Echo 2018 27-28 October SINGAPORE

Diagnosing Pulmonary Hypertension: ESC/ERS 2015

New Echo Guidelines: The Low-Down

DR. TAN JU LE
Pulmonary Hypertension Services
National Heart Centre
SINGAPORE

National Heart Centre Singapore
SingHealth

- No financial disclosures related to this talk

Outline

- Diagnosing Pulmonary Hypertension – ESC Guidelines
- The role of Echo in the diagnosis of Pulmonary Hypertension
- Summary

CLASSIFICATION of PULMONARY HYPERTENSION

- Previously classified into 2 types (old classification)
  - PRIMARY PULMONARY HYPERTENSION (PPHT)
  - SECONDARY PULMONARY HYPERTENSION
  - Depends on presence or absence of identifiable causes
  - PPHT diagnosis of exclusion
- In 1998 – 2nd World Symposium on PAH , 'Evian Classification' a new clinical based classification proposed to individualize different categories of PHT
- In 2003 - in Venice, 3rd World Symposium on PAH, modifications to Evian Classification
- 2008 – Dana Point Update (4th World symposium on PAH), further modifications
- 2013 – Latest update in NICE (5th World Symposium on PAH)
- **2015 - ESC Guidelines for the Diagnosis and Treatment of PHT**
- 2018 - 6th World Symposium in NICE on PHT, results published Oct/Nov 18

The 2015 ESC Guidelines for the diagnosis of Pulmonary Hypertension is a

- Haemodynamic Definition

2015 ESC/ERS Guidelines for the diagnosis and treatment of Pulmonary Hypertension

Table 3 Haemodynamic definitions of pulmonary hypertension*

NONE of these are Echo parameters
- **Mean PAP, PAWP, DPG and PVR are all measurements obtain from cath**

Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU‡	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU‡	

Table 5 Important pathophysiological and clinical definitions

1. Pulmonary hypertension (PH) is a haemodynamic and pathophysiological condition defined as an increase in mean pulmonary arterial pressure ≥25 mmHg at rest as assessed by right heart catheterization (Table 3). PH can be found in multiple clinical conditions (Table 4).

2. Pulmonary arterial hypertension (PAH, group 1) is a clinical condition characterized by the presence of pre-capillary PH (Table 3) and pulmonary vascular resistance >3 Wood units, in the absence of other causes of pre-capillary PH such as PH due to lung diseases, chronic thromboembolic PH, or other rare diseases (Table 4). PAH includes different forms that share a similar clinical picture and virtually identical pathological changes of the lung microcirculation (Table 4).

3. There is no sufficient data to support the definition of 'PH on exercise'.

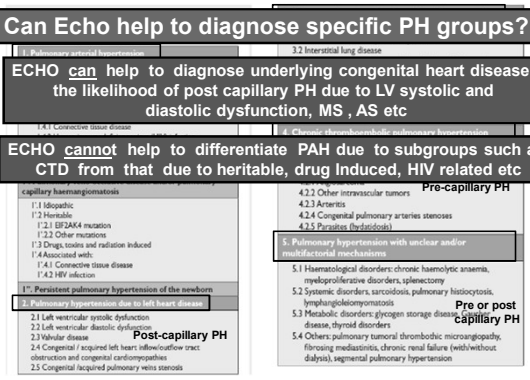
Galie *et al. EHJ ESC Guidelines Aug 2015*

2015 ESC/ERS Guidelines for the diagnosis and treatment of Pulmonary Hypertension

Can Echo help to diagnose specific PH groups?

ECHO can help to diagnose underlying congenital heart disease; the likelihood of post capillary PH due to LV systolic and diastolic dysfunction, MS , AS etc

ECHO cannot help to differentiate PAH due to subgroups such as CTD from that due to heritable, drug Induced, HIV related etc



GROUP 1 and Group 1′, 1″

Group 1: Pulmonary Arterial Hypertension

- **1.1 Idiopathic PAH**
- **1.2 Heritable PAH**
- **1.3 Drug and toxin induced**
- **1.4 Associated with**
  - **1.4.1 Connective tissue disease**
  - **1.4.2 HIV infection**
  - **1.4.3 Portal hypertension**
  - **1.4.4 Congenital heart diseases**
  - **1.4.5 Schistosomiasis**

