Pulmonary Hypertension

Echocardiography: Pearls & Pitfalls

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Ιωάννινα, 13/02/2016
• Nothing to declare
Transthoracic echocardiography is used to:
  – estimate PAP from continuous wave Doppler measurements
  – image the effects of PH on the heart
  – infer a diagnosis of PH in patients in whom multiple different
    echocardiographic measurements are consistent with this diagnosis

Echocardiography should always be performed when PH is suspected

When treatment of PH itself is being considered, echocardiography alone is not sufficient to support a treatment decision and cardiac catheterization is required.
2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

Diagnostic algorithm

Symptoms, signs, history suggestive of PH

Echocardiographic probability of PH (Table 8)

High or intermediate

Consider left heart disease and lung diseases by symptoms, signs, risk factors, ECG, PFT+DLCO, chest radiograph and HRCT, arterial blood gases (Table 9)

Low

Consider other causes and/or follow-up (Table 9)

PAH likely
Specific diagnostic tests

CTD

Drugs - Toxin

HIV

CHD

Porto-pulmonary

Schistosomiasis

Heritable PVOD/PCH

Idiopathic PVOD/PCH

Idiopathic PAH

Heritable PAH

Consider other causes

Group 5
What info are we getting from echo

1. PASP
2. RV size and function
3. Causes of PH
4. Prognosis
Accuracy of Doppler Echocardiography in the Hemodynamic Assessment of Pulmonary Hypertension

48% of the echocardiographic estimates were accurate (within 10 mm Hg of the RHC value).

DE within 1 hour of RHC

PASP Estimation Pitfalls

- PASP = 4V^2 + RAP
- Pitfalls:
  - TR Related
  - RAP Related
PASP Estimation Pitfalls I

TR related:

• The severity of tricuspid regurgitation does not correlate with degree of PH
• Overestimation
• Underestimation
• RVOT Stenosis
Shadow (Overestimation)

Reduce the Gain!
Non parallel position of the transducer (Underestimation)
Little things make a big difference!

4 chambers

4 chambers RV focused view
Suboptimal signal (Underestimation)

Contrast echocardiography (agitated saline administered i.v.) can improve the signal
Use all the available views!

The maximal velocity depends on optimal alignment with the jet.
Severe TR (underestimation)

Triangular protosystolic shape (early equalization of RV and RA pressures)
RVOT Stenosis (Non Reliable)

42 yo, male, Congenital PV Stenosis
RAP related (IVC Diameter):

- **Underestimation**
  - Left lateral position: increased intra-abdominal pressure and compression of the IVC by the liver

- **Overestimation**
  - Measurement not perpendicular to the IVC long axis
  - Dilated and may not collapse in patients on ventilators

### Right Atrial Pressure Estimation

<table>
<thead>
<tr>
<th>IVC Diameter (cm)</th>
<th>Collapse (%)</th>
<th>Estimated RA Pressure (mmHg)</th>
<th>Range (mmHg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤2.1 cm</td>
<td>&gt;50%</td>
<td>3</td>
<td>0-5</td>
</tr>
<tr>
<td>&gt; 2.1 cm</td>
<td>&lt;50%</td>
<td>15</td>
<td>10-20</td>
</tr>
<tr>
<td>Rest of cases</td>
<td></td>
<td>8</td>
<td>5-10</td>
</tr>
</tbody>
</table>
“..given the inaccuracies of RAP estimation and the amplification of measurement errors by using derived variables, we recommend using the continuous wave Doppler measurement of peak TRV (and not the estimated PASP) as the main variable for assigning the echocardiographic probability of PH.”
The TR Velocity only is not sufficient!

- Search for other echo findings that raise or reinforce suspicion of PH

<table>
<thead>
<tr>
<th>Table 8B</th>
<th>Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in Table 8A</th>
</tr>
</thead>
<tbody>
<tr>
<td>A: The ventricles&lt;sup&gt;a&lt;/sup&gt;</td>
<td>B: Pulmonary artery&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
<tr>
<td>Right ventricle/ left ventricle basal diameter ratio &gt;1.0</td>
<td>Right ventricular outflow Doppler acceleration time &lt;105 msec and/or midsystolic notching</td>
</tr>
<tr>
<td>Flattening of the interventricular septum (left ventricular eccentricity index &gt;1.1 in systole and/or diastole)</td>
<td>Early diastolic pulmonary regurgitation velocity &gt;2.2 m/sec</td>
</tr>
<tr>
<td>PA diameter &gt;25 mm.</td>
<td></td>
</tr>
</tbody>
</table>
Short AccT & Notching

56 yo, female, NYHA II FC, with scleroderma

RHC: mean PAP 38 mmHg, sPAP: 67 mmHg
Right Ventricle Assessment

Structurally complex cavity:

- complex shape
- irregular endocardial surface due to heavy trabeculation: difficult to delineate endocardial border
- location behind the sternum: inadequate image quality
- Fits no simple geometric figure:
  - failure to standardize RV volume determination
Right ventricle–focused view
Use all the available views!

