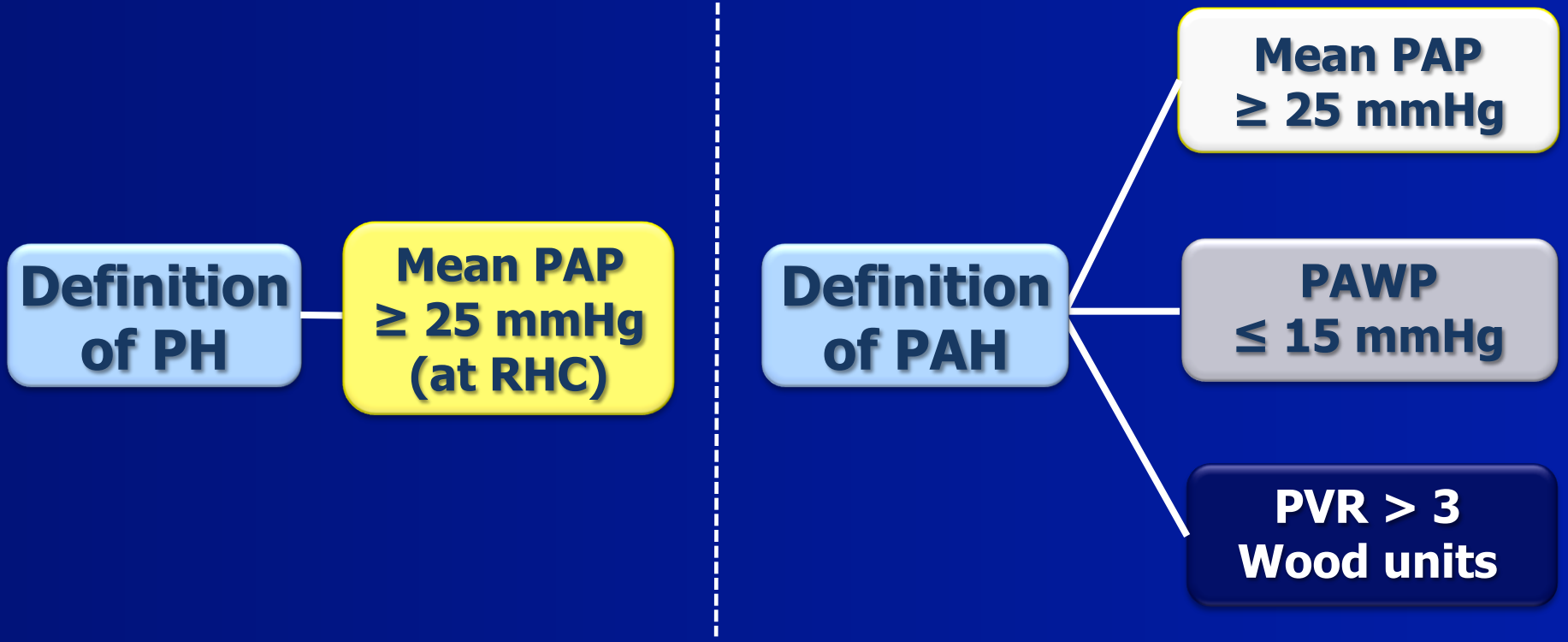
IMAGING NELL'IPERTENSIONE POLMONARE (dalla diagnosi alla prognosi)

Michele D'Alto
mic.dalto@tin.it

UOC Cardiologia
II Università degli Studi, A.O. "V. Monaldi" - Napoli

5th World Symposium on PH: Haemodynamic definition of PAH

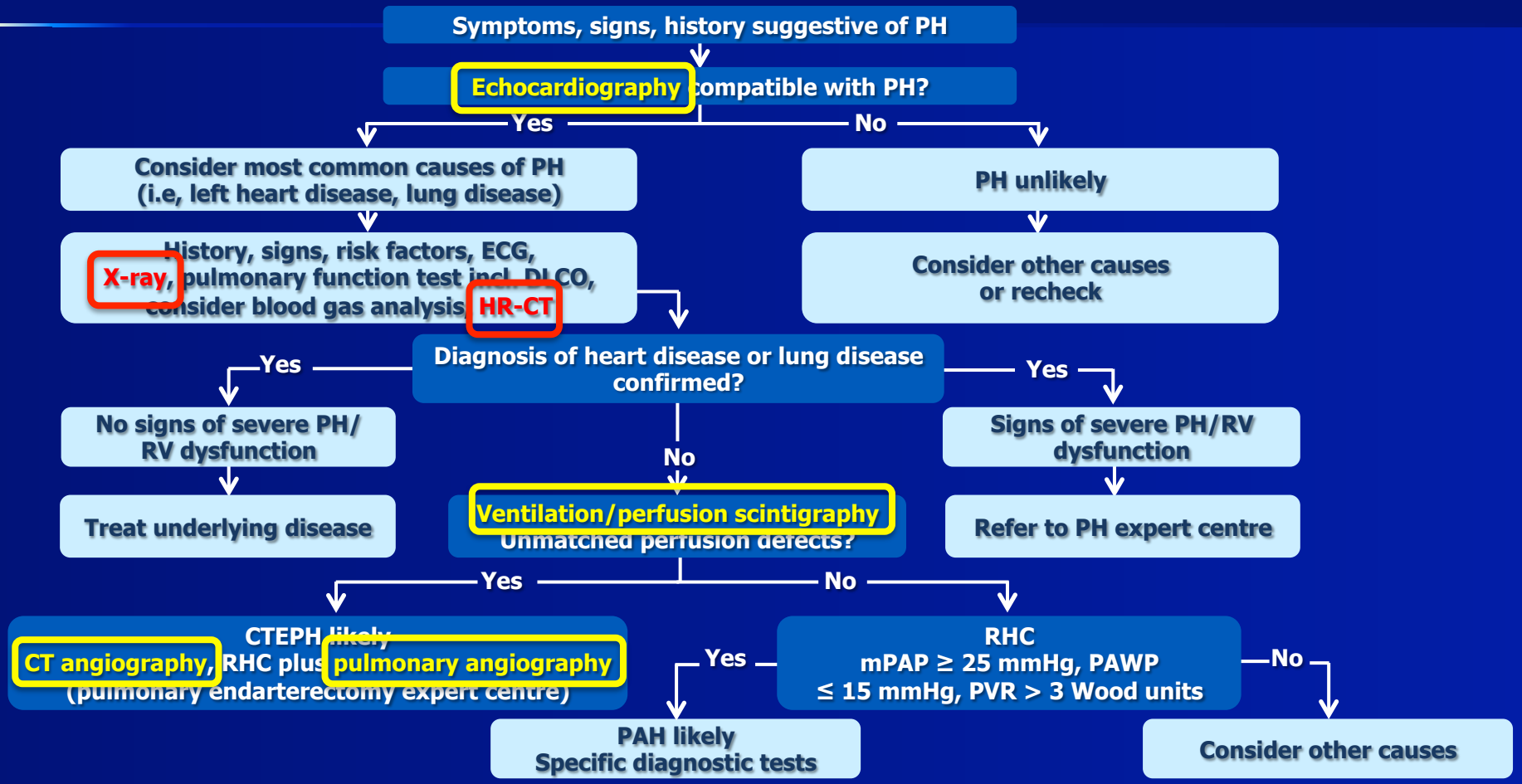


Other causes of pre-capillary PH must be excluded

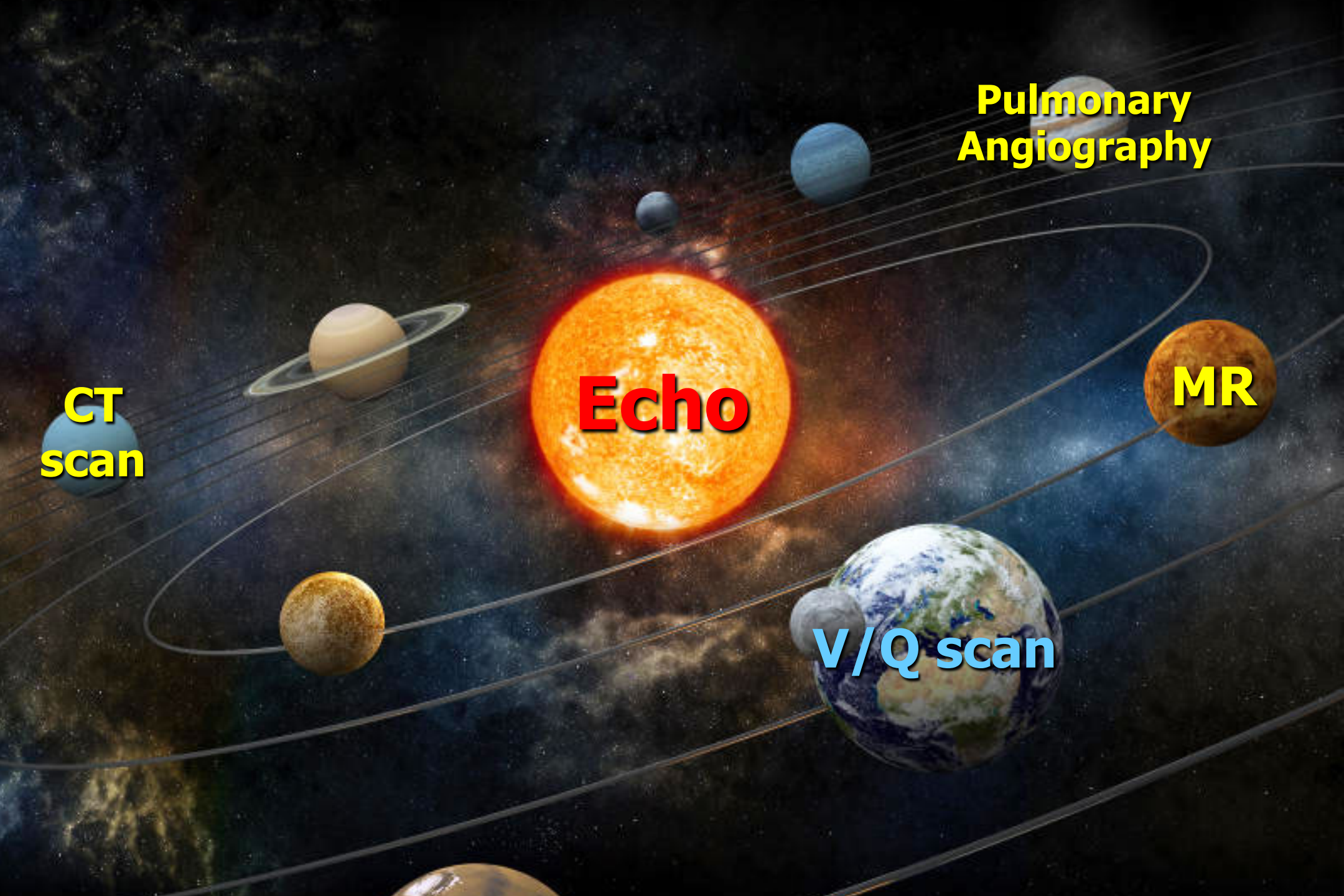
PAP: pulmonary arterial pressure; PAWP: pulmonary artery wedge pressure; PVR: pulmonary vascular resistance

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

5th World Symposium on PH: Diagnosis of PAH



Hoeper M , JACC 2013;62:D42-50


Pulmonary
Angiography
CT
scan
Echo
MR
V/Q scan

Echo for PAH diagnosis?