Group 1′ : Pulmonary veno-occlusive disease and / or pulmonary capillary haemangiomatosis

Group 1″ : Persistent PH of the newborn (PPHN)

- ECHO cannot diagnose or differentiate the different sub-groups of Group 1 PAH (ie differentiate PAH due to HIV or connective tissue disease) but
- In PAH Group 1.4.4: PAH associated with congenital heart disease, it can help to sub-divide patients into 4 sub-groups based on
  - Defect size
  - Direction of shunt
  - In post op congenital patients to exclude significant residual defect

GROUP 2

(pulmonary venous hypertension, due to left heart disease)

2.1 Left ventricular systolic dysfunction

2.2 Left ventricular diastolic dysfunction

2.3 Valvular disease

2.4 Congenital / acquired left heart inflow / outflow tract obstruction and congenital cardiomyopathies

2.5 Congenital / Acquired pulmonary vein stenosis

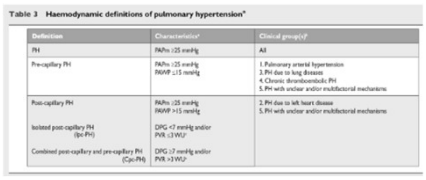


Table 3 Haemodynamic definitions of pulmonary hypertension*

Definition	Characteristics*	Clinical group(s)†
PH	PAPm ≥25 mmHg	All
Pre-capillary PH	PAPm ≥25 mmHg PAWP ≤15 mmHg	1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU‡	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU‡	

Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU‡	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU‡	

Grp 2 PH is a post capillary PH
(pulmonary venous hypertension)

GROUP 3

(due to chronic lung disease / hypoxia)

3.1 Chronic obstructive pulmonary disease
3.2 Interstitial lung disease
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4 Sleep-disordered breathing
3.5 Alveolar hypoventilation disorders
3.6 Chronic exposure to high altitude
3.7 Developmental lung diseases

GROUP 4

Chronic Thrombo-embolic Pulmonary Hypertension (CTEPH) and Other pulmonary artery obstruction (new addition in ESC 2015 guidelines)

2015 ESC/ERS Guidelines for the diagnosis and treatment of Pulmonary Hypertension

4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions
4.1 Chronic thromboembolic pulmonary hypertension
4.2 Other pulmonary artery obstructions
4.2.1 Angiosarcoma
4.2.2 Other intravascular tumors
4.2.3 Arteritis
4.2.4 Congenital pulmonary arteries stenoses
4.2.5 Parasites (hydatidosis)

Diagnosis of CTEPH:

- **To diagnosed CTEPH after PE, need at least 3 months of effective anticoagulation.**
- **To diagnose CTEPH, need to demonstrate:**
  1) Mean PAP ≥25 mmHg with PAWP ≤15 mmHg (cath haemodynamics)
  2) Mismatched perfusion defects on VQ scan and specific diagnostic signs for CTEPH seen by MDCT, CMR or pulmonary angiography, such as ring-like stenoses, webs/slits and chronic total occlusions (pouch lesions or tapered lesions)
- **Echo cannot diagnose CTEPH, can only give the probability of PH**
- **In rare cases can help to diagnose Group 4 PH due to pulmonary artery obstructions from tumour if seen in main PA, exclude congenital pulmonary artery stenosis if stenosis is in main PA or ostial branch PAs**

GROUP 5
(Unclear Multifactorial Mechanisms)

5.1 Hematologic disorders

5.2 Systemic disorders

5.3 Metabolic disorders

5.4 Others

5. Pulmonary Hypertension of Unclear Multifactorial Mechanisms

5.1 Hematologic disorders:
- Chronic hemolytic anemia
- Myeloproliferative disorders
- Splenectomy

5.2 Systemic disorders:
- Sarcoidosis
- Pulmonary histiocytosis,
- Lymphangioleiomyomatosis

5.3 Metabolic disorders:
- Glycogenstorage disease
- Gaucher disease
- Thyroid disorders

5.4 Others:
- Tumoral obstruction
- Fibrosing mediastinitis
- Chronic renal failure (with or without dialysis)
- Segmental pulmonary hypertension

