

# **SOB: Does Pulmonary Hypertension Explain Everything ?**

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# Disclosures

Consultant

-Anylam

-J&J

-Bayer

-United Therapeutics

-Merck



# Case

## History of Present Illness

EG is a 47 yo woman with h/o

- Scleroderma
- HTN

Over last 6 months progressive DOE, has to stop after climbing 2 flights of stairs



# Case

## Physical Examination

144/74 95 20 98 92% O2 sat on RA

Skin: skin sclerosis, telangectasia on upper chest

Not elevated JVP, no carotid bruits

Lungs: clear to auscultation

Heart: S1,S2, no S3 very soft murmur at left lower sternal border c/w TR

Abdomen: soft, no tenderness, no HSM

Lower extremity: no edema no cyanosis.



# Case

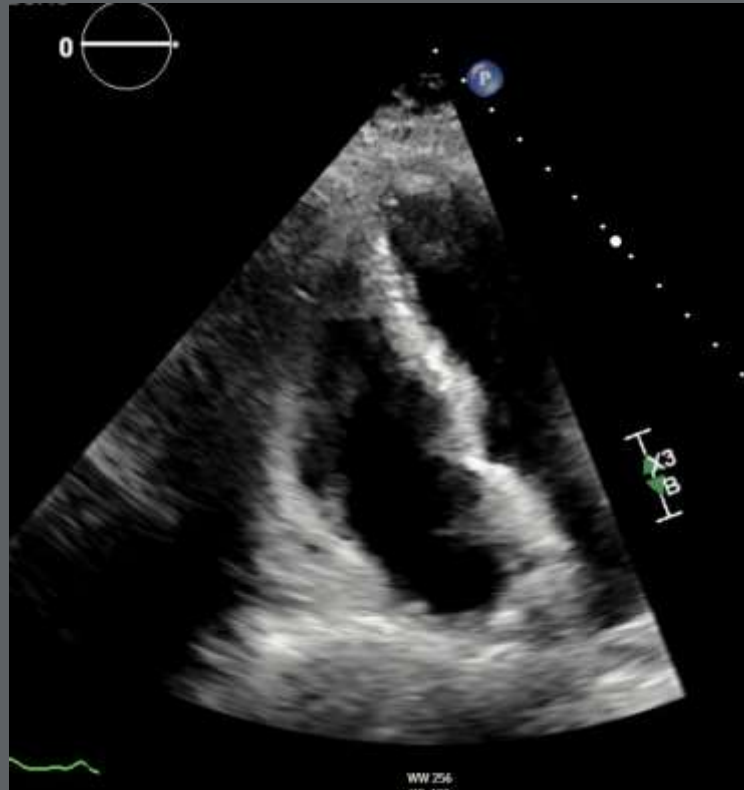
## Transthoracic Echocardiogram

- LVEF 65%, normal LV size
- No LVH
- Trace MR
- Normal RV size
- No RVH
- Normal RV function
- Normal LA and RA size
- Mild tricuspid regurgitation
- PASP 42 mm Hg



# Case 3

## Transthoracic Echocardiogram



# Laboratory Data

CBC



|     |    |      |
|-----|----|------|
| 142 | 95 | 23   |
| 3.6 | 24 | 1.10 |

Albumin 3.8

Total Bilirubin 0.8

AST/ALT 25/23

INR 1.0

BNP 68

ANA positive

Sci-70 Ab: positive



# Case

## 6 MWDT:

Patient walked 230 meters (700 feet), no stop. Sat went from 95 down to 89%. BORG scale went from 2 up to 9

**PFTs:** WNL

**Chest CT Scan:** WNL Lungs are clear. No mediastinal or axillary lymphadenopathy. No pulmonary embolism.





# Pulmonary Hypertension

## *Definition by Echocardiogram*

PA systolic < 40 mmHg: Normal

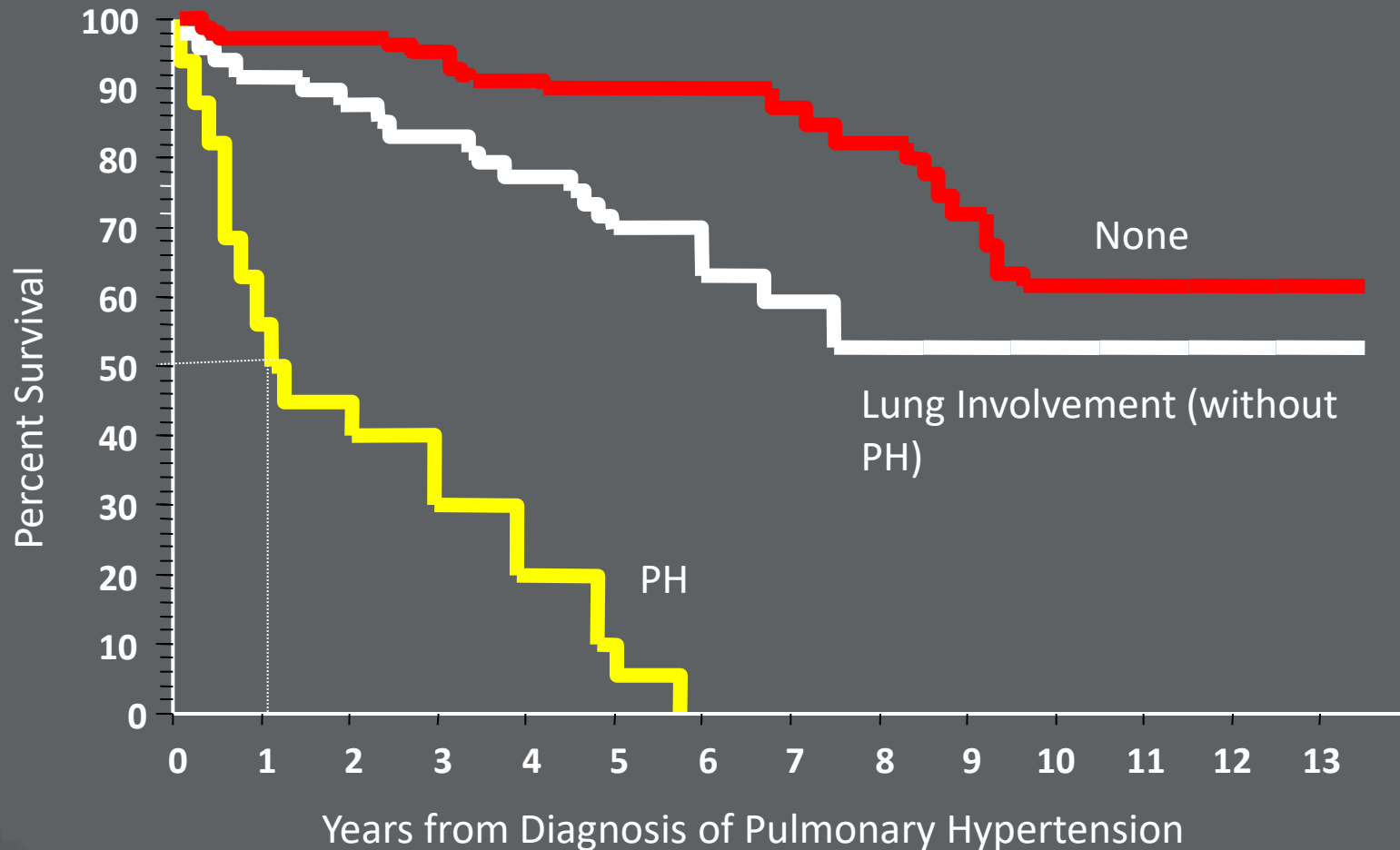
PA systolic 40-50mmHg: Mild

PA systolic 50-60mmHg: Moderate

PA systolic > 60mmHg: Severe



# Survival of Scleroderma Patients Based on Organ Involvement



# 2022 ESC/ERS Guidelines

## Clinical Classification of Pulmonary Hypertension

### 1. PAH

- 1.1 Idiopathic PAH
- 1.2 Heritable PAH
- 1.3 Drug- and toxin-induced PAH
- 1.4 PAH 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 long-term responders to calcium channel blockers
- 1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement
- 1.7 Persistent PH of the newborn syndrome

### 2. PH due to left heart disease

- 2.1 PH due to heart failure with preserved LVEF
- 2.2 PH due to heart failure with reduced LVEF
- 2.3 Valvular heart disease
- 2.4 Congenital/acquired cardiovascular conditions leading to post-capillary PH

### 3. PH due to lung diseases and/or hypoxia

- 3.1 Obstructive lung disease
- 3.2 Restrictive lung disease
- 3.3 Other lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoxia without lung disease
- 3.5 Developmental lung disorders

