

Use of Echocardiography in Pulmonary Hypertension

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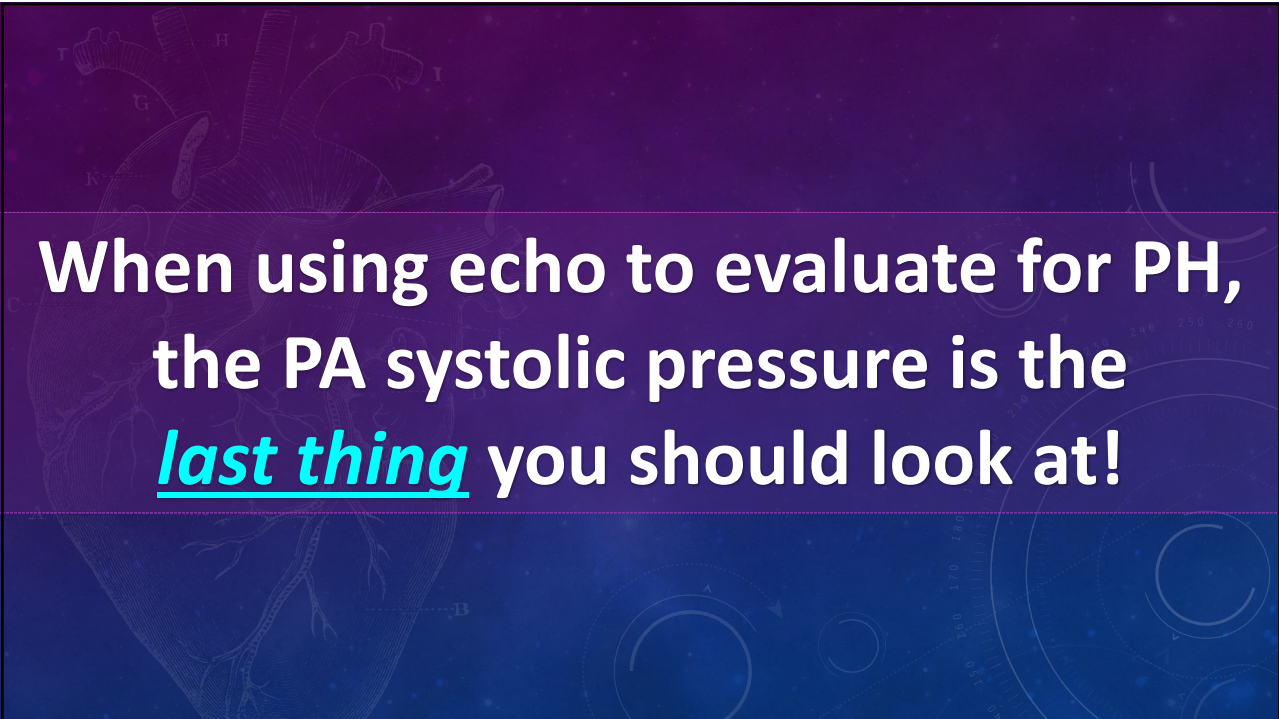
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Disclosures

- **Research funding:**
 - ✓ NIH U54 HL160273, R01 HL107577, R01 HL127028, R01 HL140731
 - ✓ AHA #16SFRN28780016, #15CVGSPD27260148
 - ✓ Actelion, AstraZeneca, Corvia, Novartis, Pfizer
- **Consulting / advisory board / steering committee:**
 - ✓ Abbott, Actelion, AstraZeneca, Amgen, Aria CV, Axon Therapies, Bayer, Boehringer-Ingelheim, Boston Scientific, Bristol-Myers Squibb, Cardiora, Coridea, CVRx, Cycleron, Cytokinetics, Edwards Lifesciences, Eidos, Eisai, Imara, Impulse Dynamics, Intellia, Ionis, Ironwood, Lilly, Merck, MyoKardia, NGMbio, Novartis, Novo Nordisk, Pfizer, Prothena, Regeneron, Rivus, Roche, Sanofi, Shifamed, Tenax, Tenaya, and United Therapeutics

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When using echo to evaluate for PH,
the PA systolic pressure is the
last thing you should look at!

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Learning objectives

- Define pulmonary hypertension (PH) and its prevalence
- Describe the diagnostic approach to PH
- Discuss role of echo and invasive hemodynamic testing in diagnosis and management of PH

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What I look for on the echo in PH

- Left heart disease causes of PH
- RV size and function function:
 - ✓ RV size (RV basal diameter) and RV wall thickness
 - ✓ RV systolic function (RV s' velocity, TAPSE, global RV function)
 - ✓ Septal flattening: systole, diastole, both?
- Pericardial effusion
- Bubble study to look for intracardiac shunt

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What I look for on the echo in PH

- Hemodynamics:
 - ✓ PA systolic pressure (peak TR gradient)
 - ✓ PA diastolic pressure (end-diastolic PR gradient)
 - ✓ RA pressure (IVC size and collapsibility)
 - ✓ LV filling pressure (E/e' ratio [use lateral e'])
 - ✓ Non-invasive PVR: peak TR velocity/RVOT VTI
- Pulmonary arterial vs. venous hypertension (PAH vs. PVH):
 - ✓ Notching in PW Doppler tracing of RV outflow?
 - ✓ LA/RA size ratio? Does the interatrial septum bow to left or right?
 - ✓ Lateral e' velocity, lateral E/e' ratio? Grade of diastolic function?

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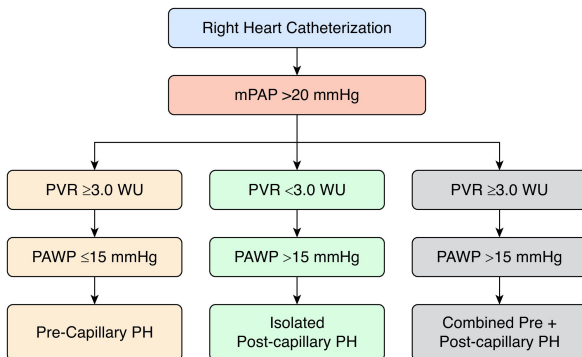
Definitions of PH

- A hemodynamic and pathophysiologic condition
- Pulmonary hypertension: not a disease itself
- Pulmonary arterial hypertension: a disease of the pulmonary microcirculation

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Classification of PH

A

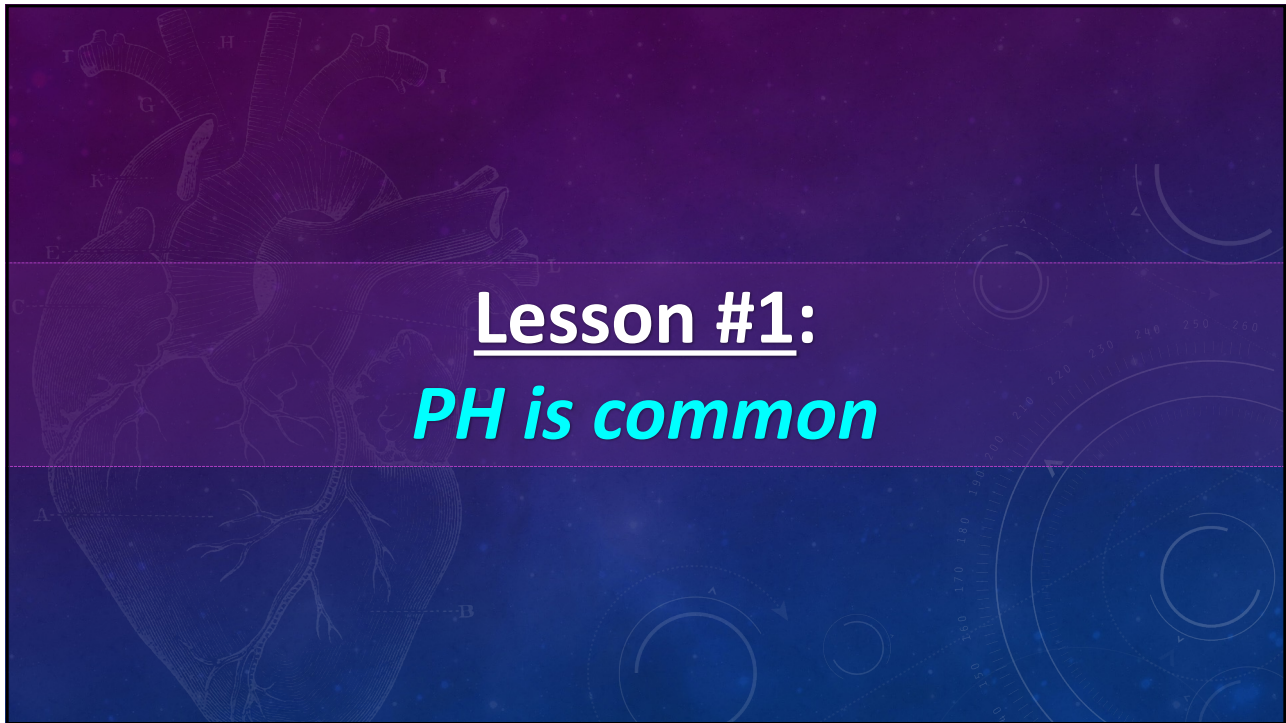


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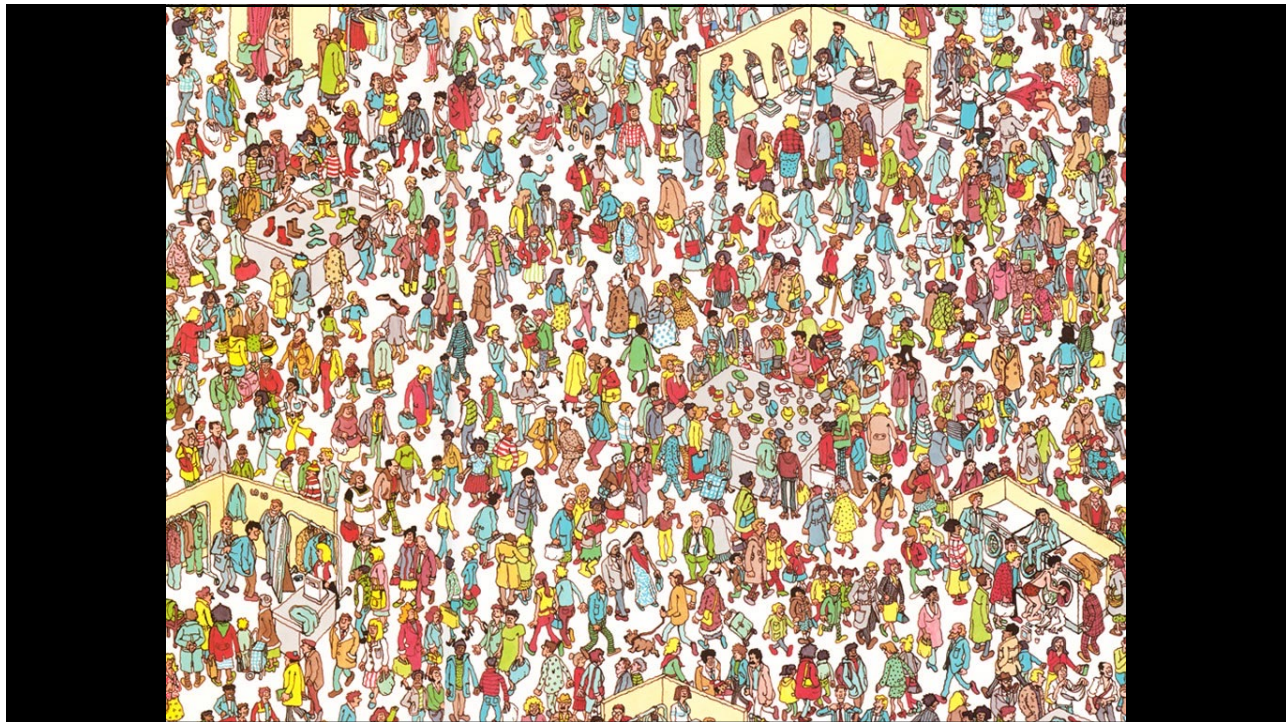
Group 1	Group 2	Group 3	Group 4	Group 5
iPAH, HPAH	Valvular Heart Disease	Lung Diseases	CTEPH	Blood disorders
PAH Associated: CTD HIV Portal HTN CHD	Some CHD	Hypoxia, No Lung Disease		Metabolic Syndromes
PVOD		Hypovent. Syndromes		Combined Diseases
Drug/toxin-induced		Developm. Lung Disorders		Some CHD