ECHO cannot diagnose Group 5 PH which can have both pre and post capillary PH haemodynamics

Outline

- **Diagnosing Pulmonary Hypertension – ESC Guidelines**
- **The role of Echo in the diagnosis of Pulmonary Hypertension**
- **Summary**

GUIDELINES:
ECHO in the DIAGNOSIS OF PulMONARY HYPERTENTION

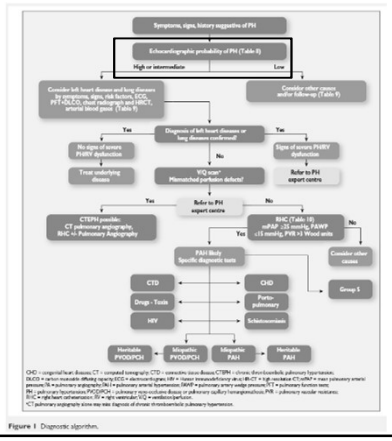


Figure 1 Diagnostic algorithm.

2015 ESC/ERS Guidelines for the diagnosis and treatment of Pulmonary Hypertension

2015 ESC Guidelines on Echo probability of PHT

Table 8A 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
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

Numbers to remember is
- **≤ 2.8 m/s**
- **> 3.4 m/s**

Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles	B: Pulmonary artery*	C: Inferior vena cava and right atrium
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm.	

PA = pulmonary artery.
*Echocardiographic signs from at least two different categories (A/B/C) from the list should be present to alter the level of echocardiographic probability of pulmonary hypertension.

2015 ESC Guidelines on Echo probability of PHT

Table 9 Diagnostic management suggested according to echocardiographic probability of pulmonary hypertension in patients with symptoms compatible with pulmonary hypertension, with or without risk factors for pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension

Echocardiographic probability of PH	Without risk factors or associated condition for PAH or CTEPH[a]	Class[b]	Level[c]	With risk factors or associated conditions for PAH or CTEPH[a]	Class[b]	Level[c]	Ref[d]
Low	Alternative diagnosis should be considered	IIa	C	Echo follow-up should be considered	IIa	C	
Intermediate	Alternative diagnosis, echo follow-up, should be considered	IIa	C	Further assessment of PH including RHC should be considered[e]	IIa	B	45, 46
	Further investigation of PH may be considered[e]	IIb					
High	Further investigation of PH (including RHC) is recommended	I	C	Further investigation of PH[f] including RHC is recommended	I	C	

CTEPH = chronic thromboembolic pulmonary hypertension; Echo = echocardiographic; PAH = pulmonary arterial hypertension; PH = pulmonary hypertension; RHC = right heart catheterization.
[a]Class of recommendation.
[b]Level of evidence.
[c]Reference(s) supporting recommendations.
[d]These recommendations do not apply to patients with diffuse parenchymal lung disease or left heart disease.
[e]Depending on the presence of risk factors for PH group 2, 3 or 5.
[f]Further investigation strategy may differ depending on whether risk factors/associated conditions suggest higher probability of PAH or CTEPH – see diagnostic algorithm.

TR velocities and PASP

- **Simplified Bernoulli equation**
  RVSP: 4 (TR Vmax)^2 + RA pressure
  RVSP = PASP in the absence of RVOT / pulmonary valve obstruction
- **RA pressure**

Table 3 Estimation of RA pressure on the basis of IVC diameter and collapse

Variable	Normal (0-5 [3] mm Hg)	Intermediate (5-10 [8] mm Hg)		High (15 mm Hg)
IVC diameter	≤2.1 cm	≤2.1 cm	>2.1 cm	>2.1 cm
Collapse with sniff	>50%	<50%	>50%	<50%

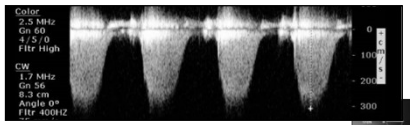
or < 20% on quiet inspiration

- **Use TR velocities for probability generation because any inaccuracies with RA pressure estimation can amplify errors**
- **Bear in mind, TR velocity depends on RV Function**

