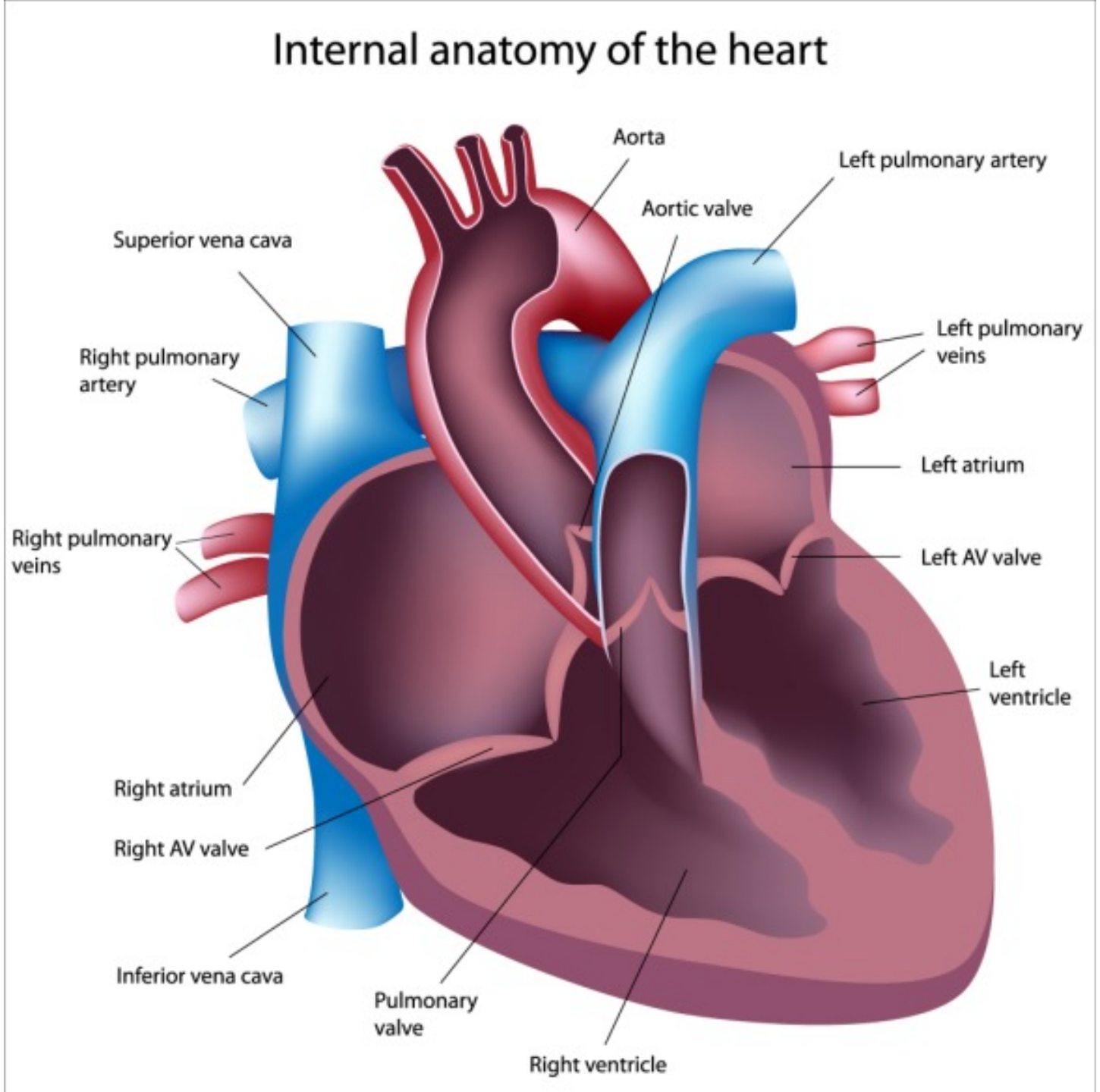
# Pulmonary Hypertension

Imaging and invasive measurements for diagnosis and prognostication

# Internal anatomy of the heart



# Pulmonary artery hypertension stratified into 5 groups by etiology

| Group | Name | Example Etiologies |
|---|---|---|
| Group 1 | Pulmonary arterial hypertension | • Heritable PAH (75% *BMPR2*)<br>• Drug or toxin PAH<br>• PAH 2/2 connective tissue disease, HIV, congenital HD, Schistosomiasis, etc.<br>• 1` – "pulmonary venoocclusive disease"<br>• 1`` – pers. pHTN of the newborn |
| Group 2 | PH from left-sided heart disease | HFpEF, HFrEF, valvular heart disease |
| Group 3 | PH from chronic hypoxic lung disease | Obstructive lung disease<br>Restrictive lung disease<br>Mixed obstructive/restrictive<br>Many others |
| Group 4 | PH from chronic thromboembolic disease | e.g. APLS, Sickle Cell |
| Group 5 | Unclear or multifactorial disease | Sarcoidosis, myeloproliferative disorders, metabolic disorders, cancer-related, dialysis, etc. |

# WHO Functional Classification System for PAH

*Circa ~2005*

## Class Description

| | |
|---|---|
| I | No limitation of usual physical activity; ordinary physical activity does not cause dyspnea, fatigue, chest pain, or presyncope |
| II | Mild limitation of physical activity; no discomfort at rest; but normal activity causes increased dyspnea, fatigue, chest pain, or presyncope |
| III | Marked limitation of activity; no discomfort at rest but less than normal physical activity causes increased dyspnea, fatigue, chest pain, or presyncope |
| IV | Unable to perform physical activity at rest; may have signs of RV failure; symptoms increased by almost any physical activity |