### 4. PH due to pulmonary artery obstructions

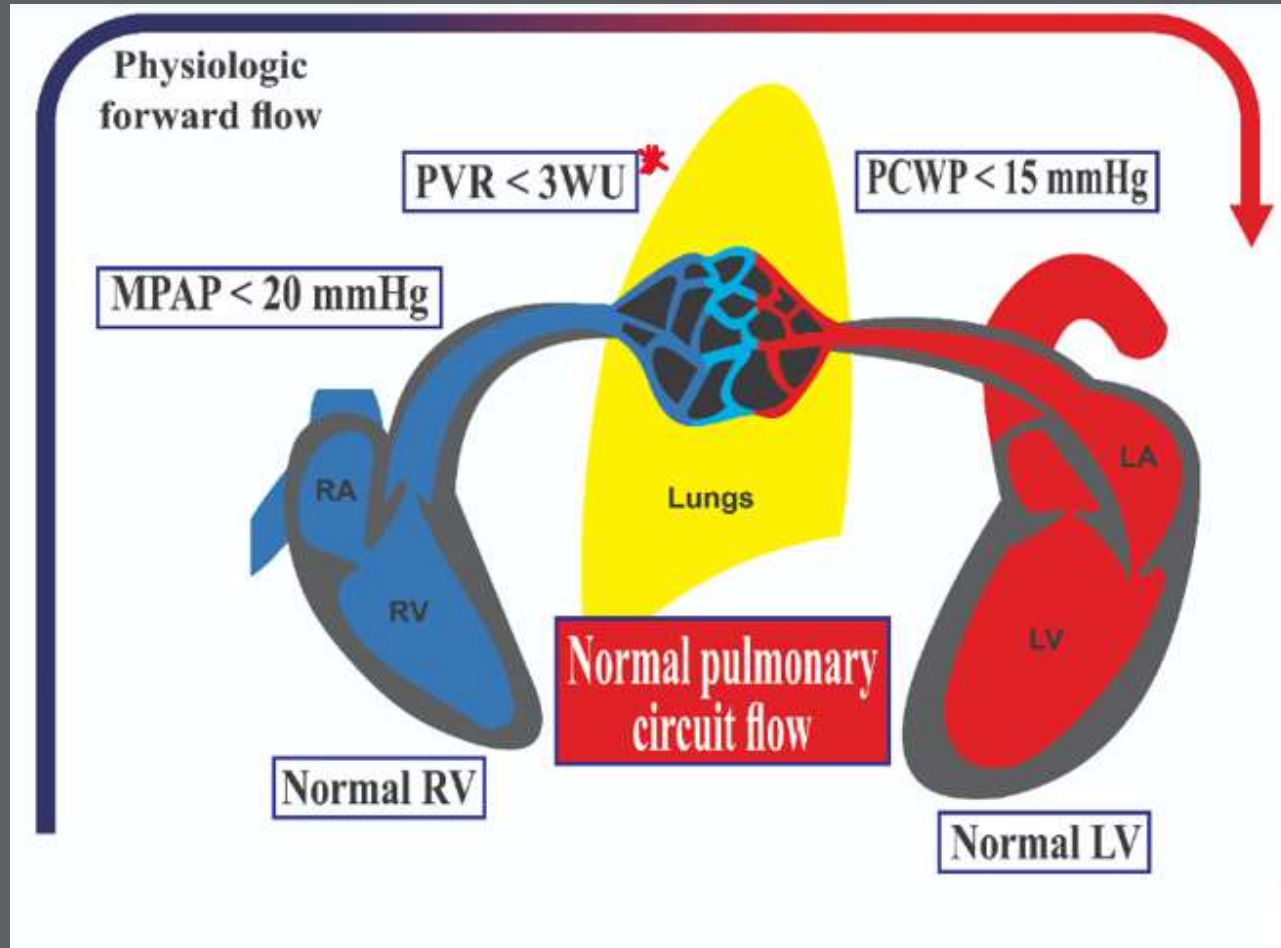
- 4.1 Chronic thromboembolic PH
- 4.2 Other pulmonary artery obstructions

### 5. PH with unclear and/or multifactorial mechanisms

- 5.1 Hematologic disorders
- 5.2 Systemic and metabolic disorders
- 5.3 Others
- 5.4 Complex congenital heart disease



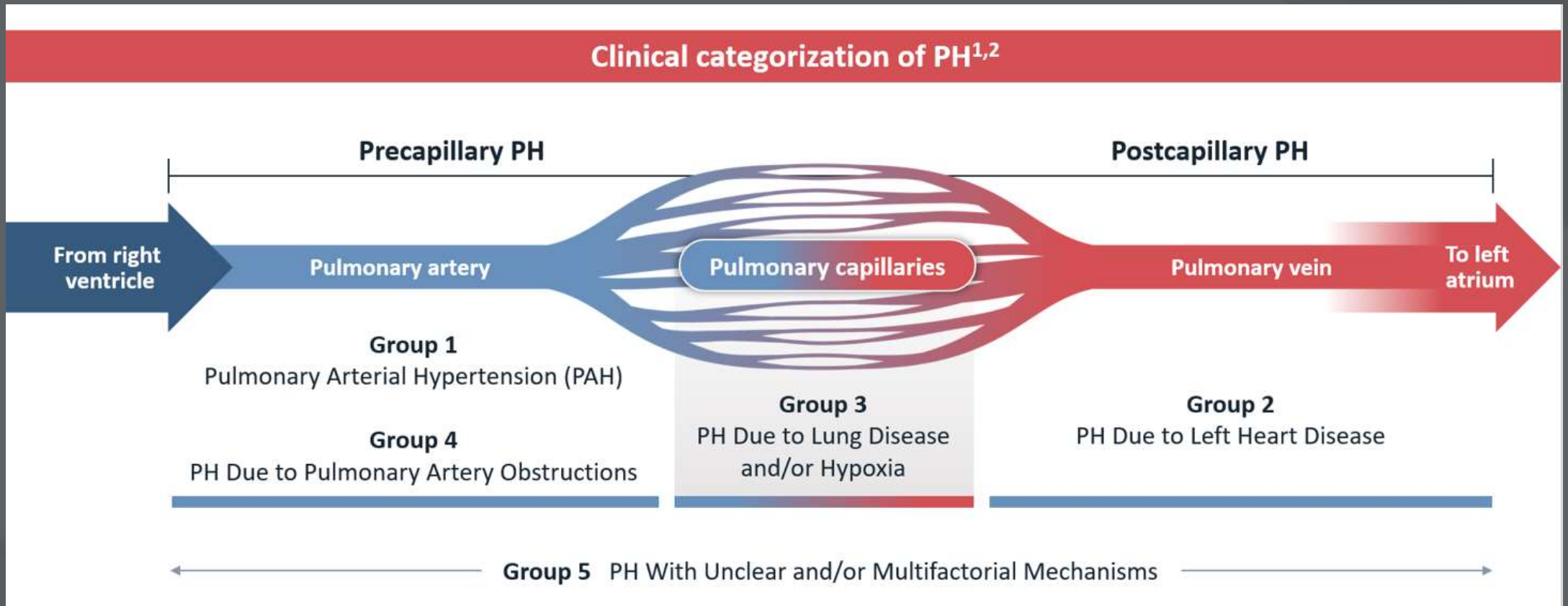
# Pulmonary Circuit Flow



Salem, M., Al-Saffar, F., & Hall, S. (2022). Management of Pulmonary Hypertension in Patients on Left Ventricular Assist Device Support. *Reviews in Cardiovascular Medicine*, 23(9), 308.



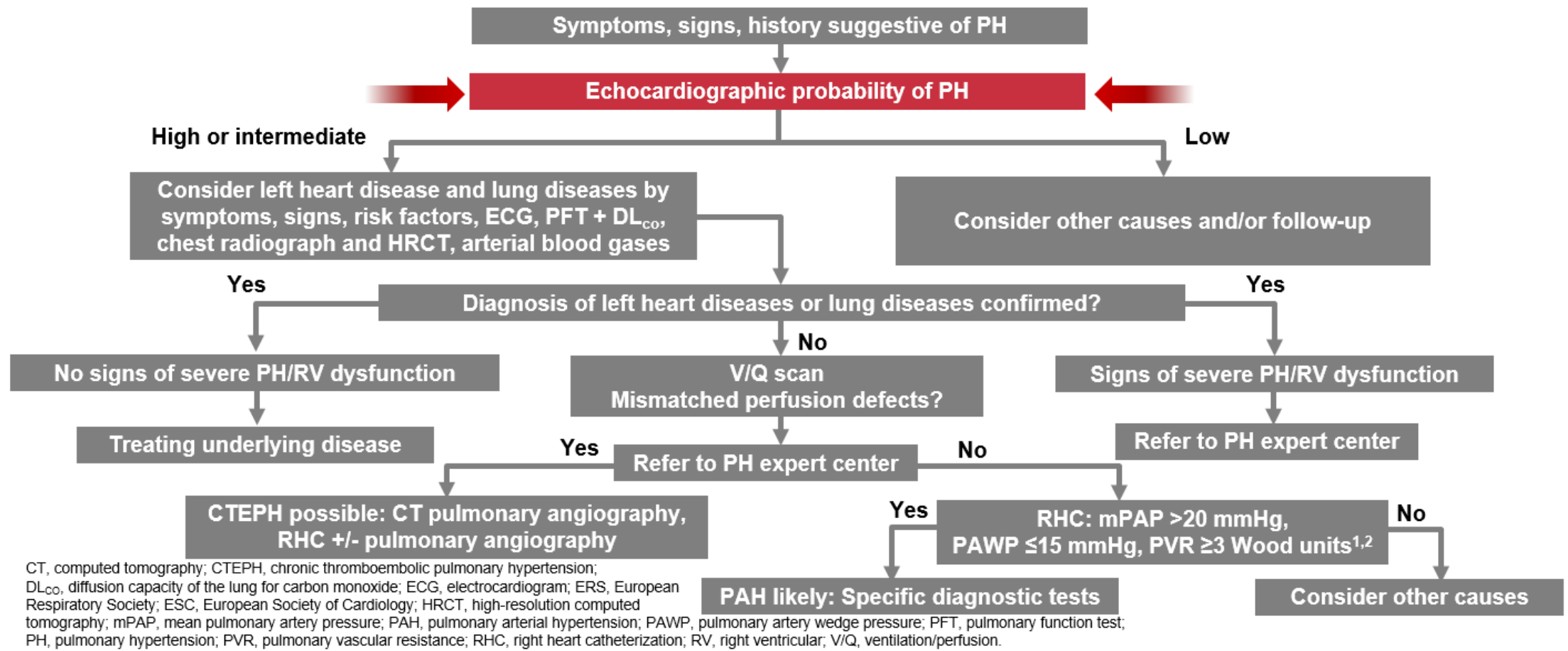
# PH is Categorized Based on its Underlying Cause



**References:** 1. Simonneau G, et al. *Eur Respir J.* 2019;53(1):1801913. 2. Kumar N, et al. Pulmonary hypertension. In: *Teaching Rounds: A Visual Aid to Teaching Internal Medicine Pearls on the Wards.* New York, NY: McGraw-Hill Education; 2016.