Point: Can Doppler Echocardiography Estimates of Pulmonary Artery Systolic Pressures Be Relied Upon to Accurately Make the Diagnosis of Pulmonary Hypertension? Yes

Rudsky LG, CHEST 2013;143:1536-39

Counterpoint: Can Doppler Echocardiography Estimates of Pulmonary Artery Systolic Pressures Be Relied Upon to Accurately Make the Diagnosis of Pulmonary Hypertension? No

Rich JD, CHEST 2013;143:1533-36

Echo provide an acceptable compromise among accuracy, safety, simplicity and cost: **best screening tool for PAH!**

Echo for PH diagnosis: pitfalls

Table 1—*Partial List of Potential Pitfalls Associated With Inaccurate DE Estimates of sPAP*

DE Pitfall	DE Underestimation of sPAP	DE Overestimation of sPAP	DE Underestimation or Overestimation of sPAP
Doppler beam not parallel with the TR jet	***		
Presence of severe TR	***		
Pressure measurements not made at end expiration	***		

DE = Doppler echocardiography; sPAP = systolic pulmonary artery pressure; TR = tricuspid regurgitation.

Rich JD, CHEST 2013

Guidelines for the Echocardiographic Assessment of the Right Heart in Adults: A Report from the American Society of Echocardiography

Endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography

Lawrence G. Rudski, MD, FASE, Chair, Wyman W. Lai, MD, MPH, FASE, Jonathan Afilalo, MD, Msc, Lanqi Hua, RDCS, FASE, Mark D. Handschumacher, BSc, Krishnaswamy Chandrasekaran, MD, FASE, Scott D. Solomon, MD, Eric K. Louie, MD, and Nelson B. Schiller, MD, *Montreal, Quebec, Canada; New York, New York; Boston, Massachusetts; Phoenix, Arizona; London, United Kingdom; San Francisco, California*

J Am Soc Echocardiogr 2010;23:685-713

From the Holy Bible of Echocardiography…

Pulmonary Systolic Pressure/RVSP. TR velocity reliably permits estimation of RVSP with the addition of RA pressure, assuming no significant RVOT obstruction. It is recommended to use the RA pressure estimated from IVC and its collapsibility, rather than arbitrarily assigning a fixed RA pressure. In general, **TR velocity > 2.8 to 2.9 m/s, corresponding to SPAP of approximately 36 mm Hg, assuming an RA pressure of 3 to 5 mm Hg**, indicates elevated RV systolic and PA pressure. SPAP may increase, however, with age and in obesity.

Guidelines for the diagnosis and treatment of pulmonary hypertension

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 of Heart and Lung Transplantation (ISHLT)

European Heart Journal
doi:10.1093/eurheartj/ehp297

ESC/ERS GUIDELINES
2009

PH possible:

- PASP 37-50 mmHg (TVR 2.9-3.4 m/s)
- additional echo variables

PH likely:

- PASP >50 (TVR > 3.4 m/s)

Table 9 Arbitrary criteria for estimating the presence of PH based on tricuspid regurgitation peak velocity and Doppler-calculated PA systolic pressure at rest (assuming a normal right atrial pressure of 5 mmHg) and on additional echocardiographic variables suggestive of PH

	Class[a]	Level[b]
Echocardiographic diagnosis: PH unlikely		
Tricuspid regurgitation velocity ≤2.8 m/s, PA systolic pressure ≤36 mmHg, and no additional echocardiographic variables suggestive of PH	I	B
Echocardiographic diagnosis: PH possible		
Tricuspid regurgitation velocity ≤2.8 m/s, PA systolic pressure ≤36 mmHg, but presence of additional echocardiographic variables suggestive of PH	IIa	C
Tricuspid regurgitation velocity 2.9–3.4 m/s, PA systolic pressure 37–50 mmHg with/without additional echocardiographic variables suggestive of PH	IIa	C
Echocardiographic diagnosis: PH likely		
Tricuspid regurgitation velocity >3.4 m/s, PA systolic pressure >50 mmHg, with/without additional echocardiographic variables suggestive of PH	I	B
Exercise Doppler echocardiography is not recommended for screening of PH	III	C

Accuracy and precision of echocardiography versus right heart catheterization for the assessment of pulmonary hypertension

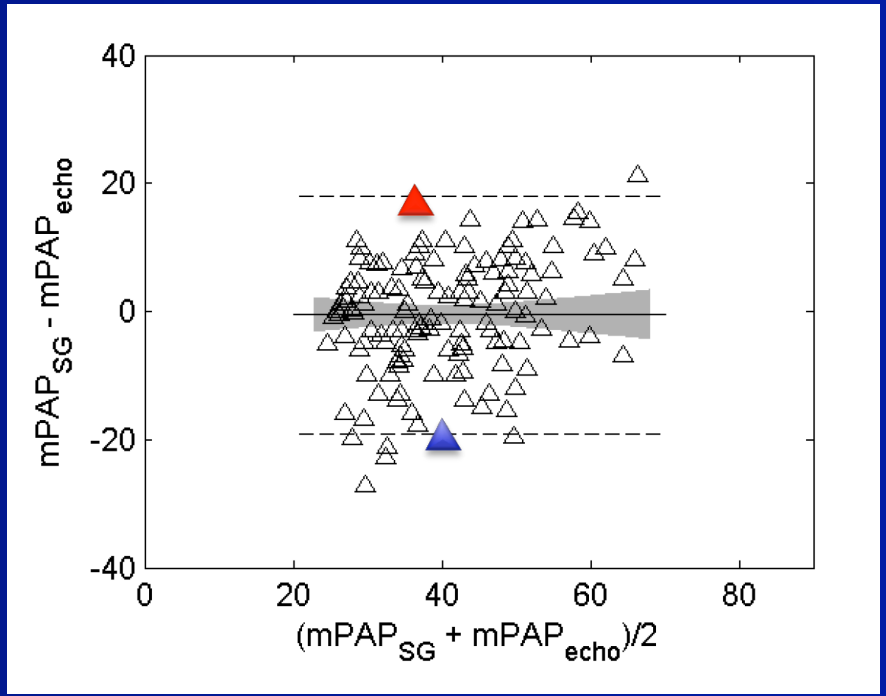
Michele D'Alto [a,*,1], Emanuele Romeo [a], Paola Argiento [a], Antonello D'Andrea [a], Rebecca Vanderpool [b], Anna Correra [a], Eduardo Bossone [c], Berardo Sarubbi [a], Raffaele Calabrò [a], Maria Giovanna Russo [a], Robert Naeije [b]

D'Alto M et al, Int J Cardiol 2013:168(4):4058-62

- Prospective study
- 161 patients referred for a suspicion of PH
- 152/161 patients with sufficient quality echo imaging
- Echo within 1 hour of an indicated RHC

Bias:
0.5±9 mmHg

Limits of agreement:
-19 to +18 mmHg



Good accuracy (low bias)
Insufficient precision (large limits of agreement)

The concept of accuracy and precision

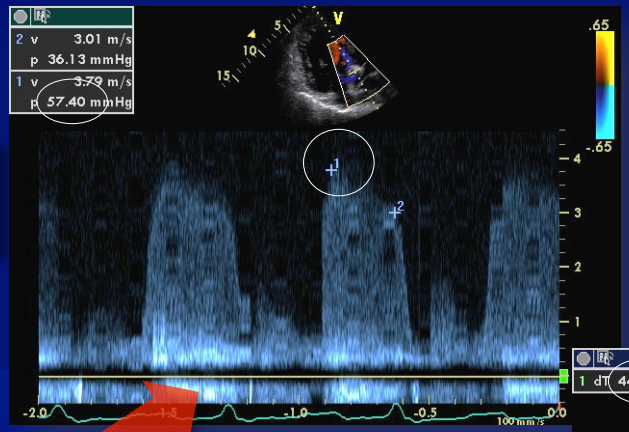
- Echo is **adequate for population studies** (good accuracy = low bias)
- Echo is **inadequate for individual diagnosis** (insufficient precision = large limits of agreement)

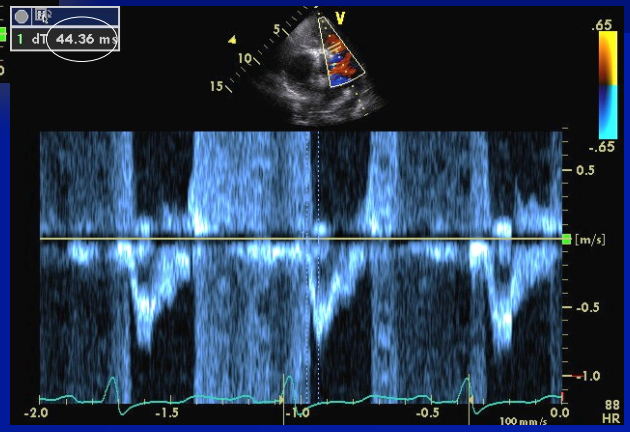
RHC

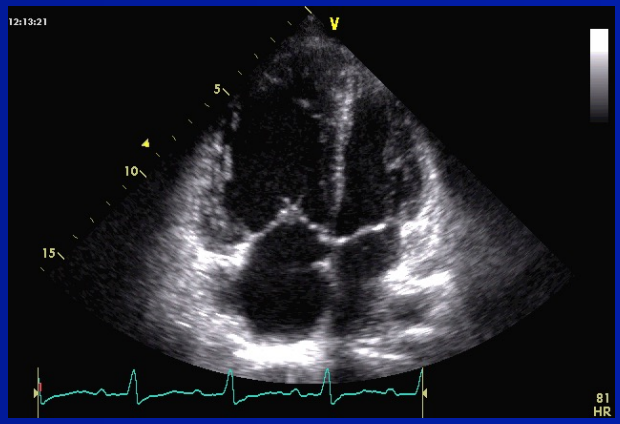
D'Alto M et al, Int J Cardiol 2013:168(4):4058-62

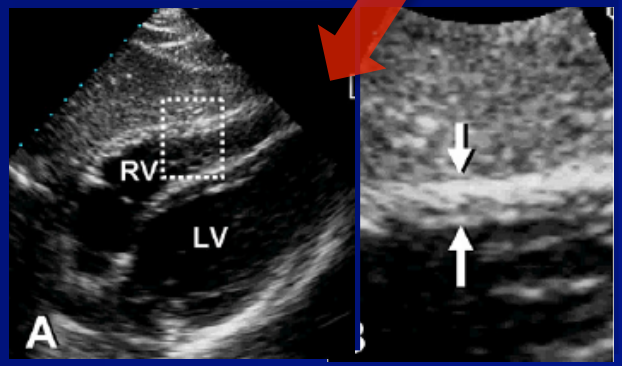
Indirect PAH signs

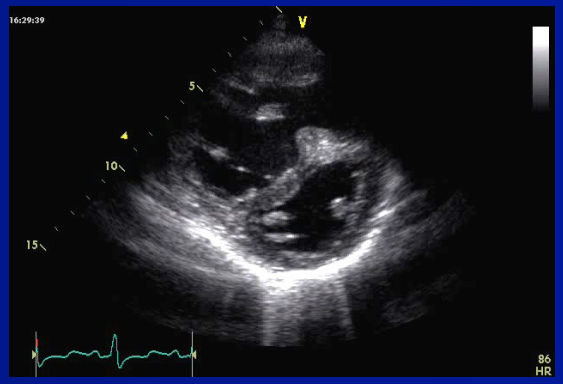
- Increased velocity PV reg (mPAP)
- Short acc. time in RVOT (mPAP)
- Right heart dilation
- Flat IV septum (LV eccentricity index)
- Increased RV wall thickness











Pre-test probability of precapillary PH

Guidelines for the diagnosis and treatment of pulmonary hypertension

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 of Heart and Lung Transplantation (ISHLT)

