV ESA (cm^2)		
Men	9 ± 3	3–15
Women	7 ± 2	3–11
RV ESA indexed to BSA (cm^2/m^2)		
Men	4.7 ± 1.35	2.0–7.4
Women	4.0 ± 1.2	1.6–6.4
RV EDV indexed to BSA (mL/m^2)		
Men	61 ± 13	35–87
Women	53 ± 10.5	32–74
RV ESV indexed to BSA (mL/m^2)		
Men	27 ± 8.5	10–44
Women	22 ± 7	8–36



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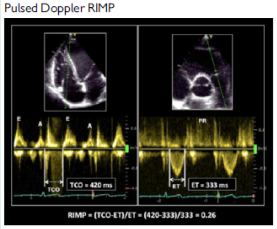
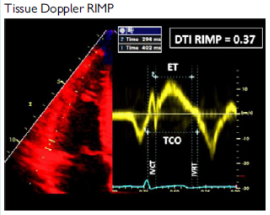
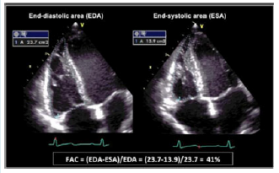
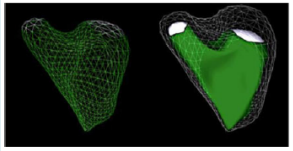
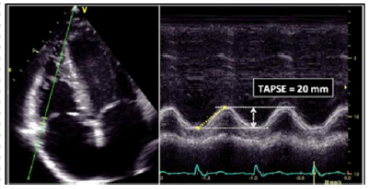
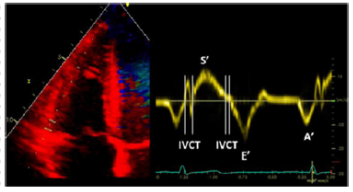
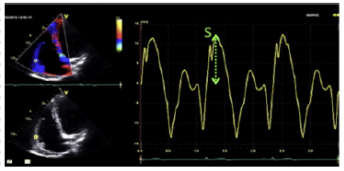
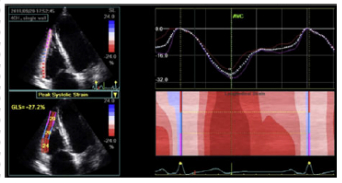
Echocardiographic imaging	Recommended methods	Advantages	Limitations
RV global function Pulsed Doppler RIMP 	RIMP (Tei index) by pulsed Doppler: RIMP = (TCO − ET)/ET	• Prognostic value • Less affected by heart rate	• Requires matching for R-R intervals when measurements are performed on separate recordings • Unreliable when RA pressure is elevated
Tissue Doppler RIMP 	RIMP by tissue Doppler: RIMP = (IVRT + IVCT)/ET = (TCO − ET)/ET	• Less affected by heart rate • Single-beat recording with no need for R-R interval matching	• Unreliable when RA pressure is elevated
RV global systolic function FAC 	RV FAC in RV-focused apical four-chamber view: RV FAC (%) = 100 × (EDA − ESA)/EDA	• Established prognostic value • Reflects both longitudinal and radial components of RV contraction • Correlates with RV EF by CMR	• Neglects the contribution of RV outflow tract to overall systolic function • Only fair inter-observer reproducibility
EF 	Fractional RV volume change by 3D TTE: RV EF (%) = 100 × (EDV − ESV)/EDV	• Includes RV outflow tract contribution to overall function • Correlates with RV EF by CMR	• Dependent on adequate image quality • Load dependency • Requires offline analysis and experience • Prognostic value not established

Echocardiographic imaging	Recommended methods	Advantages	Limitations
RV longitudinal systolic function TAPSE 	• Tricuspid annular longitudinal excursion by M-mode (mm), measured between end-diastole and peak systole • Proper alignment of M-mode cursor with the direction of RV longitudinal excursion should be achieved from the apical approach.	• Established prognostic value • Validated against radionuclide EF	• Angle dependency • Partially representative of RV global function*
Pulsed tissue Doppler S wave 	• Peak systolic velocity of tricuspid annulus by pulsed-wave DTI (cm/sec), obtained from the apical approach, in the view that achieves parallel alignment of Doppler beam with RV free wall longitudinal excursion	• Easy to perform • Reproducible • Validated against radionuclide EF • Established prognostic value	• Angle dependent • Not fully representative of RV global function, particularly after thoracotomy, pulmonary thromboendarterectomy or heart transplantation
Color tissue Doppler S wave 	• Peak systolic velocity of tricuspid annulus by color DTI (cm/sec)	• Sampling is performed after image acquisition • Allows multisite sampling on the same beat	• Angle dependent • Not fully representative of RV global function, particularly after thoracotomy, pulmonary thrombendarterectomy or heart transplantation • Lower absolute values and reference ranges than pulsed DTI S′ wave • Requires offline analysis
GLS 	• Peak value of 2D longitudinal speckle tracking derived strain, averaged over the three segments of the RV free wall in RV-focused apical four-chamber view (%)	• Angle independent • Established prognostic value	• Vendor dependent

Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'[a]	Echocardiographic probability of pulmonary hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
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PLICNÍ HYPERTENZE NEPRAVDĚPODOBNÁ

Rychlost trikuspidální regurgitace ≤ 2.8 m/s

Odhad PASP ≤36 mmHg

Bez přítomnosti hypertrofie, normální morfologie a systolická funkce pravé komory

PLICNÍ HYPERTENZE MOŽNÁ

Rychlost trikuspidální regurgitace ≤ 2.8 m/s

Odhad PASP ≤36 mmHg

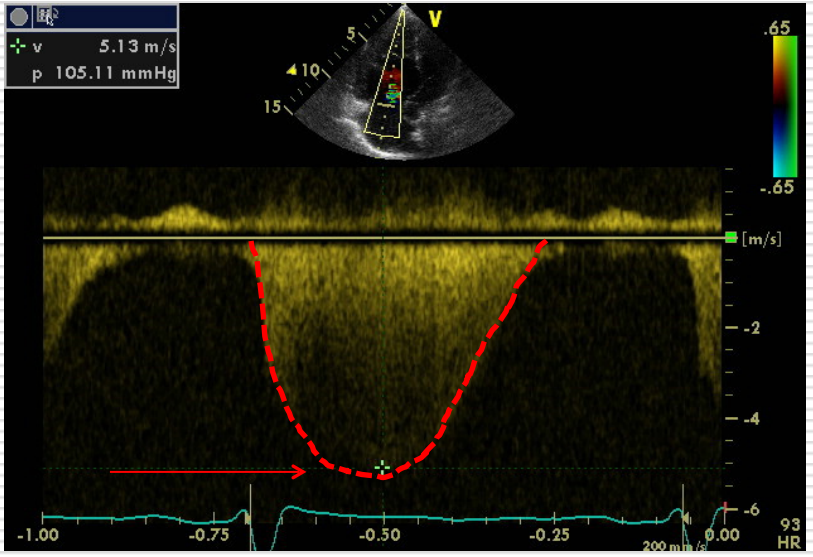
Suspektní hypertrofie, dilatace a/nebo systolická dysfunkce pravé komory

PLICNÍ HYPERTENZE PRAVDĚPODOBNÁ

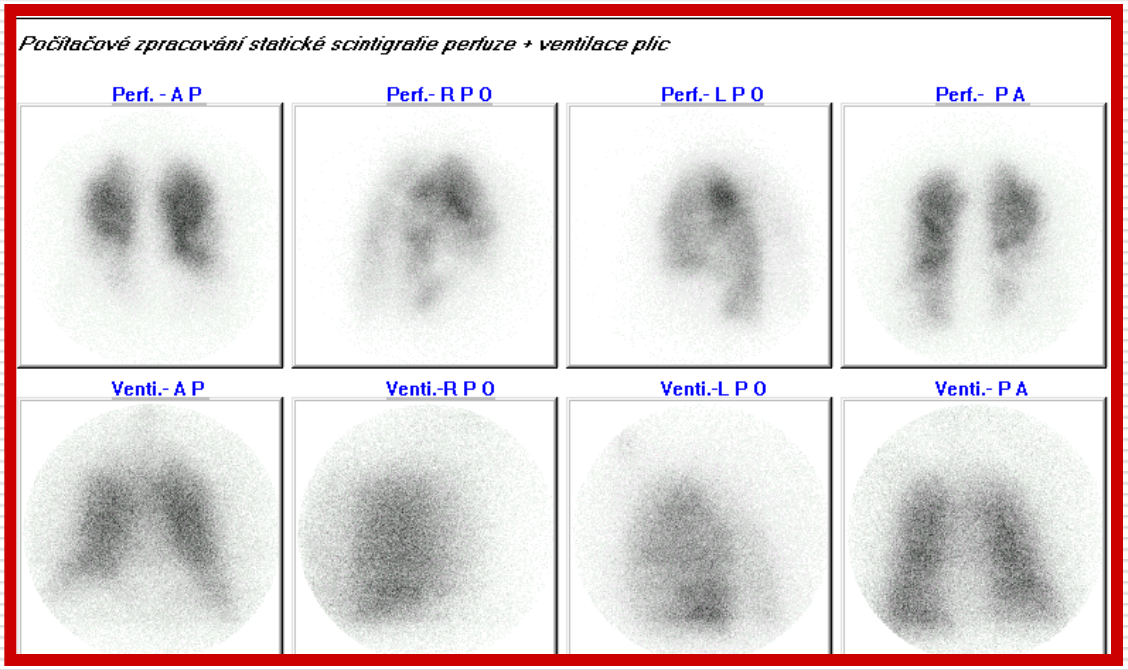
Rychlost trikuspidální regurgitace > 2.8 m/s

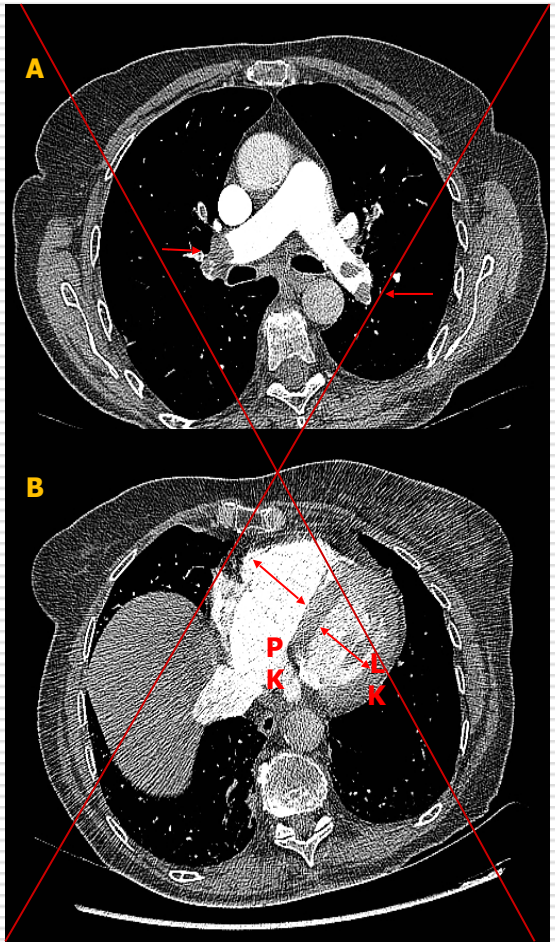
Odhad PASP >36 mmHg

Evidence/absence přítomnosti hypertrofie, dilatace a systolické dysfunkce pravé komory



Scintigrafie plic – screeningová zobrazovací metoda





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]

[1]Department of Radiology, Hammersmith Hospital, London, United Kingdom; [2]Department of Cardiology, Hammersmith Hospital, London, United Kingdom; [3]Imperial College, London, United Kingdom; and [4]Department of Nuclear Medicine, Hammersmith Hospital, London, United Kingdom

J Nucl Med 2007; 48:680–684

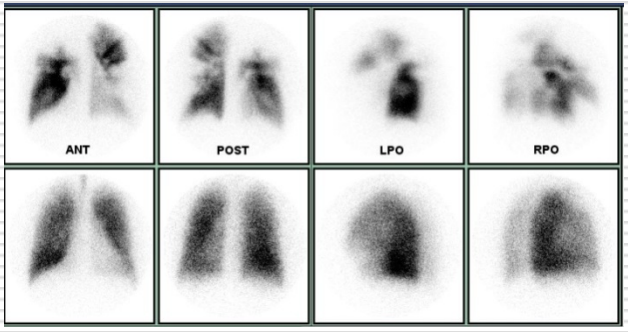
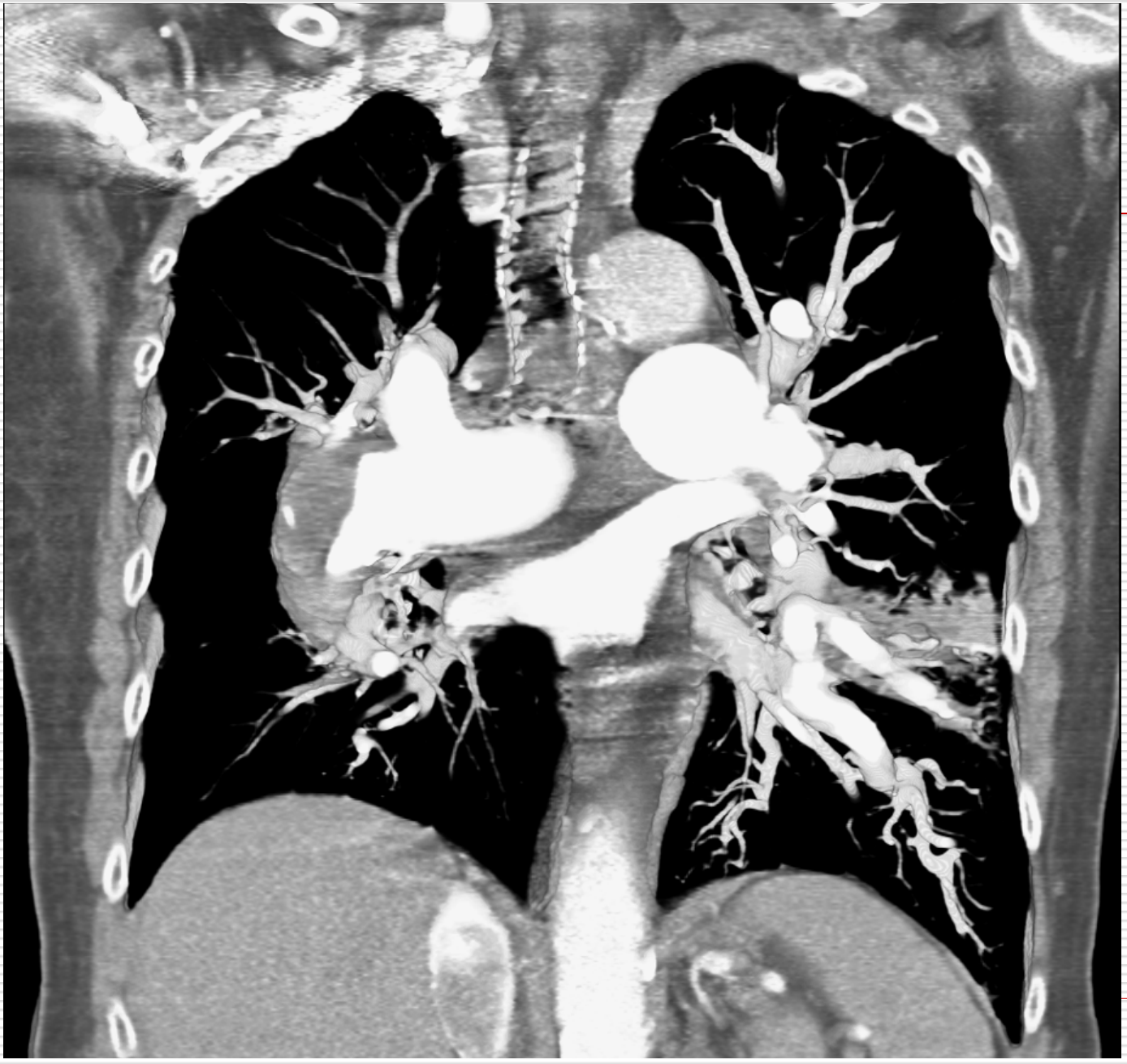


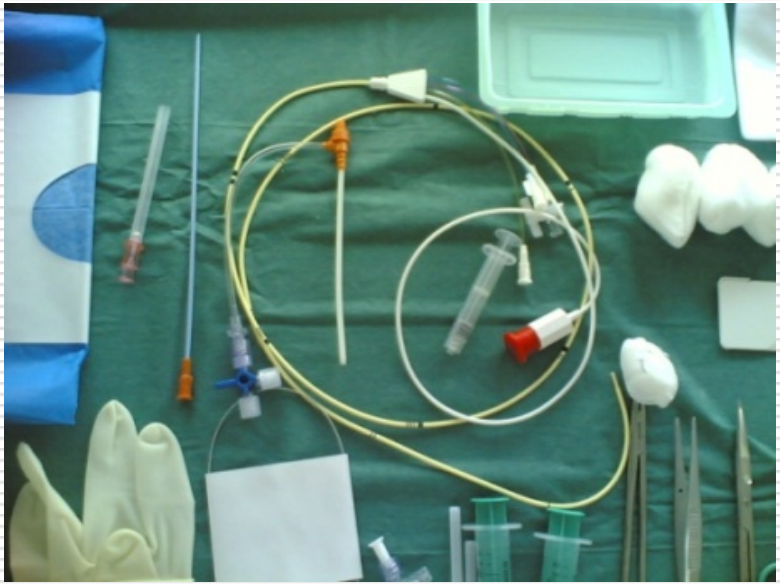
TABLE 1
Summary of V/Q Scans and CTPA Results

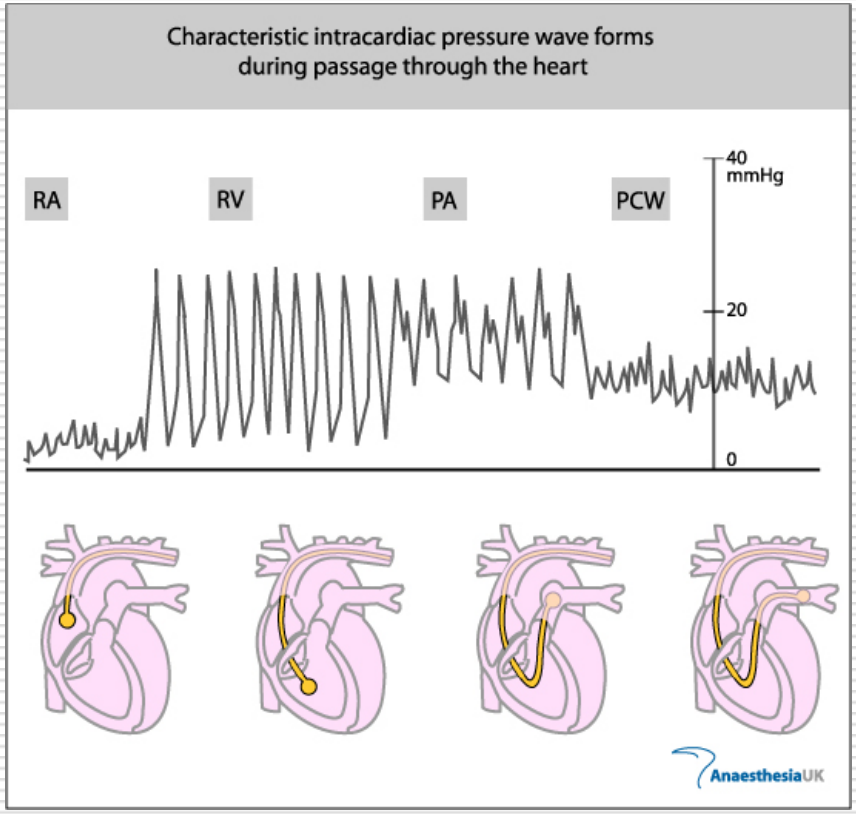
	V/Q			CTPA	
Group	Low probability	Intermediate probability	High probability	Negative	Positive
A (*n* = 78)	2	1	75	38	40
B (*n* = 149)	134	7	8	148	1



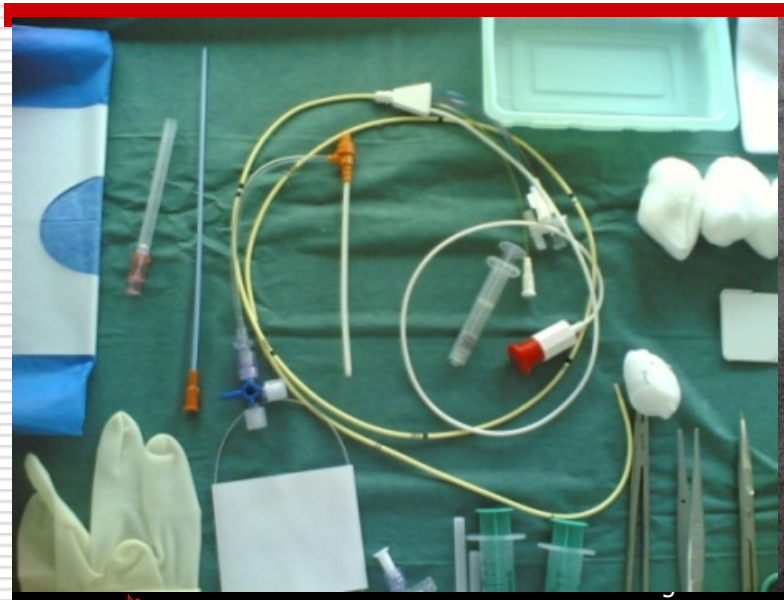
Obrázky poskytnuty laskavostí doc. Jansy