# How Echocardiography Can Guide the Identification of the Underlying Cause of PH <sup>1</sup>



# Echocardiography Is an Important Screening Tool for Patients Who Are at High Risk of PAH <sup>1,2</sup>

**Patients at high risk of PAH** include those with **SSc, CHD, or portal hypertension**, among others<sup>1</sup>



**Regular screening for PAH** is recommended for these patients<sup>1</sup>



**Echocardiography is a recommended screening tool** for high-risk patients<sup>1</sup>



***Right heart abnormality on echocardiography should trigger suspicion of PAH and proceed to comprehensive work-up, which may include RHC<sup>1,2</sup>***

1. 2  
2. Humbert M et al. Eur Heart J 2022;00:1-114



# *Early Diagnosis of PH Improves Prognosis*

- Data from large, national registries support that early diagnosis of PH is associated with better prognosis <sup>1-3</sup>
- Screening patients in high-risk groups for PAH is recommended to help with earlier diagnosis <sup>4,5,6</sup>
  - Transthoracic echocardiography is a useful non-invasive test that can be utilized to screen for PAH in high-risk patients <sup>6</sup>
- RHC is required to make the diagnosis of PAH <sup>4,5</sup>

1. Lau EM et al Nat Rev Cardiol 2015;12:143-155
2. Humbert M et al Circulation 2010;122:156-163
3. Benza RL et al. Circulation 2010;122:164-172
4. Humbert M et al. Eur Heart J 2022;00:1-114
5. Frost A et al. Eur Respir J 2019;53:1801904
6. Bossone E et al. J Am Soc Echocardiogr 2013;26:1-14





# Right Heart Catheterization to Assess Severity and Prognosis of PAH

RHC = 'Gold Standard Test'  
To establish severity and prognosis

Catheterization is required for every patient with suspected pulmonary hypertension



# Hemodynamic Definition of PH by Right Heart Catheterization

Normal mPAP  
8-16 mmHg at rest

PH  
mPAP > 20 mmHg at rest



# Key Definitions: PH

- RHC is required to make the diagnosis of PAH <sup>1,4</sup>

- **PH** refers to the presence of abnormally high pulmonary vascular pressure<sup>1,3</sup>

**Hemodynamic definition of PH<sup>1,2\*</sup>**

**mPAP >20 mmHg**

1. Humbert M et al. Eur Heart J 2022;00:1-114
2. Simonneau G et al. Eur Respir J 2019;53:1801913
3. McLaughlin VV et al. J Am Coll Cardiol 2015;65:1976-1997
4. Frost A et al. Eur Respir J 2019;53:1801904



# Over-Reliance on Doppler Estimation of PASP Detracts From Diagnostic of Echocardiography

- PASP  $\geq 40$  mmHg is consistent with PH <sup>1</sup>
- PASP is achieved by using continuous wave Doppler to determine TRV, which when applied to Bernoulli equation and added to estimated RAP provides an estimate for PASP <sup>1</sup>
- Evidence suggests that in nearly 50% of cases, PASP by Doppler differs by  $> 10$  mmHg from invasive findings <sup>2-4</sup>



1. Roberts JD, Forfia PR. *Pulm Circ* 2011;1:160-181
2. Lau EM, et al. *Nat Rev Cardiol*. 2015;12:143-155
3. Fisher MR, et al. *Am J J Respir Crit Care Med*. 2009;179:615-621
4. Rich JD, et al. *Chest* 2011;139:988-993



# Pulmonary Artery Catheter Used for RHC

## **Pulmonary artery catheter used for RHC**

- 110 cm in length
- Balloon at tip
- Thermistor near tip
- Lumens for pressure measurements
- Inserted via:
  - Right internal jugular vein
  - Left subclavian vein
  - Femoral vein



# Case Right Heart Catheterization

|   | Baseline Room Air | Normal values     |
|---|-------------------|-------------------|
| Blood Pressure (BP) (mmHg)                              | 112/52 (72)       | 110-130/60-80     |
| Heart Rate (HR) (bpm)                                   | 82                | 60-90             |
| Right Atrial Pressure (RAP) mmHg                        | 6                 | 0-6               |
| Pulm Artery Pressures (PAP) mmHg                        | 34/11             | < 30/<15          |
| Mean PAP (mPAP)   | 19                | < 20              |
| Pulm Capillary Wedge Pressure (PCWP)                    | 8                 | < 15              |
| Transpulmonary Gradient                                 | 11                | < 12              |
| PA saturation (%)                                       | 65                | 65-75             |
| AO saturation (%)                                       | 96                | >90               |
| Cardiac Output by thermodilution (L/min)                | 4.8               | 5-6               |
| Cardiac Index by thermodilution (L/min/m <sup>2</sup> ) | 2.6               | 2.5-4.2           |
| Cardiac Output by Fick (Fick CO) (L/min)                | 4.7               | 5-6               |
| Cardiac Index by Fick (Fick CI) (L/min/m <sup>2</sup> ) | 2.5               | 2.5-4.2           |
| Systemic Vascular Resistance (SVR) (WU/dynes)           | 1123              | 10-16.25/800-1300 |
| Pulm Vascular Resistance (PVR) (WU/dynes)               | 2.34              | <2/<160           |



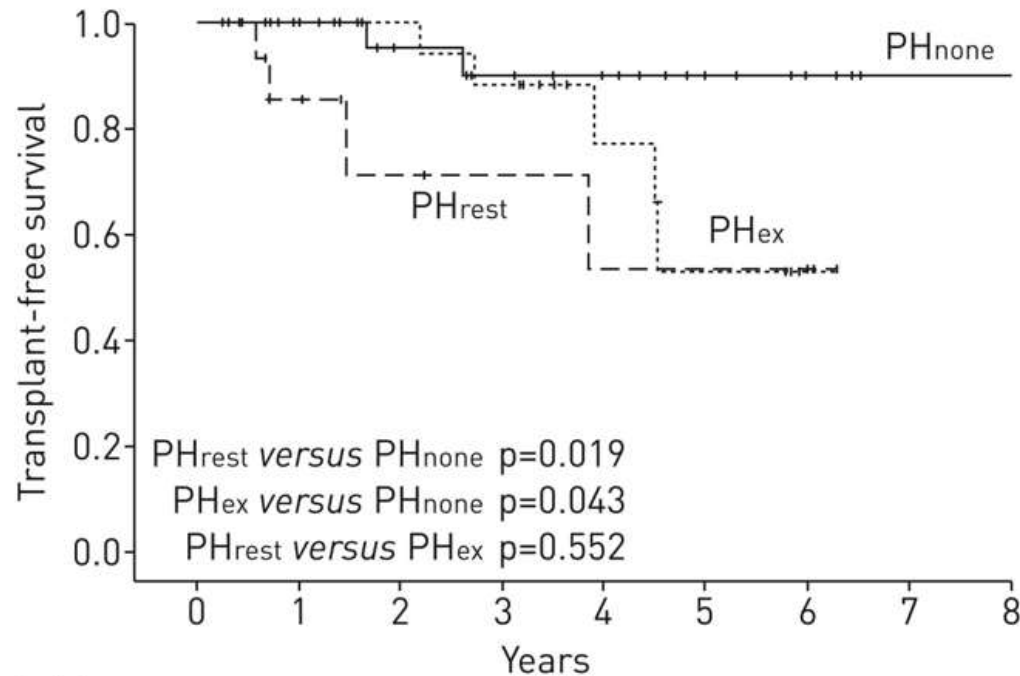
# *Early Diagnosis of PH Improves Prognosis*

- Data from large, national registries support that early diagnosis of PH is associated with better prognosis <sup>1-3</sup>
- Screening patients in high-risk groups for PAH is recommended to help with earlier diagnosis <sup>4,5,6</sup>

1. Lau EM et al Nat Rev Cardiol 2015;12:143-155
2. Humbert M et al Circulation 2010;122:156-163
3. Benza RL et al. Circulation 2010;122:164-172
4. Humbert M et al. Eur Heart J 2022;00:1-114
5. Frost A et al. Eur Respir J 2019;53:1801904
6. Bossone E et al. J Am Soc Echocardiogr 2013;26:1-14



# Prognostic relevance of pulmonary hemodynamic factors during exercise in systemic sclerosis



| Patients at risk n | 0  | 1  | 2  | 3  | 4  | 5 | 6 | 7 | 8 |
|--------------------|----|----|----|----|----|---|---|---|---|
| PH <sub>rest</sub> | 17 | 10 | 5  | 4  | 3  | 3 | 2 | 0 | 0 |
| PH <sub>ex</sub>   | 28 | 23 | 17 | 15 | 7  | 4 | 4 | 0 | 0 |
| PH <sub>none</sub> | 27 | 23 | 18 | 15 | 12 | 7 | 4 | 1 | 1 |

Among 72 SSc patients with normal mPAP at rest, an excessive increase in mPAP during exercise may indicate an early stage of pulmonary vasculopathy, associated with reduced survival similar to resting pulmonary hypertension patients.





# Exercise-Induced PH

1. Exercise-induced PH is a cause of decreased exercise capacity, and is associated with a decreased life expectancy <sup>[1]</sup>
2. Exercise-induced PH may precede the development of resting PH in a proportion of patients at risk, such as in those idiopathic PAH, sclerodermia, and HFpEF
3. Exercise-induced PH is thought to represent an intermediate stage between normal pulmonary pressure and overt PH <sup>[2],[3]</sup>
4. About 20% percent of cases with exercise-induced pulmonary hypertension progress to overt PAH in three years <sup>[2]</sup>
5. Exercise-induced PH is caused either by pulmonary vasoconstriction, pulmonary vascular remodeling, or by increased upstream transmission of pulmonary venous pressure.