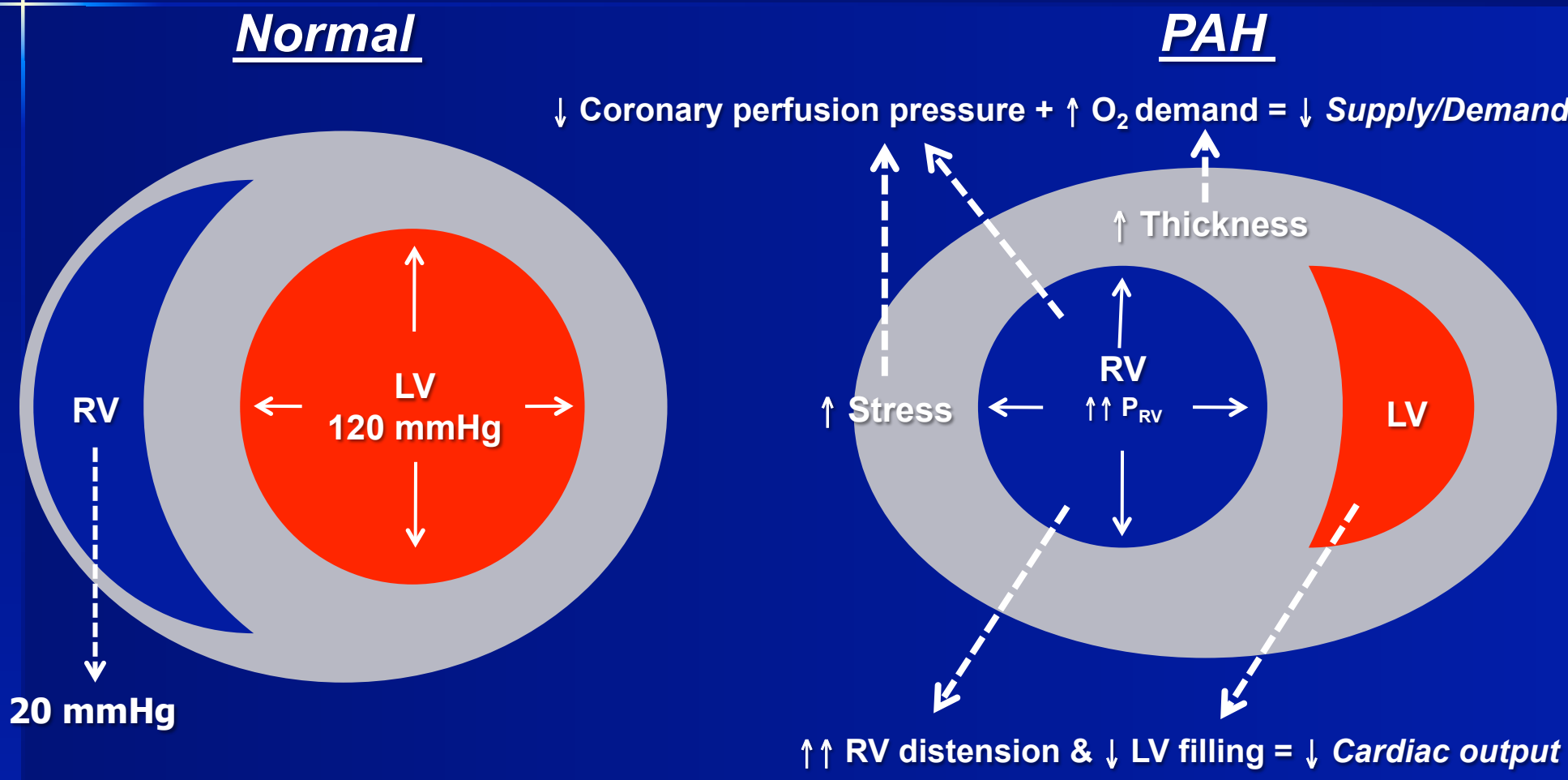
European Heart Journal
doi:10.1093/eurheartj/ehp297

ESC/ERS GUIDELINES
2009

Table 30 Factors favouring diagnosis of left ventricular diastolic dysfunction in the presence of pulmonary hypertension as assessed by Doppler echocardiography

Clinical features
Age >65
Elevated systolic blood pressure
Elevated pulse pressure
Obesity, metabolic syndrome
Hypertension
Coronary artery disease
Diabetes mellitus
Atrial fibrillation
Echocardiography
Left atrial enlargement
Concentric remodelling of the LV (relative wall thickness >0.45)
LV hypertrophy
Presence of echocardiographic indicators of elevated LV filling pressure[64,226]
Interim evaluation (after echocardiography)
Symptomatic response to diuretics
Exaggerated increase in systolic blood pressure with exercise
Re-evaluation of chest radiograph consistent with heart failure[226]

RV anatomy: normal versus PAH



Chin KM, *et al*. *Coron Artery Dis* 2005; 16:13-8

Echocardiographic Prediction of Pre- versus Postcapillary Pulmonary Hypertension

Michele D'Alto, MD, PhD, Emanuele Romeo, MD, PhD, Paola Argiento, MD, PhD, Adriana Pavelescu, MD, PhD, Christian Mélot, MD, PhD, Antonello D'Andrea, MD, PhD, Anna Correra, MD, Eduardo Bossone, MD, PhD, Raffaele Calabrò, MD, Maria G. Russo, MD, and Robert Naeije, MD, PhD, *Naples, and Salerno, Italy; Brussels, Belgium*

J Am Soc Echo, 2015

- 152 patients
- echo and RHC **within 1 hour**

Table 4 Selected echocardiographic variables of potential predictive value entered as variables in the univariate and multivariate logistic regression

		Univariate logistic regression		Multiple logistic regression			
Variable	Cutoff value	*P*	SE	β coefficient	SE	*P*	Echocardiographic score
Right > left heart chamber	1	.0018	0.34	0.30	0.48	.5158	3
EI	≥1.2	.0039	0.33	0.38	0.45	.4052	4
IVC	>20 mm, no collapse	.0076	0.39	0.96	0.48	.0464	10
E/E′ ratio	≤10	.00001	0.38	1.58	0.42	.0002	16
RV forming apex	Present	.0144	0.35	0.14	0.58	.8085	1
PA notch	Present	.2085	0.34				
Pericardial effusion	Present	.7805	0.52				
Moderate to severe left valve disease	Present	.1784	0.44				
Total score							34

Echocardiographic Prediction of Pre- versus Postcapillary Pulmonary Hypertension

Michele D'Alto, MD, PhD, Emanuele Romeo, MD, PhD, Paola Argiento, MD, PhD, Adriana Pavelescu, MD, PhD, Christian Mélot, MD, PhD, Antonello D'Andrea, MD, PhD, Anna Correra, MD, Eduardo Bossone, MD, PhD, Raffaele Calabrò, MD, Maria G. Russo, MD, and Robert Naeije, MD, PhD, *Naples, and Salerno, Italy; Brussels, Belgium*

J Am Soc Echo, 2015

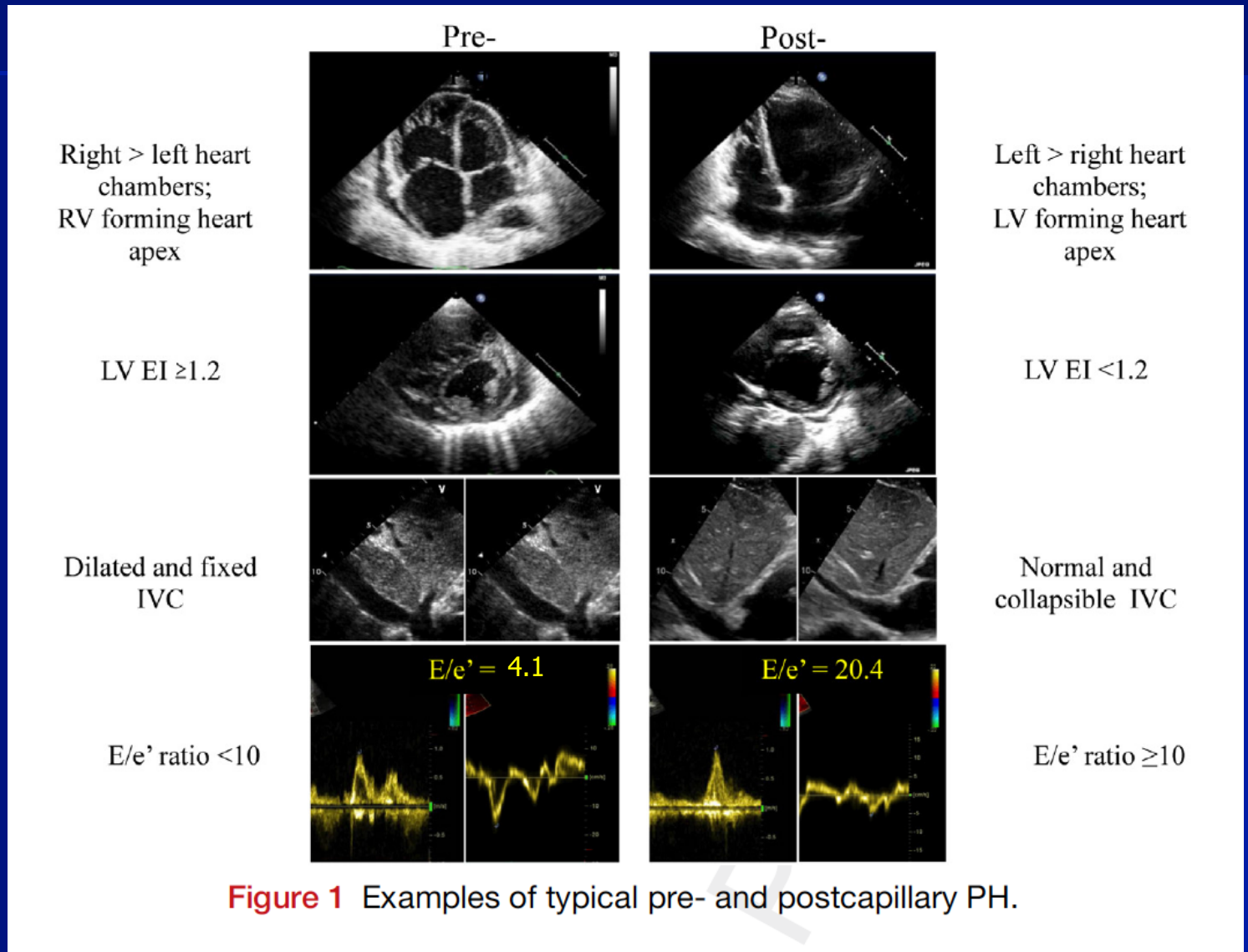
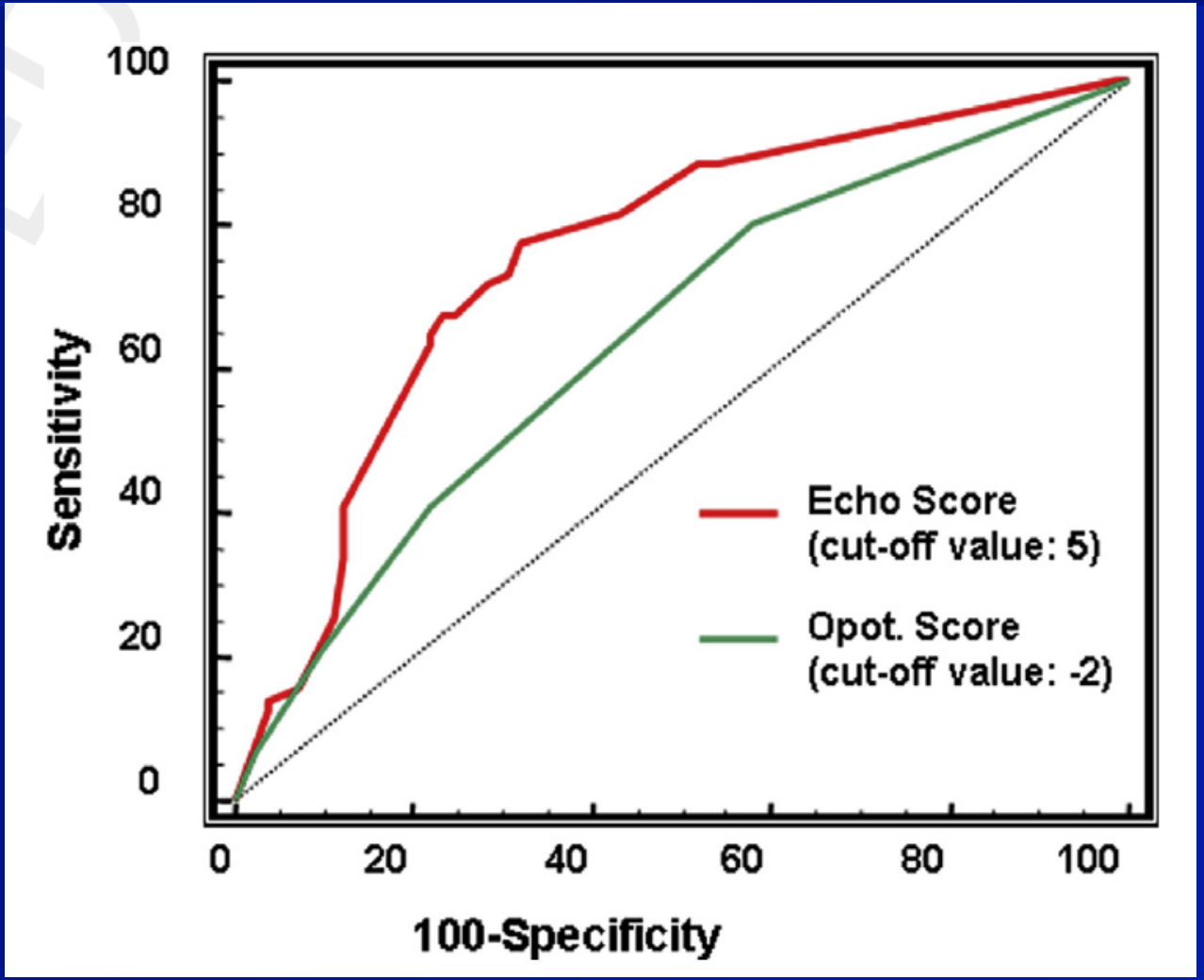