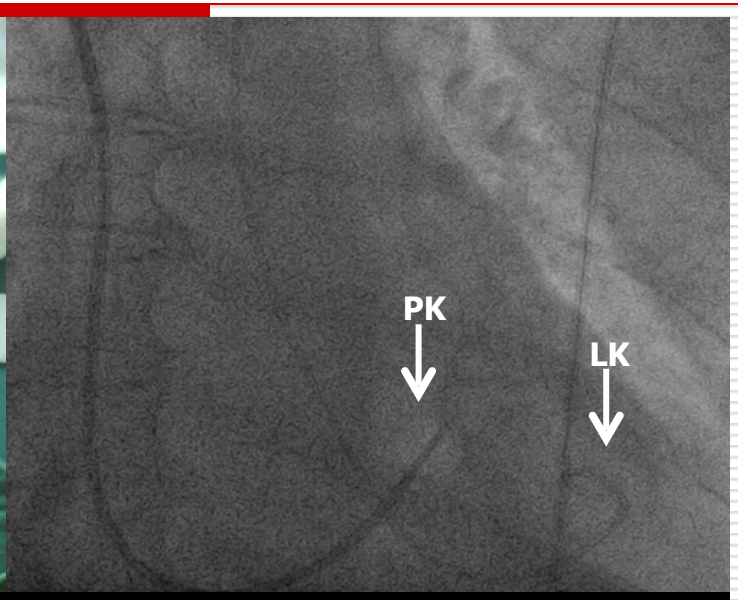
Pravostranná katetrizace - hemodynamika

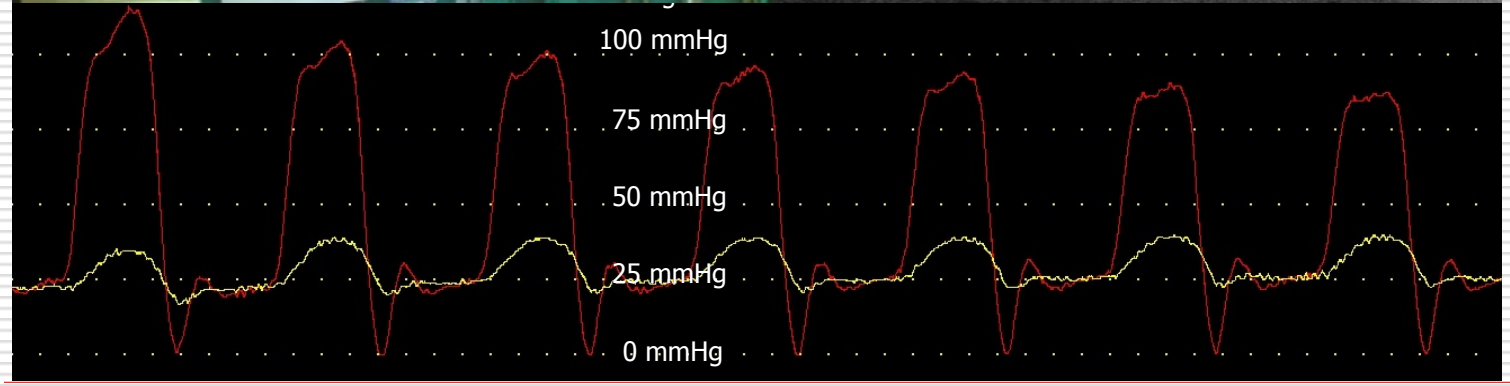




Simultánní pravo/levá katetrizace



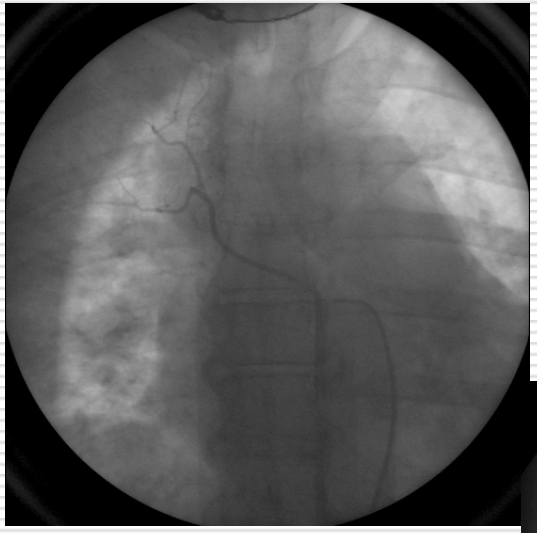


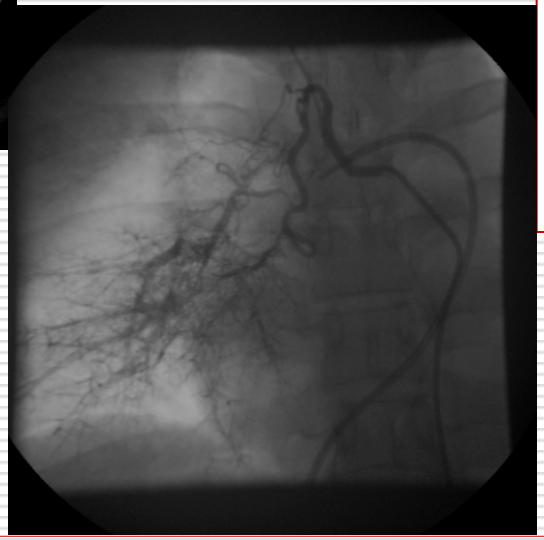


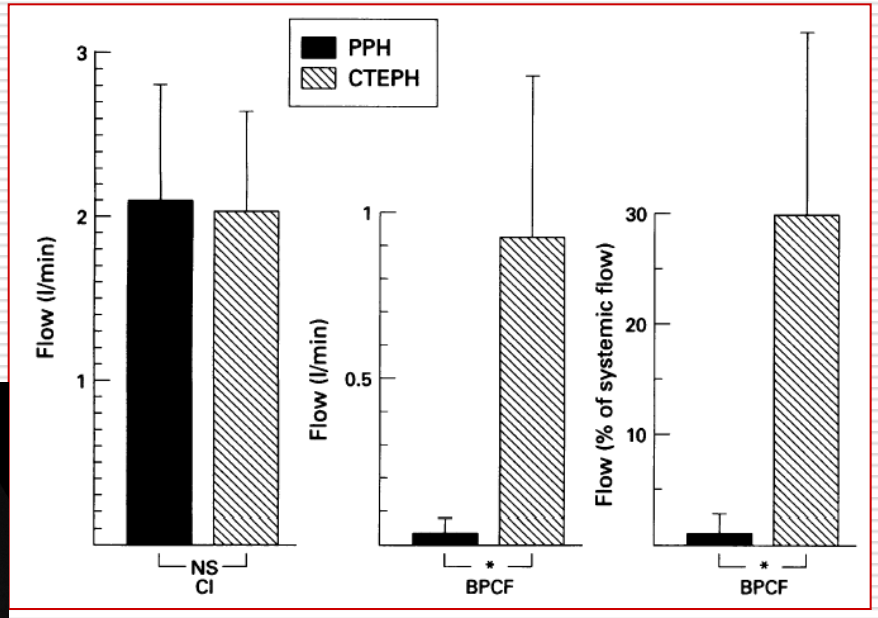
Bronchopulmonální kolaterály

Comparison of bronchopulmonary collaterals and collateral blood flow in patients with chronic thromboembolic and primary pulmonary hypertension

Jiri Endrys, Nasser Hayat, George Cherian







Heart 1997, 78: 171-6

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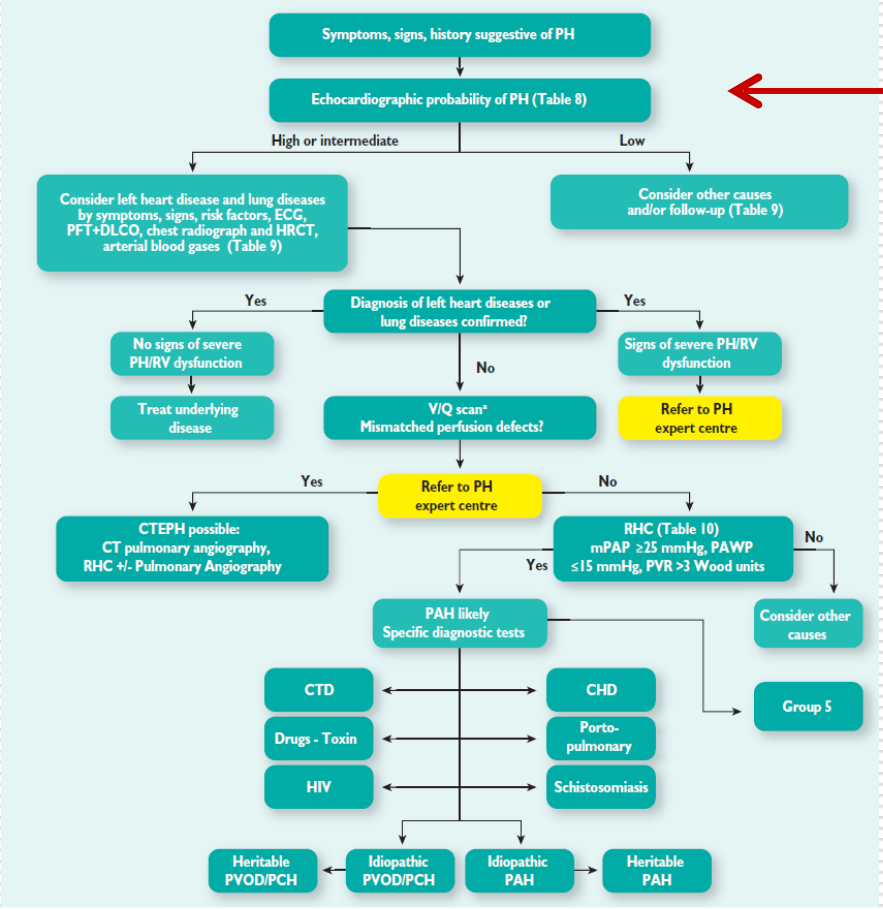
Diagnostický algoritmus



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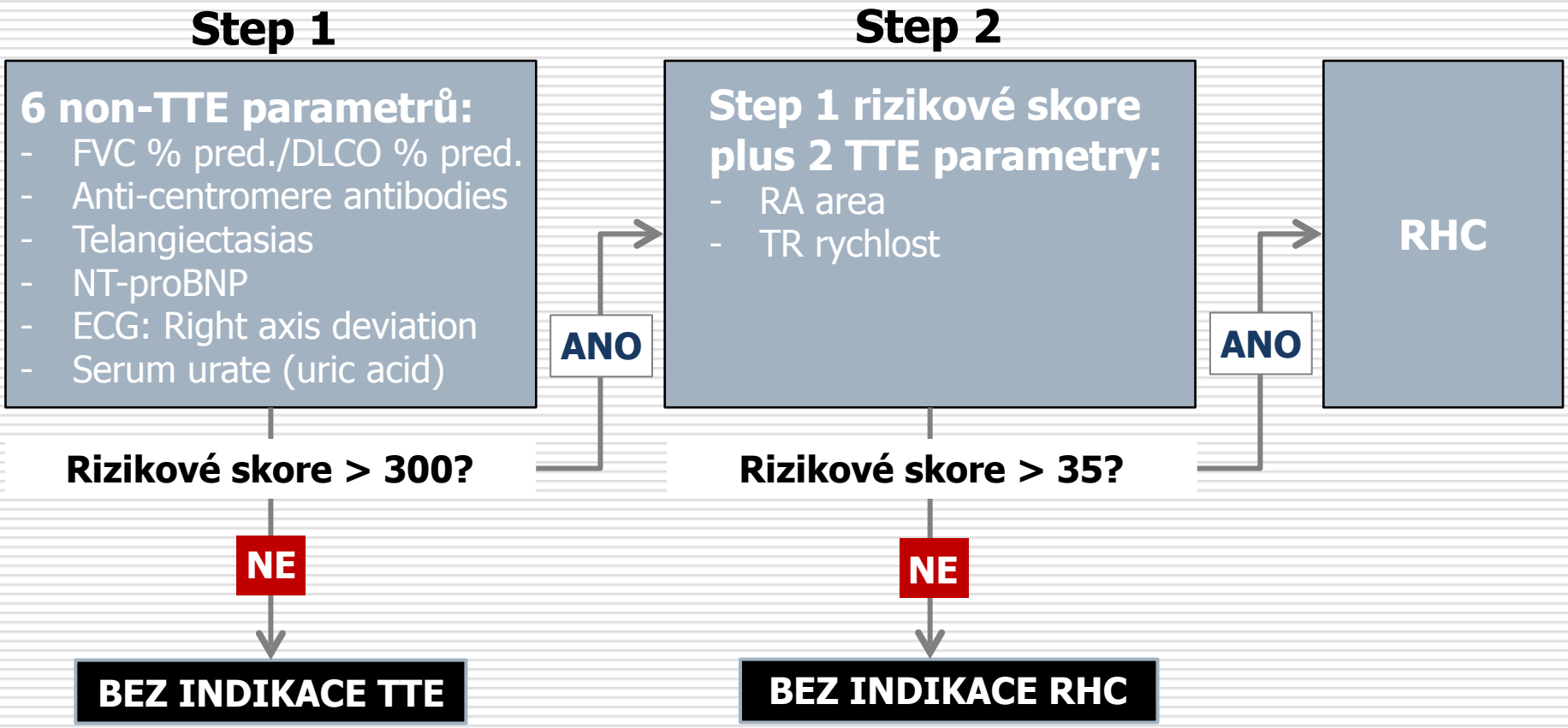
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

Echocardiographic probability of PH	Without risk factors or associated condition for PAH or CTEPH[d]	Class[a]	Level[b]	With risk factors or associated conditions for PAH or CTEPH[c]	Class[a]	Level[b]
Low	Alternative diagnosis should be considered	IIa	C	Echo follow-up should be considered	IIa	C
Intermediate	Alternative diagnosis, echo follow-up, should be considered	IIa	C	Further assessment of PH including RHC should be considered[e]	IIa	B
	Further investigation of PH may be considered[e]	IIb				
High	Further investigation of PH (including RHC[e]) is recommended	I	C	Further investigation of PH[e] including RHC is recommended	I	C

SCREENING

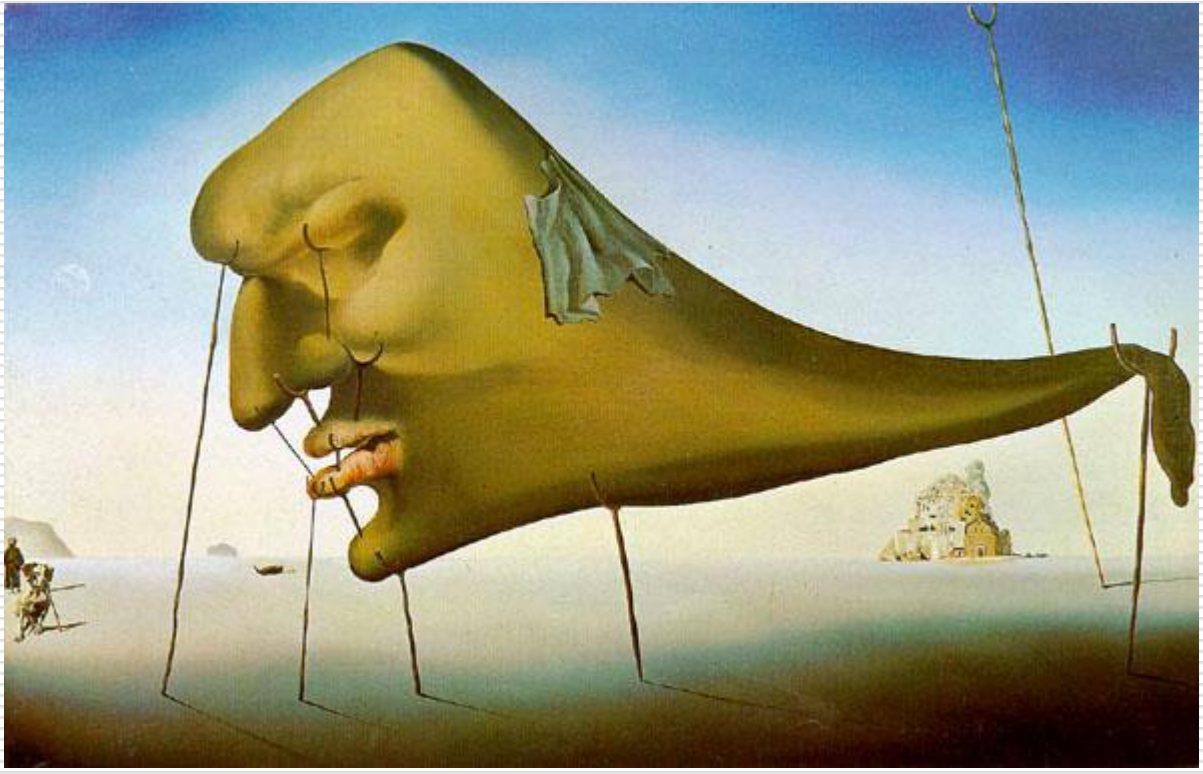
DETECT algoritmus screeningu PH u pacientů se SSc



DLCO: Diffusing capacity of the lungs for carbon monoxide; ECG: Electrocardiogram; FVC: Forced vital capacity; NT-proBNP: N-terminal prohormone brain natriuretic peptide; RHC: Right heart catheterisation; SSc: Systemic Sclerosis; TR: Tricuspid regurgitation

Coghlan JG, et al. Ann Rheum Dis 2014; 73:1340-9.

TERAPIE



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ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

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Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 ml/min/kg (35–65% pred.) VE/VCO_2 slope 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 slope ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60%

Plicní hypertenze - terapeutický algoritmus a cíle

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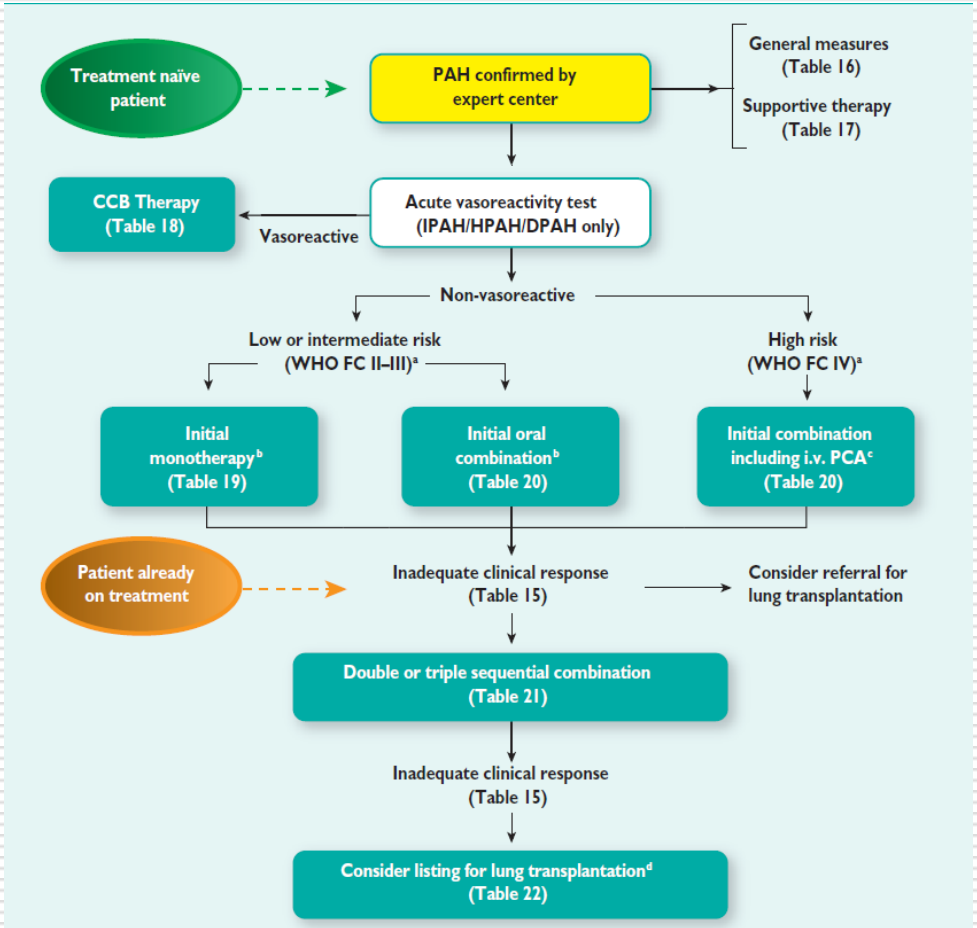

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Treatment Goals of Pulmonary Hypertension

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Functional class	I or II
Echocardiography/CMR	Normal/near-normal RV size and function
Hemodynamics	Normalization of RV function (RAP <8 mm Hg and CI >2.5 to 3.0 l/min/m^2)
6-min walk distance	>380 to 440 m; may not be aggressive enough in young individuals
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg and $EqCO_2$ <45 l/min/l/min
B-type natriuretic peptide level	Normal

CI = cardiac index; CMR = cardiac magnetic resonance; $EqCO_2$ = ventilatory equivalent for carbon dioxide; PAH = pulmonary arterial hypertension; RAP = right atrial pressure; RV = right ventricular; VO_2 = peak oxygen consumption.

