PH Grand Rounds
Journal Club

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Vera Moulton Wall Center, Stanford
04.11.17
Disclosures

• none
CME Objectives

**Topic:** Journal Club – Loss of Vascular Distensibility During Exercise Is an Early Hemodynamic Marker of Pulmonary Vascular Disease

**Session objectives:**

- Define pulmonary vascular distensibility and contrast it with the linear model for the PA Pressure-Cardiac Output relationship.
- Describe the method and clinical utility of measuring PA distensibility in identifying patients with early PVD without overt resting PH.
Background

• Pre-clinical Pulmonary Arterial Hypertension (PAH)
  • The need to identify early PAD
  • Exercise pulmonary hypertension (PH)

• Exercise PH
  • Definition. Identifying normal and diseased
  • Natural history
  • Ppa – CO relationship
    • Linear
    • Curvilinear: α (vascular distensibility)
Pulmonary Arterial Hypertension (PAH)

• Current era 1 year mortality – 9-14%
• Substantial delay in diagnosis persists
  • 2009 reports from UK and French registries:
    • ~80% of new patients are NYHA FC III-IV
    • EARLY study – Baseline PVR was 10 WU
• Earlier diagnosis and thus therapy relates to longer survival

Early PAH

• Pathologic changes prior to developing resting PH
  • Majority of (>60%) of pulmonary circulation is obstructed prior to mPAP > 25 mmHg
  • Pneumonectomy, unilateral balloon occlusion

• Exaggerated pulmonary hypertensive response to exercise related to exercise capacity

• Exercise PH may be an intermediate phase prior to resting clinically apparent PAH

Exercise Pulmonary Hypertension

• Normal subjects usually did not achieve a mPAP > 30 mmHg with graded exercise in early reports
• Peak mPAP > 30 mmHg considered abnormal
• Eventually recognized this cut-off was arbitrary and didn’t account for healthy individuals and elderly
  • PAP = CO*PVR + P_{la}
  • Need to account for flow rate and P_{la}
• In 2008, exercise PH was removed from international guidelines
Natural History of Exercise PH

• Best data in the higher risk Scleroderma patients
  • 42 pt longitudinal study of Exercise PH + SSc.
  • 19% developed resting PH over median follow up of 30 months

• Unclear whether other subgroups of patients with exercise PH remain stable or progress.

• No well defined ways of predicting prognosis in patients that have exercise PH

mPAP-CO relationship with exercise

- Initially understood using Ohms law (linear)
- $\text{mPAP} = \text{CO} \times \text{PVR} + P_{la}$
- $\Delta\text{mPAP}/\Delta\text{CO} \sim \text{total PVR}$
- Normal slope (PVR): $0.5 – 2.5 \text{ mmHg/L/min}$
- Age >50 associated with near doubling of slope
  - 1.4 to 2.5 mmHg/L/min
- Linear increase in $P_{la}$ also seen (moreso in elderly)
- At a CO of 10 L/min, peak normal mPAP $\sim 34 \text{ mmHg}$
  - Corresponding to a peak slope of $\sim 3 \text{ mmHg/L/min}$
- With multiple data points, curvilinearity appears
  - Due to natural distensibility of pulmonary arteries at higher pressures

Distensible model for Pulmonary pressure-flow curves

\[ P_a = \frac{\left[ (1 + \alpha P_v)^5 + 5\alpha R_0 \dot{Q} \right]^{1/5} - 1}{\alpha} \]  (12)

• Defined by Linehan et al in 1985
• Evaluated using explanted dog lungs with varying steady flow and differing Hct
• \( \alpha \) (distensibility) = % change in PA diameter/mmHg

Assumptions

• Linear relationship between vessel diameter and pressure
• Local vessel resistance defined by Poiseuille’s law
• Ro assumed to be TPR at rest
• Alpha is independent of vessel diameter
• Can not distinguish between recruitment of closed vessels/capillaries and distensibility

Distensibility in other species

Table 1. *Measurements relating to distensibility in normal human subjects at SL and Alt at rest and during upright cycle exercise*

