PLICNÍ ARTERIÁLNÍ HYPERTENZE

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I. INTERNÍ KLINIKA
KARDIOLOGIE
FAKULTNÍ NEMOCNICE OLOMOUC

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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)

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doi:10.1093/eurheartj/ehv317

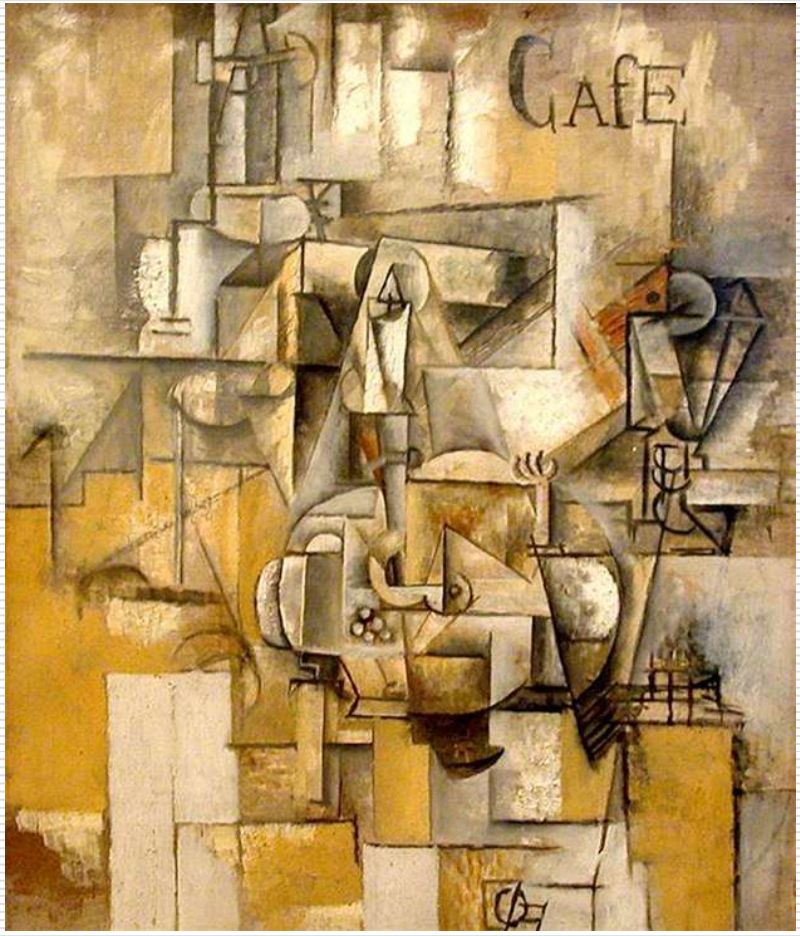
ESC/ERS GUIDELINES

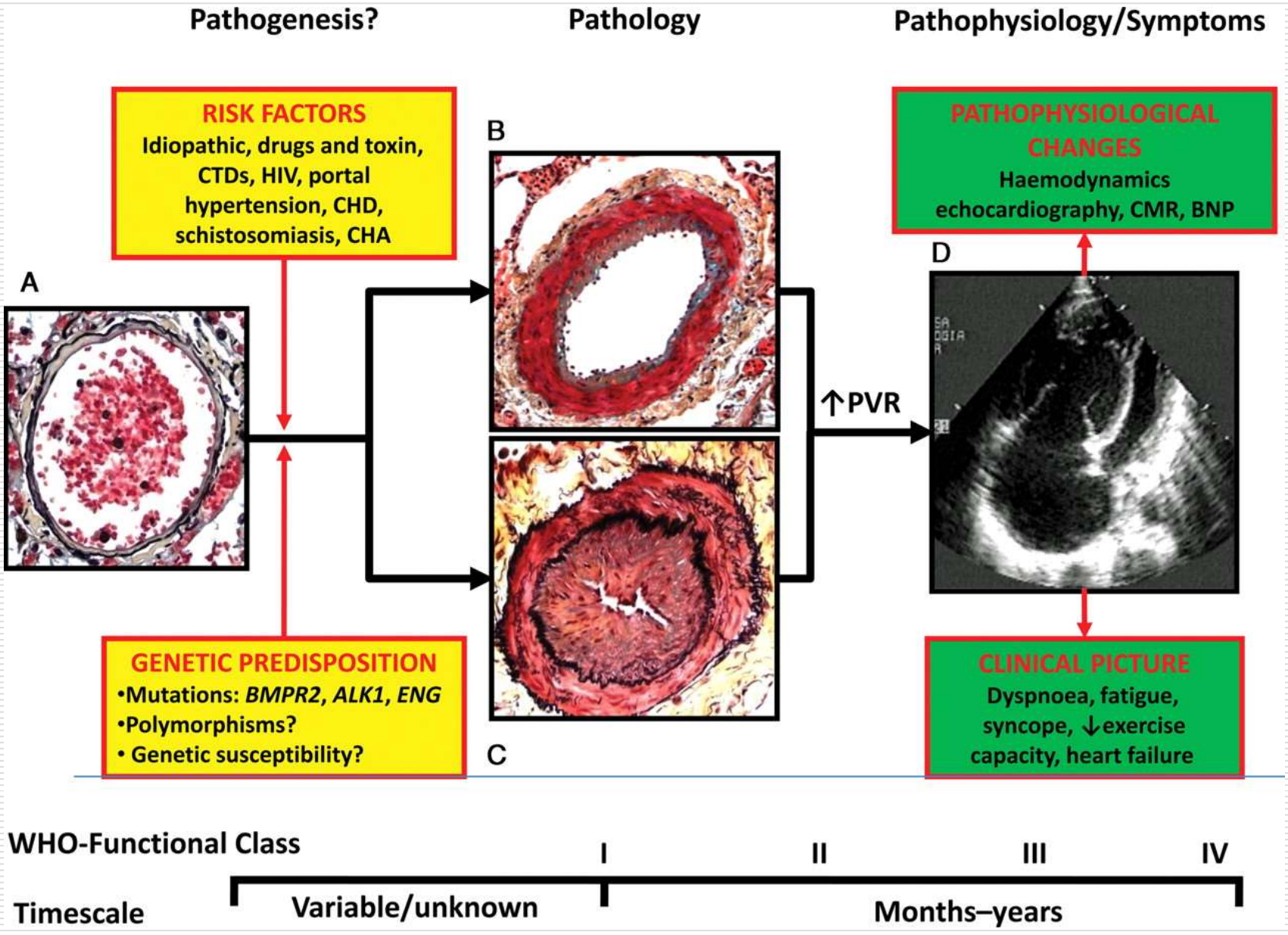
2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