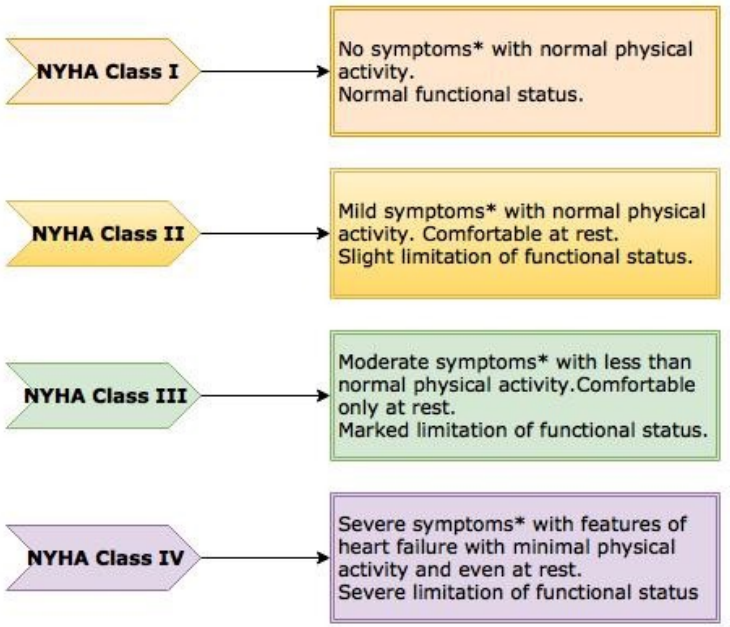
World Health Organization

Global Alliance against Chronic Respiratory Diseases
www.who.int/gard

A world where all people breathe freely

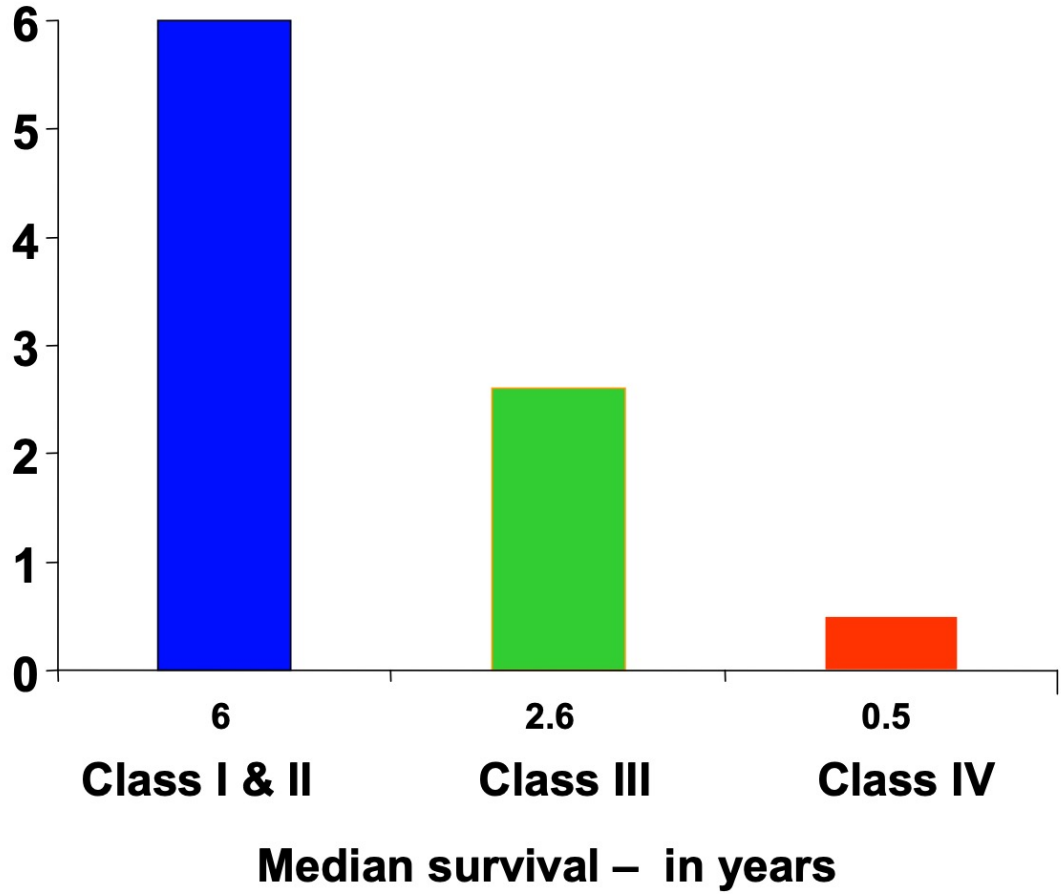
*Reminds me of…*

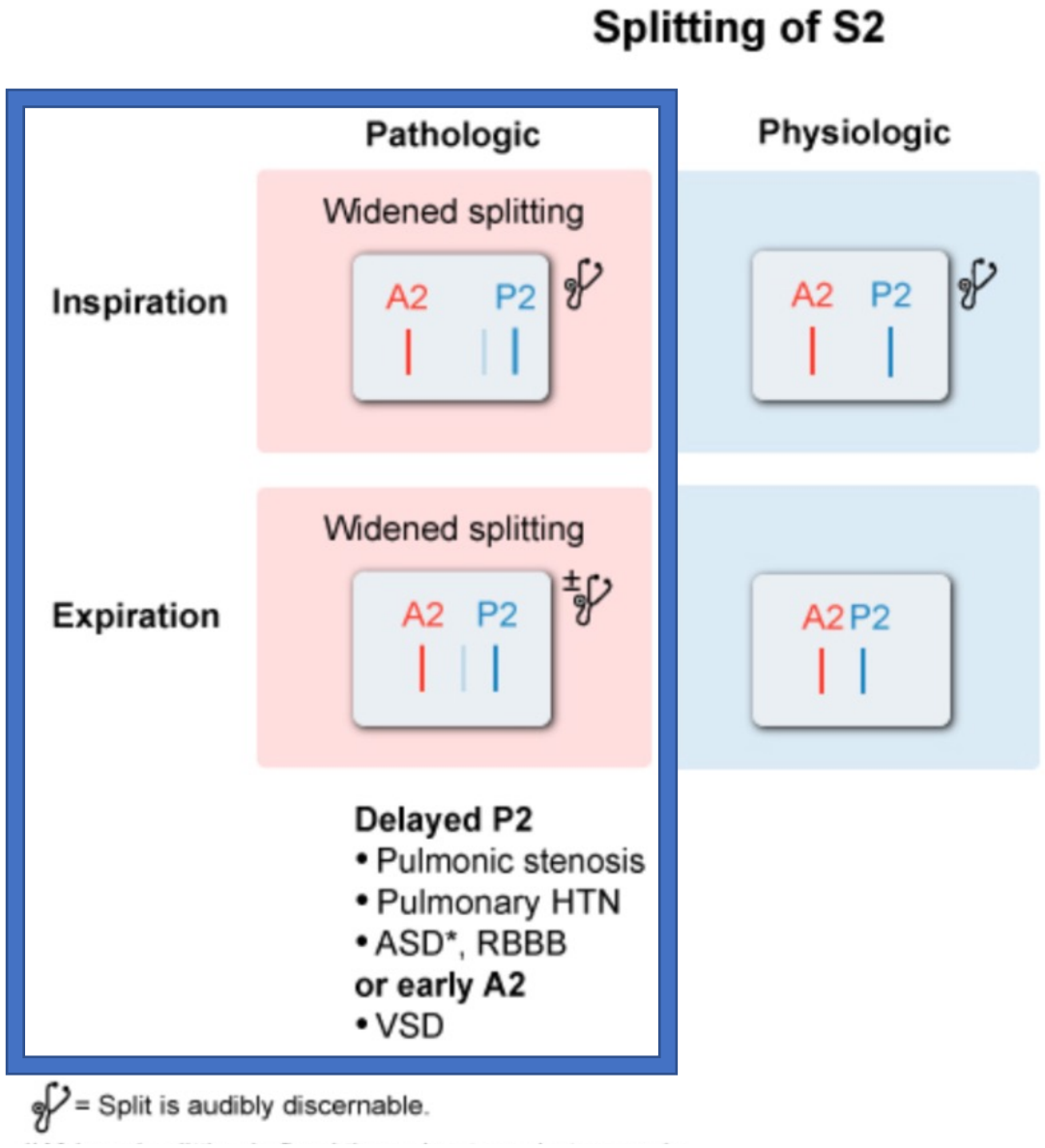
**New York Heart Association (NYHA) Classification of severity of Heart Failure**




Symptoms - Fatigue, palpitations, chest pain, dyspnea, syncope

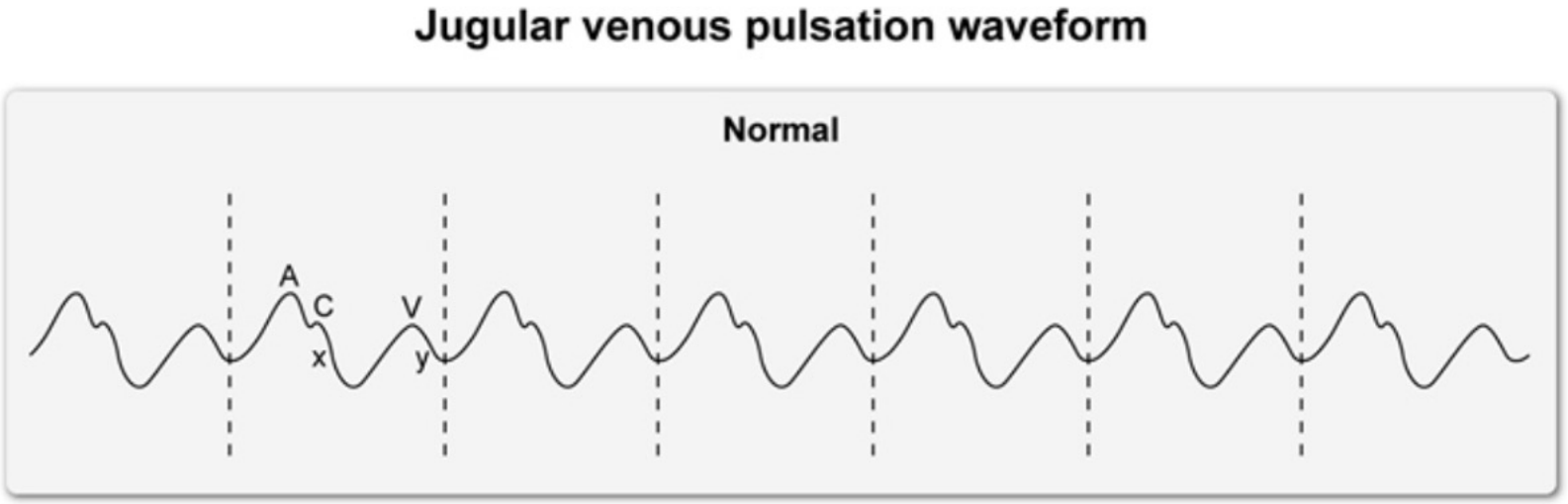
# Without treatment: survival correlates with functional class






- Increased intensity of 2nd heart sound
- Second heart sound has widened splitting in setting of RV dysfunction or failure
  - Splitting normal if RV function is preserved
- Unclear sensitivity and specificity, probably highly operator dependent

# JVP




- A waves may be prominent *(atrial contraction against overfull RV)*
- Prominent V wave would indicate tricuspid regurgitation 2/2 severe PAH

# EKG Findings

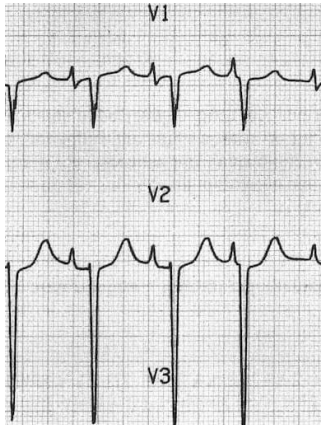
| Axis | Lead I (0°) | Lead avF (90°) |
|---|---|---|
| Normal | Positive | Positive |
| Left axis deviation | Positive | Negative |
| Right axis deviation | Negative | Positive |
| Extreme right axis deviation | Negative | Negative |

- R axis deviation;
  R atrium enlargement
- Frequently not found in early cases of disease
- **Low sensitivity overall**

***Right atrial enlargement (P pulmonale)***

Increased amplitude in II and R atrial portion of V1.
R atrial enlargement diagnosed by P wave > 2.5mm in II, II, or avF (inferior leads).

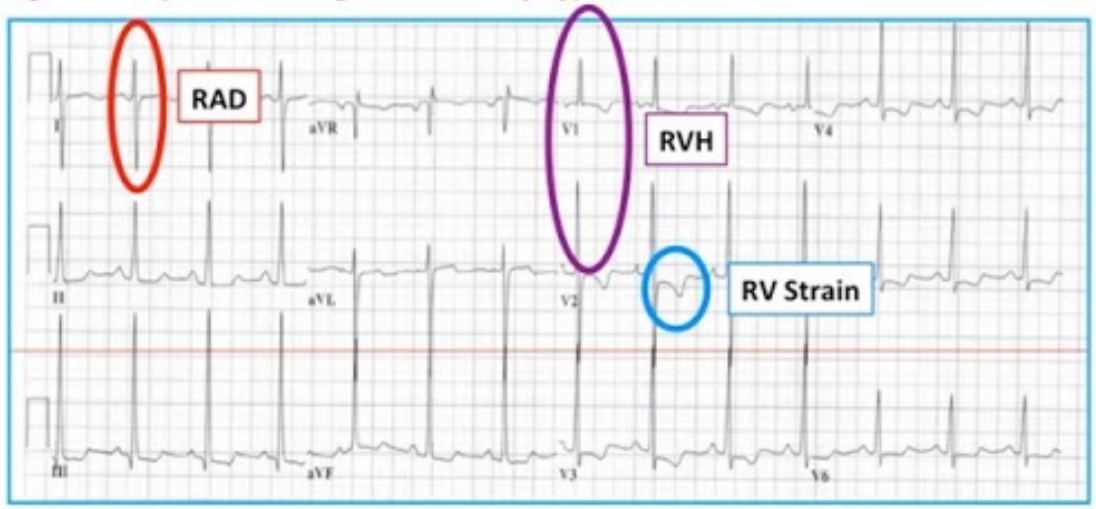
1. P waves >2.5mm in an inferior lead
2. No change in duration of p wave
3. Possible R axis deviation of P wave




| | II | V1 |
|---|---|---|
| Normal |  |  |
| RAE |  |  |
| LAE |  |  |
| RAE + LAE |  |  |

*Relatively low sensitivity and specificity*

Figure 1. Sample ECG with Signs of Pulmonary Hypertension

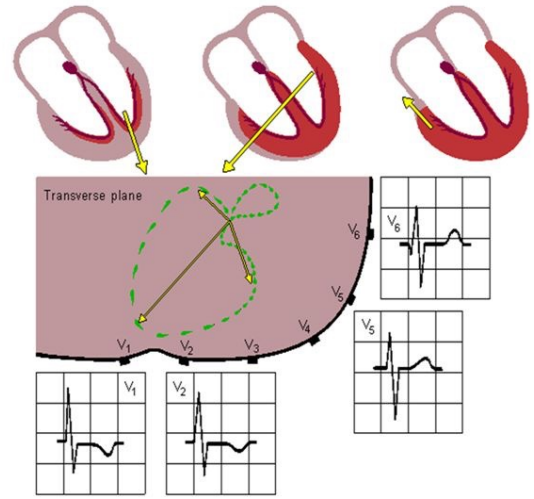



PAH, pulmonary arterial hypertension; RAD, right axis deviation; RVH, right ventricular hypertrophy; RV, right ventricle.