<table>
<thead>
<tr>
<th>Subject Number</th>
<th>R&lt;sub&gt;o&lt;/sub&gt;, mmHg·l&lt;sup&gt;-1&lt;/sup&gt;·min&lt;sup&gt;-1&lt;/sup&gt;</th>
<th>α</th>
<th>Ppa (calc-meas) Absolute Values</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>SL</td>
<td>Alt</td>
<td>SL</td>
</tr>
<tr>
<td>1</td>
<td>1.87</td>
<td>4.33</td>
<td>0.013</td>
</tr>
<tr>
<td>3</td>
<td>3.26</td>
<td>4.15</td>
<td>0.029</td>
</tr>
<tr>
<td>4</td>
<td>2.54</td>
<td>3.91</td>
<td>0.019</td>
</tr>
<tr>
<td>5</td>
<td>2.20</td>
<td>3.64</td>
<td>0.024</td>
</tr>
<tr>
<td>6</td>
<td>2.64</td>
<td>2.95</td>
<td>0.018</td>
</tr>
<tr>
<td>7</td>
<td>1.48</td>
<td>3.64</td>
<td>0.018</td>
</tr>
<tr>
<td>8</td>
<td>2.69</td>
<td>3.95</td>
<td>0.032</td>
</tr>
<tr>
<td>9</td>
<td>1.80</td>
<td>4.35</td>
<td>0.020</td>
</tr>
<tr>
<td>Mean</td>
<td>2.31</td>
<td>3.89*</td>
<td>0.022</td>
</tr>
<tr>
<td>SE</td>
<td>0.21</td>
<td>0.22</td>
<td>0.002</td>
</tr>
</tbody>
</table>

*Operation Everest II: males, ages 21–28 yr at SL and 3 wk at Alt to 6,100 m (‘87)*

<table>
<thead>
<tr>
<th>Subject Number</th>
<th>R&lt;sub&gt;o&lt;/sub&gt;, mmHg·l&lt;sup&gt;-1&lt;/sup&gt;·min&lt;sup&gt;-1&lt;/sup&gt;</th>
<th>α</th>
<th>Ppa (calc-meas) Absolute Values</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>SL</td>
<td>Alt</td>
<td>SL</td>
</tr>
<tr>
<td>1</td>
<td>3.39</td>
<td>0.019</td>
<td>1.4±0.3</td>
</tr>
<tr>
<td>2</td>
<td>1.25</td>
<td>1.95</td>
<td>0.014</td>
</tr>
<tr>
<td>3</td>
<td>2.23</td>
<td>2.04</td>
<td>0.015</td>
</tr>
<tr>
<td>4</td>
<td>4.59</td>
<td>1.74</td>
<td>0.023</td>
</tr>
<tr>
<td>5</td>
<td>2.54</td>
<td>2.03</td>
<td>0.011</td>
</tr>
<tr>
<td>6</td>
<td>1.93</td>
<td>1.90</td>
<td>0.010</td>
</tr>
<tr>
<td>7</td>
<td>2.34</td>
<td>2.57</td>
<td>0.012</td>
</tr>
<tr>
<td>8</td>
<td>1.81</td>
<td>1.86</td>
<td>0.008</td>
</tr>
<tr>
<td>Mean</td>
<td>2.51</td>
<td>1.92</td>
<td>0.014</td>
</tr>
<tr>
<td>SE</td>
<td>0.37</td>
<td>0.19</td>
<td>0.002</td>
</tr>
</tbody>
</table>

Values are means ± SE. SL, sea level; Alt, altitude; R<sub>o</sub>, total pulmonary resistance; α, distensibility; Ppa, pulmonary arterial pressure; calc, calculated; meas, measured. *Measurements at Alt differ (P < 0.05) from those at SL. Duke chamber study data obtained from R. E. Moon (personal communication).

Actual $\alpha = 0.78\%$

Idealized $\alpha = 2.0\%$

Graphs showing the relationship between Cardiac Output (L/min) and various pressures (mPAP, TPG, PAWP) with lines indicating the relationship.
Summary and features of $\alpha$

- Increases in distending pressure (mPAP) substantially reduce PVR and thus create a curvilinear relationship between flow and pressure in exercising humans.
- Distensibility ($\alpha$) defines this curvilinearity.
- $\alpha$ accounts for the influence of PAWP on mPAP.
- Independent of supine or upright exercise.
- Exercising humans have similar $\alpha$ as in vitro lungs in humans and animals.
- Normal subjects (24) undergoing exercise echo + CPET: $\alpha$ and PVRI are independent predictors of VO2max$^1$.
- Reflects vascular remodeling (chronic vs acute hypoxia).
- Values of $\alpha$ decrease with increasing age. Mixed reports of gender association.
- In all, reflects a mechanical property of PA vessels.
- An evolutionary conjecture: consistent $\alpha$ across multiple PA diameter vessels and regions leads to a simple mechanism to maintain homogenous flow distribution. Consequently, in disease, heterogeneity may also be relevant.