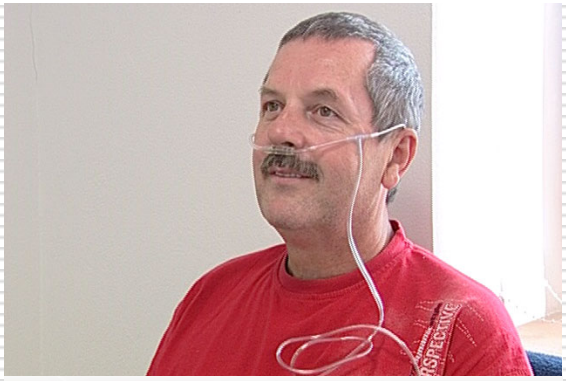
A. Konvenční a podpůrná terapie

- Blokátory kalciových kanálů

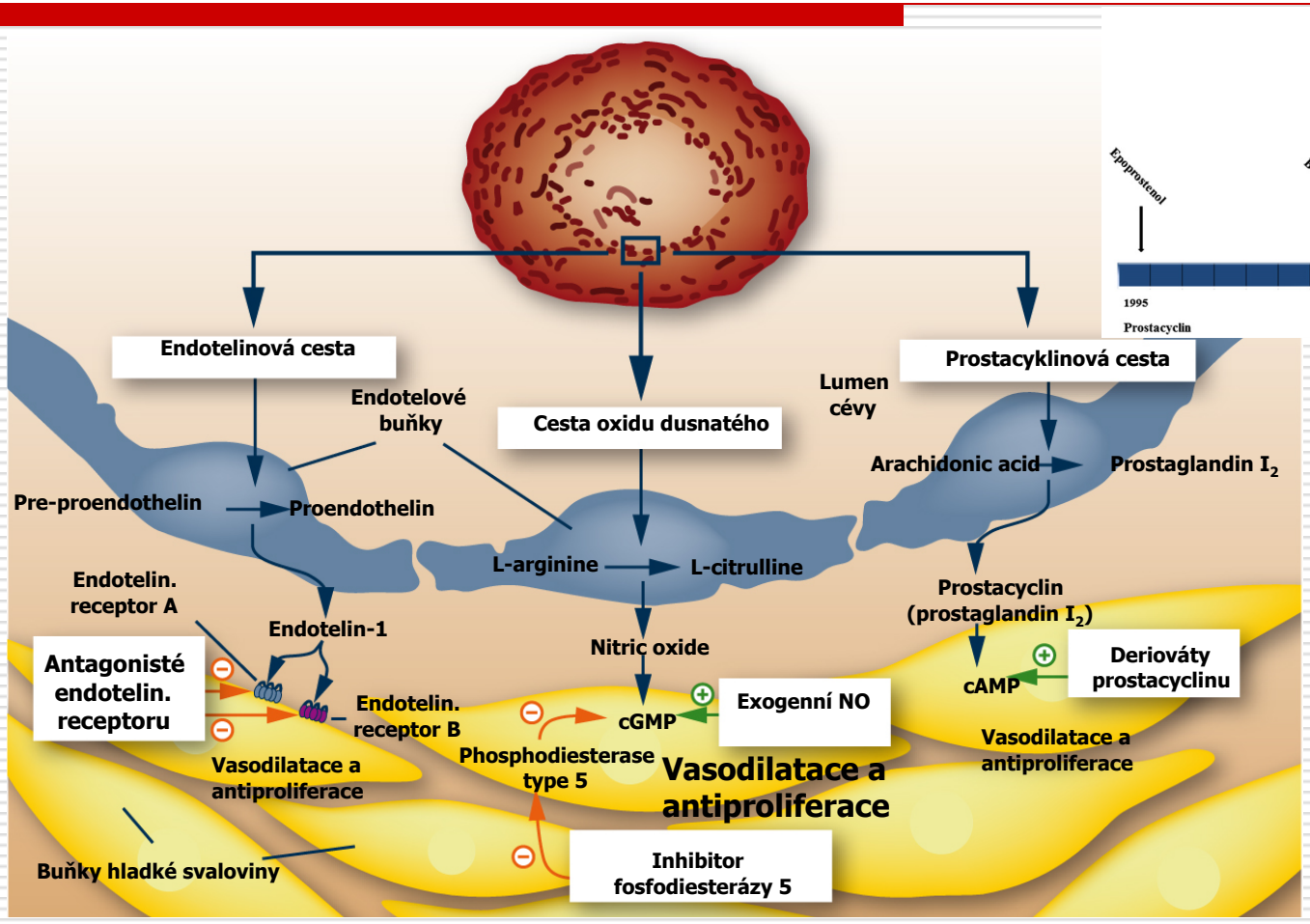
- Diuretika
- Antikoagulační terapie

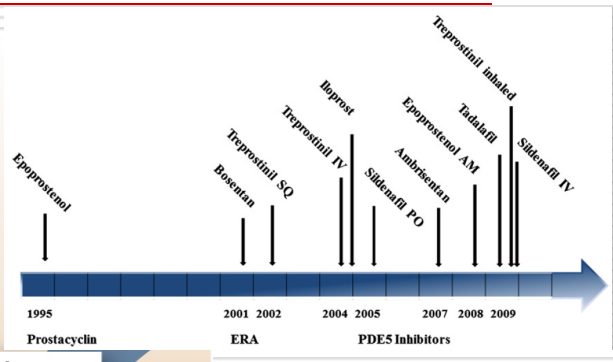
- Rehabilitace
- CAVE: těhotenství

- DDOT
- Terapie syndromu spánkové apnoe - CPAP



B. Specifická farmakoterapie – cíle





Humbert; NEJM (2004)

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Monoterapie

Measure/treatment			Class[a]-Level[b]					
			WHO-FC II		WHO-FC III		WHO-FC IV	
Calcium channel blockers			I	C[d]	I	C[d]	-	-
Endothelin receptor antagonists	Ambrisentan		I	A	I	A	IIb	C
	Bosentan		I	A	I	A	IIb	C
	Macitentan[e]		I	B	I	B	IIb	C
Phosphodiesterase type 5 inhibitors	Sildenafil		I	A	I	A	IIb	C
	Tadalafil		I	B	I	B	IIb	C
	Vardenafil[g]		IIb	B	IIb	B	IIb	C
Guanylate cyclase stimulators	Riociguat		I	B	I	B	IIb	C
Prostacyclin analogues	Epoprostenol	Intravenous[e]	-	-	I	A	I	A
	Iloprost	Inhaled	-	-	I	B	IIb	C
		Intravenous[g]	-	-	IIa	C	IIb	C
	Treprostinil	Subcutaneous	-	-	I	B	IIb	C
		Inhaled[g]	-	-	I	B	IIb	C
		Intravenous[f]	-	-	IIa	C	IIb	C
		Oral[g]	-	-	IIb	B	-	-
	Beraprost[g]		-	-	IIb	B	-	-
IP receptor agonists	Selexipag (oral)[g]		I	B	I	B	-	-

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Kombinační léčba

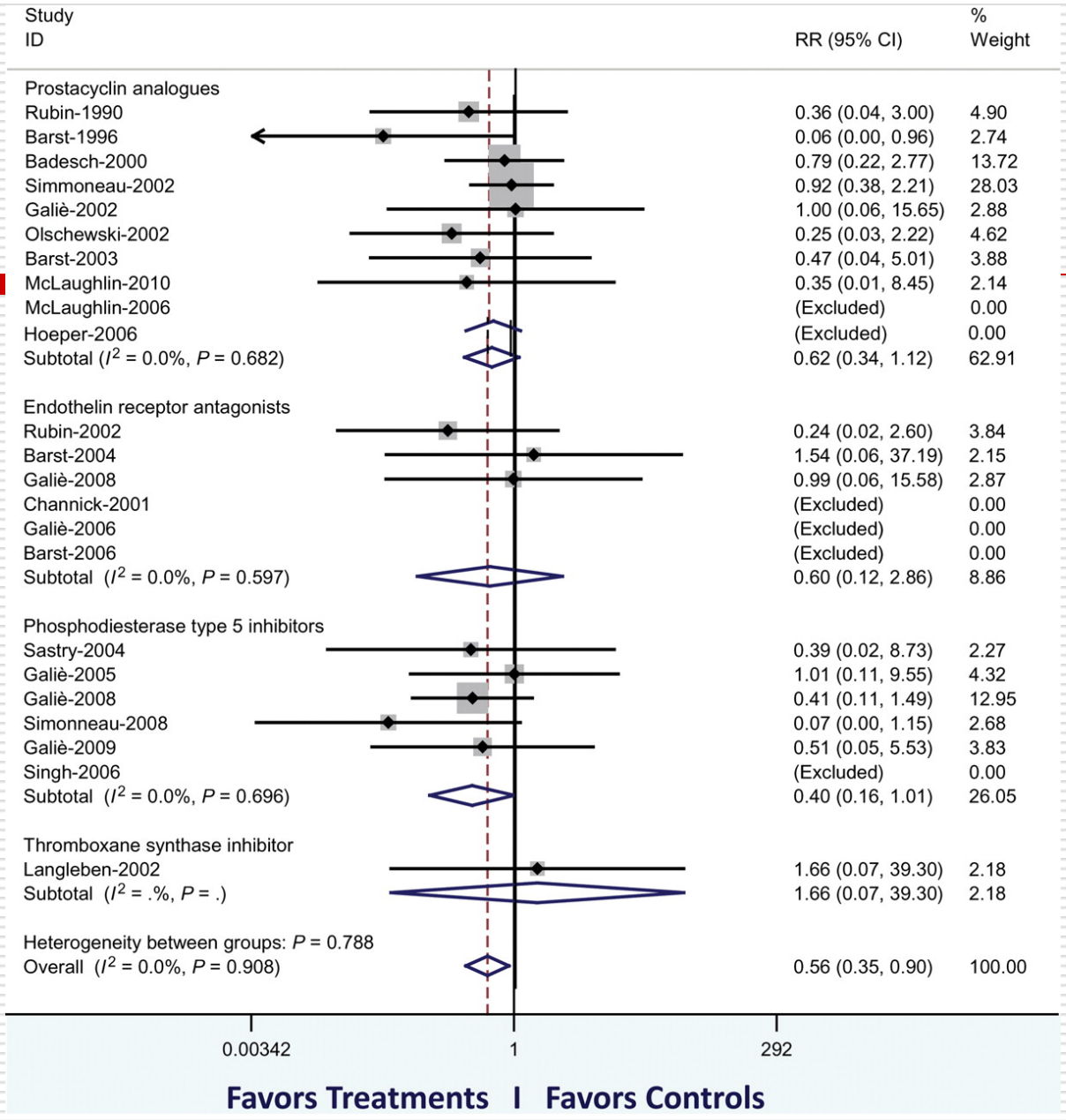
Measure/ treatment	Class[a]-Level[b]					
	WHO-FC II		WHO-FC III		WHO-FC IV	
Ambrisentan + tadalafil[d]	I	B	I	B	IIb	C
Other ERA + PDE-5i	IIa	C	IIa	C	IIb	C
Bosentan + sildenafil + i.v. epoprostenol	-	-	IIa	C	IIa	C
Bosentan + i.v. epoprostenol	-	-	IIa	C	IIa	C
Other ERA or PDE-5i + s.c. treprostinil			IIb	C	IIb	C
Other ERA or PDE-5i + other i.v. prostacyclin analogues			IIb	C	IIb	C

Measure/ treatment	Class[a]-Level[b]					
	WHO-FC II		WHO-FC III		WHO-FC IV	
Macitentan added to sildenafil[d]	I	B	I	B	IIa	C
Riociguat added to bosentan	I	B	I	B	IIa	C
Selexipag[e] added to ERA and/or PDE-5i[d]	I	B	I	B	IIa	C
Sildenafil added to epoprostenol	–	–	I	B	IIa	B
Treprostinil inhaled added to sildenafil or bosentan	IIa	B	IIa	B	IIa	C
Iloprost inhaled added to bosentan	IIb	B	IIb	B	IIb	C

Iniciální

Sekvenční

Tadalafil added to bosentan	IIa	C	IIa	C	IIa	C
Ambrisentan added to sildenafil	IIb	C	IIb	C	IIb	C
Bosentan added to epoprostenol	–	–	IIb	C	IIb	C
Bosentan added to sildenafil	IIb	C	IIb	C	IIb	C
Sildenafil added to bosentan	IIb	C	IIb	C	IIb	C
Other double combinations	IIb	C	IIb	C	IIb	C
Other triple combinations	IIb	C	IIb	C	IIb	C
Riociguat added to sildenafil or other PDE-5i	III	B	III	B	III	B



Galiè N et al. Eur Heart J 2010;31:2080-2086

Analoga prostacyklinu

Substance a aktivitou podobnou jako prostacyklin PGI_2

Vazodilatační efekt a antikoagulační působení

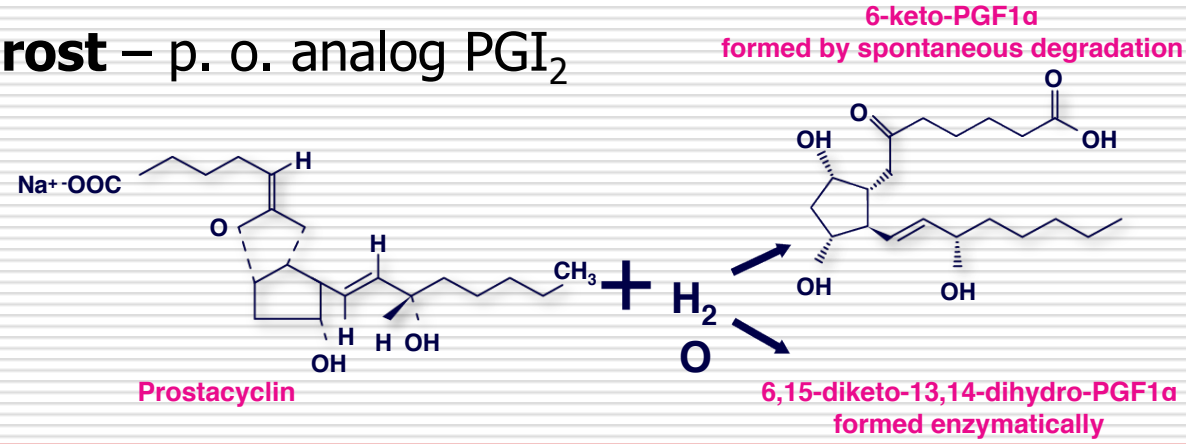
Epoprostenol (Flolan) – termolabilní, aplikace kontinuálně i.v. pumpou

Epoprostenol (Veletri) – termostabilní

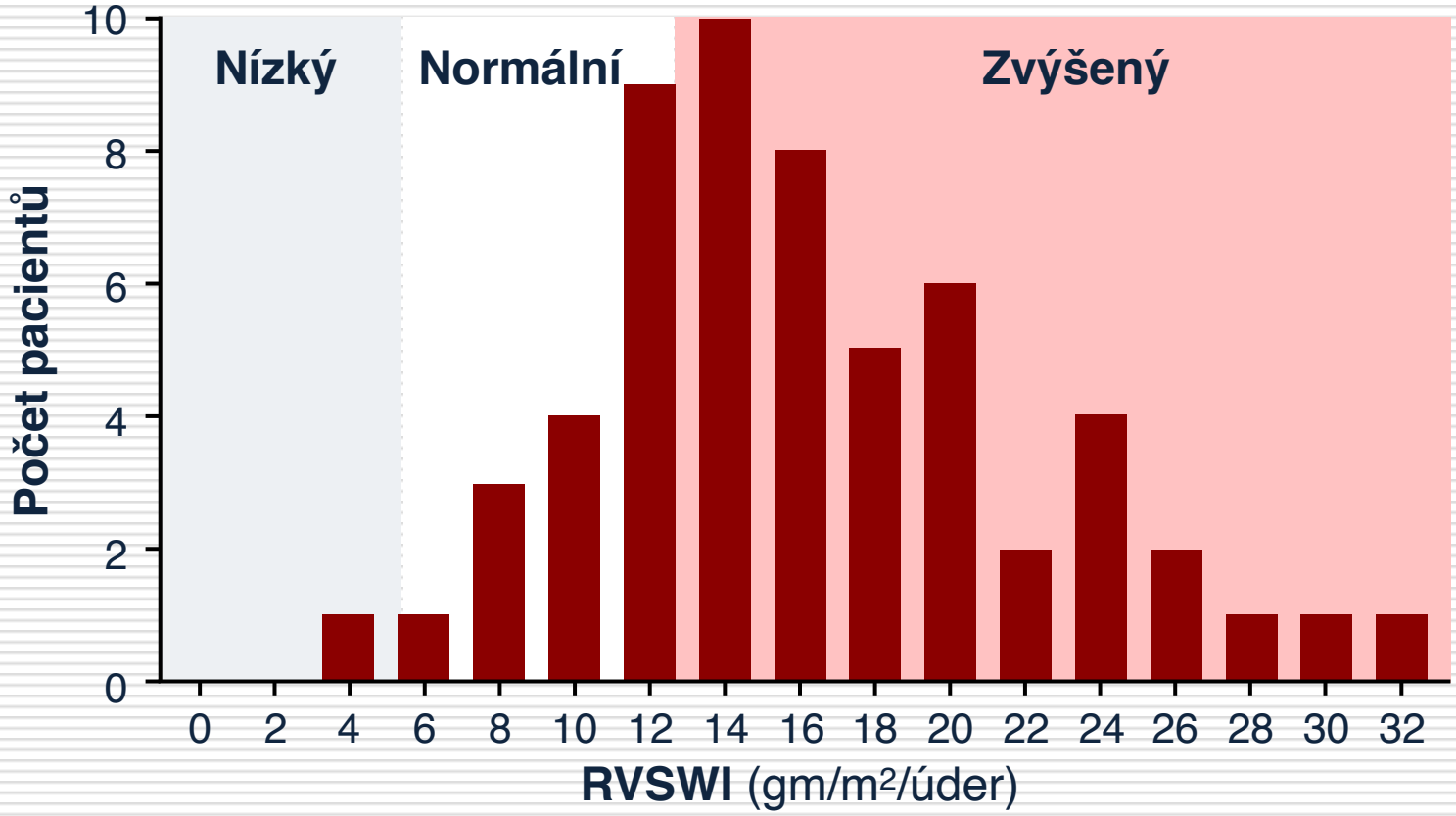
Ilopost (Ventavis) – inhalační analog PGI_2

Treprostinil (Remodulin) – s. c. analog PGI_2

Beraprost – p. o. analog PGI_2



Funkce pravé komory - PAH

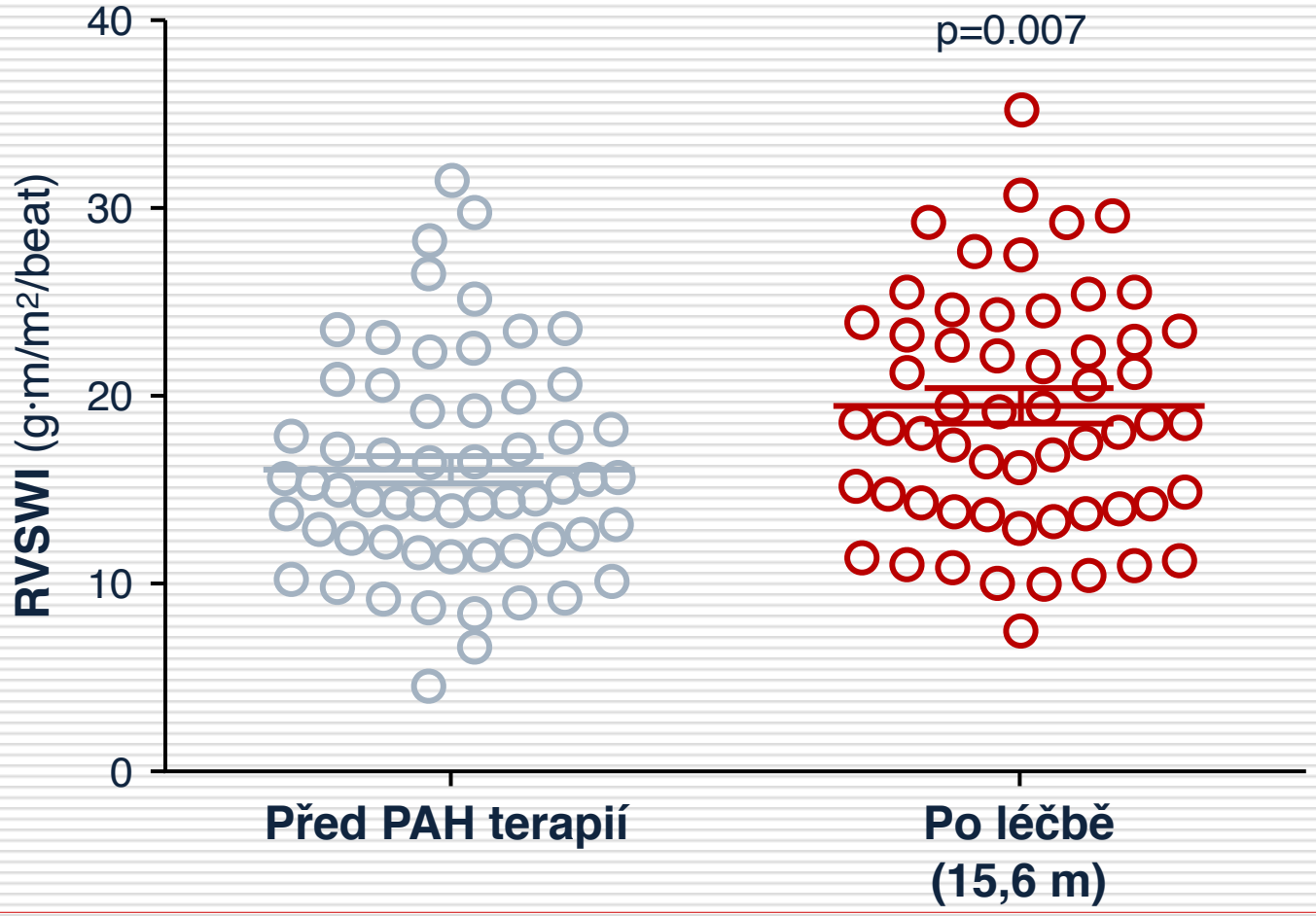


RVSWI (g·m/m²/beat) = (mean PAP – mean RAP) × (cardiac index/HR) × 0.0136

HR, heart rate; PAH, pulmonary arterial hypertension; PAP, pulmonary arterial pressure; RAP, right atrial pressure; RHC, right heart catheterisation; RV, right ventricle; RVSWI, right ventricular stroke work index

Brittain et al. *JACC Heart Fail*. 2013;1:300-307

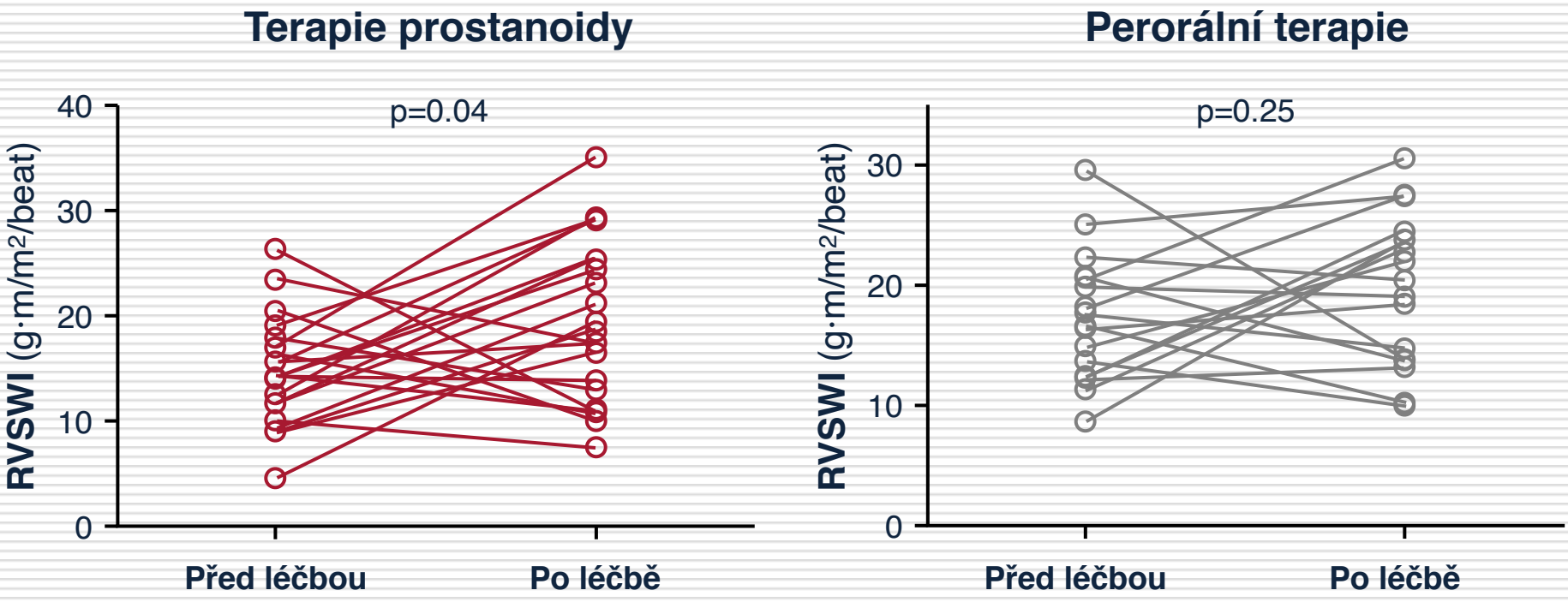
Reakce RVSWI na PAH specifickou léčbu



PAH, pulmonary arterial hypertension; RVSWI, right ventricular stroke work index

Adapted from Brittain et al. *JACC Heart Fail.* 2013;1:300-307

Reakce RVSWI podle typu PAH specifické léčby



Adapted from Brittain et al. *JACC Heart Fail.* 2013;1:300-307

I. INTERNÍ KLINIKA
KARDIOLOGIE
FAKULTNÍ NEMOCNICE OLOMOUC

Remodulin (treprostinil sodný)