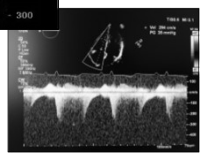
Low Downs on TR velocity:

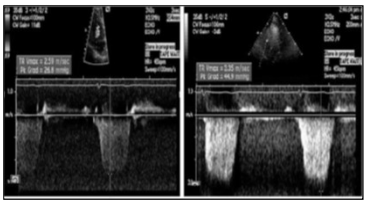
Take extra care when measuring TR velocity (not to overestimate)

- **TR velocity (to describe in report that TR jet is incomplete, see below) which means TR jet velocity is likely to be underestimated)**
  - **TR velocity can be over-estimated**



Or TR velocity under-estimated

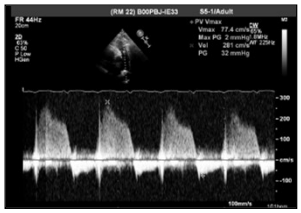




- **Measure TR signals from several windows and use the signal with the highest velocity if in sinus rhythm + sweep speeds of 100mm/s (If AF, need to average at least 5 readings)**
- **Weak signals (incomplete envelopes)**
  - Enhanced with agitated saline/ blood saline contrast
- **Measure only the well defined parts of the TR jet, most dense part**

Low Downs :
- **counter check with other parameters**

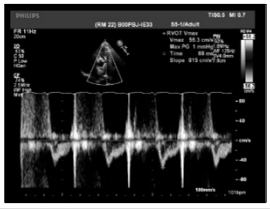
Other ways to derive estimated pressures

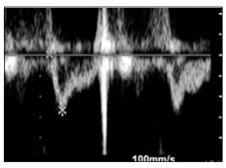


- **PA Diastolic Pressure**
  PADP: 4 (End diastolic pulmonary regurgitation velocity)^2 + RA pressure
- **Mean PAP (Masuyama)**
  MPAP: 4 (Early diastolic pulmonary regurgitation velocity)^2 + RA pressure

Other ways to derive estimated PA pressures

Mahan's formula
- Using pulmonary acceleration time by pulsed Doppler of pulmonary artery in systole
- **MPAP: 79 – (0.45 x acceleration time)** → **AT > 120ms**
- **MPAP: 90 – (0.62 x acceleration time)** → **AT < 120ms**





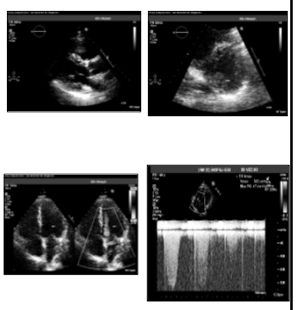
Low Downs on TR velocity : Consider Pressure and Flow

PASP = 65 + RAP mmHg

Patient A

PASP = 47 + RAP mmHg

Patient B



Low Downs: Pressure and Flow

- Patient A and Patient B are the same patient
- Patient A echo was performed a few hours before dialysis and Patient B echo was performed 12 hours post dialysis
- Volume loading in certain patient groups (eg. Patients with ESRF/ anemia / thyrotoxicosis) which can affect echo results esp TR velocity
- In this patient, the TR velocity and PASP are higher because of volume loading (increase flow); however there is still underlying PH (increase pulmonary Vascular resistance)

Pressure = Cardiac output (flow) x Resistance

Best practice:

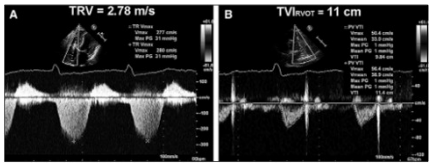
1. **Perform echo on patients after anemia or thyrotoxicosis is fully treated**
2. **For serial echo assessment of PAH patients with ESRF, request for echo to be performed 1 day after dialysis (for consistent comparison)**

Pulmonary vascular resistance (PVR)

PRESSURE change = Flow X Resistance

- PVR distinguishes elevated pulmonary pressure due to high flow from that due to pulmonary vascular disease
- PVR can be estimated using the ratio of
  - **PVR (woods) = {TR max velocity (m/s) / RVOT VTI (in cm)} X constant**

