Role of imaging in pulmonary hypertension

Evaluating hemodynamics in pulmonary hypertension

Dimitrios Tsiapras  MD FESC

Onasis Cardiac Surgery Center
<table>
<thead>
<tr>
<th>Definition</th>
<th>Characteristics</th>
<th>Clinical group(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary hypertension (PH)</td>
<td>Mean PAP ≥ 25 mmHg</td>
<td>All</td>
</tr>
<tr>
<td>Pre-capillary PH</td>
<td>Mean PAP ≥ 25 mmHg PWP ≤ 15 mmHg CO normal or reduced</td>
<td>1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms</td>
</tr>
<tr>
<td>Post-capillary PH</td>
<td>Mean PAP ≥ 25 mmHg PWP ≥ 15 mmHg CO normal or reduced</td>
<td>2. PH due to left heart disease</td>
</tr>
<tr>
<td>Passive</td>
<td>TPG ≤ 12 mmHg</td>
<td></td>
</tr>
<tr>
<td>Reactive (out of proportion)</td>
<td>TPG &gt; 12 mmHg</td>
<td></td>
</tr>
</tbody>
</table>
### Parameters with established importance for assessing disease severity, stability and prognosis in PAK

<table>
<thead>
<tr>
<th>Better Prognosis</th>
<th>Determinants of Prognosis</th>
<th>Worse Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>No</td>
<td>Clinical evidence of RV failure</td>
<td>Yes</td>
</tr>
<tr>
<td>Slow</td>
<td>Rate of progression of symptoms</td>
<td>Rapid</td>
</tr>
<tr>
<td>No</td>
<td>Syncope</td>
<td>Yes</td>
</tr>
<tr>
<td>I, II</td>
<td>WHO-FC</td>
<td>IV</td>
</tr>
<tr>
<td>Longer (&gt; 500 m)*</td>
<td>6 MWT</td>
<td>Shorter (&lt; 300 m)</td>
</tr>
<tr>
<td>Peak O₂ Consumption &gt; 15 ml/min/kg</td>
<td>Cardio-pulmonary exercise testing</td>
<td>Peak O₂ consumption &lt; 12 ml/min/kg</td>
</tr>
<tr>
<td>Better Prognosis</td>
<td>BNP/NT-proBNP plasma levels</td>
<td>Very elevated and rising</td>
</tr>
<tr>
<td>No pericardial effusion TAPSE &gt; 2.0 cm</td>
<td>Echocardiographic findings†</td>
<td>Pericardial effusion TAPSE &lt; 1.5 cm</td>
</tr>
<tr>
<td>Right atrial pressure &lt; 8 mmHg and CI ≥ 2.5 L/min/m²</td>
<td>Haemodynamics</td>
<td>RAP &gt; 15 mmHg or CI ≤ 2.0 L/min/m²</td>
</tr>
</tbody>
</table>

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Echocardiography in PAH

Syst PAP = Right ventricular systolic pressure
(in absence of pulmonary outflow obstruction)

RVSP = 4v^2 + RAP*
Cut-off RVSP Values

Large Trials results

- Prospective screening of patients with scleroderma:
  TR velocity >2.5 m/s in symptomatic patients or >3.0 m/s irrespective of symptoms ..........> 45% of cases of echocardiographic diagnoses of PH were falsely positive.

- In symptomatic patients with HIV infection:
  PH criterion based on TR velocity >2.5 m/s and >2.8 m/s was found to be a false positive in 72% and 29%, respectively.

- In systemic sclerosis patients:
  TR pressure gradient >40 mmHg (TR velocity >3.2 m/s) with an assumed right atrial pressure of 10 mmHg (thus corresponding to a systolic PAP of > 50 mmHg) selected as the cut-off value for diagnosis of PH. The Doppler diagnosis was confirmed in all patients who were submitted to RHC.