- Dostupný od roku 2002
- **Biologický poločas** 4-5 hodin
- **Stabilní** za pokojové teploty a neutrálního pH
- **Roztok** připravený v lékovce

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Prognostic factors associated with increased survival in patients with pulmonary arterial hypertension treated with subcutaneous treprostinil in randomized, placebo-controlled trials

Raymond L. Benza, MD,[a] Mardi Gomberg-Maitland, MD, MSc,[b] Robert Naeije, MD, PhD,[c] Carl P. Arneson, MStat,[d] and Irene M. Lang, MD[e]

Cílová dávka treprostinilu

Parameter	Patients (N = 811)
Patients enrolled from blinded study, *n* (%)	399 (49)
Placebo	209
SC treprostinil	190
Mean age (range), years	45 (5–83)
Female, *n* (%)	628 (77)
PAH etiology, *n* (%)	
IPAH	425 (52)
APAH	386 (48)
CTD	166 (20)
CHD	177 (22)
PoPH	43 (5)
NYHA FC, *n* (%)	
II	126 (16)
III	614 (76)
IV	71 (9)
Concomitant PAH medication, *n* (%)	186 (23)
Alternative prostanoid[a]	67 (8)
Bosentan	99 (12)
Sildenafil	24 (3)
Time (mean ± SD) since diagnosis of PAH, years	3.5 ± 6.3
Hemodynamics (mean ± SD)	
mRAP, mm Hg	9.6 ± 5.5[b]
PVR, mm Hg/liter/min	14.7 ± 8.3[c]
PVRI, mm Hg/liter/min/m^2	25.3 ± 13.3[c]
CI, liters/min/m^2	2.4 ± 0.9[d]
SVO_2, %	61.7 ± 10.5[e]
6MWD (mean ± SD), m	333 ± 83

Table 2 On-treatment Results at Week 12

Parameter	*n*	Value (mean ± SD)
Treprostinil dose, ng/kg/min	811	8.3 ± 5.9
6MWD, m	391	354 ± 89
Hemodynamics		
mRAP, mm Hg	390	9.9 ± 6.1
PVR, mm Hg/liter/min	354	14.2 ± 8.3
PVRI, mm Hg/liter/min/m^2	354	24.2 ± 12.8
CI, liters/min/m^2	379	2.4 ± 0.9
SVO_2, %	365	62.5 ± 10.2

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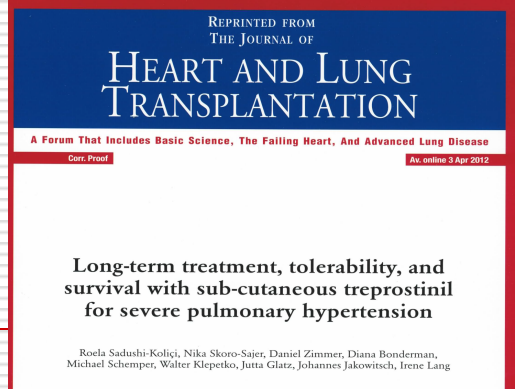
Table 6 Multivariate Predictors of Survival

Prognostic factor	Hazard ratio for death (95% CI)	*p*-value
Patient-related factors (based on available data for 811 patients)		
Age, ↑ 10 y	1.01 (0.89–1.15)	NS
BMI, ↑ 10 kg/m^2	0.52 (0.38–0.72)	<0.001
Albumin, ↑ 0.5 g/dl	0.70 (0.58–0.83)	<0.001
Sodium, ↑ 5 mmol/liter	0.90 (0.70–1.15)	NS
Total bilirubin, ↑ 0.1 mg/dl	1.02 (1.00–1.03)	0.033
Creatinine, ↑ 0.5 mg/dl	1.73 (1.31–2.28)	<0.001
On-treatment factors at Week 12		
Treprostinil dose[a] ↑ 10-ng/kg/min increments	0.64 (0.45–0.89)	0.009
6MWD[b] ↑ 20-m increments	0.86 (0.78–0.95)	0.004
PVRI[c] ↓ 10 mm Hg/liter/min/m^2	0.73 (0.44–1.21)	NS
SVO_2[d] ↑ 10% increments	0.66 (0.46–0.93)	0.018
Cardiac index[e] ↑ 1 liters/min/m^2	0.40 (0.12–1.28)	NS

Table 5 Univariate On-treatment Predictors of Survival

Prognostic factor	*n*	Hazard ratio for death (95% CI)	*p*-value
Treprostinil dose ↑ 10-ng/kg/min increments[a]	811	0.66 (0.48–0.90)	0.009
≥40 ng/kg/min[b]	230	0.29 (0.20–0.44)	<0.001
6MWD[a] ↑ 20-m increments	391	0.92 (0.86–0.99)	0.032
≥20-m increase	158	0.61 (0.38–0.98)	0.039
PVRI[a] ↓ 10 mm Hg/liter/min/m^2	335	0.70 (0.56–0.86)	<0.001
≤20 mm Hg/liter/min/m^2	354	0.40 (0.24–0.68)	<0.001
mRAP, ↑ 1 mm Hg[a]	390	1.06 (1.03–1.10)	<0.001
Cardiac index, ↑ 1 liter/min/m^2[a]	379	0.64 (0.47–0.87)	0.005

Tolerance treprostinilu



- **Celkové přežití po 1, 5 a 9 letech 84%, 53% a 33%**
- Pacienti, více než 6 měsíců na léčbě přežívali v 57% po 9 letech léčby

Table 1 Baseline Clinical and Hemodynamic Characteristics

	Patient groups			
Variables[a]	All	Treated ≥ 6 months	PAH[b]	CTEPH
Clinical data	(*n* = 111)	(*n* = 85)	(*n* = 39)	(*n* = 42)
Age, year	52 ± 17	53 ± 17	49 ± 18	61 ± 15
Sex				
Male	34 (31)	27 (32)	9 (23)	15 (36)
Female	77 (69)	58 (68)	30 (77)	27 (64)
Etiology of PH				
PAH	69 (62)	54 (64)		
CTEPH	42 (38)	31 (36)		
WHO FC				
III	62 (56)	51 (60)	21 (54)	20 (48)
IV	49 (44)	34 (40)	18 (46)	22 (52)
6MWD, meters	288 ± 108	293 ± 108	289 ± 119	269 ± 97
Borg Dyspnea Score	6 ± 2	6 ± 2	6 ± 2	7 ± 2
Time to				
Diagnosis, months	16 (3–276)	14 (2–276)	13 (3–276)	30 (6–96)
Treatment start, months	0.1 (0.1–3)	0.1 (0.1–1.2)	0.1 (0.1–1)	0.1 (0.1–3)
Hemodynamics				
Heart rate, beats/min	83 ± 18	83 ± 17	84 ± 15	81 ± 14
mRAP, mmHg	11 ± 5	11 ± 6	12 ± 6	11 ± 4
mPAP, mmHg	60 ± 16	60 ± 15	59 ± 15	56 ± 13
PCWP, mmHg	10.4 ± 4.2	10.3 ± 3.3	9.4 ± 3.3	11.2 ± 3.2
Cardiac output, liters/min	3.7 ± 1.0	3.8 ± 1.0	3.5 ± 1.0	3.6 ± 0.7
Svo_2, %	55.8 ± 8.3	59.8 ± 10.2	55.1 ± 8.1	56.1 ± 8.5
CI, liters/min/m^2	2.0 ± 0.4	2.1 ± 0.4	2.1 ± 0.4	2.0 ± 0.3
PVR, dyn/sec/cm^5	1,103 ± 519	1,075 ± 539	1,038 ± 499	1,000 ± 367

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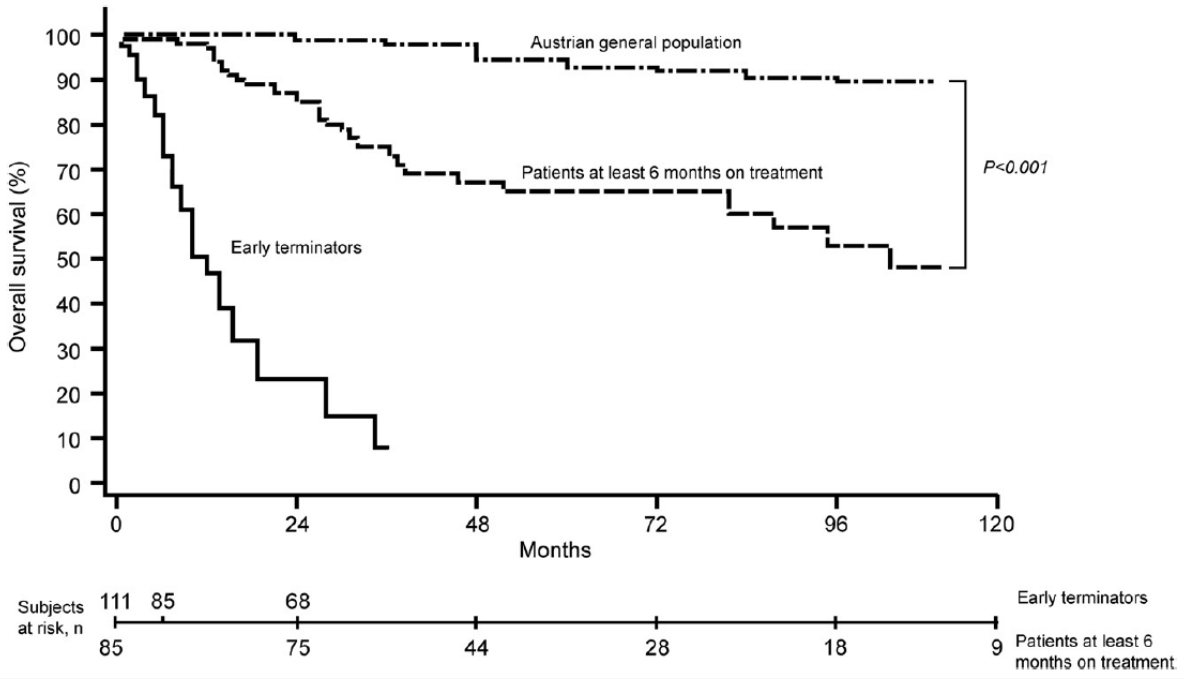
ORIGINAL CLINICAL SCIENCE

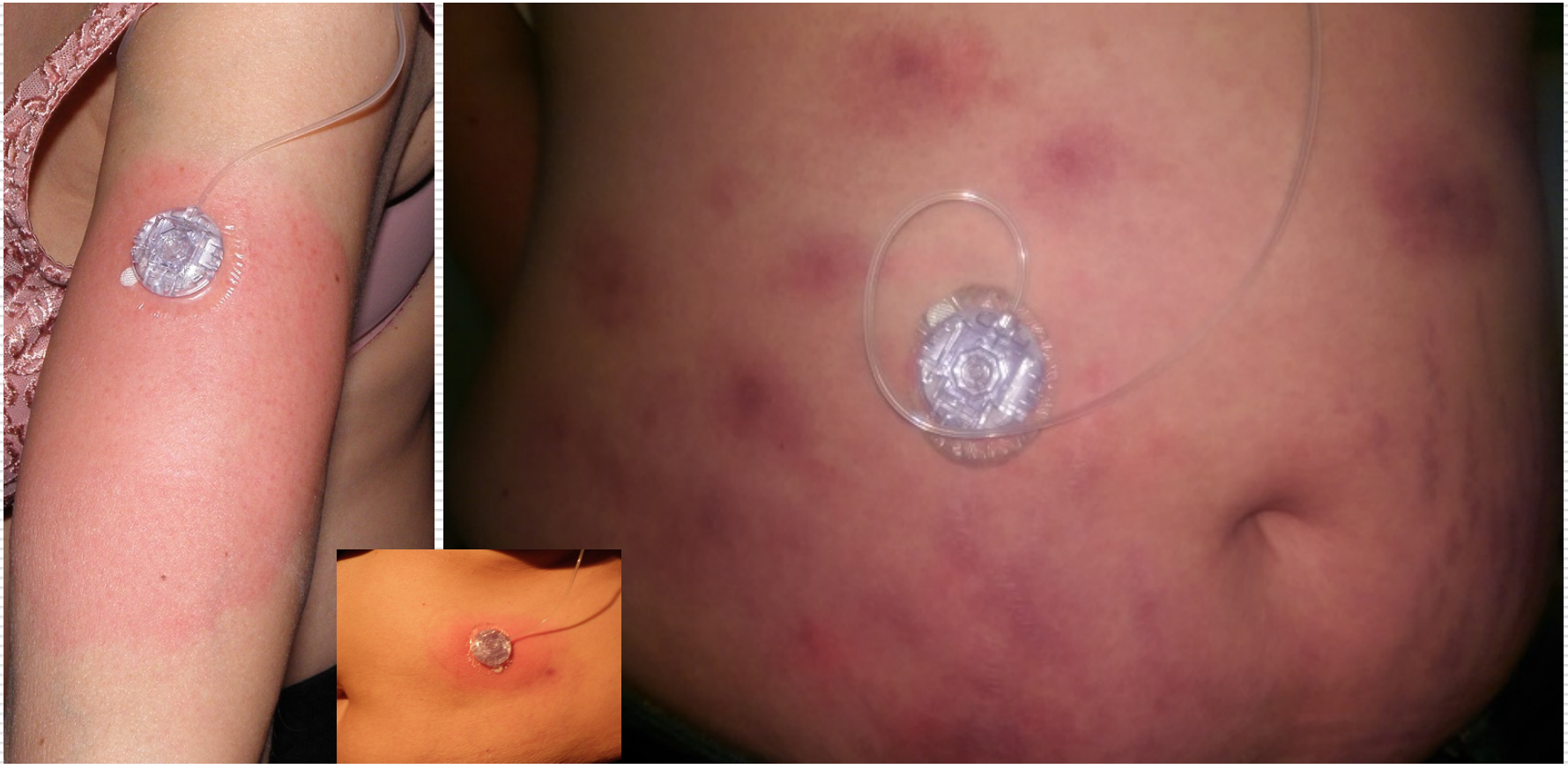
Long-term treatment, tolerability, and survival with sub-cutaneous treprostinil for severe pulmonary hypertension

Roela Sadushi-Koliçi, MD,[a] Nika Skoro-Sajer, MD,[a] Daniel Zimmer, MD,[a] Diana Bonderman, MD,[a] Michael Schemper, PhD,[b] Walter Klepetko, MD,[c] Jutta Glatz, MSc,[d] Johannes Jakowitsch, PhD,[a] and Irene Lang, MD[a]



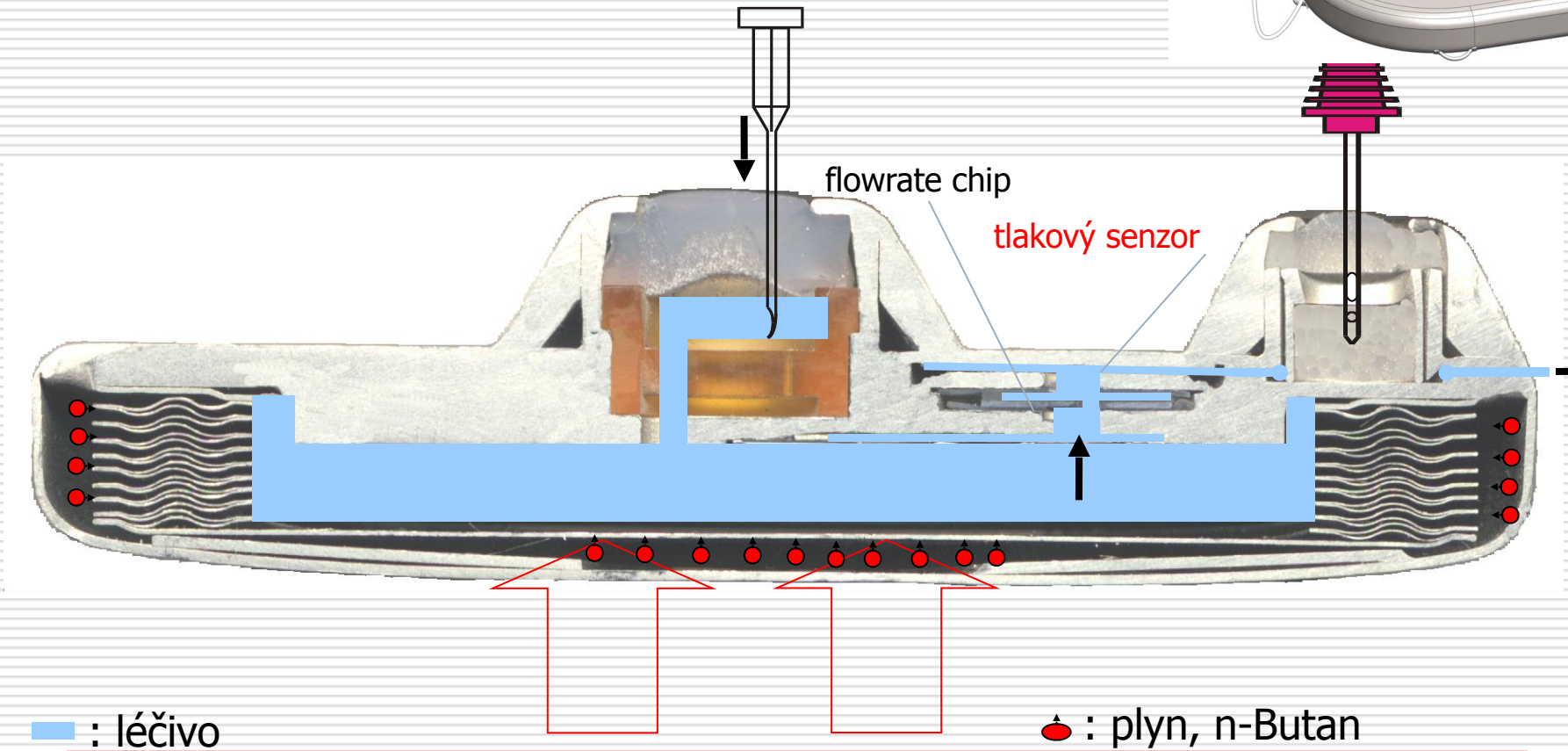
Adverse event relationship with underlying disease or treatment	No. (%)[a]
Likely	
Infusion site reaction	111 (100)
Infusion site pain	89 (80)
Diarrhea	24 (21.6)
Facial flushing	15 (13.5)
Local abscesses	3 (2.7)
Painful swelling of inguinal lymph nodes	3 (2.7)
Persistent lower limb pain	2 (1.8)
Hospitalizations for IV diuretics and/or vasopressors	31 (27.9)
Unlikely	
Major surgeries	
Urologic surgery	4 (3.6)
Hip replacement	2 (1.8)
Other orthopedic surgeries	2 (1.8)
Aortic valve replacement	1 (0.9)
Hepatic surgery	1 (0.9)
Minor surgeries	16 (14.4)
Cytostatic chemotherapy	1 (0.9)

Až 80% pacientů – reakce v místě vpichu
Nejčastější důvod přerušení léčby (15%), v prvním roce podávání
Míra bolestivosti často nekoreluje s mírou lokální reakce



Simonneau G, Barst RJ, Galie N et al., Am J Respir Crit Care Med. 2002 Mar 15;165(6):800-4.
Barst RJ, Galie N, Naeije R et al., Eur Respir J. 2006 Dec;28(6):1195-203.
Lang IM, Gomez-Sanchez M, Kneussl M et al., Chest. 2006 Jun;129(6):1636-43.