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

I. INTERNÍ KLINIKA
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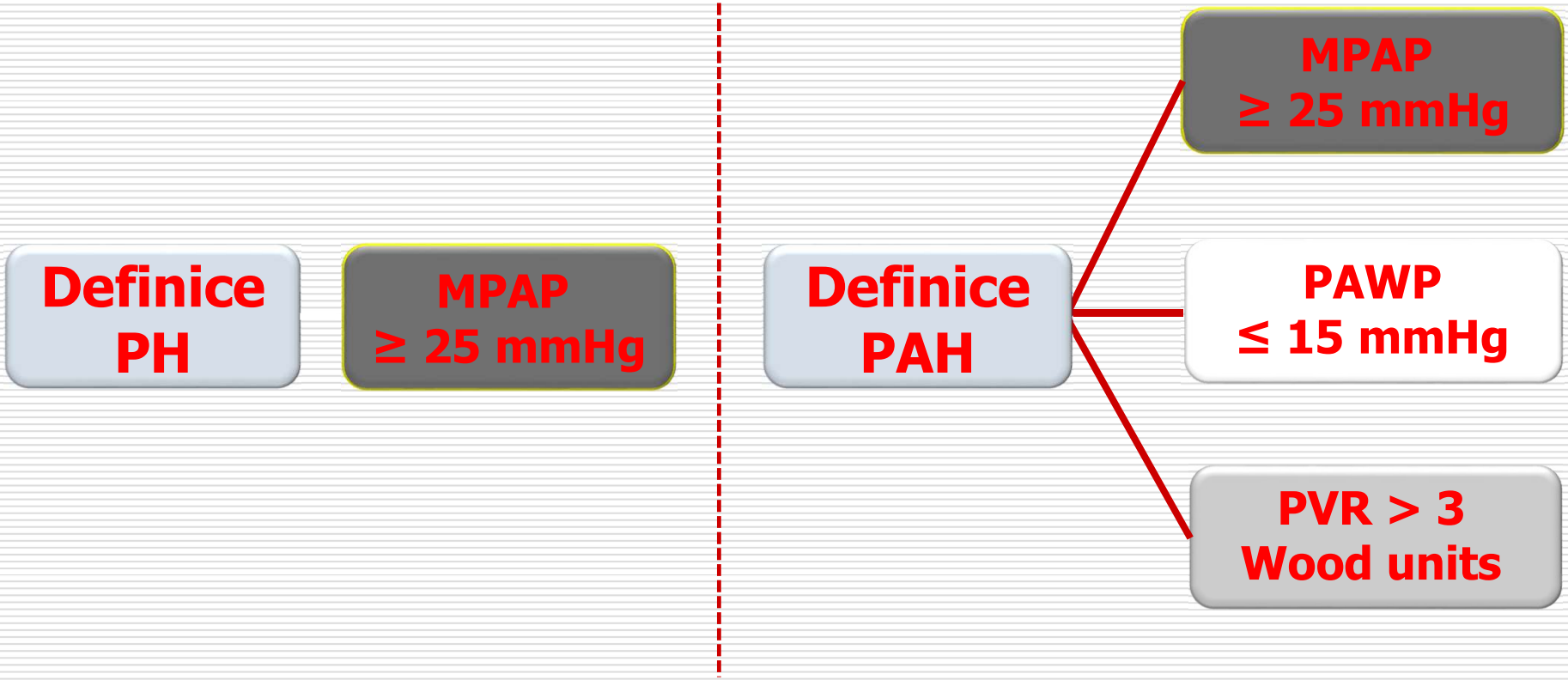
Hemodynamická definice plicní hypertenze



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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David Celermajer, MD, PHD,§ Chris Denton, MD, PHD,‖ Ardeschir Ghofrani, MD,¶
Miguel Angel Gomez Sanchez, MD,# R. Krishna Kumar, MD,** Michael Landzberg, MD,††
Roberto F. Machado, MD,‡‡ Horst Olschewski, MD,§§ Ivan M. Robbins, MD,‖‖
Rogiero Souza, MD, PHD¶¶

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

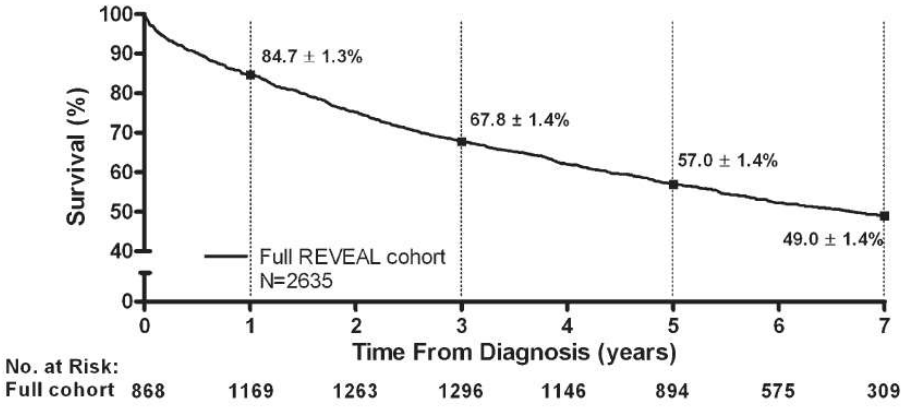


FIGURE 2. Seven-year survival from time of diagnostic right-sided heart catheterization for full REVEAL Registry cohort, using left truncation methods. ■ = estimated survival estimate ± SE at each particular time point. See Figure 1 legend for expansion of abbreviation.

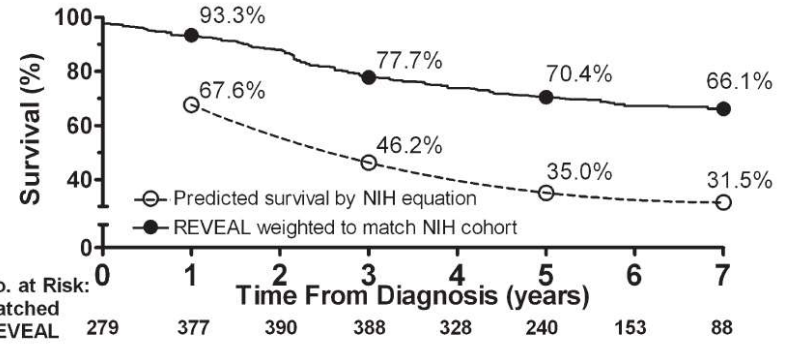


FIGURE 5. Seven-year survival from time of diagnostic RHC of REVEAL Registry cohort weighted to match age, sex, and mean pulmonary artery pressure distribution of NIH cohort. This cohort consisted of patients who met the NIH criteria (ie, had IPAH or FPAH and a pulmonary capillary wedge pressure of ≤12 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

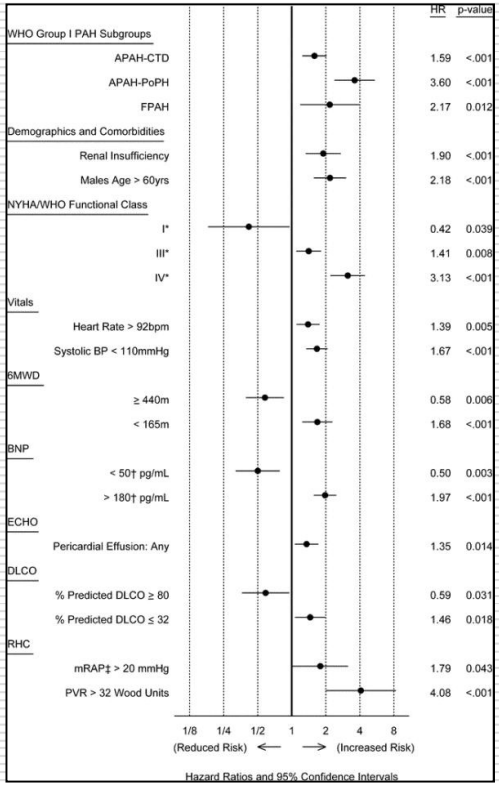
EU/REM/JUL12/248(1). Date of preparation: July 2013.

