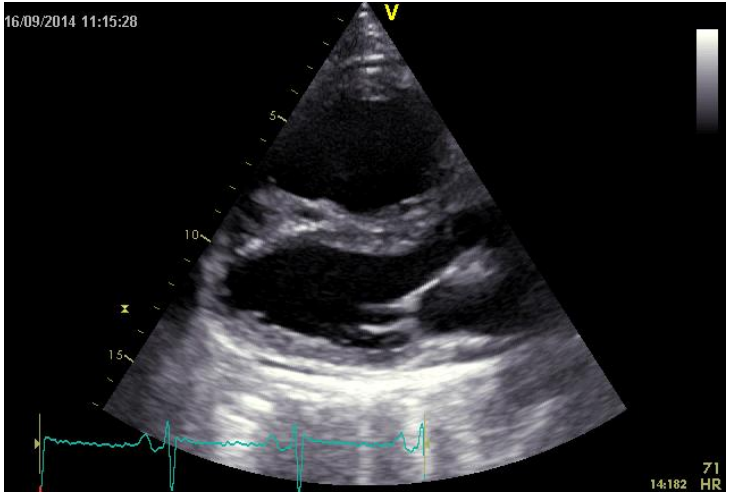
FAKULTNÍ NEMOCNICE® OLOMOUC

Lékařská fakulta
Univerzita Palackého v Olomouci

KOMPLEXNÍ KARDIOVASKULÁRNÍ CENTRUM
FAKULTNÍ NEMOCNICE OLOMOUC

Plicní hypertenze – *definice*

Martin Hutyra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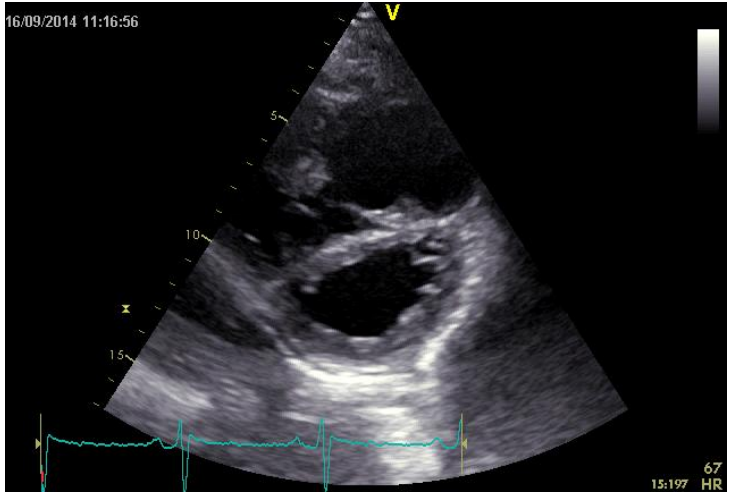
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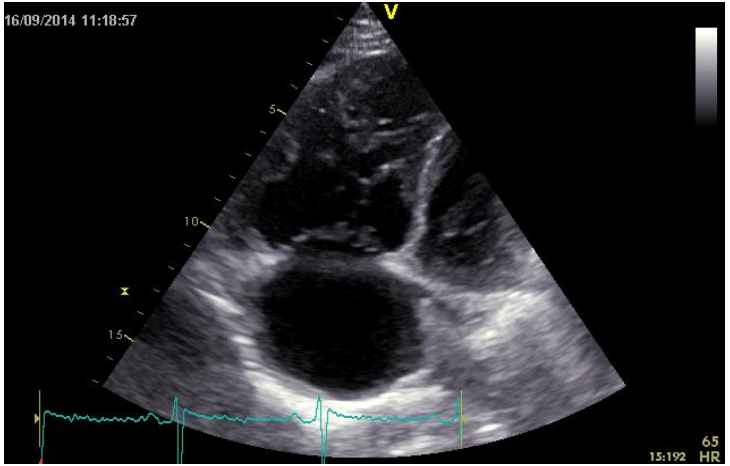
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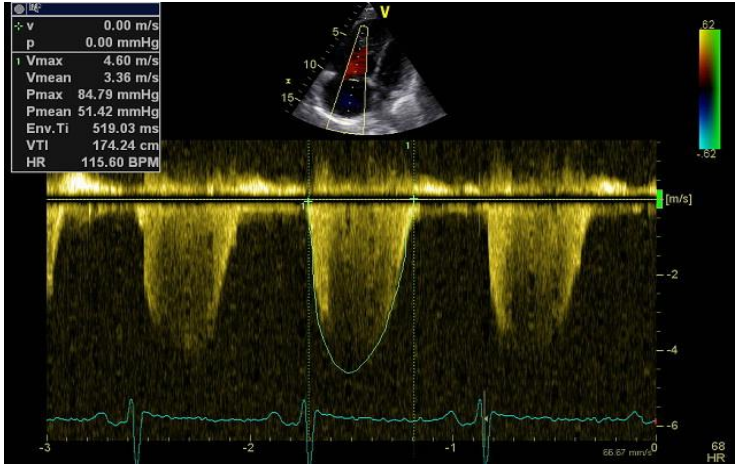
v 0.00 m/s
p 0.00 mmHg
Vmax 4.60 m/s
Vmean 3.36 m/s
Pmax 84.79 mmHg
Pmean 51.42 mmHg
Env.Ti 519.03 ms
VTI 174.24 cm
HR 115.60 BPM
[m/s]
68
HR

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).

Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG).