Figure 1 Examples of typical pre- and postcapillary PH.

Echocardiographic Prediction of Pre- versus Postcapillary Pulmonary Hypertension

Michele D'Alto, MD, PhD, Emanuele Romeo, MD, PhD, Paola Argiento, MD, PhD, Adriana Pavelescu, MD, PhD, Christian Mélot, MD, PhD, Antonello D'Andrea, MD, PhD, Anna Correra, MD, Eduardo Bossone, MD, PhD, Raffaele Calabrò, MD, Maria G. Russo, MD, and Robert Naeije, MD, PhD, *Naples, and Salerno, Italy; Brussels, Belgium*

J Am Soc Echo, 2015



Multimodality imaging in pulmonary hypertension

Canadian Journal of Cardiology 31 (2015) 440–459

Review

Multimodality Imaging in Pulmonary Hypertension

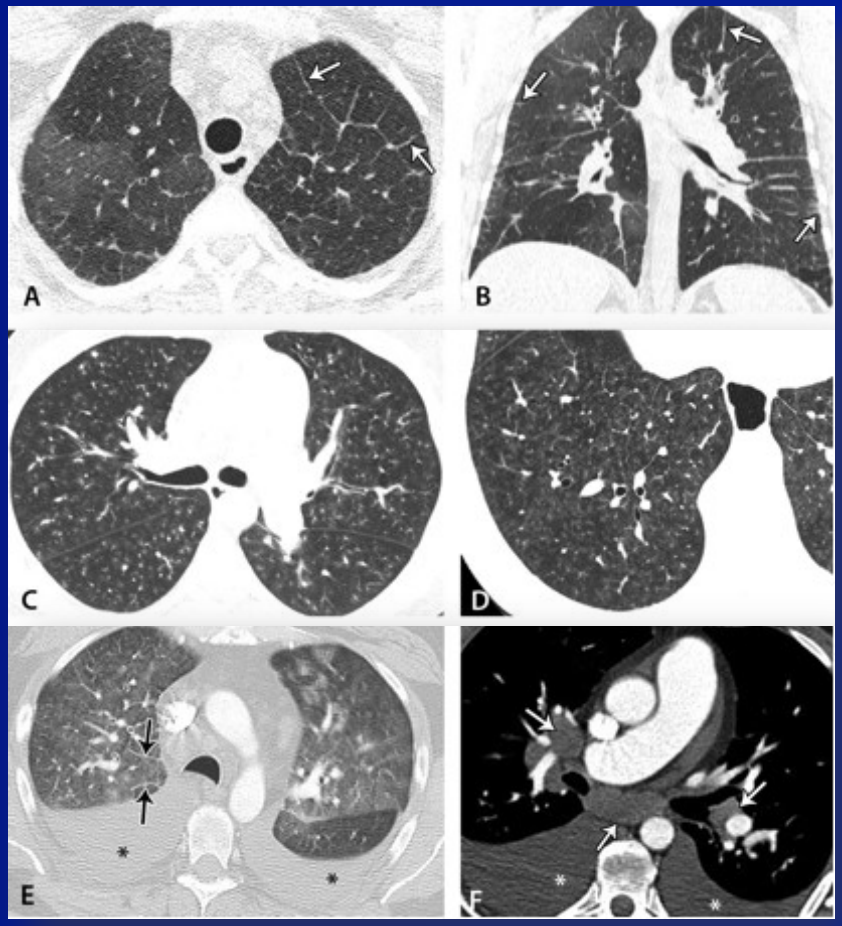
Eduardo Bossone, MD, PhD,[a,b] Santo Dellegrottaglie, MD, PhD,[c,d] Smita Patel, MBBS, MRCP,[e] Ekkehard Grunig, MD,[f] Antonello D'Andrea, MD, PhD,[g] Francesco Ferrara, MD, PhD,[b,h] Paola Gargiulo, MD,[i] Michele D'Alto, MD,[g] Andrea Soricelli, MD,[i,j] Antonio Cittadini, MD, PhD,[h] Javier Sanz, MD,[d] Pasquale Perrone-Filardi, MD, PhD,[k] and Melvyn Rubenfire, MD[l]

Canadian J Cardiol, 2015;31:440-59

HR-CT: Pulmonary veno-occlusive disease (PVOD) and pulmonary capillary hemangiomatosis (PCH)

PVOD

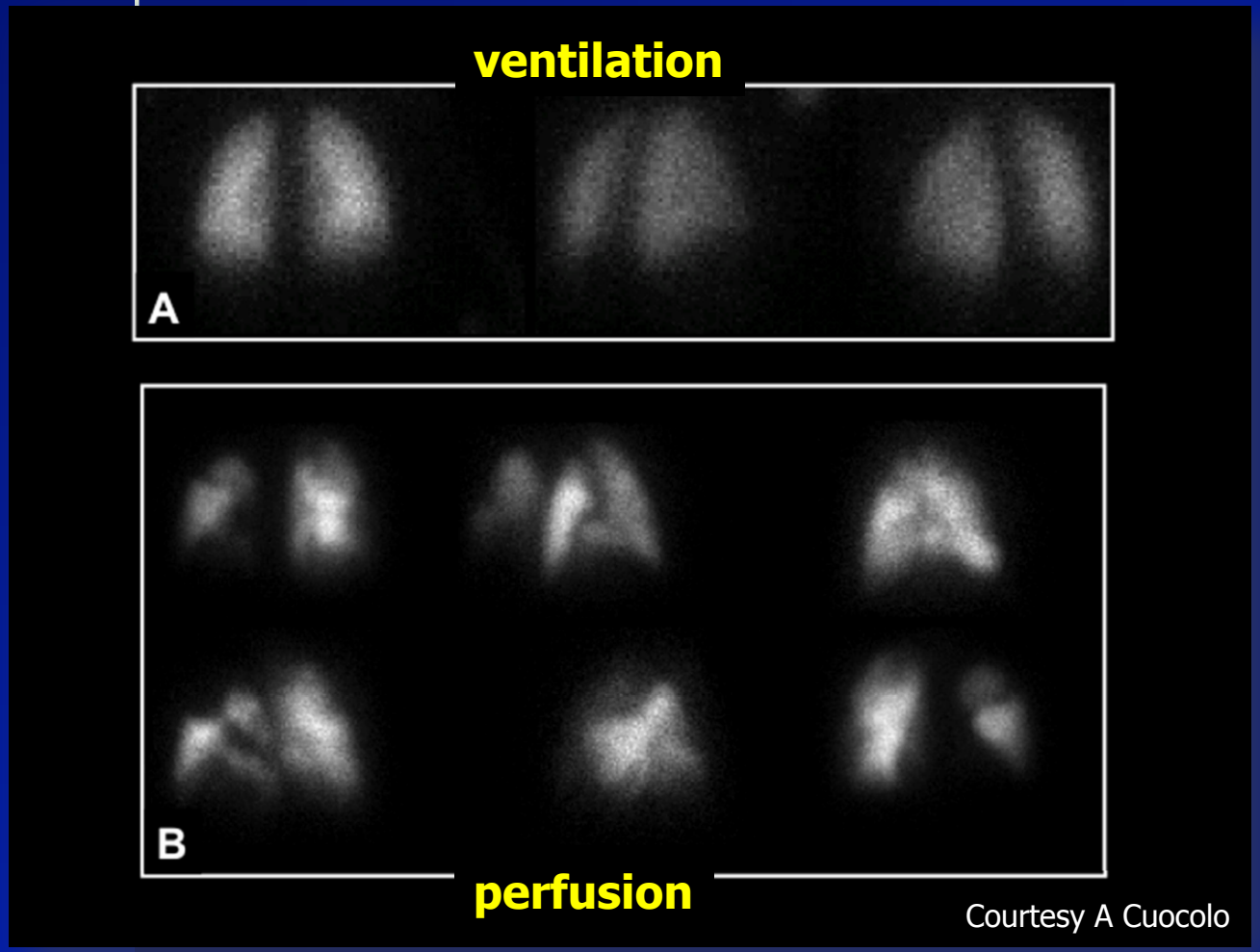
PCH



Suspected PCH/PVOD on the basis of clinical worsening while on iv prostacyclin therapy with subsequent intractable pulmonary edema

Bossone et al, Canadian J Cardiol 2015

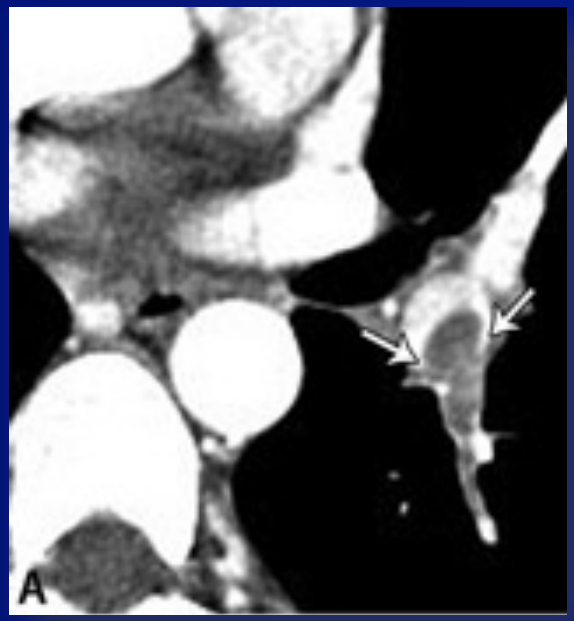
V/Q scan (scintigraphy) for ruling out CTEPH