  This relationship is not reliable in patients with very high PVR, with measured PVR > 8 Wood units
- Normal PVR is <1.5 Wood units, significant PH is defined as a PVR > 3 Wood units



Abbas et al : JASE 2013: Volume 26 Number 10

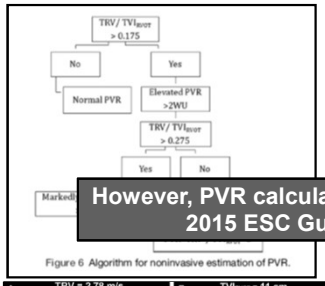
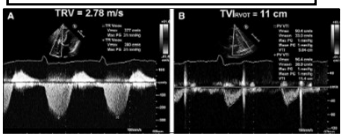


Figure 6 Algorithm for noninvasive estimation of PVR.

Resistance (PVR) = PA – LA Pressure difference / RV CO

Simplified Method to estimate / calculate PVR by Abbas et al using TR Vmax and RVOT VTI

However, PVR calculation is not in the 2015 ESC Guidelines



TRV / TVI_{rvot} = 2.78 /11 = 0.253

PVR = TRV / TVI_{rvot} x 10 = 2.53 WU

Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm.	

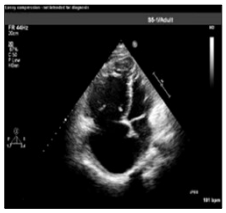
PA = pulmonary artery.
[a]Echocardiographic signs from at least two different categories (A/B/C) from the list should be present to alter the level of echocardiographic probability of pulmonary hypertension.

Echo signs suggestive of PH: If present in <u>at least 2 different categories</u> will result in higher probability of PH

Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm.	

PA = pulmonary artery.
[a]Echocardiographic signs from at least two different categories (A/B/C) from the list should be present to alter the level of echocardiographic probability of pulmonary hypertension.



Low Downs: Echo signs suggestive of Pulmonary Hypertension

- Dilated Right Ventricle
- Dilated Right Atrium
- Flatted IVS

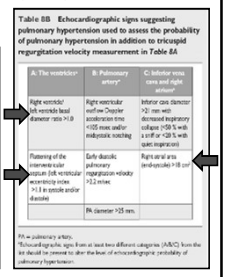


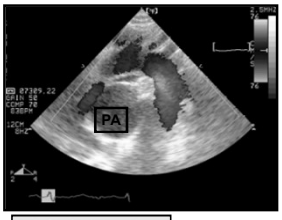
Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles*	B: Pulmonary artery*	C: Inferior vena cava and right atrium*
Right ventricle/left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or mid-systolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systolic) >18 cm²
	PA diameter >25 mm.	

PA = pulmonary artery.
*Echocardiographic signs from at least two different categories (A/B/C) from the list should be present to alter the level of echocardiographic probability of pulmonary hypertension.

AT least 1 from 2 different categories

Echo signs of Pulmonary Hypertension



- Dilated Pulmonary Artery > 25 mm (avoid measuring in an oblique angle but sometimes difficult when PA is dilated and unable to get a perpendicular axis)
- Dilated PA can also be seen on other views eg. suprasternal view

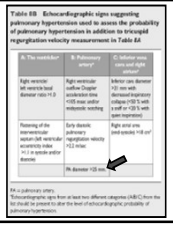
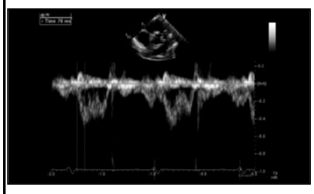


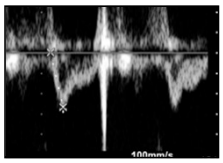
Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles*	B: Pulmonary artery*	C: Inferior vena cava and right atrium*
Right ventricle/left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or mid-systolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systolic) >18 cm²
	PA diameter >25 mm.	

PA = pulmonary artery.
*Echocardiographic signs from at least two different categories (A/B/C) from the list should be present to alter the level of echocardiographic probability of pulmonary hypertension.

