FAKULTNÍ NEMOCNICE® OLOMOUC

Lékařská fakulta
Univerzita Palackého v Olomouci

KOMPLEXNÍ KARDIOVASKULÁRNÍ CENTRUM
FAKULTNÍ NEMOCNICE OLOMOUC

PLICNÍ ARTERIÁLNÍ HYPERTENZE – definice, klasifikace, pravá komora a zobrazovací metody

Martin Hutyra

ESC European Society of Cardiology
European Heart Journal (2022) **00**, 1–114
https://doi.org/10.1093/eurheartj/ehac237

ESC/ERS GUIDELINES

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

Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS).

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Definition, classification and diagnosis of pulmonary hypertension

Gabor Kovacs[1,2], Sonja Bartolome[3], Christopher P. Denton[4], Michael A. Gatzoulis[5,6], Sue Gu[7], Dinesh Khanna[8], David Badesch[7] and David Montani[9,10,11]

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Corresponding author: Gabor Kovacs (gabor.kovacs@uniklinikum.kages.at)

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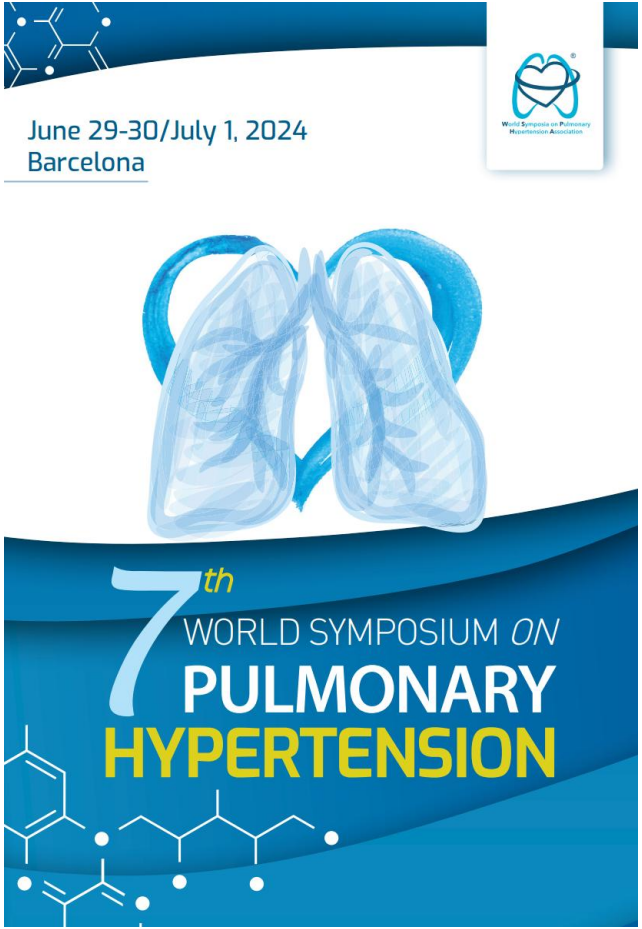
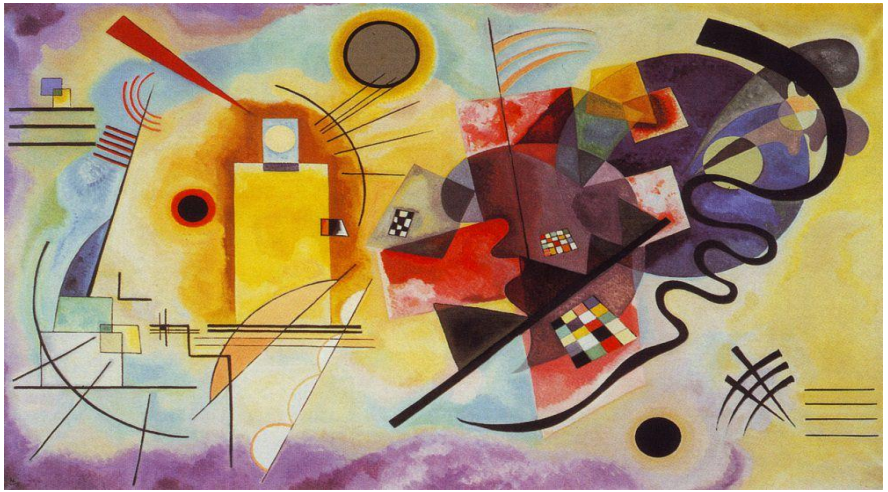
Shareable abstract (@ERSpublications)
In this article, we provide the definitions, the current clinical classification and the diagnostic algorithm for pulmonary hypertension, based on the 7th World Symposium on Pulmonary Hypertension. https://bit.ly/3W442cD

Cite this article as: Kovacs G, Bartolome S, Denton CP, *et al.* Definition, classification and diagnosis of pulmonary hypertension. *Eur Respir J* 2024; in press: 2401324 [DOI: 10.1183/13993003.01324-2024].



DEFINICE



WORLD HEALTH ORGANIZATION

ORGANISATION MONDIALE DE LA SANTE

EXECUTIVE BOARD

Twenty-eighth Session

Provisional agenda item 3.5

EB28/13 ✓
16 May 1961

ORIGINAL: ENGLISH

REPORT ON EXPERT COMMITTEE MEETINGS

Report by the Director-General

1. Introduction

In compliance with paragraph 10.6 of the regulations for expert advisory panels and committees,[1] the Director-General is here reporting on the action to be taken with reference to meetings of Expert Committees.

2. Reports

The reports of expert committee meetings which have been prepared in both working languages, since the twenty-seventh session of the Executive Board and are now available for annexation to this report are the following:

Expert Committee on Antibiotics (Standardization of Methods for Conducting Microbic Sensitivity Tests)

Expert Committee on Chronic Cor Pulmonale

Expert Committee on Professional and Technical Education of Medical and Auxiliary Personnel (Recommended Requirements for Schools of Public Health)

Expert Committee on Public Health Administration (Planning of Public Health Services)

Expert Committee on Specifications for Pharmaceutical Preparations - Sub-Committee on Non-Proprietary Names

Expert Committee on Specifications for Pharmaceutical Preparations

In 1961, a report of the WHO Expert Committee on Chronic Cor Pulmonale mentioned clearly that the mean pulmonary arterial pressure (mPAP) **does not normally exceed 15 mmHg** when the subject is at rest in a lying position, and that the value was **little affected by age** and **never exceeded 20 mmHg**.

CLINICAL PROGRESS

Chronic Cor Pulmonale

Report of an Expert Committee*

Contents

Foreword

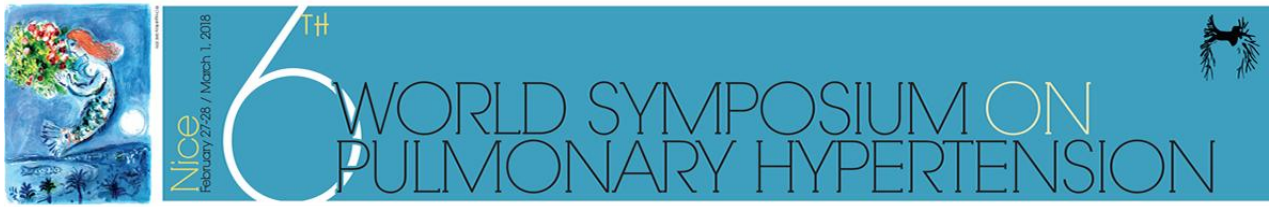
IN THE early months of 1960, the Director-General of the World Health Organization appointed an Expert Committee to inquire into and write a report on the subject of cor pulmonale.

In order to facilitate the preliminary study, as well as the actual deliberations of the Committee, two consultants were appointed, to prepare a survey of the subject: Professor H. Denolin, Chargé de cours á l'Université de Bruxelles; and Dr. C. M. Fletcher, Senior Lecturer in Medicine, Postgraduate Medical School, London. This, a sixty-page report, was put together by Drs. Denolin and Fletcher during July, 1960, and made available to Committee members shortly thereafter.

The membership of the Expert Committee was as follows: Dr. J. Dankmeijer, Professor of Anatomy, Embryology and Physical Anthropology, University of Leiden, the Netherlands; Dr. F. Herles, Professor of Medicine II Internal Clinic, Charles University, Prague, Czechoslovakia; Dr. M. Ibrahim, formerly Professor of Cardiology, Faculty of Medicine, Cairo University, Cairo, Province of Egypt, United Arab Republic; Dr. D. D. Reid, Professor of Epidemiology, Department of Medical Statistics and Epidemiology, London School of Hygiene and Tropical Medicine, London, England; Dr. D. W. Richards, Lambert Professor of Medicine, College of Physicians and Surgeons, Columbia University, New

*Reprinted by permission from the World Health Organization Technical Report Series No. 213. Reprints of the original report may be obtained for $0.30 from World Health Organization, Palais Des Nations, Geneva.

Hemodynamická definice plicní hypertenze



Definice PH — **MPAP ≥20/25 mmHg**

Definice PAH — **MPAP ≥20/25 mmHg**; **PAWP ≤15 mmHg**; **PVR >3 WU**

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.
6th World Symposium on Pulmonary Hypertension, Nice, February 27 to March 1, 2018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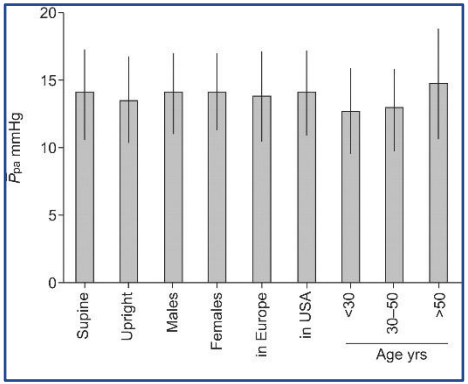
Definition	Haemodynamic characteristics
PH	**mPAP >20 mmHg**
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg **PVR >2 WU**
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg **PVR ≤2 WU**
Combined post- and pre-capillary PH	mPAP >20 mmHg PAWP >15 mmHg **PVR >2 WU**
Exercise PH	**mPAP/CO slope between rest and exercise >3 mmHg/L/min**

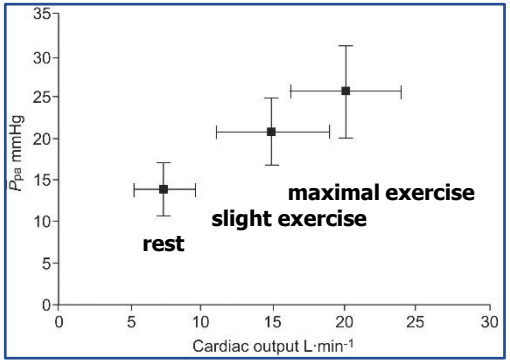
Pulmonary arterial pressure during rest and exercise in healthy subjects: a systematic review

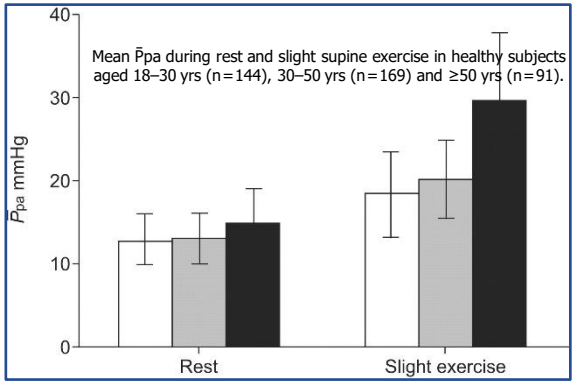
G. Kovacs*, A. Berghold#, S. Scheidl* and H. Olschewski*

Co je horní hranice normálního tlaku v plicnici?

RHC studies in healthy individuals to determine **normal values of mPAP** at rest and exercise. Data from 1187 normal subjects from 47 studies were analysed. mPAP at rest was 14.0±3.3 mmHg; this value was independent of sex and ethnicity, and was only slightly influenced by age and posture.