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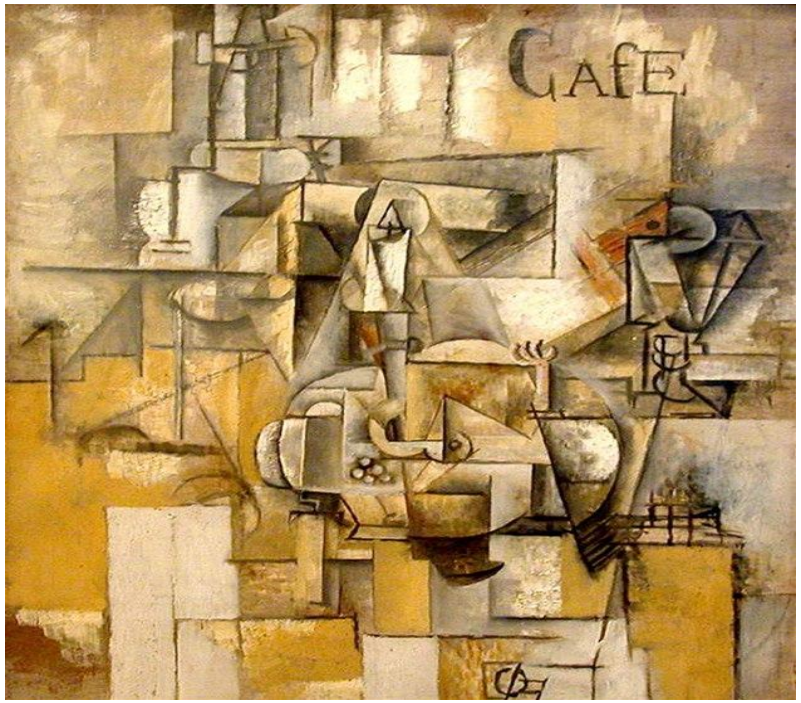
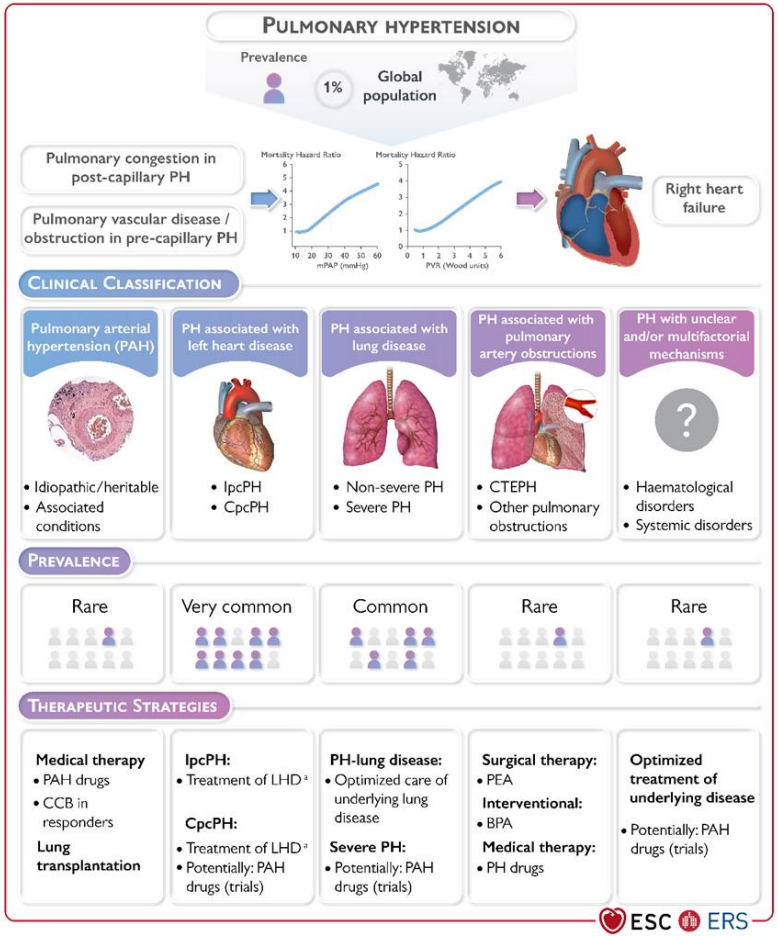
Authors/Task Force Members: Marc Humbert (France), Gabor Kovacs (Austria), Marius M. Hoeper (Germany), Roberto Badagliacca (Italy), Rolf M.F. Berger (Netherlands), Margarita Brida (Croatia), Jørn Carlsen (Denmark), Andrew J.S. Coats (United Kingdom), Pilar Escribano-Subias (Spain), Pisana Ferrari (Italy), Diogenes S. Ferreira (Brazil), Hossein Ardeschir Ghofrani (Germany), George Giannakoulas (Greece), David G. Kiely (United Kingdom), Eckhard Mayer (Germany), Gergely Meszaros (Hungary), Blin Nagavci (Germany), Karen M. Olsson (Germany), Joanna Pepke-Zaba (United Kingdom), Jennifer K. Quint (United Kingdom), Göran Rådegran (Sweden), Gerald Simonneau (France), Olivier Sitbon (France), Thomy Tonia (Switzerland), Mark Toshner (United Kingdom), Jean-Luc Vachiery (Belgium), Anton Vonk Noordegraaf (Netherlands), Marion Delcroix *† (ERS Chairperson) (Belgium), Stephan Rosenkranz *† (ESC Chairperson) (Germany), and ESC/ERS Scientific Document Group


2022 ESC-ERS Guidelines for the
Miró

ÚVOD





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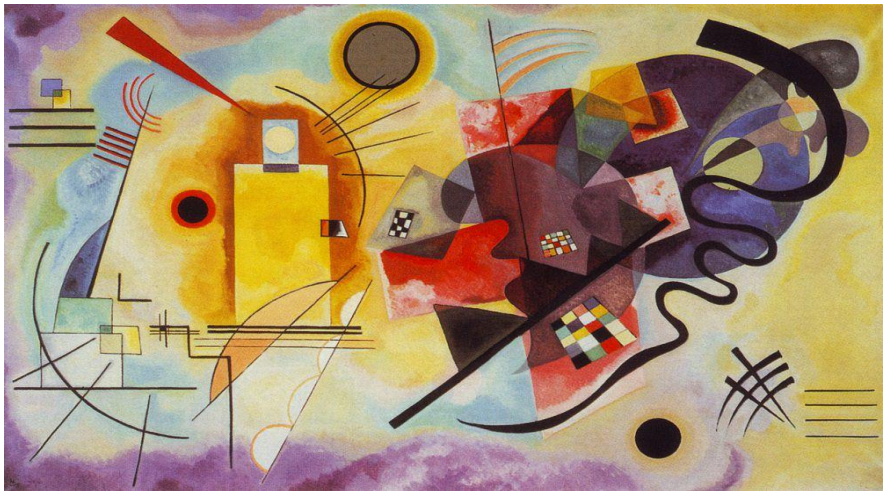
DEFINICE A KLASIFIKACE