Loss of Vascular Distensibility During Exercise Is an Early Hemodynamic Marker of Pulmonary Vascular Disease

Edmund M. T. Lau, MD, PhD; Denis Chemla, MD, PhD; Laurent Godinas, MD; Kaixian Zhu, MSc; Olivier Sitbon, MD, PhD; Laurent Savale, MD, PhD; David Montani, MD, PhD; Xavier Jais, MD; David S. Celermajer, MD, PhD; Gérald Simonneau, MD; Marc Humbert, MD, PhD; and Philippe Hervé, MD, PhD

- Retrospective review of RHC from 2008 to 2014 in French database (n-248)
- Same group as the recent Herve et al paper proposing new Exercise PH criteria
- Patients referred to dyspnea clinic and underwent invasive exercise hemodynamics
  - Controls: Normal PFT, CT and V/Q. No risk factors for PVD. Normal hemodynamics at rest and with exercise
  - PVD-PH: Pre-capillary PAH at rest. Included PAH (25) and CTEPH (6)
  - PVD-noPH: No rest PAH. Developed PAH at follow up (8), lung biopsy showed PVD (4), VQ+ and angio+ (21)

Exercise hemodynamics

• Required at least 5 data points from exercise study (mPAP, CO)

• Supine bike ergometry performed with PA pressure, TD-CO and PAWP measurements every 3-5 min

• $\alpha$ calculated for each set of data using successive iteration of $\alpha$ to get the best least square fit between calculated and measured mPAP using…

\[
P_a = \frac{[1 + \alpha p_v]^5 + 5\alpha R_0 \dot{Q}]^{1/5} - 1}{\alpha} \tag{12}
\]

Method of Calculating $\alpha$


<table>
<thead>
<tr>
<th>Demographic</th>
<th>Control Subjects (n = 26)</th>
<th>PVD-noPH (n = 33)</th>
<th>PVD-PH (n = 31)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y</td>
<td>51 ± 13</td>
<td>55 ± 13</td>
<td>51 ± 14</td>
<td>.45</td>
</tr>
<tr>
<td>Male, No. (%)</td>
<td>12 (46)</td>
<td>11 (33)</td>
<td>14 (45)</td>
<td>.18</td>
</tr>
<tr>
<td>Height, cm</td>
<td>168 ± 9</td>
<td>168 ± 8</td>
<td>167 ± 10</td>
<td>.92</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>72 ± 15</td>
<td>73 ± 15</td>
<td>67 ± 15</td>
<td>.32</td>
</tr>
<tr>
<td>BMI, kg/m²</td>
<td>26 ± 5</td>
<td>26 ± 4</td>
<td>24 ± 4</td>
<td>.16</td>
</tr>
<tr>
<td>BSA, m²</td>
<td>1.80 ± 0.19</td>
<td>1.80 ± 0.20</td>
<td>1.75 ± 0.23</td>
<td>.58</td>
</tr>
<tr>
<td><strong>Resting pulmonary hemodynamics</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>HR, bpm</td>
<td>72 ± 14</td>
<td>74 ± 11</td>
<td>78 ± 12</td>
<td>.23</td>
</tr>
<tr>
<td>mPpa, mm Hg</td>
<td>14 ± 4</td>
<td>20 ± 3</td>
<td>34 ± 10</td>
<td>&lt; .0001</td>
</tr>
<tr>
<td>Ppw, mm Hg</td>
<td>7 ± 3</td>
<td>6 ± 3</td>
<td>7 ± 3</td>
<td>.10</td>
</tr>
<tr>
<td>TPG, mm Hg</td>
<td>7 ± 3</td>
<td>14 ± 4</td>
<td>26 ± 10</td>
<td>&lt; .0001</td>
</tr>
<tr>
<td>RAP, mm Hg</td>
<td>4 ± 3</td>
<td>3 ± 2</td>
<td>5 ± 3</td>
<td>.037</td>
</tr>
<tr>
<td>CO, L/min</td>
<td>6.4 ± 1.2</td>
<td>5.5 ± 1.1</td>
<td>5.4 ± 1.3</td>
<td>.0084</td>
</tr>
<tr>
<td>SV, mL</td>
<td>91 ± 24</td>
<td>75 ± 16</td>
<td>70 ± 19</td>
<td>.0006</td>
</tr>
<tr>
<td>PVR, Wood Units</td>
<td>1.1 ± 0.6</td>
<td>2.6 ± 0.8</td>
<td>5.2 ± 2.2</td>
<td>&lt; .0001</td>
</tr>
<tr>
<td>Csv/pp, mL/mm Hg</td>
<td>6.2 ± 2.1</td>
<td>3.5 ± 1.0</td>
<td>2.2 ± 0.9</td>
<td>&lt; .0001</td>
</tr>
</tbody>
</table>
Exercise Hemodynamic Results