Echo signs of Pulmonary Hypertension





Short Acceleration Time

RVOT Pulse Wave - Mid Systolic Notch

- Notching is a very important sign, reflects PA stiffness
- The absence of RVOT notching strongly predicts pulmonary venous hypertension, with an odds ratio of 33:1
- In contrast, a notched (mid or late) Doppler pattern was present in 100% of incident cases of PAH, and predicted a PVR >3 Wood Units, with an odds ratio of 22:1

Reference: Am J Respir Crit Care Med 2011;183:268–276.

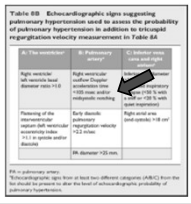
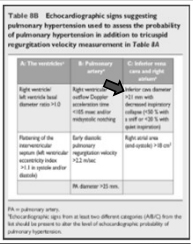


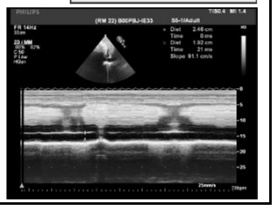
Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles*	B: Pulmonary artery*	C: Inferior vena cava and right atrium*
Right ventricle/left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or mid-systolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systolic) >18 cm²
	PA diameter >25 mm.	

PA = pulmonary artery.
*Echocardiographic signs from at least two different categories (A/B/C) from the list should be present to alter the level of echocardiographic probability of pulmonary hypertension.

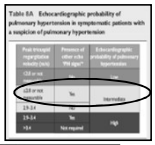
Echo signs of Pulmonary Hypertension

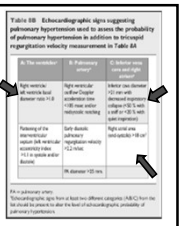




- Dilated and Plethoric Inferior Vena Cava

However, things are not always as they appear to be





- TR velocity 2.78, hence less than 2.8
- Presence of PH echo signs in 2 different categories
- Hence overall, probability of PH is INTERMEDIATE

BUT WAIT

- Patient also has severe TR, hence RA, RV dilatation and IVC plethora can be due to TR and not PH
- PH probability can also be LOW instead of INTERMEDIATE as the other signs are due to TR

Low Downs: When reading echo

When TR velocity is low (< 2.8 m/s) but other "PH signs" are present,

- Be sure that these PH signs are not due to other concomitant causes such as TR, ASD, PR, chronic AF, etc

otherwise,

- It would not be accurate to use these signs to elevate the overall probability of PH

Don't just look at specific things, look at overall picture or clinical context

2015 ESC/ERS Guidelines for the diagnosis and treatment of Pulmonary Hypertension

Table 14 Suggested assessment and timing for the follow-up of patients with pulmonary arterial hypertension

	At baseline	Every 3–6 months[a]	Every 6–12 months[a]	3–6 months after changes in therapy[a]	In case of clinical worsening
Medical assessment and determination of functional class	+	+	+	+	+
ECG	+	+	+	+	+
6MWT/Borg dyspnoea score	+	+	+	+	+
CPET	+		+		+[f]
Echo	+		+	+	+
Basic lab[b]	+	+	+	+	+
Extended lab[c]	+		+		+
Blood gas analysis[d]	+		+	+	+
Right heart catheterization	+		+[f]	+[f]	+[f]

How often to do serial echo in follow up of PH patients?

- **At baseline**
- **6-12 monthly if no change in PH therapy**
- **3-4 monthly after change of PH therapy**

2015 ESC/ERS Guidelines for the diagnosis and treatment of Pulmonary Hypertension

Table 13 Risk assessment in pulmonary arterial hypertension

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[b]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 ml/min/kg (35–65% pred.) VE/VCO_2 slope 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/ml	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60%

1. **RA size – RA area**
2. **Presence of Pericardial effusion**

What can the presence of <u>RA dilatation</u> on echo tells us?

- RA enlargement is another indirect measure of RV dysfunction
- Increased RA area index **(more than 18 cm^2)** is predictive of increased mortality in PAH
- RA dilatation occurs as a consequence of
  - **Impaired RV systolic function**
  - **Longstanding TR and RA hypertension causing RA remodeling**
  - **Progressive RA dilatation may also reflect dysfunction of the RA itself**
- RA systolic function is likely to play a critical role in supporting total right heart function in PAH

What to do when the echo reports the presence of pericardial effusion?