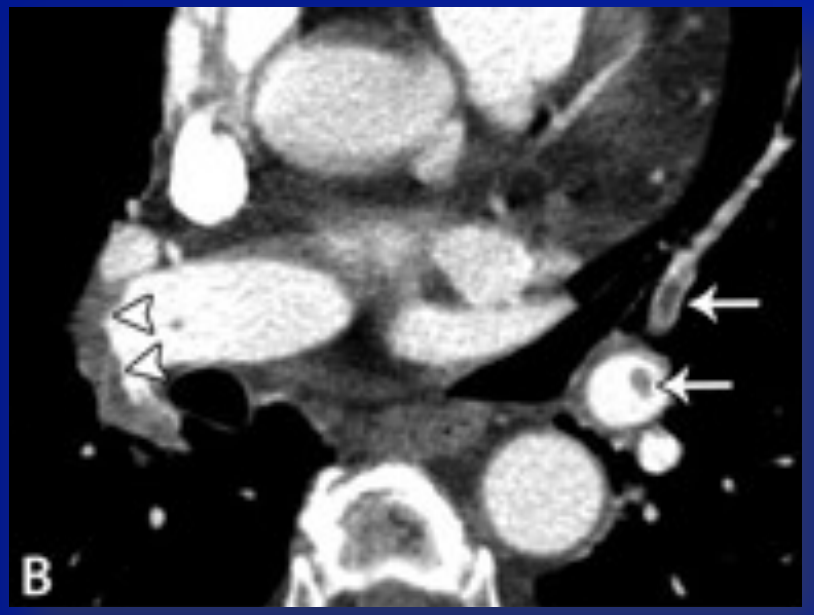
Mismatch = high probability of CTEPH

Bossone et al, Canadian J Cardiol 2015

Pulmonary CT angiography for chronic thromboembolic pulmonary hypertension (CTEPH)



Acute pulmonary embolism: filling defect forming acute angles (arrow) with the vessel wall, compatible with acute pulmonary embolism (PE).



Acute and chronic embolism: eccentric wall-adherent peripheral thrombus (arrow) with irregular contour of the intimal surface (chronic PE). Central filling defect in the left lower lobe pulmonary artery and lingula (arrowheads) due to acute PE.

Bossone et al, Canadian J Cardiol 2015

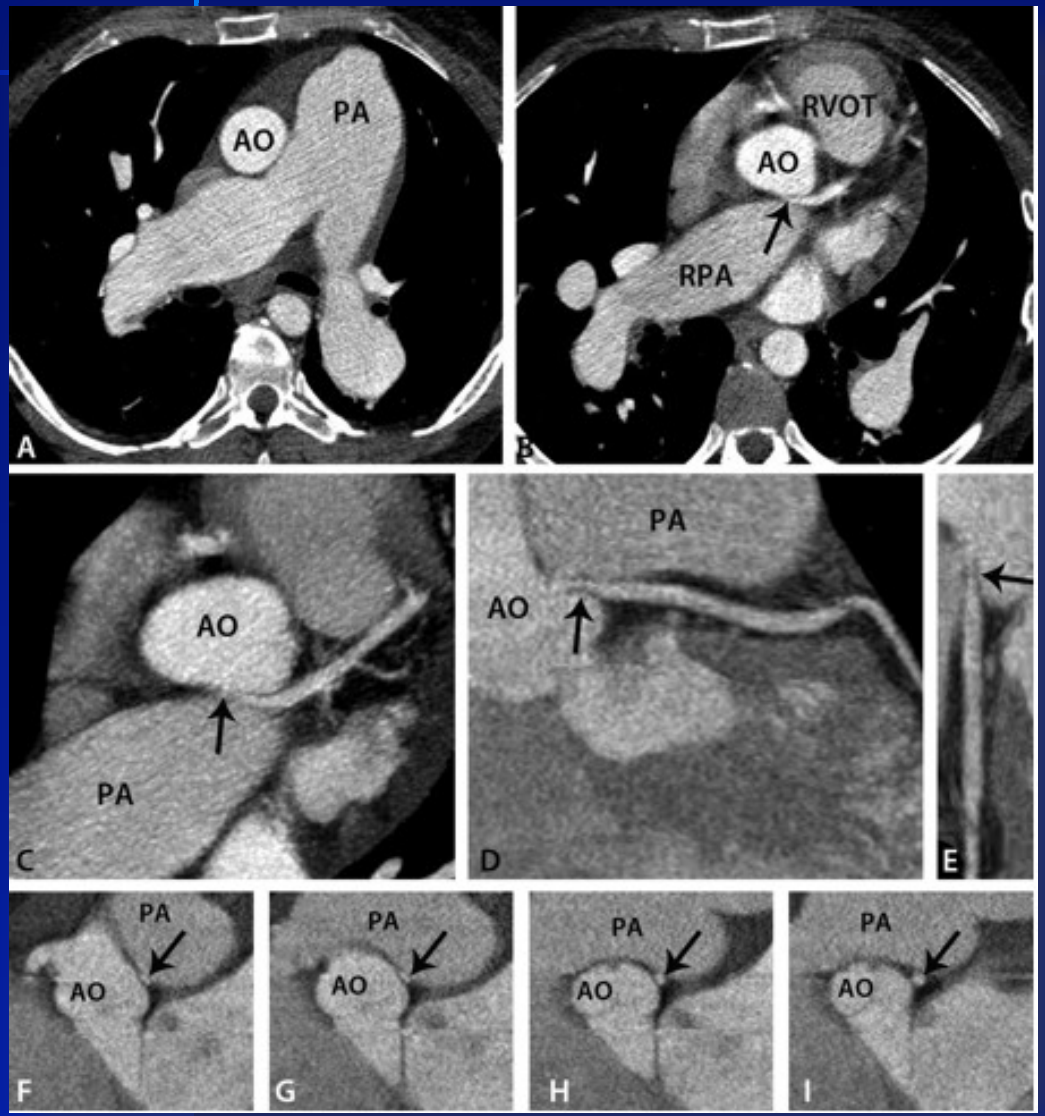
Pulmonary CT angiography before and after pulmonary endoarterectomy (PEA)

before

after

Bossone et al, Canadian J Cardiol 2015

Myocardial ischemia in PAH patients?

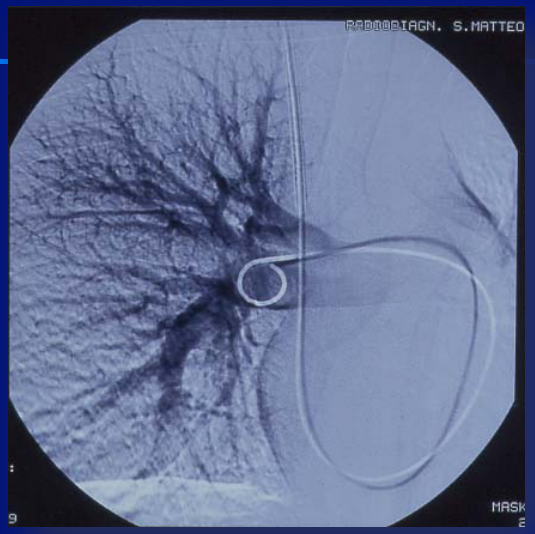


A 38-year-old female patient with pulmonary arterial hypertension and an ASD with **shortness of breath and chest pain**.

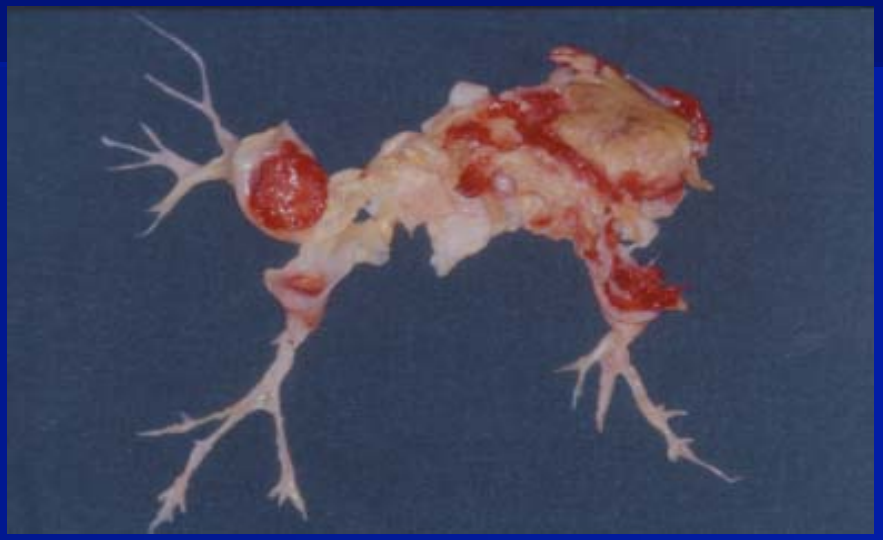
Left main compression due to enlargement of the proximal pulmonary artery. Symptoms resolved with a **left main bare metal stent**.

Bossone et al, Canadian J Cardiol 2015

Pulmonary angiogram

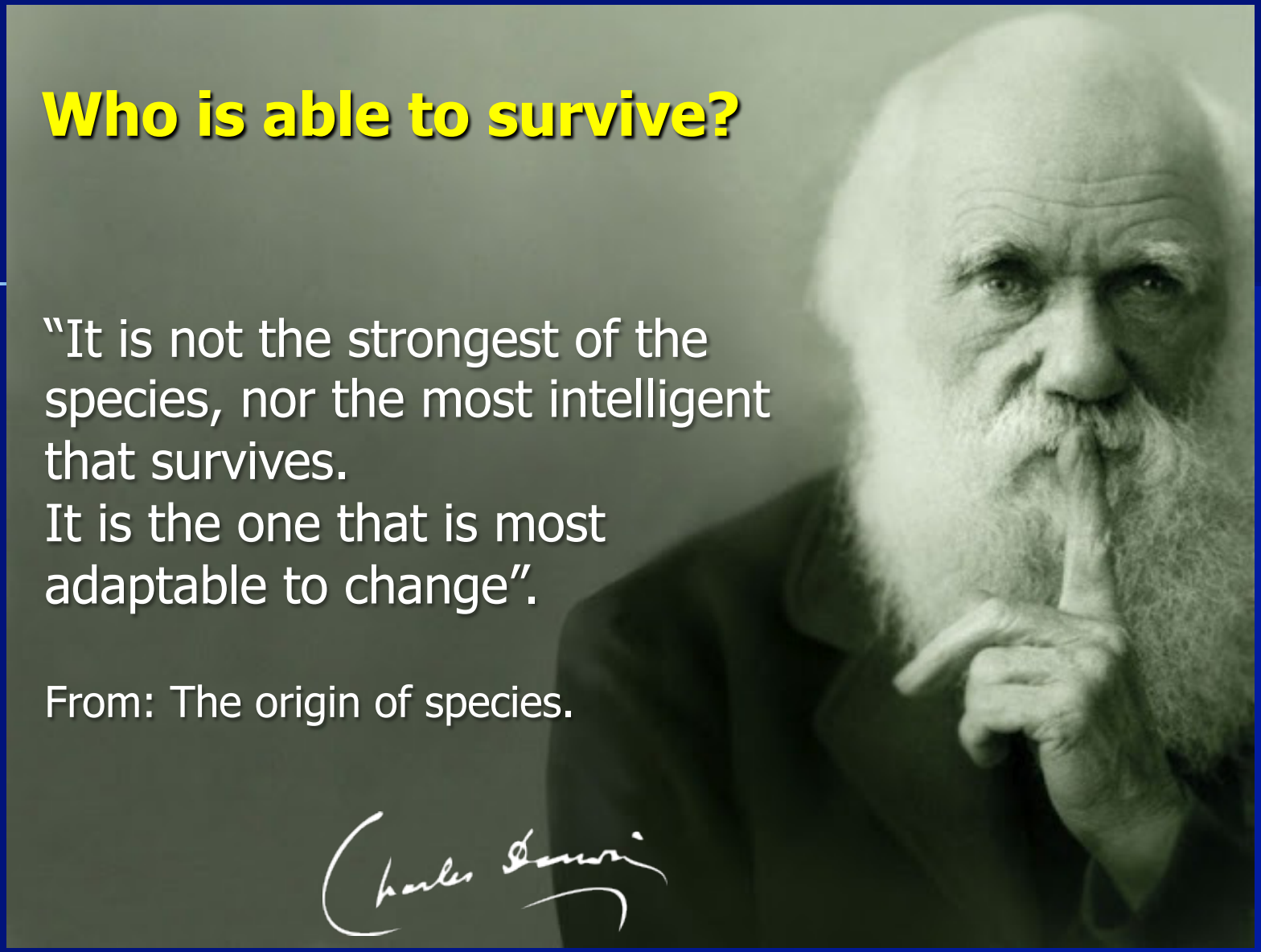


Typical surgical specimens



Courtesy A D'Armini

Who is able to survive?