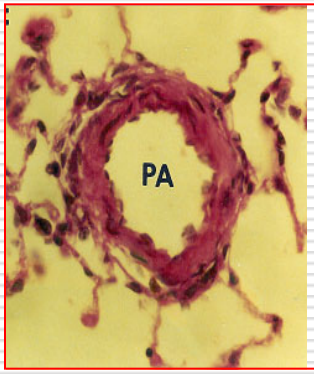
LENUSpro® pumpa



: léčivo

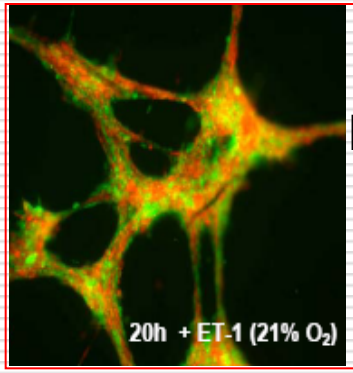
: plyn, n-Butan

Antagonisté receptorů pro endotelin (ERA)



Buňky hladké svaloviny

Vazokonstrikce, hypertrofie, proliferace[1,2]

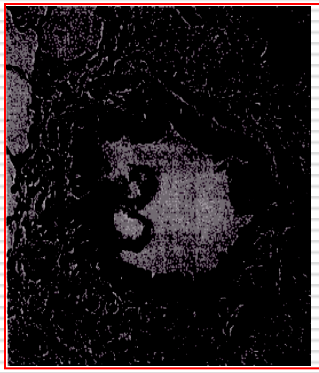


Endoteliální buňky

Hypetrofie,

dysfunkce[3-6]

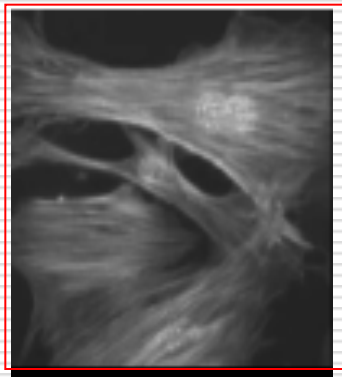
Endotelin



Makrofágy

Zánět

a uvolnění cytokinů[10-13]



Fibroblasty

Fibróza[7-9]

1. Clozel et al; J Pharmacol Exp Ther (1989), 2. Yang et al; Circulation (1999), 3. Kuhlmann et al; Acta Physiol Scand (2005), 4. Davie Univ. Colorado Health Sciences Center. With permission, 5. Girgis et al; Am J Respir Crit Care Med (2005), 6. Amiri et al; Circulation (2004), 7. Cambrey et al; Am J Respir Cell Mol Biol (1994), 8. Shi-Wen et al; J Invest Dermatol (2001), 9. Shi-Wen et al; Mol Biol Cell (2004), 10. Yang et al; Circulation (2004), 11. Wilson et al; Biochem Piophys Res Comm (2001), 12. Helset et al; Am J Physiol (1996), 13. Salani et al; Am J Pathol (2000)

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Updated Treatment Algorithm of Pulmonary Arterial Hypertension

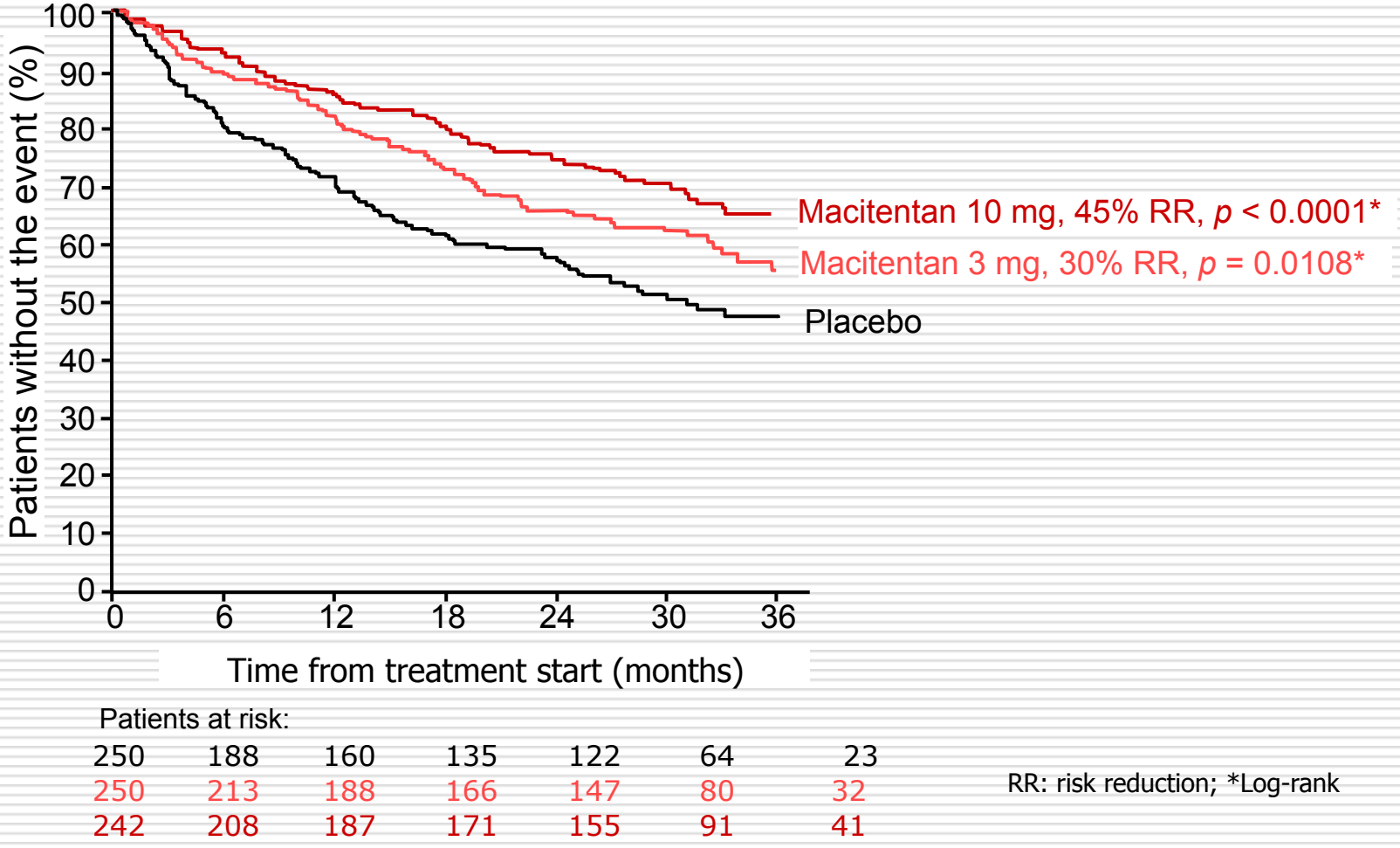
Nazzareno Galiè, MD,* Paul A. Corris, MD,† Adaani Frost, MD,‡ Reda E. Girgis, MD,§ John Granton, MD,‖ Zhi Cheng Jing, MD,¶ Walter Klepetko, MD,# Michael D. McGoon, MD,** Vallerie V. McLaughlin, MD,†† Ioana R. Preston, MD,‡‡ Lewis J. Rubin, MD,§§ Julio Sandoval, MD,‖‖ Werner Seeger, MD,¶¶ Anne Keogh, MD##

Drug(s) Tested	Study	Background	Primary Endpoint	Outcome (Secondary Endpoint)	Duration (weeks)	No. of Patients
Ambrisentan	ARIES-1	No	6MWD	TTCW (NS)	12	202
	ARIES-2	No	6MWD	TTCW	12	192
Bosentan	Study-351	No	6MWD	TTCW	12	32
	BREATHE-1	No	6MWD	TTCW	16	213
	BREATHE-2*	No	PVR	—	12	33
	EARLY	No Sildenafil (16%)	PVR, 6MWD	TTCW	24	185
	BREATHE-5	No	SaO_2, PVR	—	12	54
Macitentan†	SERAPHIN	No, PDE5i or Inhal iloprost	TTCW	Safety	100	742

*Bosentan + epoprostenol versus placebo + epoprostenol. †Approved by the FDA for PAH patients and has obtained at the time of printing the positive opinion of the Committee for Medicinal Products for Human Use of the the EMA for this indication.

6MWD = 6-min walk distance; inhal = inhalation; NS = not statistically significant; PDE5i = phosphodiesterase type-5 inhibitors; PVR = pulmonary vascular resistance; SaO_2 = finger oxygen saturation; TTCW = time to clinical worsening.

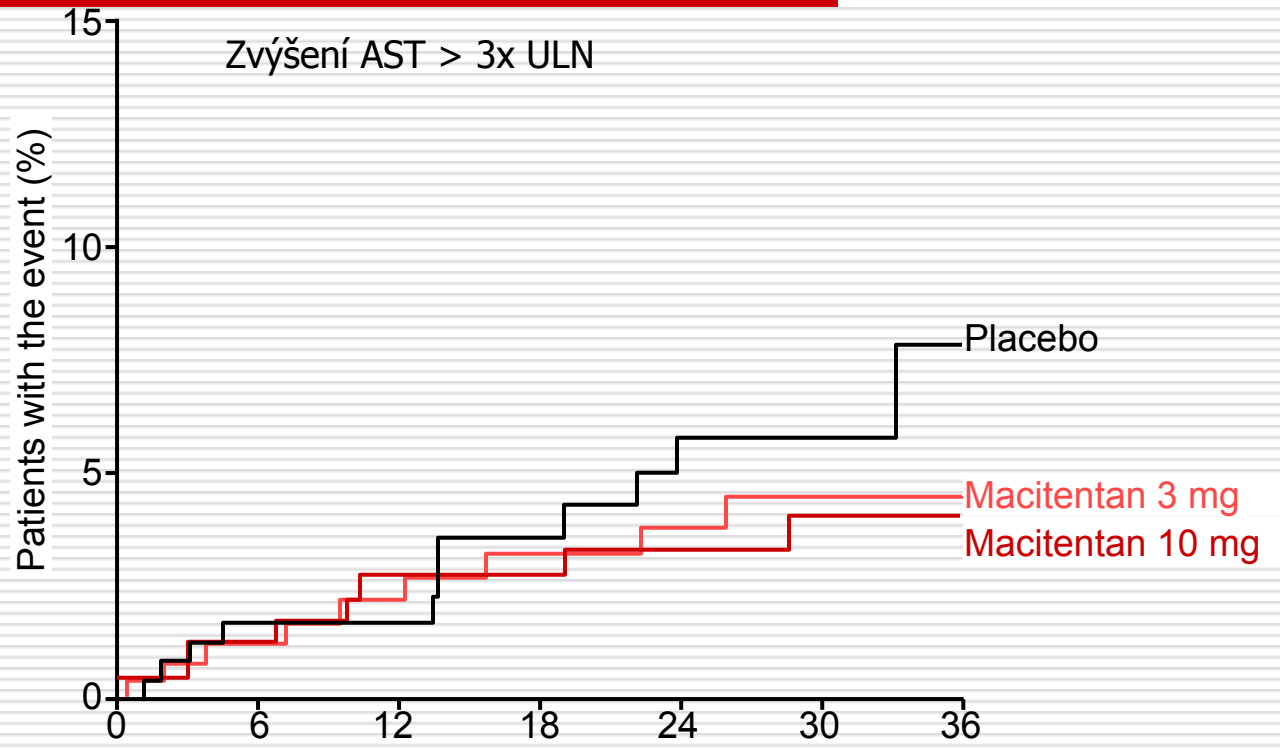
Primární endpoint: Macitentan signifikantně snižuje riziko morbidity/mortality



RR: risk reduction; *Log-rank

Pulido, et al. N Engl J Med 2013.

Indukce hepatopatie



Pulido, et al. N Engl J Med 2013.

Léky interferující s biologickou dostupností NO

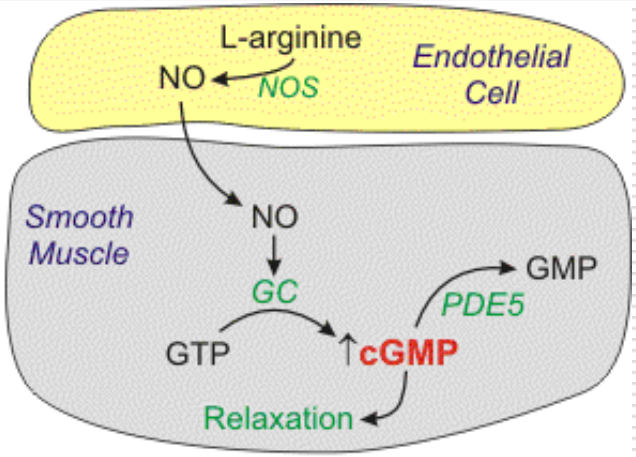


Table 4 Characteristics of Randomized Controlled Trials With Pulmonary Arterial Hypertension Drugs Interfering With the Nitric Oxide Pathway (See Text for References)

Drug(s) Tested	Study	Background	Primary Endpoint	Outcome (Secondary Endpoint)	Duration (weeks)	No. of Patients
Soluble Guanylate Cyclase Stimulators						
Riociguat*	PATENT	No bosentan or prostanoids	6MWD	TTCW	12	443
Phosphodiesterase Type-5 Inhibitors						
Sildenafil	SUPER-1	No	6MWD	TTCW (NS)	12	277
	Sastry	No	TT	—	12	22
	Singh	No	6MWD	—	6	20
	PACES	Epoprostenol	6MWD	TTCW	16	264
	Iversen	Bosentan	6MWD	—	12	20
Tadalafil	PHIRST	No or bosentan (54%)	6MWD	TTCW	16	405
Vardenafil†	EVALUATION	No	6MWD	TTCW	12	66

*Approved by the FDA for PAH and CTEPH patients and is currently undergoing the regulatory approval process by the EMA for both indications. †Not approved for pulmonary arterial hypertension. TT = treadmill test; other abbreviations as in Table 3.

Ryerson *et al. Respiratory Research* 2010, **11**:12
http://respiratory-research.com/content/11/1/12

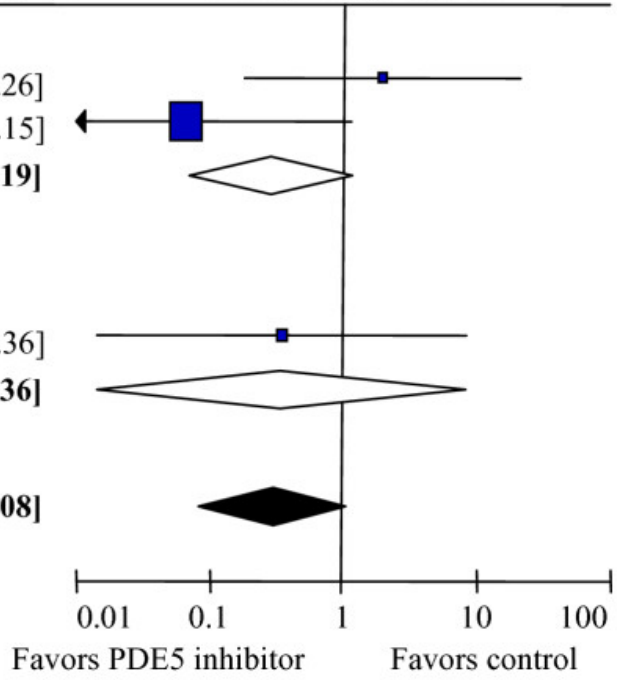
RESPIRATORY RESEARCH

RESEARCH Open Access

Pharmacotherapy in pulmonary arterial hypertension: a systematic review and meta-analysis

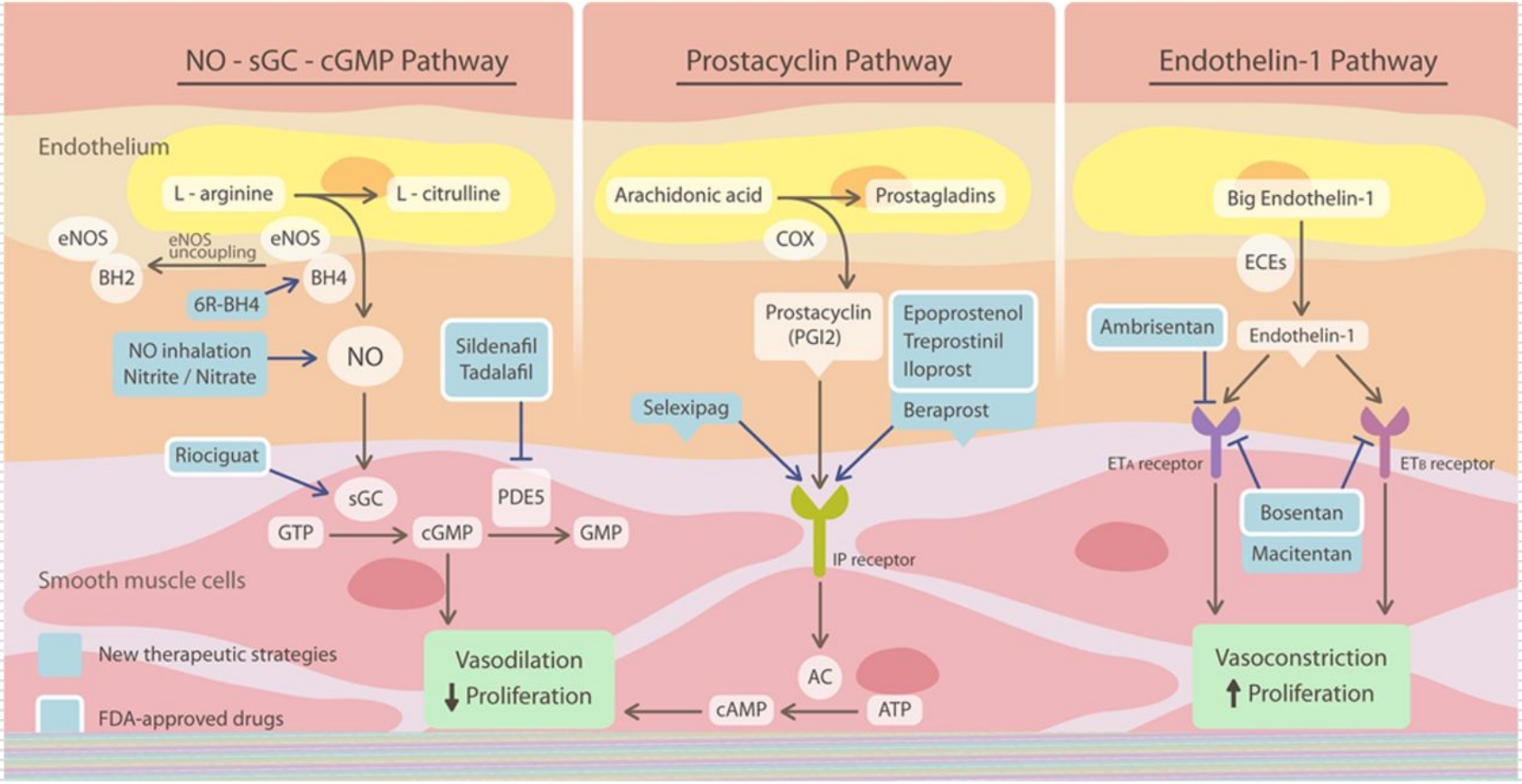
Christopher J Ryerson*, Shalini Nayar, John R Swiston, Don D Sin

Study	PDE5 Inhibitor Events	PDE5 Inhibitor Total	Placebo Events	Placebo Total	Weight	Risk Ratio M-H, Fixed, 95% CI
Sildenafil						
Galie 2005 [29]	2	71	1	70	10.1%	1.97 [0.18, 21.26]
Simonneau 2008 [30]	0	134	7	133	75.2%	0.07 [0.00, 1.15]
Subtotal (95% CI)	**2**	**205**	**8**	**203**	**85.3%**	**0.29 [0.07, 1.19]**
Heterogeneity: P = 0.06						
Tadalafil						
Galie 2009 [31]	0	79	1	82	14.7%	0.35 [0.01, 8.36]
Subtotal (95% CI)	**0**	**79**	**1**	**82**	**14.7%**	**0.35 [0.01, 8.36]**
Heterogeneity: Not applicable						
Total (95% CI)	**2**	**284**	**9**	**285**	**100.0%**	**0.30 [0.08, 1.08]**
Heterogeneity: P = 0.17						



Monoterapie vs. kombinační léčba PAH

Selexipag (GRIPHON)

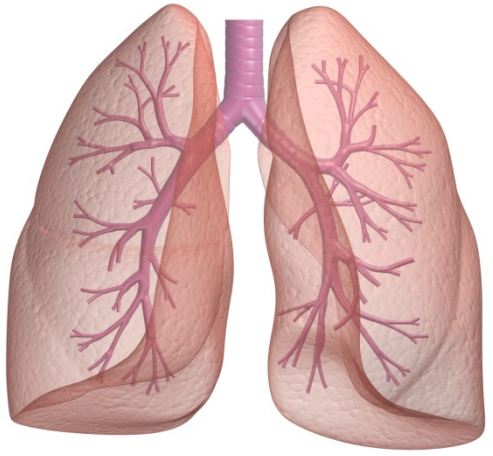


Transplantační léčba

Indikována u pacientů s neadekvátní klinickou odpovědí na zavedenou a maximalizovanou farmakoterapii PAH

Časování k vyšetření se záměrem zařazení na WL LTx je určeno nejen vstupními symptomy, následnou odpovědí na specifickou léčbu PAH, ale i dalšími prognostickými markery PAH (etiologie)

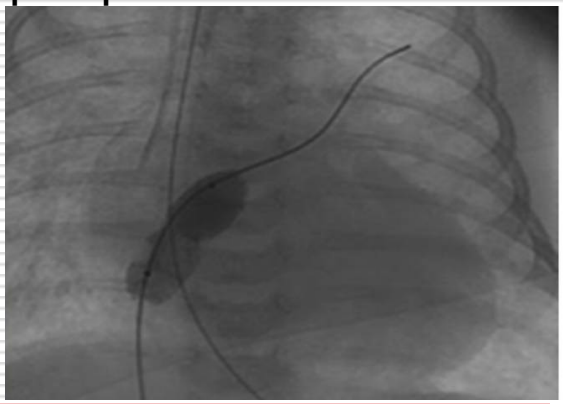
	1 year	5 years	10 years
Pittsburgh (Toyoda et al., 2008 [74])	86	75	66
Paris (Fadel et al., 2010 [75])	79	52	43
Toronto (de Perrot et al., 2012 [76])	78	60	45
Vienna (Klepetko, unpublished data, 2011)	73	71	—



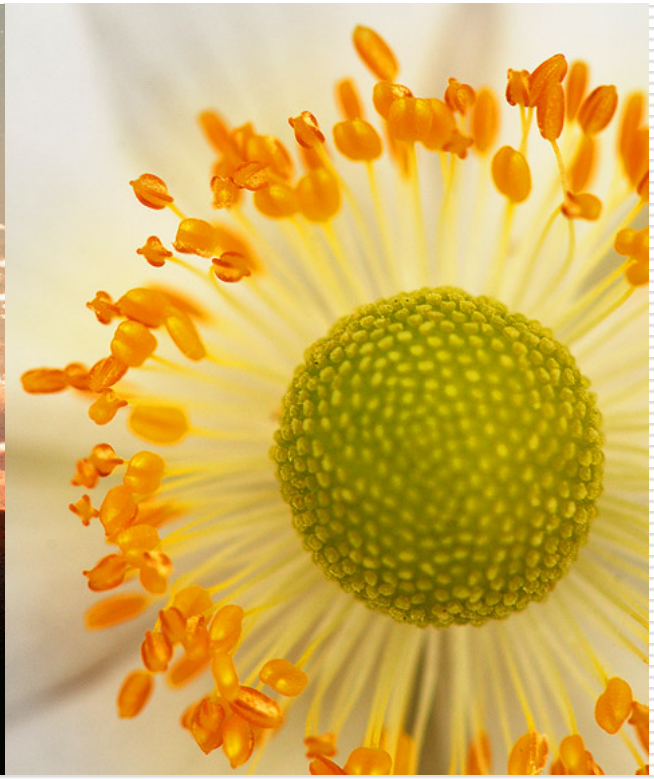
J Am Coll Cardiol. 2013;62(25_S):. doi:10.1016/j.jacc.2013.10.031

Balónková septostomie

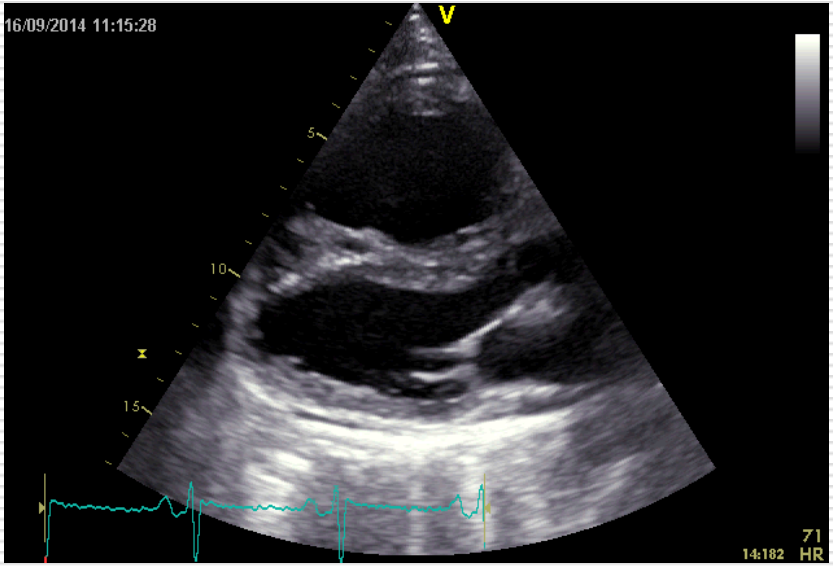
Atriální septostomie - balloon atrial septostomy (BAS) je intervenční procedura, která je **indikována** v rámci paliace nebo přemostění k LTx u pacientů s PAH ve funkčním stádiu WHO/NYHA IV s refrakterním pravostranným srdečním selháním a rekurentními synkopami.
Cílem intervence je zvýšení srdečního výdeje za cenu systémové desaturace. Saturace (spO2) by neměla po výkonu klesnout o více než 10 %.
Hemodynamickým **důsledkem** septostomie okolo 5-8 mm průměru je zvýšení srdečního výdeje o 20–25 %.
Výkon je **kontraindikován** při RAP> 20 mm Hg a při spO2 < 80 %.