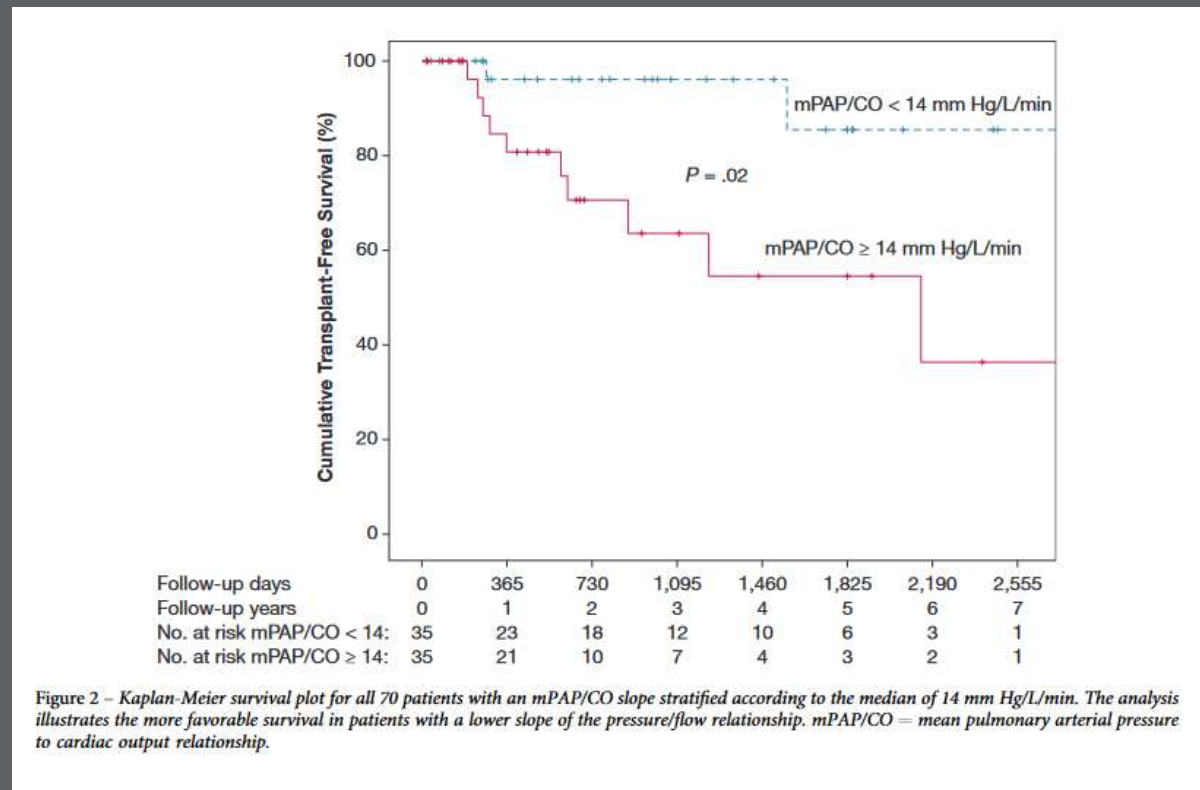
1. Hasler ED et al. Chest 2016; 150 (1): 57-67

2. R. Condliffe, et al. Am. J. Respir. Crit. Care Med., 179 (2009), pp. 151-157

3. J.J. Tolle et al. Circulation, 118 (2008), pp. 2183-2189



# Pressure-Flow During Exercise Catheterization Predicts Survival in Pulmonary Hypertension



# Hemodynamic Definitions of PH

|  | 2022 ESC/ ERS Guidelines <sup>1</sup>                 | 6th WSPH <sup>2</sup>                               | Clinical Group(s) <sup>1,2</sup>  |
|--|---|---|---|
| <b>PH</b>  | mPAP >20 mmHg   | mPAP >20 mmHg                                       | All   |
| <b>Precapillary PH</b>                                     | mPAP >20 mmHg<br>PAWP ≤15 mmHg<br>PVR >2 Wood units   | mPAP >20 mmHg<br>PAWP ≤15 mmHg<br>PVR ≥3 Wood units | 1. PAH<br>3. PH due to lung diseases<br>4. PH due to pulmonary artery obstructions<br>5. PH with unclear and/or multifactorial mechanisms |
| <b>Isolated post-capillary PH</b>                          | mPAP >20 mmHg<br>PAWP >15 mmHg<br>PVR ≤2 Wood units   | mPAP >20 mmHg<br>PAWP >15 mmHg<br>PVR <3 Wood units | 2. PH-LHD<br>5. PH with unclear and/or multifactorial mechanisms  |
| <b>Combined post-capillary and precapillary PH (CpcPH)</b> | mPAP >20 mmHg<br>PAWP >15 mmHg<br>PVR >2 Wood units   | mPAP >20 mmHg<br>PAWP >15 mmHg<br>PVR ≥3 Wood units | 2. PH-LHD<br>5. PH with unclear and/or multifactorial mechanisms  |
| <b>Exercise PH</b>   | mPAP/CO slope between rest and exercise >3 mmHg/L/min | —   |   |

\*Hemodynamics assessed by right heart catheterization.

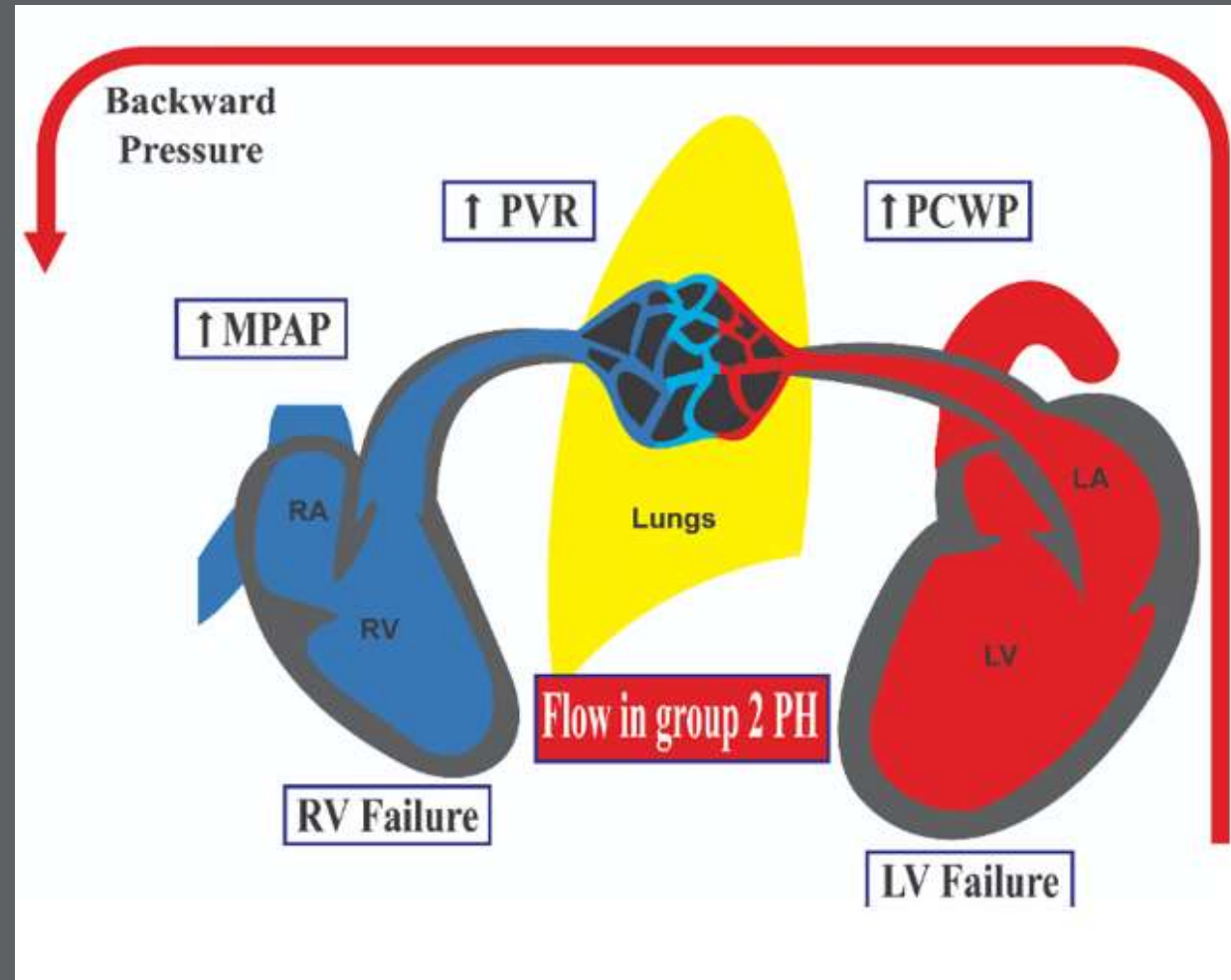
CO, cardiac output; ERS, European Respiratory Society; ESC, European Society of Cardiology; mPAP, mean pulmonary artery pressure; PAH, pulmonary arterial hypertension; PAWP, pulmonary artery wedge pressure; PH, pulmonary hypertension; PH-LHD, pulmonary hypertension associated with left heart disease;

PVR, pulmonary vascular resistance; WSPH, World Symposium on Pulmonary Hypertension.