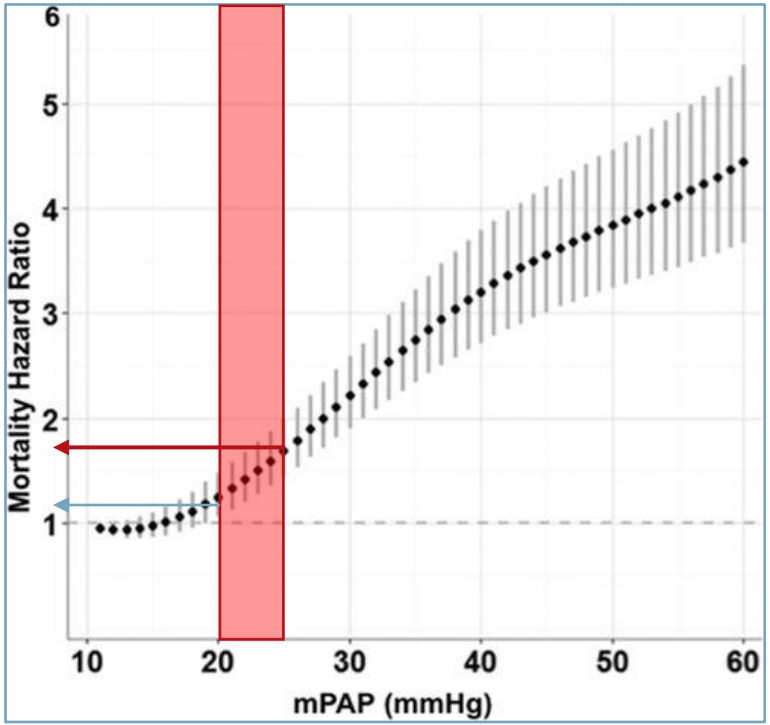


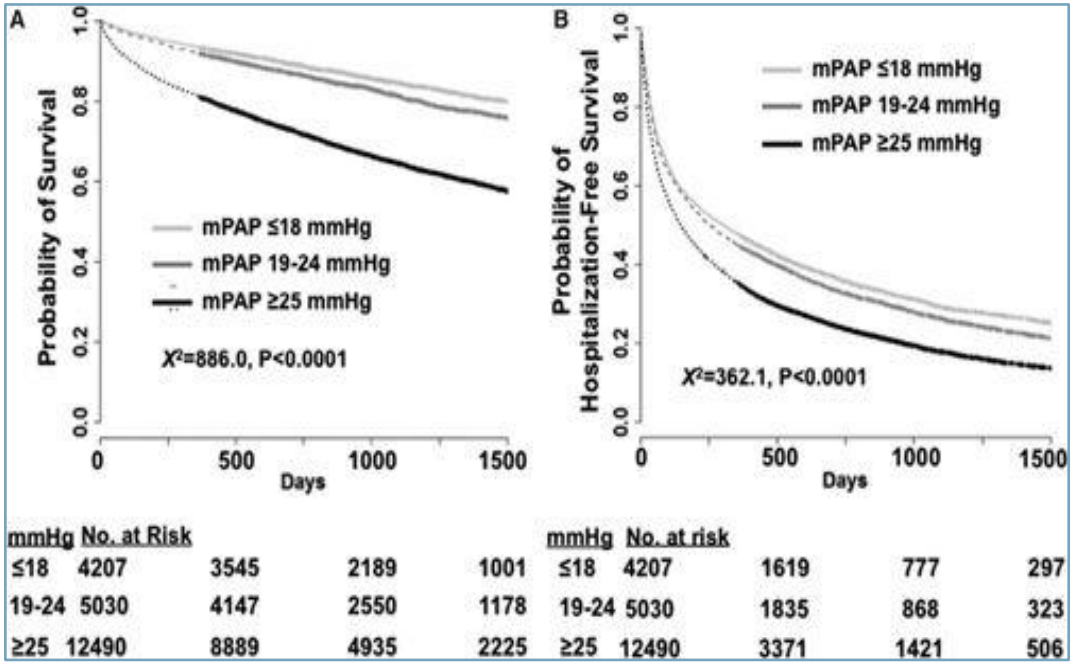


Considering this mPAP of 14 mmHg, two standard deviations would suggest mPAP >20 mmHg as above the upper limit of normal (i.e. above the 97.5th percentile).
This definition is, therefore, no longer arbitrary, but based on a scientific approach.

G. Kovacs et al. Eur Respir J 2009;34:888-894

Má hraniční mPAP vliv na prognózu?



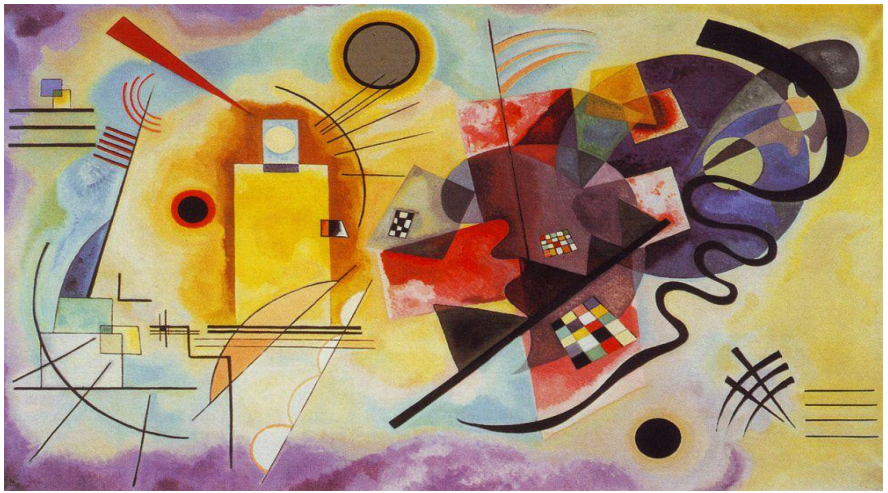


mmHg	No. at Risk			
≤18	4207	3545	2189	1001
19-24	5030	4147	2550	1178
≥25	12490	8889	4935	2225

mmHg	No. at risk			
≤18	4207	1619	777	297
19-24	5030	1835	868	323
≥25	12490	3371	1421	506

Bradley A. Maron. Circulation. Association of Borderline Pulmonary Hypertension With Mortality and Hospitalization in a Large Patient Cohort: Insights From the Veterans Affairs Clinical Assessment, Reporting, and Tracking Program, Volume: 133, Issue: 13, Pages: 1240-1248, DOI: (10.1161/CIRCULATIONAHA.115.020207)

KLASIFIKACE



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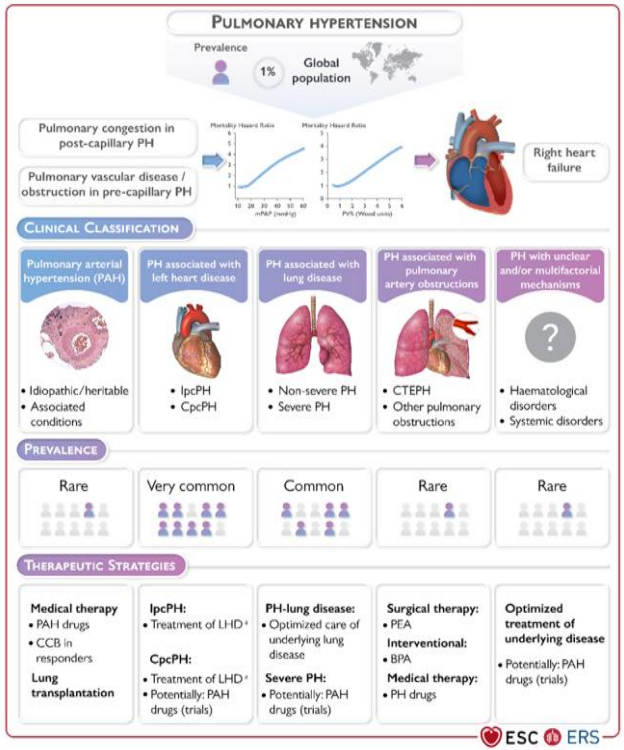

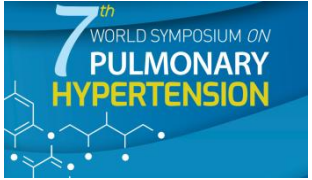

Group 1: PAH
1.1 Idiopathic
1.1.1 Long-term responders to calcium channel blockers
1.2 Heritable[#]
1.3 Associated with drugs and toxins[#]
1.4 Associated with:
1.4.1 connective tissue disease
1.4.2 HIV infection
1.4.3 portal hypertension
1.4.4 congenital heart disease
1.4.5 schistosomiasis
1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
1.6 Persistent PH of the newborn
Group 2: PH associated with left heart disease
2.1 Heart failure:
2.1.1 with preserved ejection fraction
2.1.2 with reduced or mildly reduced ejection fraction
2.1.3 cardiomyopathies with specific aetiologies[¶]
2.2 Valvular heart disease:
2.2.1 aortic valve disease
2.2.2 mitral valve disease
2.2.3 mixed valvular disease
2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH
Group 3: PH associated with lung diseases and/or hypoxia
3.1 COPD and/or emphysema
3.2 Interstitial lung disease
3.3 Combined pulmonary fibrosis and emphysema
3.4 Other parenchymal lung diseases[*]
3.5 Nonparenchymal restrictive diseases:
3.5.1 hypoventilation syndromes
3.5.2 pneumonectomy
3.6 Hypoxia without lung disease (*e.g.* high altitude)
3.7 Developmental lung diseases
Group 4: PH associated with pulmonary artery obstructions
4.1 Chronic thromboembolic PH
4.2 Other pulmonary artery obstructions[§]
Group 5: PH with unclear and/or multifactorial mechanisms
5.1 Haematological disorders[f]
5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis and neurofibromatosis type 1
5.3 Metabolic disorders[##]
5.4 Chronic renal failure with or without haemodialysis
5.5 Pulmonary tumour thrombotic microangiopathy
5.6 Fibrosing mediastinitis
5.7 Complex congenital heart disease

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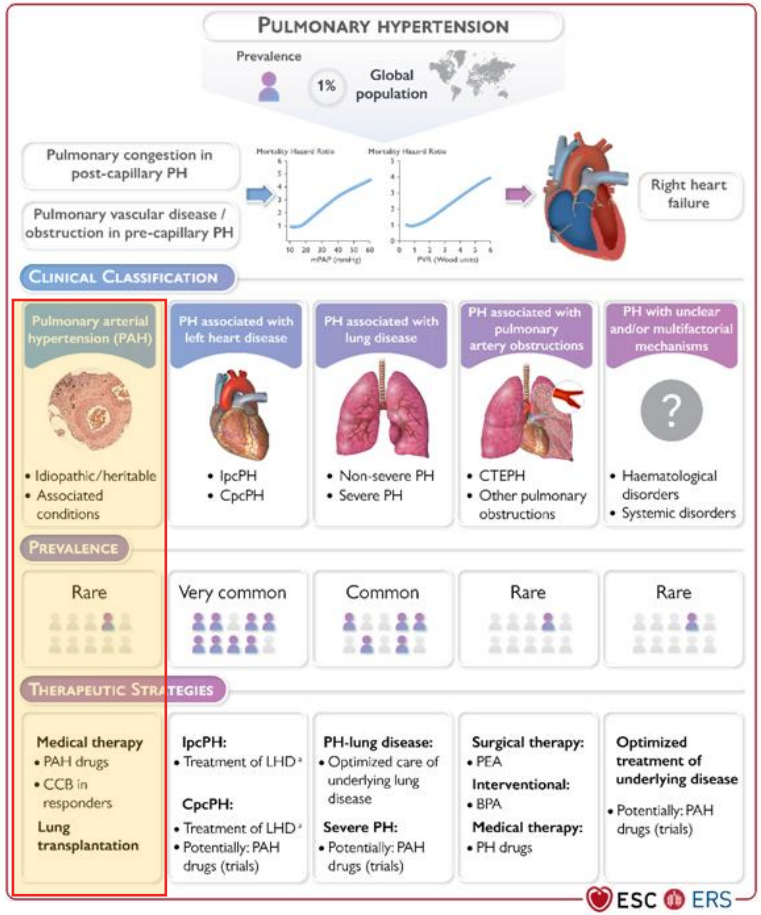
GROUP 1 Pulmonary arterial hypertension (PAH)
1.1 Idiopathic
1.1.1 Non-responders at vasoreactivity testing
1.1.2 Acute responders at vasoreactivity testing
1.2 Heritable[a]
1.3 Associated with drugs and toxins[a]
1.4 Associated with:
1.4.1 Connective tissue disease
1.4.2 HIV infection
1.4.3 Portal hypertension
1.4.4 Congenital heart disease
1.4.5 Schistosomiasis
1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
1.6 Persistent PH of the newborn
GROUP 2 PH associated with left heart disease
2.1 Heart failure:
2.1.1 with preserved ejection fraction
2.1.2 with reduced or mildly reduced ejection fraction[b]
2.2 Valvular heart disease
2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH
GROUP 3 PH associated with lung diseases and/or hypoxia
3.1 Obstructive lung disease or emphysema
3.2 Restrictive lung disease
3.3 Lung disease with mixed restrictive/obstructive pattern
3.4 Hypoventilation syndromes
3.5 Hypoxia without lung disease (e.g. high altitude)
3.6 Developmental lung disorders
GROUP 4 PH associated with pulmonary artery obstructions
4.1 Chronic thrombo-embolic PH
4.2 Other pulmonary artery obstructions[c]
GROUP 5 PH with unclear and/or multifactorial mechanisms
5.1 Haematological disorders[d]
5.2 Systemic disorders[e]
5.3 Metabolic disorders[f]
5.4 Chronic renal failure with or without haemodialysis
5.5 Pulmonary tumour thrombotic microangiopathy
5.6 Fibrosing mediastinitis