RV size estimation requires integration of multiple views and qualitative and quantitative assessment.
RV:LV Ratio - Foreshortening
LV eccentricity index
Volume vs Pressure Overload

RV volume overload
- the flattening of the septum is seen only in diastole
- Ecc Index >1 at end diastole

RV pressure overload
- the septum maintains the altered shape during the entire cardiac cycle
- Ecc Index >1 at end systole and end diastole
Longitudinal function of the right ventricle

- TAPSE
  - angle dependency
  - load dependency (significant tricuspid regurgitation)
  - LV systolic performance may influence TAPSE value due to ventricular interdependence

- Tissue Doppler
  - Ensure optimal image orientation to avoid underestimation
  - Insufficient data in the elderly
Fractional Area Change

- Quantitative estimation of RV function
- Good correlation with RVEF
- Take Care:
  - Accurate definition of endocardial limits
  - Trace the free wall beneath the trabeculations
Right Heart Dysfunction ≠ PH

- RV Infarction
- Arrhythmogenic right ventricular dysplasia (ARVC)
- Congenital heart disease
Normal RV Function does not exclude PH
Causes of PH
Be careful!

- LHD (group2) is the most common cause of PH

2. Pulmonary 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 veins stenosis
# Left ventricular heart failure and pulmonary hypertension

## Suggestive of PAH (Nice group 1)
- Younger age, familial cases, bendopnea\(^a\), risk factors for PAH: CTD, CHD, severe liver disease, portal hypertension, HIV
- RV hypertrophy, right axis, RV strain
- Enlarged right heart chambers, dilated PA, peripheral PA pruning
- Normal/mild obstructive spirometry, normal or moderately decreased DLCO\(^d\), low \(p_{\text{CO}_2}\) (\(\leq 36\) mmHg)\(^6\)
- BNP/NTproBNP elevated (not discriminate between Groups 1 and 2)
- Low \(P_{\text{ET}}\)\(\text{CO}_2\) at AT, decreasing during exercise; high V\(E/\text{VCO}_2\), increasing during exercise
- To diagnose or rule out parenchymal lung disease (not discriminate between Groups 1 and 2)
- To diagnose or rule out CTEPH (not discriminate between Groups 1 and 2)

## Suggestive of PH-LHD (Nice group 2)
- Older age, hypertension, diabetes, CAD, BMI > 30, pulmonary congestion, history of pulmonary oedema, orthopnoea
- LV hypertrophy (Sokolow–Lyon index: S in V1 + R in V6), left axis, atrial fibrillation
- Enlarged LA, LV hypertrophy, signs of systolic (EF) and/or diastolic (E/A, DT, E/E') LV dysfunction, valvular disease
- Pulmonary congestion, Kerley B lines, pleural effusions, enlargement of left heart chambers
- Normal/obstructive spirometry, normal DLCO (may be decreased due to comorbid COPD), high \(p_{\text{CO}_2}\) (> 36 mmHg)\(^6\)
- BNP/NTproBNP elevated (not discriminate between group 1 and 2)
- \(P_{\text{ET}}\)\(\text{CO}_2\) at AT normal or slightly lowered, not decreasing during exercise, V\(E/\text{VCO}_2\) not increasing during exercise
- To diagnose or rule out parenchymal lung disease (not discriminate between Groups 1 and 2)
- To diagnose or rule out CTEPH (not discriminate between Groups 1 and 2)
PH due to LHD

81 yo, Female, AF, LVH, LA/RA = 1, E'/e >15, TR = 3.3 m/sec
PH due to PAH

64 yo, Female, E<A, RV/LV > 1, RA/LA > 1, Interatrial septum bows from RA to LA, TR = 5.06 m/sec
Prognosis
### 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