15 negativních faktorů

4 protektivní faktory

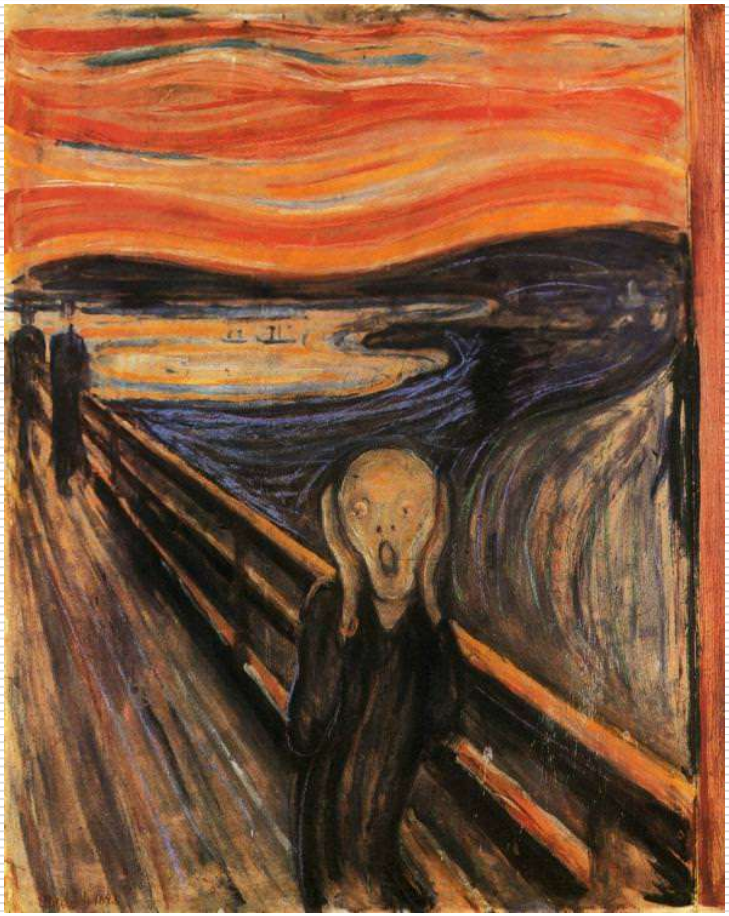
Kalk. rizikové skóre 0-22

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.

DIAGNÓZA



Screening plicní hypertenze

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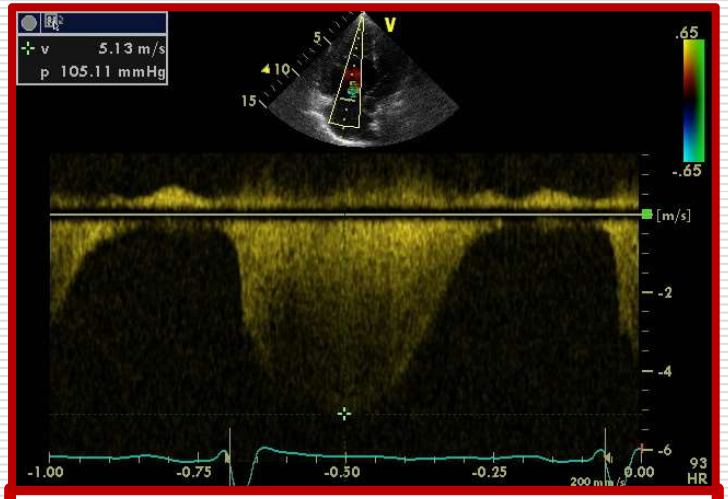
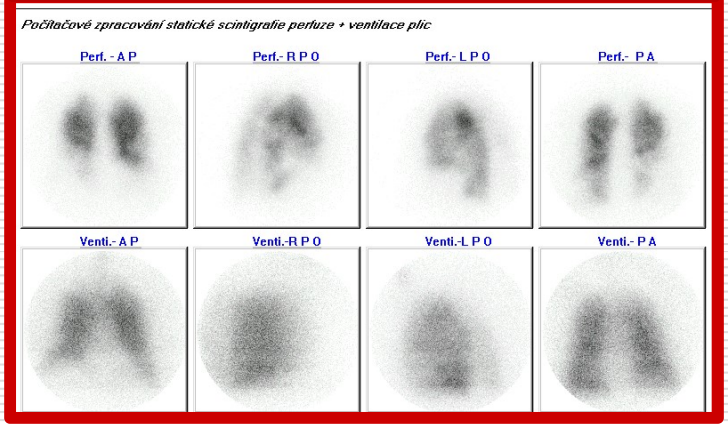
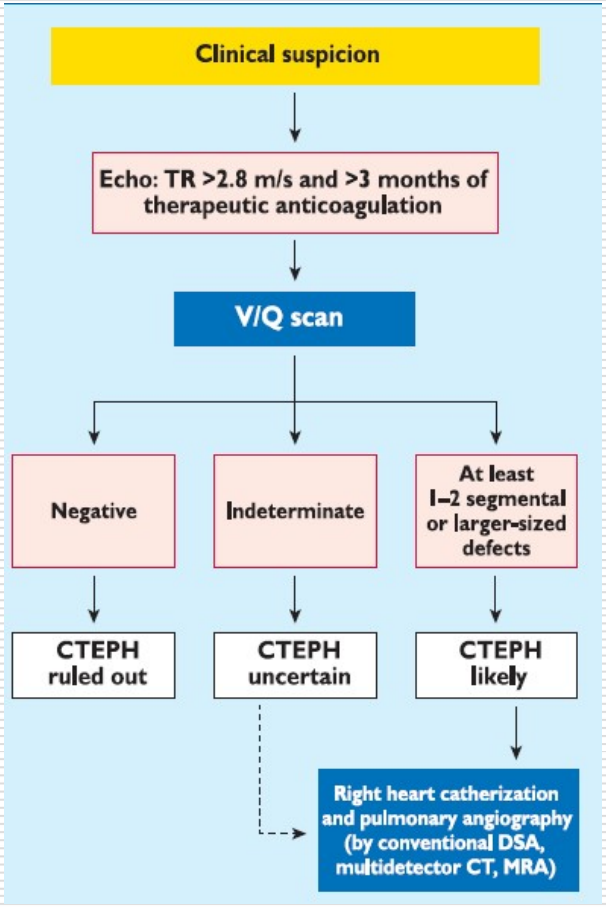
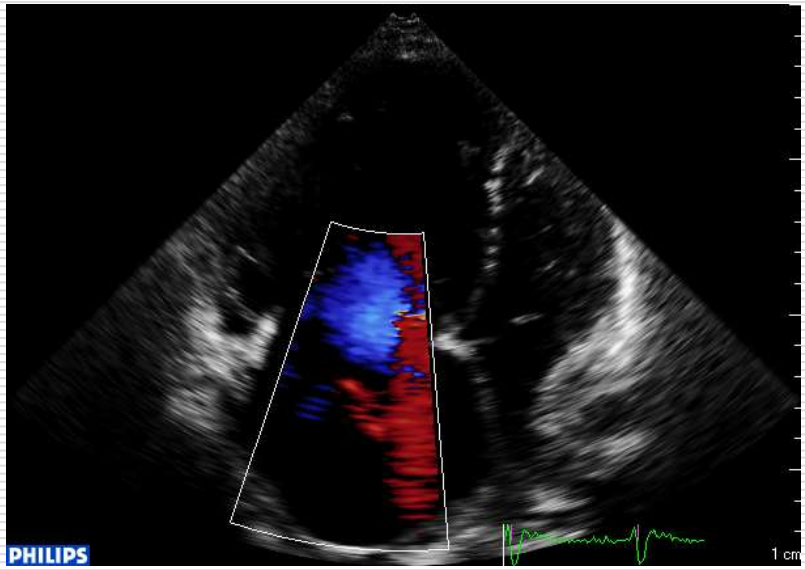
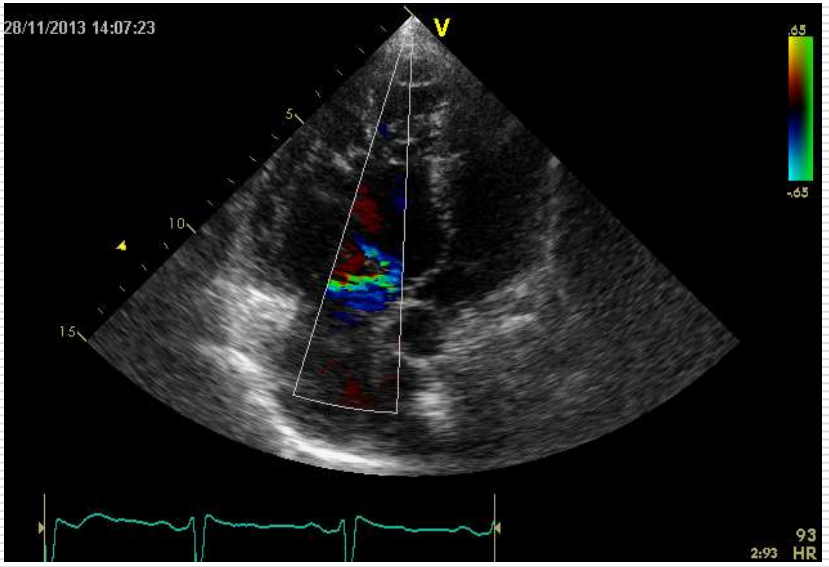