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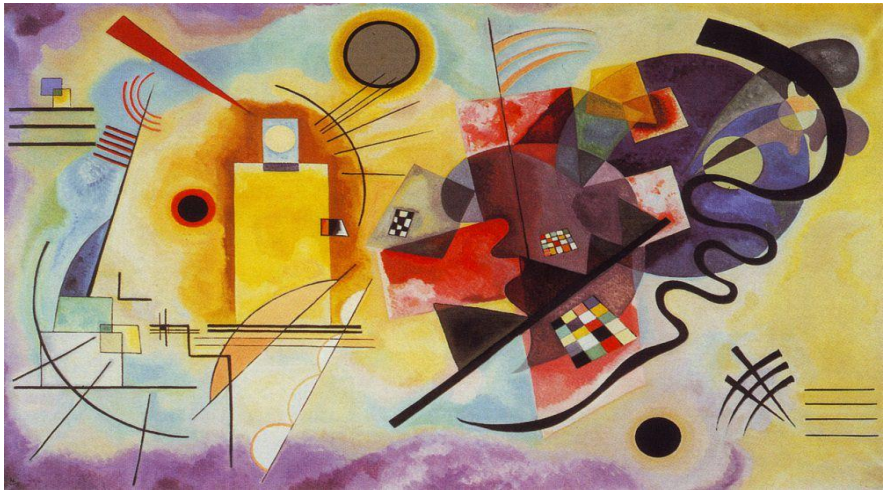

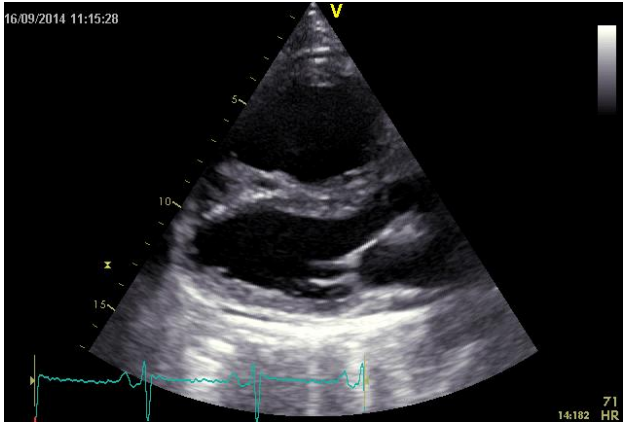
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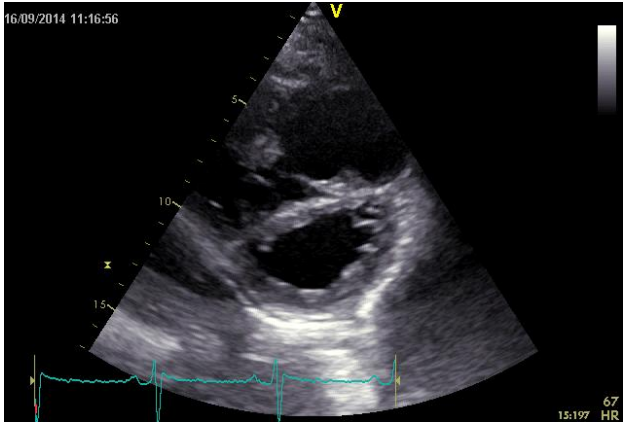
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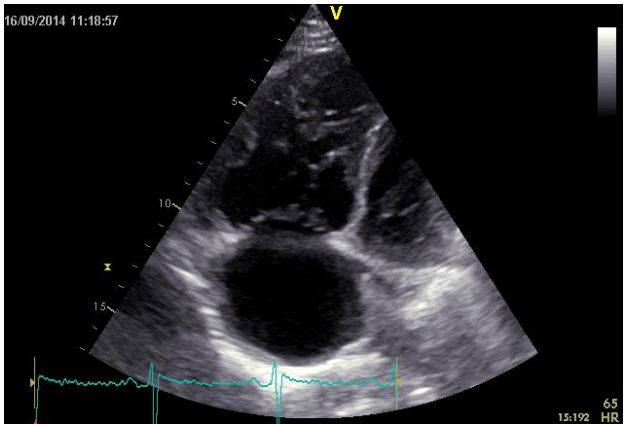
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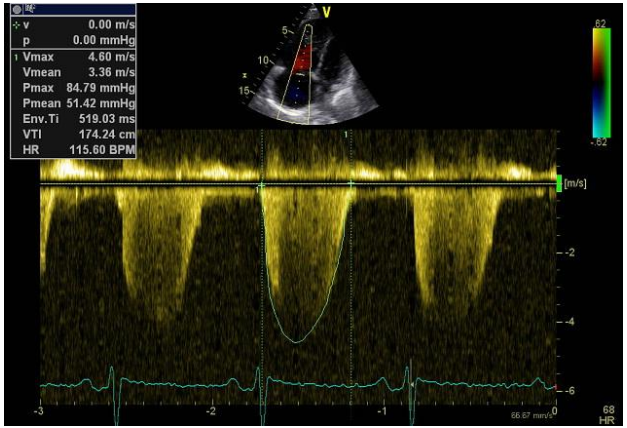
v 0.00 m/s
p 0.00 mmHg
Vmax 4.60 m/s
Vmean 3.36 m/s
Pmax 84.79 mmHg
Pmean 51.42 mmHg
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VTI 174.24 cm
HR 115.60 BPM
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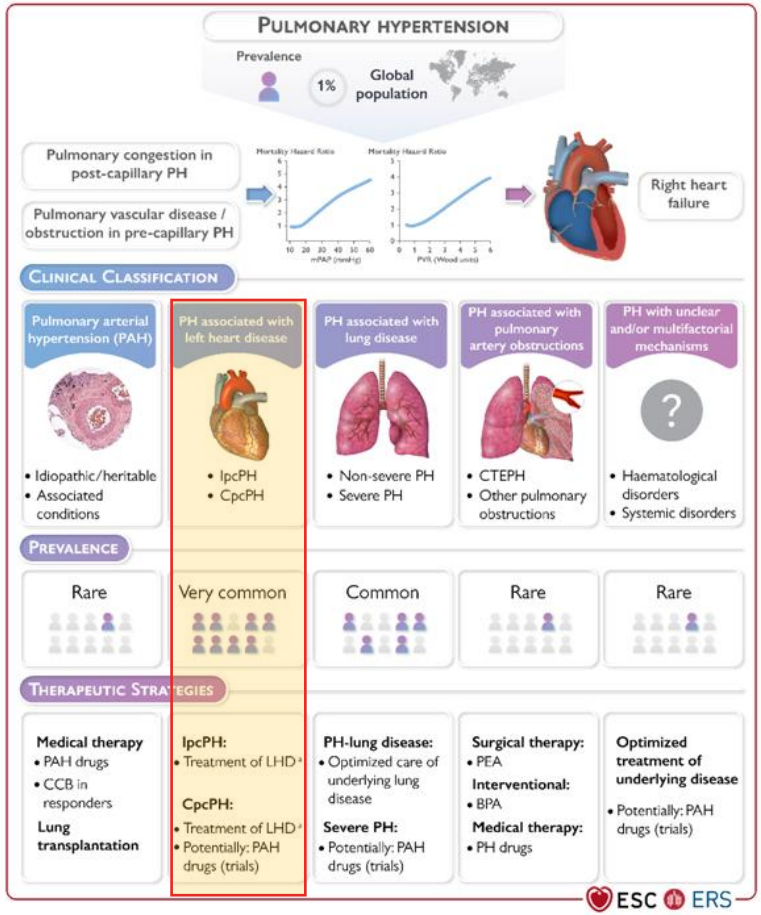
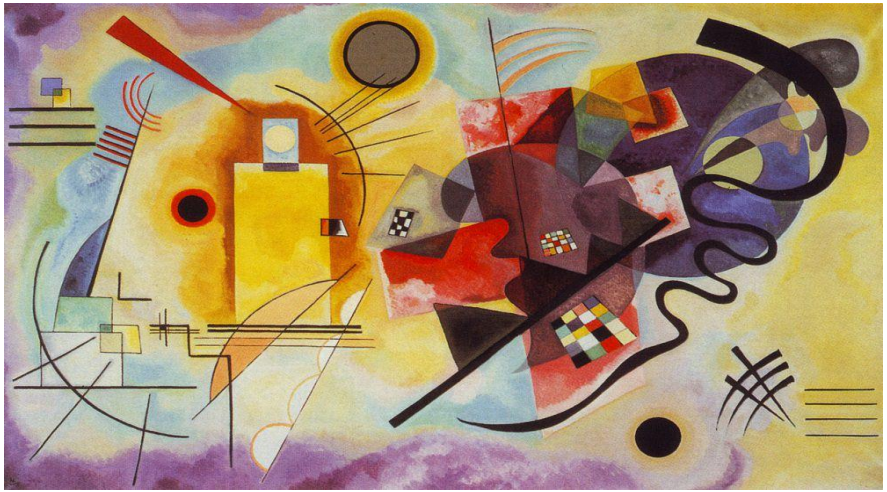
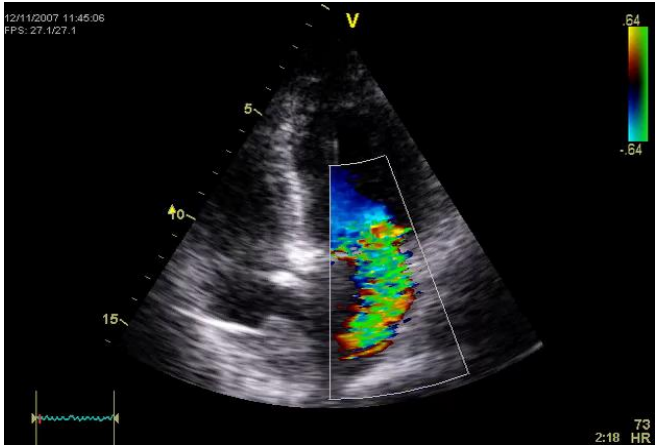
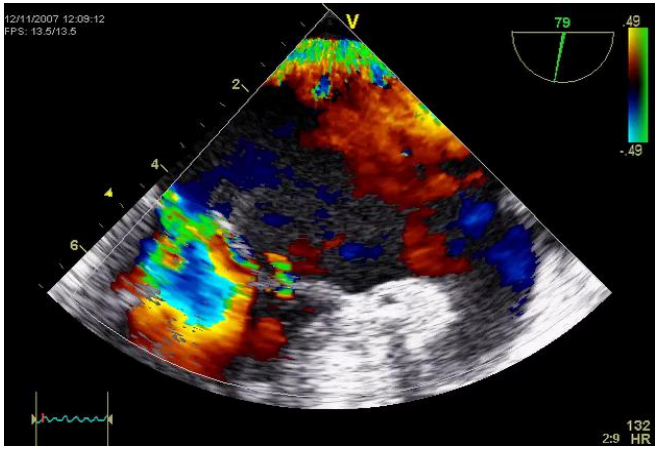
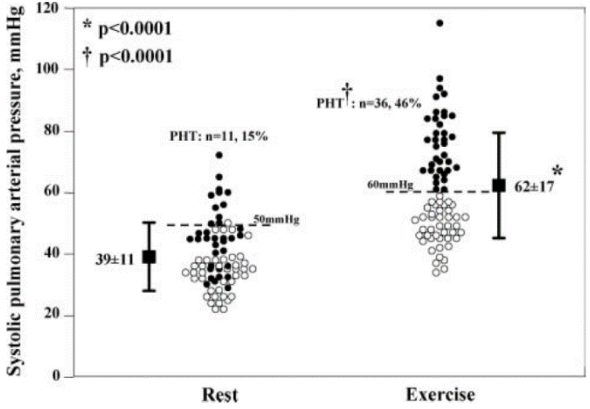