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

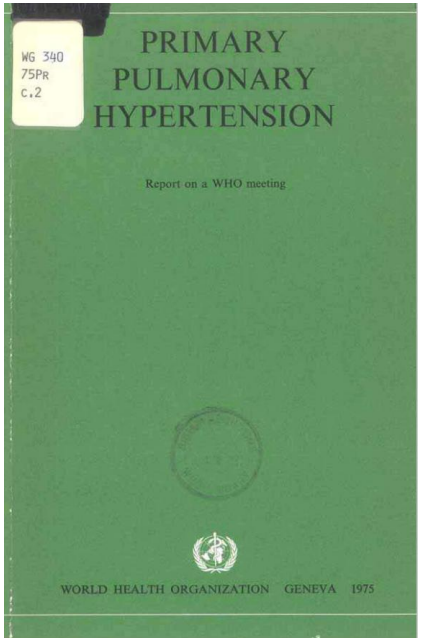
Foreword

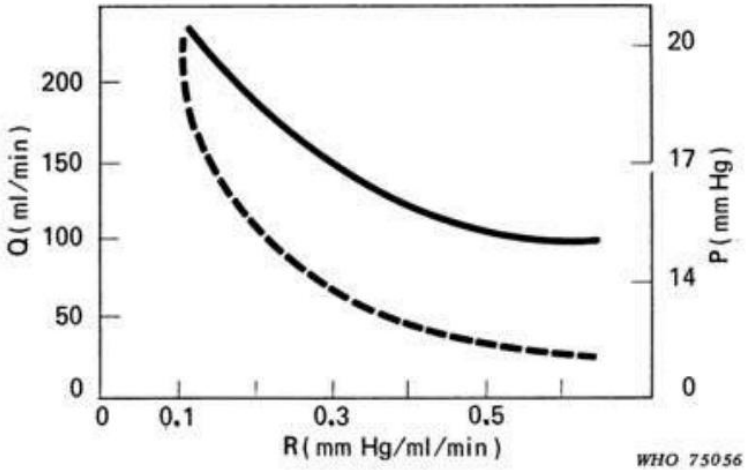
IN THE early months of 1960, the Director-General of the World Health Organization appointed an Expert Committee to inquire into and write a report on the subject of cor pulmonale.

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.



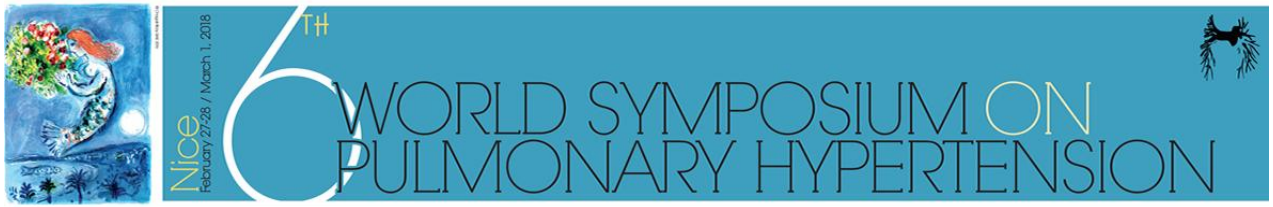


Passive changes in pulmonary vascular resistance (R) at different arterial pressures (P) and at different pulmonary blood flows (Q). Pulmonary venous pressure remains constant throughout. As flow and pressure decrease, resistance increases. The continuous line represents the relationship between P and R, and the interrupted line represents the relationship between Q and R. (Based on a figure in reference no. 4)

Since the 1st World Symposium on Pulmonary Hypertension organised by the WHO in Geneva in 1973, PH has been defined as mPAP ⩾25 mmHg measured by right heart catheterisation in the supine position at rest.
The Geneva WHO meeting was devoted to primary PH, some years after an outbreak related to the intake of the anorexic drug aminorex.
In the report of the meeting, it was recognised that this upper limit of normal mPAP of 25 mmHg was somewhat empirical and arbitrarily defined.

FISHMAN, A. P. Dynamics of the pulmonary circulation. *In:* Hamilton, W. F. & Dow, P., ed. Handbook of physiology, section 2: circulation, Washington, D. C., American Physiological Society, 1963, vol. 2, pp. 1667-1743

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

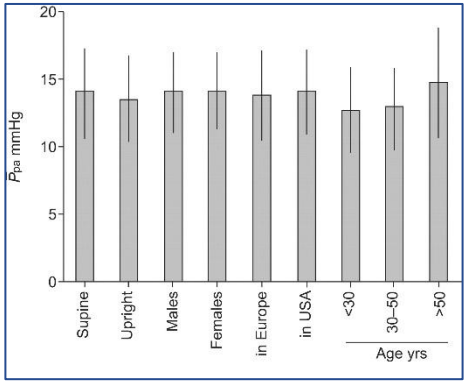
Hoeper MM, *et al. J Am Coll Cardiol* 2013; 62:D42-50.
6th World Symposium on Pulmonary Hypertension, Nice, February 27 to March 1, 2018

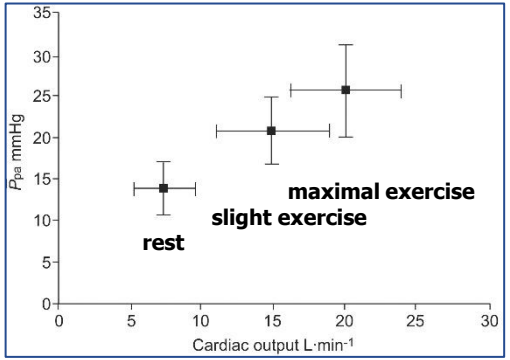
Pulmonary arterial pressure during rest and exercise in healthy subjects: a systematic review

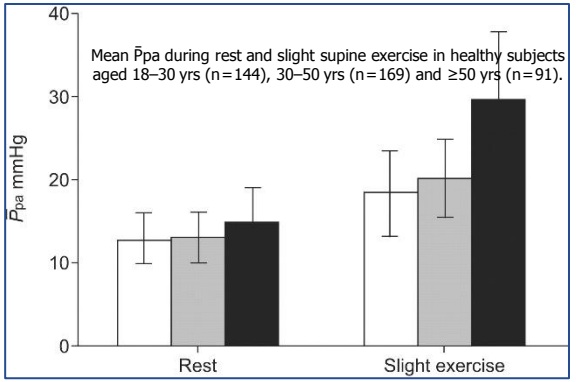
G. Kovacs*, A. Berghold#, S. Scheidl* and H. Olschewski*

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.







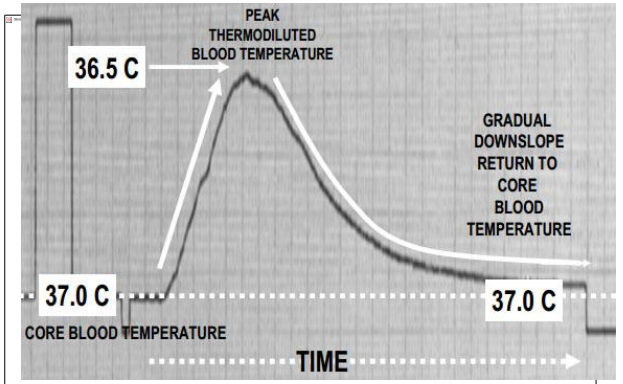
Considering this mPAP of 14 mmHg, two standard deviations would suggest mPAP >20 mmHg as above the upper limit of normal (i.e. above the 97.5th percentile).
This definition is, therefore, no longer arbitrary, but based on a scientific approach.