# ECG Criteria for RVH.

- Right axis deviation of +110 degrees or more.
- Dominant R wave in lead V1.
- R wave in lead V1 ≥7mm.

- Other supporting criteria:
- ST segment depression
- T inversion V1 - V4.
- Deep S waves V5, V6, I and aVL.




Precordial leads are also helpful; R wave progression should be disrupted:

- R wave larger than S wave in V1
- S wave larger than R wave in V6

# ACR appropriateness criteria for imaging in suspected PH

1-3: Usually not appropriate
4-6: May be appropriate
7-9: Usually appropriate

**American College of Radiology**
**ACR Appropriateness Criteria®**
**Suspected Pulmonary Hypertension**

**Variant 1:** **Suspected pulmonary hypertension.**

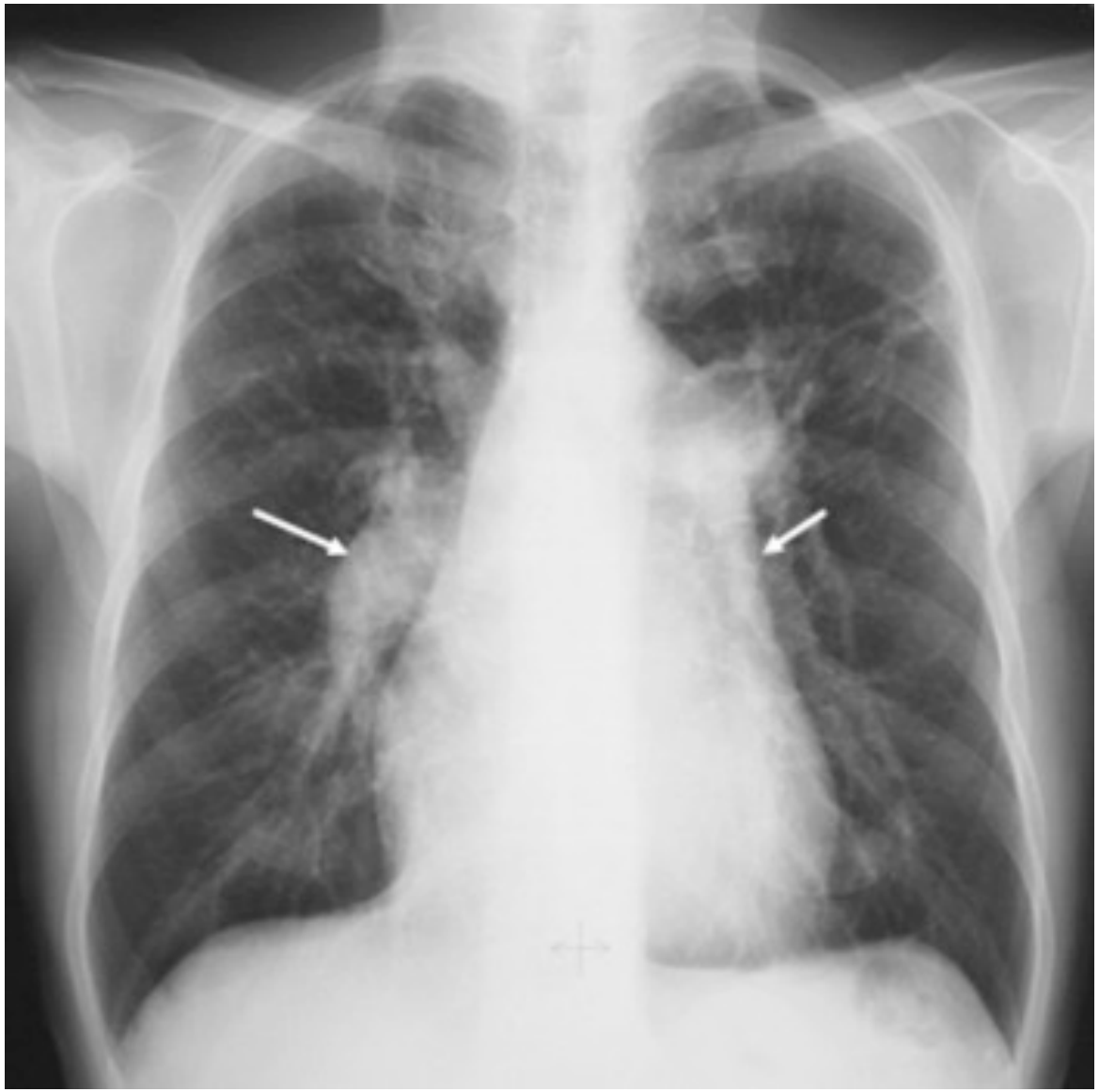
| Radiologic Procedure | Rating | Comments | RRL* |
|---|---|---|---|
| US echocardiography transthoracic resting | 9 | Catheterization and echocardiography are complementary examinations. Both should be performed. Echocardiography is typically performed before catheterization. | O |
| Catheterization right heart | 9 | Catheterization and echocardiography are complementary examinations. Both should be performed. Echocardiography is typically performed before catheterization. | ☢☢ |
| X-ray chest | 9 | X-ray chest is usually performed during the initial workup/screening of suspected pulmonary hypertension and is often the first test performed. | ☢ |
| CTA chest with IV contrast | 8 | This procedure is equivalent to CT chest with IV contrast. The examination choice of CTA chest with IV contrast or CT chest with IV contrast depends on institutional preference. | ☢☢☢ |
| V/Q scan lung | 8 | This procedure is the examination of choice to evaluate for CTEPH. | ☢☢☢ |
| CT chest with IV contrast | 7 | This procedure is equivalent to CTA chest with IV contrast. The examination choice of CTA chest with IV contrast or CT chest with IV contrast depends on institutional preference. | ☢☢☢ |
| MRI heart function and morphology without IV contrast | 6 | | O |
| MRI heart function and morphology without and with IV contrast | 6 | | O |
| MRA chest without and with IV contrast | 5 | CTA chest with IV contrast has better sensitivity for detection of CTEPH. | O |
| US echocardiography transesophageal | 5 | | O |
| CT chest without IV contrast | 4 | If there is a concern for an occult interstitial lung disease, HRCT may be appropriate. | ☢☢☢ |
| Arteriography pulmonary with right heart catheterization | 3 | Noninvasive imaging is generally performed instead. This procedure can be performed when considering surgical or percutaneous embolectomy. | ☢☢☢☢ |
| MRA chest without IV contrast | 2 | | O |
| FDG-PET/CT heart | 2 | | ☢☢☢☢ |
| CT chest without and with IV contrast | 1 | | ☢☢☢ |

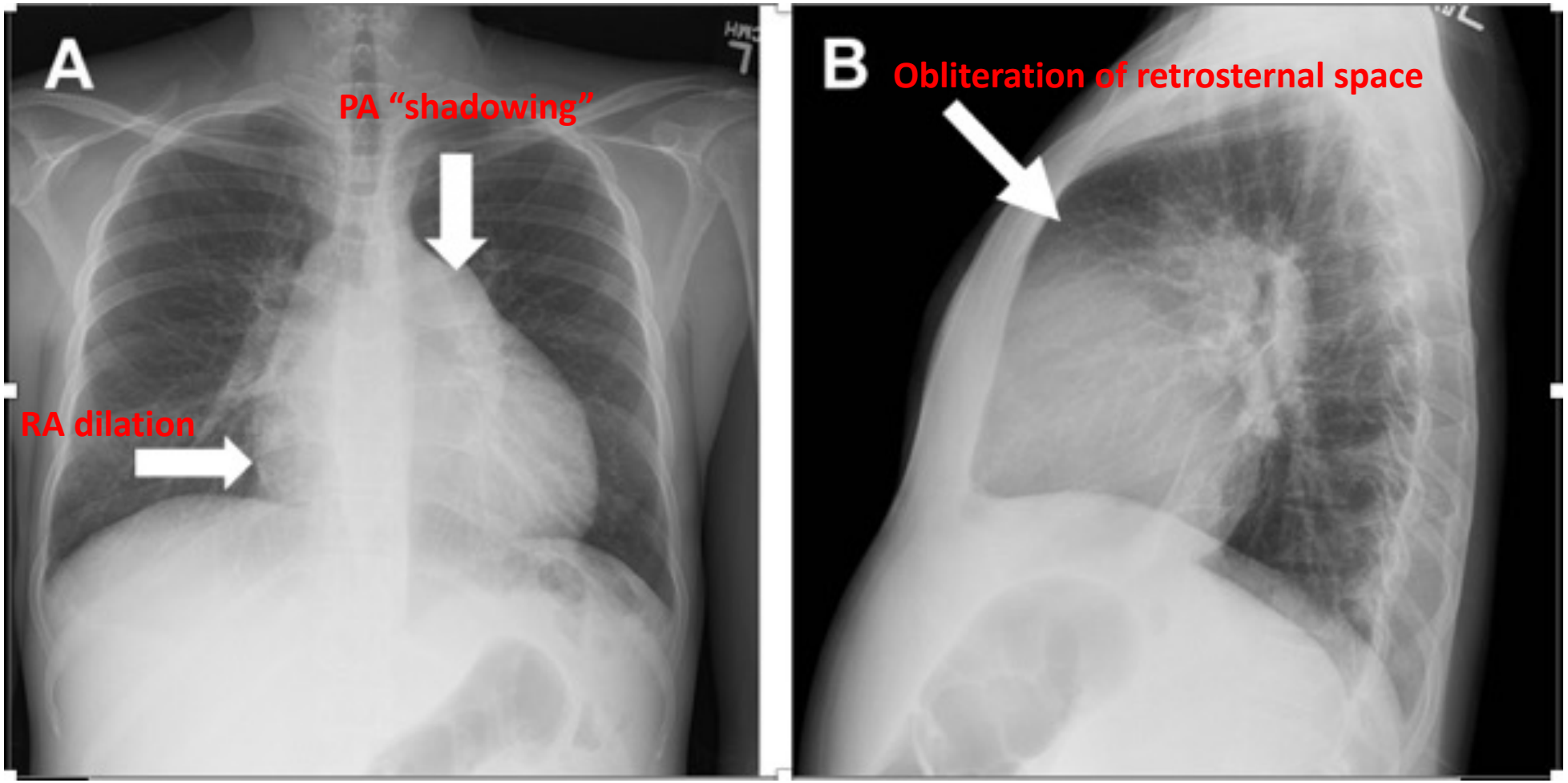
**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