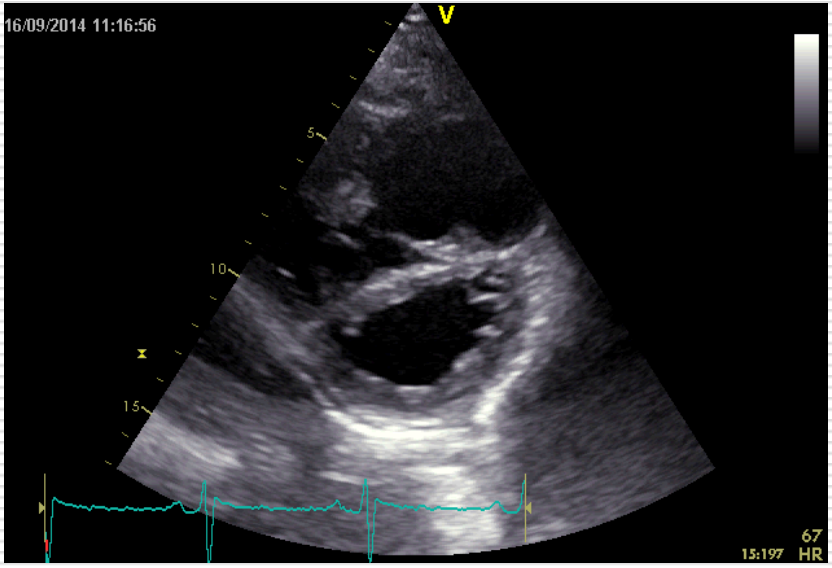


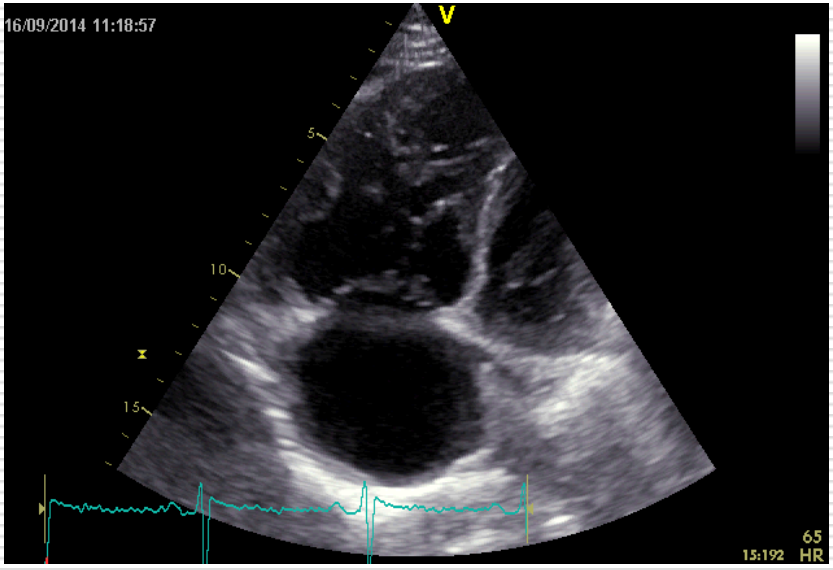
Děkuji za pozornost

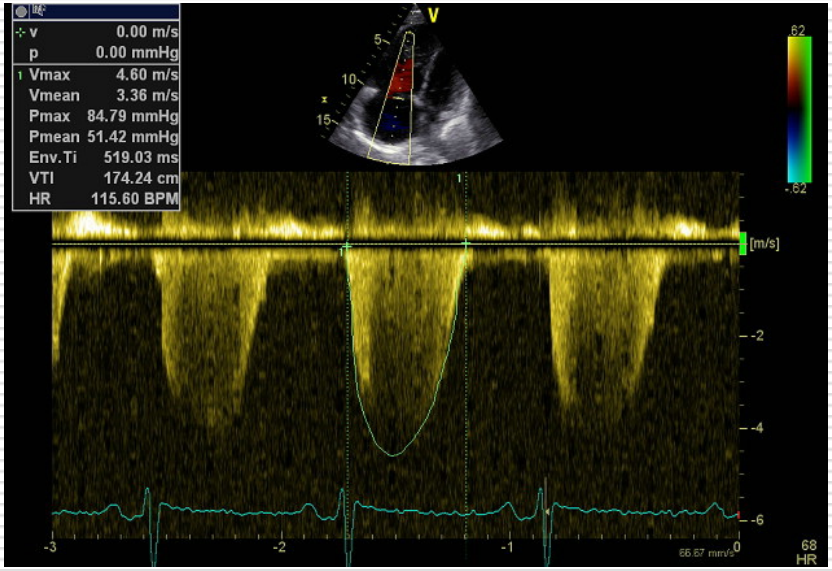


KAZUISTIKA 1






16/09/2014 11:18:57
V
5
10
15
65
15:192
HR


v 0.00 m/s
p 0.00 mmHg
Vmax 4.60 m/s
Vmean 3.36 m/s
Pmax 84.79 mmHg
Pmean 51.42 mmHg
Env.Ti 519.03 ms
VTI 174.24 cm
HR 115.60 BPM
V
82
-82
[m/s]
-2
-4
-6
-3
-2
-1
0
66.67 mm/s
68
HR

Pravostranná srdeční katetrizace:

TK arteriální 115/80 mmHg, spO2 94%, P 84/ min., SR, spO2 smíšená ven. krev 64,4%

PS: 15/9/13 mmHg, spO2 61,0%

PK: 89/12 mmHg (systolický/diastolický), spO2 60,2%

AP: 90/40/59 mmHg (systolický/diastolický/ střední), spO2 61,9%

PCWP: 11 mmHg (střední)

PVR: 13,63 WU

CO: 3,52 l/min.

CI: 1,9 l/min./m2

Závěr: Těžká fixovaná plicní arteriální hypertenze prekapilární, idiopatická plicní arteriální hypertenze, NYHA III, bez evidence intrakardiálního levo-pravého zkratu, normální plnící tlaky LK, snížený pravostranný srdeční výdej, zvýšená plicní vaskulární rezistence.

6MWT

TK 127/94, TF 82/min,

Dušnost (borgova škála) start 0, konec 8

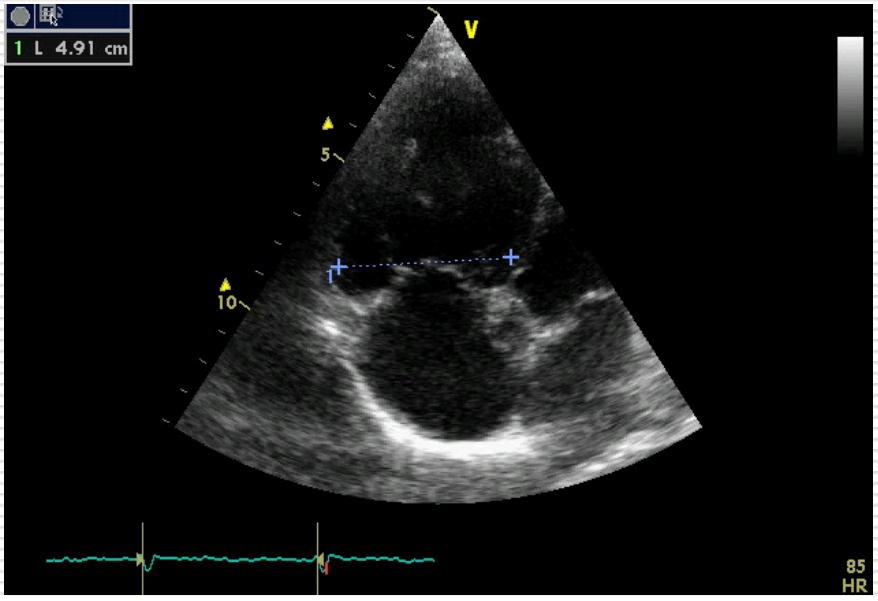
Únava (Borgova škála) start 0, konec 6

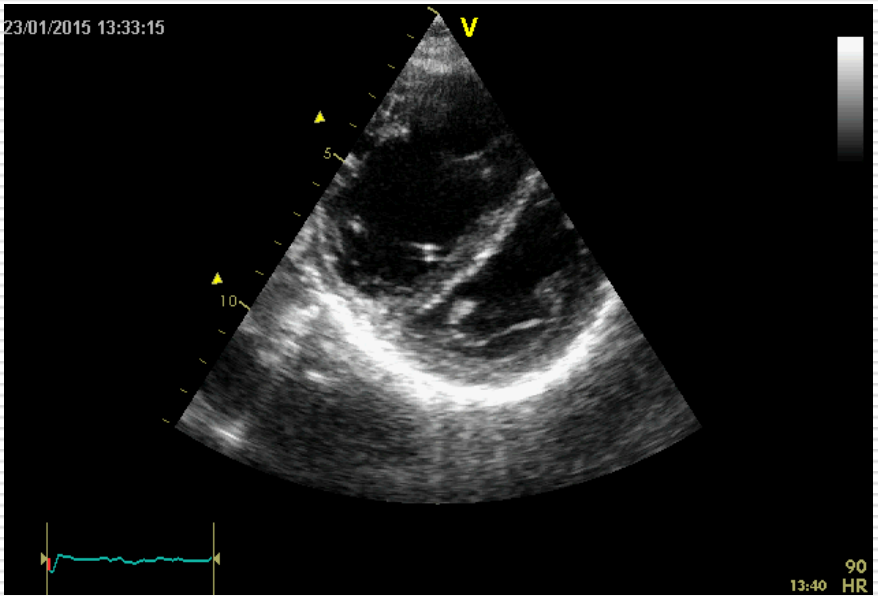
SpO2 start 87%, konec 98%

Přerušení chůze během testu+důvod? 2x krátká pauza pro výraznou dušnost

Celková vzdálenost 260m

KAZUISTIKA 2





Pravostranná srdeční katetrizace: Na katetrizačním sále pod skiaskopickou kontrolou v lokální anestezii Supracain inj. 1 amp. bez komplikací zaveden 8F sheath do VJI dx. a arteriofix AR dx. Dále pod skiaskopickou kontrolou zaveden Swan-Ganzův katétr do plicnice a distálně do zaklínění. Změřeny pravostranné srdeční tlaky, srdeční výdej termodiluční metodou, z jednotlivých srdečních oddílů provedeny odběry na oxymetrii (analyzátor CCX). Poté proveden test vazoreaktivity ve standardním eskalovaném schématu Flolan 2-12 ng/kg/min. s dekrementem 2 ng/kg/min. po 10 min., od dávky Flolanu 6 ng/kg/min. mírný flush obličeje se setvralým poklesem systémové tenze - asymptomatickým a bolesti hlavy, regredující po ukončení testu.

Měřené hodnoty klidové: TK arteriální 121/81/90 mmHg, spO2 98%, arteriální spO2 95,5%, pO2 9,32 kPa, pCO2 2,75 kPa, pH 7,46, horní dutá žíla spO2 65%, P 67/min., SR, sO2 98%

PS: 20/15/17 mmHg, spO2 62,2%

PK: 73/20 mmHg, spO2 65,3%

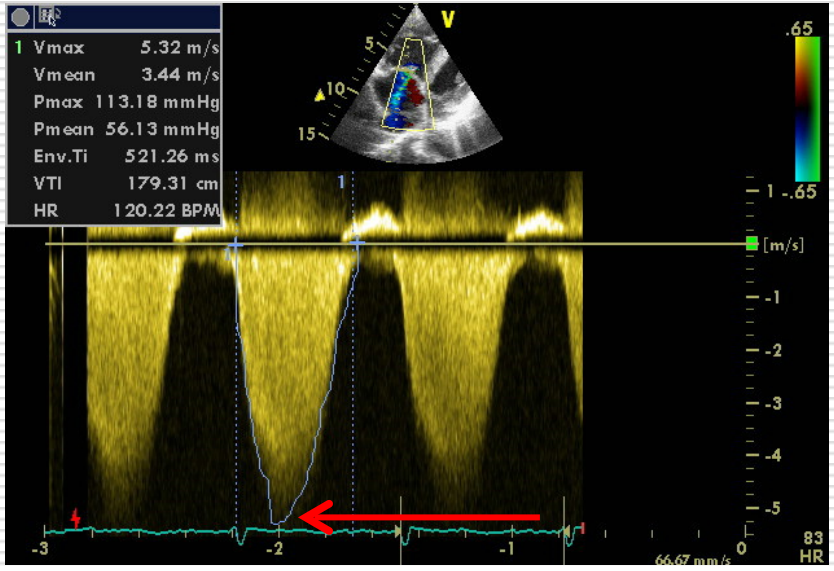
AP: 74/45/57 mmHg, spO2 69,7%

PCWP: 10 mmHg

PVR: 17,6 WU

CO: 2,67 l/min.

CI: 1,41 l/min./m2



Test vazoreaktivity:

Flolan 0 ng/kg/min.: AP 74/47/57, TK 121/81/90 mmHg

Flolan 2 ng/kg/min.: AP 77/46/58, TK 119/79/90 mmHg

Flolan 4 ng/kg/min.: AP 77/45/58, TK 116/75/89 mmHg

Flolan 6 ng/kg/min.: AP 78/46/57, TK 107/67/79 mmHg

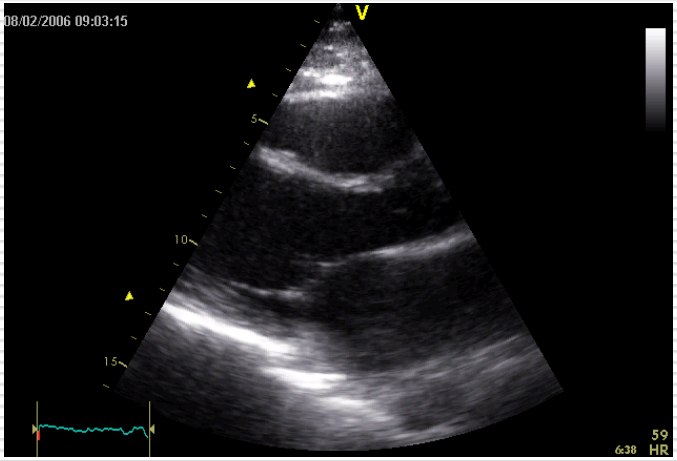
Flolan 8 ng/kg/min.: AP 76/45/57, TK 106/66/75 mmHg

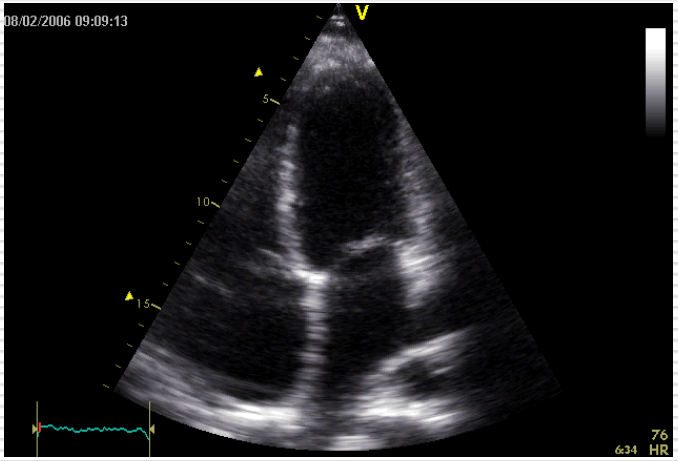
Flolan 10 ng/kg/min.: AP 75/43/56, TK 102/60/73 mmHg, CO 3,55 l/min.

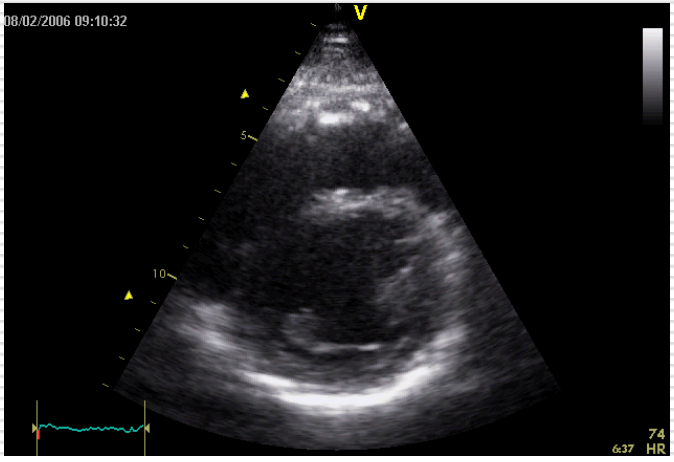
Flolan 12 ng/kg/min.: AP 80/51/60 mmHg, TK 106/64/76 mmHg, P 83/min., CO 3,2 l/min., CI 3 l/min., WP 9

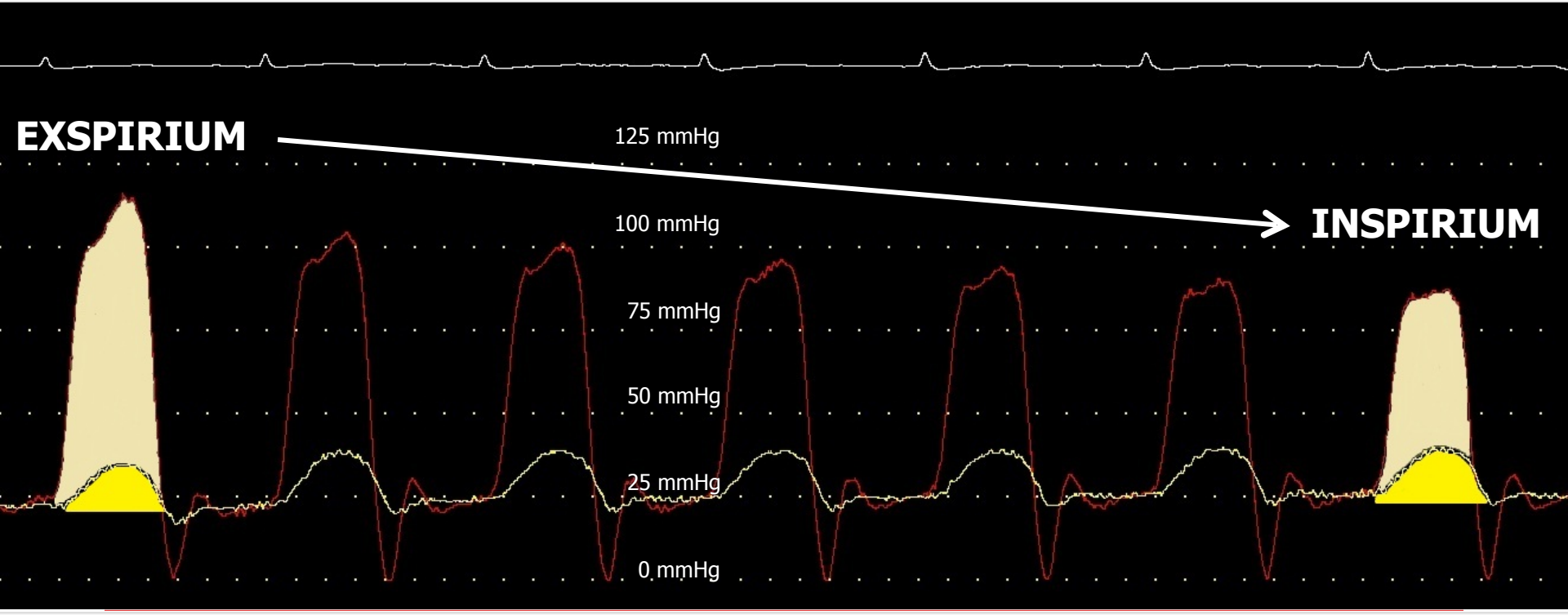
Klid po testu: AP 76/45/58 mmHg, arteriální TK 120/75/86 mmHg,P 83/min., spO2 98%

KAZUISTIKA 3








EXSPIRIUM
INSPIRIUM
125 mmHg
100 mmHg
75 mmHg
50 mmHg
25 mmHg
0 mmHg

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doi:10.1016/j.jacc.2007.09.039