K diagnostice prekapilární PH svědčící o přítomnosti plicní vaskulární nemoci by měla být zahrnuta hodnota PVR…

Including pulmonary vascular resistance (PVR=(mPAP–PAWP)/CO) in the definition of pre-capillary PH is essential, allowing discrimination of elevation of PAP due to PVD from those due to elevation of PAWP or due to high CO.

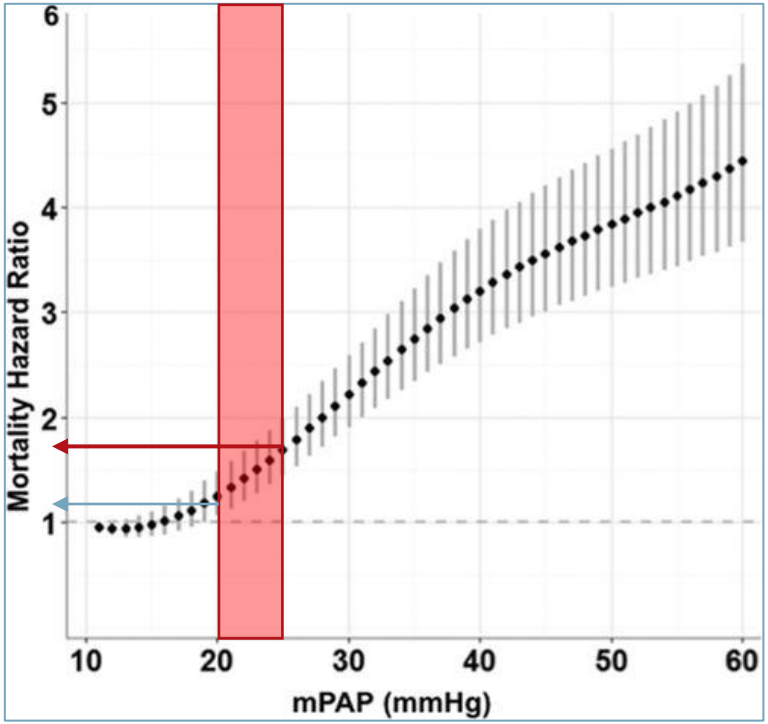
Since the **3rd WSPH held in 2003**, pre-capillary PH of group 1 (PAH) has been defined by the presence of *mPAP ⩾25 mmHg with a normal PAWP ⩽15 mmHg and elevated PVR ⩾3 Wood Units (WU)*.

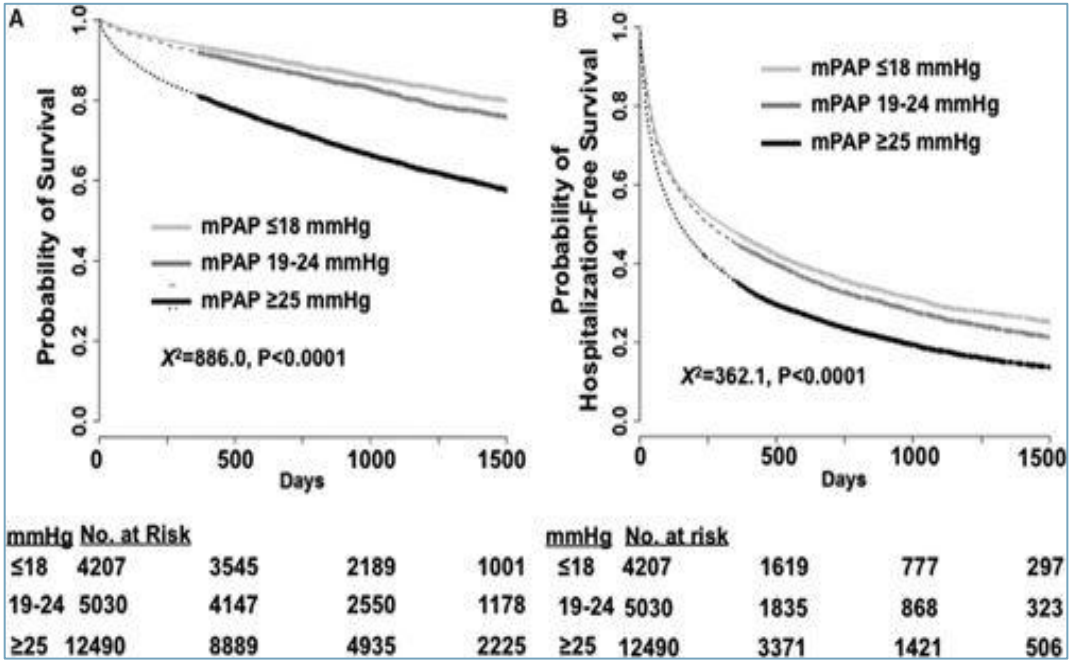
This cut-off value of **PVR ⩾3 WU** is also quite arbitrary since some recent data suggest that **PVR >2 WU** could be also considered abnormal.
The use of a cut-off value of PVR ⩾3 WU is conservative, suggesting the presence of a manifest pre-capillary PH. This value of PVR ⩾3 WU is considered clinically relevant in different clinical situations, suggesting the presence of a significant PVD.



Barst RJ, McGoon M, Torbicki A, et al. Diagnosis and differential assessment of pulmonary arterial hypertension. J Am Coll Cardiol 2004; 43: 40S–47S.
Badesch DB, Champion HC, Sanchez MAG, et al. Diagnosis and assessment of pulmonary arterial hypertension. J Am Coll Cardiol 2009; 54: S55–S66.
Hoeper MM, Bogaard HJ, Condliffe R, et al. Definitions and diagnosis of pulmonary hypertension. J Am Coll Cardiol 2013; 62: D42–D50.

Má hraniční mPAP vliv na prognózu?