Maron B, et al. *Am J Resp Crit Care Med* 2021

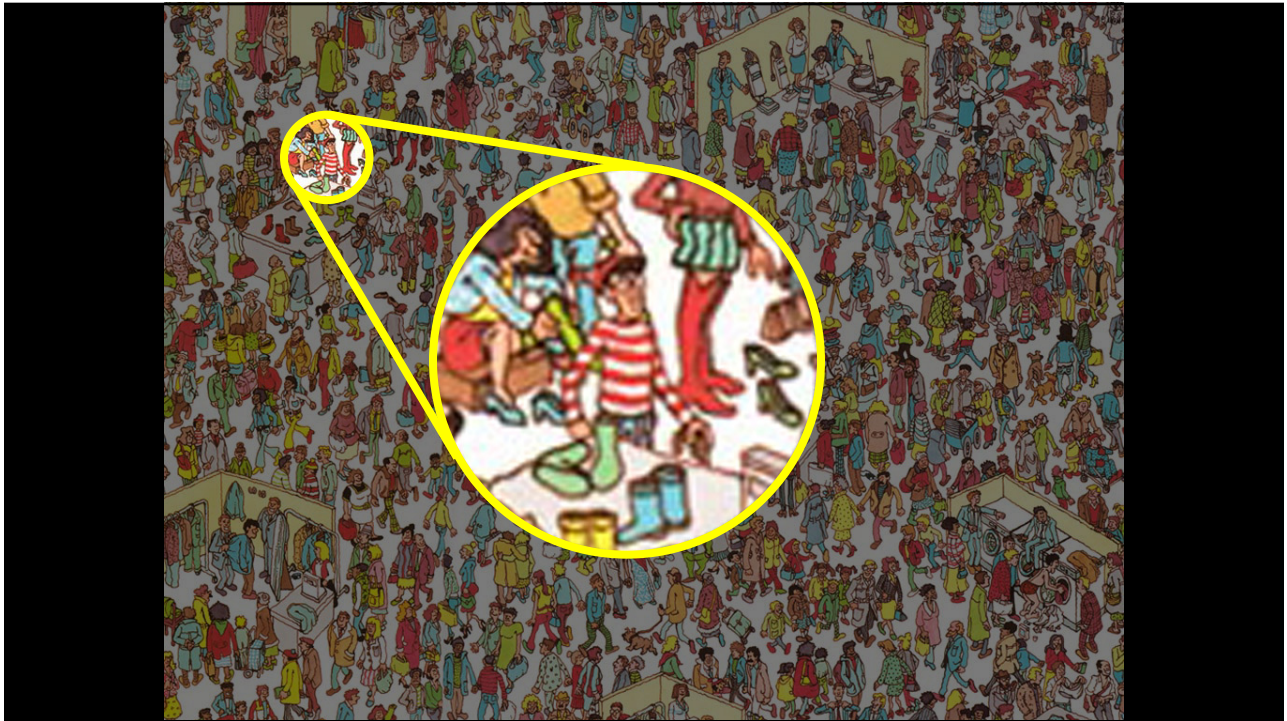
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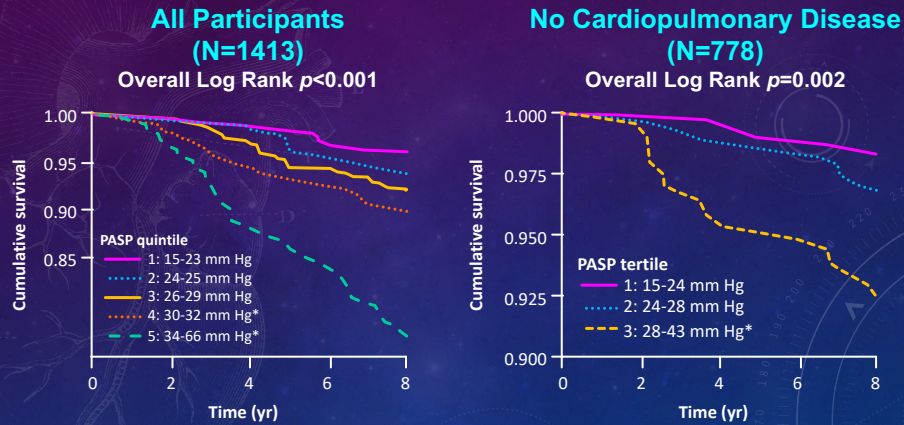


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PH in the community



Up to 20% of the US population has echo evidence of PH

Lam CSP et al. *Circulation* 2009

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PH due to left heart disease

- Most common cause of PH:
 - ✓ 80% of HFpEF: elevated PA systolic pressure on echo
- Associated with high morbidity + mortality
- Hemodynamic classification has been debated:
 - ✓ Isolated post-capillary PH (IpcPH):
 - DPG (PADP-PCWP) < 7 mmHg *and* PVR ≤ 3 WU
 - ✓ Combined pre-post capillary PH (CpcPH):
 - DPG ≥ 7 mmHg *or* PVR > 3 WU
- No approved therapies

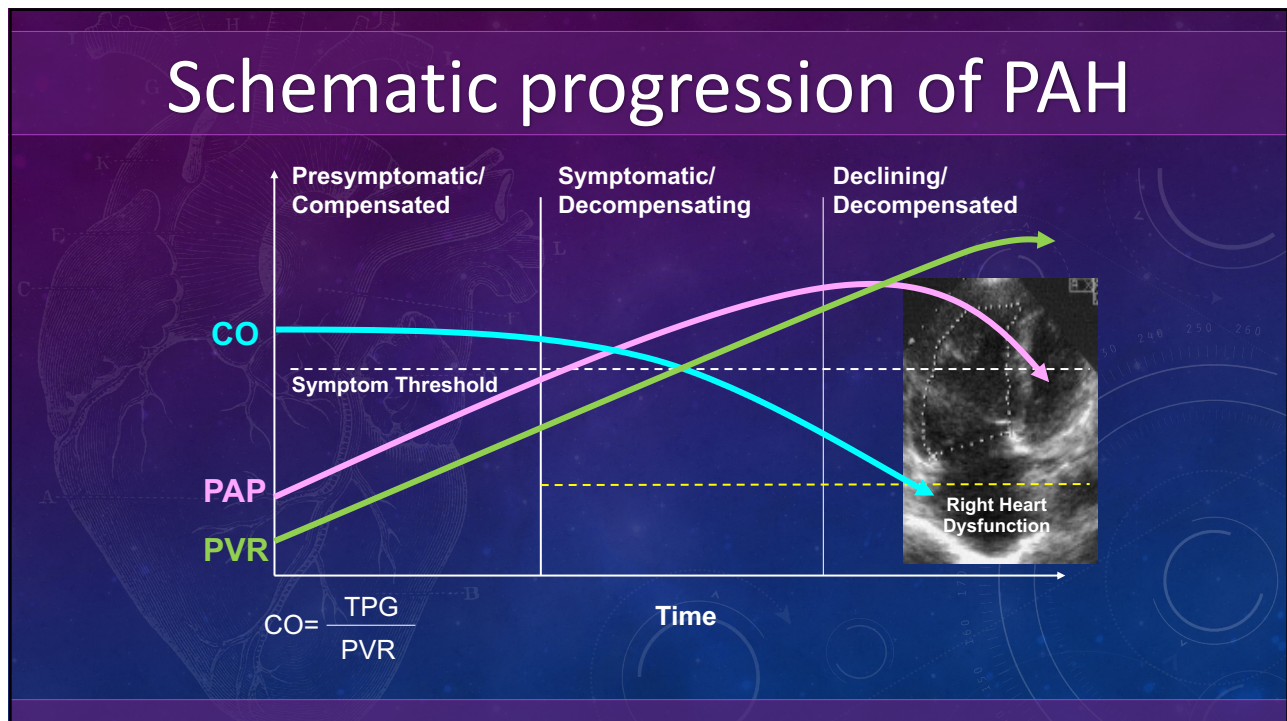
Lam CS, et al. *JACC* 2009; Rosenkranz S, et al. *Eur Heart J* 2016

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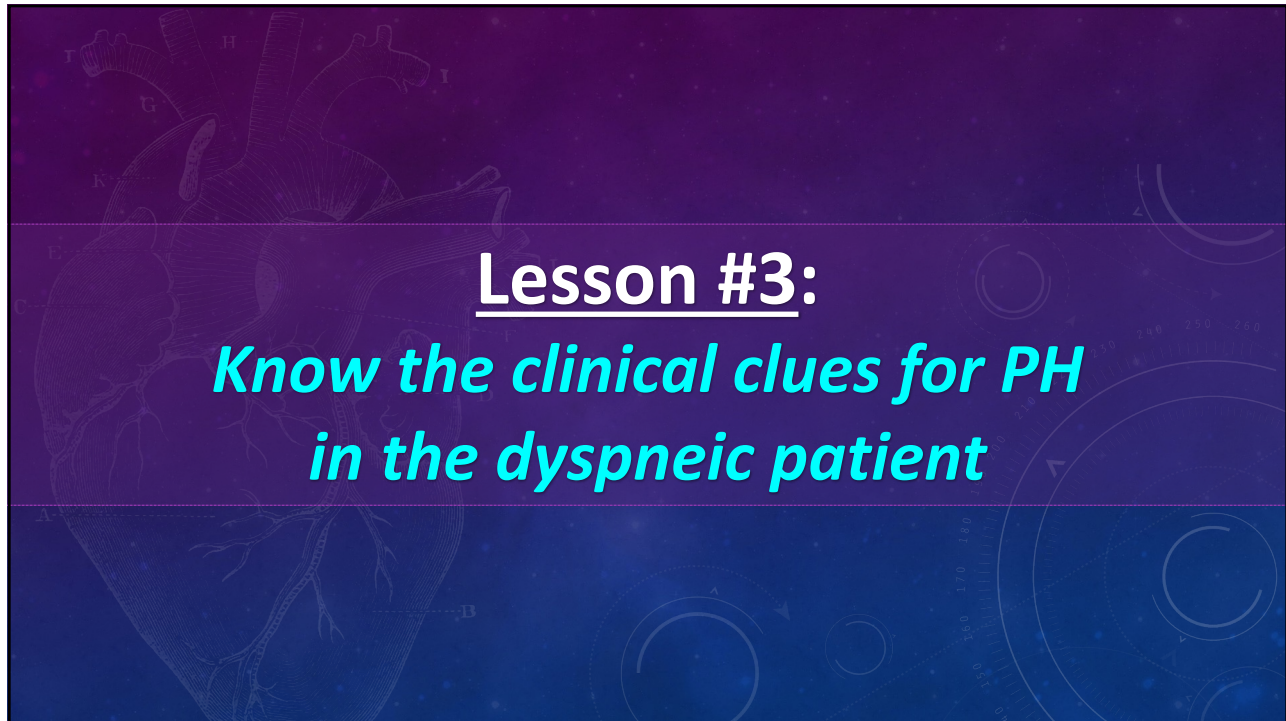
Lesson #2:

***WHO Group I PAH is rare but deadly—
make the diagnosis early when it is
treatable***

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A slide with a dark blue background featuring a faint anatomical illustration of a human heart. The text is centered and reads:

Is there a reason to suspect PAH?

- Risk factors:
 - ✓ Family history
 - ✓ Connective tissue disease
 - ✓ Congenital heart disease
 - ✓ Portal hypertension—liver transplant candidate
 - ✓ Environmental/drug factors
 - ✓ HIV
 - ✓ Lung disease / OSA
 - ✓ Chronic PE

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Is there a reason to suspect PAH?