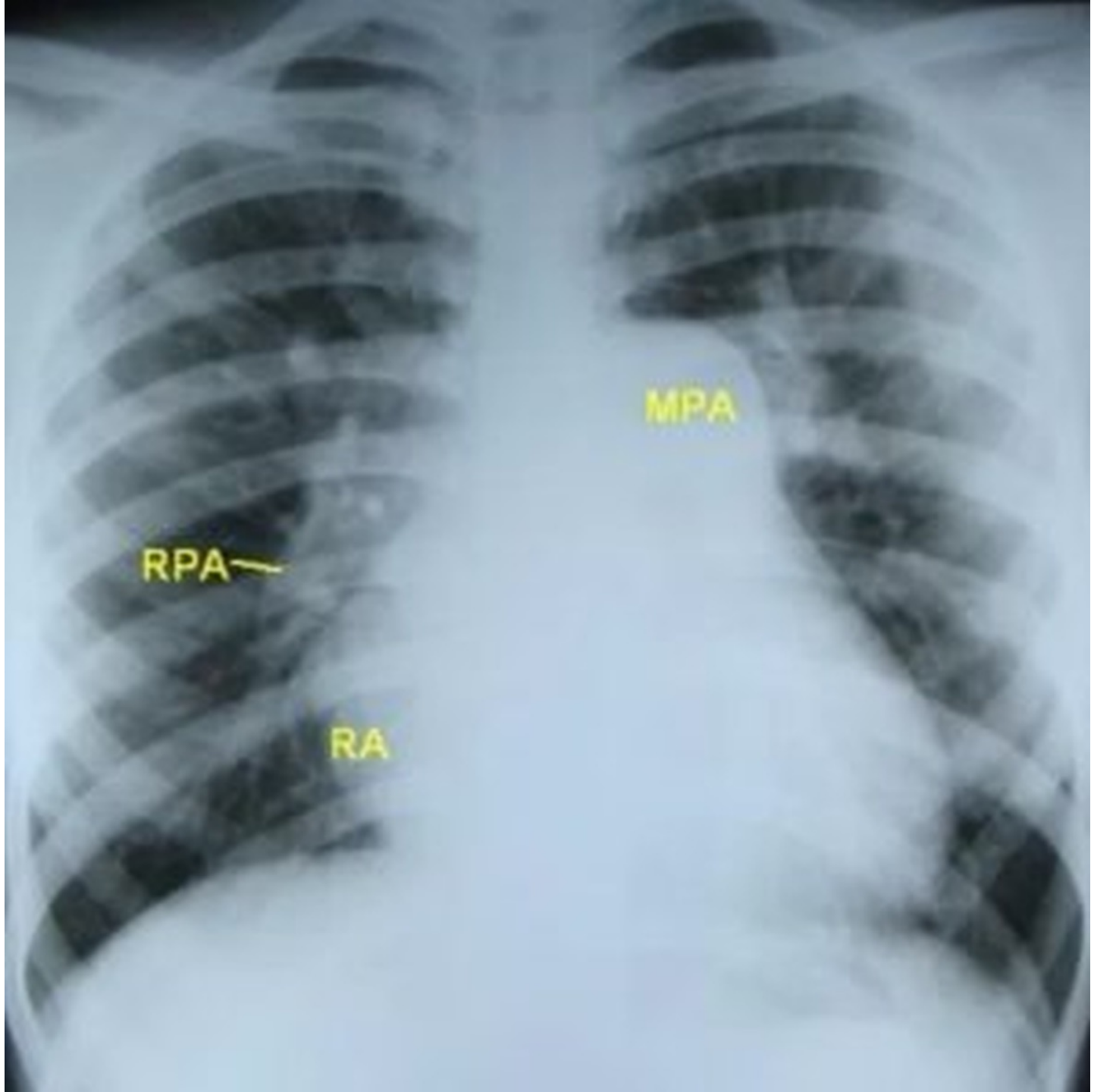
*Relative Radiation Level

# Pulmonary Hypertension Chest Radiograph

Engorgement/dilation of R and L descending pulmonary arteries




A
PA “shadowing”
RA dilation
B
Obliteration of retrosternal space


MPA
RPA
RA

# Pulmonary Hypertension Echocardiography

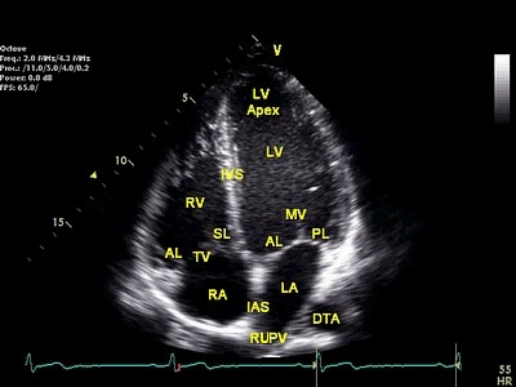
TTE is first choice

# TTE in pHTN

TTE is initial test of choice in patients with pHTN

3 major evaluations on TTE:

1. Tricuspid regurgitant jet velocity (TRJV) --> ePASP
2. RV size, wall thickness, and function
3. Assessment for contributing left heart disease




- TRJV is estimator for pulmonary artery systolic pressure (ePASP)