**TABLE 2** ] Exercise Hemodynamics of Study Subjects

<table>
<thead>
<tr>
<th>Exercise Hemodynamic</th>
<th>Control Subjects (n = 26)</th>
<th>PVD-noPH (n = 33)</th>
<th>PVD-PH (n = 31)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>mPpa-CO points, No.</td>
<td>6.1 ± 1.1</td>
<td>5.9 ± 1.0</td>
<td>5.8 ± 1.1</td>
<td>.79</td>
</tr>
<tr>
<td>Peak workload, W</td>
<td>58 ± 24</td>
<td>54 ± 22</td>
<td>47 ± 21</td>
<td>.34</td>
</tr>
<tr>
<td>Peak HR, bpm</td>
<td>112 ± 21</td>
<td>111 ± 19</td>
<td>112 ± 14</td>
<td>.99</td>
</tr>
<tr>
<td>Peak mPpa, mm Hg</td>
<td>25 ± 5</td>
<td>41 ± 8</td>
<td>58 ± 11</td>
<td>&lt; .0001</td>
</tr>
<tr>
<td>Peak Ppw, mm Hg</td>
<td>12 ± 5</td>
<td>11 ± 5</td>
<td>12 ± 4</td>
<td>.67</td>
</tr>
<tr>
<td>Peak TPG, mm Hg</td>
<td>12 ± 4</td>
<td>30 ± 7</td>
<td>46 ± 12</td>
<td>&lt; .0001</td>
</tr>
<tr>
<td>Peak CO, L/min</td>
<td>12.8 ± 2.6</td>
<td>11.1 ± 2.9</td>
<td>9.9 ± 2.7</td>
<td>.0013</td>
</tr>
<tr>
<td>Peak SV, mL</td>
<td>117 ± 29</td>
<td>96 ± 19</td>
<td>89 ± 25</td>
<td>.0002</td>
</tr>
<tr>
<td>Peak PVR, Wood Units</td>
<td>1.0 ± 0.4</td>
<td>2.7 ± 0.8</td>
<td>5.1 ± 2.3</td>
<td>&lt; .0001</td>
</tr>
</tbody>
</table>

Exercise Pressure-Flow Relationships

A 

Control

B 
PVD-noPH

C 
PVD-PH

<table>
<thead>
<tr>
<th></th>
<th>Control</th>
<th>PVD-noPH</th>
<th>PVD-PH</th>
</tr>
</thead>
<tbody>
<tr>
<td>mPAP-Q Slope</td>
<td>1.5</td>
<td>3.2</td>
<td>4.9</td>
</tr>
</tbody>
</table>

Bland Altman between calculated and measured mPAP

• In controls, $\alpha$ decreased with increasing age.

e-Figure 2. Vascular distensibility α according to thromboembolic and non-thromboembolic etiology of pulmonary vascular disease. Distensibility α was lower in PVD without PH compared to PVD with resting PH for thromboembolic etiology (0.50±0.26 vs. 0.26±0.11, \( p = 0.010 \)) and non-thromboembolic etiology (0.36±0.18 vs. 0.26±0.15, \( p = 0.15 \)).

α - Capacitance relationship

Graph A: Sens 88% and Spec of 100% at cutoff value of $\alpha = 0.76$

Graph B: Good discrimination even in patients with resting mPAP < 20

Major contributions from this study

• Distensibility model accurately describes observed mPAP-CO relationship in controls and PVD patients

• Good accuracy and precision throughout a large range of observed mPAP

• Marked attenuation in distensibility (α) in patients with PVD but without resting PAH (even with resting mPAP < 20 mmHg)
  • Remodeling of resistive vessels effecting α may be an early event in the progression of PVD

• α has moderate correlation with Capacitance (SV/PP) but better performance in distinguishing PVD-noPH
Other thoughts