- Clinical presentation:

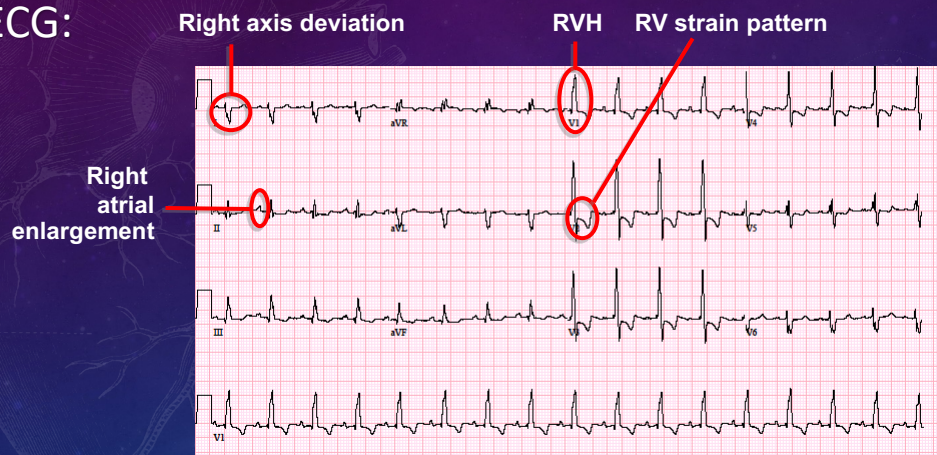
History	Exam (PH)	Exam (RV Failure)
<ul style="list-style-type: none"> Dyspnea (86%) Fatigue (27%) Chest pain (22%) Edema (22%) Syncope (17%) Dizziness (15%) Cough (14%) Palpitations (13%) 	<ul style="list-style-type: none"> Loud P2 (listen at apex) RV lift (left parasternal – fingertips) RV S3, S4 Systolic murmur (TR; ↑with inspiration) Diastolic murmur (PR) 	<ul style="list-style-type: none"> JVD; increased A wave, V wave; hepatjugular reflex Pulsatile liver Hepatomegaly Edema Ascites Low BP, low PP, cool extremities

REVEAL study. Brown LM et al. *Chest*. 2011;140:19-26.
Adapted from McLaughlin VV et al. *J Am Coll Cardiol*. 2009;53:1573-1619.

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Is there a reason to suspect PAH?

- ECG:

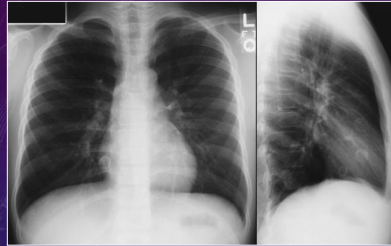


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Is there a reason to suspect PAH?

- CXR:

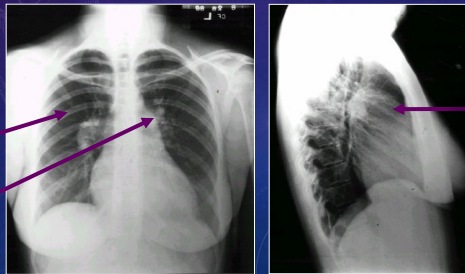
Normal



Abnormal

Peripheral hypo-vascularity (pruning)

Prominent central pulmonary artery



RV enlargement into retrosternal clear space

Adapted from McLaughlin VV et al.
J Am Coll Cardiol. 2009;53:1573-1619.

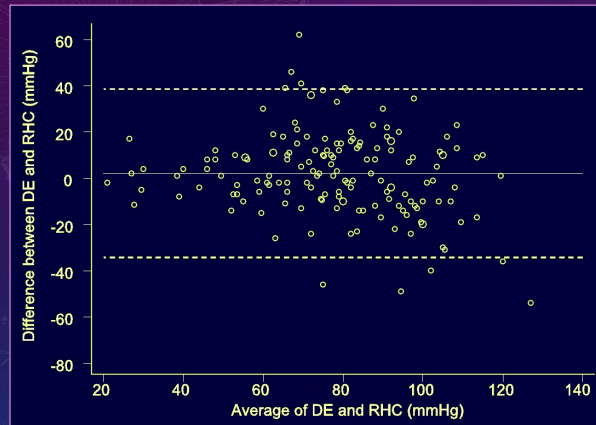
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Lesson #3:

PA systolic pressure is the last thing you should look at on echo when evaluating for PH

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PA systolic pressure: echo vs. invasive



N=160 consecutive patients with known or suspected PH

High correlation ($r=0.7$, $p<0.001$)

But...poor agreement

Rich J, Shah SJ, et al. Chest 2010

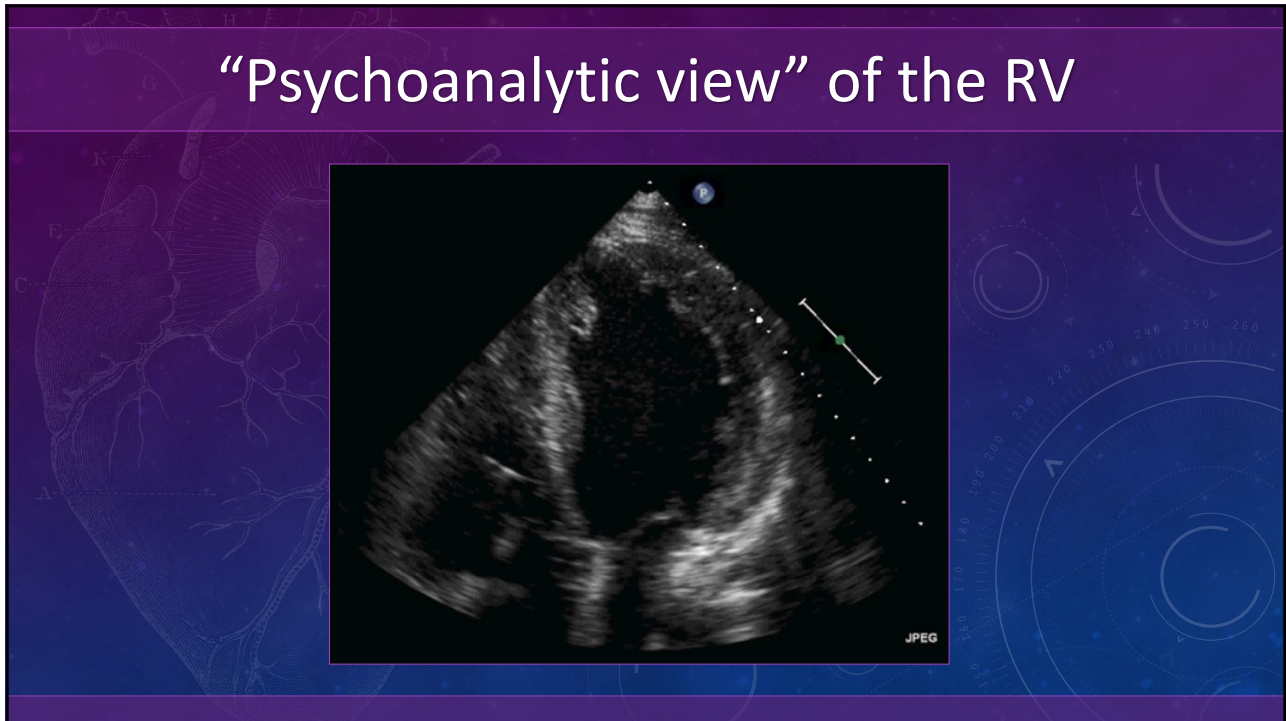
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Problems with Doppler PASP

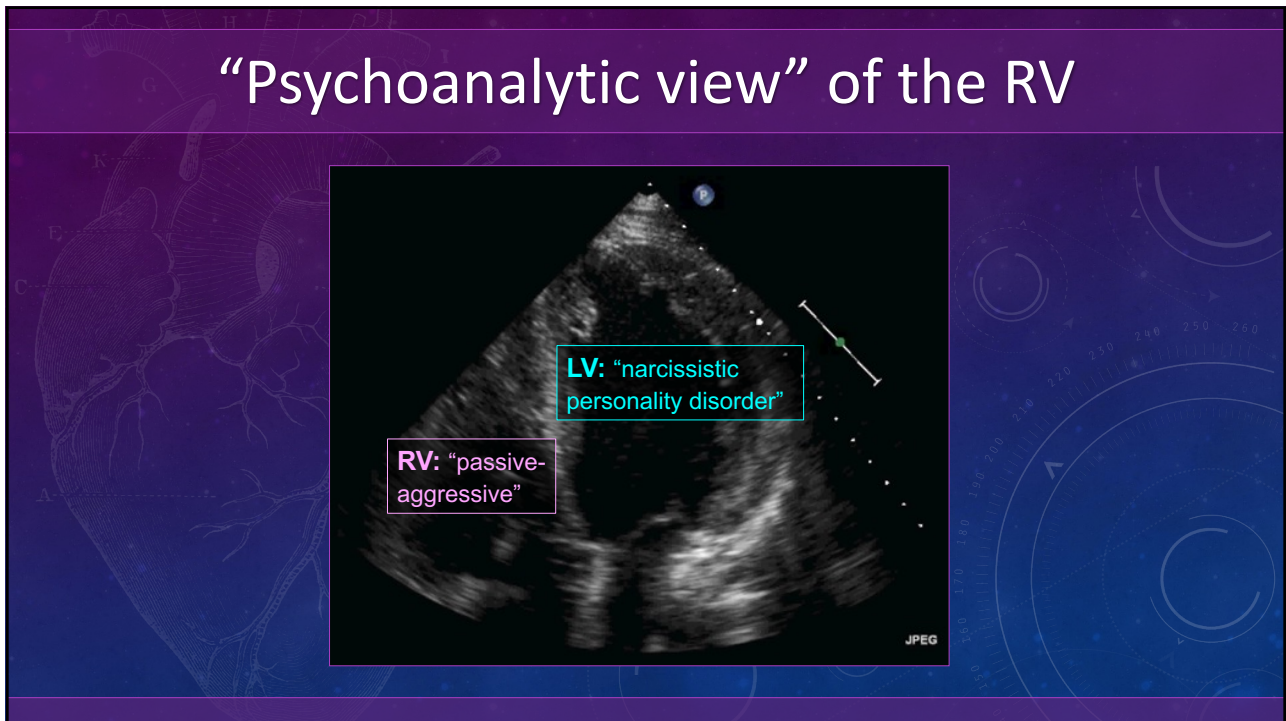
- Ultrasound / Doppler:
 - ✓ Poor acoustic windows
 - ✓ Inter-observer variability (under- and over-measured)
 - ✓ Poor Doppler beam alignment
- Bernoulli:
 - ✓ Viscosity (affected by RBC deformity, anemia, hypoxemia)
 - ✓ Pressure-recovery phenomenon
- Other causes of elevated PASP besides elevated PVR
 - ✓ Elevated left atrial pressure
 - ✓ Elevated cardiac output
 - ✓ Elevated systemic blood pressure