- **The presence of pericardial effusion is an indirect index of RV dysfunction**
- **Mild-moderate circumferential pericardial effusions are seen in 20%–50%** of patients with PAH *(JASE 1999;12:655–662)*
- **However, the presence of pericardial effusion as specific sign of severe PAH lacks sensitivity and specificity as**
  - **The presence of a pericardial does not always indicate right heart decompensation especially in PAH associated with connective tissue disease (CTD), where the effusion may reflect an underlying serositis**
- **Attempted drainage** of pericardial effusions in patients with PAH has been associated with **very poor outcomes**, with most deaths occurring within 12–24 hours of the procedure *(Southern Med J 2008;101:490–494)*
- PAH associated pericardial effusions **may progressively decrease in size or resolve** over weeks or months in response to diuresis and pulmonary vasodilator therapy

Echocardiographic Predictors

Table 2. Echocardiographic measures predictive of survival

Echocardiographic parameters	Worse prognosis
Tricuspid annular plane systolic excursion	<15 mm
Right ventricular Doppler (Tei) index	>0.88
Pericardial effusion	Present
Left ventricular eccentricity index at end-diastole	>1.7
Right atrial volume	Increasing size
Right ventricular fractional area change	Decreasing %

Howard LS. Prognostic factors in pulmonary arterial hypertension: assessing the course of the disease. Eur Respir Rev. 2011 Dec;20(122):236-42.

In reality, the survival of PAH patients is determined by their

Right Ventricle Function

and hence serial assessment of RV function is mandatory in the echo follow up of PH patients

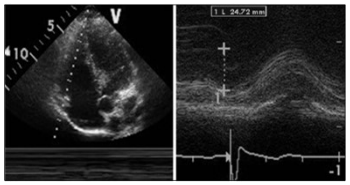
RV systolic function

- **RV systolic function has been evaluated using**
  - **TAPSE (Tricuspid annular peak systolic excursion)**
  - **RIMP (RV index of myocardial performance)**
  - **RV dP/dT**
  - **2D RV FAC (Fractional area of change)**
  - **2D RV ejection fraction (RVEF)**
  - **3D RVEF**
  - **Tissue Doppler derived tricuspid lateral annular systolic velocity (S')**
  - **Longitudinal strain and strain rate**

TAPSE (Tricuspid Annular Peak Systolic Excursion)

- **TAPSE represents longitudinal RV function**
- **TAPSE is simple, less dependent on optimal image quality and reproducible**
- **Disadvantages of TAPSE:**
  - **represents the function of a complex 3D structure**
  - **it is angle dependent and may be load dependent**
- **Recommendations:**
  - **TAPSE can be used as a simple method of estimating RV function**

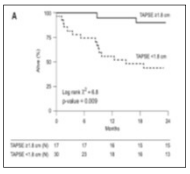
  TAPSE < 17 mm in adults suggest impaired RV longitudinal function



What does echo TAPSE tells you in PAH?

- In PAH patients, echo **TAPSE ≤1.5 cm predicts a 3X higher rate of death** or emergent lung transplant compared to subjects with a TAPSE >1.5 cm *(Int J Card 2010;140:272–278)*
- TAPSE should also be interpreted differently when TR is present. **TAPSE > 1.5 cm in a PAH patient with moderate to severe TR is associated with worse outcome** than patients with a TAPSE >1.5 cm and minimal to mild TR *(J Am Soc Echocardiogr 2002;15:1160–1164)*
- **TAPSE < 1.8 predicts RV stroke volume index (SVI) <29 mL/m2** with 87% accuracy and is associated with increased hospitalization rates for RV failure and decreased survival in patients with PAH

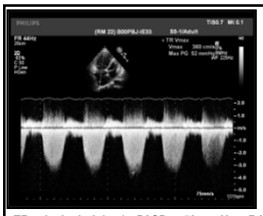
  (American Journal of Respiratory and Critical Care Medicine. 174: 1034 –1041)



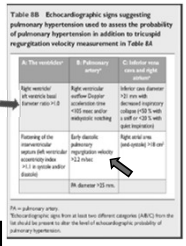
Example of a CTD patient with SOB and suspected of PAH: Let's read the echo together