Launay D, J Rheumatol 2007;34:1005-1011
# Echocardiographic probability of pulmonary hypertension in symptomatic patients with a suspicion of pulmonary hypertension according with PTRV & additional signs

<table>
<thead>
<tr>
<th>Peak tricuspid regurgitation velocity (m/s)</th>
<th>Presence of other echo “PH signs”</th>
<th>Echocardiographic probability of pulmonary hypertension</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤2.8 or not measurable</td>
<td>No</td>
<td>Low</td>
</tr>
<tr>
<td>≤2.8 or not measurable</td>
<td>Yes</td>
<td>Intermediate</td>
</tr>
<tr>
<td>2.9–3.4</td>
<td>No</td>
<td>High</td>
</tr>
<tr>
<td>2.9–3.4</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>&gt;3.4</td>
<td>Not required</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>A: The ventricles</th>
<th>B: Pulmonary artery</th>
<th>C: Inferior vena cava and right atrium</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right ventricle/ left ventricle basal diameter ratio &gt;1.0.</td>
<td>Right ventricular outflow Doppler acceleration time &lt;105 m/sec and/or midsystolic notching.</td>
<td>Inferior cava diameter &gt;21 mm with decreased inspiratory collapse (&lt;50 % with a sniff or &lt;20 % with quiet inspiration).</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>
<td>Right atrial area (end-systole) &gt;18 cm².</td>
</tr>
<tr>
<td></td>
<td>PA diameter &gt;25 mm.</td>
<td></td>
</tr>
</tbody>
</table>
Doppler estimation of SPAP

TRV should be measured in multiple views in an effort to ensure attainment of maximal TRV.

TRV is measurable in at least 75% of unselected patients. If contrast agents are used, can be obtained in more than 90% of patients.
RVOT Flow Morphology

Distribution of PASP that correspond to each RVOT spectral Doppler type.

Visual inspection of right ventricular outflow tract Doppler spectral signals showed four dynamic patterns.

López-Candales A Eur J Echocardiogr 2011
Doppler estimation of DPAP

PA end-diastolic pressure is frequently used as an estimate of pulmonary capillary wedge pressure

\[ \text{PAEDP} = 4(V_{PR-ED})^2 + \text{RVEDP} \]

\[ V_{PR-ED} = \text{peak end diastolic velocity of PR signal (m/sec)} \]
Doppler estimation of MPAP

\[ \text{MPAP} = 4V_1^2 \]

\( (V_1: \text{maximal first-diastolic velocity of pulmonary regurgitation}) \)

\[ \text{MPAP} = \frac{2}{3} \times \text{DPAP} + \frac{1}{3} \times \text{SPAP} \]

\[ \text{MPAP} = 0.61 \times \text{SPAP} + 2 \text{ mmHg} \]

Pulmonary Vascular Resistance

- PVR: TR peak pressure drop/RVOT VTI
- PVR: PASP/(HR × RVOT VTI)

Pulmonary Vascular Resistance

$PVR_{rhc} : 0.95 \times PVR_{echo} - 0.29$

- **PVR**: $PAMP_{echo} - PCWP/CO$
  - $PAMP = PASP_{echo} \times 0.61 + 2$ mmHg
    - (PASP$_{echo}$: TR peak PG+ 10 or 7 mmHg)
  - $PAMP$: $PADP + 0.33 (PASP-PAPD)$

The calculated ventricular mass index (VMI = ratio of right ventricular mass over left ventricular mass) provides an accurate and practical means of estimating pulmonary artery pressure noninvasively in pulmonary hypertension and may provide a more accurate estimate than Doppler echocardiography. Sensitivity and specificity for pulmonary hypertension were 84 and 71% respectively for the VMI compared with 89 and 57% for echocardiography.
Interventricular Septal Configuration at MR Imaging and Pulmonary Arterial Pressure in Pulmonary Hypertension

In patients with PH, systolic PAP > 67 mm Hg may be expected when leftward curvature is observed.

Roeleveld et al. Radiology 2005
Echocardiogram

Disadvantages

- Can be fooled:
  - Air trapping (COPD/Emphysema)
  - Expansion of thoracic cage
  - Alterations of position of heart
  - No estimation of LVEDP (PCWP) or CO/CI
  - Important in selection of therapy
  - Not useful for vasodilator challenges
  - Important in selection of therapy
In simultaneous DE and RHC measurements, there was moderate correlation between DE and RHC measurements of PASP (r = 0.71).

The bias for DE estimates of PASP was 8.0 mm Hg with 95% limits of agreement ranging from -28.4 to 44.4 mm Hg.