#### Table 13  Risk assessment in pulmonary arterial hypertension

<table>
<thead>
<tr>
<th>Determinants of prognosis* (estimated 1-year mortality)</th>
<th>Low risk &lt;5%</th>
<th>Intermediate risk 5–10%</th>
<th>High risk &gt;10%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical signs of right heart failure</td>
<td>Absent</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Progression of symptoms</td>
<td>No</td>
<td>Slow</td>
<td>Rapid</td>
</tr>
<tr>
<td>Syncope</td>
<td>No</td>
<td>Occasional syncope(^b)</td>
<td>Repeated syncope(^c)</td>
</tr>
<tr>
<td>WHO functional class</td>
<td>I, II</td>
<td>III</td>
<td>IV</td>
</tr>
<tr>
<td>6MWD</td>
<td>&gt;440 m</td>
<td>165–440 m</td>
<td>&lt;165 m</td>
</tr>
<tr>
<td>Cardiopulmonary exercise testing</td>
<td>Peak VO$_2$ &gt;15 ml/min/kg (&gt;65% pred.) VE/VCO$_2$ slope &lt;36</td>
<td>Peak VO$_2$ 11–15 ml/min/kg (35–65% pred.) VE/VCO$_2$ slope 36–44.9</td>
<td>Peak VO$_2$ &lt;11 ml/min/kg (&lt;35% pred.) VE/VCO$_2$ slope ≥45</td>
</tr>
<tr>
<td>NT-proBNP plasma levels</td>
<td>BNP &lt;50 ng/l</td>
<td>BNP 50–300 ng/l</td>
<td>BNP &gt;300 ng/l</td>
</tr>
<tr>
<td></td>
<td>NT-proBNP &lt;300 ng/l</td>
<td>NT-proBNP 300–1400 ng/l</td>
<td>NT-proBNP &gt;1400 ng/l</td>
</tr>
<tr>
<td>Imaging (echocardiography, CMR imaging)</td>
<td>RA area &lt;18 cm$^2$ No pericardial effusion</td>
<td>RA area 18–26 cm$^2$ No or minimal, pericardial effusion</td>
<td>RA area &gt;26 cm$^2$ Pericardial effusion</td>
</tr>
<tr>
<td>Haemodynamics</td>
<td>RAP &lt;8 mmHg CI ≥2.5 l/min/m$^2$ SvO$_2$ &gt;65%</td>
<td>RAP 8–14 mmHg CI 2.0–2.4 l/min/m$^2$ SvO$_2$ 60–65%</td>
<td>RAP &gt;14 mmHg CI &lt;2.0 l/min/m$^2$ SvO$_2$ &lt;60%</td>
</tr>
</tbody>
</table>

*Includes both right and left heart failure.

\(^a\) May be influenced by disease duration.

\(^b\) Occasional syncope defined as 1–2 episodes/year.

\(^c\) Repeated syncope defined as >2 episodes/year.
Pulmonary Hypertension Decreases the Predictive Accuracy of Echocardiographic Clues for Cardiac Tamponade*

Table 3—Sensitivity, Specificity, and Predictive Accuracy for Cardiac Tamponade in Patients Without and With Pulmonary Hypertension

<table>
<thead>
<tr>
<th></th>
<th>Normal Pulmonary Pressure</th>
<th>Pulmonary Hypertension</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Sensitivity, %</td>
<td>Specificity, %</td>
</tr>
<tr>
<td>RAC</td>
<td>82</td>
<td>67</td>
</tr>
<tr>
<td>RVDC</td>
<td>73</td>
<td>89</td>
</tr>
<tr>
<td>FVP</td>
<td>91</td>
<td>89</td>
</tr>
<tr>
<td>IVCP</td>
<td>100</td>
<td>89</td>
</tr>
</tbody>
</table>

*p<0.05 vs normal pulmonary pressure group.

Gary D. Plotnick, MD; David C. Rubin, MD; Zenaida Feliciano, MD; and Andrew A. Ziskind, MD

(Chest 1995; 107:919-24)
Tamponade?

- 71 yo, female,
- WHO-FC IV,
- Group I PH
- BP 90/60mmHg
- RVSP 105 mmHg
Pericardial involvement in PAH:

- Diagnosis of cardiac tamponade in a patient with severe PAH is challenging.
- High rightsided pressures can mask many of the typical right-sided clinical and echocardiographic findings of tamponade.
- Right-sided chamber collapse is uncommon
- Left atrial early diastolic collapse is more commonly seen.
Conclusions

✓ A detailed echocardiographic assessment must be performed in all patients with suspected PH

✓ TR velocity and other echocardiographic variables might raise or reinforce suspicion of PH

✓ Heart chambers must be evaluated for causative and prognostic information
Thank You!
Pericardial Effusion