1. Humbert M, et al. *Eur Heart J.* 2022;00:1-114. 2. Simonneau G, et al. *Eur Respir J.* 2019;53:1801913.



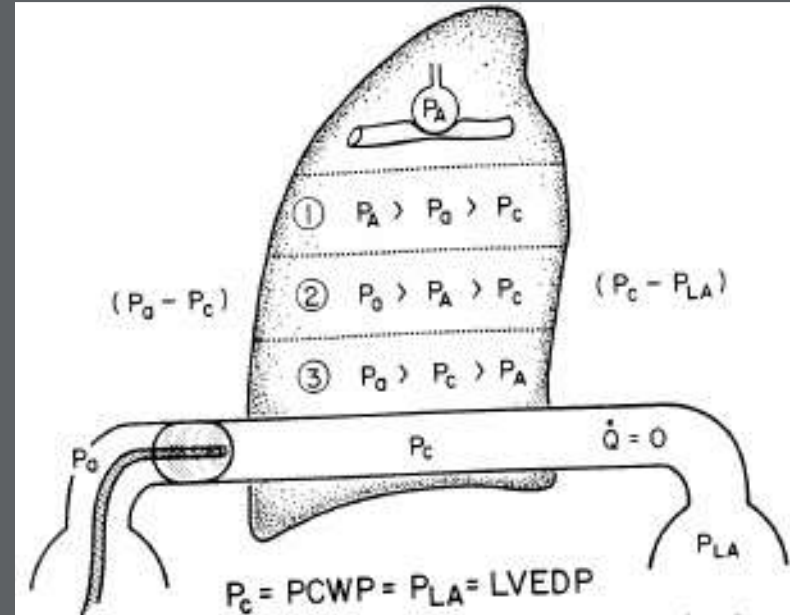
# Upstream Effects of LV Failure



Salem, M., Al-Saffar, F., & Hall, S. (2022). Management of Pulmonary Hypertension in Patients on Left Ventricular Assist Device Support. *Reviews in Cardiovascular Medicine*, 23(9), 308.



# Pulmonary Capillary Wedge Pressure



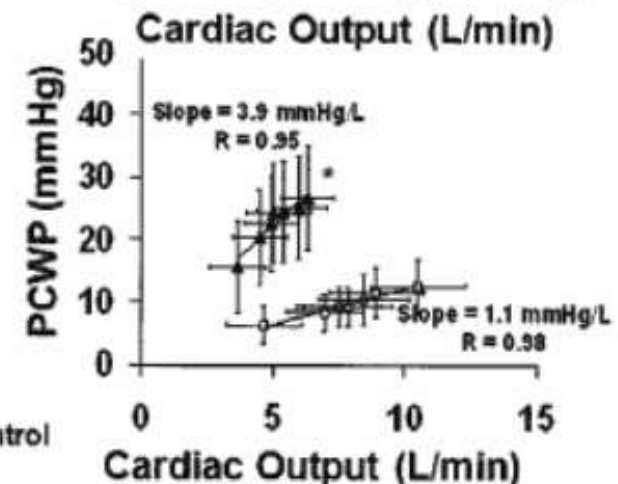
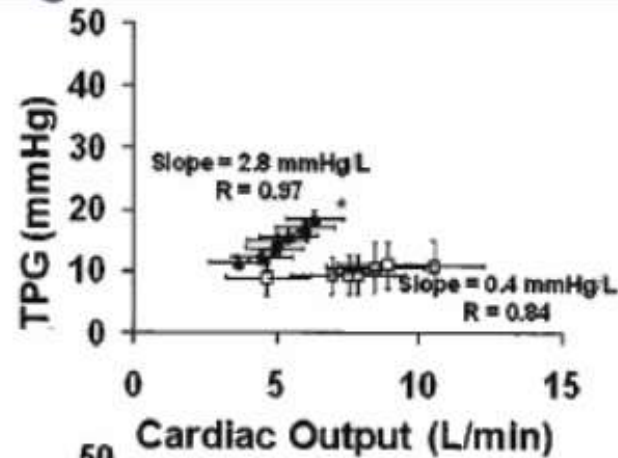
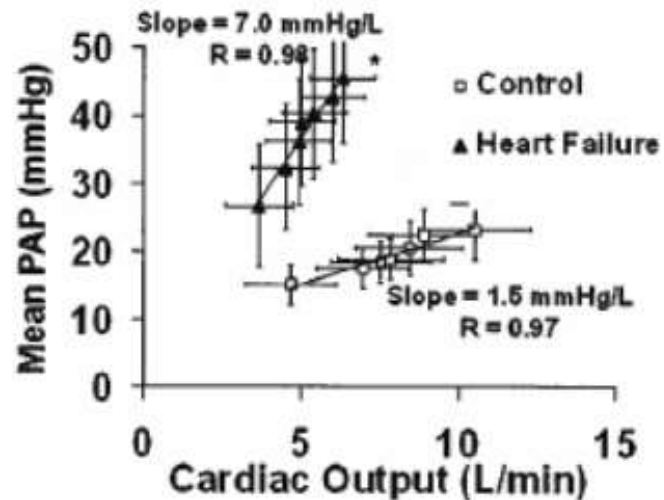
When inflated the balloon impacts into a small PA branch. In this position the balloon stops the flow, and the catheter tip senses pressure transmitted backward through the static column of blood

PCWP = closely reflects LA and left ventricular end-diastolic pressures



# Pressure Flow Relationships in the Pulmonary Vasculature During Exercise

**N=60 consecutive LVSD patients**  
Age  $60 \pm 12$ yo, NYHA II-III, standard therapy  
Excluded WHO-Group 1, 3-5 PH  
**N=19 age + sex matched normal controls**  
Preserved exercise capacity



Submitted, Under Review

\* $p < 0.01$  vs. control



# Cardio-metabolic bike exercise



# Case Right Heart Catheterization

|                            | Baseline<br>Room Air | Bike exercise<br>25 Watts | Bike exercise<br>100 Watts | Ref values<br>(at rest) |
|----------------------------|----------------------|---------------------------|----------------------------|-------------------------|
| BP                         | 112/52 (72)          | 131/74 (98)               | 160/86 (120)               | 110-130/60-80           |
| HR (ppm)                   | 82                   | 112                       | 135                        | 60-90                   |
| RAP (mmHg)                 | 6                    | 10                        | 12                         | 0-6                     |
| PA (mmHg)                  | 34/11                | 44/20                     | 66/25                      | < 35/<15                |
| PA mean (mmHg)             | 19                   | 32                        | 39                         | < 20                    |
| PCWP (mmHg)                | 8                    | 14                        | 16                         | < 15                    |
| TPG (mmHg)                 | 11                   | 18                        | 23                         | <12                     |
| PA saturation (%)          | 65                   | 48                        | 32                         | 65-75                   |
| AO saturation (%)          | 96                   | 95                        | 93                         | > 90                    |
| CO/CI by Fick (L/min)      | 4.7/2.5              | 7.4/3.93                  | 9.8/5.21                   | 5-6/2.5-4.2             |
| CO/CI by TD (L/min)        | 4.8/2.6              | 7.9/4.20                  | -                          | 5-6/2.5-4.2             |
| SVR (WU/dynes)             | 14.04/1123           | 11.90/952                 | 11.02/882                  | 800-1300                |
| PVR (WU/dynes)             | 2.34/187             | 2.43/195                  | 2.35/188                   | < 2                     |
| $\Delta$ mPA/ $\Delta$ CO  |                      |                           | 4.50                       | <3                      |
| $\Delta$ PCWP/ $\Delta$ CO |                      |                           | 1.56                       | <2                      |





# Case

## Right Heart Catheterization

$$\Delta mPA/\Delta CO = \frac{\text{mean PA max} - \text{mean PA baseline}}{\text{CO max} - \text{CO baseline}} = \frac{39 - 19}{9.8 - 4.7} = \frac{20}{5.1} = 4.50 (< 3)$$

$$\Delta PCWP/\Delta CO = \frac{\text{mean PCWP max} - \text{mean PCWP baseline}}{\text{CO max} - \text{CO baseline}} = \frac{16 - 8}{9.8 - 4.7} = \frac{8}{5.1} = 1.56 (< 2)$$



# Case

Patient started on Endothelin Receptor Antagonist + PD5-inhibitor

Reevaluation 6 months later:

**-6 MWDT:**

Patient walked 430 meters (1200 feet), no stop. Sat went from 96 down to 92%. BORG scale went from 0 up to 2