Shah SJ. JAMA 2012

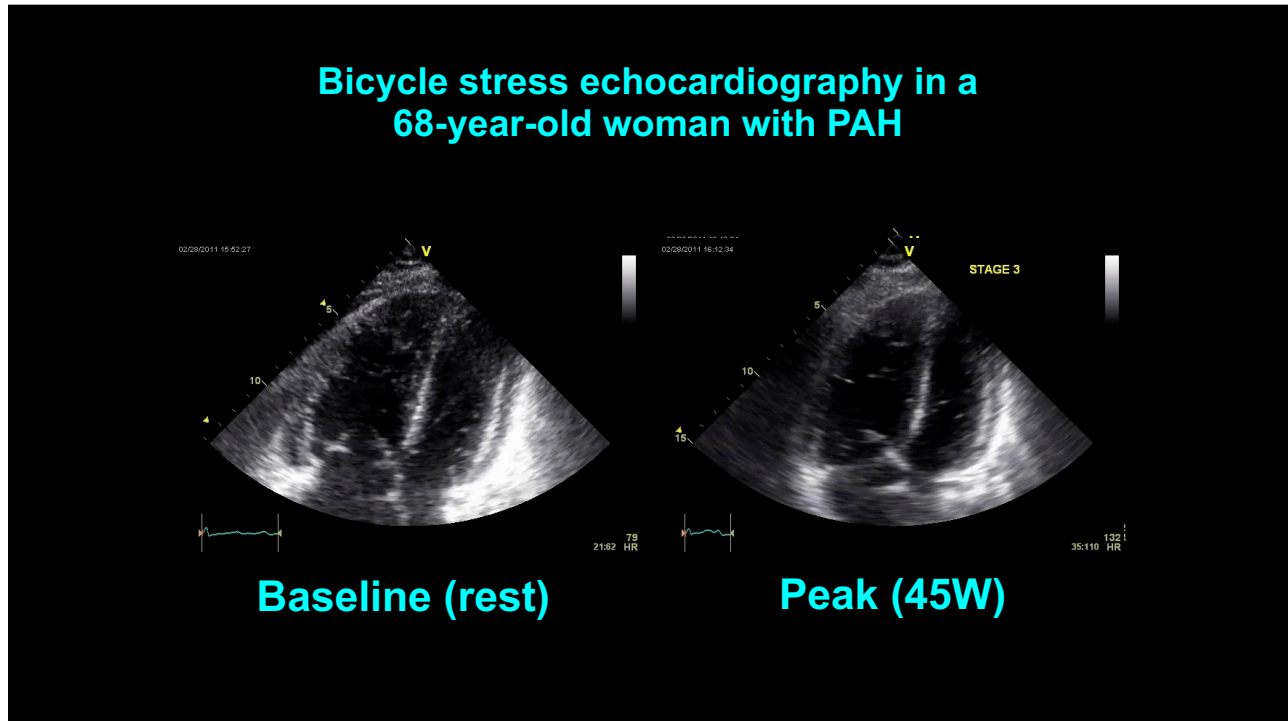
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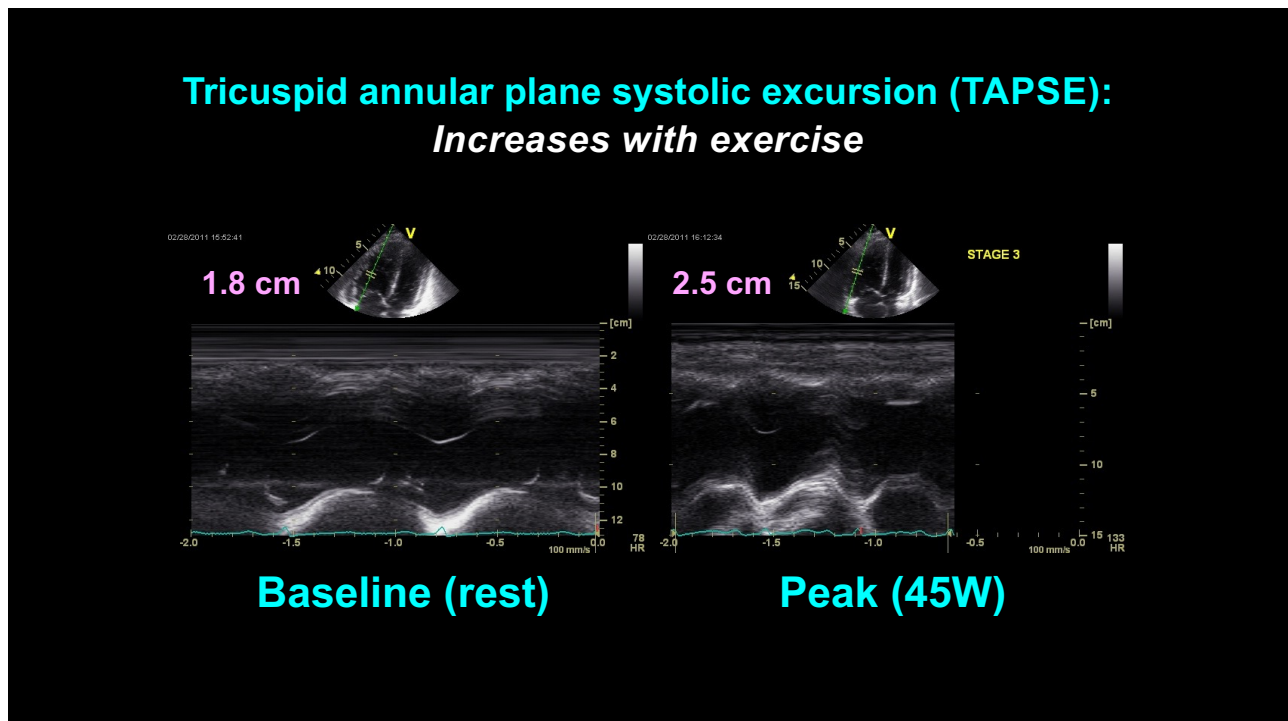
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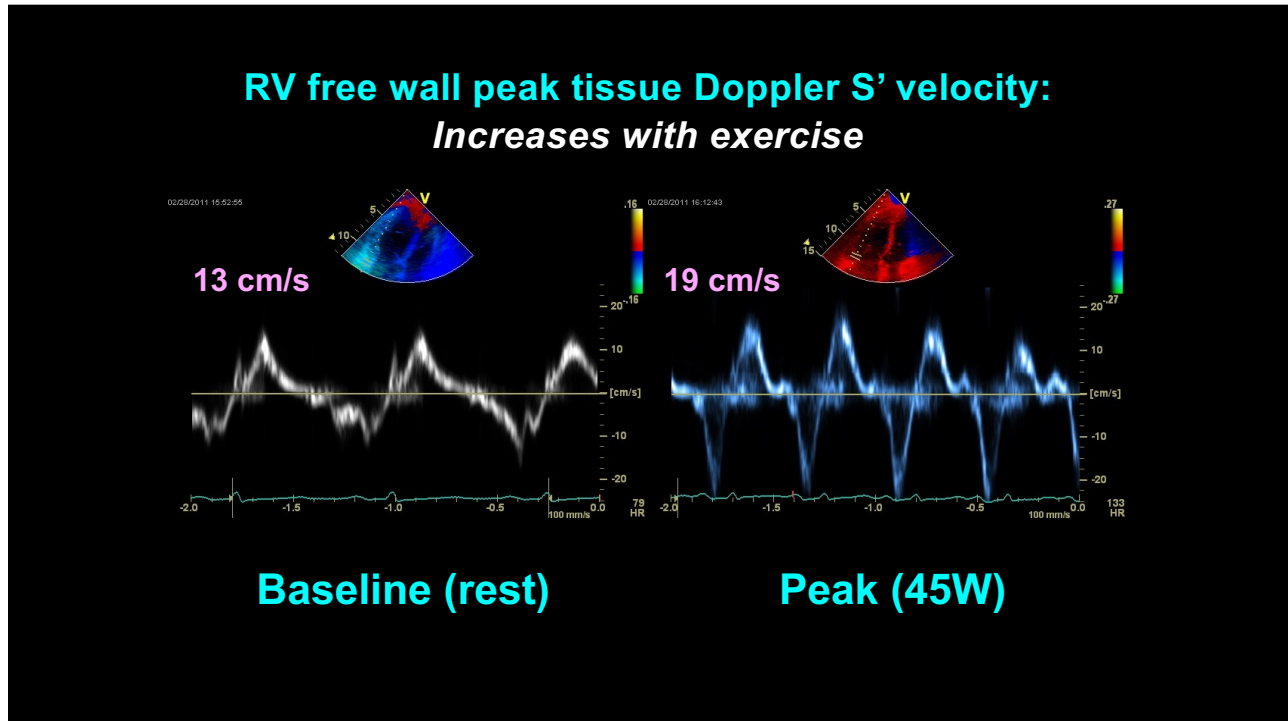
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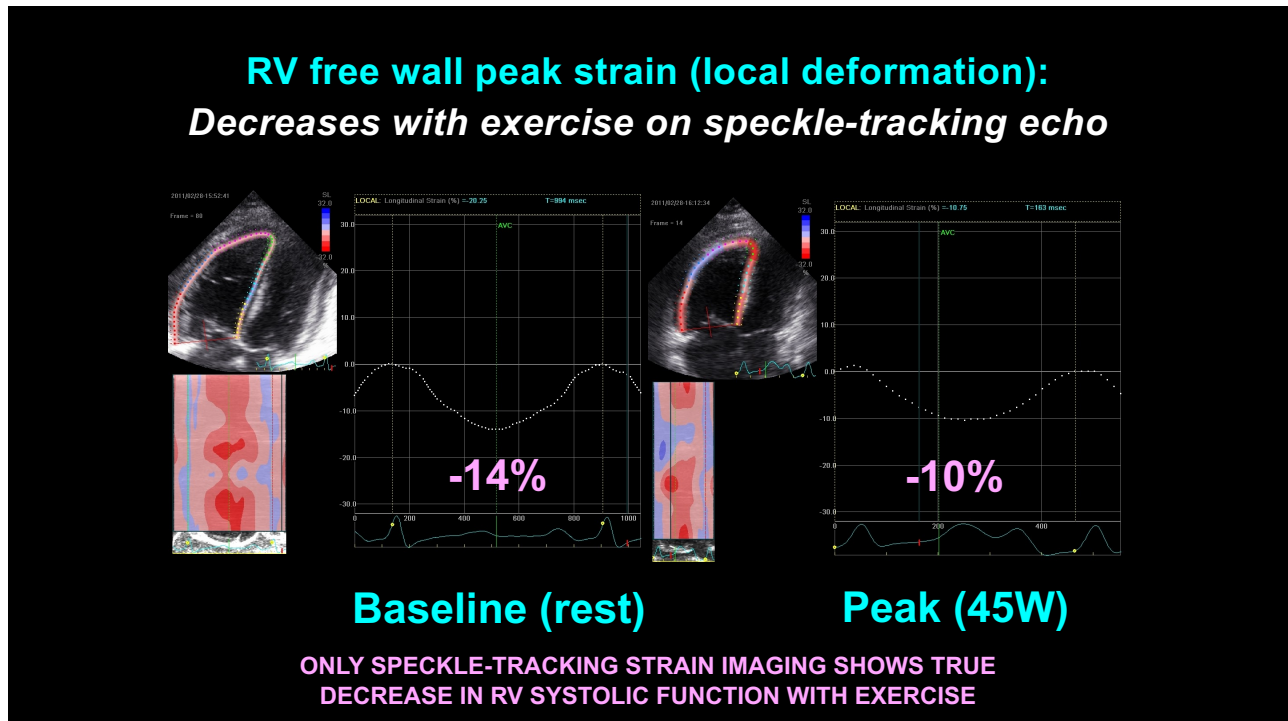
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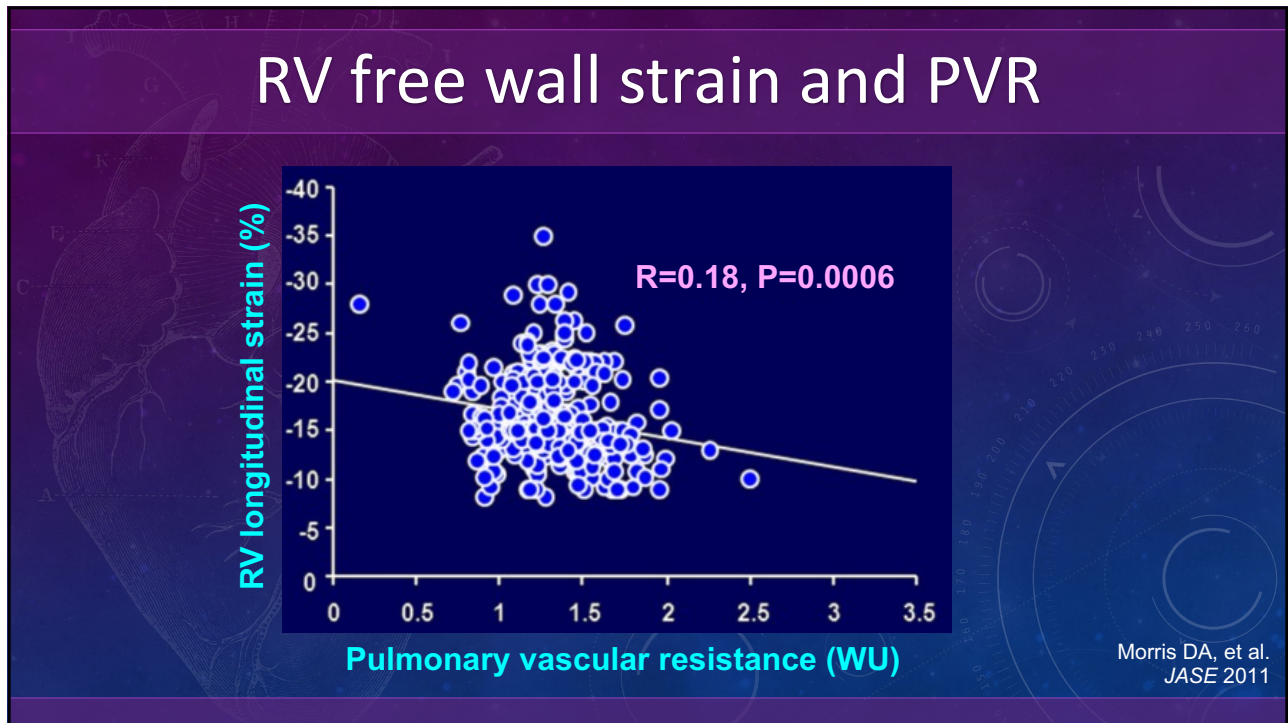
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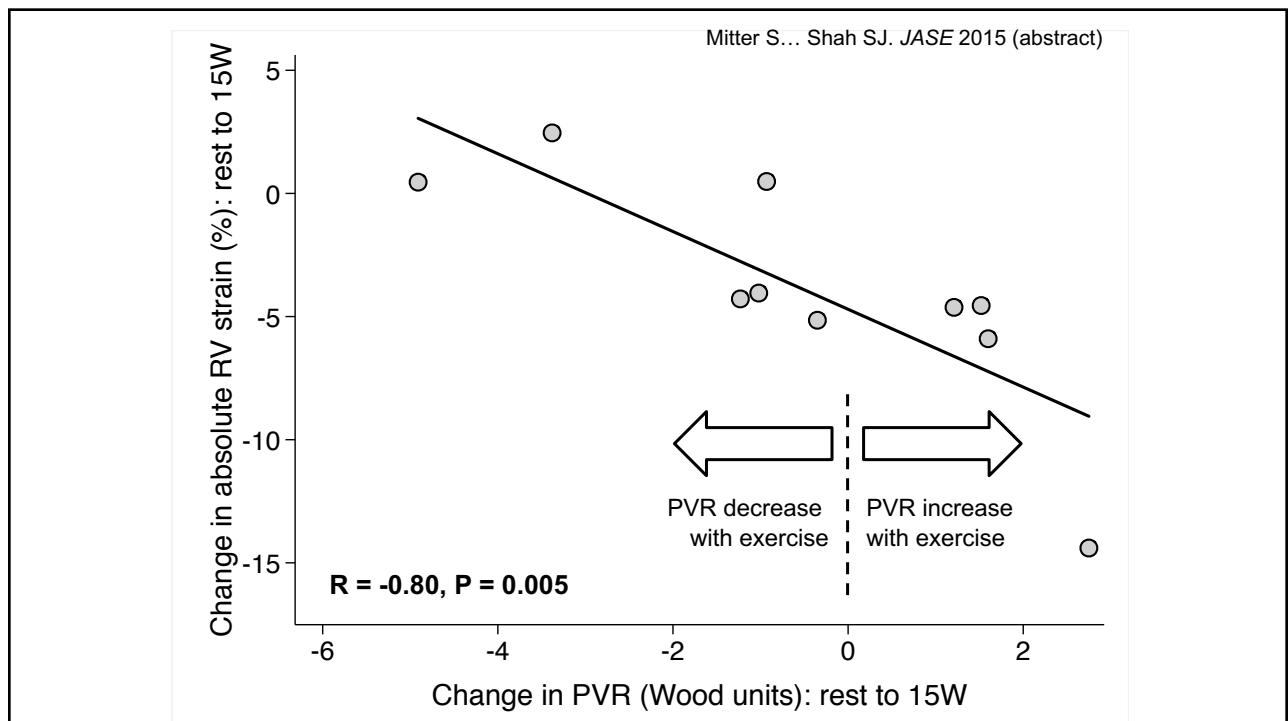
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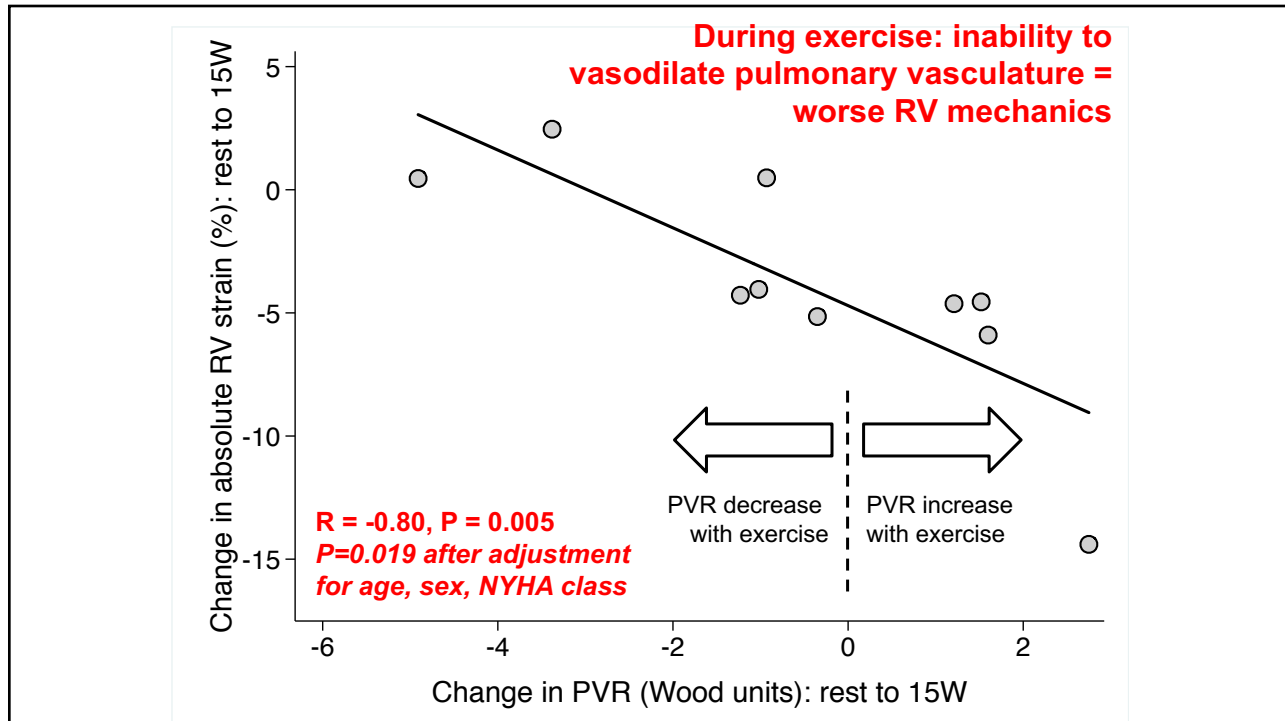
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Change in RV strain with exercise