Constrictive Pericarditis in the Modern Era

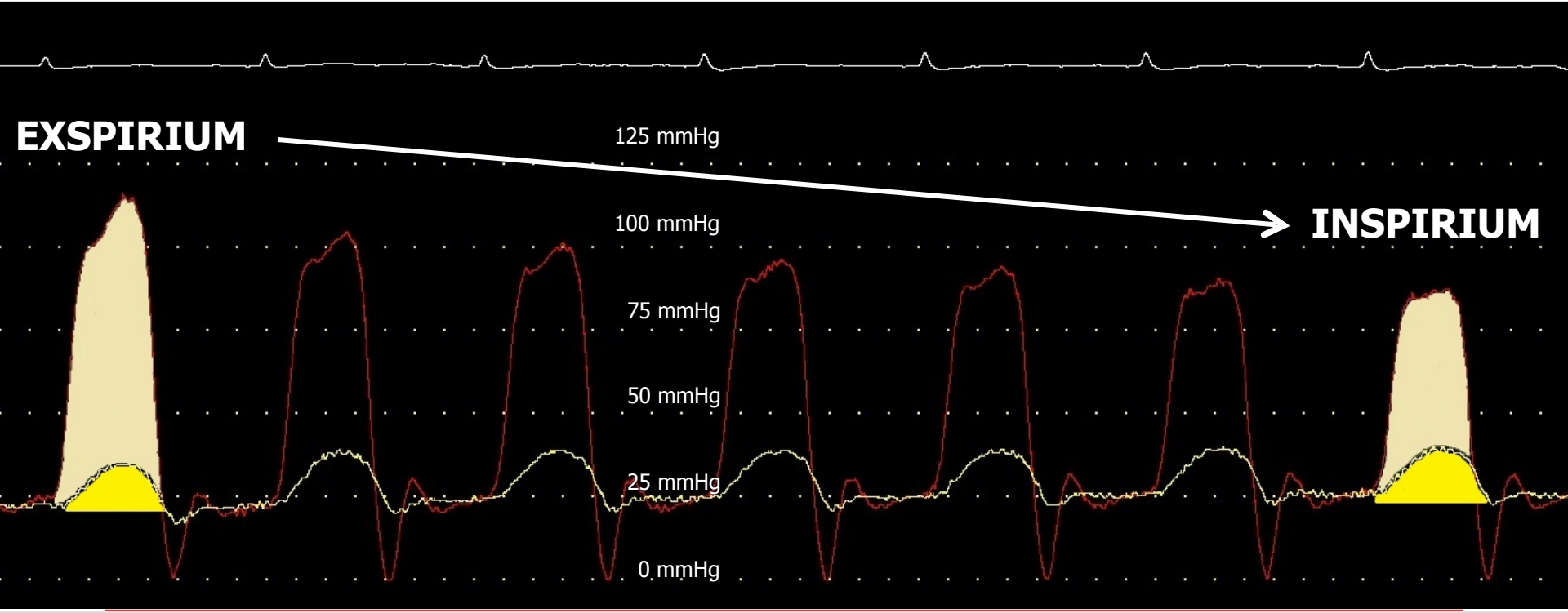
Novel Criteria for Diagnosis in the Cardiac Catheterization Laboratory

Deepak R. Talreja, MD, FACC, Rick A. Nishimura, MD, FACC, Jae K. Oh, MD, FACC, David R. Holmes, MD, FACC

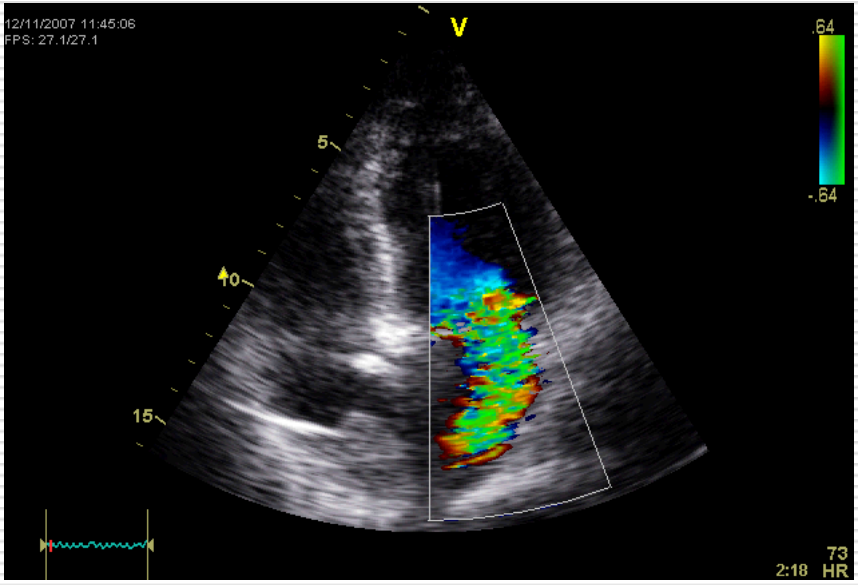
Rochester, Minnesota

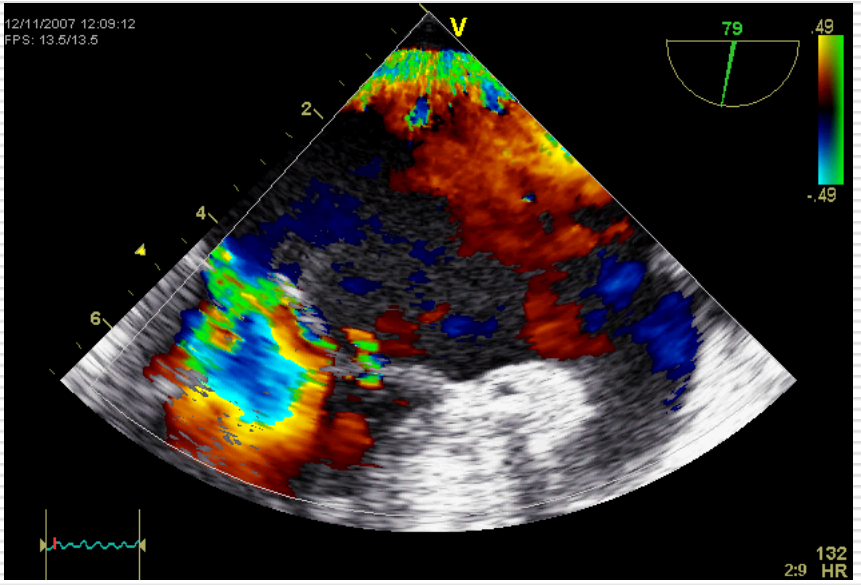
Table 2 Catheterization Criterion

Criterion	Sensitivity (%)	Specificity (%)	Positive Predictive Accuracy (%)	Negative Predictive Accuracy (%)
LVEDP − RVEDP ≤5 mm Hg	46	54	58	40
PASP <55 mm Hg	90	29	73	66
RVEDP/RVSP >1/3	93	46	71	79
LVRFW >7 mm Hg	45	44	62	42
Inspiratory decrease in RAP <5 mm Hg	71	37	62	39
Systolic area index >1.1	97	100	100	95

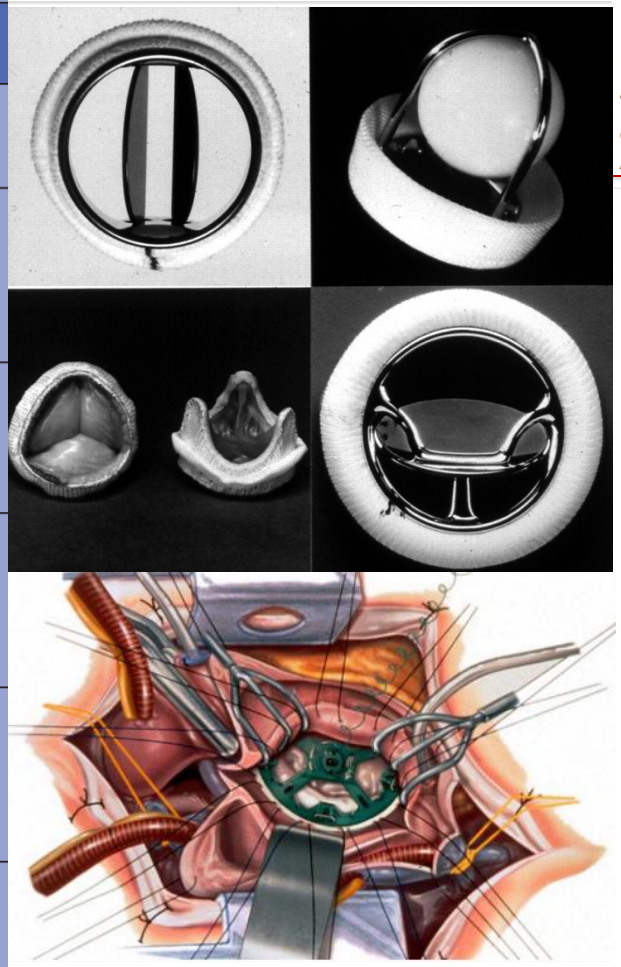


KAZUISTIKA 4





	Class [a]	Level [b]
Mitral valve repair should be the preferred technique when it is expected to be durable.	I	C
Surgery is indicated in symptomatic patients with LVEF >30% and LVESD <55 mm.	I	B
Surgery is indicated in asymptomatic patients with LV dysfunction (LVESD ≥45 mm and/or LVEF ≤60%).	I	C
Surgery should be considered in asymptomatic patients with preserved LV function and new onset of atrial fibrillation or pulmonary hypertension (systolic pulmonary pressure at rest >50 mmHg).	IIa	C
Surgery should be considered in asymptomatic patients with preserved LV function, high likelihood of durable repair, low surgical risk and flail leaflet and LVESD ≥40 mm.	IIa	C
Surgery should be considered in patients with severe LV dysfunction (LVEF <30% and/or LVESD >55 mm) refractory to medical therapy with high likelihood of durable repair and low comorbidity.	IIa	C
Surgery may be considered in patients with severe LV dysfunction (LVEF <30% and/or LVESD >55 mm) refractory to medical therapy with low likelihood of durable repair and low comorbidity.	IIb	C
Surgery may be considered in asymptomatic patients with preserved LV function, high likelihood of durable repair, low surgical risk, and: • left atrial dilatation (volume index ≥60 ml/m² BSA) and sinus rhythm, or • pulmonary hypertension on exercise (SPAP ≥60 mmHg at exercise).	IIb	C

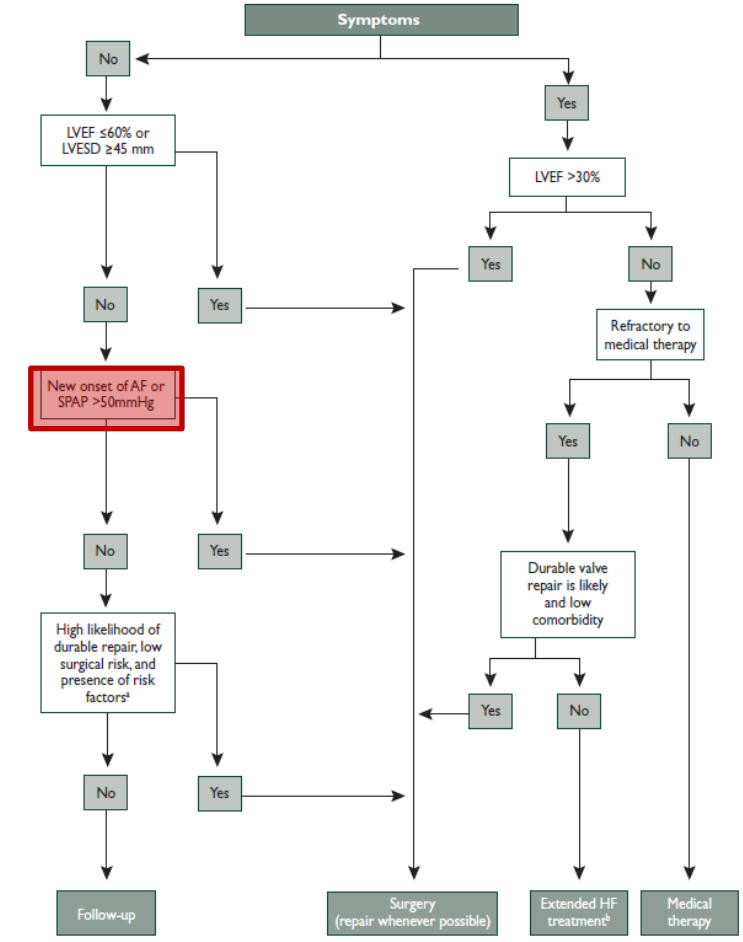


European Heart Journal (2012) 33, 2451–2496
doi:10.1093/eurheartj/ehs109

ESC/EACTS GUIDELINES

Guidelines on the management of valvular heart disease (version 2012)

The Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS)



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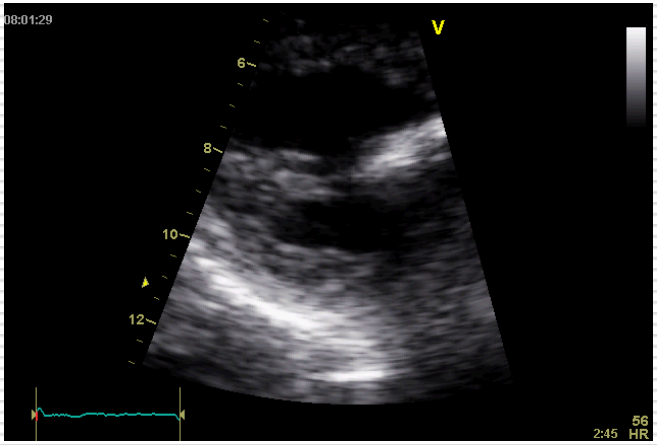

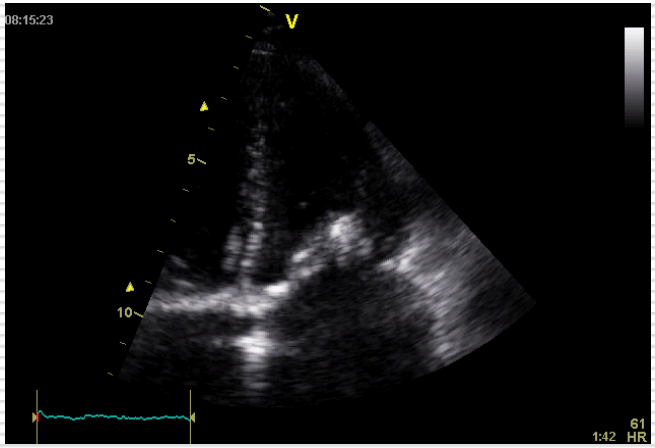

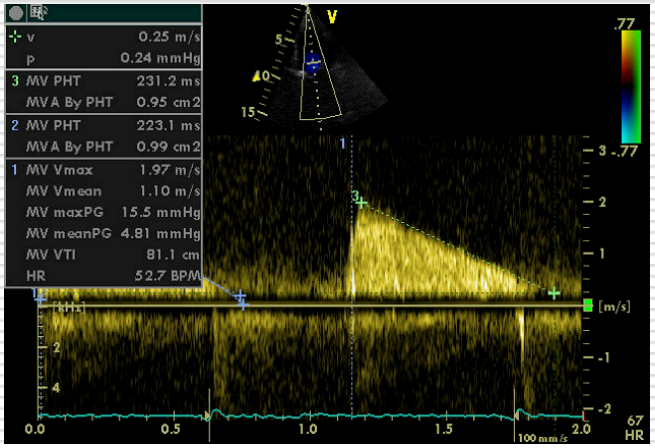

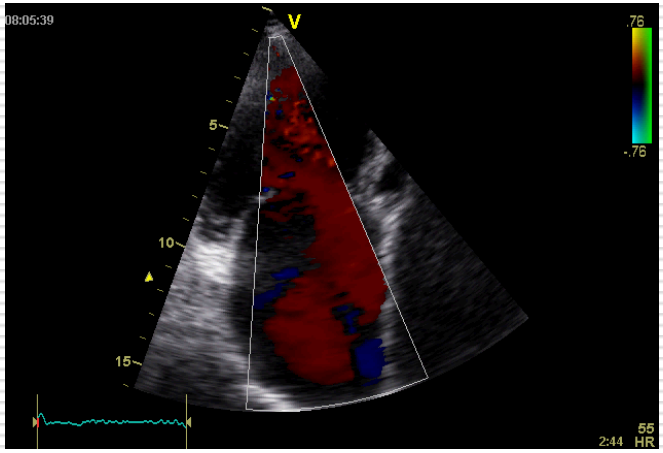

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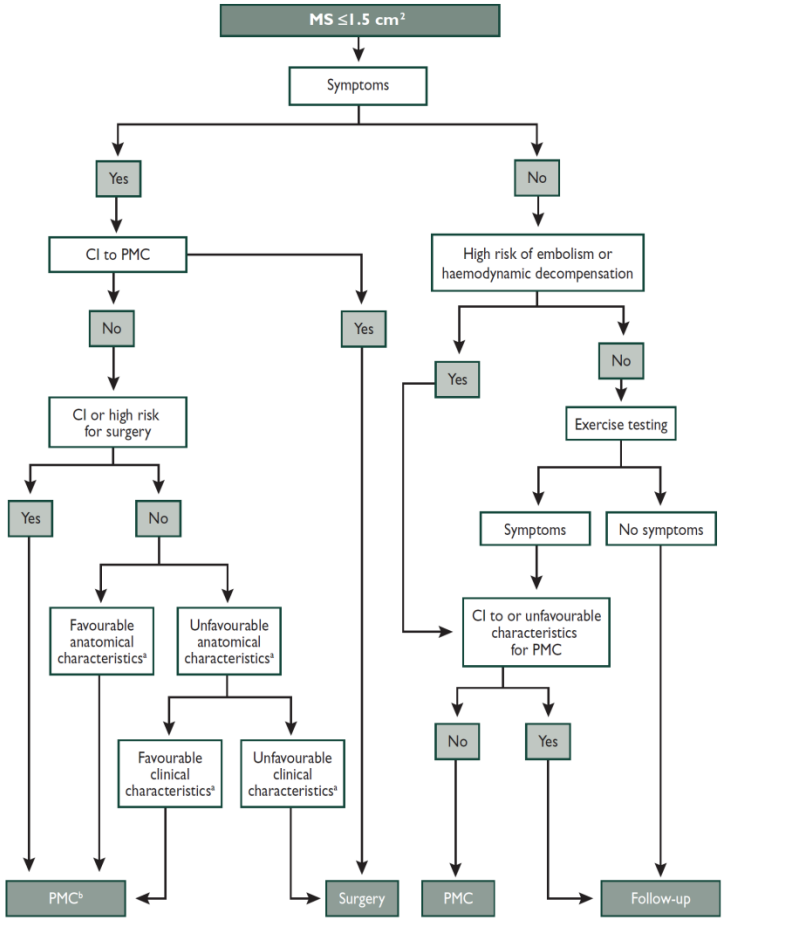
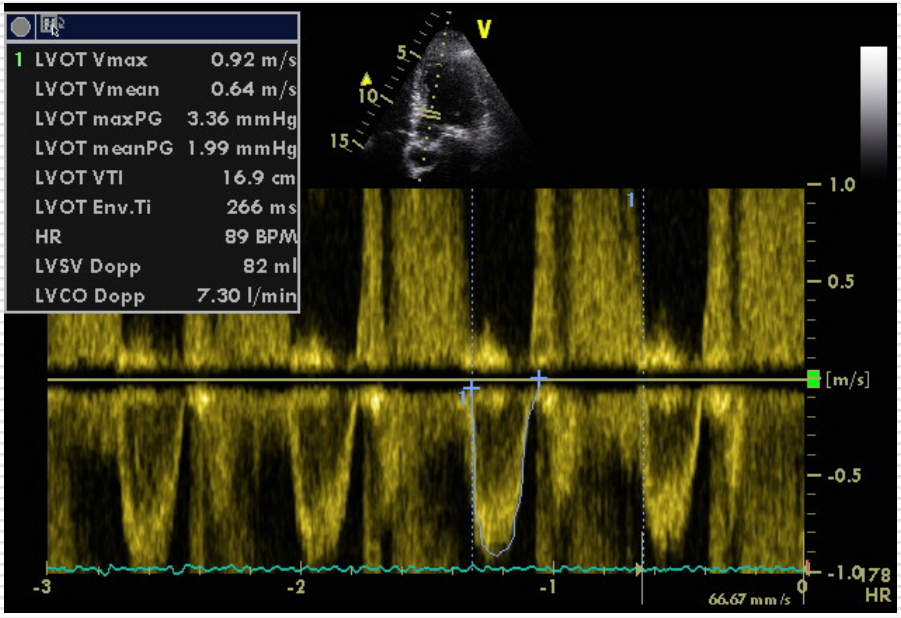
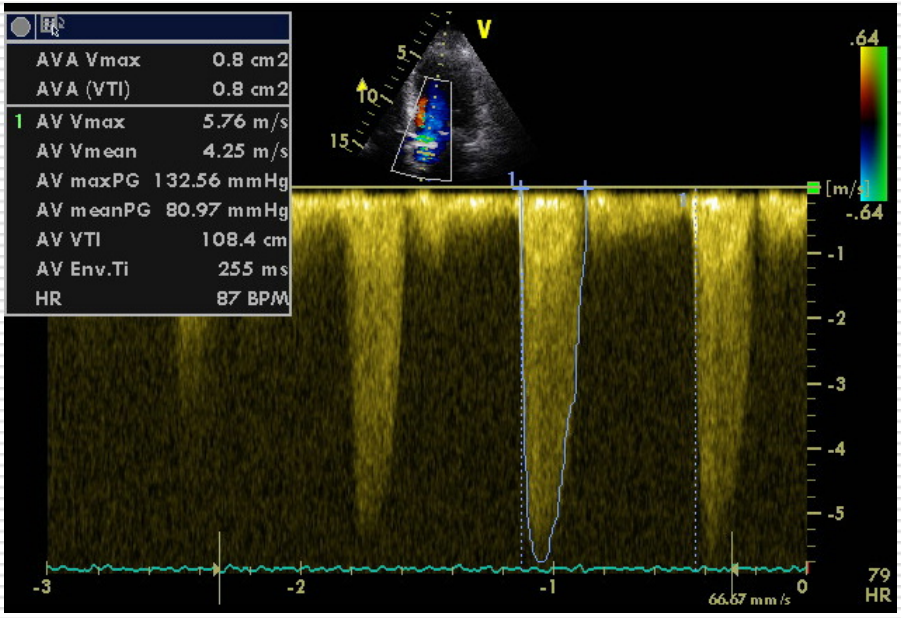
	Class [a]	Level [b]	Ref [c]
PMC is indicated in symptomatic patients with favourable characteristics.[d]	I	B	160, 170
PMC is indicated in symptomatic patients with contraindication or high risk for surgery.	I	C	
PMC should be considered as initial treatment in symptomatic patients with unfavourable anatomy but without unfavourable clinical characteristics.[d]	IIa	C	
PMC should be considered in asymptomatic patients without unfavourable characteristics[d] and • high thromboembolic risk (previous history of embolism, dense spontaneous contrast in the left atrium, recent or paroxysmal atrial fibrillation) and/or • high risk of haemodynamic decompensation (systolic pulmonary pressure >50 mmHg at rest, need for major non-cardiac surgery, desire for pregnancy).	IIa	C	



AORTÁLNÍ STENÓZA





Aortální stenóza

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION
American Heart Association

Determinants and Prognostic Significance of Exercise Pulmonary Hypertension in Asymptomatic Severe Aortic Stenosis

Patrizio Lancellotti, Julien Magne, Erwan Donal, Kim O'Connor, Raluca Dulgheru, Monica Rosca and Luc A. Pierard

Circulation. 2012;126:851-859; originally published online July 25, 2012;
doi: 10.1161/CIRCULATIONAHA.111.088427
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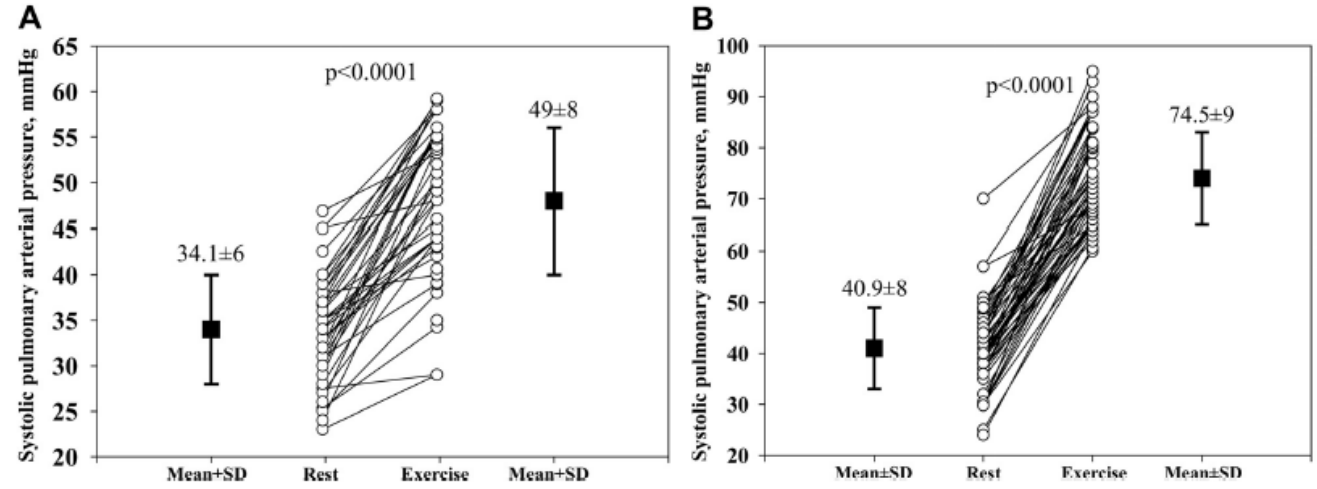

Figure 1. Impact of exercise on systolic pulmonary arterial pressure in patients without exercise pulmonary hypertension (**A**) and with exercise pulmonary hypertension (**B**).