$$ePASP = 4 \times TRV^2 + RAP$$

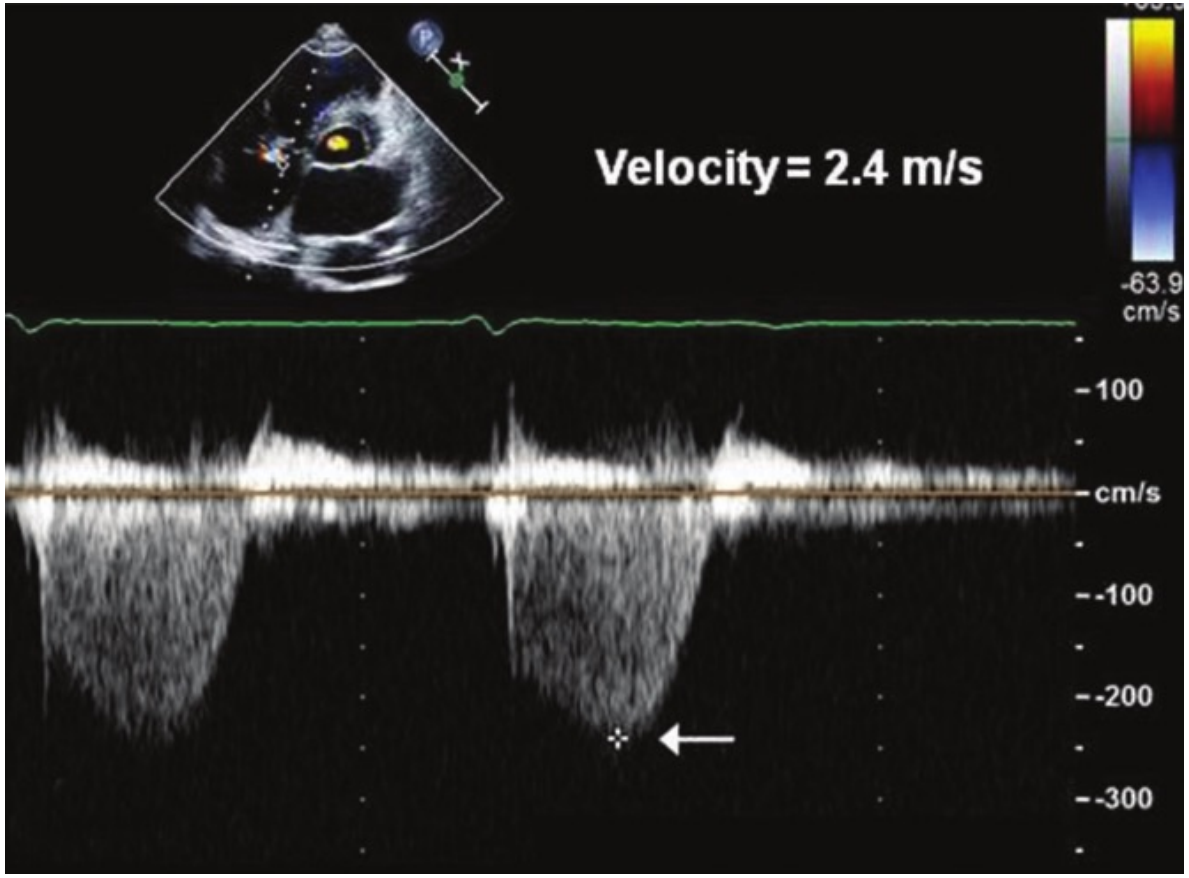


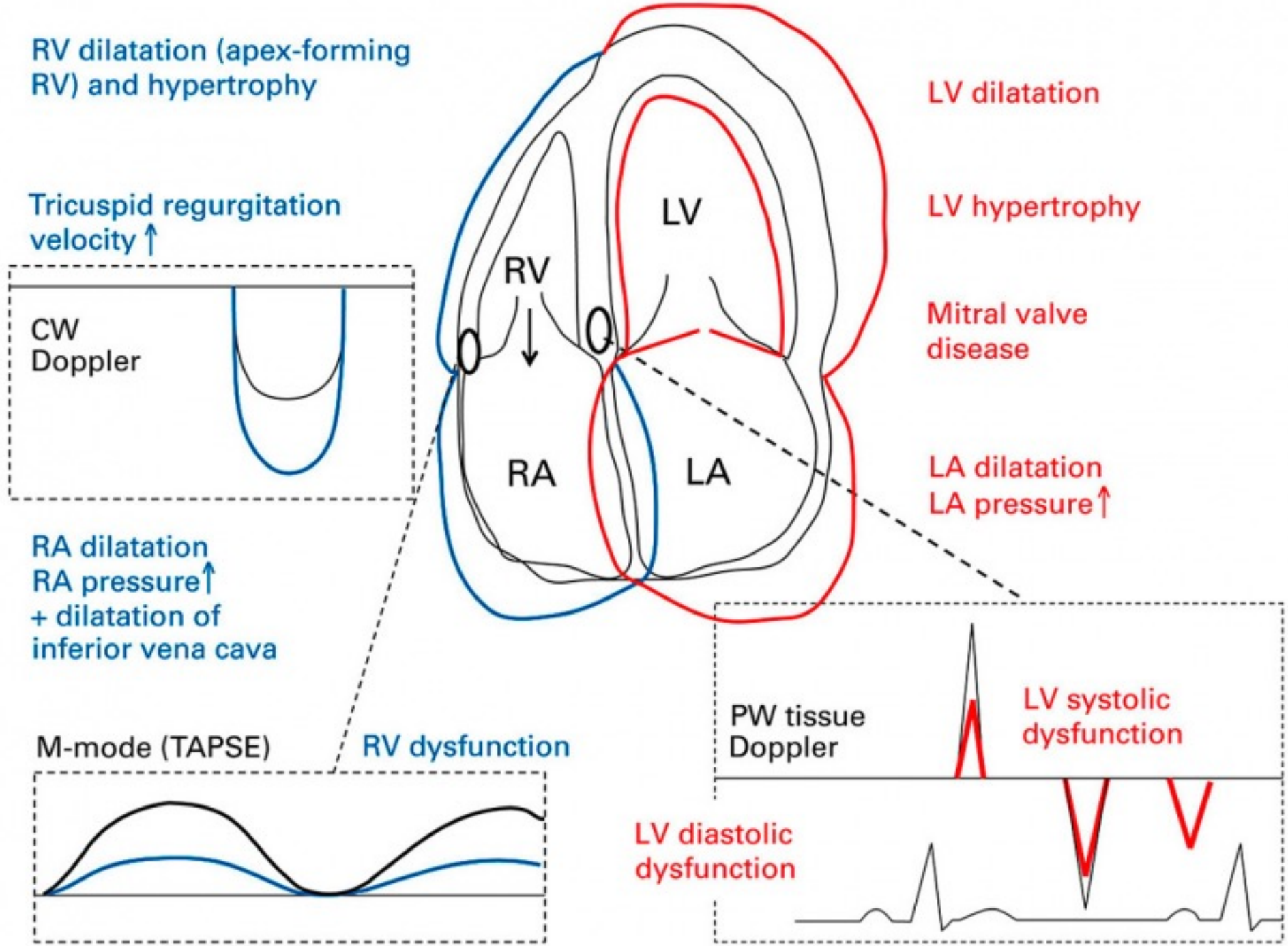

**Table A:**
**Echocardiographic probability of pulmonary hypertension in symptomatic patients with a suspicion of pulmonary hypertension**

| Peak tricuspid regurgitation velocity (m/s) | Presence of other echo "PH signs"* | Echocardiographic probability of pulmonary hypertension |
|---|---|---|
| ≤2.8 or not measurable | No | Low |
| ≤2.8 or not measurable | Yes | Intermediate |
| 2.9 to 3.4 | No | |
| 2.9 to 3.4 | Yes | High |
| >3.4 | Not required | |

ESC 2015

***2.8 and 3.4 m/s are numbers to remember***

Other echo PH signs: RV/LV diameter > 1.0, flattening of IVS during systole, RV outflow doppler acceleration, early diastolic PA regurgitation >2.2m/s, IVC diameter >21mm without inspiratory collapse, RA ES area >18cm$^2$


RV dilatation (apex-forming RV) and hypertrophy
Tricuspid regurgitation velocity ↑
CW Doppler
RA dilatation
RA pressure ↑
+ dilatation of inferior vena cava
M-mode (TAPSE)
RV dysfunction
RV
LV
RA
LA
LV dilatation
LV hypertrophy
Mitral valve disease
LA dilatation
LA pressure ↑
PW tissue Doppler
LV systolic dysfunction
LV diastolic dysfunction

If echo probability of PH is **intermediate** to **high** ***and*** PH is not explained by left-heart disease:

Guidelines recommend performing additional testing to detect underlying etiology.

**“Sufficient left heart disease PH” (Group 2)**

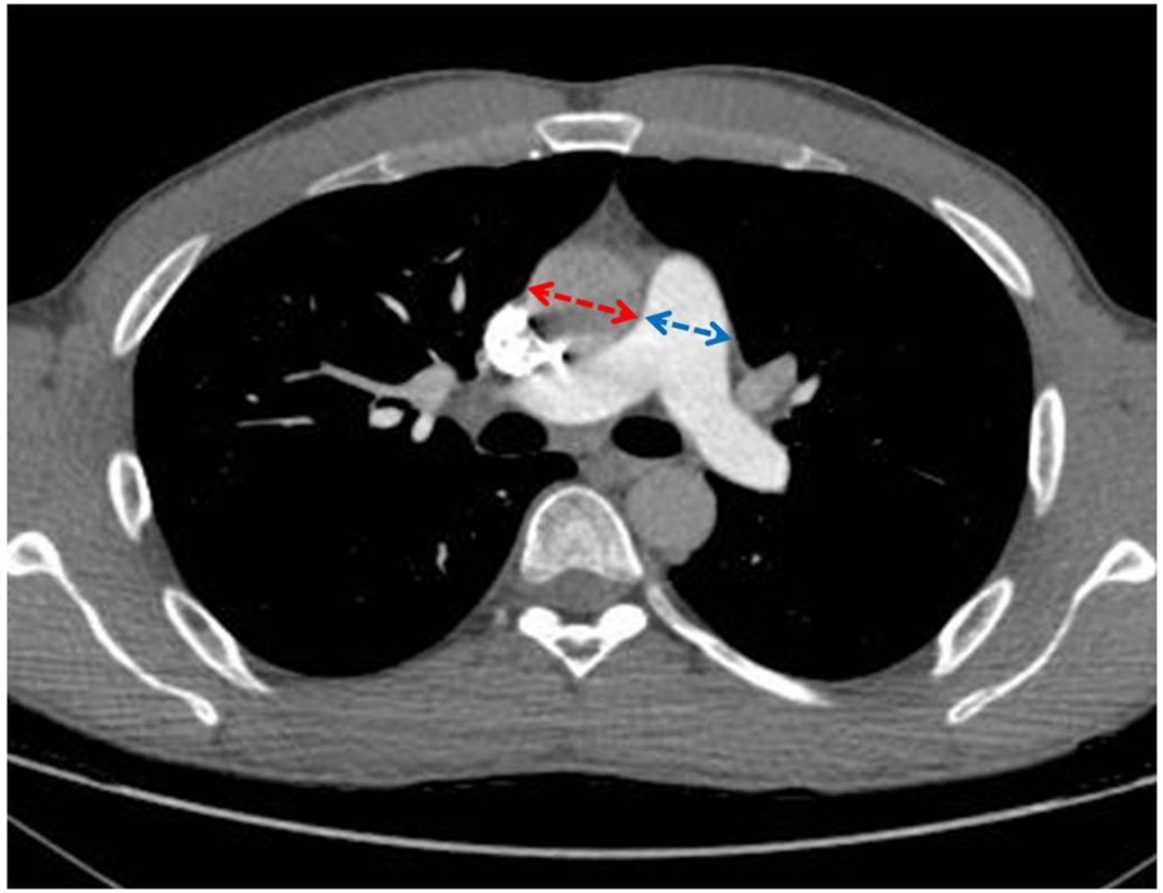
vs.

**“Insufficient left heart disease PH (1, 3, 4, 5)**

*i.e., is the PH explainable by left heart disease, or do we need to find another explanation*

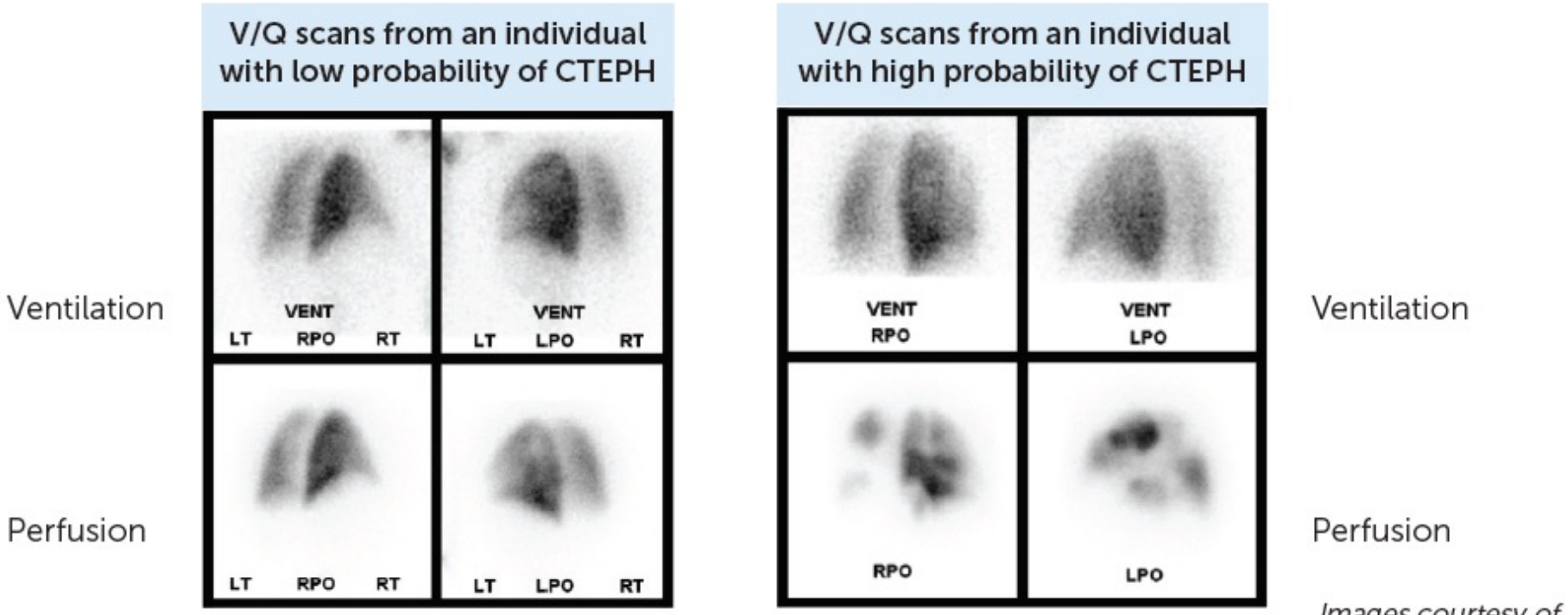
# Suspected Group 3 PH (Chronic Hypoxic Lung Disease)