ΔRV free wall strain (%-units)
Baseline to 15W exercise

P=0.016

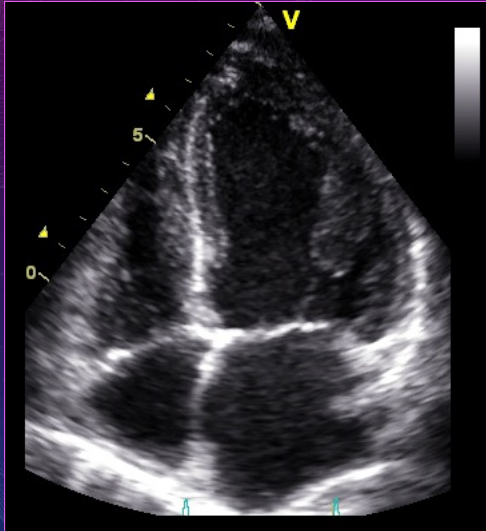
Idiopathic PAH | Scleroderma PAH

- No difference in baseline RV strain, ΔPVR or ΔZ_c between PAH subtypes
- More intrinsic RV dysfunction in scleroderma, unmasked by exercise speckle-tracking echo

Mitter S... Shah SJ. JASE 2015 (abstract)

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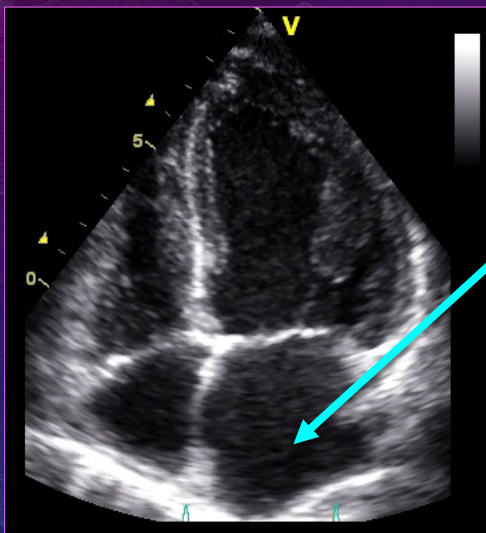
PAH vs. PVH: Practical tips on echo



Normal LVEF + \uparrow PASP?
Think PVH (HFpEF) until
proven otherwise

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PAH vs. PVH: Practical tips on echo

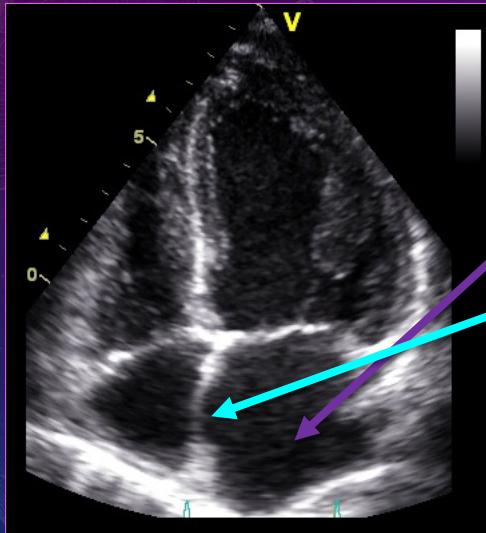


Normal LVEF + \uparrow PASP?
Think PVH (HFpEF) until
proven otherwise

Left atrial enlargement
(LA size > RA size)

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PAH vs. PVH: Practical tips on echo



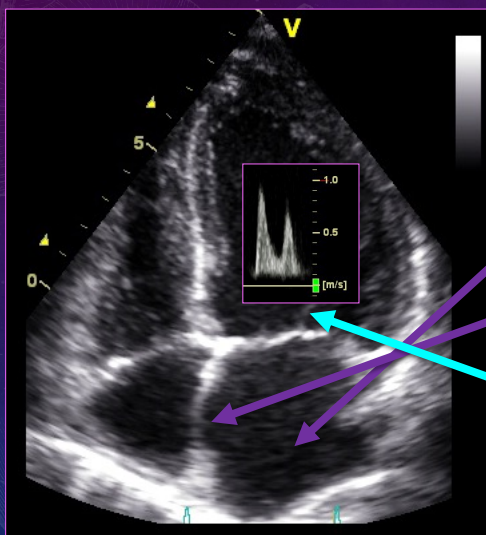
Normal LVEF + \uparrow PASP?
Think PVH (HFpEF) until
proven otherwise

Left atrial enlargement
(LA size > RA size)

Interatrial septum bows
from left to right

37

PAH vs. PVH: Practical tips on echo



Normal LVEF + \uparrow PASP?
Think PVH (HFpEF) until
proven otherwise

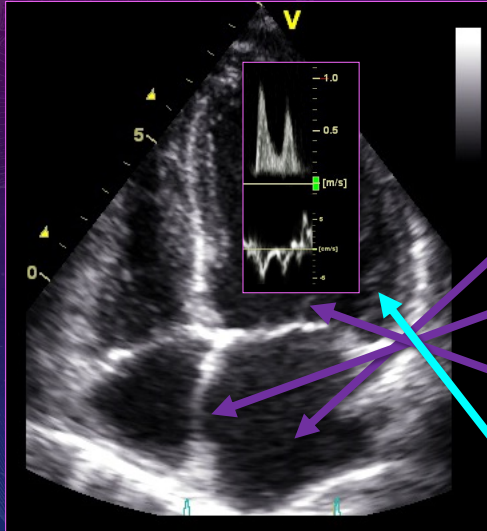
Left atrial enlargement
(LA size > RA size)

Interatrial septum bows
from left to right

Grade 2+ diastolic
dysfunction (\uparrow E/A ratio)

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PAH vs. PVH: Practical tips on echo



Normal LVEF + \uparrow PASP?
Think PVH (HFpEF) until proven otherwise

Left atrial enlargement (LA size > RA size)