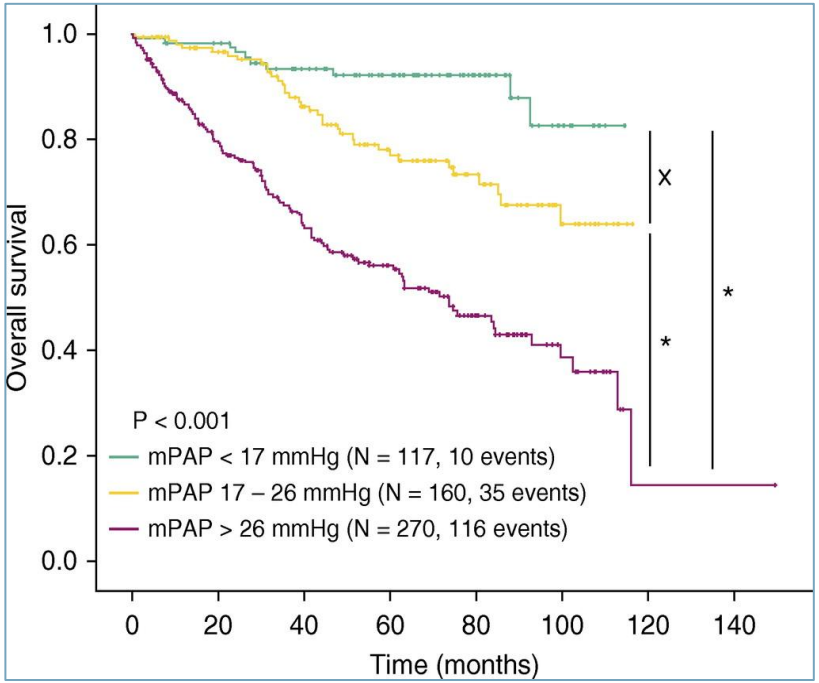


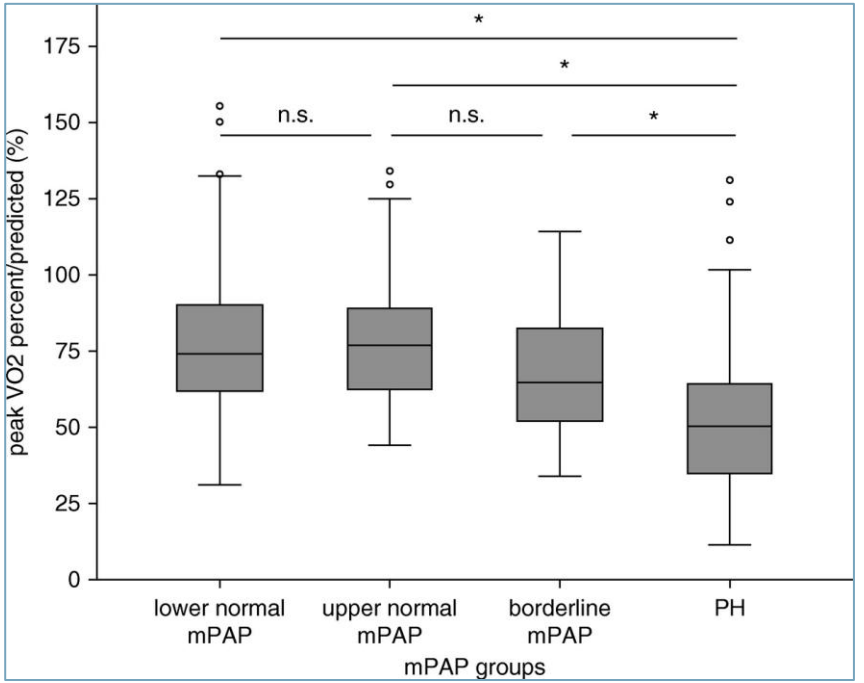
mmHg	No. at Risk			
≤18	4207	3545	2189	1001
19-24	5030	4147	2550	1178
≥25	12490	8889	4935	2225

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)

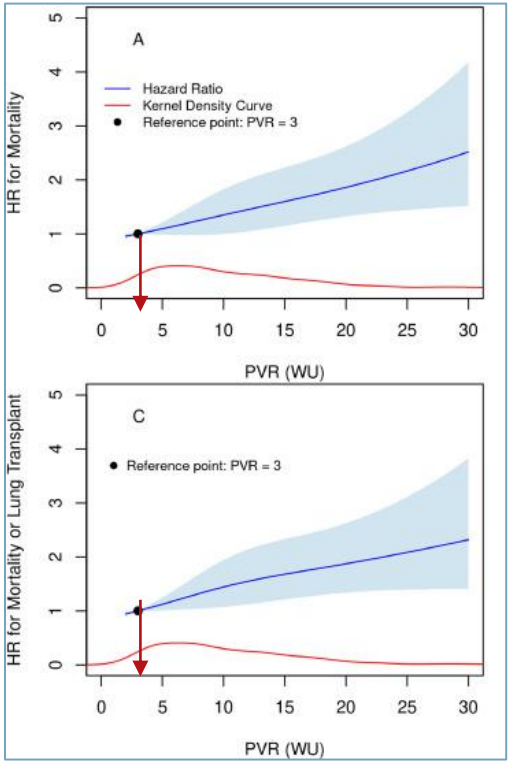
Jakou prognostickou informaci mPAP přináší?





Philipp Douschan; Gabor Kovacs; Alexander Avian; Vasile Foris; Fabian Gruber; Andrea Olschewski; Horst Olschewski; *Am J Respir Crit Care Med* 197509-516. DOI: 10.1164/rccm.201706-1215OC

Má hodnota PVR 2-3 vliv na prognózu?



The Journal of Heart and Lung Transplantation
The Official Publication of the International Society for Heart and Lung Transplantation

FULL LENGTH ARTICLE | VOLUME 40, ISSUE 7, P614-622, JULY 01, 2021

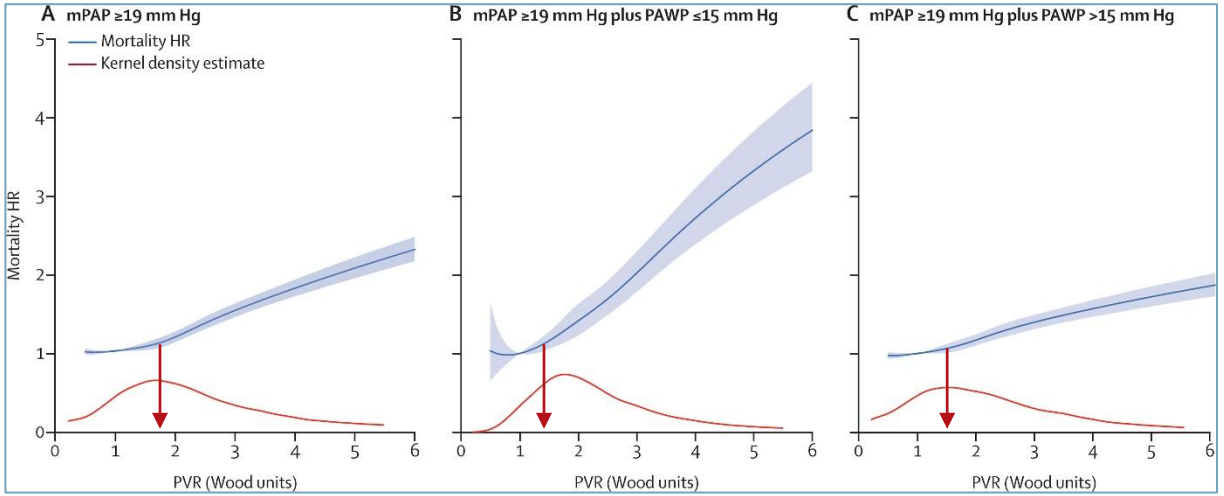
Is pulmonary vascular resistance index better than pulmonary vascular resistance in predicting outcomes in pulmonary arterial hypertension?