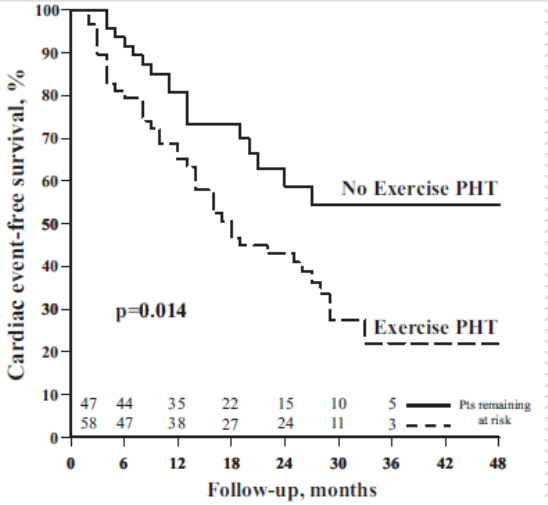

Figure 2. Cardiac event-free survival according to the presence or absence of exercise pulmonary hypertension (PHT).

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION
American Heart Association

Mitrální regurgitace

Exercise Pulmonary Hypertension in Asymptomatic Degenerative Mitral Regurgitation
Julien Magne, Patrizio Lancellotti and Luc A. Piérard

Circulation. 2010;122:33-41; originally published online June 21, 2010;
doi: 10.1161/CIRCULATIONAHA.110.938241
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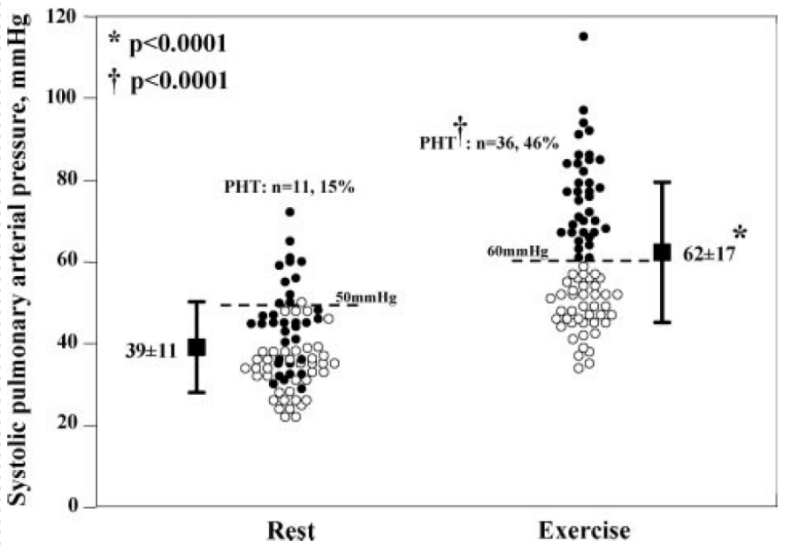


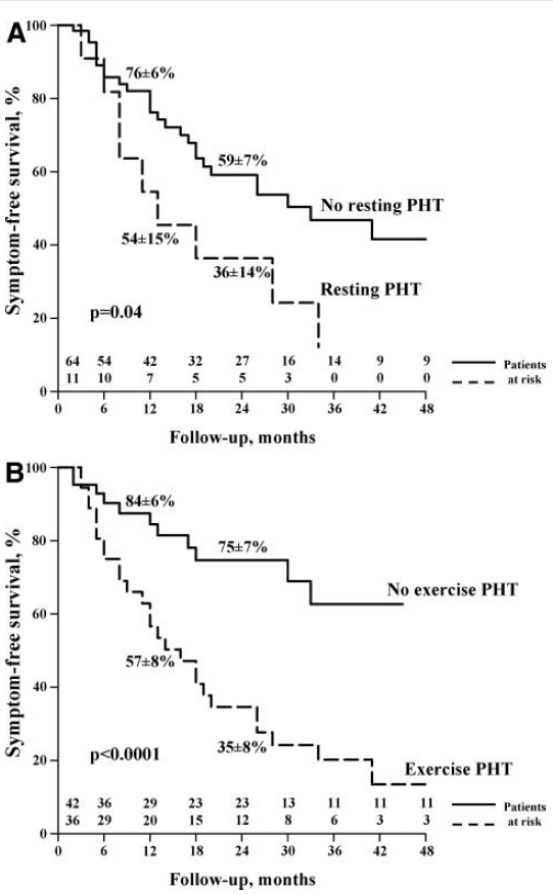
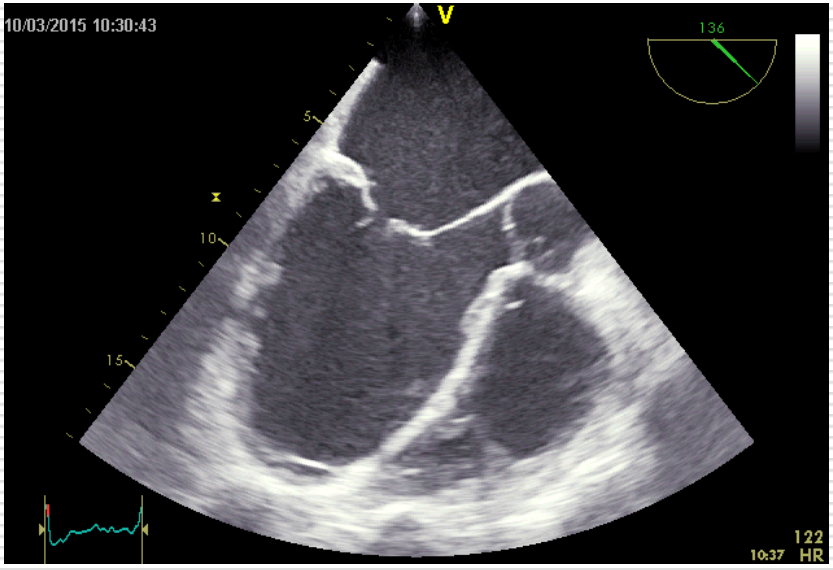
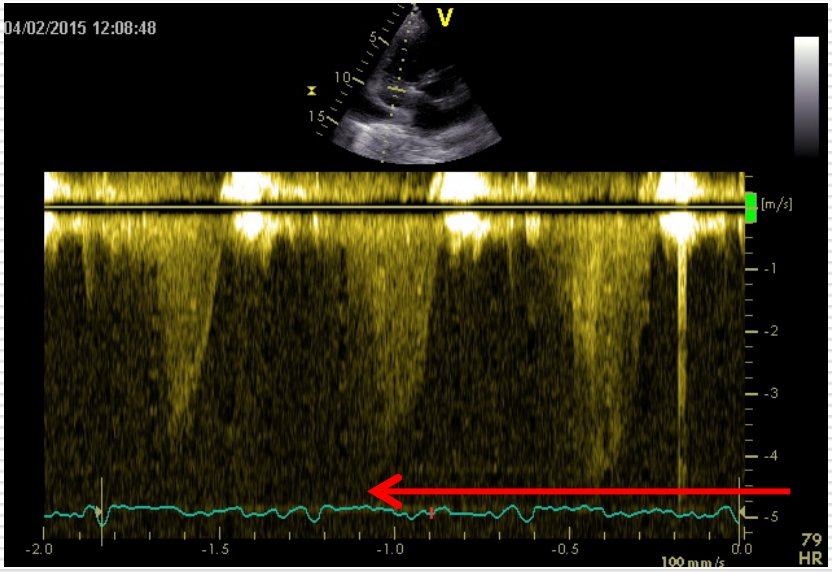


Figure 2. Symptom-free survival according to resting (A) and exercise (B) PHT.

KAZUISTIKA 5





NO: Pacient přijat na doporučení poradny pro srdeční pro globální srdeční dekompenzaci. Postupně zvětšení objemu břicha, nárůst hmotnosti. Námahová dušnost NYHA III, progrese, snížená tolerance námahy - chůze po rovince již s potížemi, musí se zastavovat. Bolesti na hrudi nemá. Ambulantně navýšena diuretika, přechodně během víkendu nastolena negativní bilance tekutin, na hmotnosti ale bez větší redukce (nemá ale doma příliš přesnou váhu)... Nové potíže nejsou.

OA: Arteriální hypertenze, stp. hepatitidě v mládí. Kardiogenní šok, UPV, vaECMO 11.7. STEMI/Q přední stěny, delay 2-3 hodiny, koronarograficky uzávěr prox. RIA, stenosa ostia RMS1 80%, kolat. uzávěr prox. ACD. Komplikováno AVB III. st. s nutností dočasné stimulace a UPV. AV ECMO femoro-femorální jako mech.podpora při kardiogenním šoku do 17.7. Revize art. femoralis bilat a vena femoralis l.dx, st.p.embolectomiitr. tibiofibularis l.sin 18.8., MODS - plíce, ledviny (myoglobimémie), játra, koagulace

Pravostranná srdeční katetrizace 1

Měřené hodnoty: TK arteriální 120/75 mmHg, P 76/min., SR, sO2 98%

PS: 21 mmHg (střední)

PK: 71/23 mmHg (systolický/diastolický)

AP: 70/34/48 mmHg (systolický/diastolický/střední)

PAWP: 30 mmHg (střední)

PVR: 5,3 WU

CO: 3,4 l/min.

CI: 1,5 l/min./m2

Závěr: Těžká čistě postkapilární plicní hypertenze, atributy hypervolémie, snížený pravostranný srdeční výdej, vysoká plicní vaskulární rezistence.

Dop.: Registrace hmostnosti, současně navýšení dávky furosemidu, zítra měření hmotnosti opět. Pokud nedojde k výraznější redukci tělesné hmotnosti, tak zítra CRRT s UF, kontrolní hemodynamika v pondělí a transfer ke zvážení LVAD/HTx.

Pravostranná srdeční katetrizace - kontroní vyšetření při "suché" váze po CRRT:

Měřené hodnoty: TK arteriální 120/80 mmHg, P 72/min., SR, sO2 98%

PS: 6 mmHg (střední)

PK: 60/7 mmHg (systolický/diastolický)

AP: 59/21/36 mmHg (systolický/diastolický/střední)

PAWP: 23 mmHg

PVR: 2,9 WU

CO: 4,49 l/min.

CI: 2,36 l/min./m2

Závěr: Středně těžká čistě postkapilární plicní hypertenze, permisivní hypovolémie, normální pravostranný srdeční výdej, vyšší plicní vaskulární rezistence.

Tekutinové přetížení - bilance

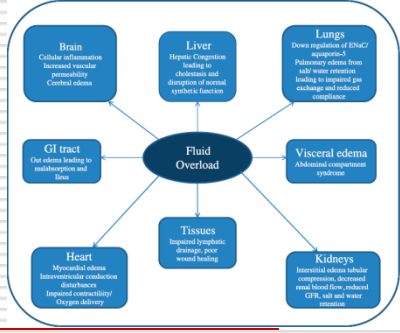
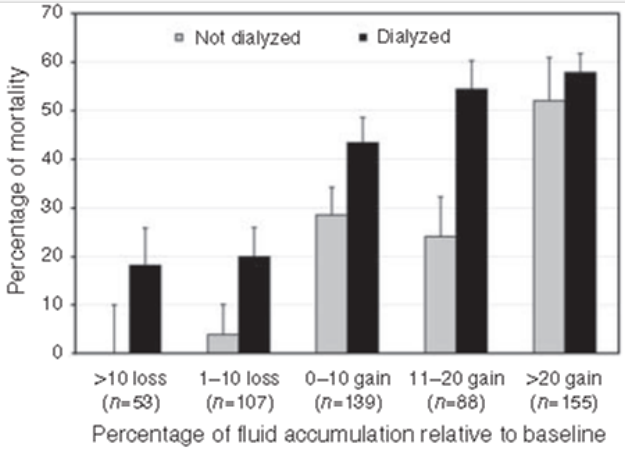


Table 1 Fluid overload at dialysis initiation and its association with mortality

Study	Year	No. of patients	Study design	Percentage of fluid overload		
				Survivors	Nonsurvivors	*P*
Goldstein *et al.* [17]	2001	21[a]	Retrospective observational	16.4 ± 13.8	34.0 ± 21.0	0.03
Foland *et al.* [15]	2004	113[a]	Retrospective observational	7.8 (2.0–16.7)	15.1 (4.9–25.9)	0.02
Gillespie *et al.* [16]	2004	77[a]	Retrospective observational	N/A	N/A	N/A
Michael *et al.* [19]	2004	26[a]	Retrospective interventional	N/A	N/A	N/A
Goldstein *et al.* [18]	2005	116[a]	Prospective observational	14.2 ± 15.9	25.4 ± 32.9	<0.03
Bouchard *et al.* [7]	2009	396[b]	Prospective observational	8.8	14.2	<0.001, 0.01[c]



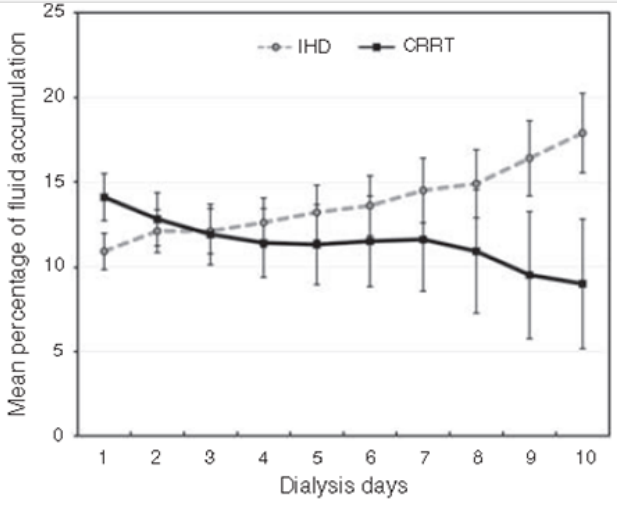


Figure 2 | Mortality rate by final fluid accumulation relative to baseline weight and stratified by dialysis status.

Bouchard et al.: Fluid accumulation, survival and recovery of kidney function in critically ill patients with acute kidney injuryFluid accumulation in acute kidney injury. Kidney International 76, 422-427 (August (2) 2009) | doi:10.1038/ki.2009.159

DOSE studie

❶ 308 pacientů s ADHF randomizovaných do větve bolusové (2x denně i.v.) a kontinuálně aplikované ve standardní dávce (ekvivalentní denní p.o. dávka) a vysoké dávce (2,5x p.o. ekvivalentu).

❷ NS rozdíly v hodnocení symptomů nebo změny kreatininu během 72 h. aplikace furosemidu bolusově/kont. ve standardní/vysoké dávce.

❸ Pacienti léčení vysokou dávkou měli příznivější výsledky sekundárních endpointů (regrese dušnosti, změna hmotnosti a diuréza) za cenu zhoršení renálních funkcí.

Tyto změny neovlivnily 60-denní prognózu…

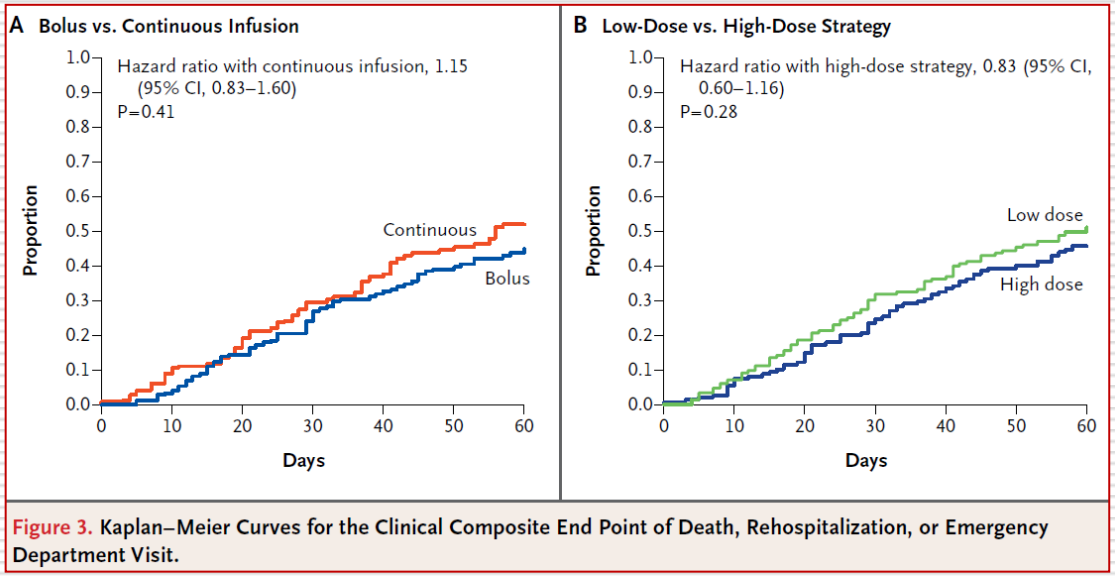
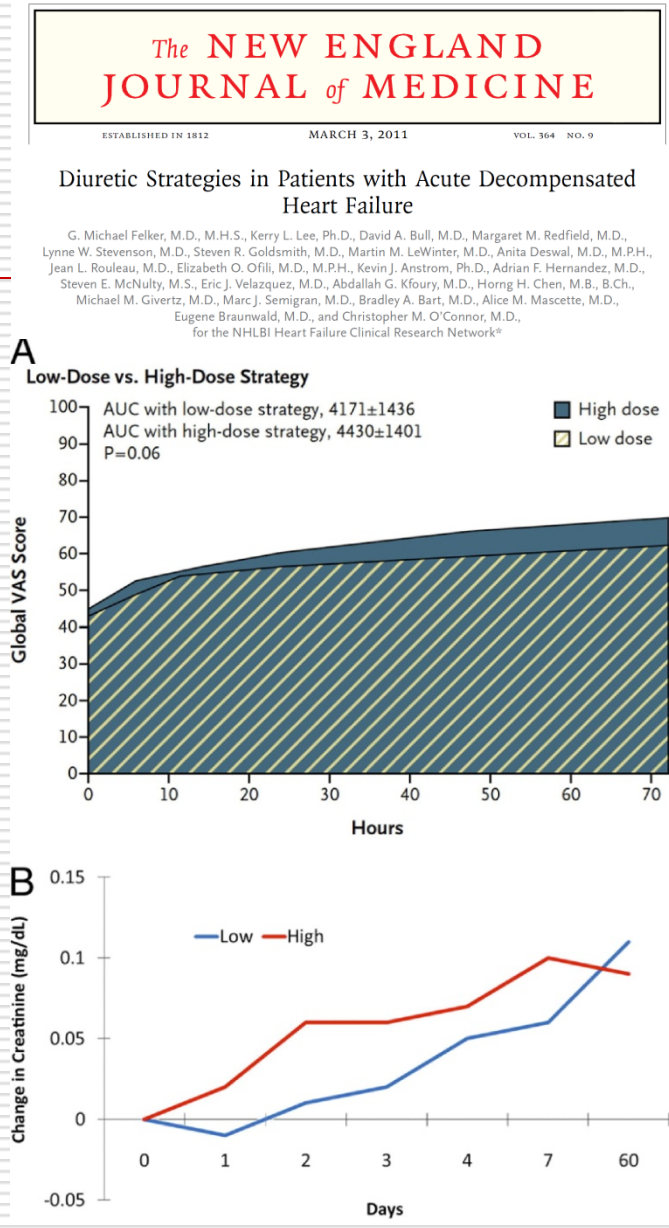


Figure 3. Kaplan–Meier Curves for the Clinical Composite End Point of Death, Rehospitalization, or Emergency Department Visit.



Felker GM, Mentz RJ. JACC 2012, 2145-2153

Ultrafiltrace