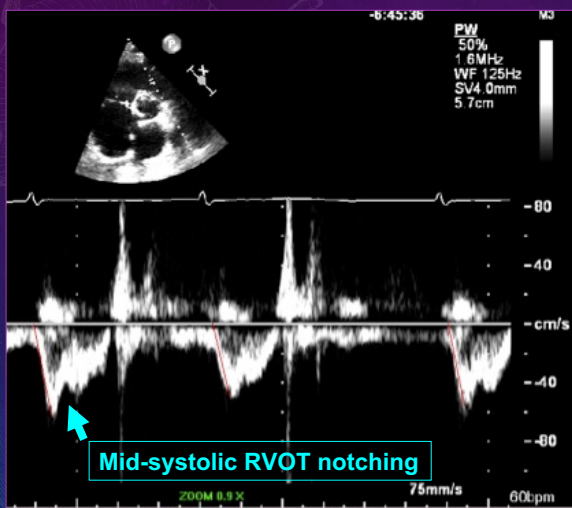
Interatrial septum bows from left to right

Grade 2+ diastolic dysfunction (\uparrow E/A ratio)

Decreased lateral e'
Elevated lateral E/e'

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RVOT notching: sign of \uparrow PVR



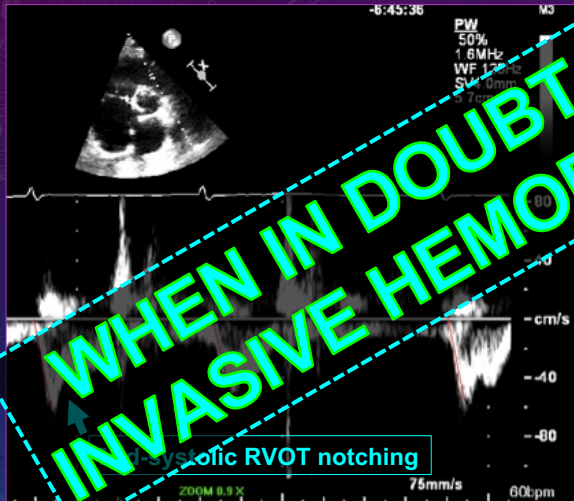
RVOT notching:
N=293 HFpEF pts

77%: no notching
6%: mid-systolic notch
3%: late-systolic notch
14%: uninterpretable

Notching associated with \uparrow PVR, \uparrow DPG (\uparrow PADP-PCWP) (P<0.01)

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RVOT notching: sign of ↑PVR



**WHEN IN DOUBT CHECK
INVASIVE HEMODYNAMICS**

Diastolic notching
Late-systolic notch
4%: uninterpretable

Notching associated
with ↑PVR, ↑DPG
(↑PADP-PCWP)
($P < 0.01$)

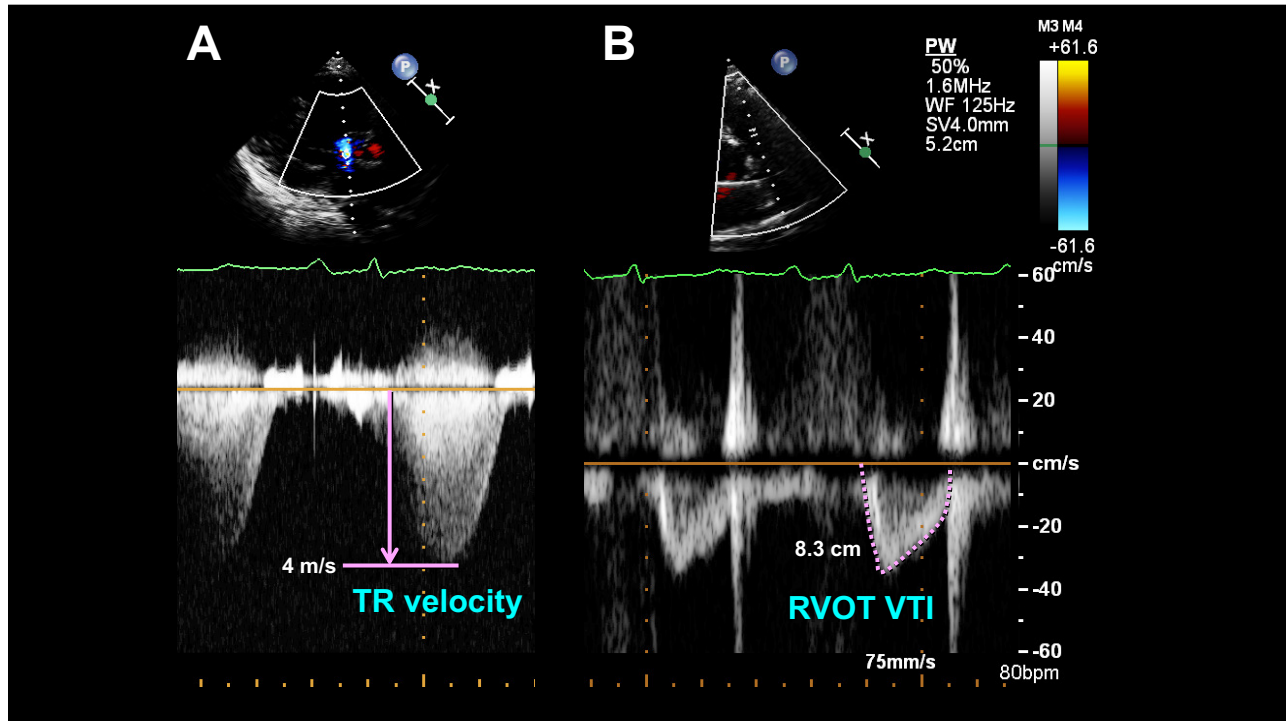
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Estimation of PVR on echo

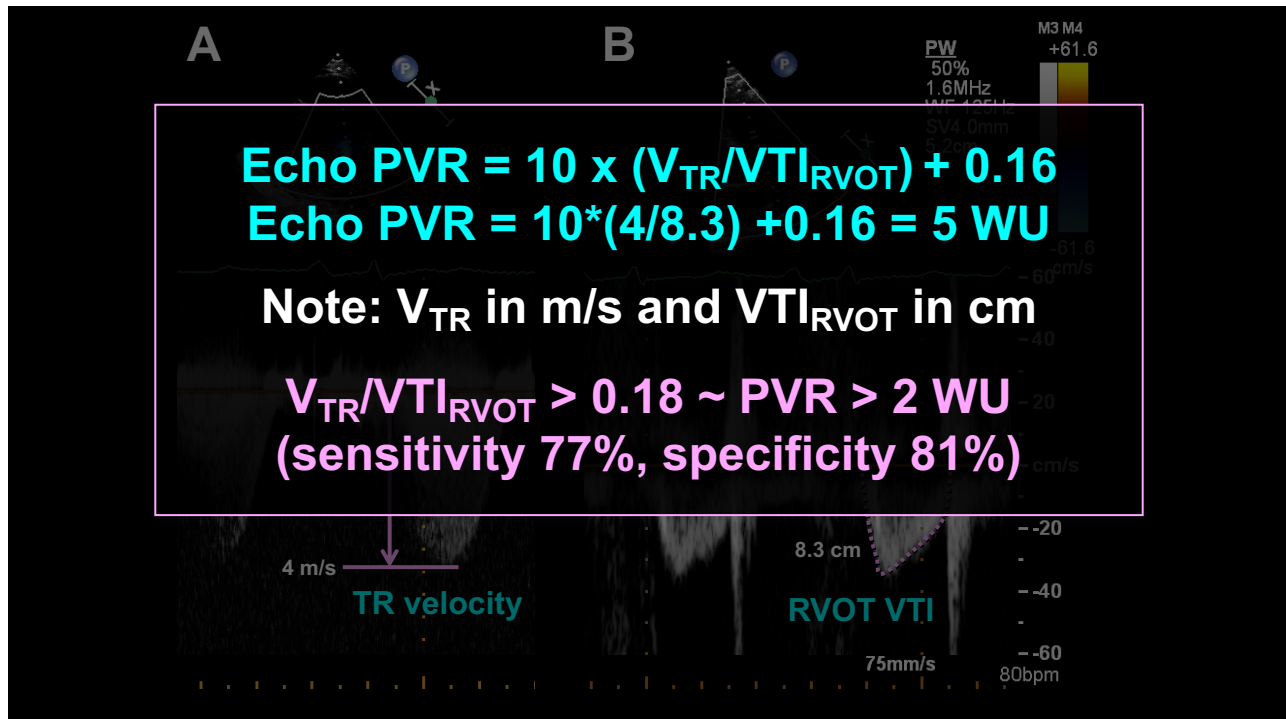
- $PVR = (mPAP - PCWP) / CO$
- Total pulmonary resistance = $mPAP / CO$
- $PVR_{echo} = [10 \times (V_{TR} / VTI_{RVOT})] + 0.16$
- Works well at identifying elevated PVR
- Underestimates $PVR_{invasive}$ at very high PVR
- Overestimates $PVR_{invasive}$ in patients with elevated PCWP
- Really a measure of TPR: total pulmonary resistance

Abbas AE, et al. J Am Coll Cardiol 2003; Farzaneh-Far R, Am J Cardiol 2008

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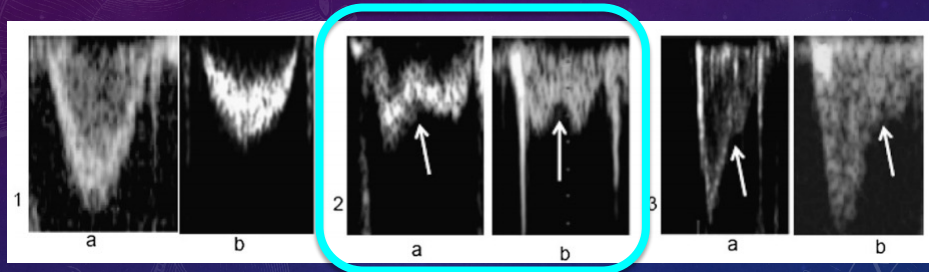


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PW Doppler of RVOT outflow notching



Sign of severe PA stiffness
↑PVR

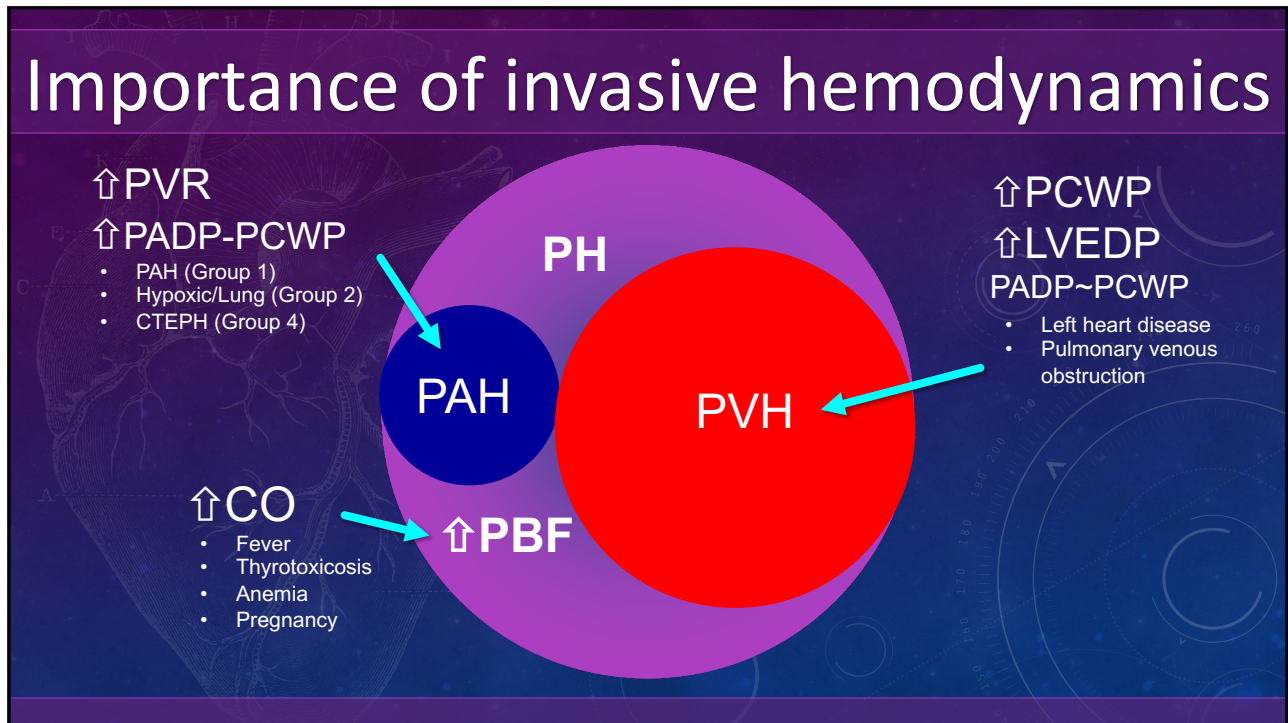
Arkles, et al. *Am J Resp Crit Care Med* 2011