"It is not the strongest of the species, nor the most intelligent that survives.
It is the one that is most adaptable to change".

From: The origin of species.

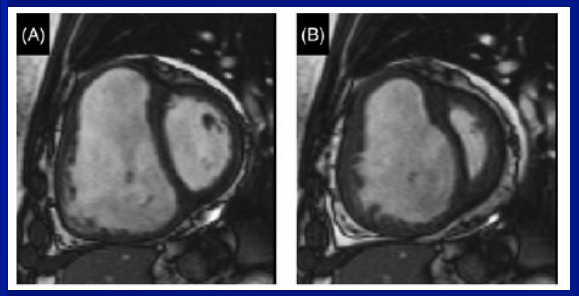
"La specie che sopravvive non è quella più forte, né quella più intelligente.
Ma quella che si adatta meglio al cambiamento".

Right ventricular remodeling in Eisenmenger syndrome

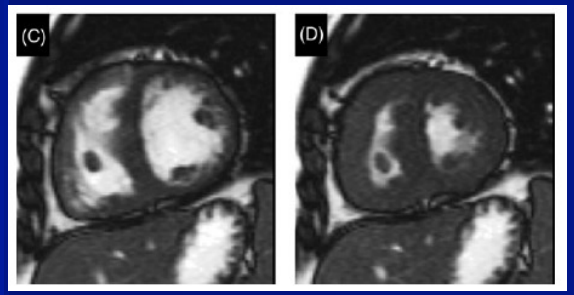
PH and GUCH Unit Monaldi

diastole systole

IPAH

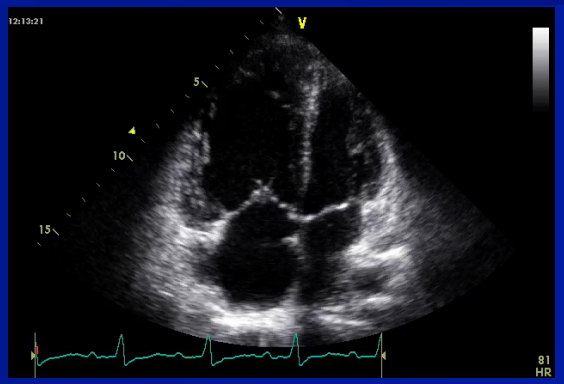


CHD-PAH

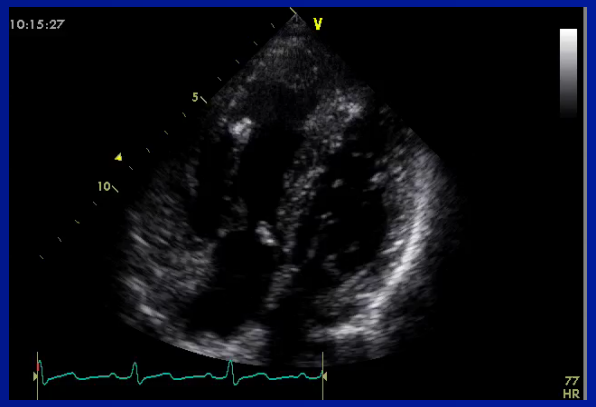


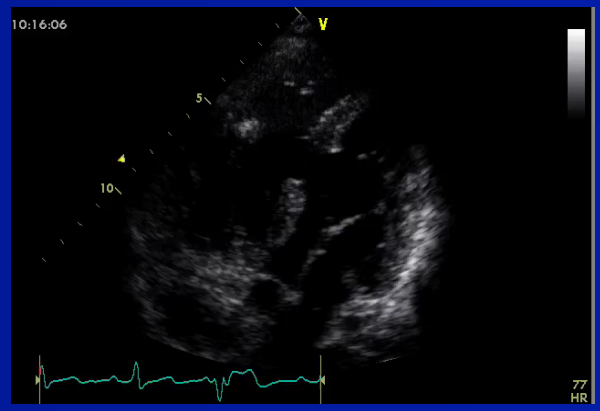
Diller, Eur Heart J 2007;H54–H60

IPAH



CHD-PAH





Optimal RV adaptation

Parameters reflecting RV function

Echocardiography

- Right atrial area[1]
- Right ventricular area[1]
- TAPSE[1,2]
- Tei index[3]
- Right ventricular fractional area change[2]
- Degree of tricuspid regurgitation[2]
- Pericardial effusion[4]
- Inferior vena cava collapsibility[2]
- Superior vena cava flow velocity pattern[2]
- Left ventricular eccentricity index[2]
- Right ventricular filling pressure[5]

MRI

- Right ventricular ejection fraction[6]
- Right ventricular stroke volume
- Right ventricular mass
- Right ventricular volume[1]

RHC

- Right arterial pressure[7]
- Cardiac index[8]

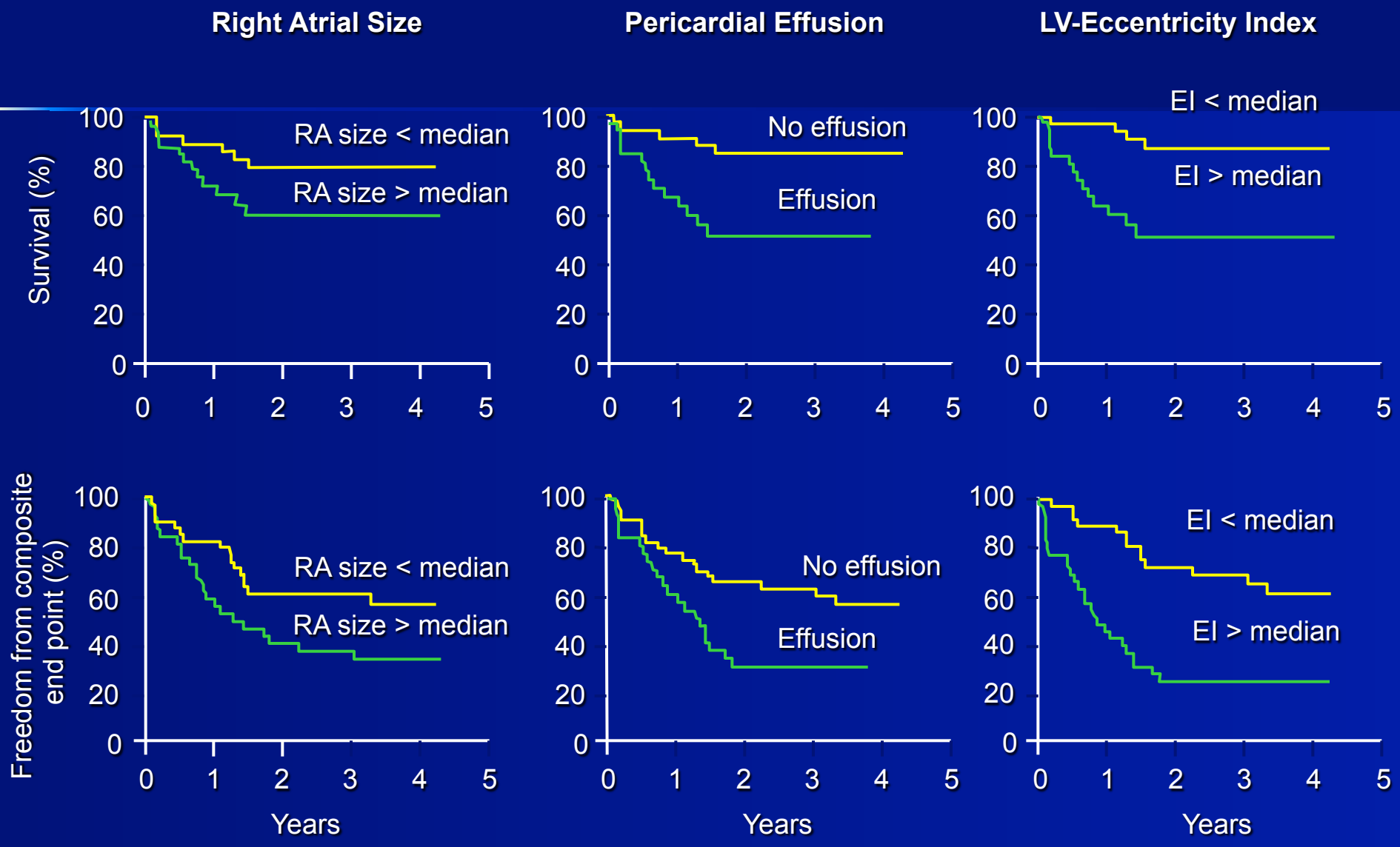
Biomarkers

- N-terminal pro-brain natriuretic peptide[9]
- Troponin T[10]

TAPSE: tricuspid annular plane systolic excursion

1. Grünig E, *et al. DMW* 2010. 2. Ghio S, *et al. Int J Cardiol* 2010.
3. Tei C, *et al. J Am Soc Echocardiogr* 1996. 4. Raymond RJ, *et al. JACC* 2002.
5. Utsunomiya H, *et al. J Am Soc Echocardiogr* 2009. 6. van de Veerdonk M, *et al. JACC* 2011.
7. McLaughlin VV, *et al. Circulation* 2002. 8. D'Alonzo GE, *et al. Ann Intern Med* 1991.
9. Nagaya N, *et al. JACC* 1998. 10. Torbicki A, *et al. Circulation* 2003.