- Example testing modalities:
  - High-resolution CT
  - DLCO
  - Pulmonary spirometry
  - Cardiopulmonary exercise testing
  - Sleep studies (OSA)



# Suspected Group 4 PH (CTEPH)

- V/Q scan
- High sensitivity (~95%) and specificity (90%) for CTEPH
- Can do CTPA or MRPA as well

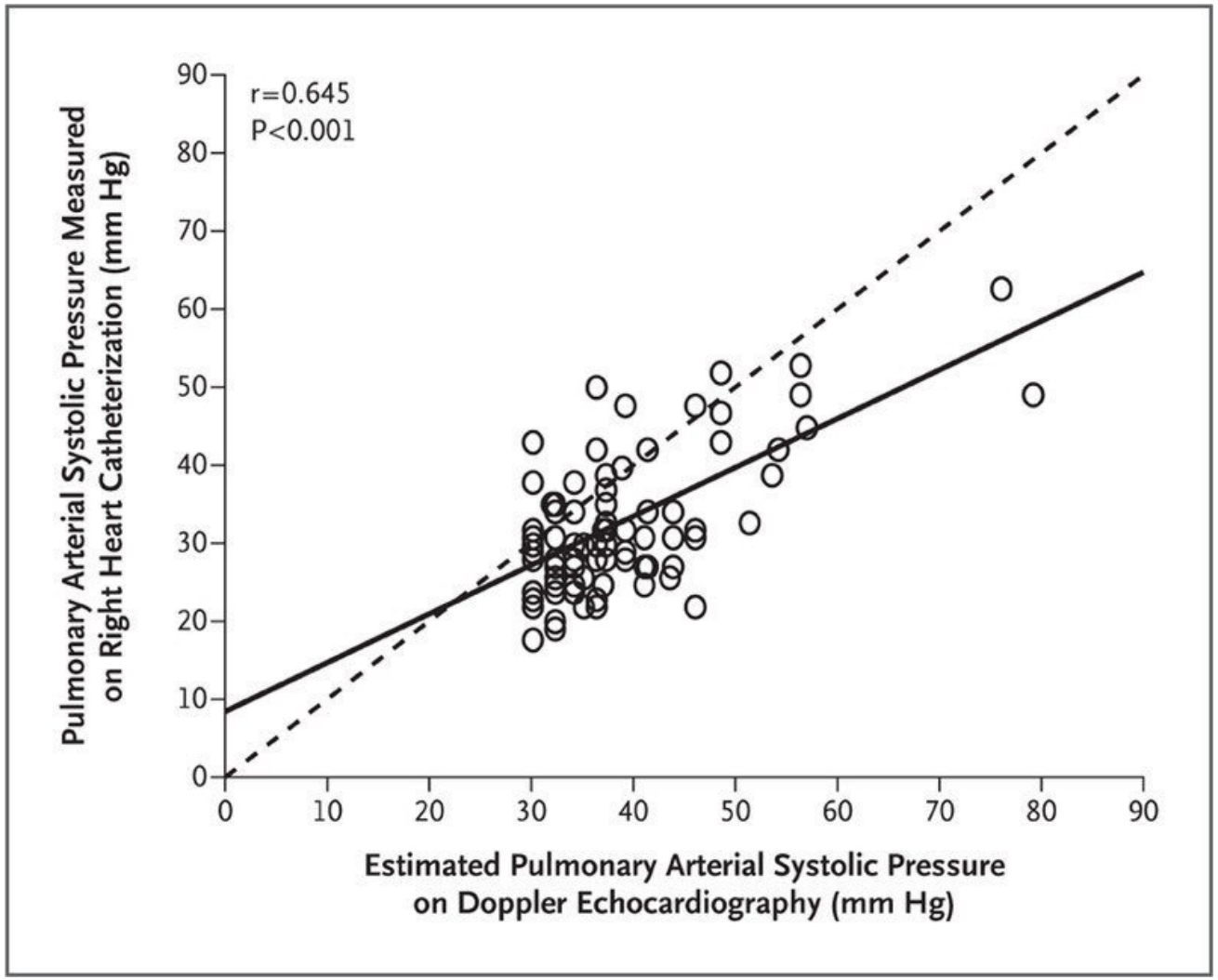


# Right Heart Catheterization (RHC)

“Gold standard”

# Why is RHC the gold standard?

ePASP PPV around 25% for pHTN diagnosis




$r^2 = 0.42$

ORIGINAL ARTICLE

A Hemodynamic Study of Pulmonary Hypertension in Sickle Cell Disease

Florence Parent, M.D., Dora Bachir, M.D., Jocelyn Inamo, M.D., François Lionnet, M.D., Françoise Driss, M.D., Gylna Loko, M.D., Anoosha Habibi, M.D., Soumiya Bennani, M.D., Laurent Savale, M.D., Serge Adnot, M.D., Bernard Maitre, M.D., Azzedine Yaïci, M.D., et al.

July 7, 2011
N Engl J Med 2011; 365:44-53
DOI: 10.1056/NEJMoa1005565

Article Figures/Media

*Many similar studies*

# Evaluation for Group 1 and 5 PH

- Diagnostic RHC should be performed after groups 2, 3, and 4 are ruled out, **or** in cases of diagnostic uncertainty

- Example diagnostic uncertainty scenarios:
  - Echo PH risk is low probability, but symptoms clinically out of proportion
  - Echo PH risk is intermediate to high, but unclear left heart contribution
  - Group 3 PH, but severity of PH not proportional to imaging findings
  - Suspected mixed etiology

- RHC also useful for baseline severity assessment in all groups, vasoreactivity testing, etc.