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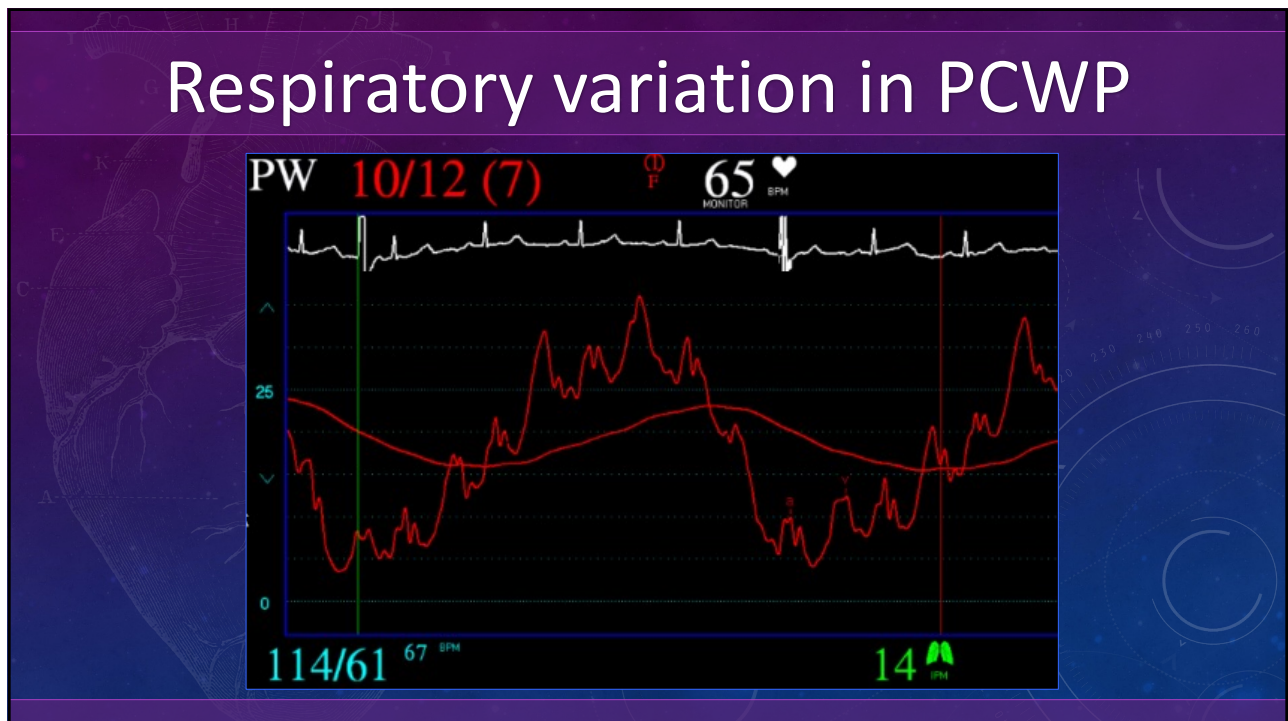
Lesson #5:

***Definitive diagnosis of PH requires
 invasive hemodynamic testing...
 done correctly!***

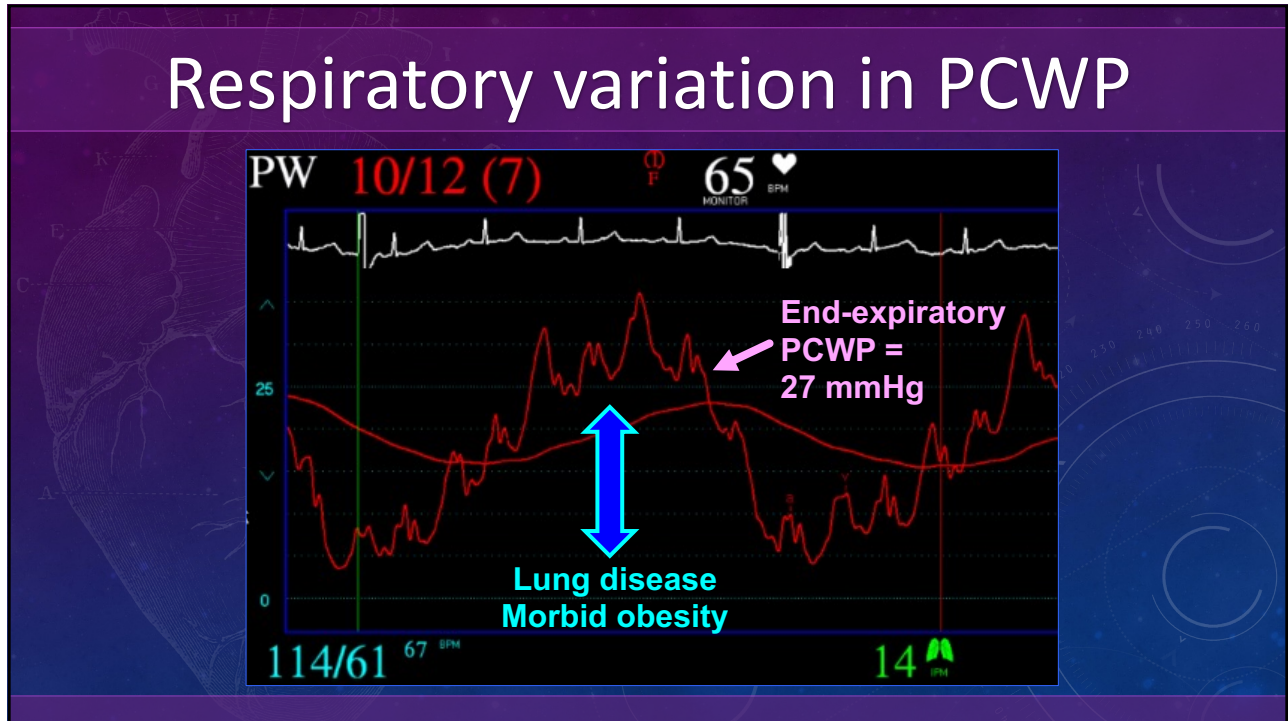
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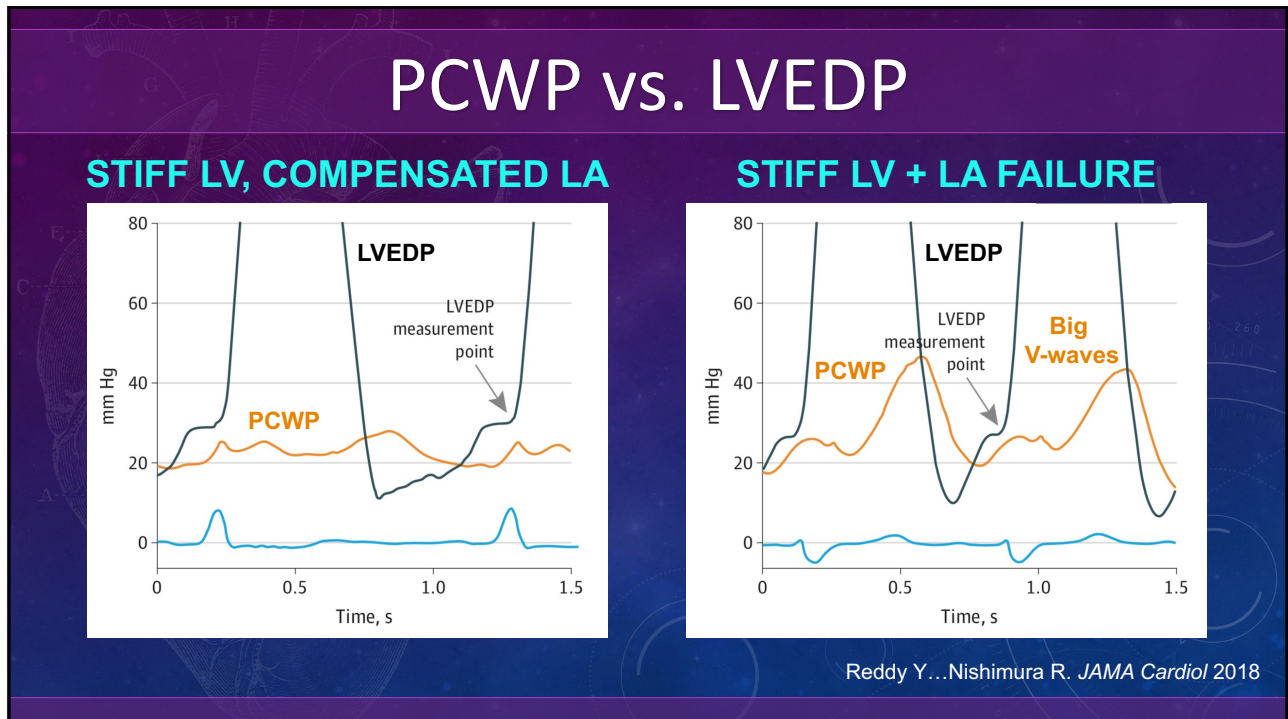
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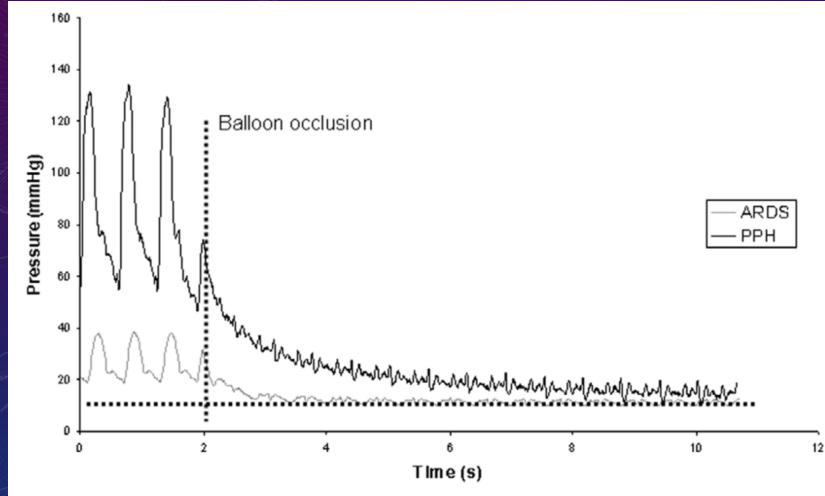


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Did you get the true PCWP?

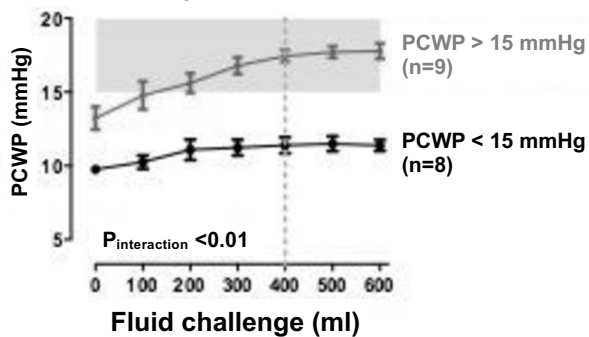


Souza R, et al. *Crit Care* 2005

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What about the fluid status?