Echocardiographic predictors in PAH



Raymond et al. J Am Coll Cardiol. 2002

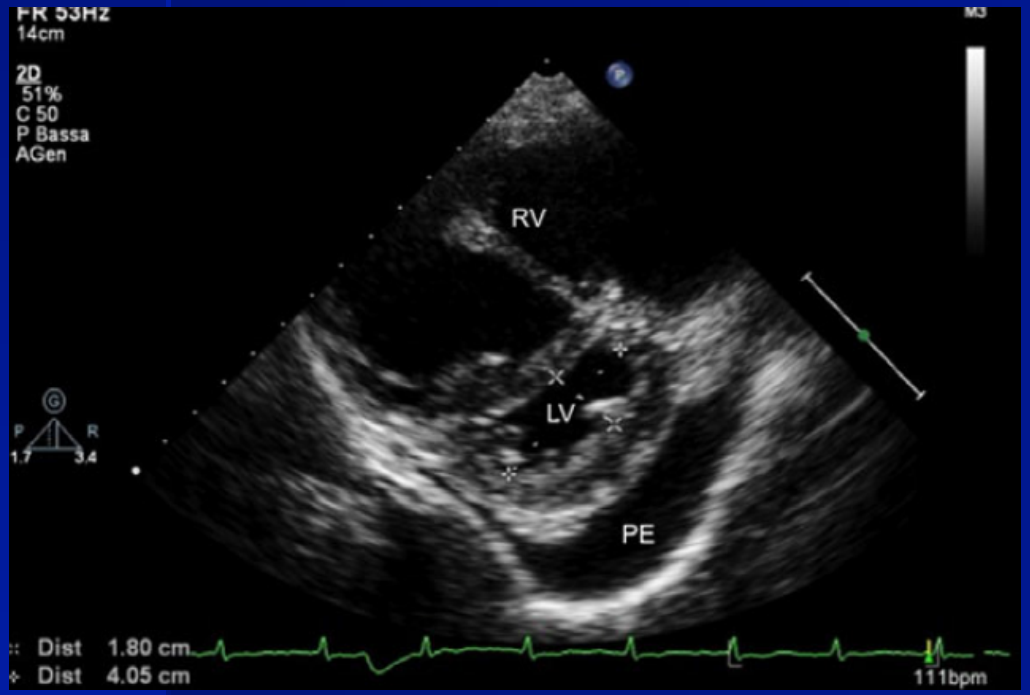
Pulmonary Arterial Hypertension: The Key Role of Echocardiography

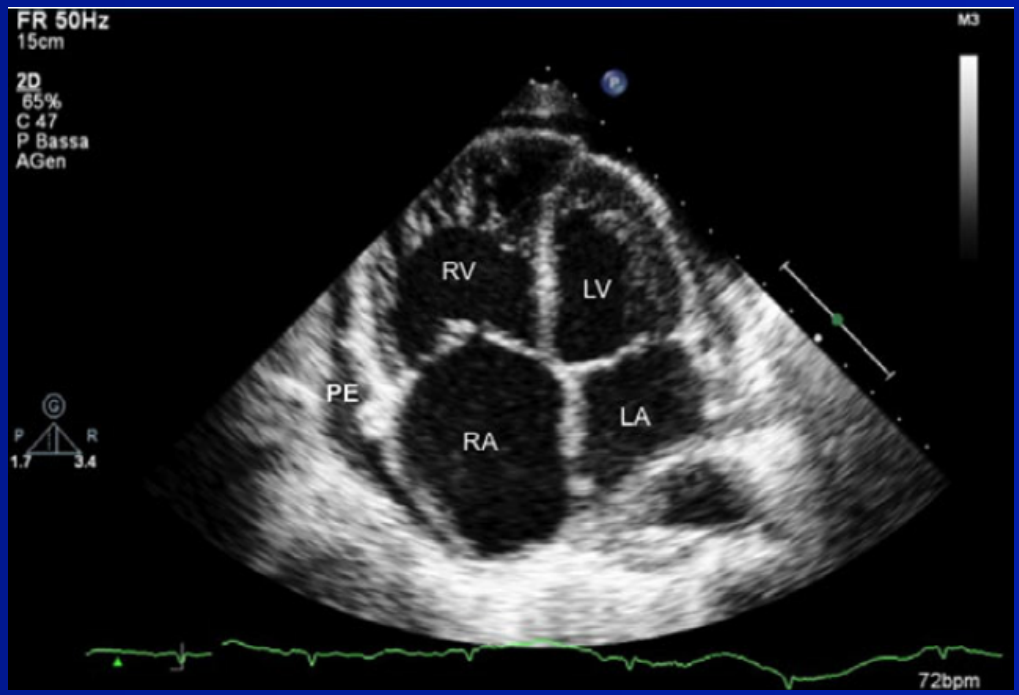
Michele D'Alto, Ph.D.,* Emanuele Romeo, Ph.D.,* Paola Argiento, Ph.D.,* Giovanni Di Salvo, Ph.D.,* Roberto Badagliacca, M.D.,† Anna P. Cirillo, M.D.,‡ Harald Kaemmerer, M.D.,§ Eduardo Bossone, M.D.,¶ and Robert Naeije, M.D.**

Echocardiography 2015

Poor prognosis in PAH if…

Pericardial effusion



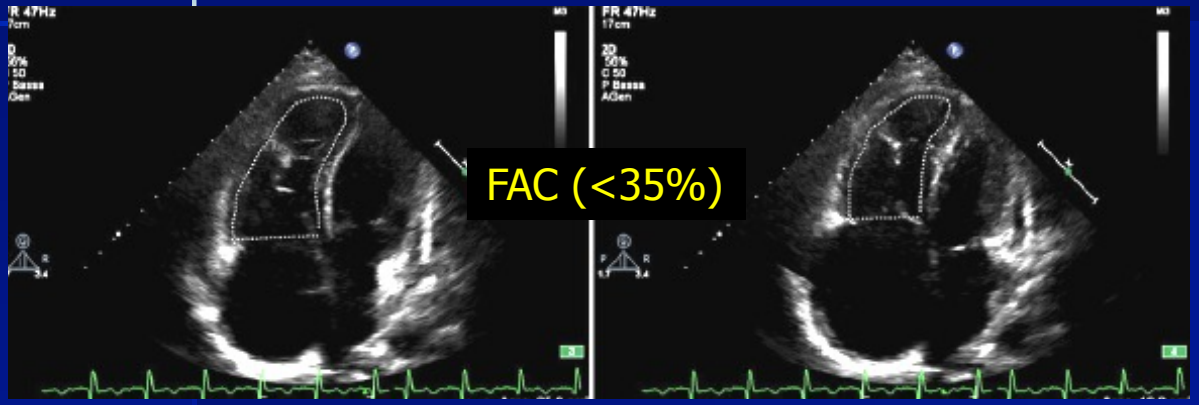


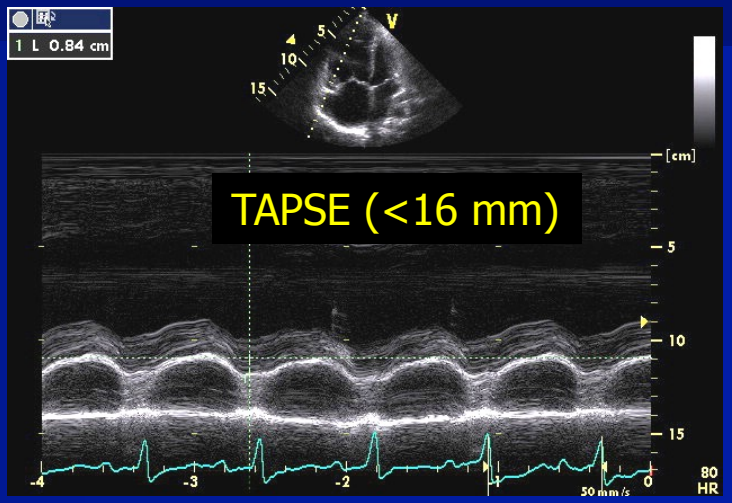
Pulmonary Arterial Hypertension: The Key Role of Echocardiography

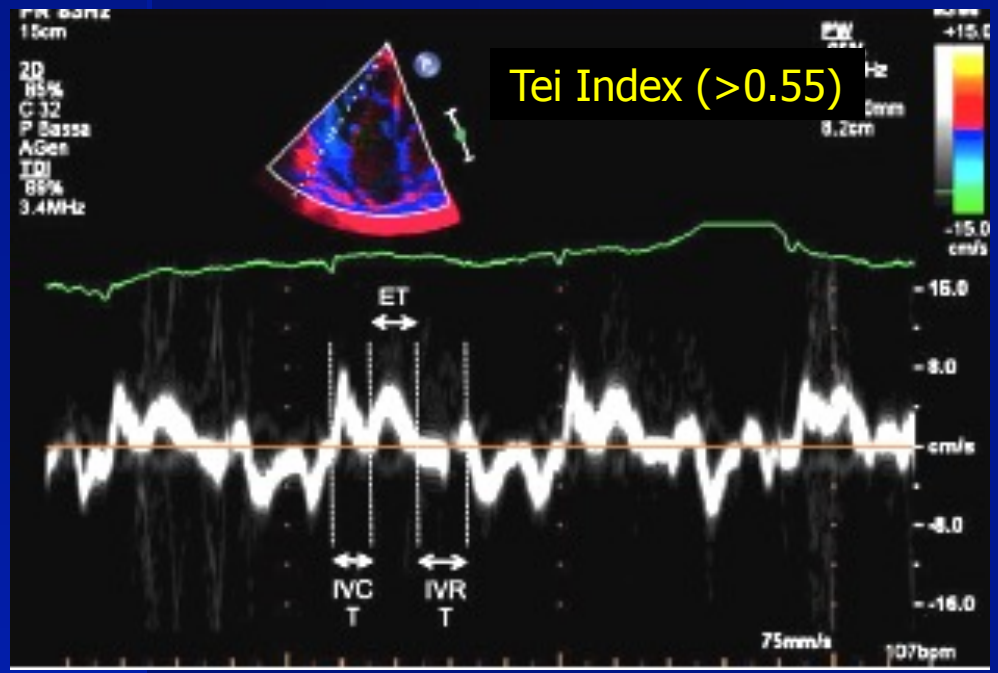
Michele D'Alto, Ph.D.,* Emanuele Romeo, Ph.D.,* Paola Argiento, Ph.D.,* Giovanni Di Salvo, Ph.D.,* Roberto Badagliacca, M.D.,† Anna P. Cirillo, M.D.,‡ Harald Kaemmerer, M.D.,§ Eduardo Bossone, M.D.,¶ and Robert Naeije, M.D.**