**-NYHA Class I**



# Exercise-induced PH

## Historical perspective

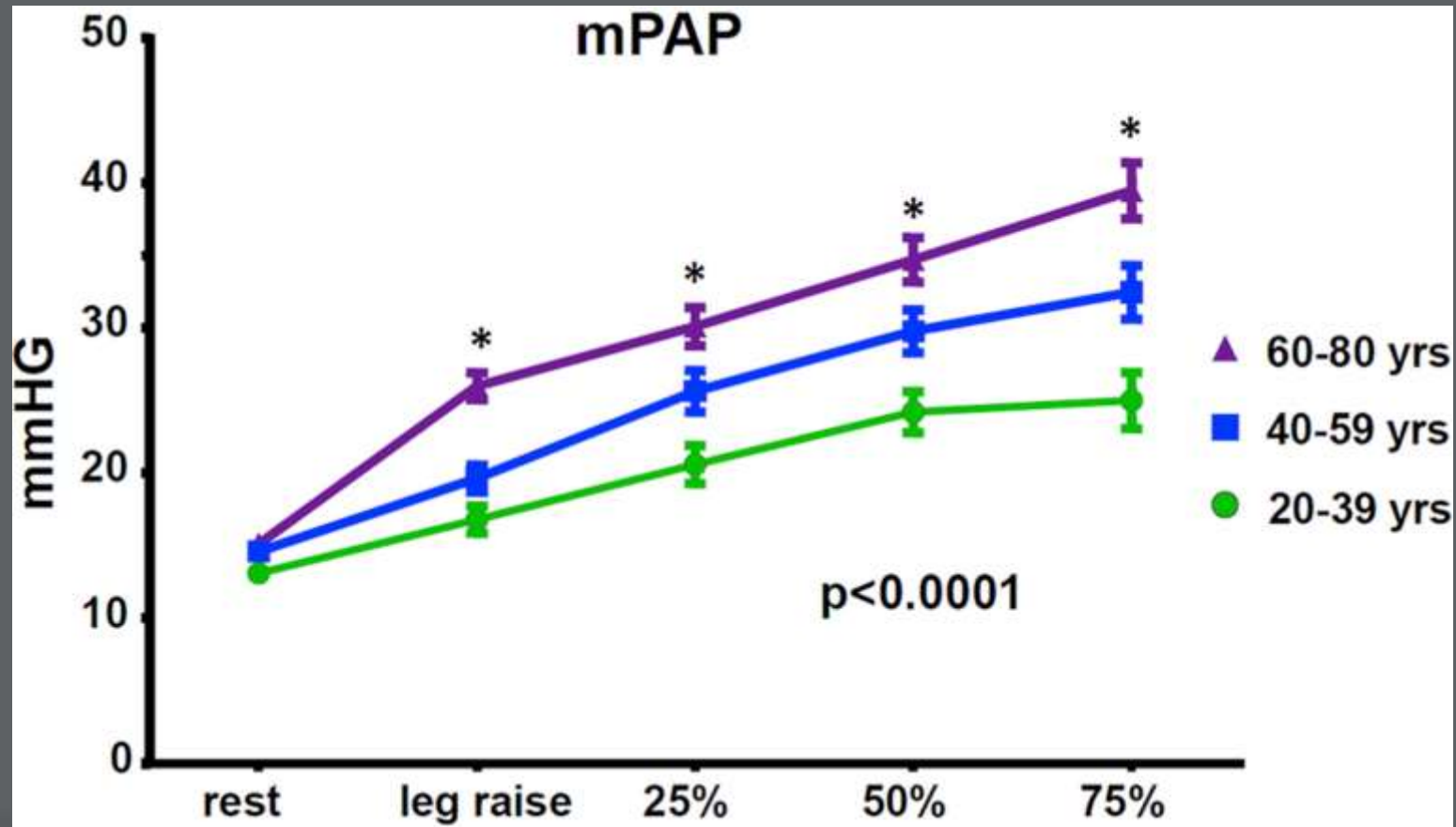
- At the first WHO meeting on primary PH in 1973 [3] it was postulated that “mean PAP (mPAP) does not normally exceed 30 mmHg during exercise” [1]
- This assumption led to the introduction of “exercise PH” defined as mPAP >30 mmHg on effort in the ESC guidelines on PH in 2004 [2]
- A few years later, it was recognised that exercise hemodynamics are strongly dependent on the level of exercise and age and that even healthy subjects frequently exceeded this threshold at high exercise levels [3,4]
- As a consequence, the term exercise PH was abandoned from the following ESC/European Respiratory Society (ERS) PH guidelines [5,6]
- Most recent ESC/ERS PH guidelines, published in August 2022, re-introduced exercise PH as part of the hemodynamic definitions of PH [7]

1. Hatano S, Strasser T, World Health Organization. <https://apps.who.int/iris/handle/10665/39094> 2. Galiè N, et al. Eur Heart J 2004; 25: 2243–2278.  
3. Kovacs G. et al. Eur Respir J 2009; 34: 888–894 4. Kovacs G, et al. Eur Respir J 2012; 39: 319–328. 5. Galie N, et al. Eur Heart J 2009; 30: 2493–2537.  
6. Galiè N, et al. Eur Respir J 2015; 46: 903–975. 8. Humbert M, et al. Eur Heart J 2022; 43: 3618–3731.



# Exercise-induced PH

## Exercise mPAP is age-related



# Exercise-induced PH

## Meta-analysis [1]

- Hemodynamic data of 1187 healthy subjects from 47 studies stratified by sex, age, geographic origin, body position and exercise level [1]
- Meta-analysis confirmed that mPAP during exercise depended on age:
  - Subjects aged  $\geq 50$  years have higher mPAP values compared with subjects aged  $< 50$  years ( $29 \pm 8$  vs.  $19 \pm 5$  mmHg,  $p < 0.001$ )
  - Subjects aged  $\geq 50$  years very frequently attained mPAP  $> 30$  mmHg at maximum exercise levels
  - Even 20% of the young subjects attained mPAP values  $> 30$  mmHg at maximum exercise, although this was associated with very high CO
- These findings showed that it is not possible to define an upper limit of exercise mPAP without relating it to CO [1]



# Exercise induced PH

## 2<sup>nd</sup> meta-analysis [1]

250 subjects from 11 studies, the mPAP/CO slope was positive in every single case and showed a significant increase with age

Mean values were:

- $0.8 \pm 0.4$  WU (ULN 1.6 WU) in subjects around 30 years of age
- $1.6 \pm 0.2$  WU (ULN 2.1 WU) in subjects around 50 years of age
- $2.4 \pm 0.5$  WU (ULN 3.3 WU) in subjects around 70 years of age

mPAP/CO slope  $>3$  WU can be considered as pathologic in most healthy subjects, even among the elderly



# Exercise-Induced PH in HFpEF: Translating Pathophysiological Concepts Into Clinical Practice

## Question

- **How much PAWP increase during exercise are considered pathological ?**

The upper limit of normal of PAWP during exercise is generally thought to be 15 -20 mm Hg, but higher values can be recorded in athletes and elderly subjects. [1-3]

Some consider 20 mm Hg a reasonable upper limit of normal.[4]

A higher cutoff value of 25 mm Hg has been proposed for the diagnosis of heart failure. [5-6]

1. Naeije R et al. Am J Respir Crit Care Med, 187 (6) (2013), pp. 576-583
2. Lewis GD, et al. Circulation, 128 (2013), pp. 1470-1479
3. Kovacs G et al. Eur Respir J, 39 (2) (2012), pp. 319-328

4. Oliveira RK *et al.* Eur Respir J, 47 (4) (2016), pp. 1179-1185.
5. Borlaug BA, et al Circ Heart Fail, 3 (5) (2010), pp. 588-595
6. Andersen MJ, *et al.* Circ Heart Fail, 5 (4) (2012), pp. 444-451

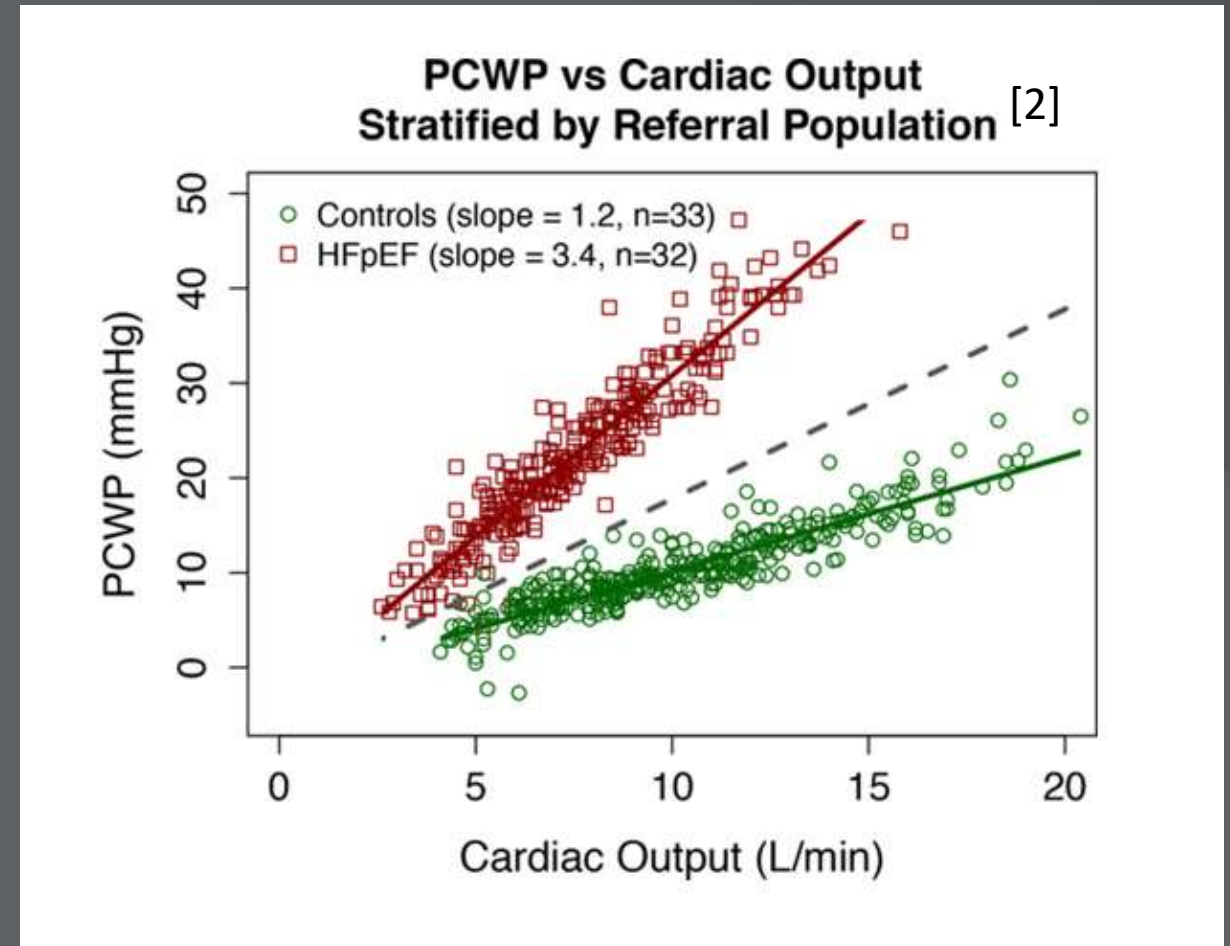


# Exercise induced PH

Pulmonary arterial wedge pressure (PAWP)/CO slope ranged from :