Occult PVH in patients with PAH at rest



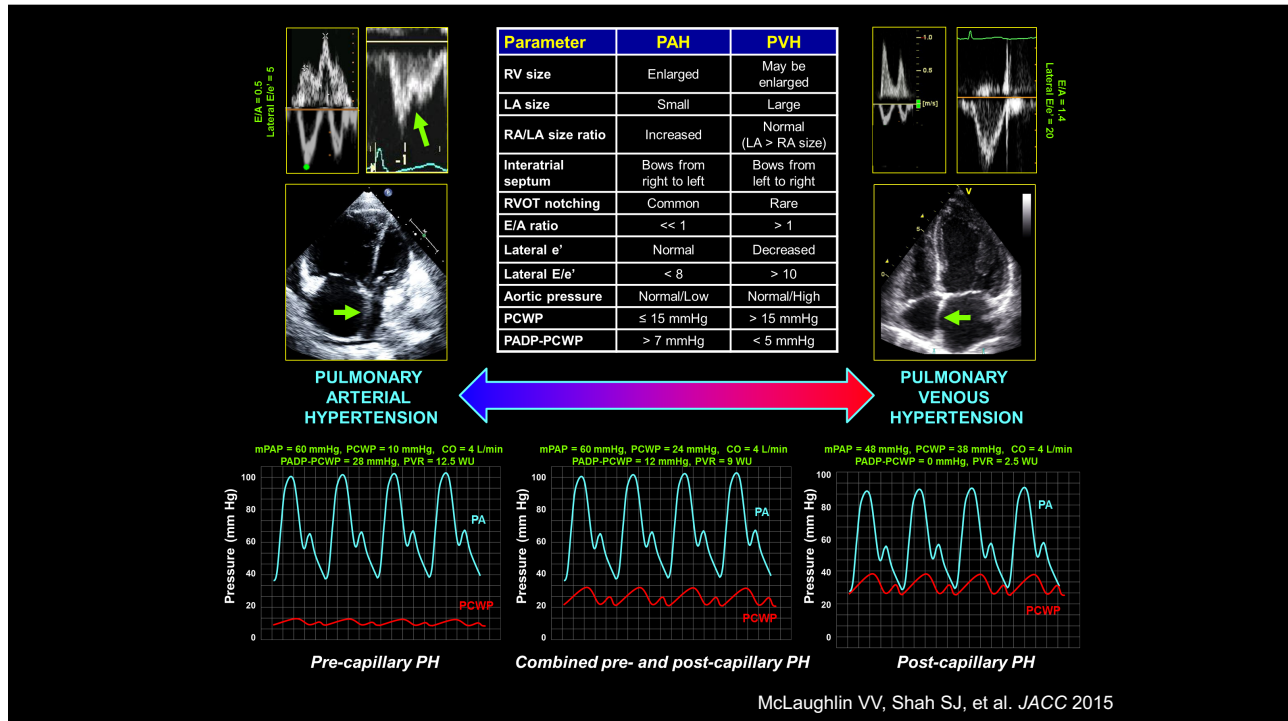
Oosterveer FPT, et al. *Eur Respir J* 2013

Predictors of occult PVH after fluid challenge in patients with PAH at rest (multivariable analysis)

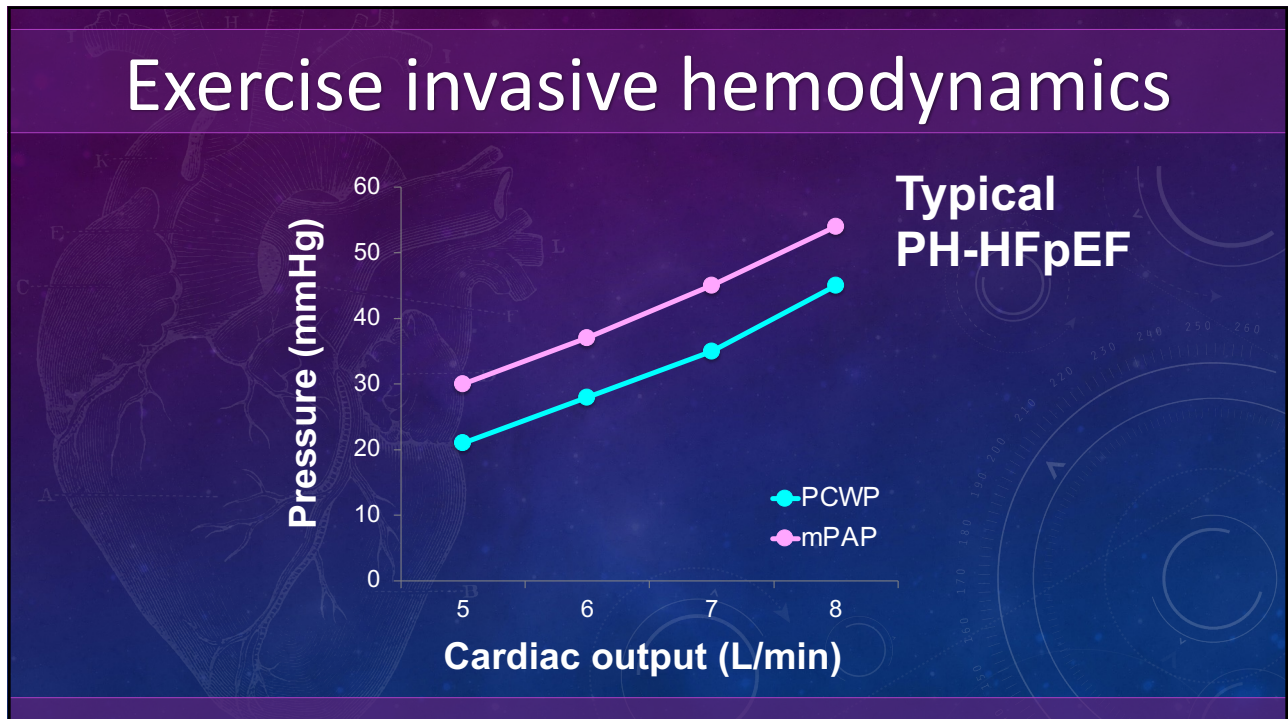
Variables	OR	95% CI	P Value
Age	1.030	1.001–1.060	0.046
HTN	2.337	1.106–4.938	0.026
BMI	1.057	1.005–1.113	0.032
LVH	1.910	0.830–4.395	0.113

Robbins IM, et al. *Circ Heart Fail* 2014

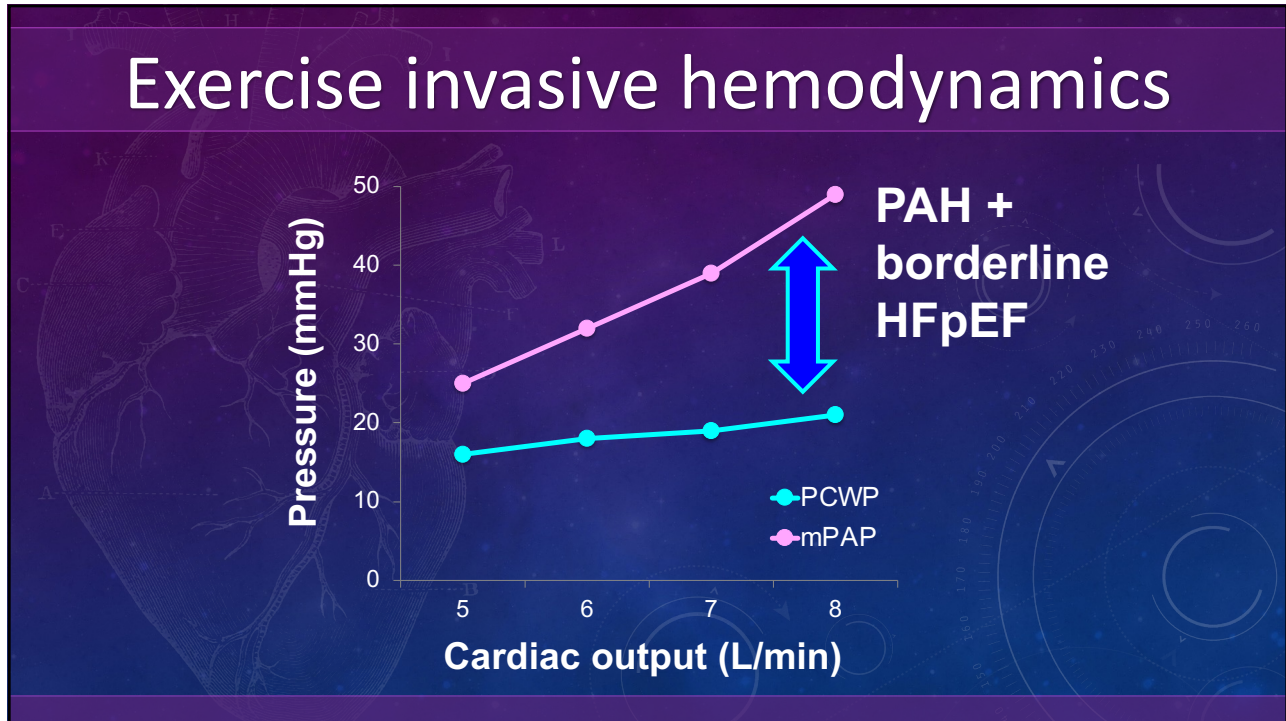
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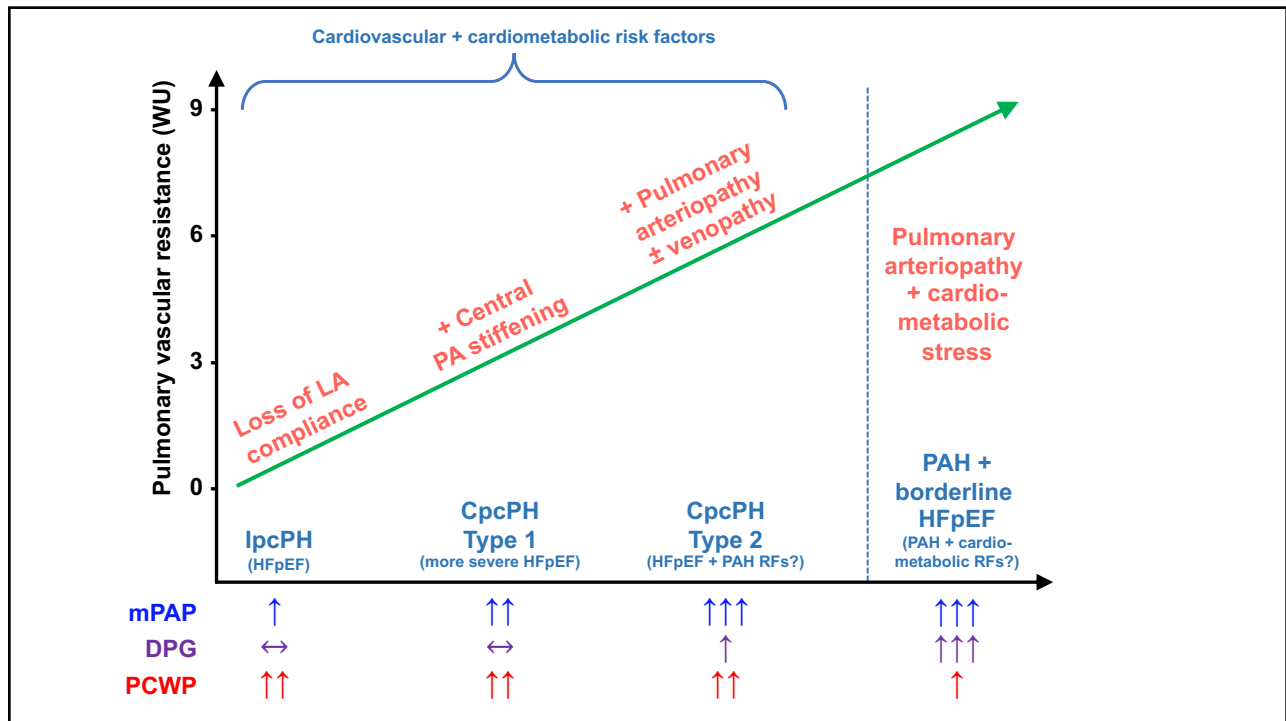
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Take home points

- PH is common, but most often due to left heart disease
- PAH is rare but deadly: outcomes have improved but diagnosis must be made as early as possible
- Know the PH clinical clues in the dyspneic patient
- Look beyond PA pressure to the health of the RV, and know the other echo signs of PH (and PAH vs. PVH)
- Definitive diagnosis of PH requires invasive hemodynamics, but don't forget to integrate the echo findings!

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