THE LANCET
Respiratory Medicine

ARTICLES | VOLUME 8, ISSUE 9, P873-884, SEPTEMBER 01, 2020

Pulmonary vascular resistance and clinical outcomes in patients with pulmonary hypertension: a retrospective cohort study

Bradley A Maron, MD · Evan L Brittain, MD · Edward Hess, MS · Stephen W Waldo, MD · Prof Anna E Barón, PhD · Shi Huang, PhD · et al. Show all authors · Show footnotes

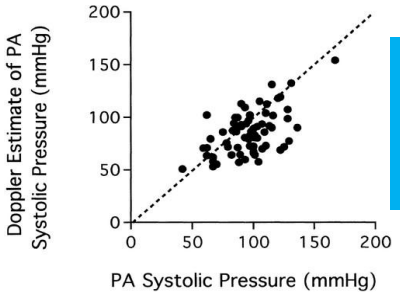


Echokardiografie je nejužitečnějším screenigovým nástrojem

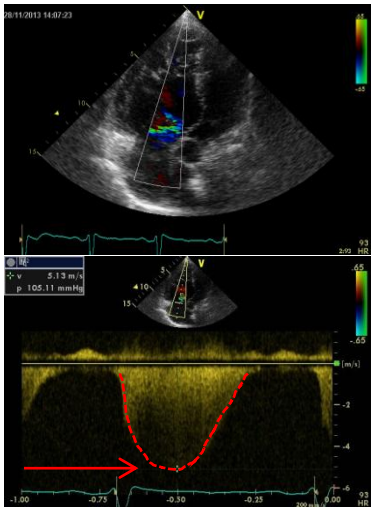
1. **Dvourozměrná echokardiografie** hodnotí funkci pravé komory (RV). Dysfunkce RV má širokou škálu etiologií, z nichž PH je nejběžnější.
2. **Dopplerovská echokardiografie** je užitečná, protože odhaduje rychlost trikuspidálního regurgitativního paprsku (TR jet), což je odhad systolického tlaku v pravé 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)

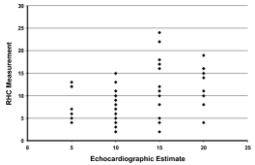


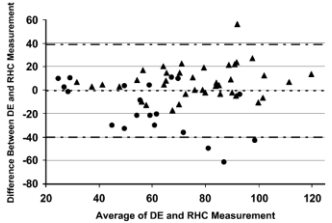
Accuracy of Doppler Echocardiography in the Hemodynamic Assessment of Pulmonary Hypertension

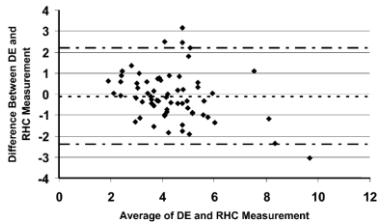
Micah R. Fisher[1]*, Paul R. Forfia[2†], Elzbieta Chamera[2], Traci Housten-Harris[1], Hunter C. Champion[2], Reda E. Girgis[1], Mary C. Corretti[2], and Paul M. Hassoun[1]

[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



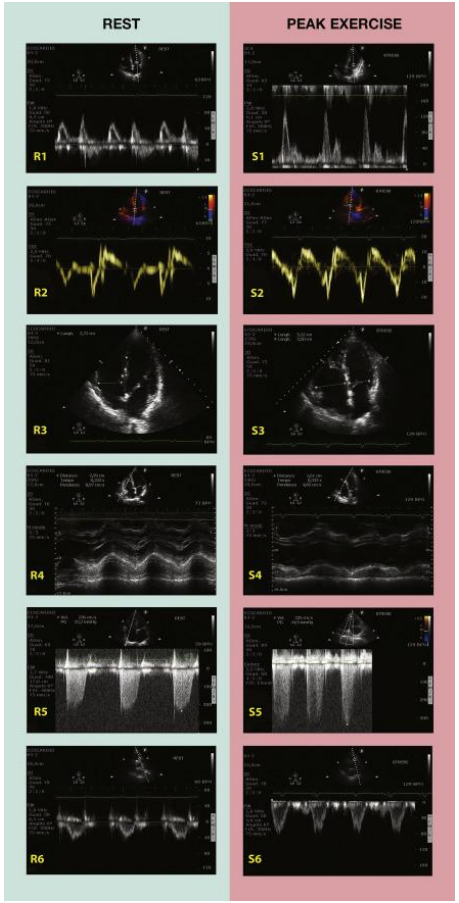




Micah R. Fisher et al. Am J Respir Crit Care 2009, Med Vol 179. pp 615–621,

Obrázek *z Interv Akut Kardiol 2018; 17(3): 164–168* a laskavostí MUDr. Martina Troubila

	Positive ExEcho N=43 (49%)	Negative ExEcho N=42 (48%)	P-value
Age (years)	54.7 ± 12	50.6 ± 13	NS
Race	White: 22 (51%) AA: 19 (44%) Other: 2 (5%)	White: 24 (57%) AA: 15 (36%) Other: 3 (7%)	NS
ANAs, %	Anti-centromere: 28% Anti-scl 70: 12% Anti-nucleolar ANA:26%	Anti-centromere: 32% Anti-scl 70: 20% Anti-nucleolar ANA:20%	NS
SSc type (limited scleroderma), %	60 %	68 %	NS
FVC % ± SD	81 ± 21	85 ± 22	NS
DLCO % ± SD	66 ± 22	65 ± 20	NS
FVC(%)/DLCO(%) ± SD	1.35 ± 0.48	1.44 ± 0.5	NS
Baseline 6MWT (m) ± SD	435 ± 104	455 ± 102	NS
Baseline RVSP (mm Hg)	33 ± 7	31 ± 6	NS
Time to follow up (years) ± SD	5 ± 2.4	4.6 ± 2.4	NS
Developed PH	**10 (23%)**	**3 (7%)**	**0.039**



DOI: https://doi.org/10.1016/j.echo.2018.02.004

Zátěžová echokardiografie může být použita k screeningu pacientů na přítomnost zátěžové plicní hypertenze.
V observační studii byla v průběhu 3 let diagnostikovaná PAH u 20% pacientů s SSc, u kterých byla identifikována zátěží indukovaná PH.