- Comparison of diagnostic performance with linear slope of mPAP-CO relationship
- Not necessarily quick to calculate this clinically
- Ultimately a diagnostic index for early PVD only. Not much discrimination in patients with PAH.
- Not sure if it provides that much additional information given the time it takes to calculate
- Correlation with direct imaging measurements of $\alpha$?
Abnormal Pulmonary Artery Stiffness in Pulmonary Arterial Hypertension: *In Vivo* Study with Intravascular Ultrasound

Edmund M. T. Lau¹,²,³*, Nithin Iyer³*, Rahn Ilsar¹, Brian P. Bailey¹, Mark R. Adams¹,³, David S. Celermajer¹,³*

¹Department of Cardiology, Royal Prince Alfred Hospital, Camperdown, Australia, ²Department of Respiratory and Sleep Medicine, Royal Prince Alfred Hospital, Camperdown, Australia, ³Sydney Medical School, University of Sydney, Camperdown, Australia

**Abstract**

**Background:** There is increasing recognition that pulmonary artery stiffness is an important determinant of right ventricular (RV) afterload in pulmonary arterial hypertension (PAH). We used intravascular ultrasound (IVUS) to evaluate the mechanical properties of the elastic pulmonary arteries (PA) in subjects with PAH, and assessed the effects of PAH-specific therapy on indices of arterial stiffness.

**Method:** Using IVUS and simultaneous right heart catheterisation, 20 pulmonary segments in 8 PAH subjects and 12 pulmonary segments in 8 controls were studied to determine their compliance, distensibility, elastic modulus and stiffness index $\beta$. PAH subjects underwent repeat IVUS examinations after 6-months of bosentan therapy.

**Results:** At baseline, PAH subjects demonstrated greater stiffness in all measured indices compared to controls: compliance $(1.50\pm0.11\times10^{-2}$ mm$^2$/mmHg vs $4.49\pm0.43\times10^{-2}$ mm$^2$/mmHg, $p<0.0001$), distensibility $(0.32\pm0.03\%$/mmHg vs $1.18\pm0.13\%$/mmHg, $p<0.0001$), elastic modulus $(720\pm64$ mmHg vs $198\pm19$ mmHg, $p<0.0001$), and stiffness index $\beta$ $(15.0\pm1.4$ vs $11.0\pm0.7$, $p=0.046$). Strong inverse exponential associations existed between mean pulmonary artery pressure and compliance ($r^2=0.82$, $p<0.0001$), and also between mean PAP and distensibility ($r^2=0.79$, $p=0.002$). Bosentan therapy, for 6-months, was not associated with any significant changes in all indices of PA stiffness.

**Conclusion:** Increased stiffness occurs in the proximal elastic PA in patients with PAH and contributes to the pathogenesis of RV failure. Bosentan therapy may not be effective at improving PA stiffness.

Pulmonary Vascular Distensibility Predicts Pulmonary Hypertension Severity, Exercise Capacity, and Survival in Heart Failure

Rajeev Malhotra, MD; Bishnu P. Dhakal, MD; Aaron S. Eisman, BS; Paul P. Pappagianopoulos, MEd; Ashley Dress, MS; Rory B. Weiner, MD; Aaron L. Baggish, MD; Marc J. Semigran, MD; Gregory D. Lewis, MD

• CPET+invasive hemodynamics
• In controls (30), HFrEF (55), HFpEF(48) and PAH (18) patients
• Looking mainly at HF patients and what determines survival
### Table 3. Multivariable Model for Predicting Peak VO₂ in All Patients With HF (n=103)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Normalized β-Coefficient</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y</td>
<td>−0.25</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Male sex</td>
<td>0.16</td>
<td>0.008</td>
</tr>
<tr>
<td>Hb, g/dL</td>
<td>0.20</td>
<td>0.001</td>
</tr>
<tr>
<td>Resting cardiac index, L/min per m²</td>
<td>0.29</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Resting PAWP, mm Hg</td>
<td>−0.22</td>
<td>0.003</td>
</tr>
<tr>
<td>Resting mPAP, mm Hg</td>
<td>−0.19</td>
<td>0.018</td>
</tr>
<tr>
<td>Distensibility, % per mm Hg</td>
<td>0.25</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Hb indicates hemoglobin; HF, heart failure; mPAP, mean pulmonary arterial pressure; and PAWP, pulmonary arterial wedge pressure.
Figure 3. Pulmonary vascular distensibility is associated with change in right ventricular ejection fraction (RVEF) with exercise. A scatterplot of change in RVEF versus distensibility of the 4 patient groups are depicted. Spearman rank correlations were determined. Pulmonary vascular distensibility is a strong determinant of right ventricular function with exercise.