A

B

No. at risk

Follow-up, years

Pericardial effusion

No pericardial effusion

No. at risk

Follow-up, years

Moderate or greater effusion

Small effusion

No effusion

No effusion

Effusion

≥ Mod effusion

No effortion

CHEST 2013; 144(5):1530–1538
Prognostic information

• right atrial area
• eccentricity index
• Pericardial effusion
• Tei index
• TAPSE
Right Heart Dysfunction ≠ PH

• Ischaemic right ventricular disease and right ventricular failure RV ischaemia complicates up to 50% of inferior myocardial infarction
  – RV free wall hypokinesia or akinesia detected by echocardiography is a qualitative and sensitive parameter for RV dysfunction
Right Heart Dysfunction ≠ PH

• Arrhythmogenic right ventricular dysplasia (ARVC)
  – Regional RV akinesia, dyskinesia, or aneurysm
  The triangle of dysplasia: a region including the subtricuspidal RV wall, the RV apex, and the RV outflow tract
Right Heart Dysfunction ≠ PH

- Congenital heart disease
  - chronic volume overload
  - intracardiac shunts (atrial septal defect),
  - anomalies of the pulmonary valve, and arteries (pulmonary atresia)
  - RV is pumping in the systemic circulation (transposition of the great arteries, single ventricle).
Also evaluate

- Pulmonary Artery
  - AcT (<105 msec) (HR 60-100 b/min)
  - Notching (midsystolic)
  - Diameter (> 25 mm)
  - Early diastolic Pulmonary Velocity Regurgitation (>2,2m/sec)
  - Mean PAP

- IVC diameter and Respiratory Variation

- Right Atrial Dimensions
Regional RV Strain and Strain Rate

- identify the early signs of RV dysfunction
- follow patients under therapy
- probably accounts for the RV global dysfunction, given the fibre arrangement
- RV longitudinal strain <19% has been described as an independent predictor of all-cause mortality
PAH vs PVH: Echocardiographic Differentiation

Mc Laughlin et al. JACC 2015; 65. NO 18.
# Checklist for the Echocardiographic Evaluation of PH

<table>
<thead>
<tr>
<th>Completed?</th>
<th>Action Item</th>
<th>Notes</th>
</tr>
</thead>
</table>
|           | Record estimated PASP | - Underestimated when Doppler beam alignment is poor or when TR jet is minimal  
|           |             | - Overestimated in patients with significant anemia or in some cases of agitated saline-enhanced TR jet on continuous wave Doppler (due to feathering)  
|           |             | - Assumes absence of pulmonic stenosis  
|           |             | - Echocardiographic PASP does not equal mean PA pressure (definition of PH per guidelines is on the basis of invasive hemodynamics: mean PA pressure >25 mm Hg) |
|           | Evaluate RV size and function | - Signs of RV enlargement (apical 4-chamber view): RV shares apex with LV; RV bigger than LV; RV basal diameter >4.2 cm  
|           |             | - RV hypertrophy (subcostal view): RV end-diastolic wall thickness >5 mm  
|           |             | - RV systolic dysfunction: RV fractional area change <35%; TAPSE <1.6 cm; RV tissue Doppler s' velocity <10 cm/s at base of the RV free wall (tricuspid annulus)  
|           |             | - Septal flattening: in systole = RV pressure overload and in diastole = RV volume overload |
|           | Evaluate for signs of elevated PVR | - RVOT notching on pulse-wave Doppler profile is a sign of elevated PVR  
|           |             | - Peak TR velocity (m/s)/RVOT VTI (cm) <0.18: unlikely PVR is elevated |
|           | Estimate volume status | - Use size and collapsibility of IVC (during sniff maneuver) to determine RA pressure  
|           |             | - Hepatic vein flow: systolic flow reversal can be a sign of severe TR, RV overload, and/or increased RV stiffness  
|           |             | - Signs of RA overload/enlargement: RA area >18 cm²; interatrial septum bows from right to left |
|           | Evaluate severity of TR | - Features suggestive of severe TR include dense TR jet on continuous-wave Doppler, V-wave cutoff sign, and systolic flow reversal on hepatic vein pulse-wave Doppler imaging |
|           | Evaluate for pericardial effusion | - In patients with PAH, the presence of a pericardial effusion = poor prognostic sign |
|           | Evaluate for causes of PH (left heart disease, shunt lesions) | - Left heart disease: look for overt LV systolic dysfunction, grade 2 or worse diastolic dysfunction, severe aortic or mitral valvular disease, and less common abnormalities of the left heart (e.g., hypertrophic cardiomyopathy, cor triatriatum)  
|           |             | - Shunt lesions: perform agitated saline bubble study |
|           | Differentiate PAH from PVH | - Signs favoring PVH: LA enlargement (LA size >RA size); interatrial septum bows from left to right; E/A ratio >1.2; E/e' (lateral) > 11; lateral e' <8 cm/s;  
|           |             | - In patients with significantly elevated PASP at rest: grade 1 diastolic dysfunction pattern (E/A ratio <0.8) favors PAH diagnosis because of underfilled LA and decreased LV compliance due to RV/LV interaction (extrinsic compression of LV by RV).  
|           |             | - See also Figure 1 |