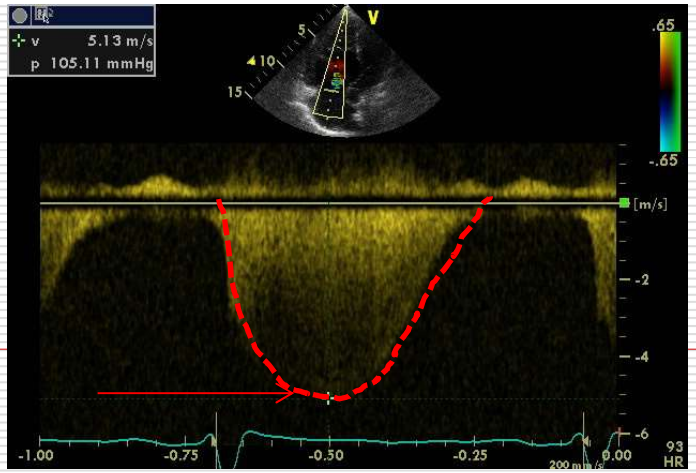




Echokardiografie - klíčové screeningové vyšetření







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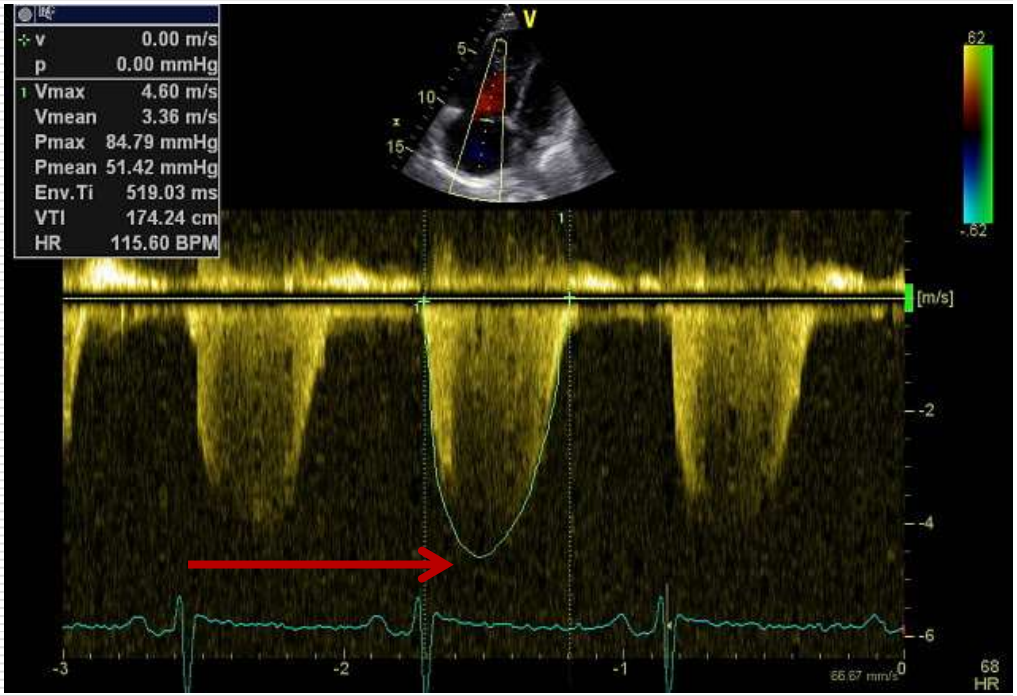
A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<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

European Heart Journal – Cardiovascular Imaging (2015) **16**, 233–271
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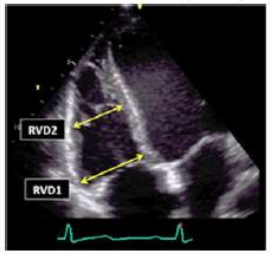
POSITION PAPER

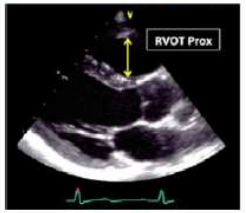
Recommendations for Cardiac Chamber Quantification by Echocardiography in Adults: An Update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging

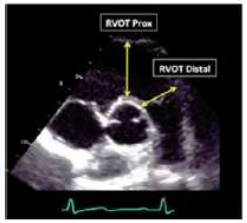
Rozměry DUTINY pravé komory

RV linear dimensions (inflow)*

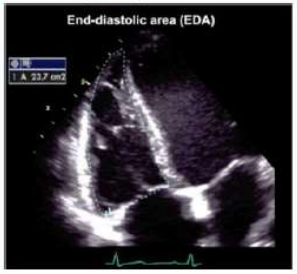


RV linear dimensions (outflow)*

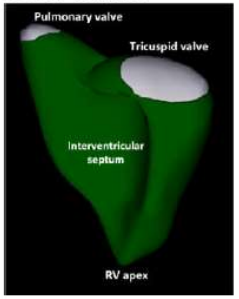




RV areas (inflow)



3DE RV volumes



RV wall thickness

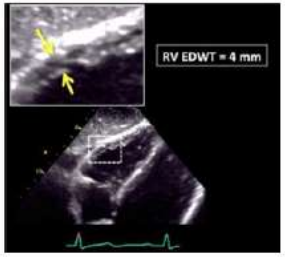
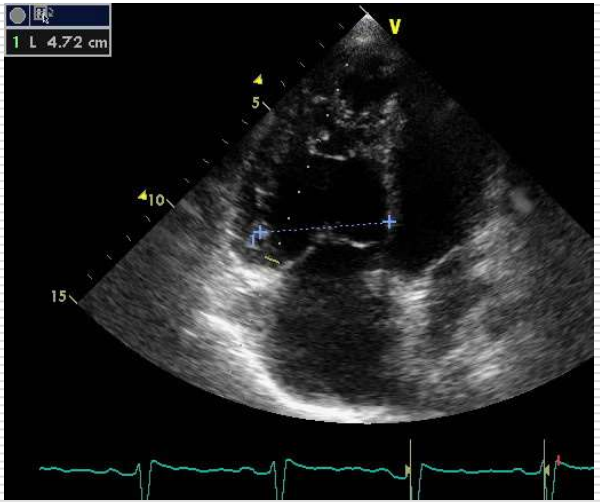