KAZUISTIKY 2









Circulation
American Heart Association.
Exercise Pulmonary Hypertension in Asymptomatic Degenerative Mitral Regurgitation
Julien Magne, Patrizio Lancellotti and Luc A. Pierard
Circulation. 2010;122:33-41; originally published online June 21, 2010;
doi: 10.1161/CIRCULATIONAHA.110.958241
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 0009-7322. Online ISSN: 1524-4539

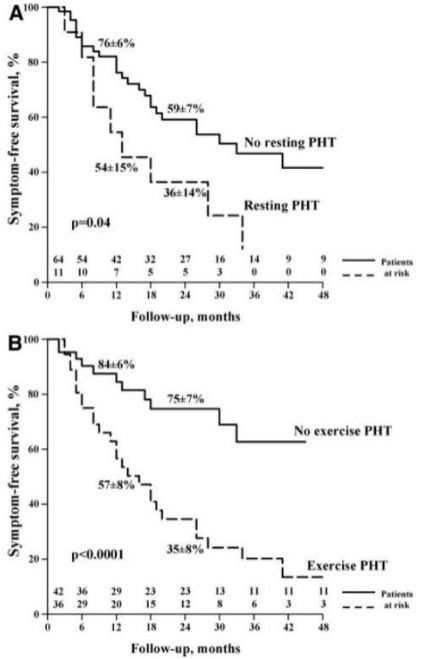
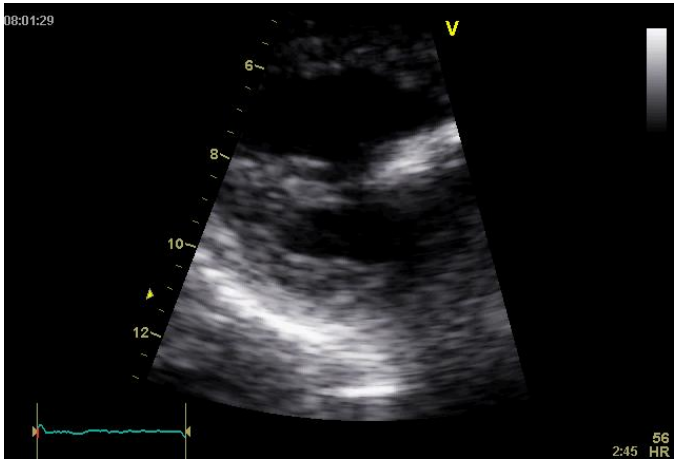


Figure 2. Symptom-free survival according to resting (A) and exercise (B) PHT.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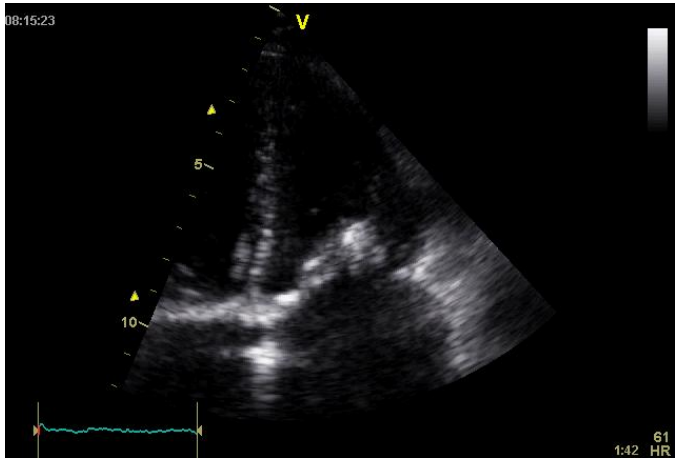
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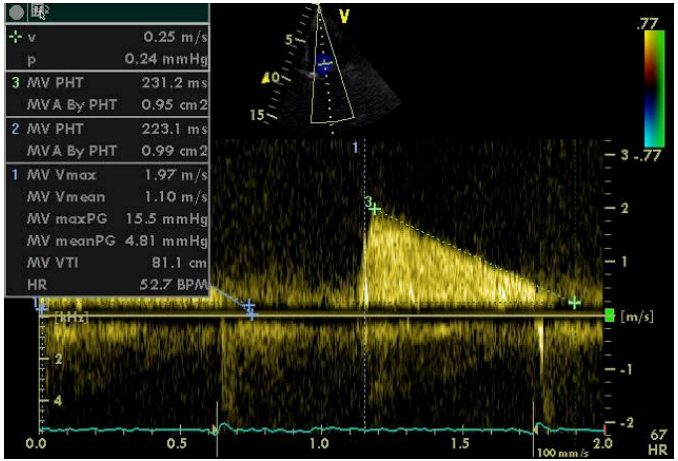
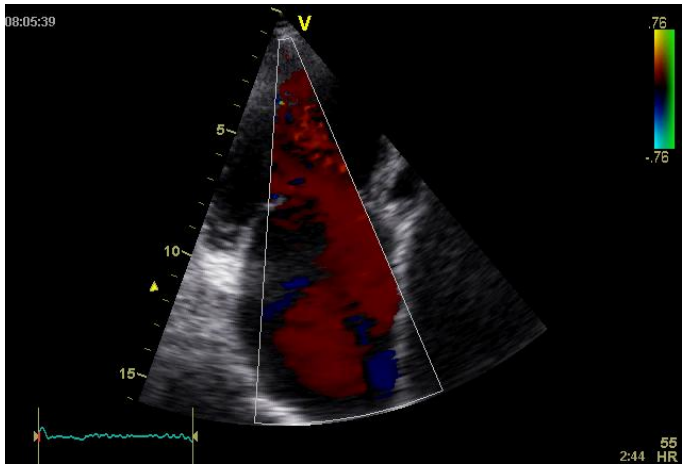
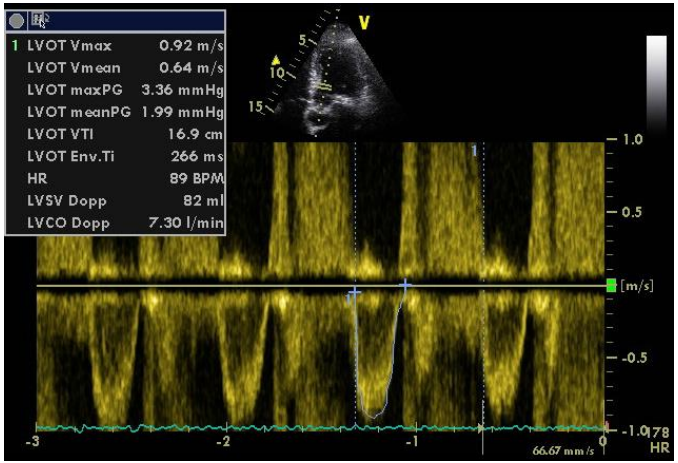
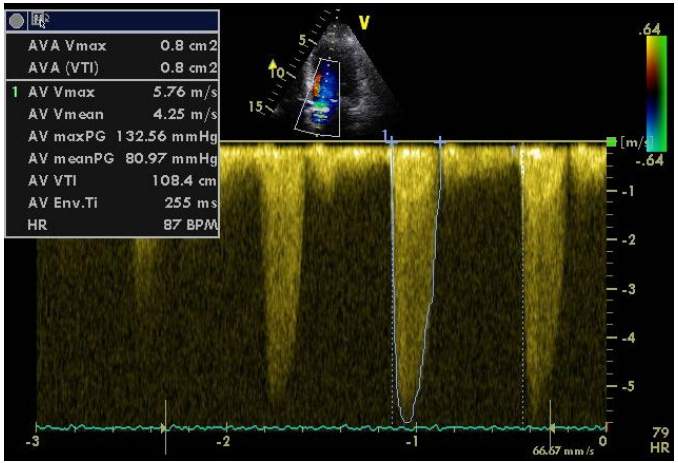

v 0.25 m/s
p 0.24 mmHg
3 MV PHT 231.2 ms
MVA By PHT 0.95 cm2
2 MV PHT 223.1 ms
MVA By PHT 0.99 cm2
1 MV Vmax 1.97 m/s
MV Vmean 1.10 m/s
MV maxPG 15.5 mmHg
MV meanPG 4.81 mmHg
MV VTI 81.1 cm
HR 52.7 BPM
V
5
10
15
.77
-.77
3
2
1
-1
-2
[m/s]
0.0
0.5
1.0
1.5
2.0
100 mm/s
67
HR


08:05:39
V
5
10
15
.76
-.76
55
2:44
HR





Circulation
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

Print ISSN: 0009-7322. Online ISSN: 1524-4539

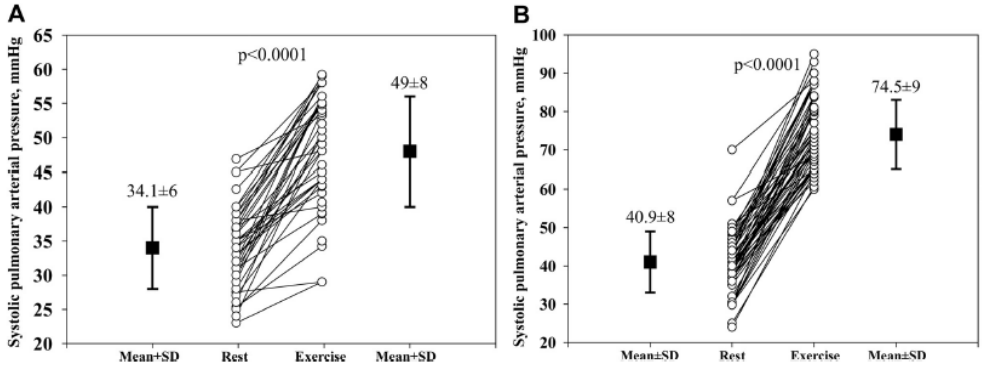


Figure 1. Impact of exercise on systolic pulmonary arterial pressure in patients without exercise pulmonary hypertension (**A**) and with exercise pulmonary hypertension (**B**).

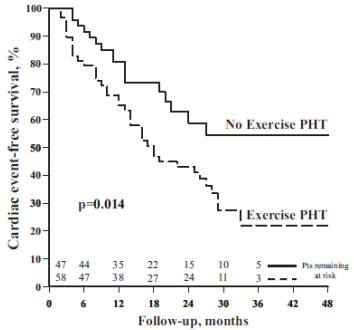
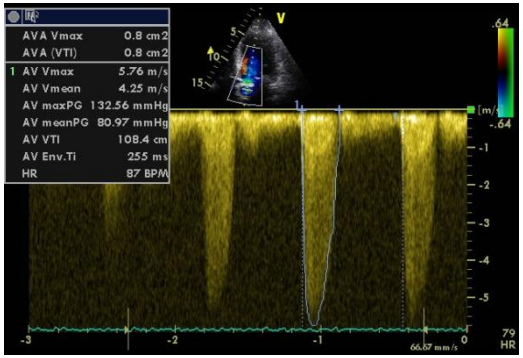


Figure 2. Cardiac event-free survival according to the presence or absence of exercise pulmonary hypertension (PHT).

C) Asymptomatic patients with severe aortic stenosis (refers only to patients eligible for surgical valve replacement)		
SAVR is indicated in asymptomatic patients with severe aortic stenosis and systolic LV dysfunction (LVEF <50%) not due to another cause.	I	C
SAVR is indicated in asymptomatic patients with severe aortic stenosis and an abnormal exercise test showing symptoms on exercise clearly related to aortic stenosis.	I	C
SAVR should be considered in asymptomatic patients with severe aortic stenosis and an abnormal exercise test showing a decrease in blood pressure below baseline.	IIa	C
SAVR should be considered in asymptomatic patients with normal ejection fraction and none of the above-mentioned exercise test abnormalities if the surgical risk is low and one of the following findings is present: • Very severe aortic stenosis defined by a V_{max} >5.5 m/s • Severe valve calcification and a rate of V_{max} progression ≥0.3 m/s/year • Markedly elevated BNP levels (>threefold age- and sex-corrected normal range) confirmed by repeated measurements without other explanations • Severe pulmonary hypertension (systolic pulmonary artery pressure at rest >60 mmHg confirmed by invasive measurement) without other explanation.	IIa	C



Indications for surgery in asymptomatic aortic stenosis

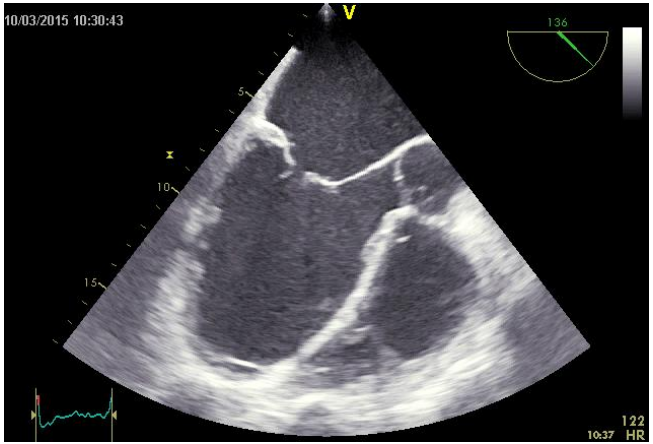
New IIa C recommendation:

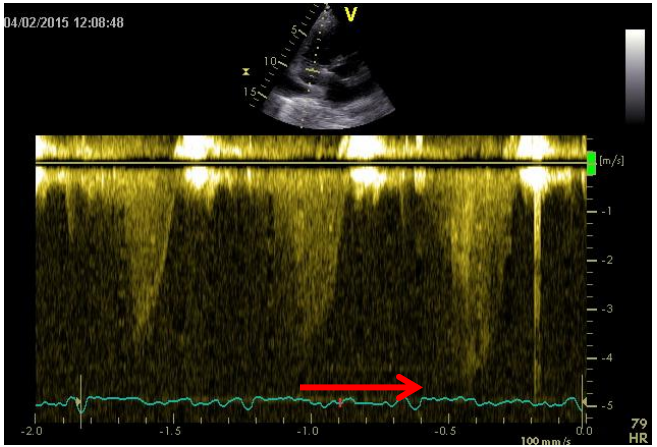
Severe pulmonary hypertension (systolic pulmonary artery pressure at rest >60 mmHg confirmed by invasive measurement) without other explanation.