**Schwan-Ganz Catheter**

*Sits in RA*

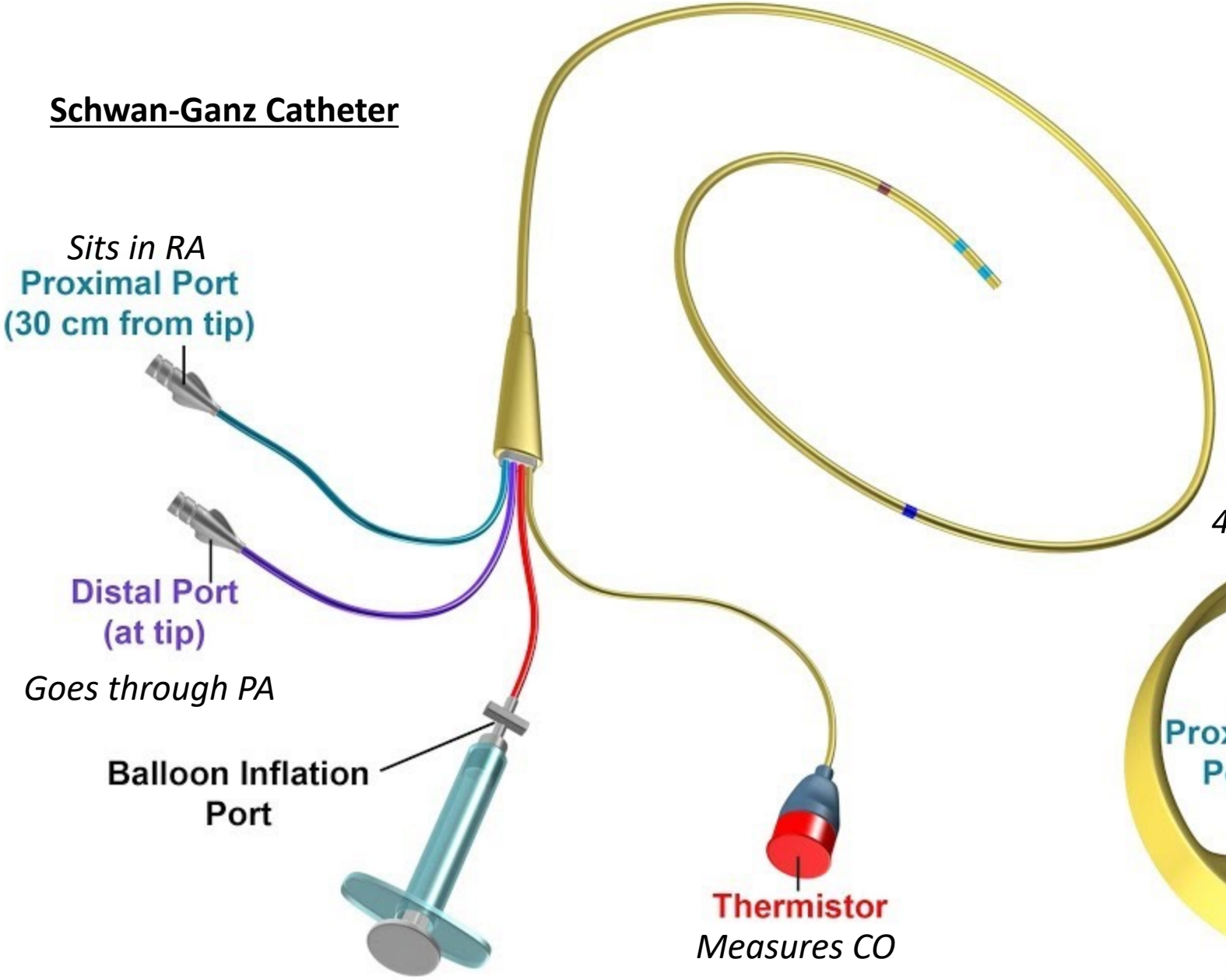
**Proximal Port**
**(30 cm from tip)**

**Distal Port**
**(at tip)**

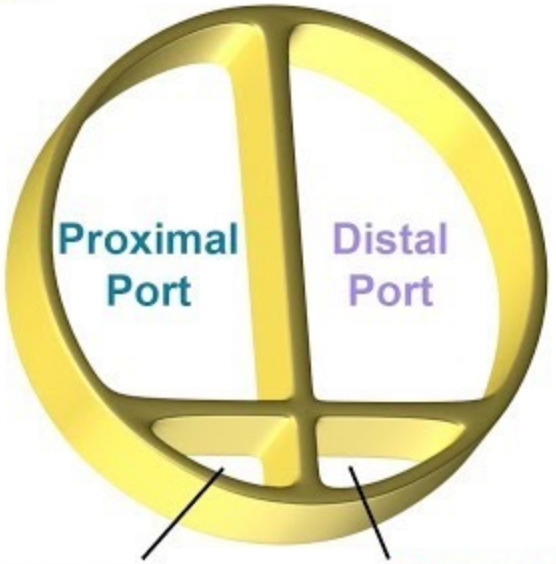
*Goes through PA*

**Balloon Inflation Port**

**Thermistor**
*Measures CO*

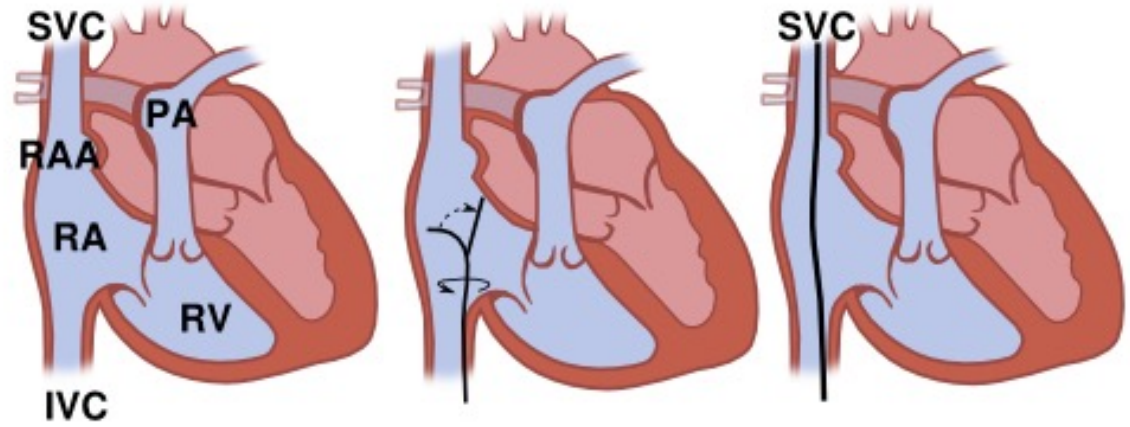


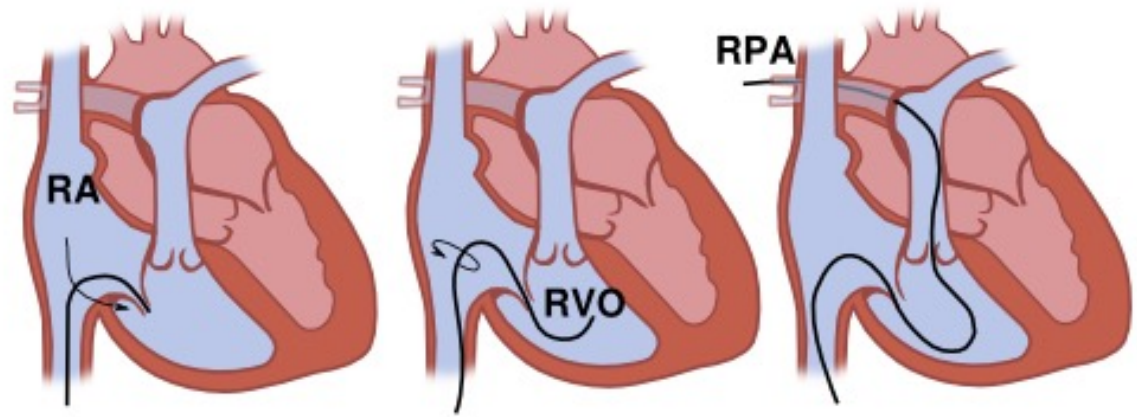
*4 major components*

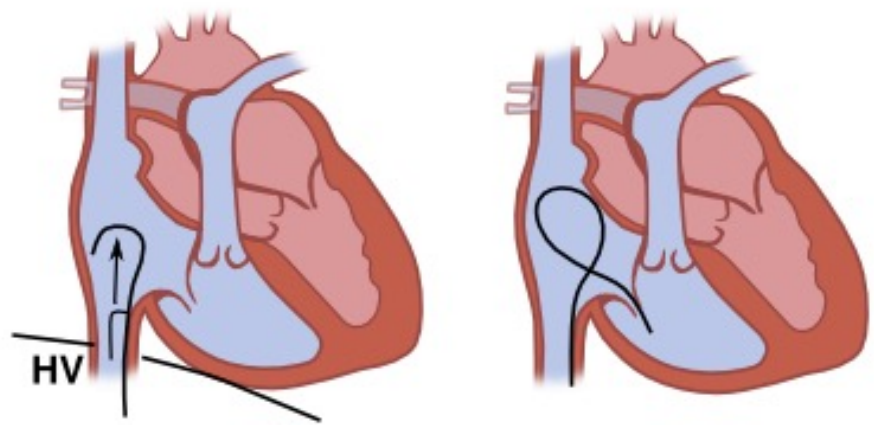



*For PCWP*

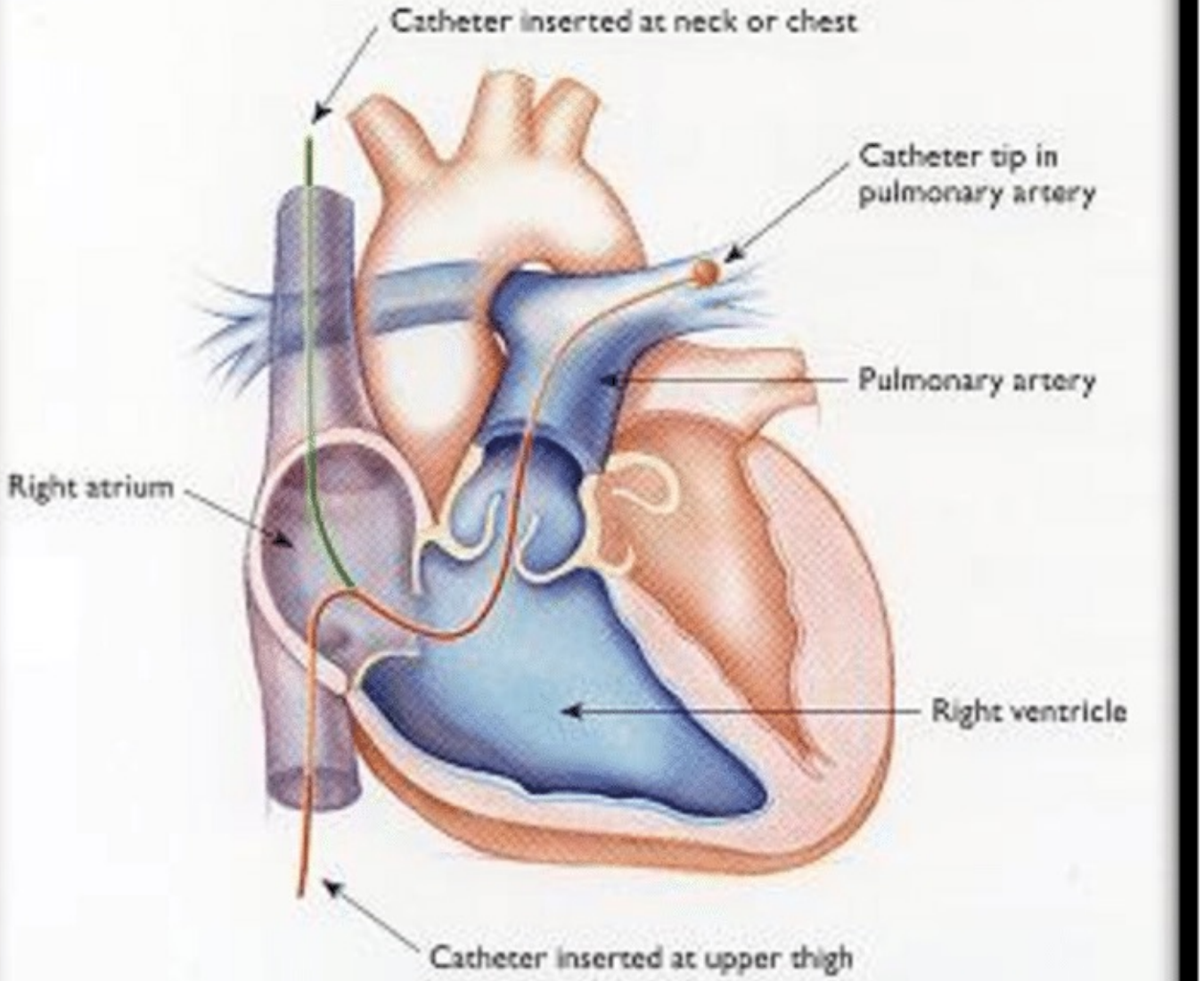
Example Femoral Approach

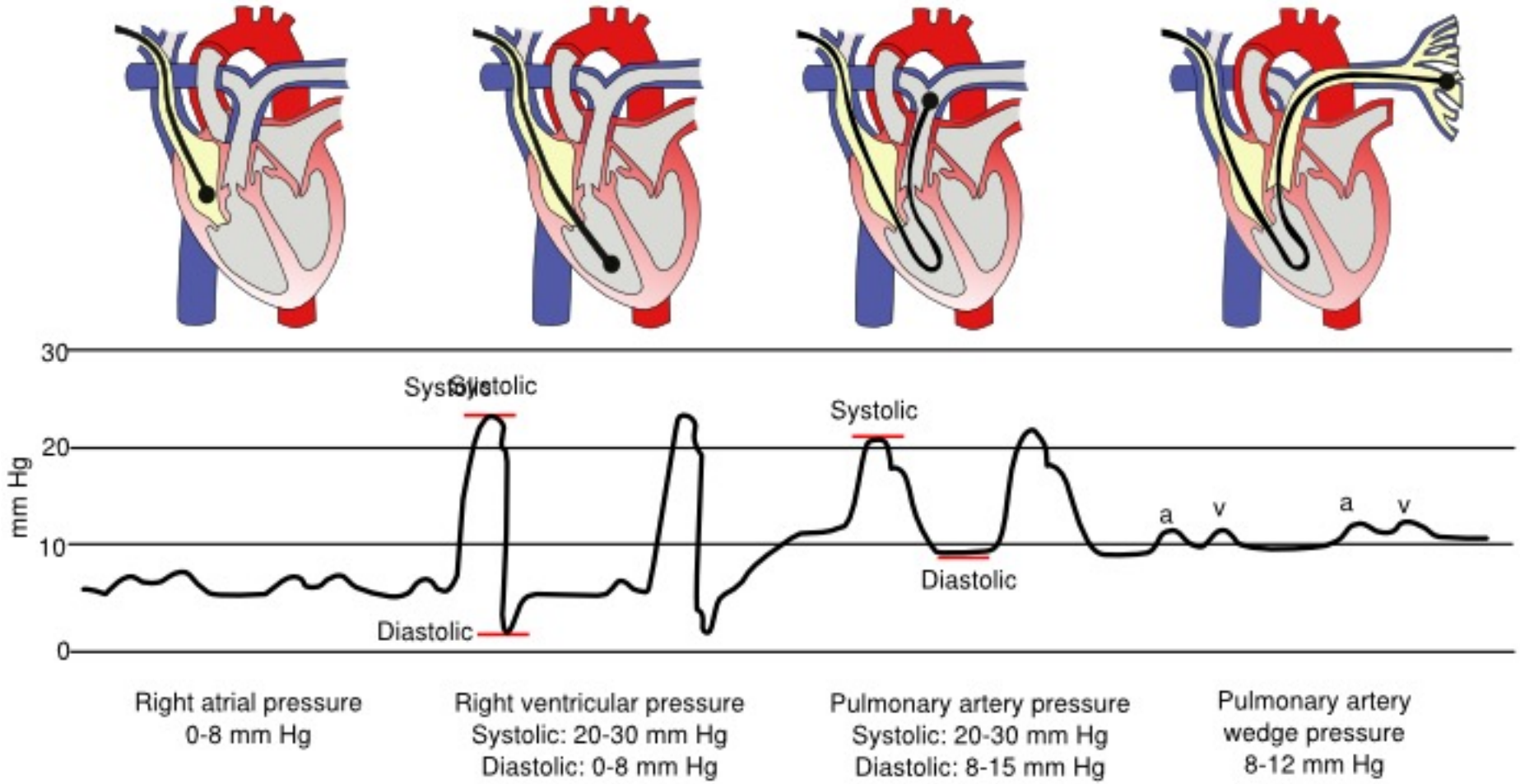


***Approach via RIJ more common***




30
20
10
0
mm Hg
Systolic
Systolic
Diastolic
Systolic
Diastolic
a
v
a
v
Right atrial pressure
0-8 mm Hg
Right ventricular pressure
Systolic: 20-30 mm Hg
Diastolic: 0-8 mm Hg
Pulmonary artery pressure
Systolic: 20-30 mm Hg
Diastolic: 8-15 mm Hg
Pulmonary artery
wedge pressure
8-12 mm Hg

3 different pressures are essential to PH diagnosis:

1. Pulm. a. MAP
2. PAWP (PCWP)
3. PVR

| Pressures | Average (mm Hg) | Range (mm Hg) |
|---|---|---|
| Right atrium | | |
| a wave | 6 | 2-7 |
| v wave | 5 | 2-7 |
| Mean | 3 | 1-5 |
| Right ventricle | | |
| Peak systolic | 25 | 15-30 |
| End-diastolic | 4 | 1-7 |
| Pulmonary artery | | |
| Peak systolic | 25 | 15-30 |
| End-diastolic | 9 | 4-12 |
| Mean | 15 | 9-19 |
| Pulmonary capillary | | |
| Wedge | | |
| Mean | 9 | 4-12 |
| Left atrium | | |
| a wave | 10 | 4-16 |
| v wave | 12 | 6-21 |
| Mean | 8 | 2-12 |
| Left ventricle | | |
| Peak systolic | 130 | 90-140 |
| End-diastolic | 8 | 5-12 |
| Central aorta | | |
| Peak systolic | 130 | 90-140 |
| End-diastolic | 70 | 60-90 |