**Figure 4.** Pulmonary vascular (PV) distensibility predicts cardiovascular mortality in patients with heart failure (HF). Kaplan–Meier survival curves of patients with HF (n=103, both HF with preserved ejection fraction [HFpEF] and HF with reduced ejection fraction [HFrEF]) are depicted, dichotomized by PV distensibility value. Compared with those with lower PV distensibility, HF patients with a distensibility ≥0.70% per mmHg exhibit reduced cardiovascular mortality (P=0.03).

**Table 4.** Pulmonary Vascular Distensibility Predicts Cardiovascular Survival in All Patients With HF (n=103)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Cox Hazard Ratio</th>
<th>95% Confidence Interval</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y</td>
<td>1.01</td>
<td>0.98–1.05</td>
<td>0.48</td>
</tr>
<tr>
<td>Male sex</td>
<td>2.85</td>
<td>1.22–6.66</td>
<td>0.015</td>
</tr>
<tr>
<td>Peak VO₂, mL/kg per min</td>
<td>0.79</td>
<td>0.70–0.88</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Distensibility, % per mmHg</td>
<td>0.30</td>
<td>0.10–0.93</td>
<td>0.036</td>
</tr>
</tbody>
</table>

HF indicates heart failure.
RV Dysfunction In Pulmonary Hypertension Is Independently Related To Pulmonary Artery Stiffness

Gerin R. Stevens, MD, PhD,* Ana Garcia-Alvarez, MD, MSC,‡§|| Sheila Sahni, MD,† Mario J. Garcia, MD,* Valentin Fuster, MD, PhD,‡§ Javier Sanz, MD‡

New York, New York; and Madrid and Barcelona, Spain

• RHC + CMR performed within 1 week (124 patients)
• Known or suspected PH
• RV performance indices related to measures of PA stiffness

**Table 2. Patient Hemodynamic Characteristics**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Median (IQR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrial pressure, mm Hg</td>
<td>6.0 (3.0–12.0)</td>
</tr>
<tr>
<td>Systolic PA pressure, mm Hg</td>
<td>64.5 (45.5–80.0)</td>
</tr>
<tr>
<td>Mean PA pressure, mm Hg</td>
<td>40.0 (29.3–50.0)</td>
</tr>
<tr>
<td>PA pulse pressure, mm Hg</td>
<td>39.0 (26.0–50.0)</td>
</tr>
<tr>
<td>Pulmonary capillary wedge pressure, mm Hg</td>
<td>10.0 (7.0–13.0)</td>
</tr>
<tr>
<td>Transpulmonic gradient, mm Hg</td>
<td>27.0 (17.3–40.0)</td>
</tr>
<tr>
<td>PVRI, Wood units·m⁻²</td>
<td>8.7 (5.0–14.2)</td>
</tr>
<tr>
<td>Cardiac index, l/min·m⁻²</td>
<td>3.4 (2.5–3.8)</td>
</tr>
<tr>
<td>RVSWI, g·m⁻²/m²/beat</td>
<td>16.7 (10.5–22.8)</td>
</tr>
<tr>
<td>PA oxygen saturation, %</td>
<td>66.0 (57.0–71.0)</td>
</tr>
<tr>
<td>Heart rate, beats/min</td>
<td>79.0 (70.0–91.0)</td>
</tr>
<tr>
<td>Mean systemic arterial pressure, mm Hg</td>
<td>88.0 (80.0–99.8)</td>
</tr>
</tbody>
</table>

Values are median (interquartile range).

PA = pulmonary artery; PVRI = pulmonary vascular resistance index; RVSWI = right ventricular stroke work index.

Summary:
Pulmonary Arterial Distensibility ($\alpha$)

- Definition: (D change/resting D)/pulse pressure
- Methods of calculating – directly measure $\alpha$ using imaging?
  - Exercise hemodynamics
  - MRI
  - Gated CT angiogram
  - Echocardiography
  - IVUS
- Normal values: 0.15 - 0.2 (%/mmHg)
- Correlation:
  - Between different methods of calculating
  - With other hemodynamic parameters
  - With disease subtypes and disease progression
  - Functional capacity
  - Survival
  - Treatment response
- Performance compared with other exercise hemodynamic markers
Thank You