Indications for intervention in asymptomatic severe primary mitral regurgitation

New additional statement:

If pulmonary hypertension (SPAP >50 mmHg at rest) is the only indication for surgery, the value should be confirmed by invasive measurement.



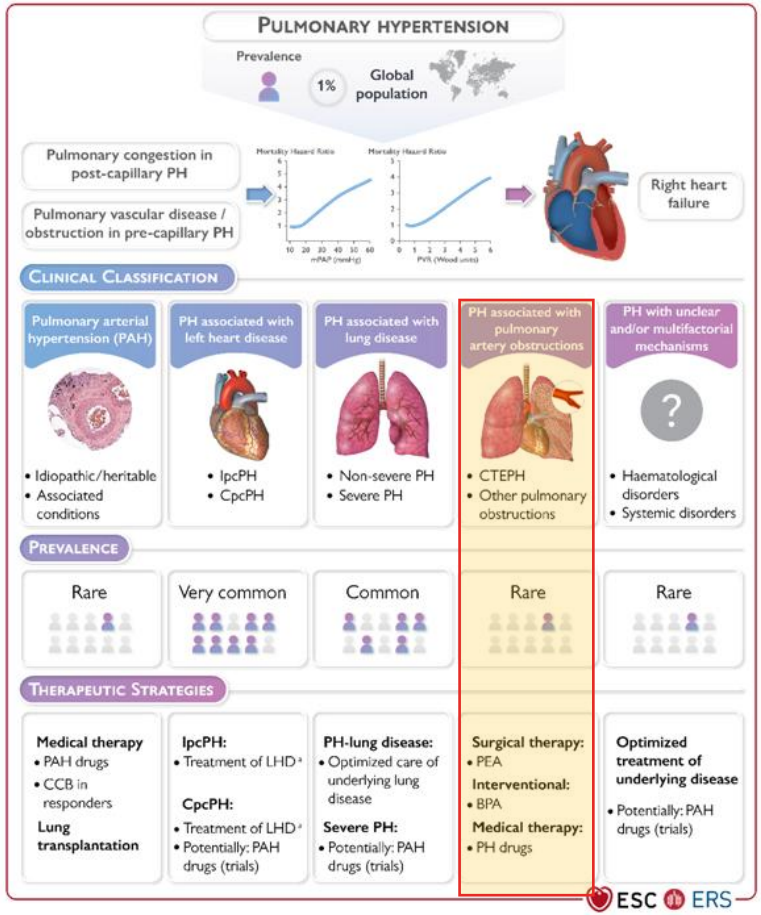


HTx

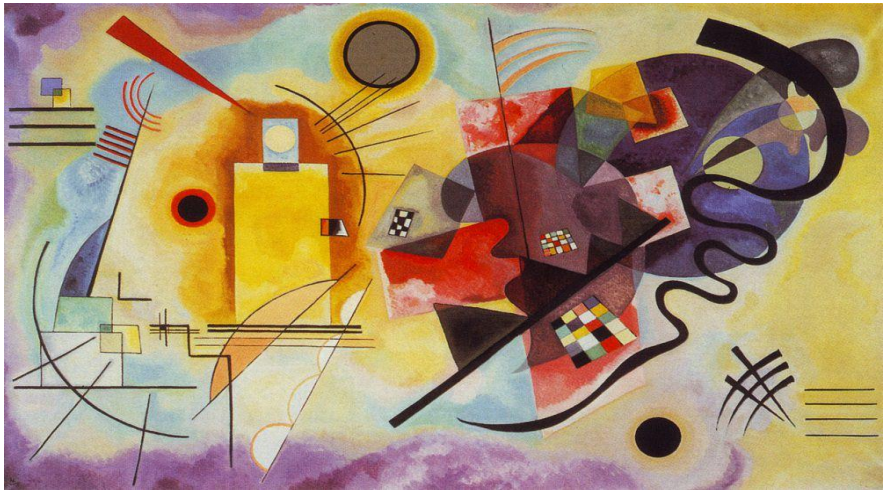
Patients to consider	End-stage HF with severe symptoms, a poor prognosis, and no remaining alternative treatment options. Motivated, well informed, and emotionally stable. Capable of complying with the intensive treatment required postoperatively.
Contra-indications	Active infection. Severe peripheral arterial or cerebrovascular disease. Pharmacologically irreversible pulmonary hypertension (LVAD should be considered with a subsequent re-evaluation to establish candidacy). Cancer (a collaboration with oncology specialists should occur to stratify each patient as to their risk of tumour recurrence). Irreversible renal dysfunction (e.g. creatinine clearance <30 mL/min). Systemic disease with multi-organ involvement. Other serious co-morbidity with poor prognosis. Pre-transplant BMI >35 kg/m² (weight loss is recommended to achieve a BMI <35 kg/m²). Current alcohol or drug abuse. Any patient for whom social supports are deemed insufficient to achieve compliant care in the outpatient setting.

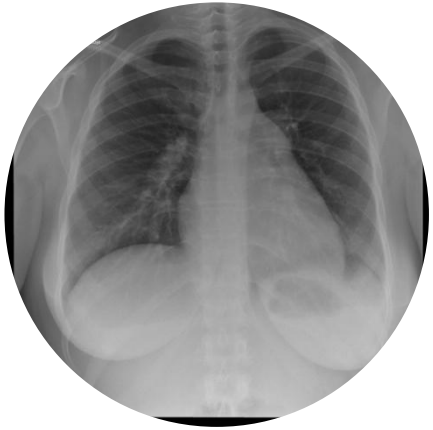
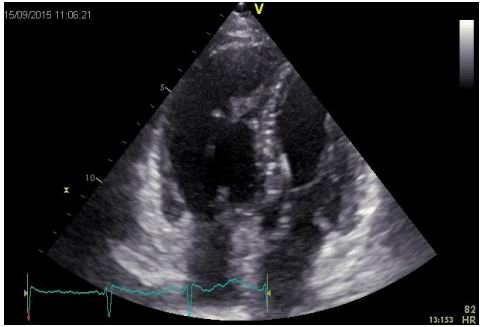
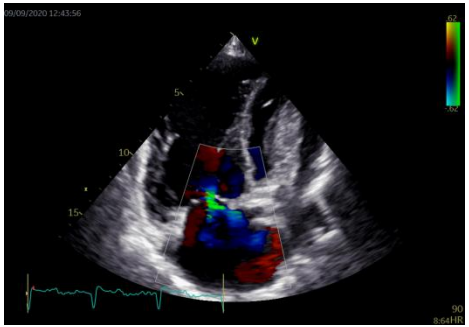
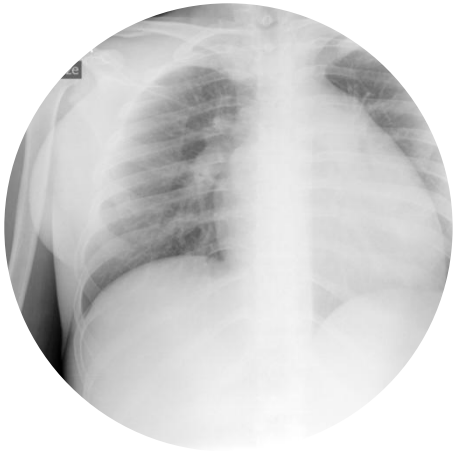
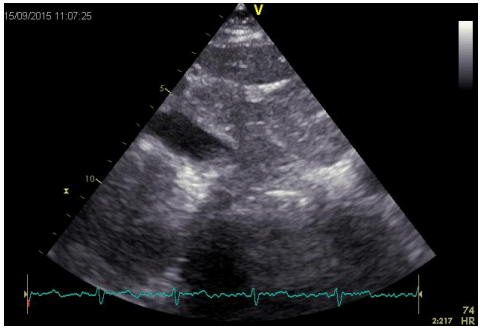
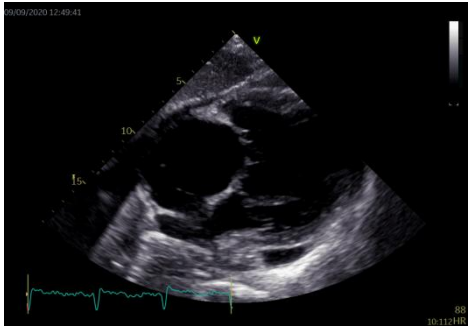
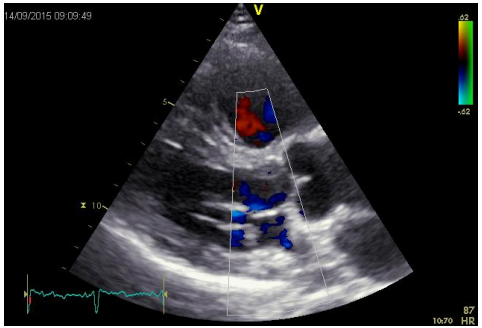
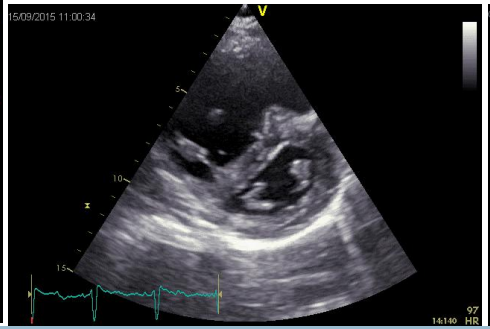
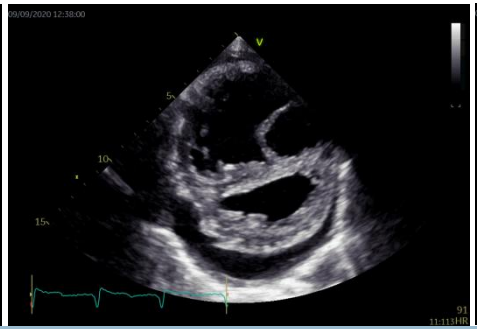
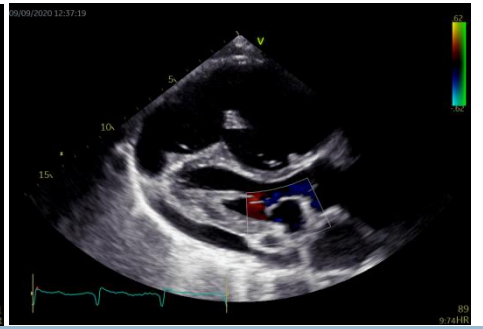
LVAD

Patients with >2 months of severe symptoms despite optimal medical and device therapy and more than one of the following:
LVEF <25% and, if measured, peak VO_2 <12 mL/kg/min.
≥3 HF hospitalizations in previous 12 months without an obvious precipitating cause.
Dependence on i.v. inotropic therapy.
Progressive end-organ dysfunction (worsening renal and/or hepatic function) due to reduced perfusion and not to inadequate ventricular filling pressure (PCWP ≥20 mmHg and SBP ≤80–90 mmHg or CI ≤2 L/min/m²).
Absence of severe right ventricular dysfunction together with severe tricuspid regurgitation.