DE estimates of PASP are inaccurate in patients with PH and should not be relied on to make the diagnosis of PH or to follow the efficacy of therapy.
Mean pulmonary artery pressure and cardiac output relationship

Mean PA pressure

- Normal
- Mild to moderate
- Severe

Cardiac Output [L/min]

REST       EXCERCISE
Cases of Wrong Decisions

PVR : (mPAP - PAWP)/cardiac output

• In a prospective multicenter study* of 403 patients with SCD…
• 95 of patients had PAH (23.5%) using the criterion of TRV . 2.5 m/s
  • In RHC 75 patients were found to have normal mPAP;
• >> DE in this population resulted in false-positives in 75% of the cases.
  • In the remaining 24 patients, 13 had elevated PAWP.
• In 5 patients, the PASP was indeed elevated because of increased cardiac output, and the PVR was normal.

>>>>>>> The presence of true PAH is only 1.6% .

Right heart catheterisation

Characteristic intracardiac pressure waveforms during passage through the heart

RA  RV  PA  PCW

40 mmHg

20
What do we want to know?

- The true pressures?
- RA, RV, PA, PCWP
- How much blood is exiting the heart?
- Cardiac output, Cardiac Index
- What is the resistance?
- PVR
- SVO2
- Shunt?
- RA: 0-6 mmHg
- RV: 30/0-6 mmHg
- PA: 30/6-12 mmHg  PA mean: 15-20 mmHg
- PCWP: 5-15 mmHg
# Right heart catheterization in pulmonary hypertension

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>RHC is recommended to confirm the diagnosis of pulmonary arterial hypertension (Group 1) and to support treatment decisions.</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>In patients with PH, it is recommended to perform RHC in expert centres (Table 34) as it is technically demanding and may be associated with serious complications.</td>
<td>I</td>
<td>B</td>
</tr>
<tr>
<td>RHC should be considered in pulmonary arterial hypertension (Group 1) to assess the treatment effect of drugs (Table 12).</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>RHC is recommended in patients with congenital cardiac shunts to support decisions on correction (Table 23).</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>RHC is recommended in patients with PH due to left heart disease (Group 2) or lung disease (Group 3) if organ transplantation is considered.</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>When measurement of PAWP is unreliable, left heart catheterization should be considered to measure LVEDP.</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>RHC may be considered in patients with suspected PH and left heart disease or lung disease to assist in the differential diagnosis and support treatment decisions.</td>
<td>IIb</td>
<td>C</td>
</tr>
<tr>
<td>RHC is indicated in patients with Chronic Thromboembolic Pulmonary Hypertension (Group 4) to confirm the diagnosis and support treatment decisions.</td>
<td>I</td>
<td>C</td>
</tr>
</tbody>
</table>
6 minute walk test (6MWT)

- Measure of patients’ functional limitations
- Simple, inexpensive, convenient
- Correlate with WHO FC

ECHO: KEY DIAGNOSTIC ROLE!!

- Normal
- Further Diagnostic tests?
- 12 months F-U

- Pulmonary Hypertension
  - Aetiology?
    - PFT, X-ray, V/Q, CT
  - RHC
  - Treatment

Hoeper, ERS 2003
The Ten Commandments

1. Right heart catheterization is recommended to confirm the diagnosis of pulmonary arterial hypertension (PAH - Group 1) and to support treatment decisions.

2. **Vasoreactivity testing** performed during right heart catheterization is recommended in patients with idiopathic PAH, heritable PAH and PAH induced by drugs or toxins use to detect patients who can be treated with high doses of a calcium channel blocker.

3. It is recommended to evaluate the severity of PAH patients with a panel of data derived from clinical assessment, exercise tests, biochemical markers, and echocardiographic and haemodynamic evaluation and to perform regular follow-up assessments every 3-6 months in stable patients.

4. It is recommended to avoid pregnancy in patients with PAH.

5. It is recommended for referral centres to provide care by a multi-professional team (cardiology and respiratory medicine physicians, clinical nurse specialist, radiologists, psychological and social work support, appropriate on-call expertise).
Evaluating hemodynamics in pulmonary hypertension!!

Thank you for your Attention!!