LV Eccentricity Index at end diastole = 6.28/3.67 = 1.7

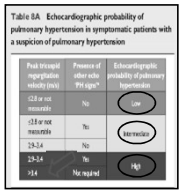
LV Eccentricity Index at end systole =4.15/3.03 =1.4



TR velocity is 3.6 m/s; PASP = 52 mmHg + RAP

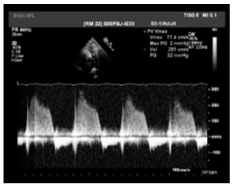


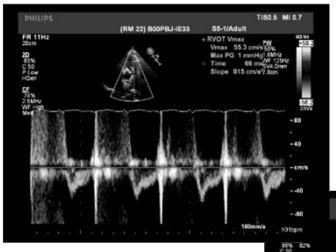
Peak PR velocity is at least 2.81 m/s, hence mean PAP at least 32 mmHg + RAP



As this patient's TR velocity is already greater than 3.4 m/s, the echo probability of pulmonary hypertension is **HIGH**

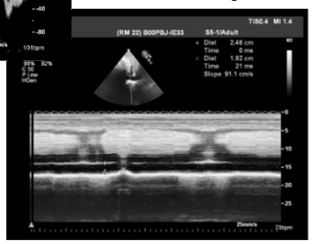
The presence of other echo PH signs will not add further to the probability of PH



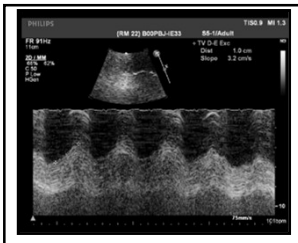


TR velocity is 3.6 m/s;
PASP = 52 mmHg + RAP
= 52 + 15
= 67 mmHg

Dilated and plethoric IVC, estimated RAP at least 15 mmHg



Short ACT of 66 ms with mid-systolic notch present



TAPSE = 1 (reduced)
TAPSE ≤1.5 cm predicts a 3X higher rate of death

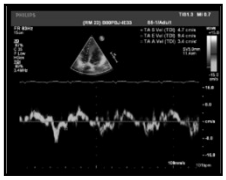
TR velocity/PASP depends on function of RV

<u>Caution</u>:
When RV function deteriorates, TR velocity and hence PASP falls

- **This indicates progression of disease and not improvement of condition**
- **TR velocity/PASP should be interpreted together with RV function**

- **TAPSE and TDI 's' values suggest that the RV function is depressed in this patient**
- **Hence, the PASP of 67 mmHg would have underestimated the severity of the PH in this patient. For this pressure of 67 mmHg, RV CO is likely low and hence PVR high**
  (Pressure = CO x Resistance)

TDI 's' wave = 4.7



In conclusion, this echo should read

- **Echo probability of pulmonary hypertension is HIGH**
- **RV is dilated with depressed function and there is moderate to severe degree of tricuspid regurgitation due to increased annular dilation. This suggest there is already chronic RV remodelling**
- **LV is compressed, RVOT flow showed mid-systolic notching. Again, features are consistent with high likelihood of pulmonary arterial hypertension**
- **Patient should be advised to undergo right and left heart catheterisation to calculate PVR and confirm the diagnosis of PAH**

Outline

- **Diagnosing Pulmonary Hypertension – ESC Guidelines**
- **New Echo Guidelines**
- **The role of Echo in the diagnosis of Pulmonary Hypertension**
- **Summary**

IN SUMMARY:

- **Echo is commonly ordered for patients suspected to have pulmonary hypertension**
- **Echo is a very useful tool (first step) to help categorize patients into low, intermediate or high probability of pulmonary hypertension (2015 ESC Guideline on the echo probability for the diagnosis of pulmonary hypertension)**
- **The echo probability of PH is based on TR velocity + other PH signs (and not estimated PASP or derived PVR)**
- **Be wary of potential pitfalls in measurement of TR velocity and in assessing 'other PH signs'**
- **Echo cannot confirm PH as catheter based haemodynamic data is required for confirmation**
- **In established PH patients- echo parameters assessing RV function (eg TAPSE), LV deformity (LV eccentricity index), presence of pericardial effusion, etc provide additional prognostic information**

Thank You