$0.3 \pm 0.2$  WU (ULN 0.6 WU) in subjects around 30 years old

to  $1.4 \pm 0.2$  WU (ULN 1.8 WU) in 70-year-old subjects <sup>[1]</sup>



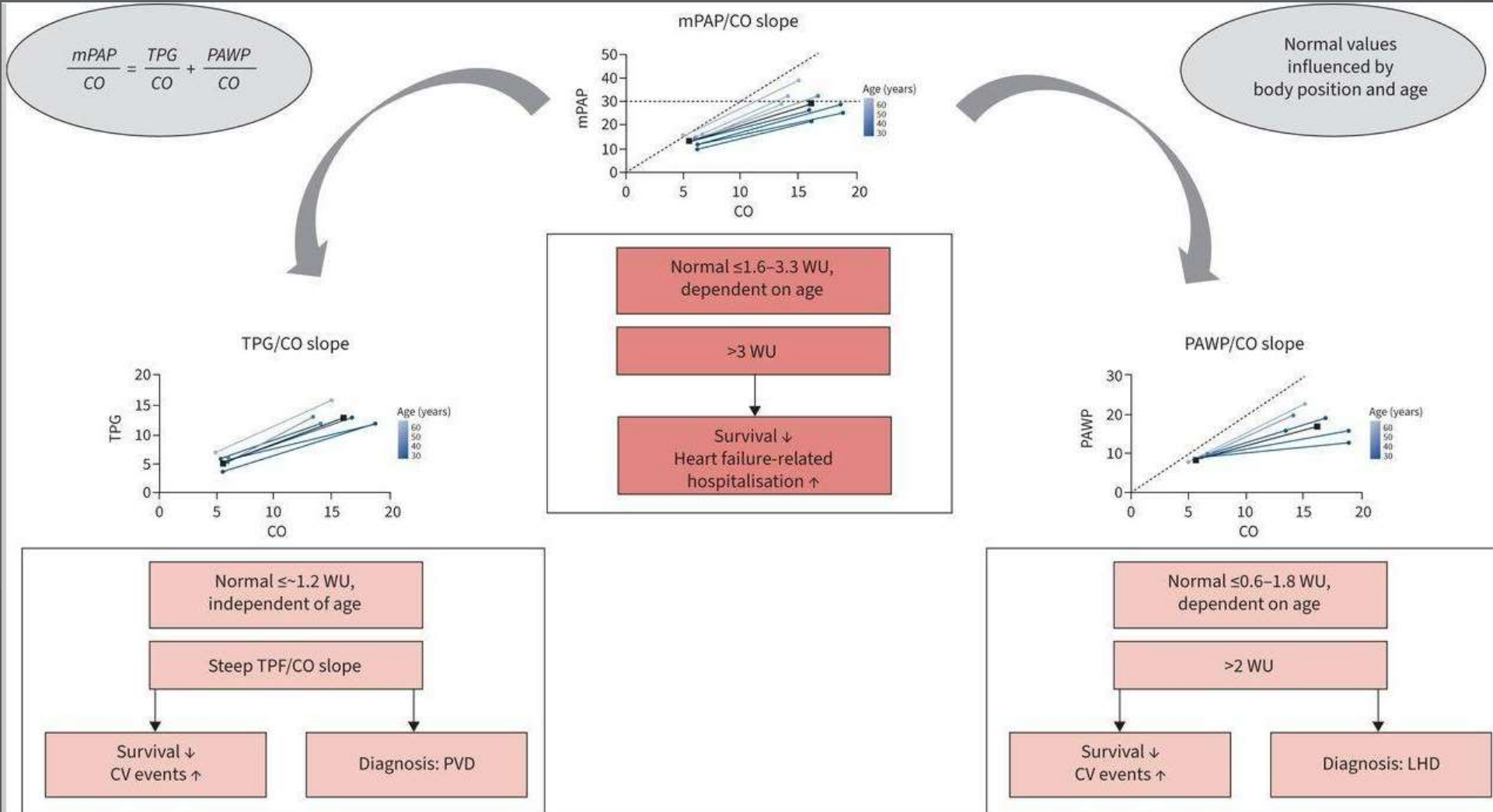
1. Zeder K, Banfi C, Steinrissler-Alex G, et al. Eur Respir J 2022; 60: 2103181

2. Eisman AS, Shah RV, Dhakal BP, et al. Circ Heart Fail 2018; 11: e004750.





# Exercise induced PH



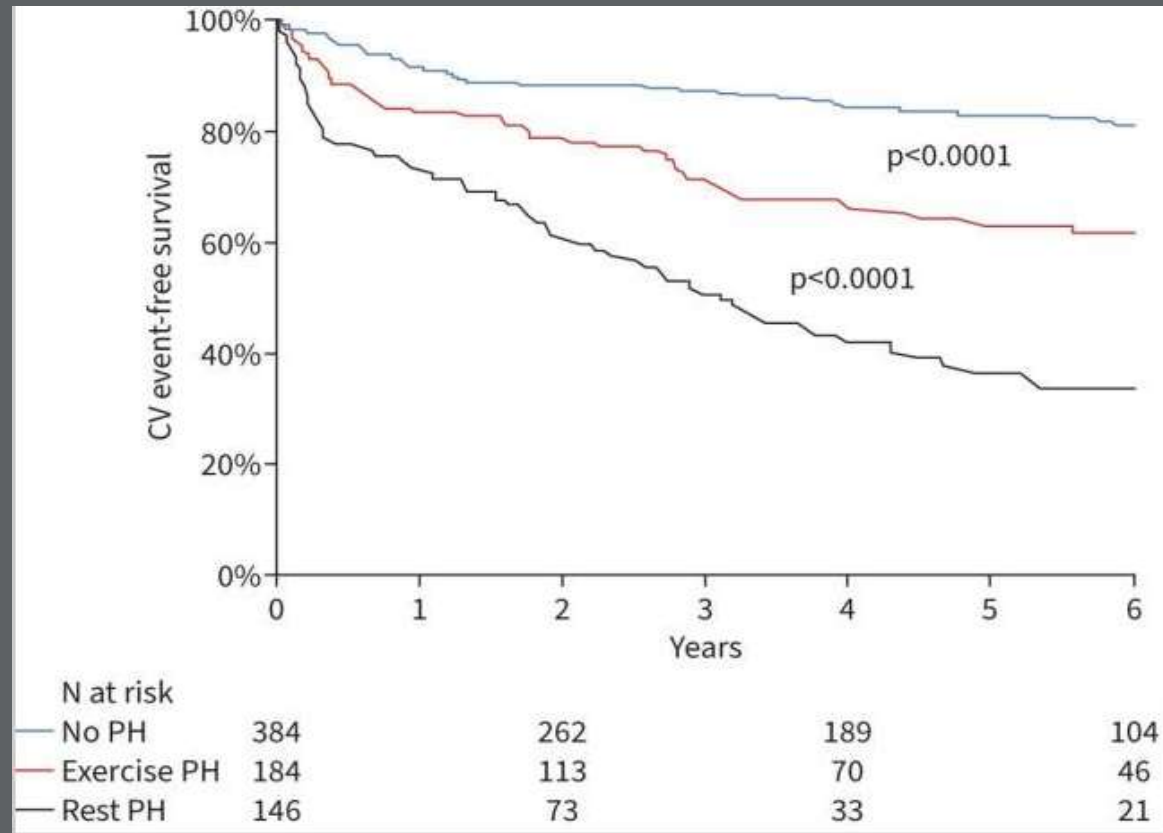
# Exercise pulmonary hypertension

## Summary

- Exercise mPAP is age-related and frequently exceeds 30 mmHg, especially in elderly individuals, which makes it difficult to define normal mPAP values during exercise
- The mPAP/CO slope emerged as a simple and consistent variable characterising pulmonary hemodynamic changes during exercise



# Prognostic relevance of pulmonary hemodynamics during exercise [1]



Cardiovascular (CV) event-free survival among individuals with dyspnea by PH status.