*Mc Laughlin et al. JACC 2015; 65. NO 18.*
Echocardiographic Predictors of Adverse Outcomes in Primary Pulmonary Hypertension

- Right atrial size
- Pericardial effusion
- Eccentricity index
Non Conventional echocardiography

• RT3DE
  – Accurate volume analysis independent of RV size and shape
  – enables unique views to better understand specific causes of PH (i.e., septal defects, complex congenital pathology, left-sided valvular or ventricular heart disease)
  – investigate RV functional and morphologic changes

• Two-Dimensional Strain
  – identify the early signs of RV dysfunction
  – follow patients under therapy
  – <19% an independent predictor of all-cause mortality

Bossone et al. JACC 2013; 26. NO 1.
TR ≠ PH

• TR occurs as the result of the effect of increased RV afterload on RV dilatation and function.
• TR is usually caused by tricuspid annular dilation, altered RV geometry, and apical displacement of the tricuspid leaflets.
• The degree of TR cannot be used as a surrogate for the degree of PAP elevation.
Evaluation of the RV Function

• Subjective:
  – Thickening of the RV wall
  – Motion (Inward/radial/longitudinal)
  – Regional dysfunction
  – Integrate findings from various views

• RV fractional area change (FAC)

• TAPSE
• Tissue Doppler
• Tei Index
Echo inaccurate in 57.4% of PASP estimates (>10 mmHg higher or lower than RHC)

Farber et al. Congest Heart Fail. 2011 Mar-Apr;17(2):56-64
Use all the available views!

- The maximal velocity depends on optimal alignment with the jet
PASP Estimation Pitfalls

• The volume of tricuspid regurgitation does not correlate with the velocity
  – The absence of TR (moderate or severe) does not exclude PH
  – Severity of TR does not predict severity of PH

• Overestimation
  – Detecting shadow

• Underestimation
  – Non parallel position of the transducer
  – Suboptimal signal
  – “Free” TR
  – Eccentric tricuspid regurgitation jet

• RVOT Stenosis
IVC Diameter

Not reliable in:
• Normal young athletes (dilated in the presence of normal pressure)
• In patients on ventilators (TEE at the cavoatrial junction)
PH Pathophysiology

RV function and size are indicators of the severity and chronicity of pulmonary hypertension impose an additional cause of symptoms and reduced longevity. Right ventricular function is the most important determinant of longevity in patients with pulmonary arterial hypertension.
Right Atrium Evaluation

- Apical 4-chamber view
- End-systole
- Significant dilatation if $> 18\text{cm}^2$
- Right atrial enlargement is a manifestation of high right atrial pressure due to functional tricuspid regurgitation or elevated right ventricular diastolic pressure, both consequences of right ventricular failure

Accuracy of Doppler Echocardiography in the Hemodynamic Assessment of Pulmonary Hypertension

65 patients, various forms of PH, DE within 1 hour of a clinically indicated right-heart catheterization to compare noninvasive hemodynamic estimates with invasively measured values

• 48% of the echocardiographic estimates were accurate (within 10 mm Hg of the RHC value).
• the magnitude of underestimation was greater than the overestimation (38% pressure overestimates > 20 mm Hg 80% pressure underestimates > 20 mm Hg).
• 83% underestimates were associated with a fair or poor quality Doppler jet.
TAPSE

• Longitudinal function of the right ventricle

• Several limitations
  – angle dependency
  – load dependency (significant tricuspid regurgitation)
  – 1D (Regional RV myocardial abnormalities are neglected)
  – LV systolic performance may influence TAPSE value due to ventricular interdependence
Tissue Doppler

- Assumes that the function of a single segment represents the function of the entire right ventricle
- Normal value $S’ > 10\text{cm/sec}$
The TR Velocity only is not sufficient!

- Search for other echo findings that raise or reinforce suspicion 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  
Right ventricle/ left ventricle basal diameter ratio >1.0 | B: Pulmonary artery  
Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching | C: Inferior vena cava and right atrium  
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 cm² |
| PA diameter >25 mm. |