| Mean | 85 | 70-105 |

| Vascular Resistance | Mean (dyne-sec · cm $^{-5}$) | Range (dyne-sec · cm $^{-5}$) |
|---|---|---|
| Right atrium | | |
| Systemic vascular resistance | 1100 | 700-1600 |
| Total pulmonary resistance | 200 | 100-300 |
| Pulmonary vascular resistance | 70 | 20-130 |

Normal mean PA pressure <20

Normal PAWP < 15

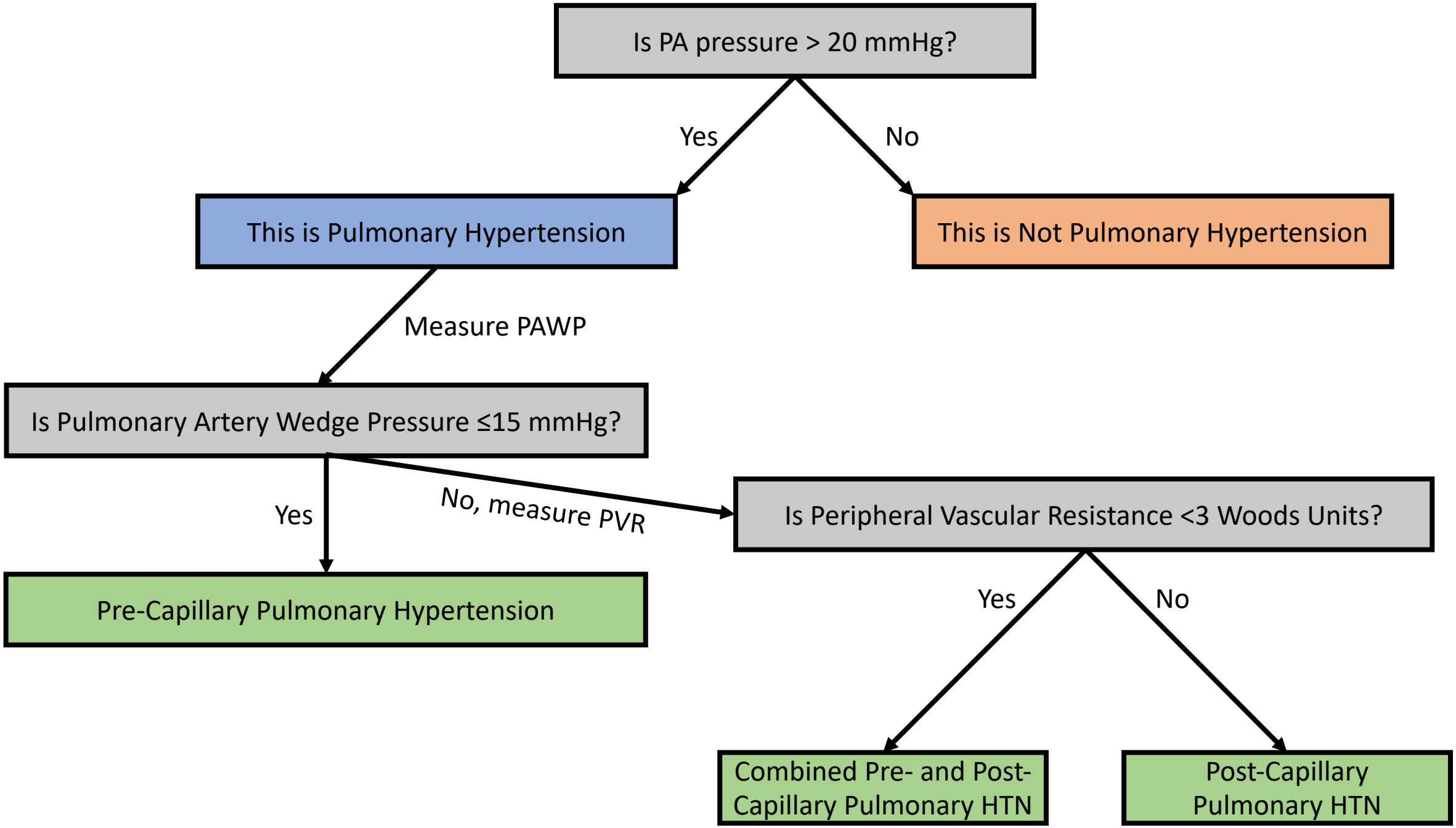
Normal pulmonary vascular resistant <3 woods units

## Hemodynamic definitions of pre- and post-capillary pulmonary hypertension

| Definitions | Characteristics | Clinical groups* |
|---|---|---|
| Pre-capillary PH | mPAP >20 mmHg<br>PAWP ≤15 mmHg<br>PVR ≥3 WU | 1, 3, 4, and 5 |
| Isolated post-capillary PH (IpcPH) | mPAP >20 mmHg<br>PAWP >15 mmHg<br>PVR <3 WU | 2 and 5 |
| Combined pre- and post-capillary PH (CpcPH) | mPAP >20 mmHg<br>PAWP >15 mmHg<br>PVR ≥3 WU | 2 and 5 |

PH: pulmonary hypertension; mPAP: mean pulmonary arterial pressure; PAWP: pulmonary arterial wedge pressure; PVR: pulmonary vascular resistance; WU: Wood units.

* Group 1: PAH; group 2: PH due to left heart disease; group 3: PH due to lung diseases and/or hypoxia; group 4: PH due to pulmonary artery obstructions; group 5: PH with unclear and/or multifactorial mechanisms.


Is PA pressure > 20 mmHg?
Yes
No
This is Pulmonary Hypertension
This is Not Pulmonary Hypertension
Measure PAWP
Is Pulmonary Artery Wedge Pressure ≤15 mmHg?
Yes
No, measure PVR
Is Peripheral Vascular Resistance <3 Woods Units?
Pre-Capillary Pulmonary Hypertension
Yes
No
Combined Pre- and Post-Capillary Pulmonary HTN
Post-Capillary Pulmonary HTN

Questions?