KAZUISTIKA 3

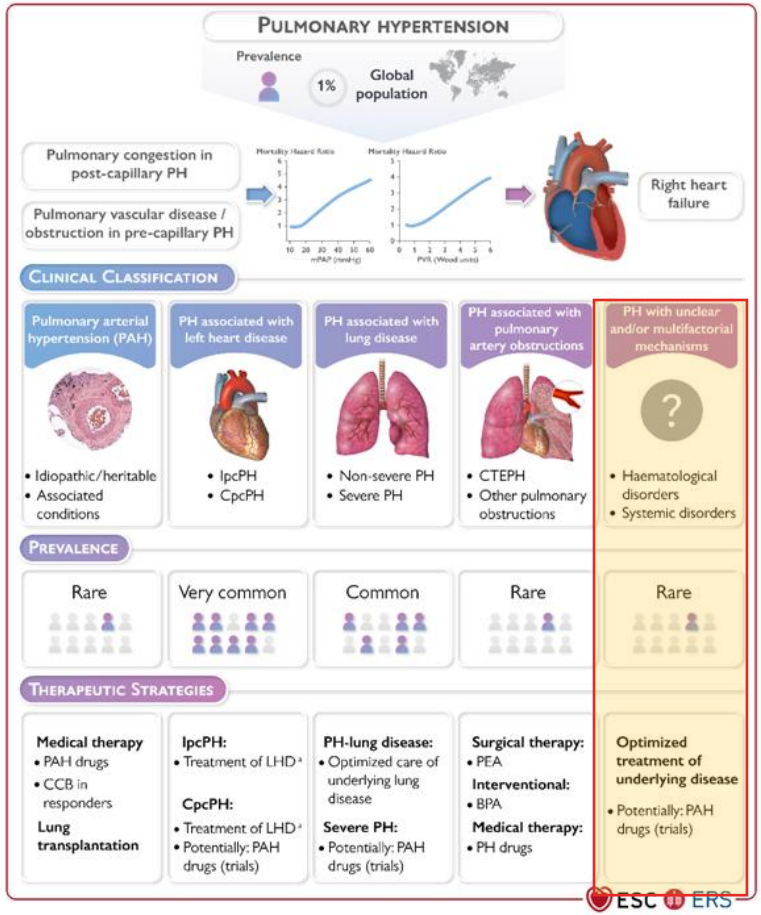


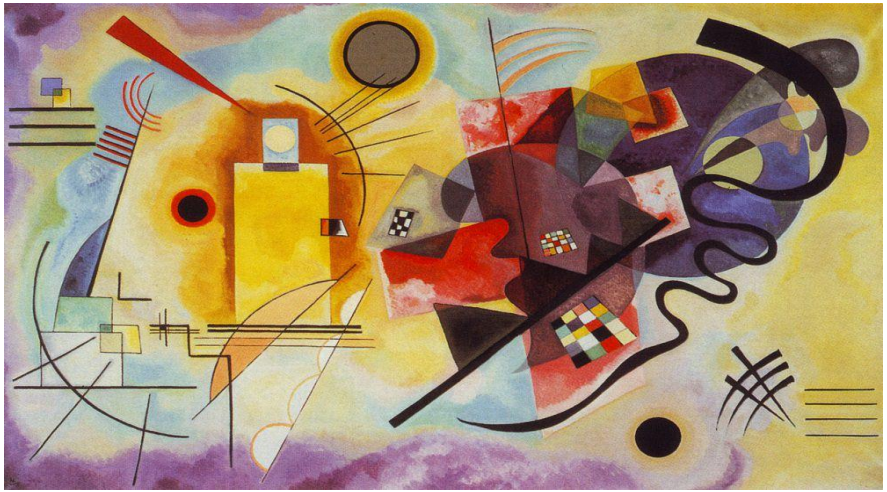
KOMPLEXNÍ KARDIOVASKULÁRNÍ CENTRUM
FAKULTNÍ NEMOCNICE OLOMOUC

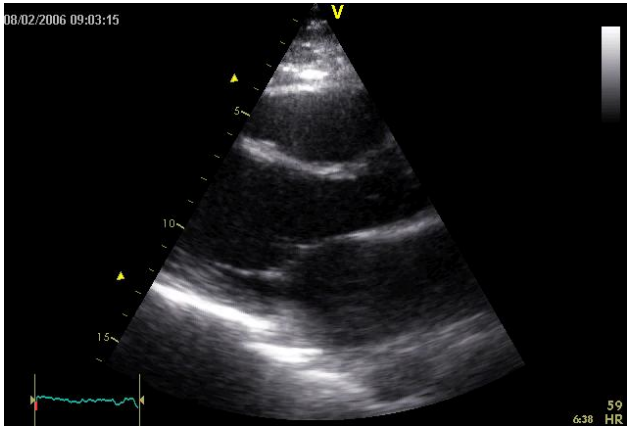
Lékařská fakulta
Univerzity Palackého
v Olomouci

I. INTERNÍ KLINIKA
KARDIOLOGICKÁ
FAKULTNÍ NEMOCNICE OLOMOUC



KAZUISTIKA 4

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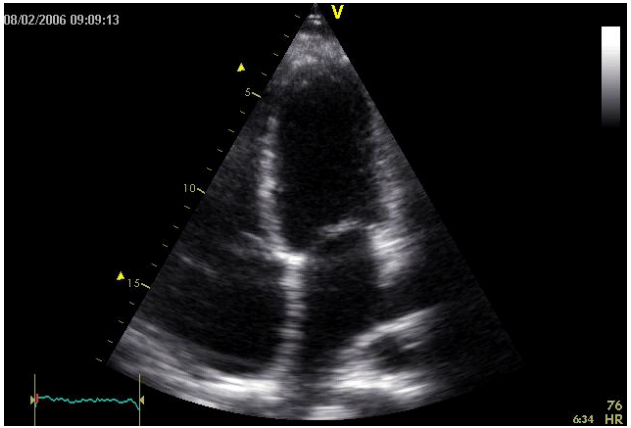
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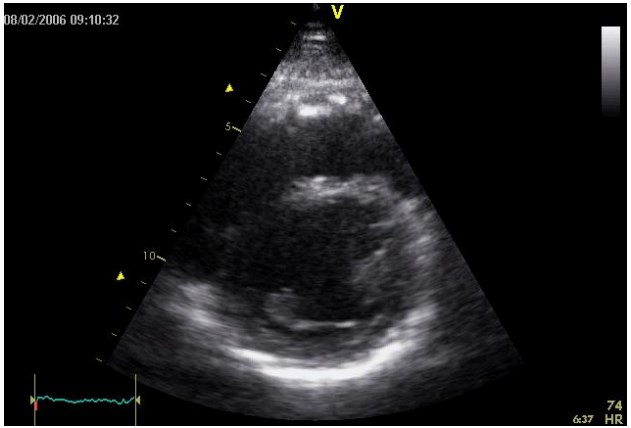
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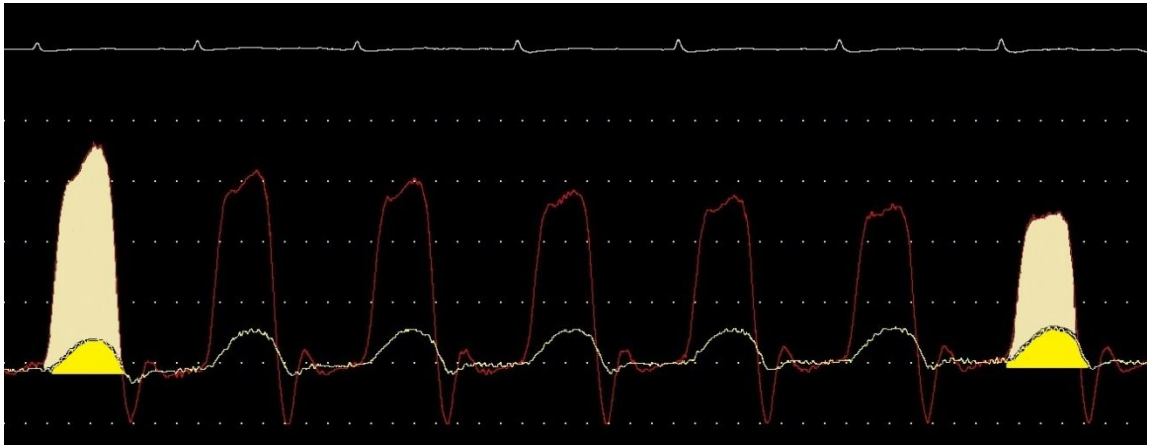
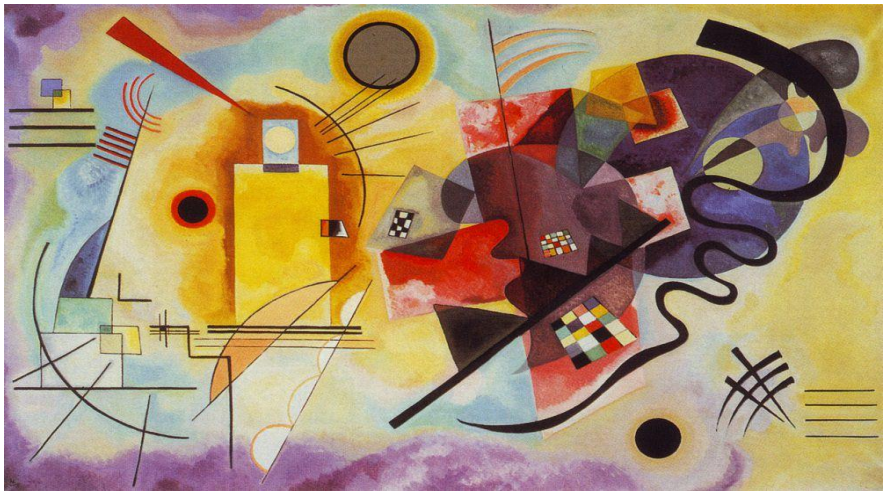
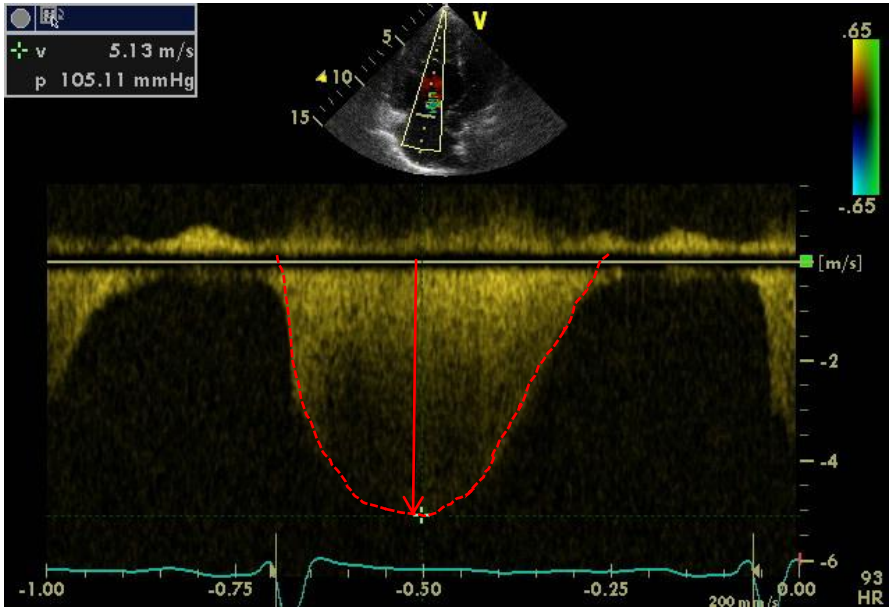
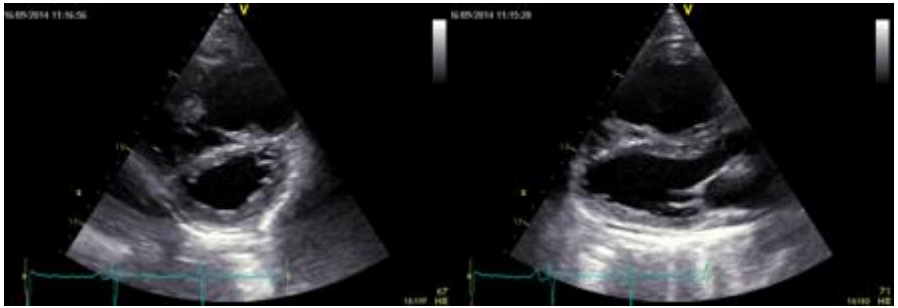
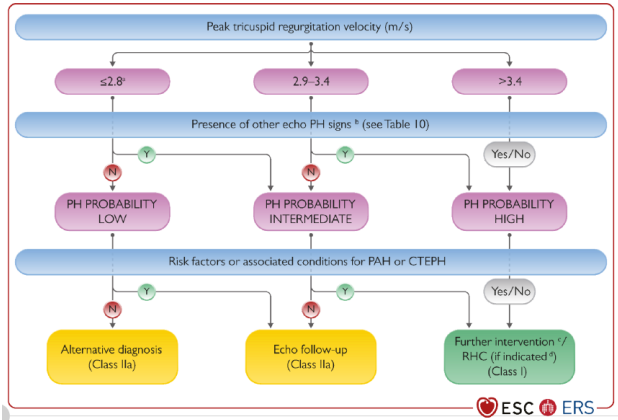