Chang B, Schachna L, White B, et al. Natural history of mild-moderate pulmonary hypertension and the risk factors for severe pulmonary hypertension in scleroderma. J Rheumatol 2006; 33:269.
Gargani L, Pignone A, Agoston G, et al. Clinical and echocardiographic correlations of exercise-induced pulmonary hypertension in systemic sclerosis: a multicenter study. Am Heart J 2013; 165:200.
Kovacs G, Avian A, Wutte N, et al. Changes in pulmonary exercise haemodynamics in scleroderma: a 4-year prospective study. Eur Respir J 2017; 50.

Studie 54 pacientů s SSc s rizikem rozvoje PAH prokázala u 44% zvýšení PASP o ≥20 mmHg po zátěži, z nichž 81% mělo klidovou nebo záětží indukovanou PH při RHC.

Steen V, Chou M, Shanmugam V, et al. Exercise-induced pulmonary arterial hypertension in patients with systemic sclerosis. Chest 2008; 134:146.

Abnormální zátěžová odpověď korelovala s abnormální spirometrií, přítomností ILD, vyšším věkem a srdeční dysfunkcí, což naznačuje, že zátěžová PH u SSc není specifická pro PAH.

Steen V, Chou M, Shanmugam V, et al. Exercise-induced pulmonary arterial hypertension in patients with systemic sclerosis. Chest 2008; 134:146.

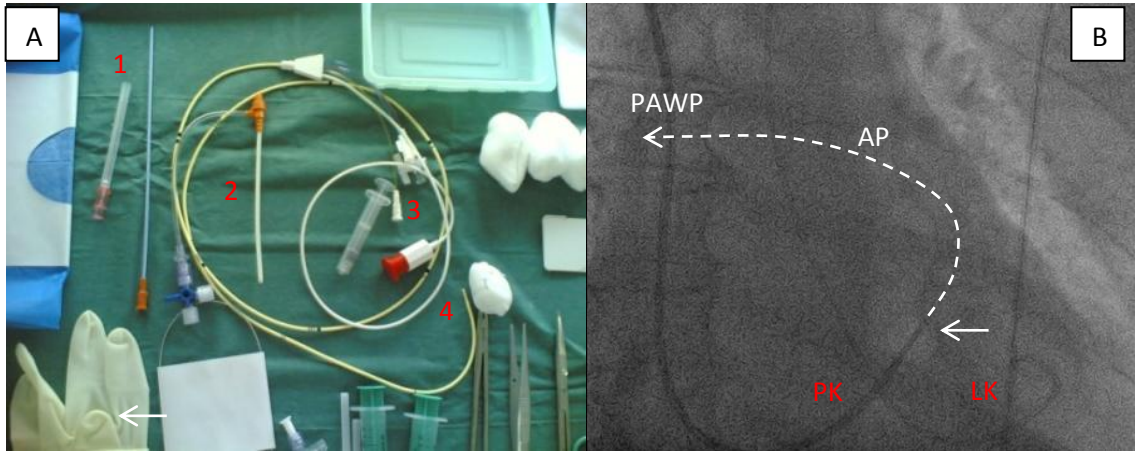
V další čtyřleté studii u pacientů s SSc, kteří neměli PAH na počáteční echokardiografii, se hemodynamika plicního cvičení časem zhoršila, ale pouze u 3% se vyvinula skutečná PAH.

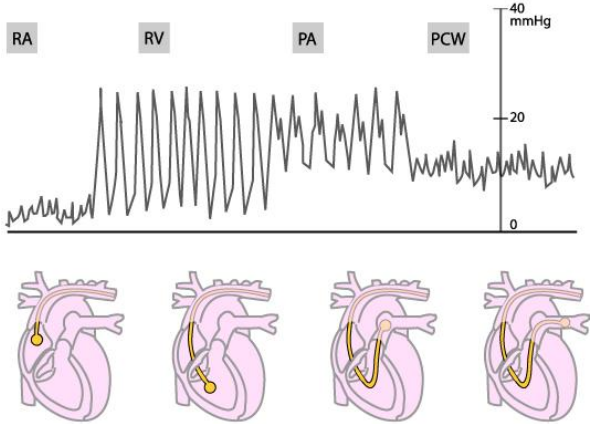
Kovacs G, Avian A, Wutte N, et al. Changes in pulmonary exercise haemodynamics in scleroderma: a 4-year prospective study. Eur Respir J 2017; 50.

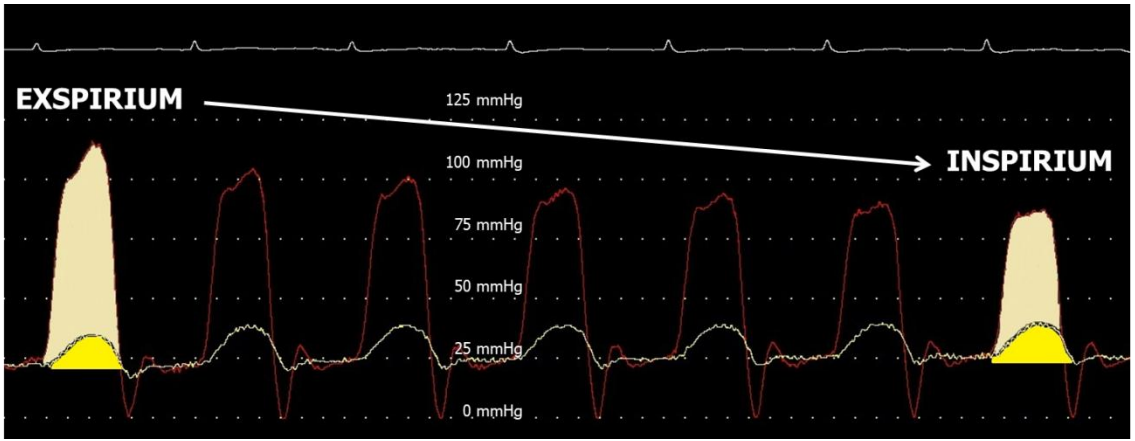
Prospektivní studie u 85 pacientů zjistila, že zátěží indukovaná PH může predikovat budoucí rozvoj PH. Většina pacientů však měla trvale pozitivní EE bez progrese do klidové PH.