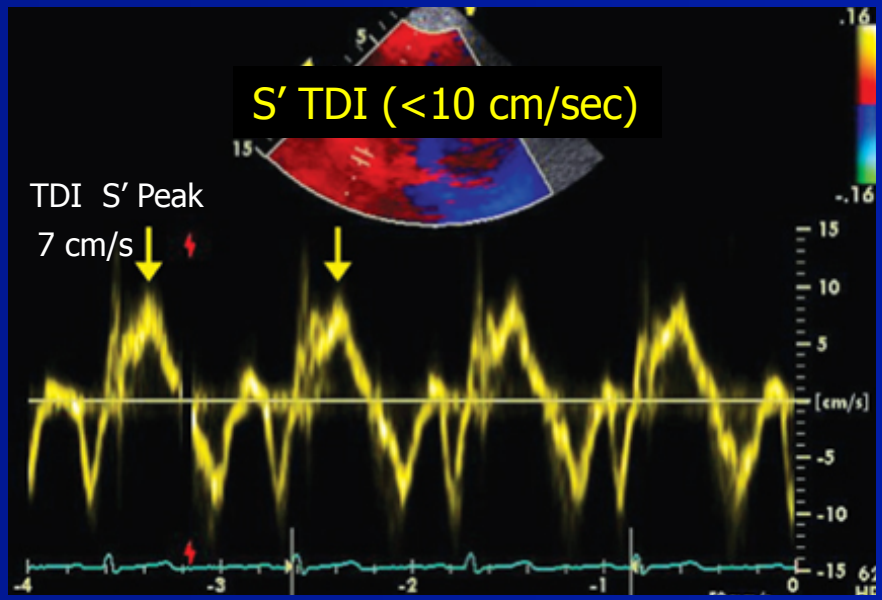
Echocardiography 2015

Poor prognosis in PAH if…

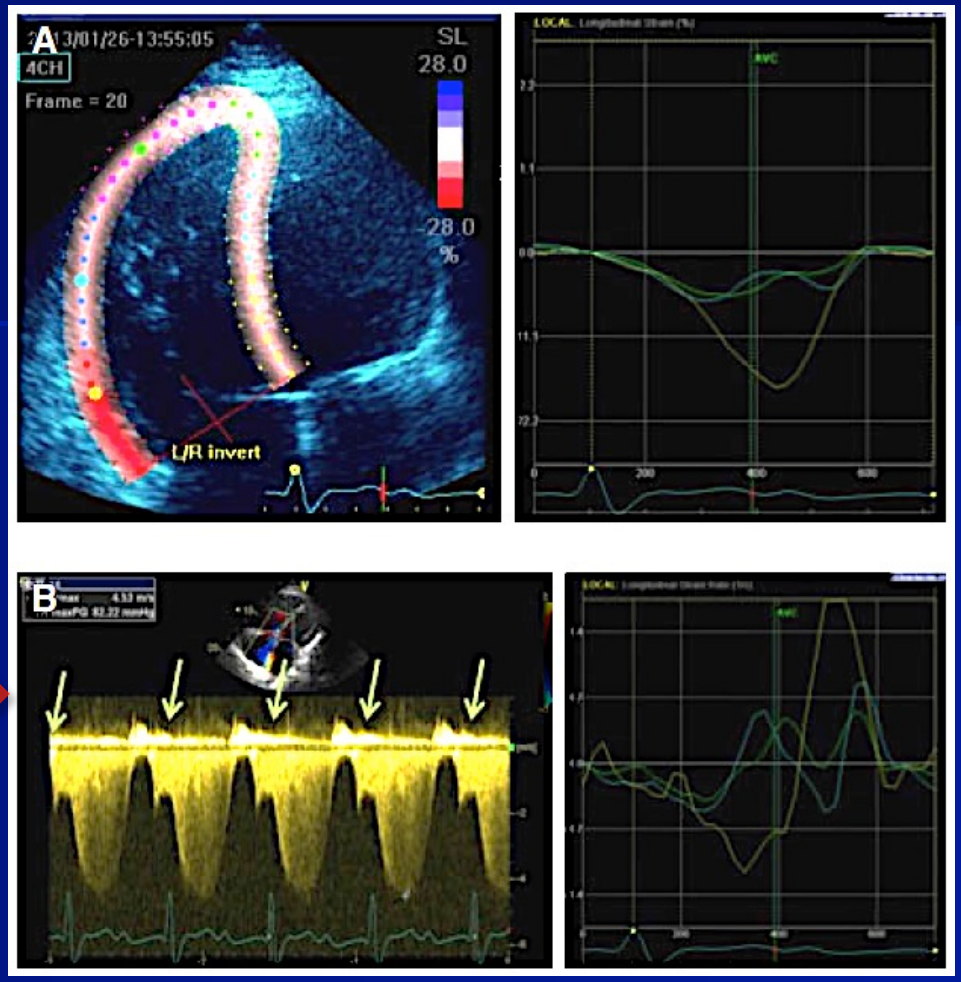








Evidence of presystolic tricuspid regurgitation



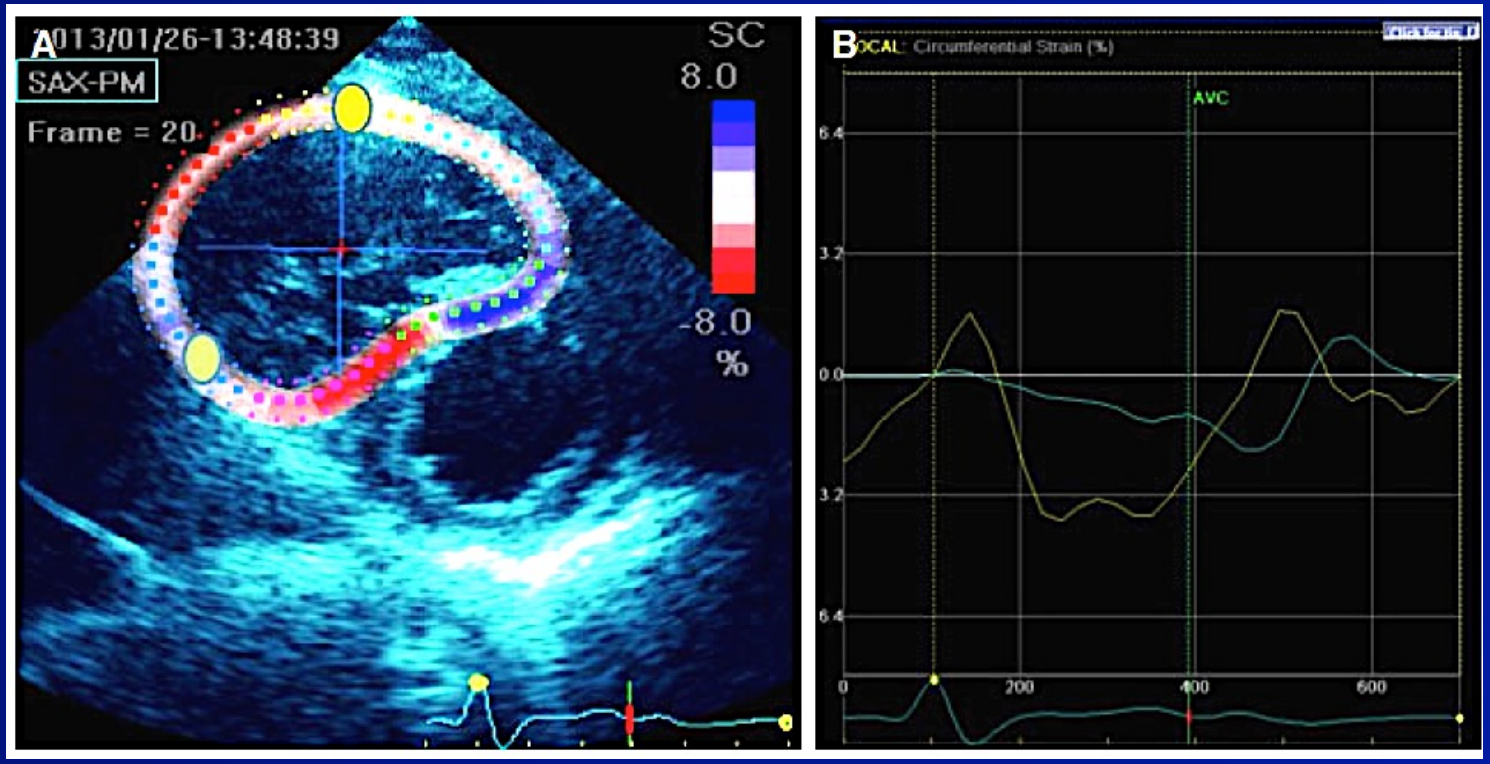
Severely **reduced myocardial deformation properties** (A. strain; B. strain rate). Myocardial deformation was more severely compromised at mid and apical right ventricular free wall, while basal segment was less involved, suggesting a **different regional response to pressure overload**.

D'Alto M et al, Echocardiography 2015

Speckle tracking, an angle independent technique, allows the assessment of circumferential myocardial deformation.

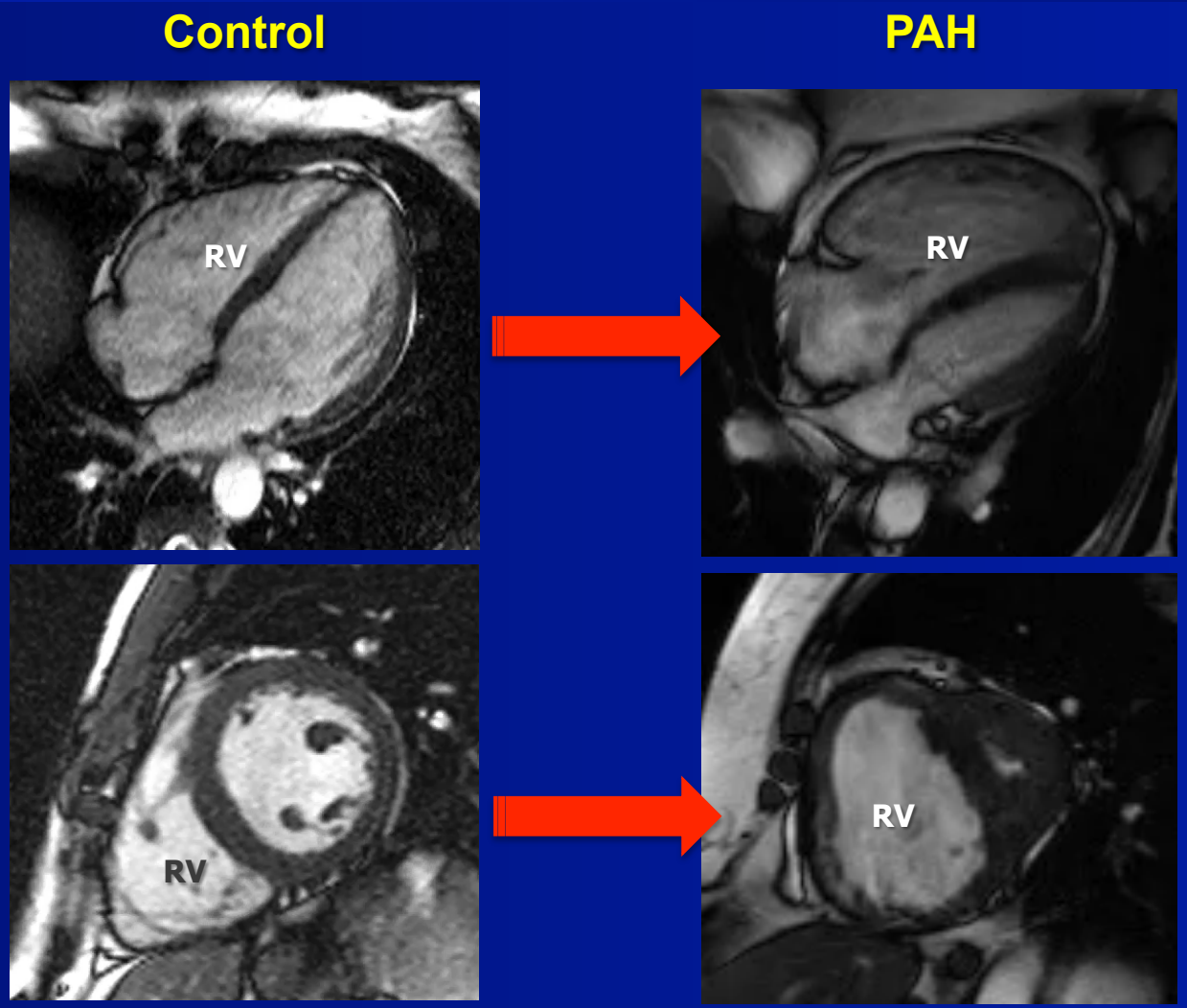
Asynchronous peak deformation

Severe reduction of circumferential strain



D'Alto M et al, Echocardiography 2015

Cardiac MRI: Four-chamber and short-axis in a healthy subject and in a PAH patient



Take-at-home message - 1

- **Multimodality imaging** is a must in PAH diagnosis and management.
- Echo plays a key-role in **screening**, **differential diagnosis** and **follow-up**.
- Echo does not provide "magic numbers": **multi-parametric evaluation** (**echo as part of clinic evaluation!**).
- Echo does **not allow a certain diagnosis** in individual patients (insufficient precision = large limits of agreement): **gold standard** for PAH diagnosis remains **right heart catheterization**.

Take-at-home message - 2

- **HR-CT scan** provides important information on **lung diseases**.
- **V/Q scan** is indicated for **ruling-out CTEPH**.
- **Pulmonary angio-CT scan and pulmonary angiography** are mandatory for **CTEPH evaluation**.
- **Cardiac MRI** is important for studying **RV remodeling and function** and identifying **pulmonary vessels anomalies**.

Multitasking PAH-imaging expert