PRAVÁ KOMORA A ZOBRAZOVACÍ METODY





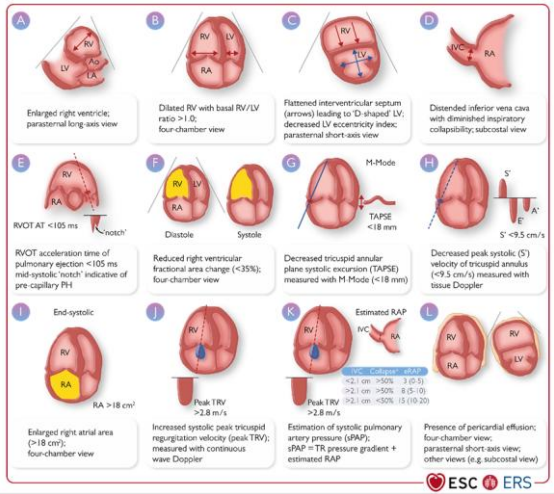




Same threshold for TRV
(≤ 2.8 m/s, 2.9 – 3.4, > 3.4 m/s)

Presence of other PH signs
(TAPSE/PAPs for coupling)

Presence of risk factors
(echo follow-up)

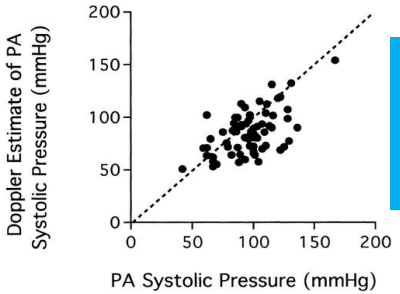


Echokardiografie je nejpoužívanějším screenigovým nástrojem

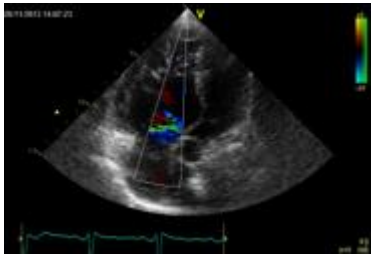
1. **Dvourozměrná echokardiografie** hodnotí funkci pravé komory (RV). Dysfunkce RV má širokou škálu etiologií, z nichž PH je nejběžnější.
2. **Dopplerovská echokardiografie** je užitečná, protože odhaduje rychlost trikuspidálního regurgitativního paprsku (TR jet), což je odhad systolického tlaku v pravé komoře (eRSVP) / systolického tlaku v plicní tepně (ePASP).
3. **Zátěžová echokardiografie** detekuje plicní hypertenzi vyvolanou zátěží, její použití pro screening PAH u pacientů se SSc zůstává do značné míry výzkumnou.

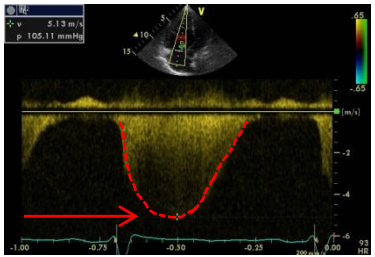
Několik studií pacientů s SSc se známými rizikovými faktory pro PAH i bez nich uvádí míru zvýšeného odhadovaného PASP v rozmezí od 11-14%. Existují však falešně pozitivní i negativní výsledky, které naznačují, že pro screening není echokardiografie vhodná samotná.

Wigley FM, Lima JA, Mayes M, et al. The prevalence of undiagnosed pulmonary arterial hypertension in subjects with connective tissue disease at the secondary health care level of community-based rheumatologists (the UNCOVER study). Arthritis Rheum 2005; 52:2125.
Hesselstrand R, Ekman R, Eskilsson J, et al. Screening for pulmonary hypertension in systemic sclerosis: the longitudinal development of tricuspid gradient in 227 consecutive patients, 1992-2001. Rheumatology (Oxford) 2005; 44:366.
Gladue H, Steen V, Allanore Y, et al. Combination of echocardiographic and pulmonary function test measures improves sensitivity for diagnosis of systemic sclerosis-associated pulmonary arterial hypertension: analysis of 2 cohorts. J Rheumatol 2013; 40:1706.



Alan L. Hinderliter. Circulation. Effects of Long-term Infusion of Prostacyclin (Epoprostenol) on Echocardiographic Measures of Right Ventricular Structure and Function in Primary Pulmonary Hypertension, Volume: 95, Issue: 6, Pages: 1479-1486, DOI: (10.1161/01.CIR.95.6.1479)



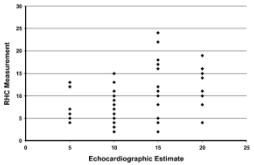


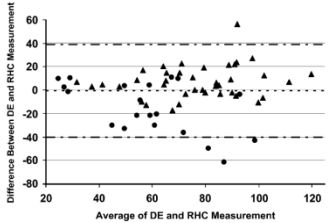
Accuracy of Doppler Echocardiography in the Hemodynamic Assessment of Pulmonary Hypertension

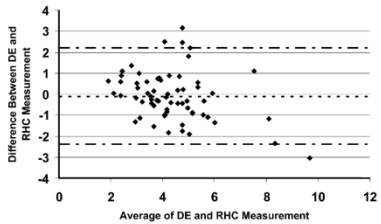
Micah R. Fisher[1]*, Paul R. Forfia[2†], Elzbieta Chamera[2], Traci Housten-Harris[1], Hunter C. Champion[2], Reda E. Girgis[1], Mary C. Corretti[2], and Paul M. Hassoun[1]

[1]Division of Pulmonary and Critical Care Medicine; [2]Division of Cardiology, Department of Medicine, Johns Hopkins University, Baltimore, Maryland

Right-Heart Catheterization	n	Mean	SD
RAP, mm Hg	65	9.4	5.0
PASP, mm Hg	65	68.5	23.9
mPAP, mm Hg	65	41.4	14.6
CO, L/min	65	4.4	1.7
Echocardiogram			
RAP, mm Hg	65	12.4	4.7
RVSP, mm Hg	59	70.2	25.1
CO, L/min	64	4.3	1.4







Odhad systolického tlaku v plicnici - trikuspidální regurgitační gradient

základní parametr pro screening PH pomocí echokardiografie

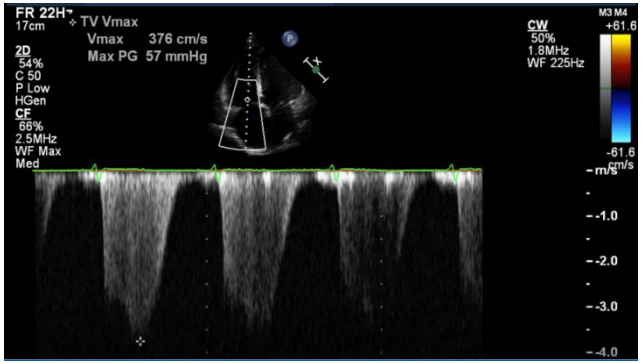
pro **hodnocení prognózy a rozhodnutí o terapii nemá význam**

vzestup odhadovaného sPAP nemusí nutně ukazovat progresi onemocnění, pokles odhadovaného sPAP nemusí nutně odpovídat zlepšení PAP

invazivně měřený mPAP má rovněž poměrně malý prognostický význam (kromě responderů akutní vazoreaktivity)

Tabulka 4A – Pravděpodobnost zjištění plicní hypertenze echokardiografickým vyšetřením symptomatických pacientů s podezřením na plicní hypertenzi

Maximální rychlost proudu krve při nedomykavosti trojcípé chlopně (m/s)	Přítomnost jiných „známek PH" při echokardiografickém vyšetření[a]	Možnost plicní hypertenze podle echokardiografického vyšetření
≤ 2,8 nebo neměřitelná	Ne	Nízká
≤ 2,8 nebo neměřitelná	Ano	Středně vysoká
2,9–3,4	Ne	
2,9–3,4	Ano	Vysoká
> 3,4	Není nutno provádět	



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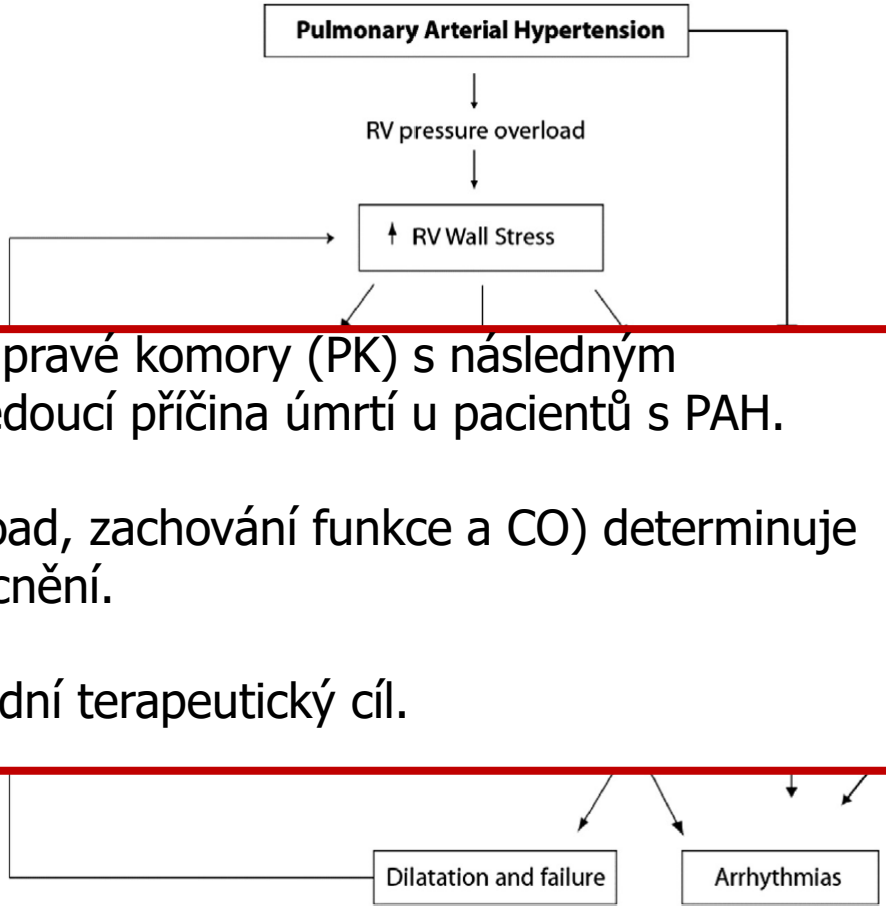
Pravá komora u PAH

- An **afterload mismatch** - increased RV afterload, driven by increased PVR, leads to right heart failure.
- At an early stage, the RV adapts to the increased afterload to preserve stroke volume (**homeometric adaptation**), followed by an **heterometric**

- Vyčerpání kompenzačních mechanismů pravé komory (PK) s následným pravostranným srdečním selháním je vedoucí příčina úmrtí u pacientů s PAH.
- Funkce PK (adaptace na zvýšený afterload, zachování funkce a CO) determinuje funkční status a klinický průběh onemocnění.
- Zachování/zlepšení funkce PK jako zásadní terapeutický cíl.

- In contrast, *maladapted ventricles* usually are characterized by **uncoupling of the RV to the pulmonary circulation**



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Reverzní remodelace PK

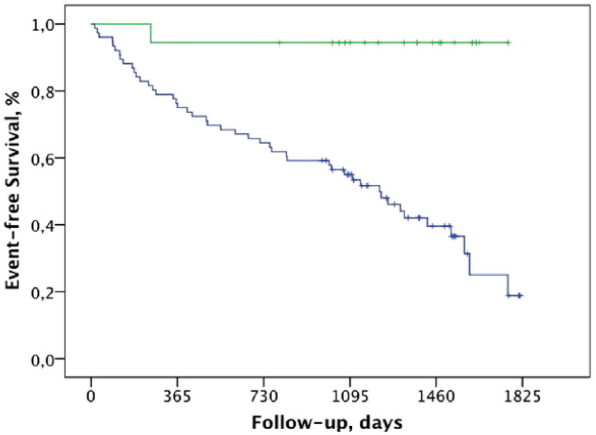

Figure 6 Event-free survival of patients with right heart reverse remodeling (green line; echo score, 4–4.5) compared with patients without right heart reverse remodeling (blue line; echo score, < 4): 94%, 94%, and 94% vs 75%, 55%, and 24% after 1, 3, and 5 years of follow-up, respectively, from the 1-year re-evaluation (p = 0.0001).