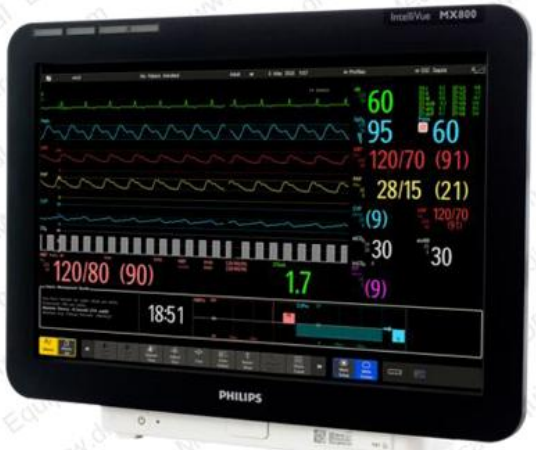
Quinn KA. Exercise Echocardiography Predicts Future Development of Pulmonary Hypertension in a High-Risk Cohort of Patients with Systemic Sclerosis. J Rheumatol 2020; 47:708.

Quinn KA. Exercise Echocardiography Predicts Future Development of Pulmonary Hypertension in a High-Risk Cohort of Scleroderma Patients. *Arthritis Rheumatol.* 2016; 68 (suppl 10).









ESC guidelines. Eur Heart J 2009;30:2493-2537
Jansa P. Chronicka plicni hypertenze. Cor Vasa 2011;53(3)

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).

Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG).

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Supplementary data

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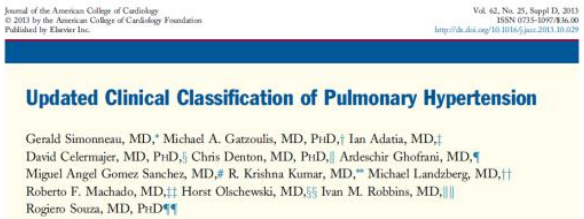
Authors/Task Force Members: Marc Humbert (France), Gabor Kovacs (Austria), Marius M. Hoeper (Germany), Roberto Badagliacca (Italy), Rolf M.F. Berger (Netherlands), Margarita Brida (Croatia), Jørn Carlsen (Denmark), Andrew J.S. Coats (United Kingdom), Pilar Escribano-Subias (Spain), Pisana Ferrari (Italy), Diogenes S. Ferreira (Brazil), Hossein Ardeschir Ghofrani (Germany), George Giannakoulas (Greece), David G. Kiely (United Kingdom), Eckhard Mayer (Germany), Gergely Meszaros (Hungary), Blin Nagavci (Germany), Karen M. Olsson (Germany), Joanna Pepke-Zaba (United Kingdom), Jennifer K. Quint (United Kingdom), Göran Rådegran (Sweden), Gerald Simonneau (France), Olivier Sitbon (France), Thomy Tonia (Switzerland), Mark Toshner (United Kingdom), Jean-Luc Vachery (Belgium), Anton Vonk Noordegraaf (Netherlands), Marion Delcroix (ERS Chairperson) (Belgium), Stephan Rosenkranz (ESC Chairperson) (Germany), and ESC/ERS Scientific Document Group

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**

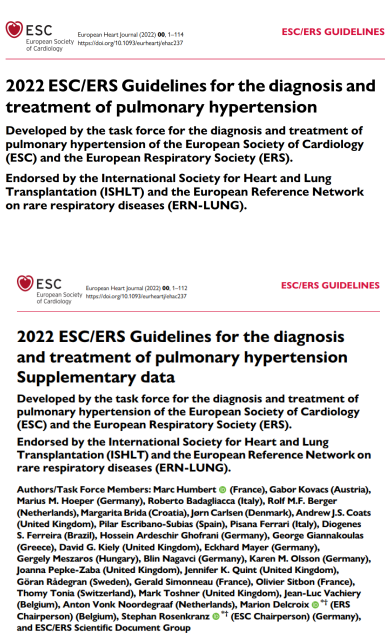
Plicní hypertenze - klasifikace a epidemiologie

Definition	Characteristics[a]	Clinical groups
Pre-capillary PH	**mPAP >20 mmHg** PAWP ≤15 mmHg PVR ≥3 WU	1. PAH 3. PH due to lung diseases 4. CTEPH 5. PH with unclear and/or multifactorial mechanisms
Ipc-PH	**mPAP >20 mmHg** PAWP >15 mmHg PVR <3 WU	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Cpc-PH	**mPAP >20 mmHg** PAWP >15 mmHg PVR ≥3 WU	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms



Prevalence of PAH in the general population
15–50 cases per million (0.0015–0.0050%)

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%



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

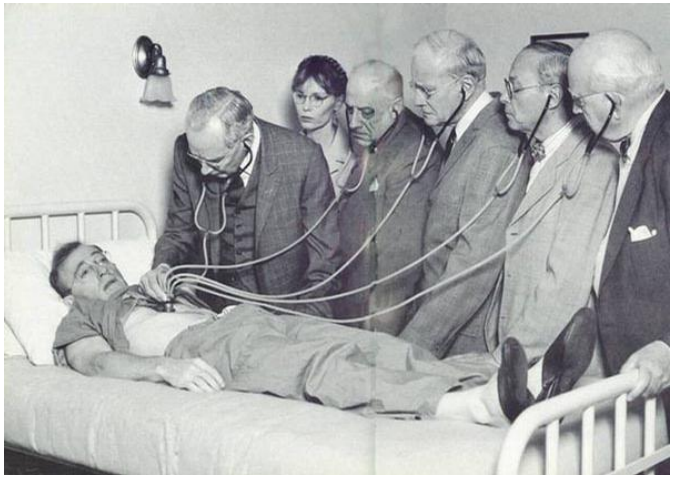
GROUP 3 PH associated with lung diseases and/or hypoxia
- 3.1 Obstructive lung disease or emphysema
- 3.2 Restrictive lung disease
- 3.3 Lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoventilation syndromes
- 3.5 Hypoxia without lung disease (e.g. high altitude)
- 3.6 Developmental 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

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

ZÁVĚRY PRO PRAXI



Plicní hypertenze (PH) je definována hemodynamicky zvýšením středního tlaku v plicnici nad 20 mmHg.

Nově definice PH obsahuje termín zátěžové plicní hypertenze

Hodnota PAWP diskriminující mezi prekapilární a postkapilární PH zůstala na hodnotě 15 mmHg

Z definice PH zmizel DPG a TPG, nově tam byla zařazena PVR s cut-off hodnotou 2 WU.

Definition	Haemodynamic characteristics
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Exercise PH	**mPAP/CO slope between rest and exercise >3 mmHg/L/min**

DĚKUJEME ZA POZORNOST

FAKULTNÍ NEMOCNICE OLOMOUC

KOMPLEXNÍ
KARDIOVASKULÁRNÍ CENTRUM
FAKULTNÍ NEMOCNICE OLOMOUC