Table 8 Normal values for RV chamber size

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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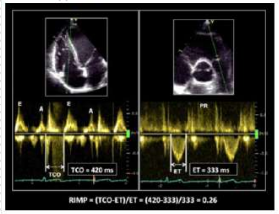
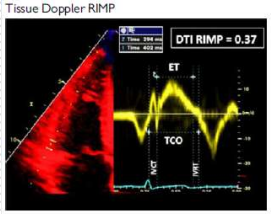
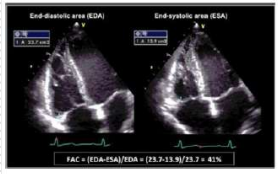
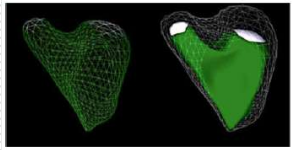
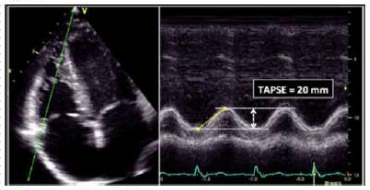
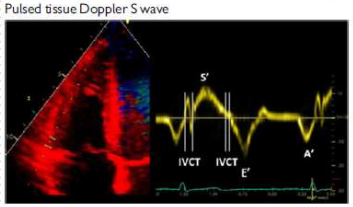
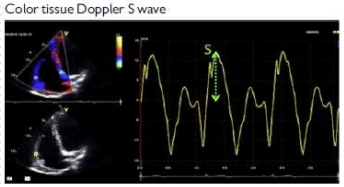
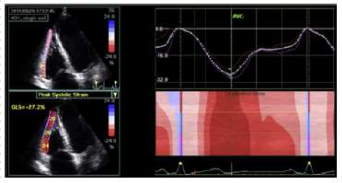
Echocardiographic imaging	Recommended methods	Advantages	Limitations
RV global function Pulsed Doppler RIMP  	RIMP (Tei index) by pulsed Doppler: RIMP = (TCO − ET)/ET	• Prognostic value • Less affected by heart rate	• Requires matching for R-R intervals when measurements are performed on separate recordings • 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'*	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
>3.4	Not required	

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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

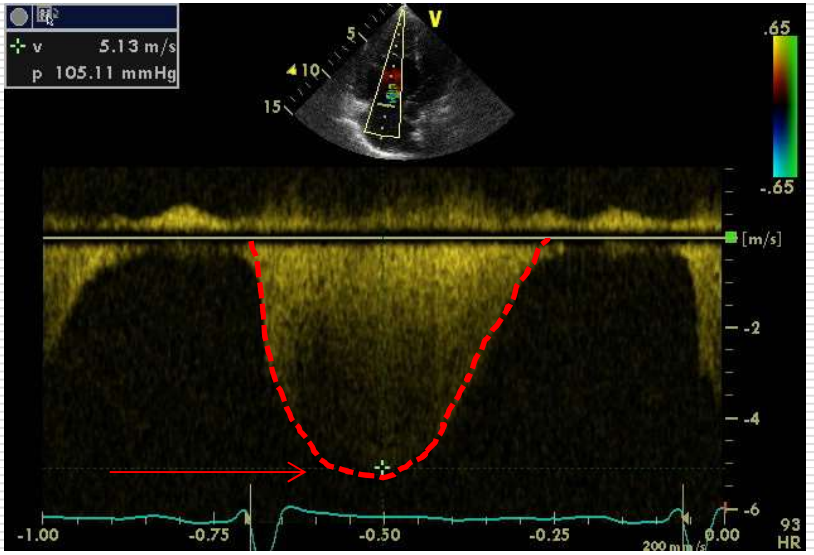
Suspektní hypertrofie, dilatace a/nebo systolická dysfunkce pravé komory

PLICNÍ HYPERTENZE PRAVDĚPODOBNÁ

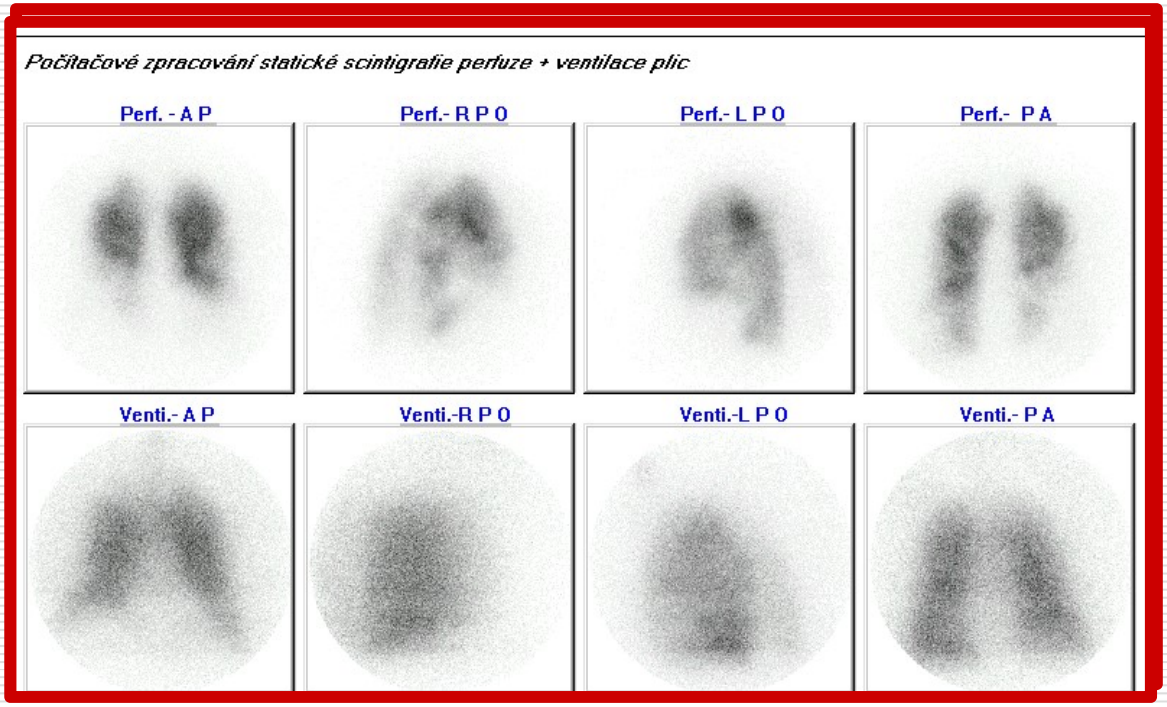
Rychlost trikuspidální regurgitace > 2.8 m/s