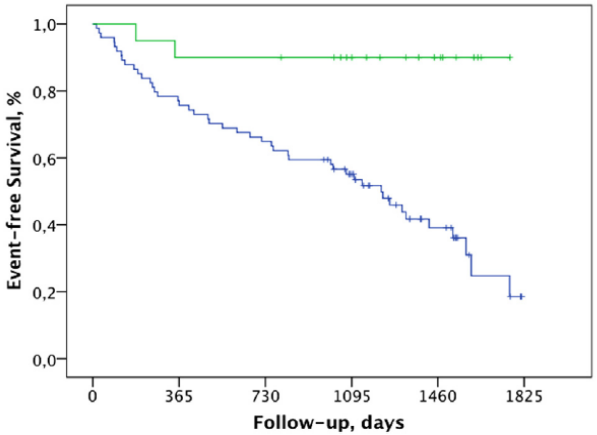

Figure 7 Event-free survival of patients with right heart reverse remodeling (green line) compared with patients without right heart reverse remodeling RHRR (blue line), defined by the presence of the upper tertile of all 3 echo parameters: 88%, 83%, and 83% vs 76, 55%, and 23% after 1, 3, and 5 years of follow-up, respectively, from the 1-year reevaluation (p = 0.001).

decrease in RV enddiastolic area (RVEDA) [-2,45 cm^2 (sensitivity 93 %; specificity 40 %)], **right atrial (RA) area** [-1.30 cm^2 (sensitivity 75 %; specificity 63 %)], **and left ventricular systolic eccentricity index (LV-EIs)** [-0.12 (sensitivity 88 %; specificity 44 %)]

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Prognostic relevance of right heart reverse remodeling in idiopathic pulmonary arterial hypertension

Roberto Badagliacca, MD, PhD • Roberto Poscia, MD, PhD • Beatrice Pezzuto, MD • ...
Roberto Torre, RN, MSN • Francesco Fedele, MD • Carmine Dario Vizza, MD • Show all authors

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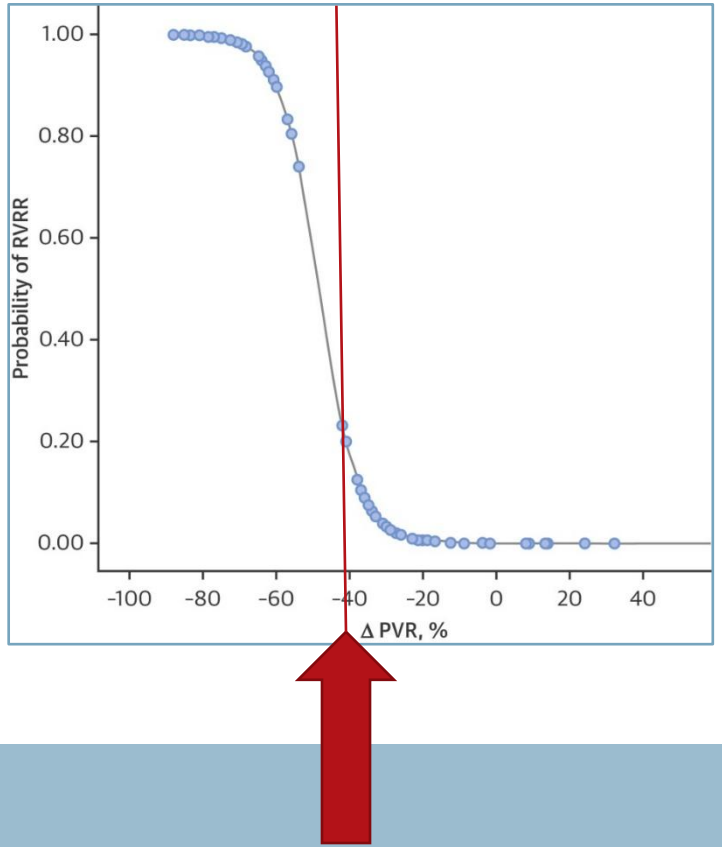

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Pulmonary Hypertension

Progressive Right Ventricular Dysfunction in Patients With Pulmonary Arterial Hypertension Responding to Therapy

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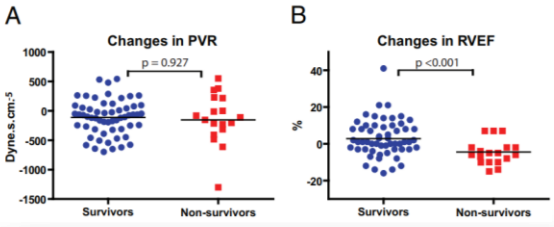
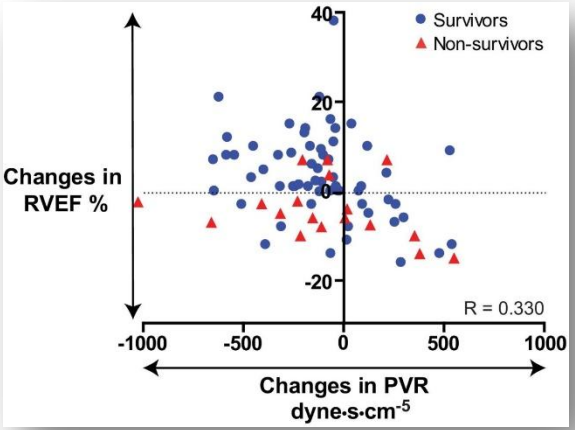
Amsterdam, the Netherlands; and Richmond, Virginia





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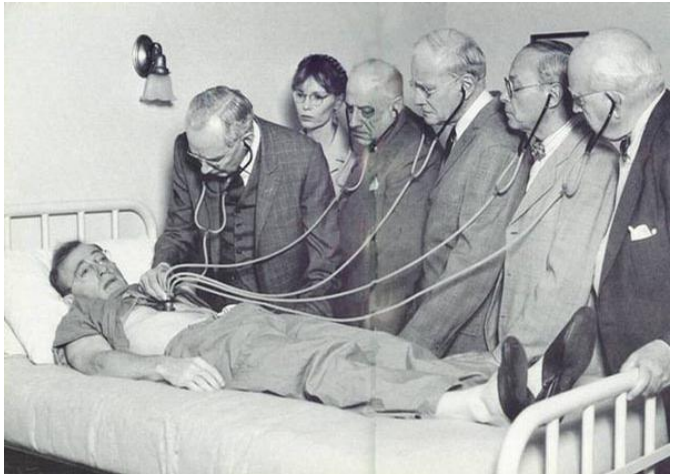
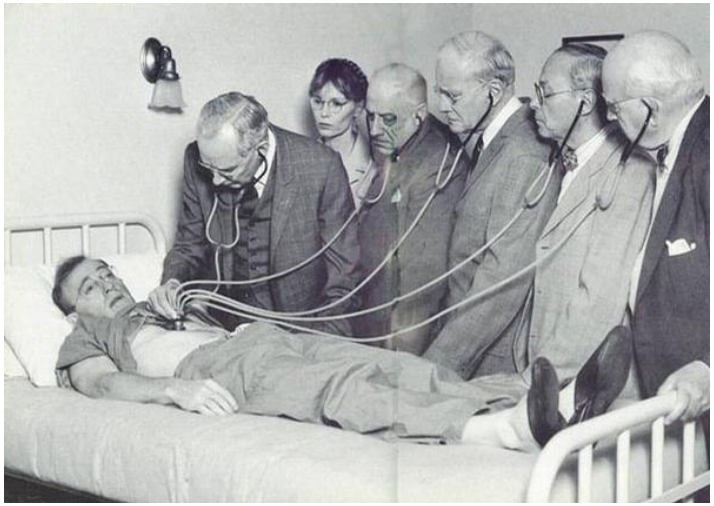


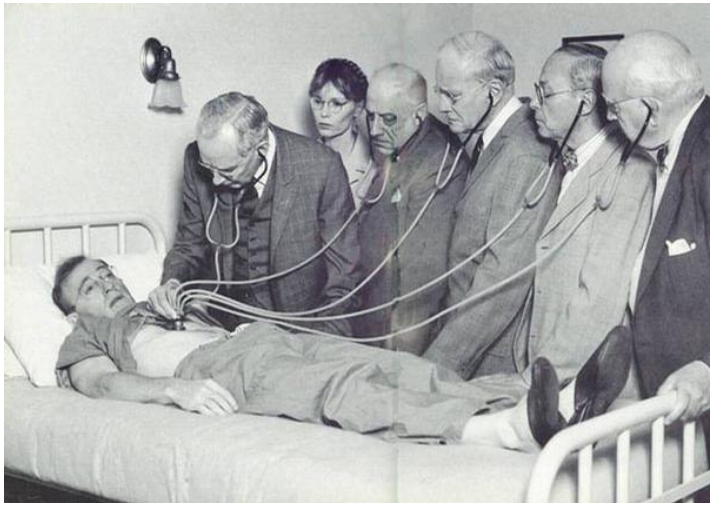
Co lze říci závěrem?

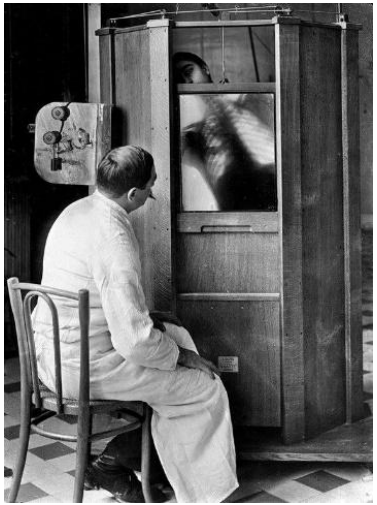
1. Plicní arteriální hypertenze je vzácné onemocnění se **závažnou prognózou**



Co lze říci závěrem?

1. Plicní arteriální hypertenze je vzácné onemocnění se **závažnou prognózou**
2. Podezření na PAH je založeno na **kompetentním zhodnocení** symptomů, objektivního nálezu a korelující funkční/strukturální abnormality plic…





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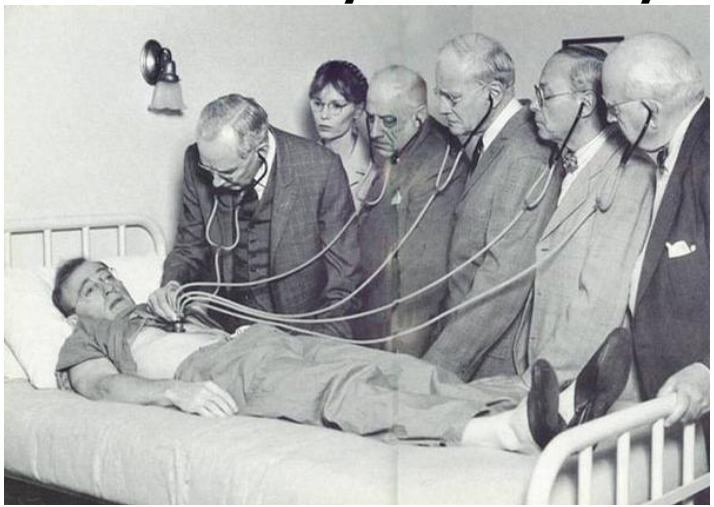
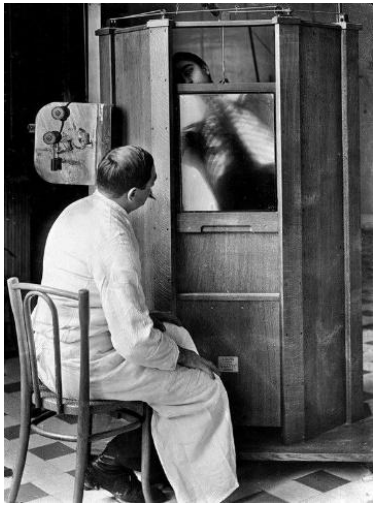
3. Při diferenciální diagnóze dušnosti je nutná **screeningová echokardiografie** a zvážení **hemodynamického vyšetření** a **kauzální léčby**.





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