# Exercise pulmonary hypertension

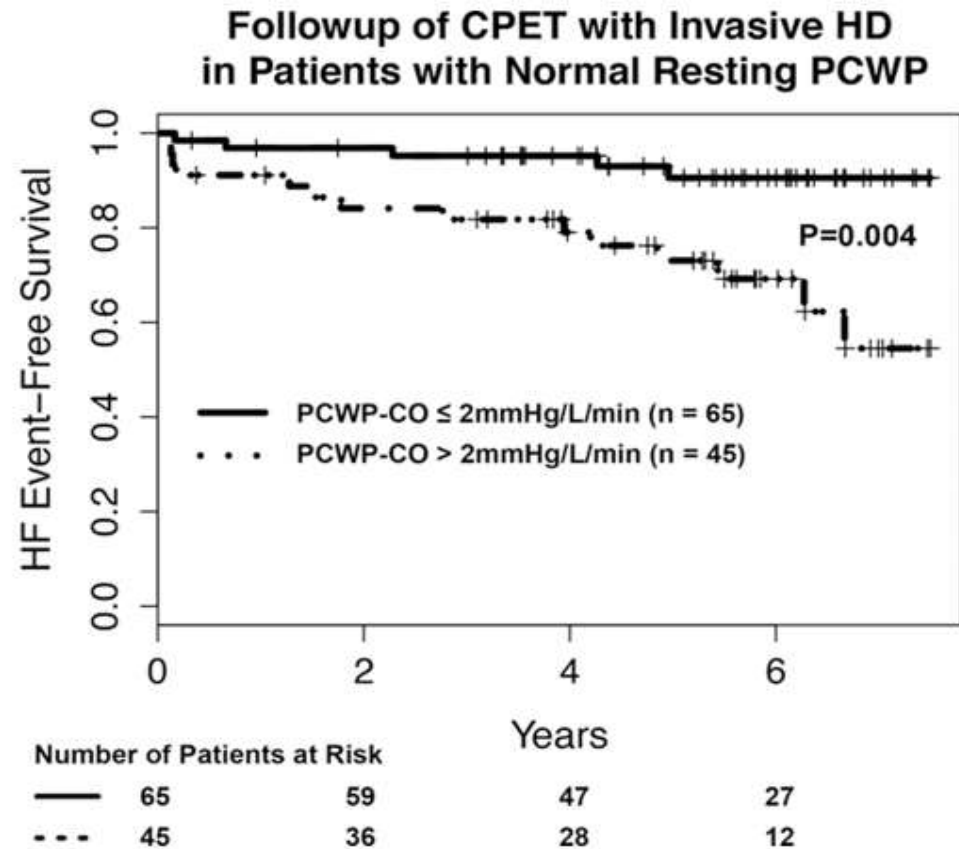
## Summary

- Exercise-induced PH has been shown to be a major risk factor for the development of resting PH in patients with systemic sclerosis <sup>[1-3]</sup> and in healthy carriers of a BMPR2 mutation <sup>[4]</sup>
- The PAWP/CO slope with a threshold >2 mmHg/L/min is consistent with HFpEF

1. R. Condliffe, D.G. Kiely, A.J. Peacock, *et al.* Am J Respir Crit Care Med, 179 (2) (2009), pp. 151-157
2. R. Saggar, D. Khanna, D.E. Furst, *et al.* Arthritis Rheum, 62 (12) (2010), pp. 3741-3750
3. C. Nagel, P. Henn, N. Ehlken, *et al.* Arthritis Res Ther, 17 (2015), p. 165
4. K. Hinderhofer, C. Fischer, N. Pfarr, *et al.* PLoS One, 9 (3) (2014), p. e91374



# Prognostic relevance of pulmonary hemodynamic factors during exercise in suspected HFpEF



- 110 patients with dyspnea and suspected heart failure with preserved ejection fraction (HFpEF) but normal PAWP and left ventricular ejection fraction at rest.
- A PAWP/CO slope  $>2$  WU was associated with poor clinical outcomes, defined as cardiovascular death, or hospitalisation due to heart failure [1]





# Case 2

65 year old woman

Presents with progressive DOE over last 2 years, after climbing a flight of stairs is out of breath.

No abdominal distension, no lower extremity edema

PMH

- HTN x 20 years
- Hips osteoarthritis

Social

- Tobacco: none
- ETOH: social



## Case 2

### Physical Examination


- BP 128/72 mmHg, HR 72 bpm, RR 22/min, BMI 31.09
- Sat 95 on RA
- Neck: Not elevated JVP, Positive HJR
- Heart: S1, S2, no murmur of TR
- Abdomen: soft, no tenderness, no HSM
- Lower extremity: no edema



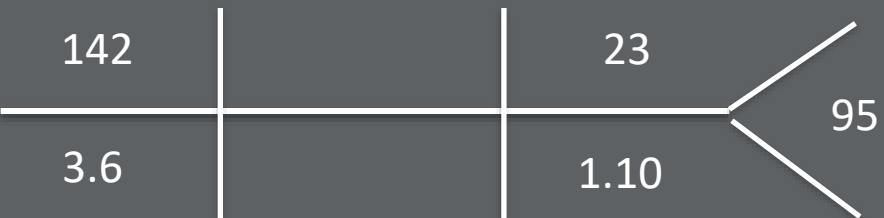


# Laboratory Data

CBC: 8.0 12.1 352 38.3

A diagram for CBC data showing a central horizontal line with four values: 8.0 on the left, 12.1 above the line, 352 on the right, and 38.3 below the line. The line has diagonal branches at both ends.

BMP: 142 23 95 3.6 1.10

A diagram for BMP data showing a central horizontal line with five values: 142 above the line, 23 above the line, 95 on the right, 3.6 below the line, and 1.10 below the line. The line has diagonal branches at the right end and vertical lines separating the values.

Albumin 4.1

Total Bilirubin 0.6

AST/ALT 23/19

INR 1.1

BNP 60



## Case 2

# Transthoracic Echocardiogram

- LVEF 65%, normal LV size
- No LVH
- Trace MR
- Normal RV size
- No RVH
- Normal RV function
- Normal LA and RA size
- Trace tricuspid regurgitation
- PASP 30 mm Hg



# Case 2

**PFTs:** WNL

**Nuclear stress test:** WNL, no evidence of ischemia

**V/Q Scan:** Low probability of CTEPH

**Chest CT Scan:** WNL Lungs are clear. No mediastinal or axillary lymphadenopathy. No pulmonary embolism.

**6 MWDT:**

Patient walked 230 meters (700 feet), no stop. Sat went from 95 down to 89%.  
BORG scale went from 2 up to 9



# Case 2 Right Heart Catheterization

|                            | Baseline<br>Room Air | Bike exercise<br>25 Watts | Bike exercise<br>50 Watts | Ref values<br>(at rest) |
|----------------------------|----------------------|---------------------------|---------------------------|-------------------------|
| BP                         | 120/60 (83)          | 149/74 (98)               | 170/95 (120)              | 110-130/60-80           |
| HR (ppm)                   | 72                   | 112                       | 128                       | 60-90                   |
| RAP (mmHg)                 | 5                    | 12                        | 15                        | 0-6                     |
| PA (mmHg)                  | 33/10                | 53/25                     | 75/27                     | < 35/<15                |
| PA mean (mmHg)             | 18                   | 35                        | 42                        | < 20                    |
| PCWP (mmHg)                | 9                    | 22                        | 25                        | < 15                    |
| TPG (mmHg)                 | 9                    | 13                        | 17                        | <12                     |
| PA saturation (%)          | 66                   | 48                        | 31                        | 65-75                   |
| AO saturation (%)          | 97                   | 95                        | 93                        | > 90                    |
| CO/CI by Fick (L/min)      | 5.75/2.87            | 8.4/4.01                  | 9.06/4.32                 | 5-6/2.5-4.2             |
| CO/CI by TD (L/min)        | 5.8/2.9              | 8.91/4.25                 | -                         | 5-6/2.5-4.2             |
| SVR (WU/dynes)             | 11.65/932            | 11.90/952                 | 12.47/998                 | 800-1300                |
| PVR (WU/dynes)             | 1.56/124             | 1.54/123                  | 1.87/150                  | < 2                     |
| $\Delta$ mPA/ $\Delta$ CO  |                      |                           | 7.25                      | <3                      |
| $\Delta$ PCWP/ $\Delta$ CO |                      |                           | 4.83                      | <2                      |



# Case 2

Patient started on ARNI (Angiotensin receptor-Neprilysin inhibitor Sacubutril/Valsartan) + SGLT-2 inhibitor + Spironolactone

Reevaluation 16 weeks later:

**-6 MWDT:**

Patient walked 340 meters (1200 feet), no stop. Sat went from 96 down to 92%.  
BORG scale went from 1 up to 3

-NYHA Class II-III

-Not fluid overloaded, 6 cm JVD



# Conclusions

- PH is a fatal and progressive disease
- RHC remains the gold standard test to diagnose PH
- Exercise-induced PH defined by an mPAP/cardiac output (CO) slope  $>3$  mmHg/L/min has been re-introduced, and is associated with impaired prognosis
- The PAWP/CO slope with a threshold  $>2$  mmHg/L/min may best differentiate between pre- and post-capillary causes of exercise PH

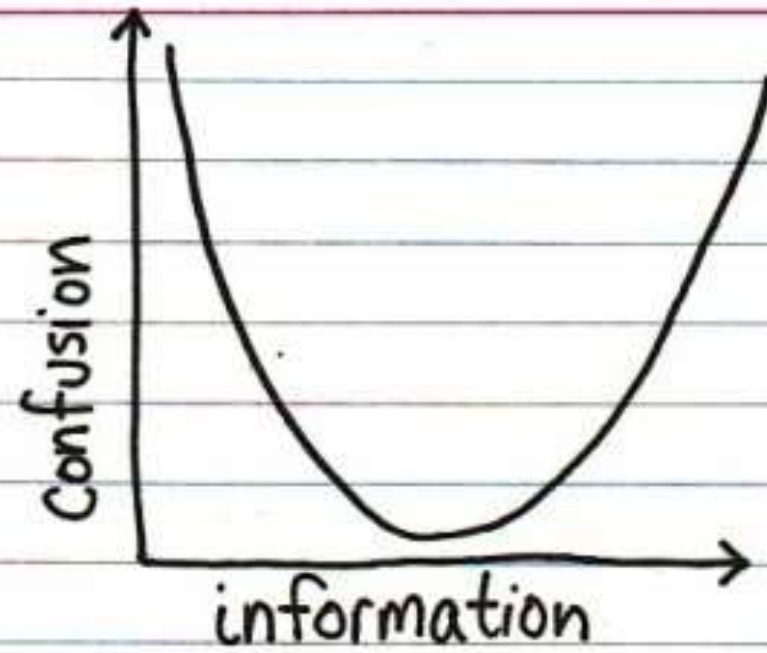


# Conclusions

SOB  $\neq$  PH

If SOB: Think Pulmonary  
Hypertension in your  
Differential Diagnosis







# Aknowledgement

Heart Failure Group under Dr Lowes' direction

The Cath Lab staff

Teya Tsai, Erin Traut and Nico Kavan



**Thank you**