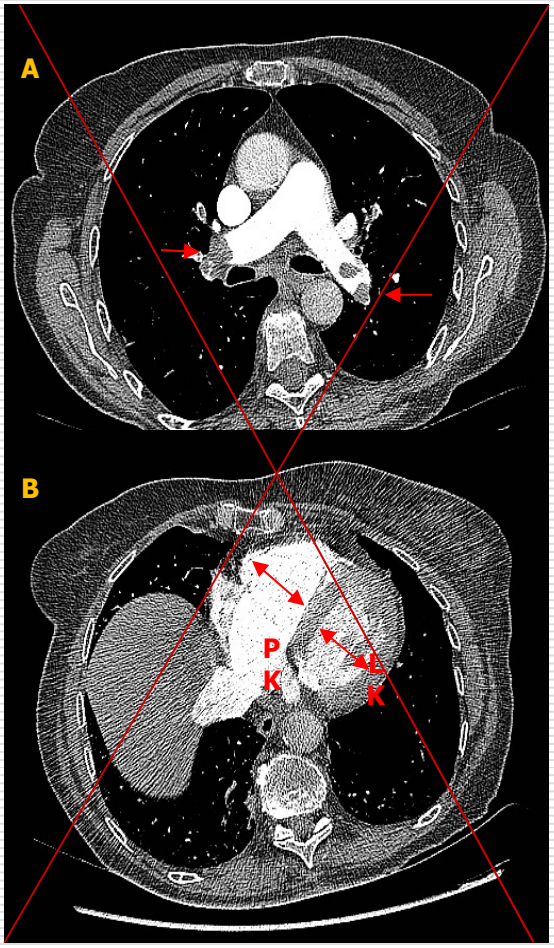
Odhad PASP >36 mmHg

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



Ventilačně-perfuzní scintigrafie plic –
základní screeningové vyšetření CTEPH (nikoliv CTA plicnice)





I. INTERNÍ KLINIKA
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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

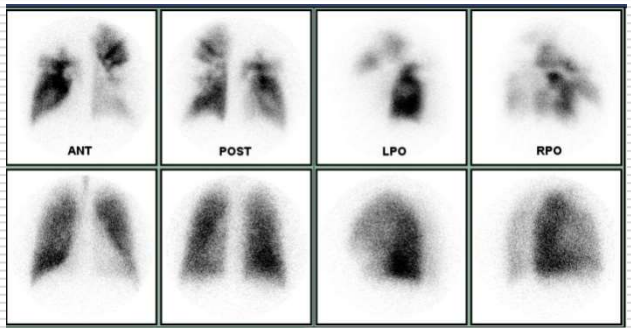

TABLE 1
Summary of V/Q Scans and CTPA Results

	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

Diagnostický algoritmus

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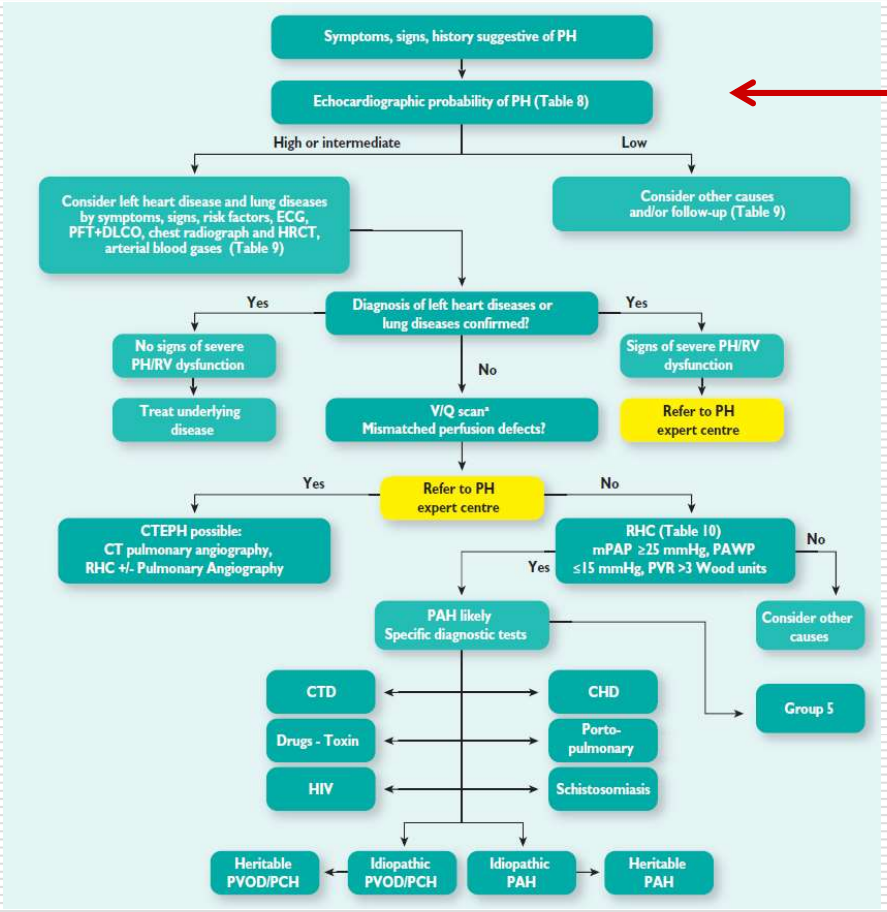
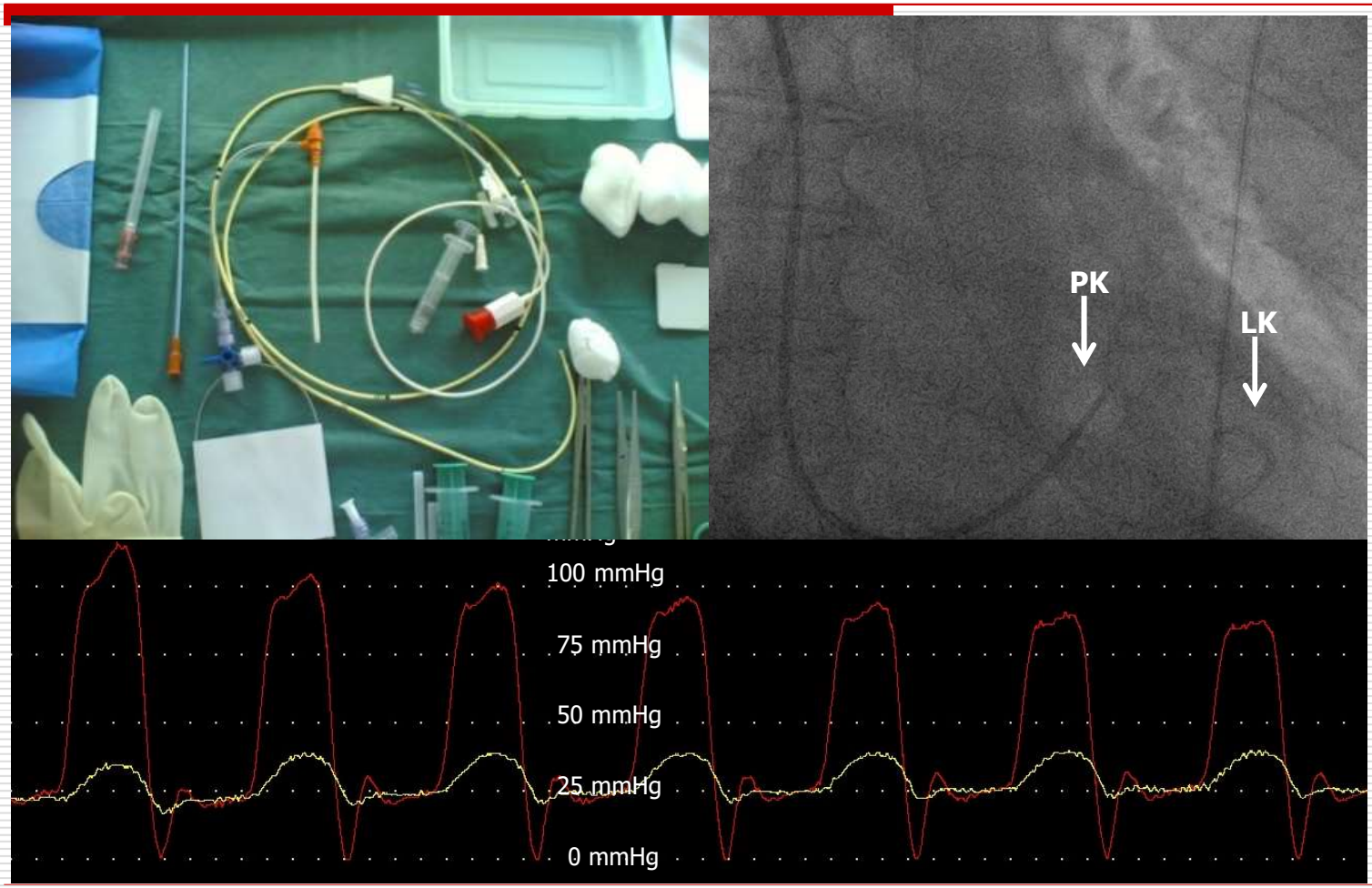


Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'[a]	Echocardiographic probability of pulmonar hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

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

Pravostranná katetrizace - test akutní vazoreaktivity



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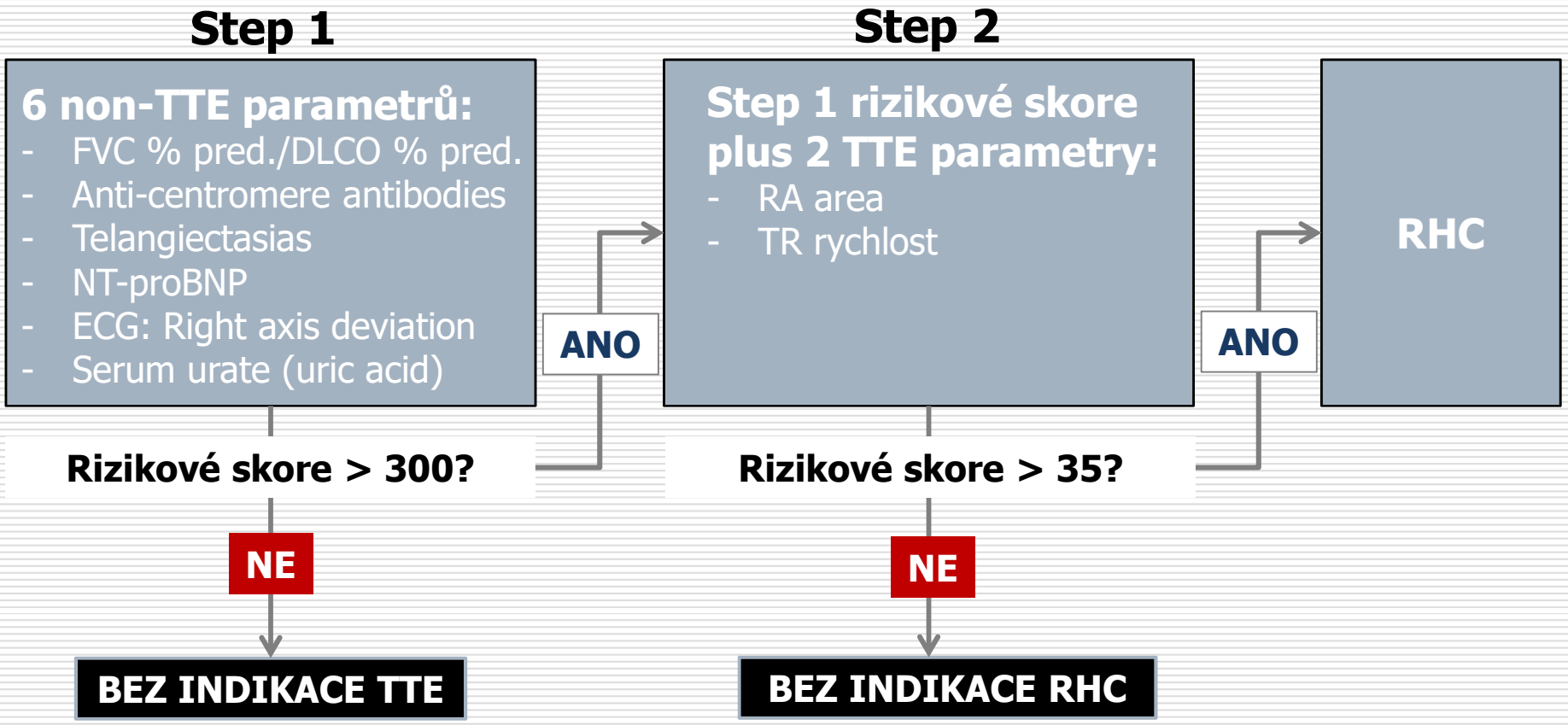
SCREENING

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.

I. INTERNÍ KLINIKA
KARDIOLOGIE
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TERAPIE

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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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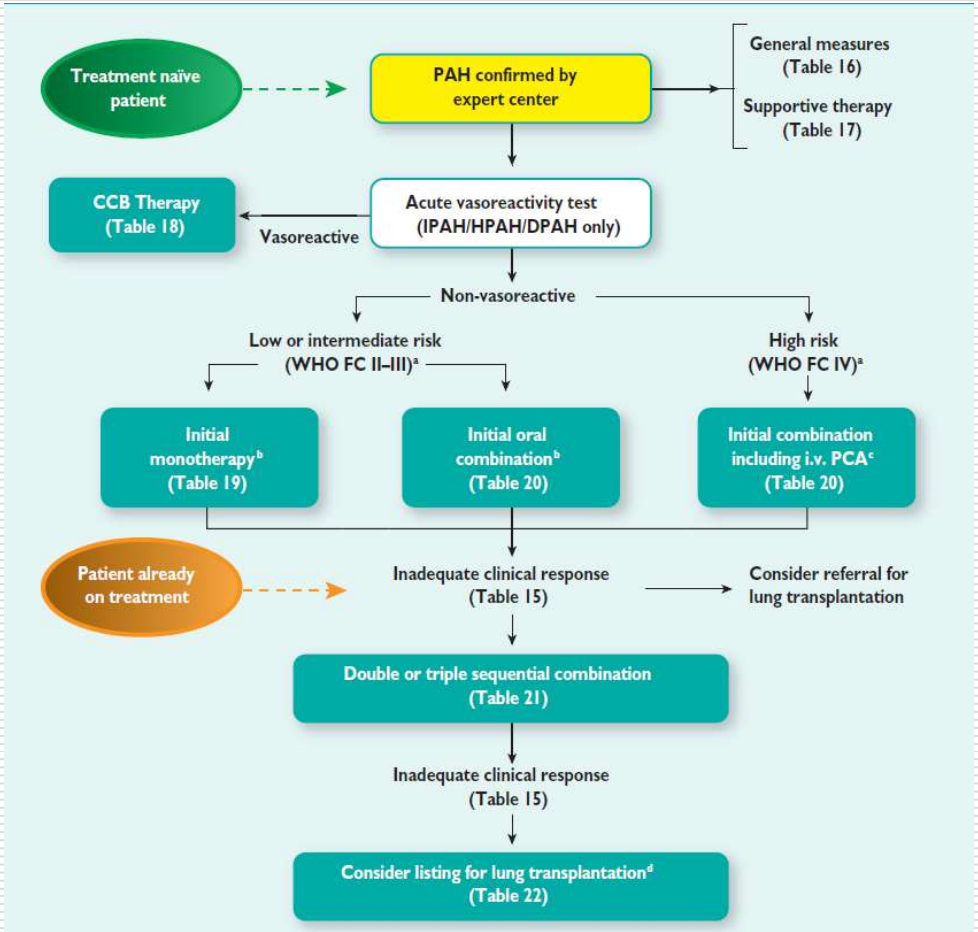

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Treatment Goals of Pulmonary Hypertension

Vallerie V. McLaughlin, MD,* Sean Patrick Gaine, MD, PHD,† Luke S. Howard, DPHIL,‡ Hanno H. Leuchte, MD,§ Michael A. Mathier, MD,‖ Sanjay Mehta, MD,¶ Massimillano Palazzini, MD,# Myung H. Park, MD,** Victor F. Tapson, MD,†† Olivier Sitbon, MD, PHD‡‡

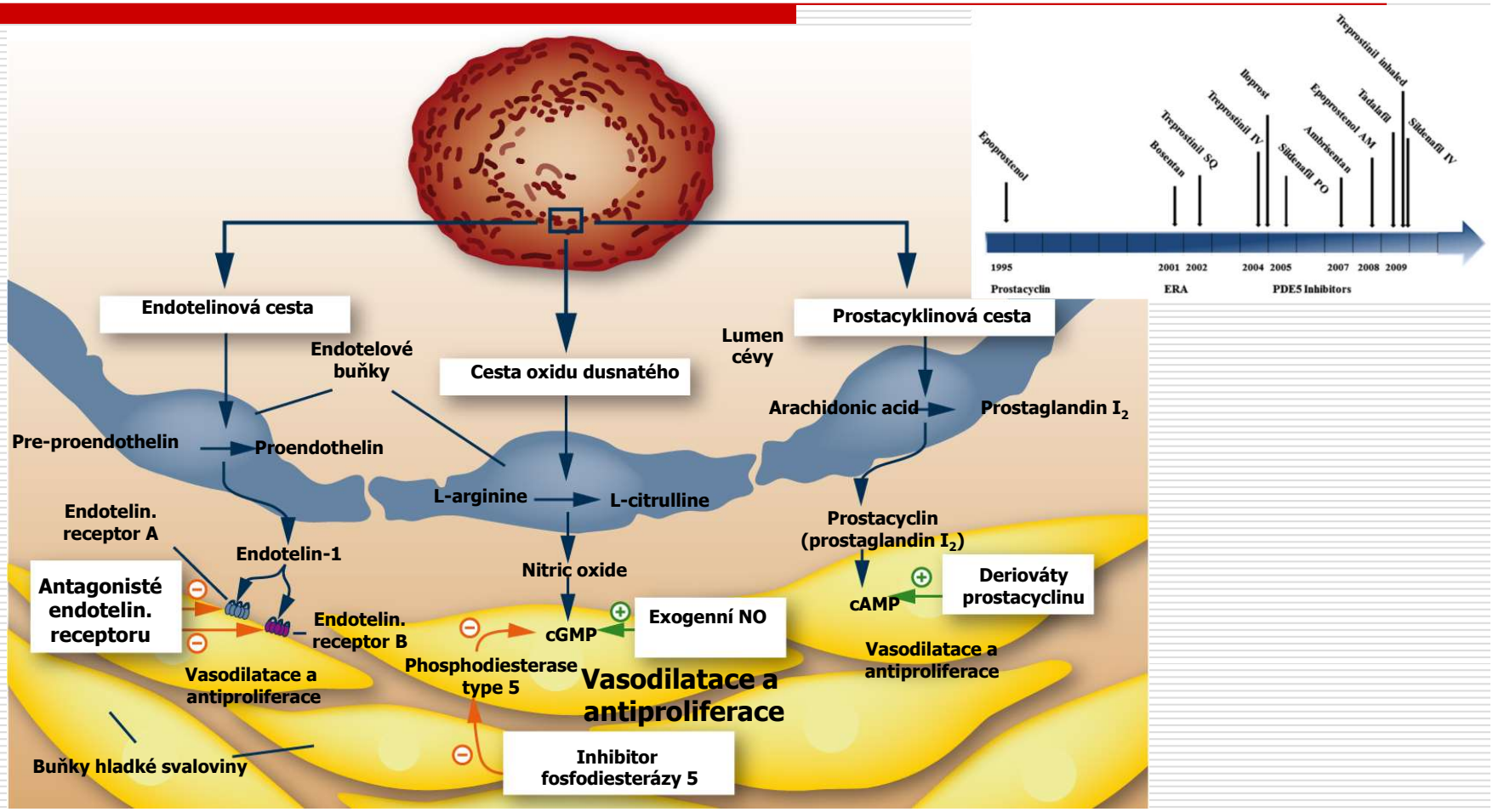
Functional class
I or II
Echocardiography/CMR
Normal/near-normal RV size and function
Hemodynamics
Normalization of RV function (RAP <8 mm Hg and CI >2.5 to 3.0 l/min/m^2)
6-min walk distance
>380 to 440 m; may not be aggressive enough in young individuals
Cardiopulmonary exercise testing
Peak VO_2 >15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

Iniciální

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

Sekvenční

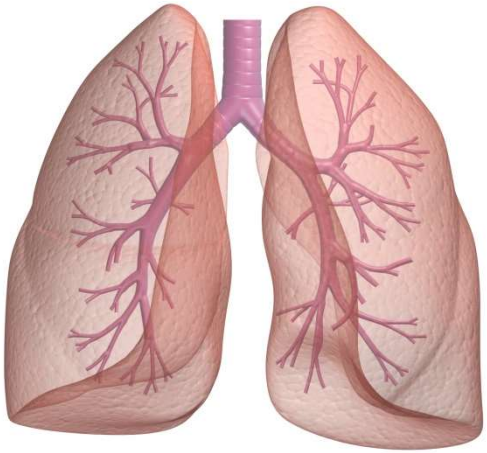
Tadalafil added to bosentan	IIa	C	IIa	C	IIa	C
Ambrisentan added to sildenafil	IIb	C	IIb	C	IIb	C
Bosentan added to epoprostenol	–	–	IIb	C	IIb	C
Bosentan added to sildenafil	IIb	C	IIb	C	IIb	C
Sildenafil added to bosentan	IIb	C	IIb	C	IIb	C
Other double combinations	IIb	C	IIb	C	IIb	C
Other triple combinations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

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)

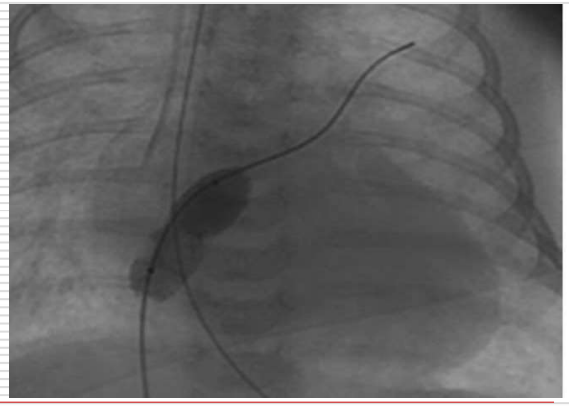
	1 year	5 years	10 years
Pittsburgh (Toyoda et al., 2008 [74])	86	75	66
Paris (Fadel et al., 2010 [75])	79	52	43
Toronto (de Perrot et al., 2012 [76])	78	60	45
Vienna (Klepetko, unpublished data, 2011)	73	71	—



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 %.



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